RDS Rheumatologic Dermatology Society North America

2025 RDS Annual Meeting Abstract Book

Oral Presentations
Session I

ADVANCED THERAPEUTICS IN CHILD-BEARING WOMEN AND PREGNANCY

ANIFROLUMAB USE IN PREGNANCY: MATERNAL AND NEONATAL OUTCOMES IN SYSTEMIC LUPUS ERYTHEMATOSUS PATIENTS

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Systemic lupus erythematosus (SLE) is an autoimmune disease that disproportionately affects women of childbearing age. Pregnancy in SLE is associated with disease flares, preeclampsia, fetal loss, and preterm birth. Anifrolumab, a monoclonal antibody targeting the type I interferon receptor, has demonstrated efficacy in reducing SLE disease activity in nonpregnant individuals. Limited data is available on the safety and efficacy of anifrolumab in pregnancy as pregnant patients were excluded from clinical trials. We performed a retrospective review of pregnant patients with SLE treated with anifrolumab at Mass General Brigham. The study included three patients ranging from 29 to 36 years in age. All participants had SLE with varying degrees of disease activity. The number of anifrolumab infusions prior to conception varied from 7 to 13, with two of the patients receiving anifrolumab within six weeks of the estimated date of conception. The patients received a mean of 4 infusions during pregnancy. All patients required induction of labor that resulted in vaginal deliveries. One case of severe fetal growth restriction was observed in the fetus whose mother had persistent serologic and clinical disease activity throughout pregnancy. No congenital malformations were reported and neonatal Apgar scores ranged from 3 to 8 at 1 minute and from 7 to 8 at 5 minutes. Maternal complications included one case of mild postpartum hemorrhage requiring medical intervention in a patient on enoxaparin for antiphospholipid antibody syndrome. This preliminary data suggests that anifrolumab may be well-tolerated in pregnancy and provides early evidence that anifrolumab use in pregnancy may not significantly increase maternal or fetal complications. Given the limited therapeutic options for pregnant individuals with SLE, further prospective studies investigating the safety of anifrolumab in pregnancy are warranted.

Category: Lupus

Session II:

SCLEROSING AND OTHER CONNECTIVE TISSUE

DISEASES

LASER-ASSISTED DRUG DELIVERY OF HYALURONIDASE FOR THE TREATMENT OF SCLERODERMA-INDUCED MICROSTOMIA

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Scleroderma-induced microstomia (SIM) is a debilitating consequence of pathological deposition of dermal collagen in the setting of systemic sclerosis. Both intradermal injections of hyaluronidase and ablative CO2 laser have shown efficacy in small cohorts. In this pilot study we combined these modalities, utilizing laser-assisted drug delivery (LADD) of hyaluronidase to more evenly disperse the medication to the target areas. Three female patients were enrolled at the University of Nebraska Medical Center to receive 3 treatments with ablative CO2 laser (settings: deep, 5% density fractional coverage, 40mJ) followed by immediate topical application of 150U (1mL) of hyaluronidase. Oral aperture measurements including maximum interincisal (IID), intercommisural (ICD), and intervermillion (IVD) distances were obtained at each visit using a digital caliper. Surveys including interval Mouth Handicap in Systemic Sclerosis (MHISS) and pre-and post-study Short Form 36 Quality of Life (SF-36 QOL) tracked changes in perceived function during the study. Laser sessions were completed in 4–6-week intervals with final measurements recorded 3 months following the final session. The intervention was well-tolerated by all three patients without adverse effects aside from mild stinging pain and erythema. Mean IID increased from 32.6mm to 37.2mm; ICD increased from 46.7mm to 47.9mm; and IVD increased from 49.8mm to 57.2mm. There was a reduction in median MHISS from 26.0 (IQR: 22.0-36.0) to 18.0 (IQR: 10.0-25.0) at the conclusion of the study. SF-36 QOL emotional domain means increased slightly from 52.9 to 53.2 and physical domain means increased notably from 26.0 to 37.0, far exceeding the established minimal clinically important difference in patients with diffuse systemic sclerosis. Based on this experience with our cohort, we feel LADD of hyaluronidase is a promising modality to increase oral aperture size and improve microstomia-associated quality of life with patients with systemic sclerosis.

Category: Sclerotic skin disease (e.g. morphea, systemic sclerosis, etc.)

SINGLE-CELL AND SPATIAL PROFILING ANALYSIS IMPLICATES RESIDENT MEMORY T CELLS AND TREM2$^+$ MACROPHAGES AS DRIVERS OF SKIN FIBROSIS

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Cutaneous fibrosis in morphea and eosinophilic fasciitis (EF) remains poorly understood and lacks FDA-approved treatments. Our prior transcriptional profiling identified cytotoxic T cell activation in fibrotic skin lesions, while immunostaining showed no increase in total T cell numbers, suggesting that resident memory T cells (TRM) sustain chronic injury. We also found that vascular injury in fibrotic lesions involved T cell-driven endothelial cell necroptosis, an immunogenic form of cell death. To further define these immune programs, we analyzed single-cell RNA sequencing (scRNA-Seq) data from 27 morphea and 17 healthy skin (HS) biopsies. We also performed Xenium digital spatial profiling (DSP) on inflamed fibrotic morphea and EF lesions and HS. ScRNA-Seq showed no difference in T cell frequency between morphea and HS, but revealed two CD8$^+$ TRM (*CD69*$^+$*ITGAE*$^+$) clusters that expressed cytotoxic effectors (*GZMA*, *GZMB*, *IFNG*), consistent with a capacity for tissue injury. DSP demonstrated infiltration of CD8$^+$ T cells marked by *IFNG*, *CCR5*, and *KLRG1*, indicating antigen-driven inflammation. DSP revealed two *TREM2*$^+$ macrophage populations: one co-expressing complement *C3* and the other *SPP1*. TREM2 senses lipids, including those released by necroptotic cells, and TREM2$^+$ macrophages are known mediators of fibrosis in other organs. Here, we implicate TREM2$^+$ macrophages in morphea and EF. The *C3*$^+$ subset suggests that complement engagement may amplify macrophage-fibroblast interactions. Spatial colocalization analysis revealed a cytotoxic niche defined by CD8$^+$ T cells clustered with activated antigen-presenting cells, and IFNγ likely induces CXCR3 ligands that amplify injury. There was also a fibrotic niche defined by *C3*$^+$*TREM2*$^+$ macrophages colocalized with fibrotic fibroblasts. These interactions define a profibrotic axis of T cell cytotoxicity, tissue injury, and macrophage-fibroblast crosstalk. We hypothesize that infiltrating CD8$^+$ T cells in inflamed lesions establish

residency and perpetuate chronic fibrosis. Our findings suggest that inhibition of TREM2 and necroptosis represent rational targeted therapeutic strategies in morphea and EF.

Teaching point: Morphea and EF pathogenesis involves an immune circuit in which cytotoxic T cells drive tissue injury and TREM2^{+} macrophages contribute to fibroblast activation.

Category: Sclerotic skin disease

THE ROLE OF MAGNETIC RESONANCE IMAGING IN THE EVALUATION OF DISEASE PROGRESSION IN EOSINOPHILIC FASCIITIS: A RETROSPECTIVE COHORT STUDY

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Word count: 290

Eosinophilic fasciitis (EF) is a rare sclerosing disorder clinically characterized by painful skin induration, limb swelling, and peripheral eosinophilia and classically involving underlying fascia. The gold standard for the diagnosis of EF is a full-thickness wedge biopsy; however, more recent studies have shown the non-inferiority of magnetic resonance imaging (MRI) in EF detection. Standard EF treatment includes high-dose steroids, which are used as first-line therapy for managing acute flares and relapses. Despite their effectiveness, prolonged steroid use results in adverse systemic effects. Therefore, an objective measure of disease progress in EF is essential to avoid unnecessary steroid exposure. No study to date has evaluated the utility of MRI in evaluating EF disease progression. We performed a retrospective review of all patients with EF who presented to our institutions between 1/1/2000 and 7/1/2025. Out of 127 patients who were identified as having EF, 52 patients underwent a diagnostic MRI which established a baseline; eighteen of these patients had additional MRI imaging to evaluate progress and were identified as subjects for our study. The most common reasons for undergoing MRI imaging were symptom flares (12/18, 66.7%), routine maintenance (4/18, 22.2%), and re-evaluation of refractory EF (2/18, 11.1%)—with steroid dose hinging on MRI results. Median time for a follow-up MRI was 27.5 months, with a range of 101.9 months. Six of 18 patients were male. The median age was 59.7 years. We found that steroid dose escalation was curbed in 15 patients (83%) since MRI revealed either no active fasciitis or diminished fascial inflammation and therefore, physicians more confidently and safely could continue tapering steroids and/or maintain stable regimens. Our study indicates early evidence that the routine use of MRI throughout EF treatment can help guide treatment management, preventing prolonged steroid exposure.

Category: Sclerotic skin disease

USE OF THE ORAL JAK INHIBITOR TOFACITINIB IN THE TREATMENT OF MORPHEA: A RETROSPECTIVE STUDY

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Morphea is a rare autoimmune disease affecting the skin and connective tissues, with diverse clinical presentations and no standard treatment. A study by Damsky et al. suggested Janus Kinase inhibitors (JAK-I) may be effective. This retrospective study evaluated the efficacy and tolerability of oral tofacitinib in morphea treatment. We reviewed patient records meeting these criteria: ≥6 months on tofacitinib, active disease at initiation, and no concurrent systemic therapy. Patients' data, including clinical scores such as the localized scleroderma skin severity index (LoSCAT), localized scleroderma activity index (LoSAI), localized scleroderma damage index (LoSDI), Physician Global Assessment of Activity (PGA-A), and Physician Global Assessment of Damage (PGA-D), were extracted. Twenty-one patients (mean age 38 ± 16 years) were included, with 96% being female. The mean treatment duration with tofacitinib was 8.2±2 months. More than half of the patients (52%) had plaque-type morphea. The most common (33%) site of involvement was scalp and neck. 15 patients were switched from methotrexate to tofacitinib due to no response or adverse effects. Nineteen patients (91%) showed improvement. The mean clinical scores of LoSCAT, LoSAI, LoSDI, PGA-A, and PGA-D were decreased at the last visit compared to the tofacitinib start visit. However, only LoSAI and PGA-A showed a statistically significant decrease (P-value<0.05). We performed a detailed examination of the LoSDI components in all subjects. Patients' dermal atrophy, dyspigmentation and skin thickness decreased after starting treatment with tofacitinib. Patients with generalized morphea showed a greater decrease in dermal atrophy and skin thickness. Also, subcutaneous atrophy decreased in patients with generalized and linear morphea, whereas in patients with plaque morphea, it increased. Regarding side effects, two patients experienced acne, which was managed with antibiotics. In conclusion, tofacitinib shows promise as a treatment for morphea, particularly in patients unresponsive to methotrexate, and could be considered as a first-line treatment.

Abstract category: Sclerotic skin disease (Morphea)

EFFICACY OF JANUS KINASE INHIBITORS IN MORPHEA AND EOSINOPHILIC FASCIITIS: A RETROSPECTIVE COHORT STUDY

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Word count: 300/300

Dysregulated immune signaling within the JAK/STAT pathway has recently been implicated in the pathogenesis of morphea and eosinophilic fasciitis (EF). We conducted a retrospective cohort study to assess the efficacy and side effects of JAK inhibitors (JAKis) in treating morphea and EF. The Mass General Brigham Research Patient Data Registry identified patients with active morphea and/or EF who were prescribed a JAKi. 20 patients met inclusion criteria. Mean age was 57.3 (±17.6) years. 55% were male and 80% were non-Hispanic White. 11 patients (55%) had morphea, 7 (35%) had EF, and 2 (10%) had EF with morphea overlap. 70% had extracutaneous features of disease, including restricted range of motion (45%), joint pain (25%), and muscle involvement (25%). Mean number of prior treatments trialed was 4.0 (±2.1), with mycophenolate mofetil (70%), intravenous corticosteroids (60%), and methotrexate (50%) as the most common. 16 (80%) and 4 (20%) patients were started on oral and topical JAKis, respectively. Mean number of concomitant medications at time of JAKi initiation was 1.7 (±1.4). Of those on oral JAKis, 44% demonstrated complete response, defined as clinical improvement with cessation of disease activity; 44% had partial response, defined as improvement but with persistent activity; and 13% had no response. Among patients on topical JAKis, 25% demonstrated complete response, 50% had no response, and 25% were lost to follow-up. Oral JAKis were well-tolerated, and over half of the cohort (9/16) experienced no side effects. 25% of patients were able to taper off all other medications to maintain disease control, with the mean number of concomitant medications decreasing to 1.5 (±0.8). This is, to our knowledge, the largest cohort to date assessing the role of JAKis in cutaneous fibrotic disorders. Overall, JAKis represent a promising treatment option for patients with morphea and EF, especially those with recalcitrant or progressive sclerosis.

Category: Sclerotic skin disease

MRI PATTERNS AND NEUROLOGIC MANIFESTATIONS IN ADULT EN COUP DE SABRE: INSIGHTS FROM THE LARGEST REPORTED COHORT

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Abstract

En Coup de Sabre (ECDS) is a rare subtype of linear morphea that primarily affects the frontoparietal forehead and scalp. This subtype is associated with neurologic involvement in the form of neurological symptoms or brain lesions detectable on neuroimaging.[1–6] Since ECDS is rare and mainly presents in childhood, literature is largely limited to pediatric reports,[1,3,7,8] leaving neurologic manifestations in adults poorly characterized. Our study examines MRI ordering patterns, imaging findings, and their relationship to neurologic symptoms in an adult ECDS cohort. Neurologists reviewed MRIs to identify abnormalities, assess their potential relationship to ECDS, and determine whether they could be associated with neurologic symptoms. Using the Research Patient Data Registry, we identified 94 adult patients diagnosed with ECDS by a dermatologist or a rheumatologist at two large academic centers (mean age 47.1 years, 87.2% female). Sixty-four patients (68.1%) underwent MRI. The most common abnormalities detected were white matter hyperintensities (27 patients, 42.2%), microhemorrhages (10 patients, 15.6%), and vascular irregularities (6 patients, 9.4%). MRI images were available for review in 42 cases. Of these, 31% were classified as likely related to ECDS, 10% possibly related, 38% unlikely related, and 21% showed no abnormalities. Neurologic symptoms were reported by 53 patients (56.4%). Headache was the most frequently reported symptom (86.8%), followed by seizures (13.2%) and sensory loss (13.2%). Among symptomatic patients, 30 underwent MRI; of these, 27% were likely related, 10% possibly related, and 63% unlikely related. Larger studies are needed to clarify the role of MRI in characterizing neurologic involvement in ECDS and to establish evidence-based guidelines for appropriate imaging. Guidelines could improve diagnostic yield, reduce unnecessary costs, and prevent overutilization. As the largest cohort evaluating neurologic manifestations in adult ECDS, our study offers novel insight into MRI utilization, symptom correlation, and importance of multidisciplinary collaboration in managing these cases.

Abstract Category: Sclerotic skin disease

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Digital Intradermal Hyaluronidase Injections for Sclerodactyly in Systemic Sclerosis: Investigating A Novel Approach to Restoring Digital Mobility

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Sclerodactyly is a disabling manifestation of systemic sclerosis (SSc), characterized by progressive dermal fibrosis that limits digit mobility and predisposes to joint contractures and digital ulcers. Standard medical and rehabilitative therapies often fail to restore function. While intradermal hyaluronidase is under investigation for SSc-associated microstomia, its use for sclerodactyly has not, to our knowledge, been previously described. We report a case of serial digital intradermal hyaluronidase (Hylenex) injections leading to improved digit mobility in SSc-related sclerodactyly. A 55-year-old woman initially presented for microstomia limiting her quality of life. She had a long-standing history of diffuse SSc (ANA+, anti–Scl-70+) managed with a stable dose of mycophenolate mofetil. She had remarkable improvement with intradermal hyaluronidase for microstomia, achieving more than a 1 centimeter improvement in mouth opening. Given this improvement, she was eager to try intradermal hyaluronidase injections for sclerodactyly. Following local nerve block with lidocaine, 150 units of hyaluronidase (1.0 mL) was injected intradermally into the volar and dorsal aspects of one digit. Injections were repeated approximately every 4 weeks over 6 months, progressively including additional digits bilaterally based on clinical response. The patient reported substantial improvement in range of motion, reduction in ulcers, and softening of skin in the treated digits. She also experienced increased independence with activities of daily living and improved quality of life. No adverse events were observed. A post-procedure video and photographs document this response. This case highlights the potential utility of digital intradermal hyaluronidase injections for the management of SSc-related sclerodactyly. Further studies are warranted to investigate the efficacy, optimal dosing, and long-term outcomes.

Word count: 259/300

Teaching Point: Digital Intradermal hyaluronidase injections may offer a novel therapeutic option for SSc-related sclerodactyly.

Category: Clinical case

Clinical images and video attached

Session III:

LUPUS ERYTHEMATOSUS

EFFICACY AND SAFETY OF FIRST-IN-CLASS ORAL SMALL MOLECULE TOLL-LIKE RECEPTOR 7/8 INHIBITOR ENPATORAN IN PATIENTS WITH LUPUS ERYTHEMATOSUS AND ACTIVE CUTANEOUS MANIFESTATIONS: RESULTS OF THE PHASE II WILLOW STUDY

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Cutaneous lupus erythematosus (CLE) may present with or without systemic lupus erythematosus (SLE) and has high unmet treatment needs. Toll-like receptor (TLR) 7/8 signaling has a role in regulating interferon (IFN) induction; elevated type I IFN gene transcripts in lupus skin lesions highlight the importance of the IFN signaling pathway in disease pathogenesis. WILLOW (NCT05162586), a Phase II, randomized, double-blind, placebo-controlled dose-finding parallel adaptive study evaluated the efficacy, safety, and effect on IFN gene signature (IFN-GS) of enpatoran (an oral small molecule TLR7/8 inhibitor) in adults with CLE and/or SLE receiving standard of care. In Cohort A, patients with active cutaneous manifestations (Cutaneous Lupus Disease Area and Severity Index-Activity [CLASI-A] score ≥8 and/or SLE with [British Isles Lupus Assessment Group [BILAG] 2004 ≤1 B, C, D) were randomized 1:1:1:1 to receive one of three enpatoran doses or placebo for 24 weeks (W). The primary objective was dose-response relationship of enpatoran in reducing disease activity, based on change from baseline (BL) in CLASI-A

score at W16. Cohort B, enrolled patients with moderate/severe SLE (BILAG 2004 ≥1A or 2B). Secondary/exploratory endpoints included safety, CLASI-A improvement ≥50%/70% (CLASI-50/70; Cohort A and Cohort B subgroup [CLASI-A score ≥8 at BL]), and change from BL in IFN-GS levels (both cohorts) through W24. Patients were randomized (N=102) to Cohort A, with 100 included in the efficacy analysis. Most patients were female (77%), had CLE only (58%), and had moderate-to-severe disease (71%). Enpatoran demonstrated a significant dose-response in reducing CLASI-A score at W16 (p=0.0002). At W24, 87%/69.6% of enpatoran-treated patients achieved CLASI-50/70 responses versus 30.8%/23.1% for placebo, respectively. Similar trends were seen in Cohort B (Table 1). Enpatoran reduced IFN-GS levels as early as W2, sustained through W24 (Figure 1). Enpatoran was well tolerated across studied doses and no new safety concerns were identified in these participants. Enpatoran showed significant dose response, improved CLASI scores, and reduced IFN-GS levels, supporting TLR7/8 involvement in patients with CLE/and or SLE with predominantly active cutaneous manifestations. Enpatoran was well tolerated; safety profile consistent with that observed in previous studies.

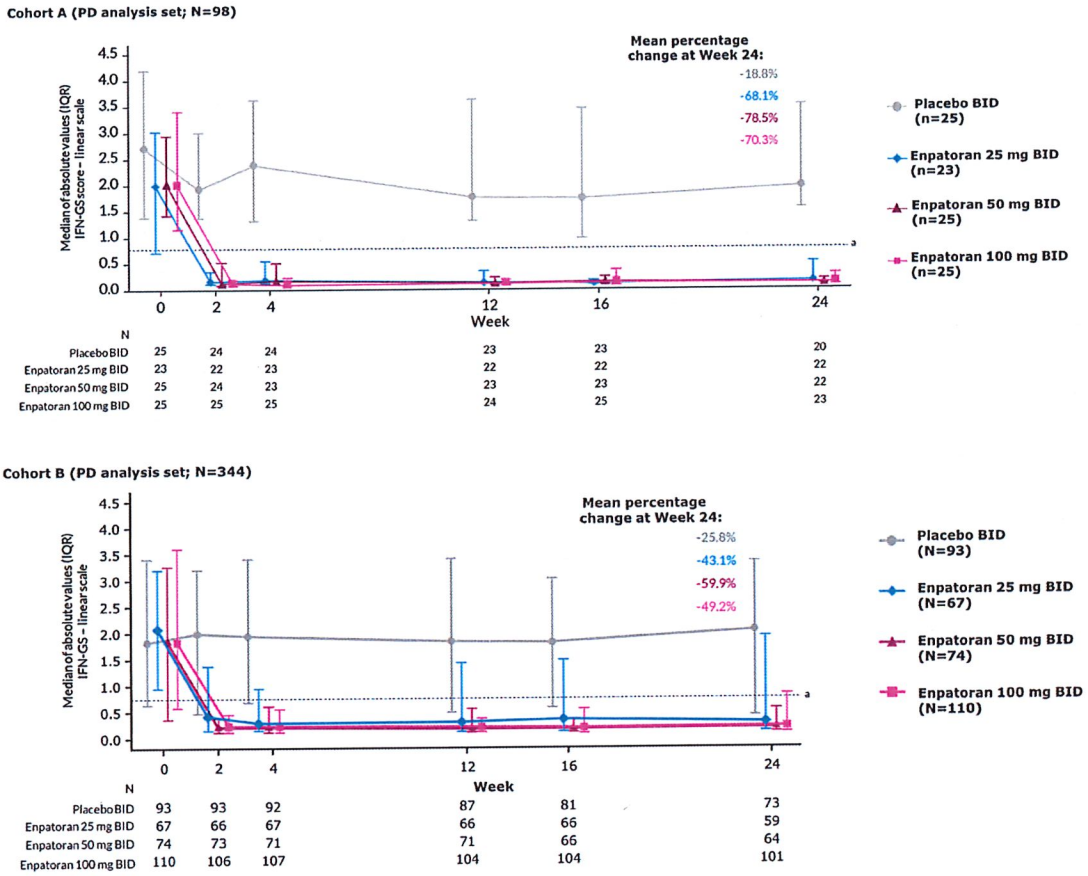
Table 1. Dose-response relationship at Week 16 and CLASI 50/70 response at Week 24 of enpatoran (Cohort A: FAS; N=100; Cohort B: N=162)

Cohort A	Placebo (n=26)	Enpatoran dose			Nominal p- value[a]
		25 mg BID (n=23)	50 mg BID (n=25)	100 mg BID (n=26)	
Primary analysis: Based on MCP-Mod[b]					
Detection of a dose-response relationship			P=0.0002		
Selected model			Log-linear (E0=-44.3, Delta=-6.0)		
Adjusted means in change from BL in CLASI-A score at Week 16, % (95%CI)	-44.3 (-55.1, -33.4)	-63.9 (-70.0, -57.7)	-67.9 (-74.8, -61.1)	-72.0 (-80.1, -64.0)	-
CLASI 50/70 response at Week 24, %	30.8/23.1	87.0/69.6	72.0/52.0	73.1/65.4	0.007/<0.001
Cohort B	**Placebo (n=41)**	**Enpatoran dose**			**Nominal p- value[a]**
		25 mg BID (n=29)	**50 mg BID (n=38)**	**100 mg BID (n=54)**	
CLASI 50/70 response at Week 24, %	41.5/26.8	58.6/41.4	78.9/60.5	81.5/51.9	0.0002/0.0148

[a]Nominal p-value for overall treatment effect of enpatoran versus placebo based on logistic regression. CLASI-50 and CLASI-70 responses were defined as a decrease in CLASI of ≥50% or ≥70%, respectively, from BL values.
[b]MCP-Mod adjusted for CLASI-A at BL, region and disease diagnosis (CLE only vs CLE + SLE).

BID, twice a day; BL, baseline; CI, confidence interval; CLE, cutaneous lupus erythematosus; CLASI-A: Cutaneous Lupus Disease Area and Severity Index-Activity; FAS, full analysis set; MCP-Mod, Multiple Comparison Procedure-Modelling; SLE, systemic lupus erythematosus.

Figure 1. Change from BL in type I IFN-GS score



[a]Dotted line represents the cut-off between participants with low IFN-GS (<0.71) and high IFN-GS (≥0.71) at baseline.
BID, twice a day; BL, baseline; IFN-GS, interferon gene signature; IQR, interquartile range; PD, pharmacodynamic

Category: Lupus

MODULATION OF TYPE I INTERFERON SIGNALING BY ANIFROLUMAB ALTERS THE SPATIAL IMMUNE LANDSCAPE IN CUTANEOUS LUPUS ERYTHEMATOSUS AND DERMATOMYOSITIS

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Reliable and effective treatments for refractory cutaneous lupus erythematosus (CLE) remained elusive until the arrival of anifrolumab, a monoclonal antibody targeting type I interferon receptor subunit 1. To investigate CLE immunopathogenesis and anifrolumab-induced changes, we conducted high-resolution spatial and single-cell analysis from paired lesional and non-lesional skin before and after treatment. Punch biopsies from patients with refractory CLE were collected at baseline and three months post-anifrolumab. Using Chromium Single Cell Gene Expression Flex and Xenium In Situ, we analyzed 27 and 17 formalin-fixed paraffin-embedded biopsies, respectively, identifying 53 distinct cell types. Comparing single cell data from lesional and non-lesional skin biopsies revealed a significant increase in immune cell infiltration, which correlated with elevated levels of cells exhibiting a type I interferon response and proliferative phenotypes (Fig1, A-H). Additionally, pro-inflammatory clusters, including CXCL10-expressing macrophages and cytokine-enriched papillary fibroblasts, emerged in lesional skin (Fig1, E-F). Cell composition analysis demonstrated prevalence of plasmacytoid dendritic cells (pDCs), B cells, plasma cells, and CXCL10+ macrophages exclusively in lesional skin (Fig1, E-F). Spatial mapping unveiled two distinct clusters of immune aggregates in CLE lesional skin: one highly enriched mregDCs and pDCs near basal keratinocytes, and another with prominent perivascular macrophages (Fig2, A). Furthermore, following anifrolumab, reduced type I interferon signaling led to the disappearance of disease-associated clusters and depletion of spatially-organized immune aggregates (Fig2, B-D). Building on these findings, an analysis of three pre- and post-treatment dermatomyositis samples, which is another connective tissue disease driven by Type I interferon, is forthcoming. In conclusion, type I interferon signaling inhibition induces a shift of lesional skin transcriptome towards the non-lesional phenotype, characterized by resolution of pro-inflammatory clusters and spatially-distinct immune aggregates. Notably, despite clinical improvement, treatment with anifrolumab did not result in the conversion of either the lesional or non-lesional skin transcriptome into the healthy phenotype.

Category: Lupus and Dermatomyositis

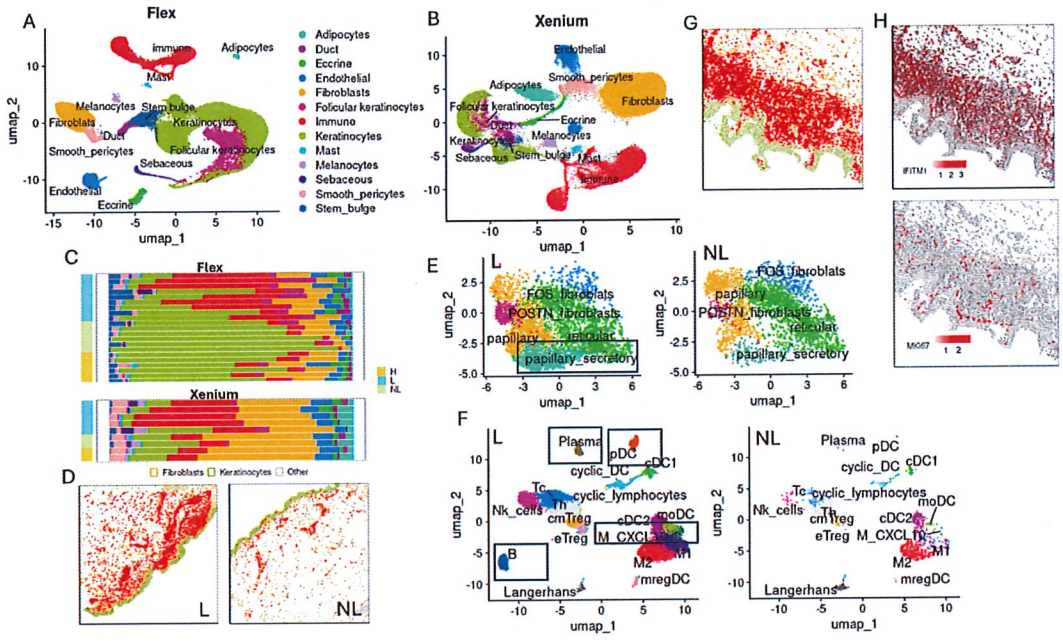

Figure1

(A) Uniform Manifold Approximation and Projection (UMAP) plot of cells from 27 skin biopsies, identified by single cell FFPE sequencing (scFFPE-seq). Sample composition: 9 lesional untreated (L), 6 healthy (H), 6 non-lesional untreated (NL), 3 lesional treated (TL), and 3 non-lesional treated (TNL). **(B)** UMAP plot of cells from 17 skin biopsies, identified by Xenium, clustered into 13 major cell types. Sample composition: 5 lesional untreated (L), 2 healthy (H), 3 non-lesional untreated (NL), 4 lesional treated (TL), and 3 non-lesional treated (TNL). **(C)** Proportion of each cell type in skin samples sequenced by scFFPE-seq and Xenium. **(D)** Cell type map from Xenium data. Colors: green - keratinocytes, yellow – fibroblasts, red – immune cells, grey – other cells. **(E-F)** UMAP analysis of fibroblast and immune cell transcriptomes in lesional (L) and non-lesional (NL) skin. **(G)** Cell type map from Xenium data in lesional skin. Colors: green - keratinocytes, yellow – fibroblasts, red – immune cells, grey – other cells. **(H)** Xenium spatial plot showing normalized expression of interferon-response marker IFITM1 and proliferative marker MKI67 in lesional skin.

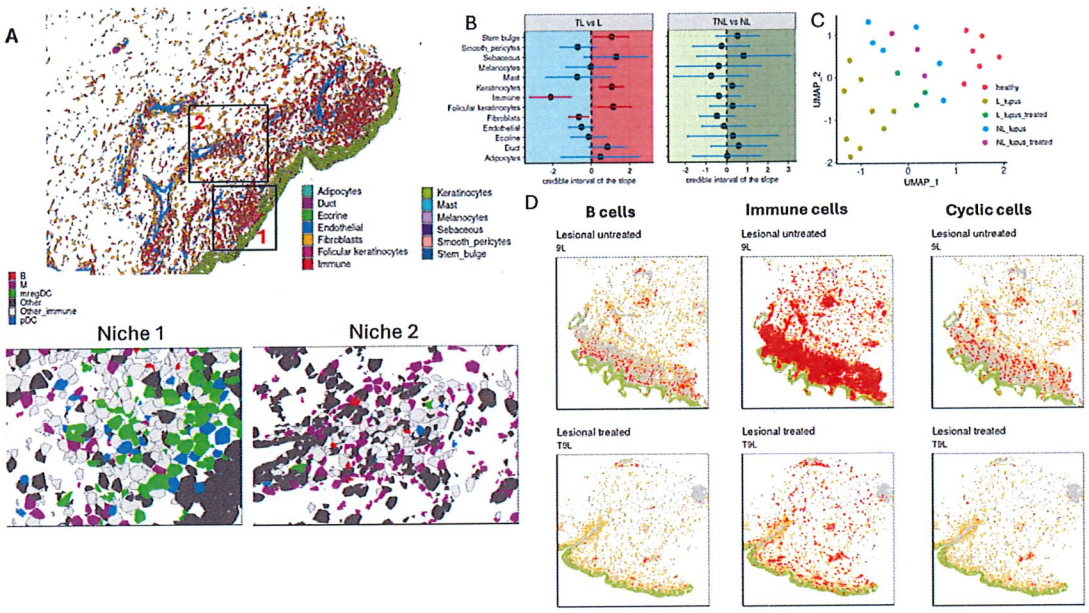

Figure 2

(A) Cell type map from Xenium data in lesional skin with two representative immune niches. **(B)** A plot of estimates of differential composition analysis for major cell types between treated and untreated lesional and non-lesional skin samples. The error bars represent 95% confidence intervals. The error bar is blue if the composition of the target cell type is insignificant and red if significant. **(C)** UMAP of Multicellular Factor Analysis factor scores showing the variability of all cell types captured by single cell Flex sequencing. **(D)** Cell type map from Xenium data in lesional skin before and after treatment. Colors: green - keratinocytes, yellow - fibroblasts, red - B cells/immune cells/cells with a proliferative phenotype.

PREVALENCE OF ANTIPHOSPHOLIPID ANTIBODIES IN CUTANEOUS LUPUS ERYTHEMATOSUS WITH AND WITHOUT SYSTEMIC DISEASE

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The prevalence of antiphospholipid antibodies (APLAs) in systemic lupus erythematosus (SLE) has been extensively investigated. However, only a few studies have assessed their prevalence in cutaneous LE (CLE) with discordant findings. This retrospective cross-sectional study analyzed 288 patients in our institutional review board-approved CLE database at the University of Pennsylvania who received APLA testing between January 2007 and June 2025. CLE and SLE were diagnosed according to clinical and pathological criteria. We evaluated the presence of the immunoglobulin G (IgG) isotype of lupus anticoagulant (LA), anticardiolipin antibody (ACA), and anti-β2-glycoprotein I antibody (anti-β2GPI). The APLA positivity rates in CLE patients with and without SLE were compared using chi-square and Fisher's exact tests with $\alpha = 0.05$. One or more of the APLAs were found in 9% (10/116) of the patients with CLE only and in 25% (43/172) of those with both CLE and SLE, $p = 0.0004$. LA and ACA were significantly less common in the CLE-only group (LA 4/84 [5%], ACA 5/113 [4%]) than in the CLE/SLE group (LA 23/151 [15%], ACA 27/171 [16%]), $p = 0.02$ and 0.003 respectively. No significant difference in APLA prevalence was found between CLE-only patients with and without antinuclear antibodies (ANA) (5/68 [7%] vs 5/46 [11%], $p = 0.5$). Among the CLE-only patients, 8/10 (80%) of those with APLA(s) had discoid LE (DLE) whereas 68/116 (59%) of the entire group had DLE, $p = 0.3$. Out of the 10 CLE-only patients with APLA(s), 1 had antiphospholipid syndrome with a history of thrombotic events. The prevalence of APLAs was significantly lower in our CLE-only patients compared to our CLE patients with SLE, regardless of the presence of ANA. Therefore, it may be of limited value to routinely screen for APLAs in CLE-only patients in the absence of compelling clinical indications.

Category: Lupus

UNBIASED IDENTIFICATION AND VALIDATION OF RNA MARKERS FOR CUTANEOUS LUPUS ERYTHEMATOSUS

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Distinguishing cutaneous lupus erythematosus (CLE) from inflammatory mimics remains diagnostically challenging, particularly when histopathologic features overlap. While CD123 immunohistochemistry identifies plasmacytoid dendritic cells which can be used as a diagnostic adjunct, it requires adequate dermis and provides limited information about interferon pathway activation. We identified and validated RNA in situ hybridization (RISH) markers that directly visualize the type I interferon signature characteristic of CLE pathophysiology. Transcriptomic analysis of published data identified interferon-stimulated genes *ISG15* and *IFI6* among the most significantly upregulated transcripts in discoid lupus. We performed RISH on 11 CLE samples, 8 normal controls, and 11 inflammatory dermatoses (atopic dermatitis, psoriasis, lichen planus) from our dermatopathology archives. Two blinded dermatopathologists scored staining intensity (0-4+) and distribution patterns. CLE demonstrated significantly higher mean staining intensity ($P < 0.05$) for both markers compared to all other conditions: *ISG15* showed 3.2±0.3 epidermal and 3.1±0.3 dermal scores in CLE versus ≤1.5 in comparators. Pattern analysis revealed diagnostic signatures unique to CLE: basal keratinocyte involvement (91-100%), follicular positivity (45-55%), and most importantly, deep dermal staining (64-73%). Deep dermal staining was exclusively observed in CLE and represents a specific diagnostic feature. While lichen planus exhibited basal staining consistent with interface dermatitis, it lacked the widespread dermal positivity characteristic of CLE. Principal component analysis confirmed clear separation of CLE from other diagnoses based on combined intensity and pattern features. These markers offer practical advantages over CD123, providing assessment across multiple tissue compartments even in superficial biopsies. Beyond diagnosis, RISH for *ISG15* and *IFI6* may identify candidates for interferon-targeted therapies like anifrolumab and enable monitoring of treatment response. This approach provides an objective diagnostic tool that can be implemented in routine pathology laboratories for challenging cases where clinical and histopathologic features are ambiguous.

Category: Lupus

CONCOMITANT INTRAVENOUS IMMUNOGLOBULIN AND ANIFROLUMAB IN LUPUS AND DERMATOMYOSITIS PATIENTS: REAL-WORLD DATA FROM A RETROSPECTIVE COHORT STUDY

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Word count: 300
Category: lupus, dermatomyositis

Anifrolumab, a monthly infusion FDA-approved for systemic lupus erythematosus (SLE), is increasingly being utilized for cutaneous LE (CLE) and dermatomyositis (DM).[1,2] Intravenous immunoglobulin (IVIG) is another infusion therapy used for these conditions, with several cases reporting patient improvement on concomitant treatment.[3,4,5] However, the timing of IVIG (typically administered every 2 or 4 weeks) with respect to anifrolumab remains to be addressed. We examined real-world anifrolumab and IVIG administration to identify patterns in timing, associated adverse effects, and treatment response. In a cohort of 80 patients receiving anifrolumab at two U.S.-based academic medical centers between January 2022 and April 2025, 9 patients received concomitant IVIG. Of these 9 patients, 89% were female with a mean age of 39 years. Indication for anifrolumab treatment was most commonly CLE (55%), followed by DM (33%), and SLE/systemic sclerosis overlap (11%). Six patients received IVIG every 4 weeks, while 3 received it every 2 weeks. IVIG timing after anifrolumab infusion was variable, ranging from the same day to 25 days later. IVIG was discontinued in one patient due to difficulty coordinating infusions, one was discontinued due to disease improvement, and another was discontinued due to a pulmonary embolism. The most common adverse effect of IVIG was headache in 6 patients (66%), all of whom had experienced these before starting anifrolumab. 8 of 9 patients experienced improvement in their skin disease. One patient reported worsening of headaches during the first IVIG infusion post-anifrolumab initiation (separated by 25 days), which did not recur on subsequent infusions. Rates of discontinuation of anifrolumab among patients on both IVIG and anifrolumab did not differ significantly from those on anifrolumab alone (33% versus 32%). This limited cohort indicates that in patients on both IVIG and anifrolumab, the relative timing of infusions varies widely but may not significantly impact adverse effects or efficacy.

1. Shaw K, Sanchez-Melendez S, Taylor D, *et al.* Assessment of clinical response to anifrolumab in patients with refractory discoid lupus erythematosus. *JAMA Dermatol.* 2023;159(5):560-3.
2. Shaw KS, Hashemi KB, Castillo RL, *et al.* Anifrolumab in recalcitrant cutaneous dermatomyositis: a multicenter retrospective cohort study. *J Am Acad Dermatol.* 2024;91(6):1217-9.
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OUTCOMES IN DISEASE ACTIVITY IMPROVE UP TO TWO YEARS OF STANDARD-OF-CARE TREATMENT IN PATIENTS WITH ACTIVE CUTANEOUS LUPUS ERYTHEMATOSUS

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The Cutaneous Lupus Erythematosus Disease Area and Severity Index activity (CLASI-A) score is an established clinical trial endpoint measuring cutaneous lupus erythematosus (CLE) disease activity. Long-term studies of disease outcomes beyond six-months are lacking in patients with active discoid lupus erythematosus (DLE) or subacute cutaneous lupus erythematosus (SCLE), who are commonly studied in clinical trials. A cohort of 48 adult CLE patients on standard-of-care (SoC) therapies with baseline CLASI-A≥8 were recruited from outpatient dermatology clinics at University of Texas Southwestern and Parkland Health between 01 January 2009 to 30 June 2024. Additional inclusion criteria were at least one follow-up visit within six-months and/or two-years [+/- three-months] and a diagnosis of DLE and/or SCLE. This study evaluated observed outcomes using change in CLASI-A at 6-months and 2-years from initial visit, CLASI-A improvement ≥50% (CLASI-50) and ≥70% (CLASI-70), and CLASI-A≤3. Our population was 35.4% White, non-Hispanic and 43.8% Black, non-Hispanic. Median CLASI-A change was -5.00 (IQR: -10.00, -1.00) at six-months (n=38) and -8.00 (IQR: -9.75, -1.50) at two-years (n=22). At six-months, 39.50% of patients achieved CLASI-50 (95% confidence interval (CI): 25.60%–55.28%) while at two-years, 50.00% of patients achieved CLASI-50 (95% CI: 30.72%–69.28%). CLASI-70 was achieved by 26.30% (95% CI: 14.97%–42.01%) of patients at six-months and 40.90% (95% CI: 23.26%–61.27%) of patients at two-years. 21.10% (95% CI: 11.07%–36.35%) of patients achieved CLASI-A≤3 at six-months and 36.40% (95% CI: 19.73%–57.05%) at two-years. Limitations include narrow geographic distribution and missed follow-up visits. Overall, these results provide insight into expected real-world improvement on SoC therapies in the short- and long-term for CLE patients. Despite receiving SoC treatments, at two-years, less than half of active patients achieved CLASI-70 or CLASI-A≤3, which demonstrates

the unmet need for novel therapies. These results provide important disease prognosis information for CLE patients under SoC treatments.

Category: Lupus

ZOSTER RISK AND VACCINATION STATUS IN LUPUS AND DERMATOMYOSITIS PATIENTS ON ANIFROLUMAB: REAL-WORLD DATA FROM A RETROSPECTIVE COHORT STUDY

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Word count: 300
Category: lupus, dermatomyositis

Anifrolumab is a type-I interferon receptor antagonist FDA-approved for the treatment of systemic lupus erythematosus (SLE), with preliminary data demonstrating promising results in cutaneous lupus erythematosus (CLE) and dermatomyositis (DM).[1,2] Increased risk of herpes zoster (HZ) in patients with CLE and DM has been previously shown,[3] and anifrolumab is independently associated with an increased HZ risk.[4,5] Despite this, there are currently no guidelines for HZ prevention or prophylaxis amongst those receiving anifrolumab.[6] Our study sought to examine real-world anifrolumab use and HZ risk in CLE and DM patients. We present a cohort of 80 patients treated with anifrolumab at two U.S.-based academic medical centers between January 2022 and April 2025 whose HZ vaccination, prophylaxis, and infectious status were retrospectively collected. Our cohort were 89% female and 56% White, with a mean age of 44 years (SD 14) at the time of anifrolumab initiation. Among this cohort, 40 (50%) never received HZ vaccination, 35 (44%) received at least one HZ vaccine dose prior to initiation of anifrolumab, and 5 (6%) received their first HZ vaccine after initiation. Of the 80 patients, 18 (23%) had a history of HZ prior to anifrolumab, two (11%) of whom were on prophylactic valacyclovir. 51 (64%) received concomitant immunosuppressants during anifrolumab treatment. Three patients (4%) developed HZ, two of whom had a history of it. One had been fully vaccinated prior to initiation, while one received a single dose of recombinant HZ vaccine after initiating anifrolumab. All three were receiving anifrolumab for CLE. One patient was receiving no concomitant immunosuppressants, one was on systemic steroids, and one was receiving both mycophenolate and low dose steroids at the time of infection. Our study contributes additional reassuring real-world data suggesting that even with relatively low rates of vaccination and prophylaxis, the rate of HZ remained low in those treated with anifrolumab.

1. Shaw K, Sanchez-Melendez S, Taylor D, *et al.* Assessment of clinical response to anifrolumab in patients with refractory discoid lupus erythematosus. *JAMA Dermatol.* 2023;159(5):560-3.
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ARYL HYDROCARBON RECEPTOR EXPRESSION IN CUTANEOUS LUPUS: A CLINICOPATHOLOGIC CORRELATION

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Abstract:

Aryl hydrocarbon receptor (AHR) is a ligand-activated transcription factor expressed by multiple cell types that plays an important role in skin immune response, barrier function, and homeostasis. It has gained attention as a potential target for many inflammatory and autoimmune diseases, and in systemic lupus erythematosus (SLE), has been found to inhibit the B-cell chemoattractant CXCL13, a chemokine induced by type I interferon. AHR has not been thoroughly explored in discoid lupus erythematosus (DLE). This study aimed to determine whether AHR is expressed in patients with DLE. Skin biopsies of DLE (n=6), psoriasis for positive control (n=4), and normal ex-vivo skin (NS) controls (n=4) were obtained from patients seen at the University of Miami. Immunohistochemistry staining with anti-AHR antibody produced in rabbit was performed. Quantification of positive cells per mm^2 was conducted with QuPath analysis software. Two-way ANOVA followed by Turkey's multiple comparison test were performed for statistical analysis ($p < .05$). Fourteen total biopsies were stained and analyzed. Compared to NS, there was a significant difference between epidermal and dermal AHR expression in both DLE and psoriasis, with decreased AHR expression in the epidermis, and increased expression of AHR in the dermis, localizing to areas of lymphocytic infiltration. There were no significant differences between DLE and psoriasis. The cause of decreased expression of AHR in the epidermis is unclear, however, the similar expression pattern to psoriasis may indicate potential benefit with medications that target this pathway. These findings suggest that AHR may play a role in DLE pathogenesis, and may be a viable therapeutic target.

Category: Lupus

EFFECT OF LITIFILIMAB ON CUTANEOUS LUPUS ERYTHEMATOSUS DISEASE AREA AND SEVERITY INDEX–ACTIVITY (CLASI-A) SUBCOMPONENTS AND PHYSICIAN GLOBAL ASSESSMENT–SKIN (PGA–SKIN) IN PATIENTS WITH CUTANEOUS LUPUS ERYTHEMATOSUS (CLE) IN A PHASE 2 STUDY

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Word count body: max 300 words (294/300)

The randomized, placebo-controlled Phase 2 LILAC study of litifilimab (NCT02847598), Part B (active CLE with/without systemic lupus erythematosus [SLE]) met its primary endpoint of percentage change from baseline in CLASI-A score at Week 16, with a greater proportion achieving a CLASI-50 response (≥50% decrease in CLASI-A score) with litifilimab than placebo.[1] The CLASI-A measures disease activity in CLE based on Erythema, Scale/Hypertrophy, Non-scarring Alopecia, Recent Hair Loss (past 30 days), and Mucous Membrane Lesions.[2] These post-hoc analyses assessed individual CLASI-A subcomponent contributions to overall score change, and the physician-reported PGA–Skin was also analyzed. Erythema and scale/hypertrophy were categorized by baseline score quartiles; other subcomponents were analyzed as recorded. PGA–Skin was rated as mild, moderate, or severe. For treatment failures, the baseline value or value from the last visit before treatment failure was carried forward, whichever was worse; post-discontinuation data were censored. At baseline, across groups, significant activity was present in 72.7–84.0% for erythema, 73.1–85.4% for scale/hypertrophy, 60.4–76.9% for alopecia, 45.5–61.5% for recent hair loss, and 12.1–29.2% for mucous membrane lesions (**Table 1**). Litifilimab-treated groups had consistently greater improvements versus placebo for erythema, scale/hypertrophy, and alopecia at all post-baseline visits (**Figure 1** and **Table 1**). Due to low baseline incidence, no conclusions were drawn for mucous membrane lesions. For the PGA–Skin at Week 16, 28.0% of placebo-treated participants and 45.8–60.0% of litifilimab-treated participants were rated with mild disease (**Figure 2**). These results suggest that erythema, scale/hypertrophy, alopecia, and recent hair loss contributed to CLASI-A improvements at Week 16, with greater improvements in litifilimab-treated participants. Combined with a greater proportion of participants achieving mild overall skin condition in the litifilimab groups, these findings support further development of litifilimab in CLE.

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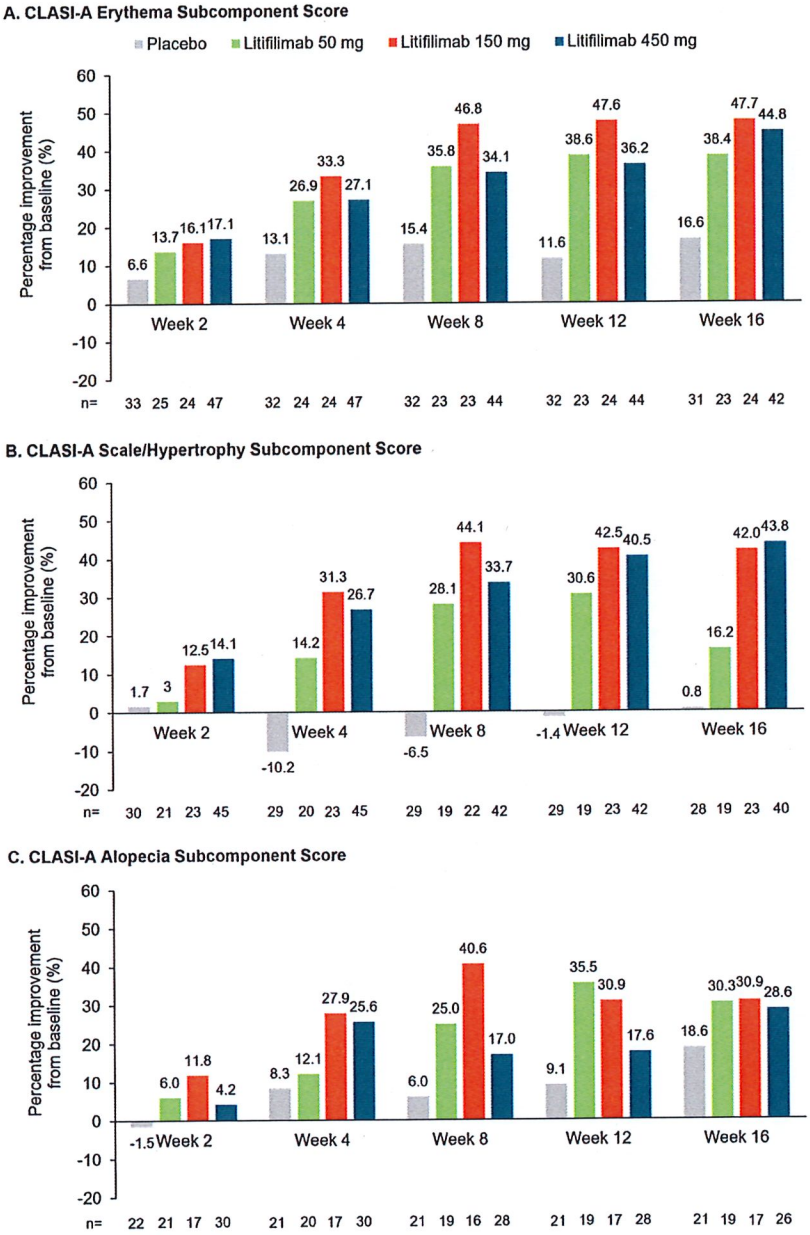
First presented at ACR 2024.

Abstract Category: Lupus

Figure 1. Percentage improvement from baseline in CLASI-A subcomponents scores by visit (MITT population)



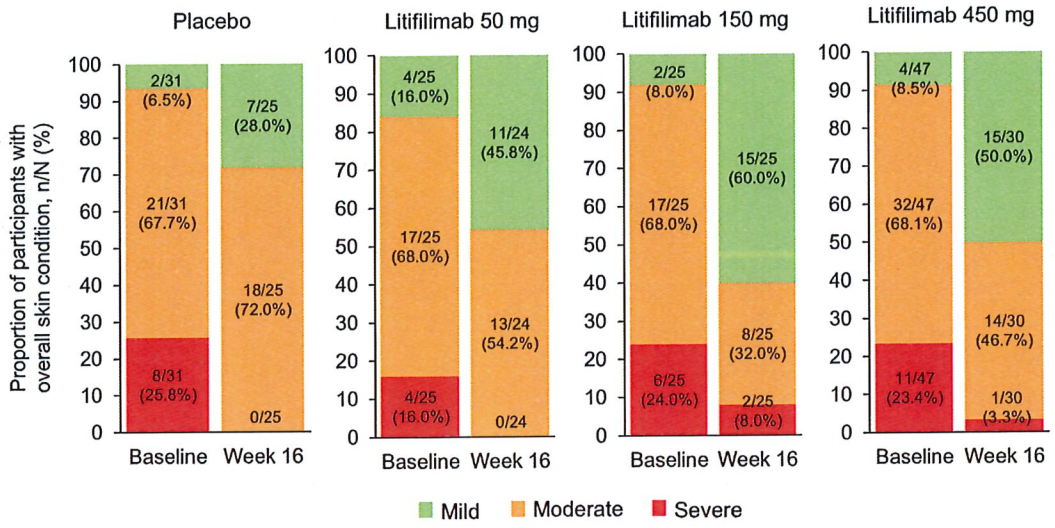
A CLASI-A erythema score ≥2 was an enrollment criterion for Part B of the LILAC study.
For participants from Part B protocol version 1 who completed treatment up to Week 12 but could not reconsent to protocol version 2, Week 16 data were imputed using the predicted value from the mixed model for repeated measures model of absolute value.
CLASI-A, Cutaneous Lupus Erythematosus Disease Area and Severity Index–Activity; MITT, modified intention-to-treat.

Table 1. CLASI-A subcomponents scores at baseline and Week 16 (MITT population)

CLASI-A Subcomponents	Placebo		Litifilimab 50 mg		Litifilimab 150 mg		Litifilimab 450 mg	
	BL	Wk 16	BL	Wk 16	BL	Wk 16	BL	Wk 16
Erythema*								
n	33	31	26	23	25	24	48	42
Score <5.5 (%)	27.3	35.5	26.9	69.6	16.0	58.3	27.1	73.8
5.5≤ score <8.5 (%)	24.2	25.8	30.8	13.0	24.0	16.7	22.9	4.8
8.5≤ score <13 (%)	18.2	19.4	30.8	8.7	20.0	12.5	27.1	4.8
Score ≥13 (%)	30.3	19.4	11.5	8.7	40.0	12.5	22.9	16.7
Scale/Hypertrophy*								
n	33	31	26	23	25	24	48	42
Score <2 (%)	18.2	29.0	26.9	30.4	16.0	50.0	14.6	47.6
2≤ score <3 (%)	12.1	9.7	23.1	39.1	20.0	8.3	16.7	19.0
3≤ score <6 (%)	36.4	32.3	30.8	13.0	24.0	33.3	39.6	16.7
Score ≥6 (%)	33.3	29.0	19.2	17.4	40.0	8.3	29.2	16.7
Alopecia								
n	33	31	26	23	25	24	48	42
Score = 0, absent (%)	36.4	38.7	23.1	34.8	36.0	54.2	39.6	52.4
Score = 1, diffuse, non-inflammatory (%)	12.1	9.7	19.2	21.7	24.0	16.7	22.9	14.3
Score = 2, focal or patchy in 1 quadrant (%)	12.1	6.5	7.7	4.3	4.0	0	4.2	2.4
Score = 3, focal or patchy in more than 1 quadrant (%)	39.4	45.2	50.0	39.1	36.0	29.2	33.3	31.0
Recent Hair Loss								
n	33	31	26	23	25	24	48	42
Score = 0, 'no' (%)	54.5	77.4	38.5	73.9	44.0	66.7	47.9	71.4
Score = 1, 'yes' (%)	45.5	22.6	61.5	26.1	56.0	33.3	52.1	28.6
Mucous Membrane Lesions								
n	33	31	26	23	25	24	48	42
Score = 0, 'absent' (%)	87.9	96.8	84.6	100	80.0	91.7	70.8	81.0
Score = 1, lesion or ulceration (%)	12.1	3.2	15.4	0	20.0	8.3	29.2	19.0

*Erythema and scale/hypertrophy are presented in 4 categories based on the quartiles of the baseline scores of the subcomponents across all LILAC Part B participants.
A CLASI-A erythema score ≥2 was an enrollment criterion for Part B of the LILAC study.
For participants from Part B protocol version 1 who completed treatment up to Week 12 but could not reconsent to protocol version 2, Week 16 data were imputed using the predicted value from the mixed model for repeated measures model of absolute value.
BL, baseline; CLASI-A, Cutaneous Lupus Erythematosus Disease Area and Severity Index–Activity; MITT, modified intention-to-treat; Wk, week.

Figure 2. Proportions of participants with overall PGA-Skin condition of mild, moderate, or severe at baseline and Week 16 (MITT population)



Data scores are presented as recorded.
BL, baseline; MITT, modified intention-to-treat; PGA–Skin, Physician Global Assessment–Skin.

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BASELINE CLE DISEASE ACTIVITY INFLUENCES THE PATIENT PERSPECTIVE OF CLINICALLY MEANINGFUL IMPROVEMENT

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The Cutaneous Lupus Erythematosus Disease Area and Severity Index Activity score (CLASI-A) has been used as an outcome measure assessing disease activity in clinical trials for cutaneous lupus erythematosus (CLE). However, the threshold of change in CLASI-A corresponding to clinically meaningful improvement in patient- and physician-reported outcomes has not been well-characterized. Thus, we performed a prospective study to determine the CLASI-A threshold associated with clinically meaningful improvement in different outcome measures using anchor-based methods. 107 CLE patients were recruited from outpatient dermatology clinics at University of Texas Southwestern Medical Center, Parkland Health, and University of Pennsylvania between July 2018 and November 2023. CLASI-A scores and outcome measures including SKINDEX-29+3, Patient Impression of Disease Progression (PIDP), and Physician's Global Assessment of Disease Activity (PGA-A) were collected at baseline and after 6 months. Linear regression analyses were performed between absolute and percent change in CLASI-A versus absolute change in SKINDEX-29+3, PIDP, and PGA-A after 6 months. Analyses were stratified by baseline CLASI-A severity, with CLASI-A 8-19 and CLASI-A $\geq$ 20 corresponding to moderate and severe activity, respectively. Threshold of CLASI-A improvement was derived from the quotient of previously published anchor-based changes in outcome measures and their corresponding regression slopes. For patients with upper mild-moderate baseline activity (n=61), the threshold of CLASI-A change ranged from 66.2%-100.2% and 9.7-11.6 (PIDP=100.2%, 11.6; SKINDEX symptoms=66.2%, 9.7; PGA-A=76.2%, 10.6). In contrast, the threshold of change in CLASI-A for severe baseline activity (n=29) ranged from 34.3%-53.1% and 10.5-15.7 (PIDP=53.1%, 15.7; SKINDEX symptoms=34.3%, 10.5; PGA-A=41.1%, 12.6). These findings suggest that the threshold of meaningful change in CLASI-A may be influenced by the severity of baseline disease activity, which has important implications for the future design of CLE clinical trials.

Category: Lupus

Session IV:

DERMATOMYOSITIS

Title: Rapid Improvement in Skin-Predominant Dermatomyositis with Brepocitinib: Clinical and Molecular Results from a Phase 2 Clinical Trial

Authors: Aaron Mangold*, Ruth Ann Vleugels*, Alisa Femia, Julie J. Paik, Rundong Jiang, Jeffrey R. Gehlhausen, Rochelle L. Castillo, Jennifer Fox, Rachael Bogle, Bryce Roberts, Scott Penner, Xing Li, Zeni Ramirez, J. Michelle Kahlenberg, Lam C. Tsoi, Katharina S. Shaw, Matthew D. Cascino, Paul N. Mudd Jr, Lisa Christopher-Stine, Victoria P. Werth**, Johann E. Gudjonsson**

Background: Dermatomyositis (DM) is characterized by overactivation of type I and II interferon (IFN) and additional inflammatory cytokines that signal via the JAK-STAT pathway. In skin, this proinflammatory microenvironment is driven by multiple cell types, including T cells, myeloid cells, and keratinocytes (KCs). We sought to evaluate brepocitinib, a once-daily (QD), oral TYK2/JAK1 inhibitor, in DM patients with skin-predominant disease, assessing both clinical endpoints and disease-modifying effects at the cellular level.

Methods: We conducted a 12-week, open-label, Phase 2 trial of brepocitinib in adult DM patients with skin-predominant disease (NCT06433999) and utilized single-cell and spatial transcriptomics to identify key immune pathways linked to treatment response. Eligible patients had moderate-to-severe skin disease (defined as Cutaneous Dermatomyositis Disease Area and Severity Index-Activity [CDASI-A] score ≥14) and no-to-minimal muscle involvement (defined as Manual Muscle Test-8 score >142). Brepocitinib 30 mg QD was given for 12 weeks. The primary outcome was change in CDASI-A score, for which the minimal clinically important difference has been reported as a 4-point or 40% decrease from baseline. Lesional and non-lesional skin biopsies were obtained at weeks 0 and 4 for spatial and single-cell RNA sequencing.

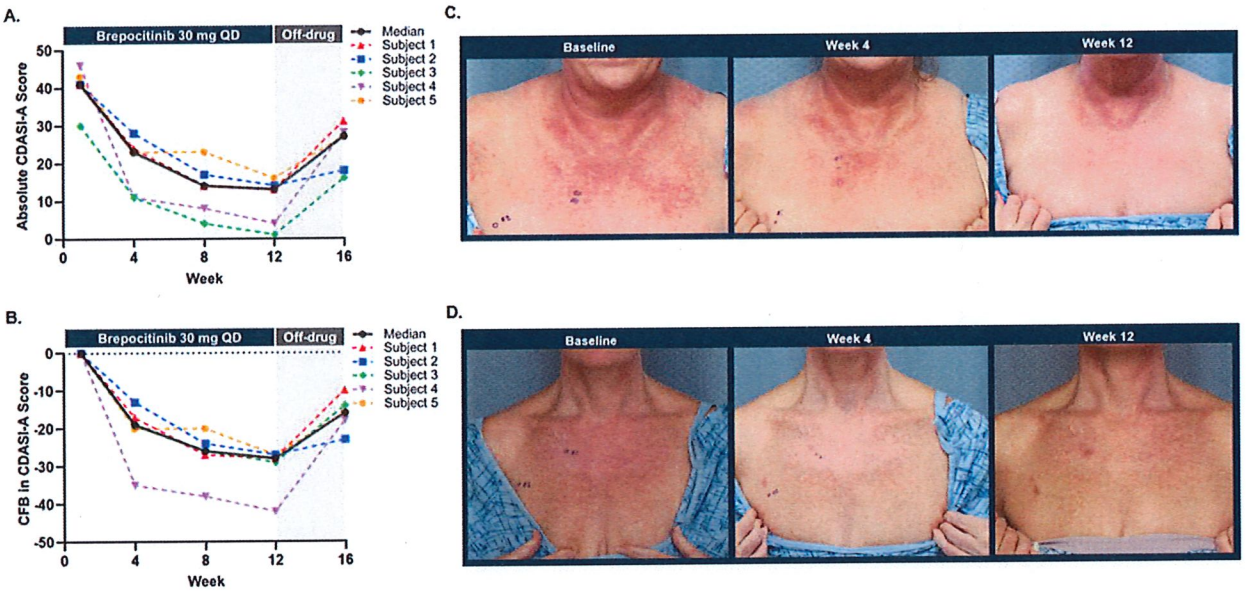
Results: Five patients (mean age, 53 years; mean [range] baseline CDASI-A score, 40.2 [30-46]) were enrolled. At week 4, CDASI-A mean change [SD] was -20.8 [8.4] and mean percentage change [SD] was -51.8% [17.8%], with additional improvement observed by week 12 (Fig 1). At baseline, lesional skin demonstrated increased type I/II IFN signaling (Fig 2A,C) and infiltrating immune cells (Fig 3A-D). By week 4, there was marked attenuation of type I/II IFN signaling (Fig 2B,D), particularly at the dermal-epidermal junction, and reductions in CD4+ and CD8+ T cells, NK cells, myeloid dendritic cells, and B cells (Fig 3A-D). Among cell types isolated from lesional skin, KCs were the most transcriptionally responsive, with basal and cycling subsets showing broad downregulation of IFN-stimulated genes and chemokine production (Fig 3E).

Conclusions: Brepocitinib induced rapid and clinically meaningful improvement in severe, skin-predominant DM within 4 weeks of treatment. Integrated spatial and single-cell

transcriptomic analyses revealed marked reductions in type I/II IFN signaling, decreased immune cell infiltration, and resolution of the proinflammatory microenvironment. Collectively, these findings support the potential of brepocitinib as a rapid-acting, targeted, disease-modifying oral therapy for DM and provide confirmation of the molecular basis for the observed clinical benefits.

Figure 1. Clinical Responses to Brepocitinib in Skin-Predominant Dermatomyositis

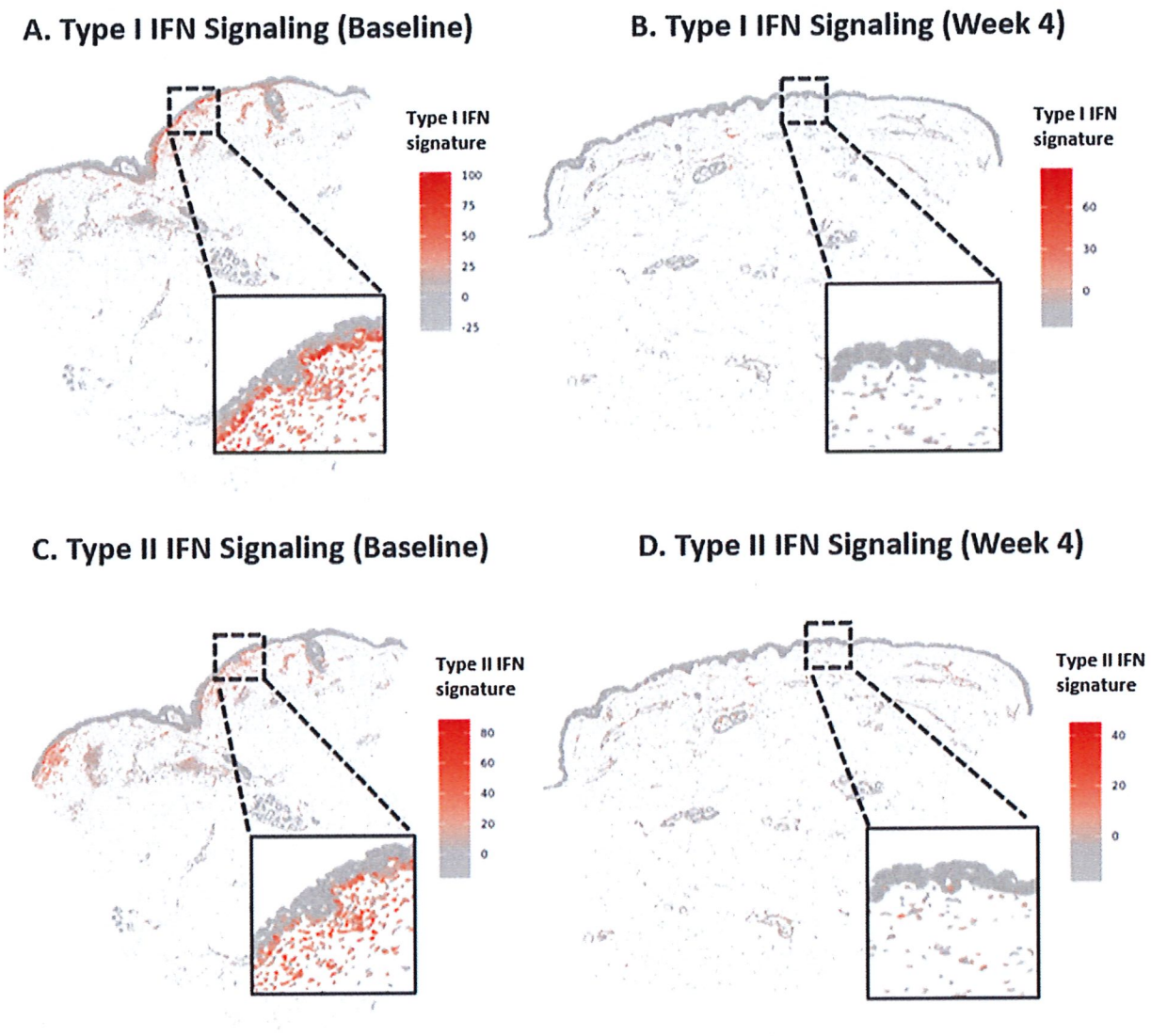
Absolute change **(A)** and change from baseline **(B)** in CDASI-A are shown for individual patients (colored dashed lines) and the group median (black solid line). Weeks 0-12 represent the treatment period with brepocitinib 30 mg once-daily; weeks 12-16 represent the 4-week off-drug follow-up period. Rapid and clinically meaningful improvements were observed during the treatment period, with partial loss of response after drug discontinuation. Representative clinical images from two study participants **(C, D)** demonstrate improvement in characteristic cutaneous dermatomyositis with brepocitinib.



Abbreviations: CDASI-A = Cutaneous Dermatomyositis Disease Area and Severity Index – Activity (CDASI-A scores range from 0-100 with higher scores indicating more severe skin disease activity); CFB = change from baseline

Figure 2. Marked Attenuation of Type I and II IFN Signaling in Lesional Dermatomyositis Skin by Week 4 of Brepocitinib Treatment

Spatial RNA sequencing of lesional dermatomyositis skin biopsies obtained at baseline demonstrated prominent aggregation of **(A)** type I and **(C)** type II interferon (IFN) signals in the superficial dermis. By week 4 of brepocitinib treatment, marked decreases in **(B)** type I and **(D)** type II interferon signaling were observed.

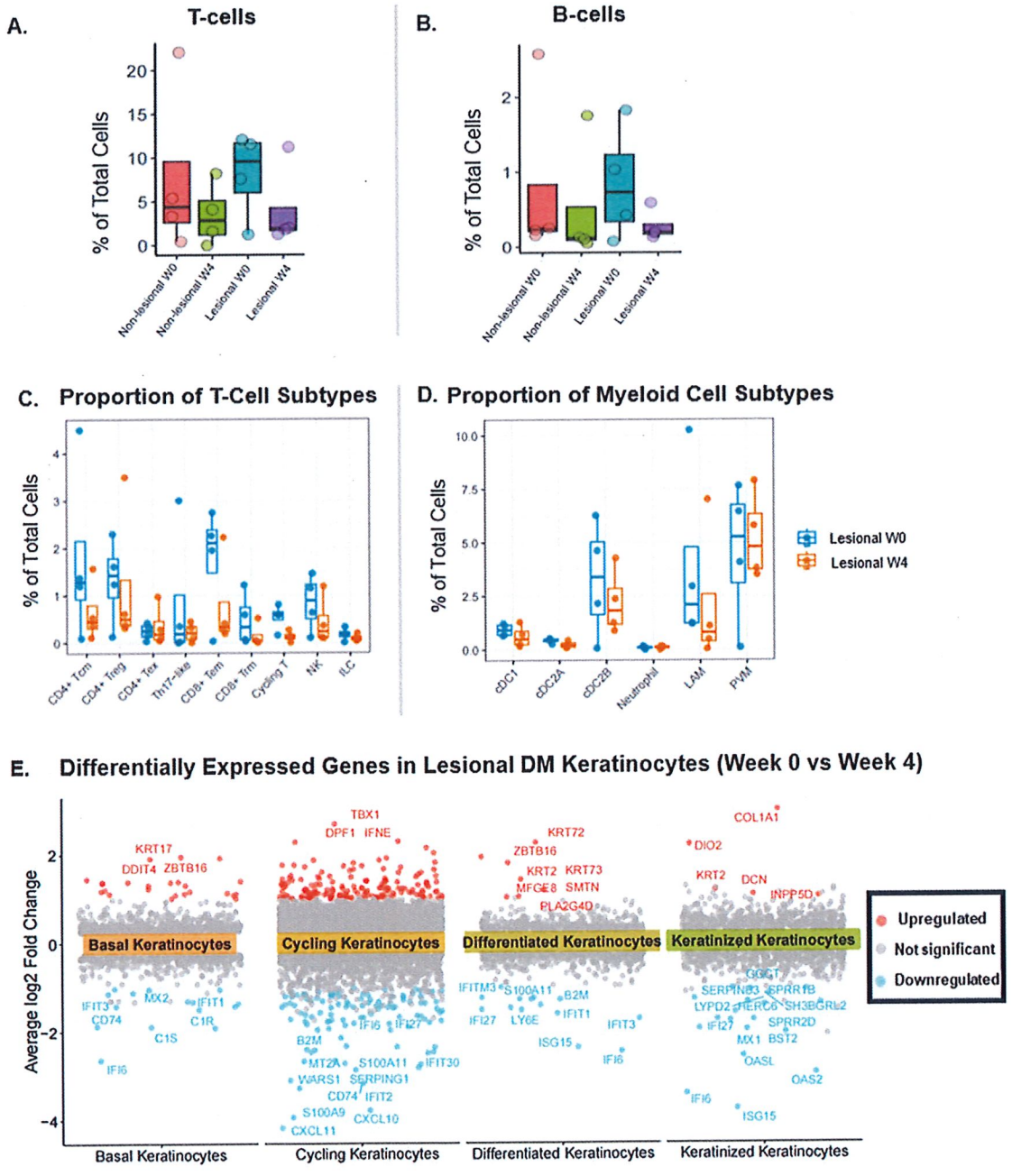


Representative spatial transcriptomic images of lesional dermatomyositis skin from a single patient are shown.

Figure 3. Early Cellular Responses in Dermatomyositis Skin With Brepocitinib

Single-cell RNA sequencing demonstrated reduced proportions of T cells **(A)** and B cells **(B)** in non-lesional and lesional dermatomyositis (DM) skin by week 4 of brepocitinib treatment. Of the profiled lymphocyte **(C)** and myeloid cell populations **(D)**, CD4+ central memory T cells, CD4+ regulatory T cells, CD8+ effector memory T cells, NK cells, and cDC2B cells were enriched in lesional DM skin at baseline and were substantially reduced following treatment with brepocitinib. Notably, differential gene expression analysis identified keratinocytes as the most transcriptionally responsive cell type, with basal and cycling keratinocytes

demonstrating marked downregulation of IFN-stimulated genes and reductions in type I and II IFN signaling **(E)**.



Abbreviations: W0 = week 0; W4 = week 4; Tcm = central memory T cell; Treg = regulatory T cell; Tex = exhausted T cell; Tem = effector memory T cell; NK = natural killer cell; ILC = innate lymphoid cell; cDC1 = conventional dendritic cell 1; cDC2A = conventional dendritic cell 2A;

cDC2B = conventional dendritic cell 2B; LAM = Langerhans cell; PVM = perivascular macrophage; DM = dermatomyositis

Transcriptional Profiling Reveals JAK/STAT-Dependent Inflammation in Dermatomyositis and Adaptive Immune Bias in Lupus

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Dermatomyositis (DM) and cutaneous lupus erythematosus (CLE) share histopathologically identical features. However, they differ in clinical presentation and response to therapy, notably to JAK inhibitors, which show reliable efficacy in DM but inconsistent benefit in CLE. To define the molecular programs underlying this divergence, we performed gene expression profiling and Ingenuity Pathway Analysis on skin biopsies from patients with CLE, DM, and healthy controls. DM gene expression changes had many similarities to those observed in CLE: 89/143 differentially expressed genes (DEG), 5/16 canonical pathways, and 64/88 upstream regulators were shared. Compared to healthy skin (HS), both diseases demonstrated strong type I interferon activation but differed in the immune profiles that accompanied this response. DM lesions showed prominent upregulation of nucleic acid sensing genes (*DDX58, TRIM14*) and JAK/STAT pathway components (*STAT1*, *STAT2*, *TYK2*). Activated upstream regulators unique to DM included type I interferon-related molecules (IFNAR2, JAK1, and IRF1). These findings support a model of JAK/STAT-dependent, type I IFN-driven inflammation in DM, mediated in part through cytosolic DNA sensing via the STING1 axis. In contrast, CLE lesions exhibited unique expression of genes related to B-cells (*CD22*, *BLNK*), T-cells (*CD3E*, *ZAP70*, *ITK*), and lymphocyte trafficking (*S1PR4*, *ITGB7*), with predicted activation of T cells (TCR, CD28) and innate pathways independent of JAK signaling (C5, CD14). Our findings suggest that CLE involves a broader adaptive immune program with additional inflammatory pathways beyond JAK/STAT, whereas inflammation in DM is more tightly coupled to JAK/STAT pathway activation. Based on our studies, an immunohistochemistry panel of PSMB8, TRIM14, and CTSC may help distinguish DM from CLE, while CD22, S1PR4, and TIGIT may aid in recognizing CLE-specific immune features. Our findings offer a path toward improved diagnostic resolution in routine pathology and support the use of JAK inhibitors in DM.

Abstract category: Dermatomyositis

Teaching point: Dermatomyositis shows a JAK/STAT-dominant inflammatory profile, whereas CLE has a broader adaptive and inflammatory immune profile, which may explain their differential responses to JAK inhibitors.

LONGITUDINAL CARDIOVASCULAR OUTCOMES IN DERMATOMYOSITIS: A PROPENSITY-MATCHED TRINETX COHORT STUDY

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Dermatomyositis (DM) may confer elevated cardiovascular (CV) risk, but long-term event rates and biomarker trajectories remain poorly defined. Using the TriNetX Global Network (2018–2023), we identified adults with ≥2 DM codes (ICD-10 M33.1/M33.9) and constructed a 1:1 propensity-matched cohort to adults seen for routine general medical examination without DM. Matching used age, sex, race/ethnicity, hypertension, hyperlipidemia, and type 2 diabetes, yielding 11,596 patients per group. Outcomes included myocardial infarction (MI), ischemic stroke, pulmonary embolism (PE), venous thromboembolism (VTE), and atrial fibrillation (AF). Biomarkers included first elevated troponin (≥0.04 ng/mL) or BNP (≥100 pg/mL) at 6 months, 3 years, and 5 years. At 5 years, DM had higher absolute risks than controls—MI 3.6% vs 2.1% (risk difference [RD] 1.5%, risk ratio [RR] 1.74), PE 3.1% vs 1.3% (RD 1.8%, RR 2.37), VTE 3.8% vs 1.8% (RD 2.0%, RR 2.16), stroke 3.0% vs 2.5% (RD 0.5%, RR 1.18), and AF 7.5% vs 5.5% (RD 2.0%, RR 1.36). Troponin (2.9% vs 0.9%) and BNP (4.5% vs 3.1%) elevations were also more frequent in DM, suggesting persistent myocardial injury and stress. In a seborrheic keratosis negative-control cohort (n=624,953 matched pairs; post-match age SMD 0.004), CV outcomes were neutral, supporting a true DM-specific signal. Limitations include reliance on coded data, nonstandardized labs, and absence of treatment exposure or follow-up duration data, precluding assessment of whether therapies such as IVIG or JAK inhibitors mitigate or contribute to CV risk. These findings support incorporating CV risk assessment and aggressive management into DM care, and motivate prospective studies capturing disease activity, treatment exposures, and standardized CV outcomes.

Teaching Point: Dermatomyositis is associated with sustained elevations in thrombotic and arrhythmic events and myocardial stress biomarkers, underscoring the need for routine cardiovascular risk assessment and prevention in DM care.

Category: Dermatomyositis

ASSESSING CANCER RISK BEYOND THE HIGH-RISK WINDOW IN DERMATOMYOSITIS PATIENTS

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Dermatomyositis (DM) is an autoimmune disease marked by skin involvement and muscle weakness, with an elevated risk of developing cancer within five years of DM diagnosis. The mechanism between the immune dysregulation and cancer formation remains unclear. Our study investigates cancer risks for patients with DM who do not develop malignancy within the five-year high-risk window, compared to matched population. We conducted a retrospective analysis using TriNetX, a de-identified electronic medical record database. The DM group was defined as patients with no prior history of malignancy and those that remained cancer-free up to five years following DM diagnosis. A comparator group was matched for age, sex, race, immunosuppressant use, and nicotine dependence (N=13,915). We evaluated cancer risks post high-risk window for the cancers commonly associated with DM including breast, lung, pancreatic, and gastric. we identified 93 cases of breast cancer, 71 cases for lung cancer, 27 for pancreatic cancer, and 18 gastric cancers in the DM cohort. Corresponding counts in the control cohort were 201, 148, 35, and 19, respectively. Compared with matched controls, the DM cohort had a decreased risk of malignancy of breast (N= RR=0.456, $p< 0.0001$), and lung (RR=0.478, $p< 0.0001$) while having normalized risk of pancreatic (RR= 0.771, $p< 0.3073$) and gastric cancers (RR= 0.948, p= 0.8716). Our findings indicate that for DM patients who remain cancer-free for five years post-diagnosis, the risk for several malignancies appears decreased in comparison to age matched peers without DM. We hypothesize that DM patients who escape malignancy during the high-risk period have an innately enhanced anti-tumor immune response. The genetic factors influencing the anti-cancer response in these patients requires further investigation which might elucidate the interplay between autoimmunity and cancer biology in DM.

Teaching Point: Dermatomyositis may be protective against cancer after the initial high-risk window (>5 years after diagnosis)

Category: Dermatomyositis

VENOUS THROMBOEMBOLIC RISK OF COMBINATION THERAPY WITH IVIG AND JANUS KINASE INHIBITORS IN DERMATOMYOSITIS: A RETROSPECTIVE STUDY

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Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by proximal muscle weakness and classic cutaneous manifestations. Although traditional therapies include systemic corticosteroids, methotrexate, and mycophenolate mofetil, many patients require additional therapies including intravenous immunoglobulin (IVIG) and Janus kinase (JAK) inhibitors. While the efficacy of IVIG and JAK inhibitors have been demonstrated in refractory DM, both therapies have been independently associated with an increased risk of venous thromboembolism (VTE) and both carry a boxed warning for VTE on their labels. Data is lacking on the use of IVIG and JAK inhibitors concurrently in DM. We performed a retrospective review of patients with DM treated with combination IVIG and JAK inhibitors at Mass General Brigham between 1/1/2012 and 5/15/2025. Twenty-one patients were identified. Most patients had previously failed at least two corticosteroid-sparing agents. Five of 21 (23.8%) of patients were male. The median duration of combination therapy was 12 months although notably several patients were treated up to 79 months at the time of study. Tofacitinib was the most commonly prescribed JAK inhibitor in 13 patients (61.9%), followed by upadacitinib in 6 patients (28.6%) and ruxolitinib in 2 patients (9.5%). In all patients, JAK inhibition was added to the patient's regimen given partial but incomplete efficacy of IVIG. Importantly, combination therapy was well-tolerated, with no VTE attributed to therapy. While larger prospective studies are needed, our study provides early evidence that combination therapy with IVIG and JAK inhibitors is a reasonable approach in patients with refractory DM.

Category: Dermatomyositis

COMPARISONS BETWEEN ADULT AND JUVENILE DM PERIPHERAL BLOOD MONONUCLEAR CELL GENE EXPRESSION MAY IDENTIFY CANCER-ASSOCIATED GENE SIGNATURES

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Dermatomyositis (DM) is an uncommon idiopathic autoimmune disease that causes muscle and skin inflammation. Adults with DM are at increased risk for malignancy, most notably of lung, bladder, colon, ovary, and breast. Cancer is often occult, and better biomarkers are needed to optimize screening. Juvenile DM (JDM) is a similarly uncommon disease that presents with comparable muscle and skin manifestations, yet, unlike adult DM (ADM), there is no increased cancer risk. From a molecular perspective, the primary commonality between these DM subtypes is a striking type I interferon signature; otherwise, these diseases are relatively unique. We utilized both in-house generated and publicly available peripheral blood mononuclear cell (PBMC) RNA sequencing data from ADM (n=10), JDM (n=32), and control (adult: n=5; juvenile: n=12) donors, to identify genes that 1) distinguish ADM from JDM; and 2) are indicative of cancer risk in ADM patients. Differential gene expression (DGE) between both ADM:adult healthy controls and JDM:juvenile healthy controls were utilized as input for Reactome pathway analysis (ReactomePA). Interestingly, terms related to DDX58/IFIH1 (RIGI/MDA5) signaling were significant only in ADM, while interleukin signaling was significant only in JDM ($p \leq 0.05$; **Fig. 1**). As expected, type I interferon signaling was significant in both DM subgroups ($p \leq 0.05$; **Fig. 1A**). DGE performed directly between these DM subgroups highlighted 21 and 14 significantly differentially expressed genes in ADM and JDM, respectively ($p \leq 0.05$; **Fig. 2A-B**). The top differentially expressed gene in the JDM group was *IL6*; a finding that supports the implication of overactive IL6/STAT3 signaling in the disease pathogenesis. In the ADM group, the genes *FAM83A* and *GUC2YC* were of particular interest as literature has indicated a link between their overexpression and lung and colorectal cancer, respectively (**Fig. 2C**), wherein gene expression is increased in the local tumor microenvironment, as well as in circulating tumor cells (CTCs) and in circulating mRNA. Continued study will allow us to highlight distinctions between ADM and JDM and may lead to more granular discovery of malignancy-associated biomarkers in ADM.

Category: Dermatomyositis

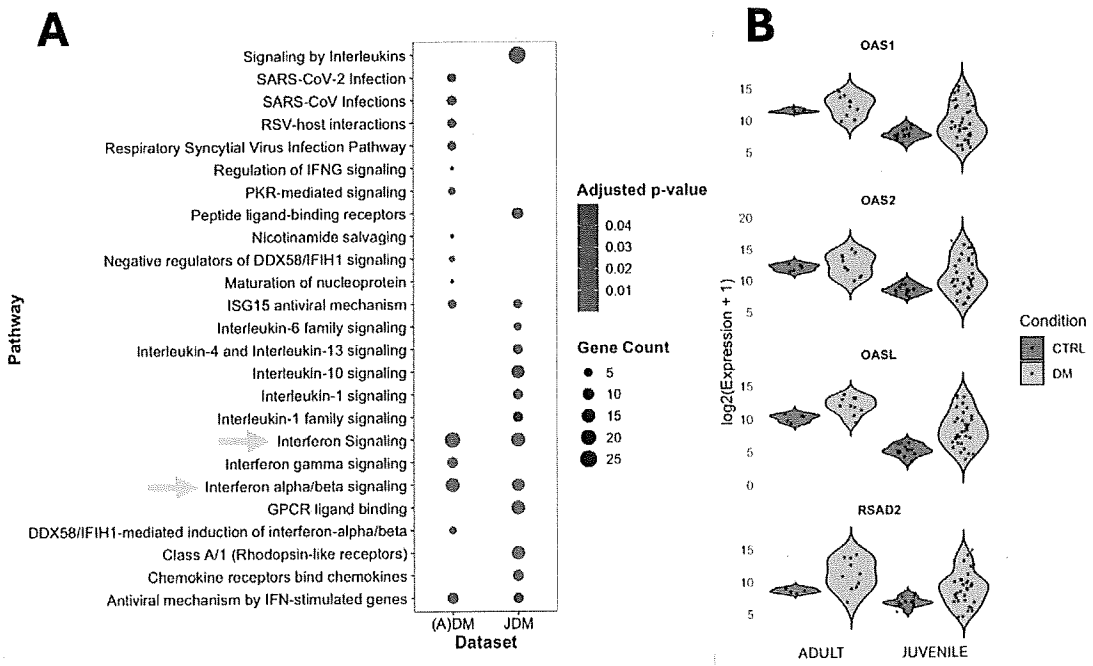

Figure 1. The type 1 interferon signature is the primary molecular commonality between ADM and JDM PBMCs. A. Faceted dot plot of significant ReactomePA pathways. Arrows indicate similarly significant interferon signaling pathways. **B.** Violin plots showing log-normalized expression counts for various interferon-stimulated genes: *OAS1*, *OAS2*, *OASL*, and *RSAD2*.

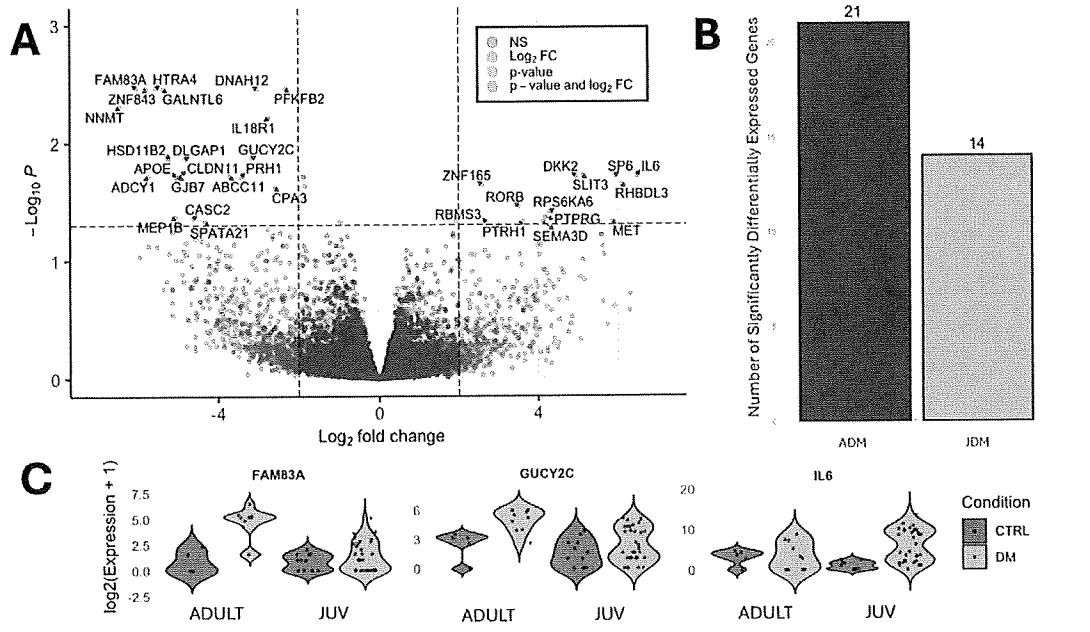

Figure 2. Differentially expressed genes in ADM and JDM PBMCs. A. Volcano plot of differential gene expression (DGE) between ADM and JDM PBMCs. Negative $\log_2$FC indicates stronger effect in ADM, positive $\log_2$FC indicates stronger effect in JDM. **B.** Total number of significantly differentially expressed genes in ADM and JDM ($\log_2\text{FC} \geq 2$ and padj ≤ 0.05). **C.** Violin plot showing log-normalized expression counts of all samples for the genes *FAM83A*, *GUCY2C*, and *IL6*.

EVALUATION OF GLP1-RECEPTOR AGONIST USE IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES: A RETROSPECTIVE ANALYSIS

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Glucagon-like peptide-1 receptor agonists (GLP1-RAs), widely used for diabetes and obesity, may potentially alleviate muscle inflammation[1,2], but data in idiopathic inflammatory myopathies (IIMs) are limited. This retrospective analysis evaluated their safety by assessing disease flare outcomes in a myositis population. Fifty-two patients from a dual-institution database with a confirmed myositis diagnosis who had used a GLP1-RA were analyzed. The patient population was predominantly female (75%), with a mean age of 57.7 years at GLP1-RA initiation. The most common myositis variants were dermatomyositis (48.1%), anti-synthetase syndrome (23.1%), and polymyositis (13.5%). We compared the presence of active skin, muscle, or lung involvement before (average: 2.87 months) and after (average: 25.4 months) GLP1-RA initiation. Before GLP1-RA initiation, active involvement was present in the skin of 13 patients (25.0%), muscles of 30 (57.7%), and lungs of 14 (26.9%). After initiation, these decreased to 9 (17.3%), 23 (44.2%), and 13 (25.0%) patients, respectively. McNemar's test demonstrated no statistically significant change for skin ($\chi^2 = 0.90$, $p = 0.34$), muscle ($\chi^2 = 1.56$, $p = 0.21$), or lung ($\chi^2 = 0.00$, $p = 1.00$). There was an average change in BMI of -2.95 kg/m^2 and in HbA1c of -0.79% with GLP1-RA use. Thirteen patients (25%) discontinued GLP1-RAs, primarily due to gastrointestinal side effects or insurance issues. Only one case of increased muscle weakness was attributed to GLP1-RA use. Our findings suggest that the addition of GLP1-RAs to standard treatments is associated with a trend toward improvement in skin, muscle, and lung disease in patients with IIM. They were well-tolerated and did not appear to increase disease activity or flares. Limitations include a small sample size and frequent issues of adherence with GLP1-RAs. Further multi-institutional, prospective studies are needed to investigate the impact of GLP1-RAs on disease activity in this population.

Teaching Point: GLP1-RAs may be associated with improvements in skin, muscle, and lung disease in patients with IIM and appear to be well-tolerated, without exacerbation of disease activity or flares.

Abstract category: Dermatomyositis

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SPATIAL TRANSCRIPTOMIC ANALYSIS OF CALCINOSIS CUTIS IN DERMATOMYOSITIS UNCOVERS DISEASE-ASSOCIATED PATHWAYS INVOLVING IL-6, TISSUE REMODELING, AND OSTEOPONTIN

Cassie Parks[1,3], Xinyi Sun[2], Marissa Mojena[1], York Wang[3], Lisa Christopher-Stine[3], Jemima Albayda[3], Joel Sunshine[1], Shira Ziegler[2], Christopher Mecoli[3]

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Calcinosis cutis is a highly morbid condition affecting up to 20% of adults with dermatomyositis (DM), with current management largely limited to surgical excision despite the risk of recurrence. To better understand the underlying pathogenesis of this devastating complication, we performed Visium 10X spatial transcriptomics on four active calcinosis samples from adult DM patients with anti-MDA5, PM-Scl, Ro, and Jo-1 autoantibodies meeting 2017 ACR/EULAR criteria and three healthy adult skin biopsies. Data were analyzed using Seurat v5.0.0 and DESeq2 v. 1.44.0. We found that DM calcinosis samples have a distinct inflammatory RNA signature compared to healthy controls, characterized by increased *IL6*, macrophage markers, *CCL3*, immunoglobulins, and extracellular matrix remodeling enzymes. Osteopontin (*SPP1*), which promotes resorption of pathologic calcification, was also robustly elevated in lesional samples. We next categorized spatial transcriptomic spots based on their distance from the calcinotic lesion; *SPP1* expression was found to be highest near the calcinosis, while professional antigen-presenting cell (APC) markers, immunoglobulin genes, and complement genes were enriched 100–150 μm from the lesion. Additionally, *MMPs 2,9,* and *13* were highest at 100-150 μm from the lesion, while *TIMP1* (an MMP regulator) was highest 50-100 μm from the lesion. To functionally evaluate IL-6's role in pathological calcification, we cultured normal human dermal fibroblasts in a pro-calcific media ± IL-6. We found that IL-6 increased the RNA expression and enzymatic activity of tissue non-specific alkaline phosphatase (TNAP), a known master regulator of extracellular ATP metabolism that is integral to physiologic and pathologic calcification. This study provides the first detailed spatial molecular map of calcinosis in adult DM, which implicates macrophage-driven inflammation, possible local antibody production, and osteopontin, and provides evidence for a mechanistic role for IL-6 and extracellular ATP metabolism in calcinosis cutis.

Category: Dermatomyositis

AN INITIAL GENOME-WIDE ASSOCIATION ANALYSIS OF DERMATOMYOSITIS IN ~2.1 MILLION INDIVIDUALS FROM THE DERMATOMYOSITIS GENETICS CONSORTIUM (DMGC)

Lujia Song[1], on behalf of Dermatomyositis Genetics Consortium (DMGC)

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Dermatomyositis (DM) is a severe autoimmune disease manifesting as muscle weakness and distinctive skin rashes. Intriguingly, DM patients have a very high probability of progressing to cancer within 5 years of DM diagnosis. Like many other autoimmune diseases, DM carries heritable risk factors. Yet the genetic basis for DM is understudied, and little progress has been made in improving our understanding of DM genetics. Given the low prevalence of DM, international collaboration would be critical for aggregating sample sizes and improving the power for genetic studies. To this end, we establish the Dermatomyositis Genetics Consortium (DMGC) and conducted an initial genome-wide association study (GWAS) meta-analyses integrating data from 11 biobanks around the globe (Figure 1). Patients with DM were identified with International Classification of Disease (ICD)-9 and ICD-10 codes of DM, dermatopolymyositis, and juvenile dermatomyositis (JDM). We have thus far assembled dataset with 3084 cases and 2,095,955 controls, making our study magnitudes larger than existing DM GWAS. An initial analysis of DMGC data uncovered 18 loci, harboring 2,149 SNPs with $p < 5 \times 10^{-8}$ (Figure 2). Within the MHC region, we also performed HLA imputation and identified top HLA alleles associated with either protection against or susceptibility to DM, including HLA-B*08:01:01 (logOR = -0.42, $p = 8.26 \times 10^{-21}$), HLA-DRB1*12:02:01 (logOR = 0.12, $p = 8.72 \times 10^{-12}$), and HLA-DRB3*03:01:03 (logOR = 0.13, $p = 3.67 \times 10^{-14}$). Outside the MHC region, the study also identifies 11 loci, including PHLPP1 and CUL5, which is involved in regulating the fate and function of CD4+ T cells and T_{reg}. In conclusion, this large-scale study presents the comprehensive translational genomics of DM, casting light on the etiology of DM and offering potential targets for future disease-modifying therapies. Subsequent analyses and collaborations may further illuminate the relationships between DM, cancer and inflammatory lung disease.

Abstract Category: Dermatomyositis

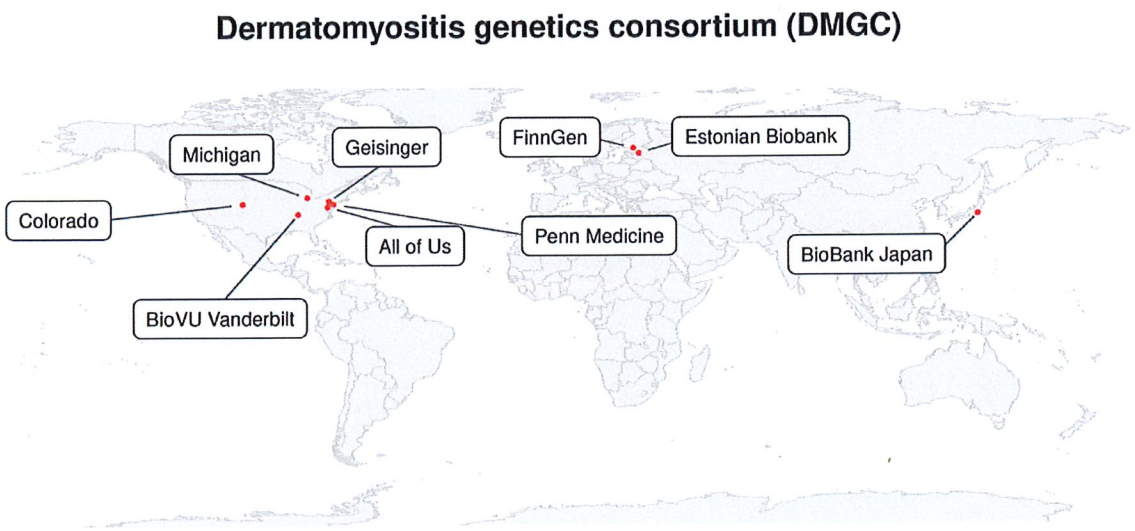

Figure 1. Geographic distribution of participating cohorts in the Dermatomyositis Genetics Consortium (DMGC).

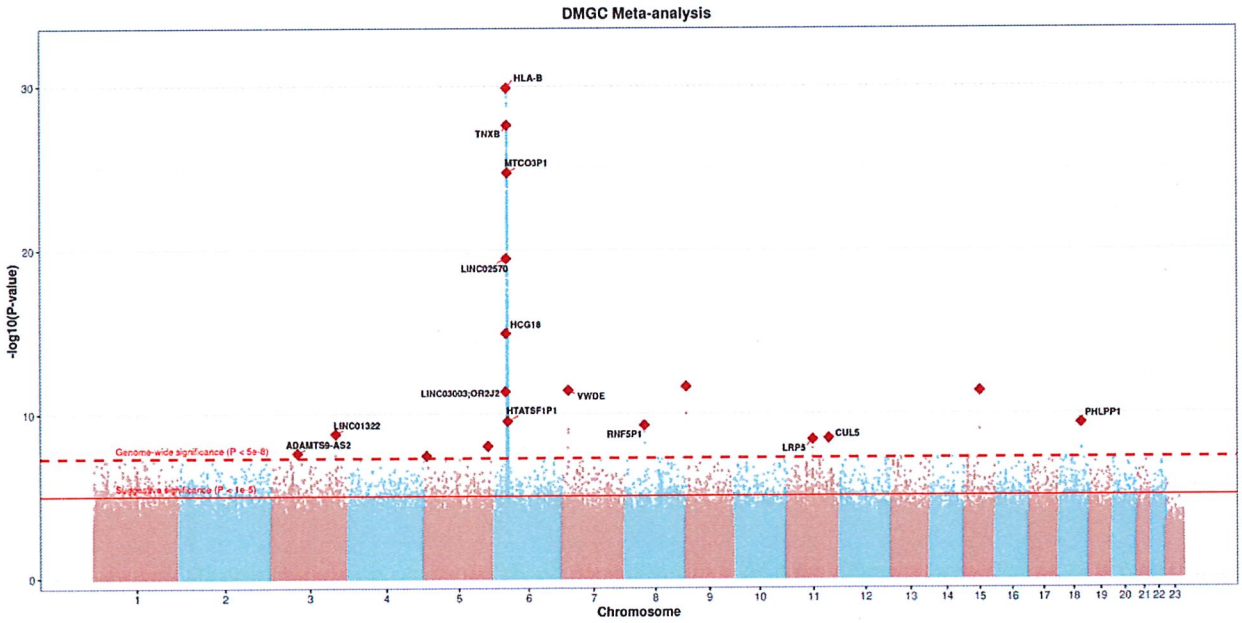

Figure 2. Manhattan plot of the DMGC genome-wide association meta-analysis.

The Effects of Hydroxychloroquine and Quinacrine on cGAS–STING, RIG-I/MDA5, TLR4, and NLRP3 Inflammasome Activation in Dermatomyositis Cell Types

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Dermatomyositis (DM) is an autoimmune disease of the skin, muscles, and lungs. Treatment options for DM include antimalarials like Hydroxychloroquine (HCQ) and Quinacrine (QC). We aimed to compare the effects of HCQ, QC, and their combination (HCQ-QC) on DM PBMCs stimulated with dsDNA transfection for cGAS-STING pathway, dsRNA transfection for RIG1/MDA5 pathway, LPS for TLR4 pathway, and LPS+ATP for NLRP3 inflammasome pathway. Cells were stained and analyzed with Flow Cytometry. QC monotherapy and HCQ+QC, but not HCQ monotherapy, significantly inhibited the levels of pSTING activation in myeloid dendritic cells (mDCs), plasmacytoid dendritic cells (pDCs), monocyte-derived dendritic cells (MoDCs), and Macrophages following dsRNA-RIG1/MDA5 and LPS-TLR4 stimulation, and in pDCs, MoDCs. and macrophages following dsDNA-cGAS stimulation. Interestingly, only the HCQ-QC combination could significantly inhibit LPS-TLR4-mediated IFNβ, and to a lesser extent (along with QC monotherapy), dsDNA-cGAS and dsRNA-RIG1/MDA5-mediated IFNβ release in pDCs— posing a possible mechanism of HCQ/QC synergism in DM. Notably, pDCs had the highest percent increase in IFNβ and pNF-κB across cell types, following LPS-TLR4 stimulation. Neither HCQ nor QC nor HCQ-QC inhibited pNF-κB activation by any pathway in pDCs. Percent increase in pSTING activation was higher in mDCs than all cell types, following dsDNA stimulation, and was greatly reduced with either HCQ or QC. However, only QC could inhibit pSTING activation by LPS, dsRNA, and LPS+ATP — suggesting a mechanism of pSTING inhibition distinct from HCQ. Unlike QC, HCQ seems least effective in reducing LPS-TLR4 and dsRNA-RIG1/MDA5 inflammation, whereas both drugs reduced NLRP3-inflammasome activity. Lastly, IFNβ reduction with HCQ in pDCs was higher in classic DM than amyopathic DM. In summary, Quinacrine demonstrated broad-spectrum inhibitory effects, intercepting various type I IFN signaling pathways across multiple cell types, comparable to HCQ-QC combination. Given its efficacy and lower risk of retinopathy, Quinacrine could be considered as monotherapy in DM, while HCQ-QC combination can provide additional benefit in certain contexts.

Abstract Category: Dermatomyositis

OBINUTUZUMAB AS AN EMERGING THERAPY IN CONNECTIVE TISSUE DISEASE: A RETROSPECTIVE COHORT STUDY

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Word count: 300/300

Obinutuzumab is a glycoengineered type II anti-CD20 monoclonal antibody with enhanced antibody-dependent cellular cytotoxicity and direct B-cell killing compared to rituximab. Initially approved for follicular lymphoma and chronic lymphocytic leukemia, it is increasingly used in rheumatology, with a phase III trial in systemic lupus erythematosus demonstrating a complete renal response benefit and prompting Food and Drug Administration (FDA) acceptance of a supplemental biologics license application for lupus nephritis. Obinutuzumab's potential in the management of other connective tissue diseases (CTD) remains underexplored. In this retrospective review, a search of the Research Patient Data Registry at Mass General Brigham identified 14 patients with dermatomyositis (DM) or systemic sclerosis (SSc) and ongoing treatment with obinutuzumab for various indications. Seven patients had active DM and/or SSc; after excluding patients who had not seen a dermatologist or rheumatologist prior to or after the initiation of obinutuzumab, five patients (three DM, one SSc, one SSc/DM overlap) were included for analysis. For patients with DM, symptoms at the time of first obinutuzumab dose included weakness (4/4), rash (3/4), myalgia (2/4), and dysphagia (2/4). For the two patients with SSc, both had sclerosis and Raynaud's. Indications for obinutuzumab included lymphoma (2/5), leukemia (1/5), cryoglobulinemia-associated glomerulonephritis (1/5), and refractory DM (1/5). All obinutuzumab doses were 900-1000mg IV, with total treatment cycle numbers depending on indication. Four of the five patients experienced improvement in their DM and SSc, two of whom experienced near to complete resolution of their symptoms. Treatment was well tolerated, with only one case of CMV-associated colitis and esophageal ulcerations. Ongoing concomitant therapies included systemic steroids (3/5), hydroxychloroquine (2/5), intravenous immunoglobulin (2/5), methotrexate (1/5), and mycophenolic acid (1/5). An additional analysis of a larger cohort of cutaneous lupus erythematosus (CLE) patients receiving obinutuzumab is ongoing. These findings suggest obinutuzumab may be a promising therapeutic option for CTD.

Category: Dermatomyositis, sclerotic skin disease (e.g. morphea, systemic sclerosis, etc.)

Session V:

MISCELLANEOUS & CLINICAL CASES OF THE YEAR

CHARACTERIZING PERNIO: CLINICAL FEATURES, WORKUP, AND OUTCOMES IN A LARGE U.S. COHORT

Authors: Zasca-aisha Ristianto BS[1,2]; Kaitlin Martins, MS[1]; Sophia Manduca, BS[1,2]; Camille Scandurro, BS[2]; Alisa N. Femia, MD[1]

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Pernio is a rare inflammatory condition affecting acral skin, often triggered by cold, damp environments. Its pathogenesis and clinical patterns remain poorly characterized. This retrospective chart review of 463 patients at a large academic center (median age: 41 years; 70.2% female) explores presentation, comorbidities, and treatment outcomes of pernio. Most cases (80%) occurred during cold, wet months. Pernio affected only the feet in 49%, only the hands in 23.3%, and both in the remainder. Discoloration (80.1%) and tenderness (76.4%) were the most common symptoms; 84.4% had multiple symptoms. Among 325 patients evaluated for autoimmune, hematologic, or viral etiologies, 73% had idiopathic pernio. Immune-mediated disease was found in 16.4%, infection in 9.3%, with 41 of 43 infectious cases attributed to COVID-19, and 1% had drug-induced pernio. Of 241 patients tested for ANA, 107 were positive, although only 76 had connective tissue disease. Among patients with follow-up (mean: 13 months), 190 achieved full resolution. Of those, 65.8% responded to conservative measures alone (e.g., dressing warmly, avoiding cold, quitting smoking). Pharmacologic treatment was required in the remainder, most commonly with corticosteroids, topical tacrolimus, calcium channel blockers, and/or hydroxychloroquine. The average number of treatments needed to reach complete response was 1.27 ($p = 1.33e-10$). This largest single-center analysis of pernio confirms seasonal prevalence, female predominance, and favorable outcomes with non-pharmacologic treatment. While most cases were idiopathic, a subset was associated with autoimmune or post-viral syndromes, particularly COVID-19. The distinct seasonal pattern observed in COVID-19-associated cases, nearly half of which presented during warmer months, suggests a unique clinical subset consistent with the previously documented "COVID toes" phenotype. These cases were also more likely to involve the feet alone, further supporting a distinct presentation compared to idiopathic pernio. This large cohort offers valuable insight into patterns of pernio presentation, comorbidity, and management. Further research should identify predictors of systemic disease to guide individualized testing and management.

Category: Miscellaneous rheumatic skin disease

JAK INHIBITORS FOR TREATMENT OF SAPHO SYNDROME: A SYSTEMATIC REVIEW OF 72 CASES

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Word count: 300

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Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome usually involves bone, joints, and skin. In the lack of known pathogenesis and clinical trials, there is no standard treatment for SAPHO patients. Janus kinase inhibitors (JAK-I) are a group of small-molecule drugs with a

wide range of effects on inflammatory and autoimmune pathways. A systematic search was conducted using MeSH terms/keywords related to JAK-I and SAPHO Syndrome through PubMed/Medline, Scopus, Web of Science, and Embase until September 8th, 2024. The inclusion criteria were a diagnosed SAPHO syndrome patient who received at least one JAK-I. We excluded reviews and animal studies. Out of 287 initially researched articles, we included 34 articles. These 34 articles involved 72 patients with a mean age of 39.36 years and a female (78%) predominance. All patients had bone or joint involvement, most commonly in the anterior chest wall (62.5%) and vertebrae (45.8%). Skin involvement was seen in 64 patients, with 52 (72.2%) presenting palmoplantar pustulosis (PPP). Nearly all (97.2%) had prior treatments, with nonsteroidal anti-inflammatory drugs (NSAIDs), corticosteroids, and methotrexate (MTX) being the most common. Tofacitinib was the most used JAK-I (54/72, 75%), followed by baricitinib (15/72, 20.8%) and upadacitinib (3/72, 4.2%). 18 (25%) patients received concomitant treatment with another medication; of these, 7 received NSAIDs and 6 received MTX. Different variants were used as responses to treatment in these studies, which were as follows: alleviating or clearing symptoms, significant changes in imaging, a decrease in laboratory inflammatory markers, or a Visual Analogue Scale (VAS). Almost all patients (71/72, 98.6%) showed a good to complete response. Adverse effects occurred in ten patients (13.9%), all on tofacitinib; only one led to drug discontinuation. Lab abnormalities were seen in 6.9% without clinical symptoms. In conclusion, JAK-I seems to be a promising treatment for SAPHO syndrome with manageable adverse effects.

Abstract category: Miscellaneous rheumatic skin disease

POINT-OF-CARE RISK FACTORS FOR SYSTEMIC DISEASE IN PATIENTS PRESENTING WITH SMALL VESSEL VASCULITIS OF THE SKIN

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Small vessel vasculitis (SVV) presenting in the skin may be skin-limited or a manifestation of systemic disease. However, no evidence-based protocols exist to identify patients at risk for systemic involvement. This study aimed to identify clinical features at initial evaluation that predict systemic disease in patients with cutaneous SVV, to support early risk stratification and management. This multi-center case-control study included 430 adults with biopsy-confirmed cutaneous SVV, identified from pathology databases at Massachusetts General Hospital, Brigham and Women's Hospital, and the Hospital of the University of Pennsylvania. Aligned with the Chapel Hill consensus criteria, cases were defined as systemic if SVV was associated with a systemic vasculitis diagnosis, connective tissue disease, or caused objective organ dysfunction. Demographic, clinical, and laboratory features at presentation were assessed. Univariate and multivariate logistic regression analyses estimated odds ratios (ORs) and 95% confidence intervals (CIs), with variable selection guided by elastic net regression and 10-fold cross-validation. Among 430 patients (mean [SD] age, 55 [17.7] years; 55.5% male), 87 had systemic disease and 343 had skin-limited SVV. Multivariate analysis identified nausea/vomiting (OR 3.92; 95% CI 1.58–9.77), ulcerating or necrotic lesions (OR 3.26; 95% CI 1.24–8.59), abdominal pain/cramping (OR 2.71; 95% CI 1.19–6.18), and fatigue/malaise/lethargy (OR 2.61; 95% CI 1.28–5.32) as significantly associated with systemic disease. In a subset with urinalysis data (n=388), dipstick hematuria (3+) was associated (OR 2.40; 95% CI 1.23–4.68) and recent antibiotic use was inversely associated (OR 0.54; 95% CI 0.29–0.98). Younger age (OR 0.74 per SD increase; 95% CI 0.58–0.93) was associated in exclusively univariate analysis. In this multi-institutional study of SVV, we identified key risk factors–ascertainable at initial presentation–that may aid in identifying high-risk patients requiring further evaluation and closer monitoring, potentially improving outcomes through earlier detection and management of critical complications.

Category: Vasculitis

ANTI-MDA5-LIKE ULCERATED GOTTRON'S PAPULES IN ANTI-NXP2 DERMATOMYOSITIS

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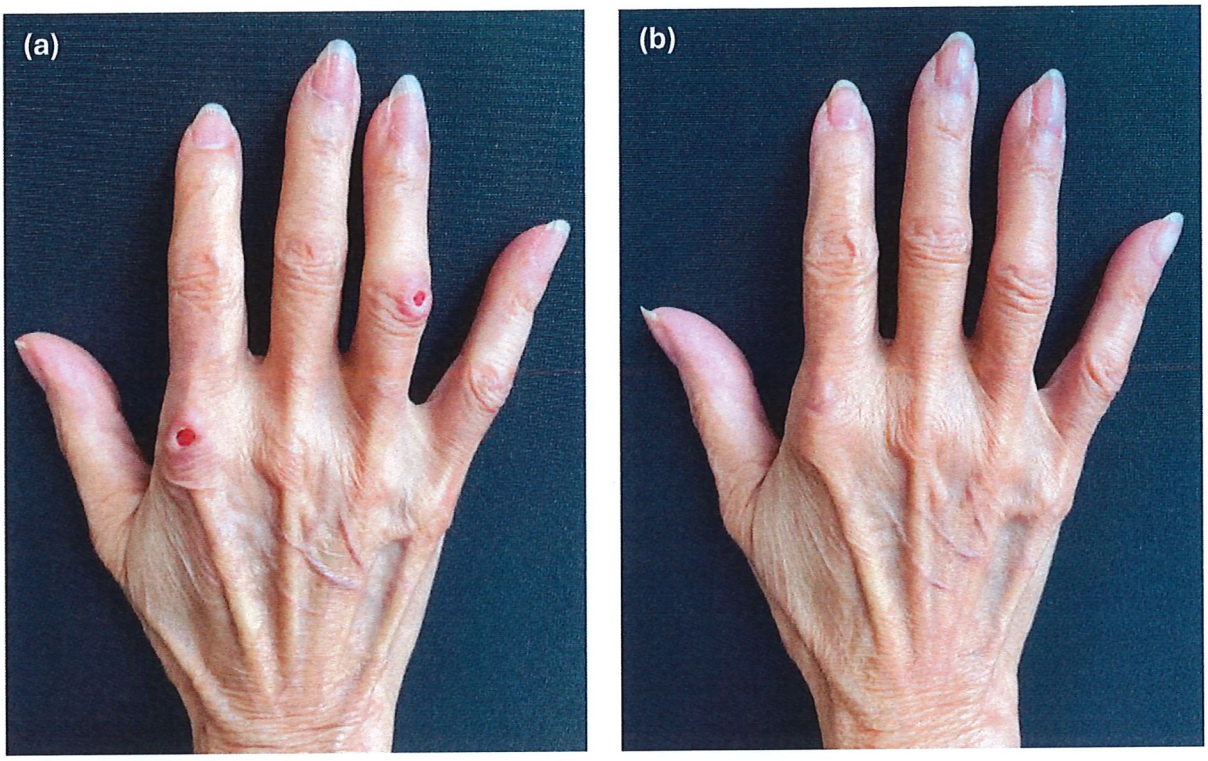
Ulcerated Gottron's papules due to vasculopathy are characteristic of anti-melanoma differentiation-associated gene 5 (MDA5) dermatomyositis (DM).[1] We report the case of a 78-year-old Puerto Rican woman with DM presenting with anti-MDA5-like ulcers overlying the metacarpophalangeal and proximal interphalangeal joints (a), in addition to prominent myositis and otherwise mild skin disease. Her hand x-rays were negative for calcinosis. Although her myositis had improved with prednisone and azathioprine, her ulcers had not. Unexpectedly, she tested positive for the anti-nuclear matrix protein 2 (NXP2) antibody at two laboratories (ARUP and Oklahoma Medical Research Foundation). Although her ulcerated Gottron's papules were atypical of anti-NXP2 DM, her severe myositis and otherwise mild cutaneous symptoms were consistent with this DM subtype.[2,3] Treatment with oral sildenafil 20 mg three times daily for six months led to resolution of the ulcers (b). This response to vascular-directed therapy, along with prior reports of extracutaneous vasculopathy in anti-NXP2 DM,[4] suggests that her ulcerated Gottron's papules were also vasculopathic in etiology, and that such findings may occur beyond anti-MDA5 DM.

Teaching Point: Although classically associated with anti-MDA5 DM, ulcerated Gottron's papules may also occur in anti-NXP2 DM and respond to vascular-directed therapy.

Category: Clinical Case



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SUCCESSFUL TREATMENT OF RECALCITRANT AMYOPATHIC DERMATOMYOSITIS WITH DEUCRAVACITINIB IN ADDITION TO INTRAVENOUS IMMUNOGLOBULIN

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Abstract:

Dermatomyositis (DM) is an autoimmune disease of skin and muscle; amyopathic DM presents with characteristic cutaneous findings without clinical myopathy. Standard care often combines photoprotection, topical corticosteroids or calcineurin inhibitors, antimalarials, classical immunosuppressants, and Janus kinase (JAK) inhibitors, with intravenous immunoglobulin (IVIG) the only U.S. Food and Drug Administration–approved therapy. We report a 60-year-old man with pruritic, photosensitive violaceous facial plaques (heliotrope) and poikilodermatous trunk lesions for 6 months. Two biopsies showed interface dermatitis with superficial periadnexal lymphocytes; myositis panel was anti–Mi-2–positive with normal creatine kinase, and malignancy/pulmonary screenings were unrevealing. Despite strict photoprotection and sequential systemic therapy—prednisone 1 mg/kg daily, hydroxychloroquine 200 mg twice daily, mycophenolate mofetil 1500 mg twice daily (later methotrexate 20 mg weekly), and monthly IVIG 2 g/kg—cutaneous disease remained active and prednisone could not be tapered below 20 mg. Deucravacitinib 6 mg daily, a selective tyrosine kinase 2 (TYK2) inhibitor, was added to ongoing IVIG and topical therapy, leading over 4 months to resolution of periorbital edema and violaceous plaques with symptomatic relief; prednisone was tapered off without relapse, and no adverse events occurred. TYK2 inhibition may benefit DM via modulation of type I interferon–driven signaling while sparing interleukin-15/natural killer cell pathways, supporting a favorable malignancy-related safety profile. This case suggests that deucravacitinib can be an effective adjunct to IVIG in recalcitrant amyopathic DM, enabling corticosteroid discontinuation; controlled studies are warranted to define efficacy, durability, and safety.

Teaching Point: Adding deucravacitinib (a selective TYK2 inhibitor) to IVIG achieved steroid-free remission in recalcitrant amyopathic dermatomyositis.

Abstract Category: Clinical Case

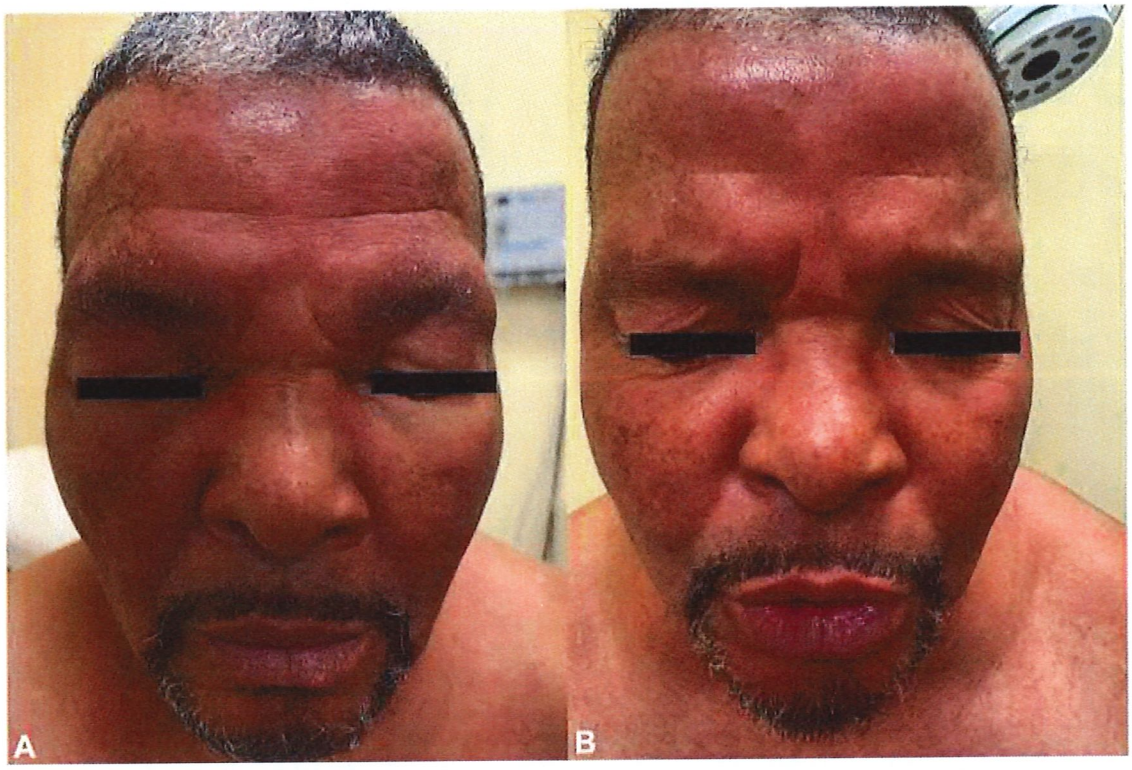

Figure 1: **A,** Facial dermatomyositis presenting with ill-defined violaceous plaque and periorbital edema, consistent with heliotrope sign that (**B**) improved after the addition of 4 months of 6 mg daily deucravacitinib.

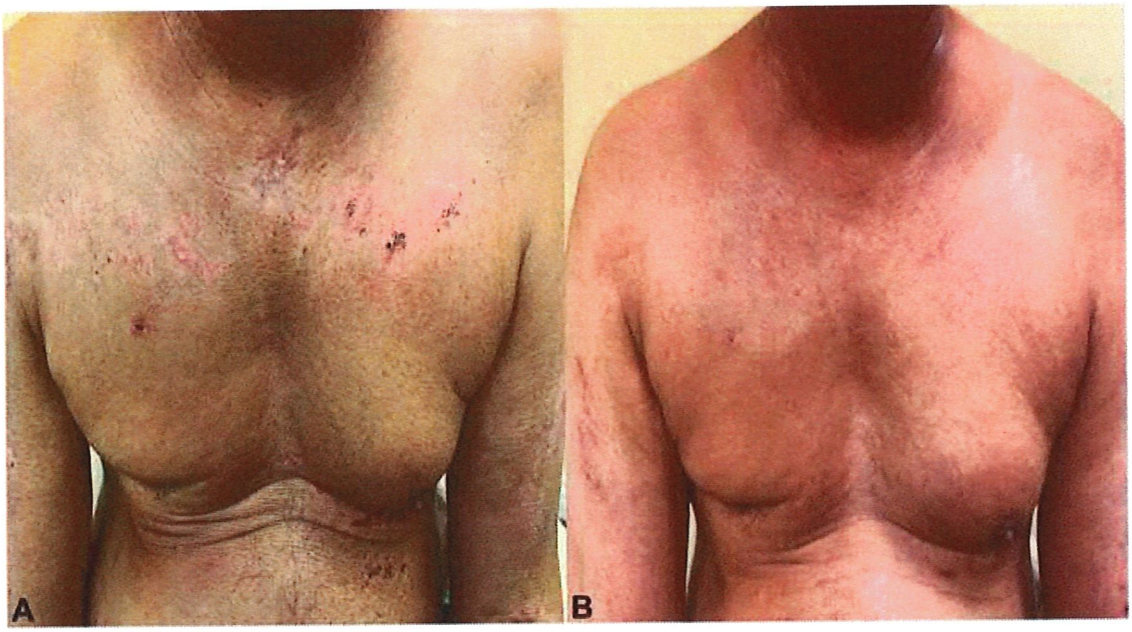

Figure 2: Violaceous poikilodermaotus papules and plaques on the chest (V-sign) that (**B**) improved after the addition of 4 months of 6 mg daily deucravacitinib.

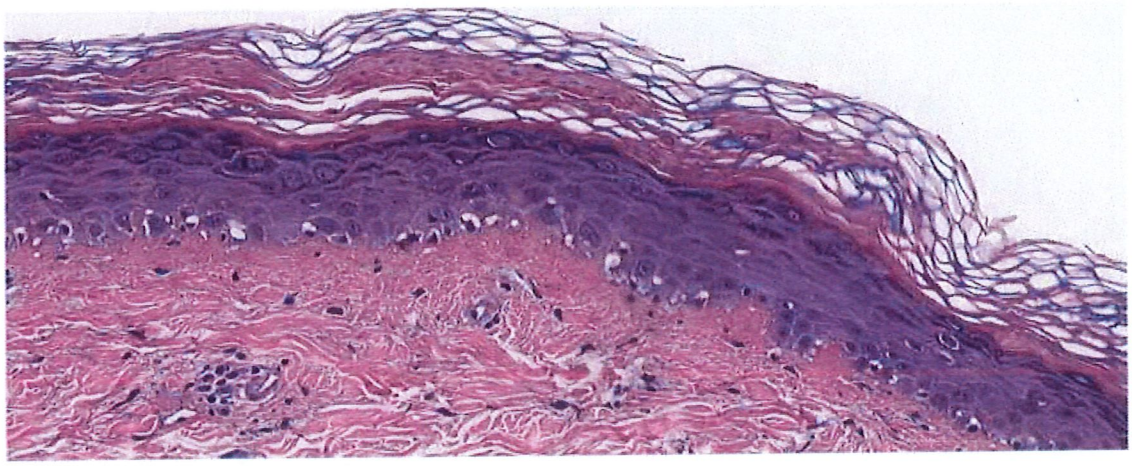

Figure 3: H&E 8×, punch biopsy of the chest lesion showed interface dermatitis and sparse periadnexal infiltrates. *H&E,* Hematoxylin and eosin.

Title: Zilucoplan-induced morphea successfully treated with methotrexate and UVA1 phototherapy

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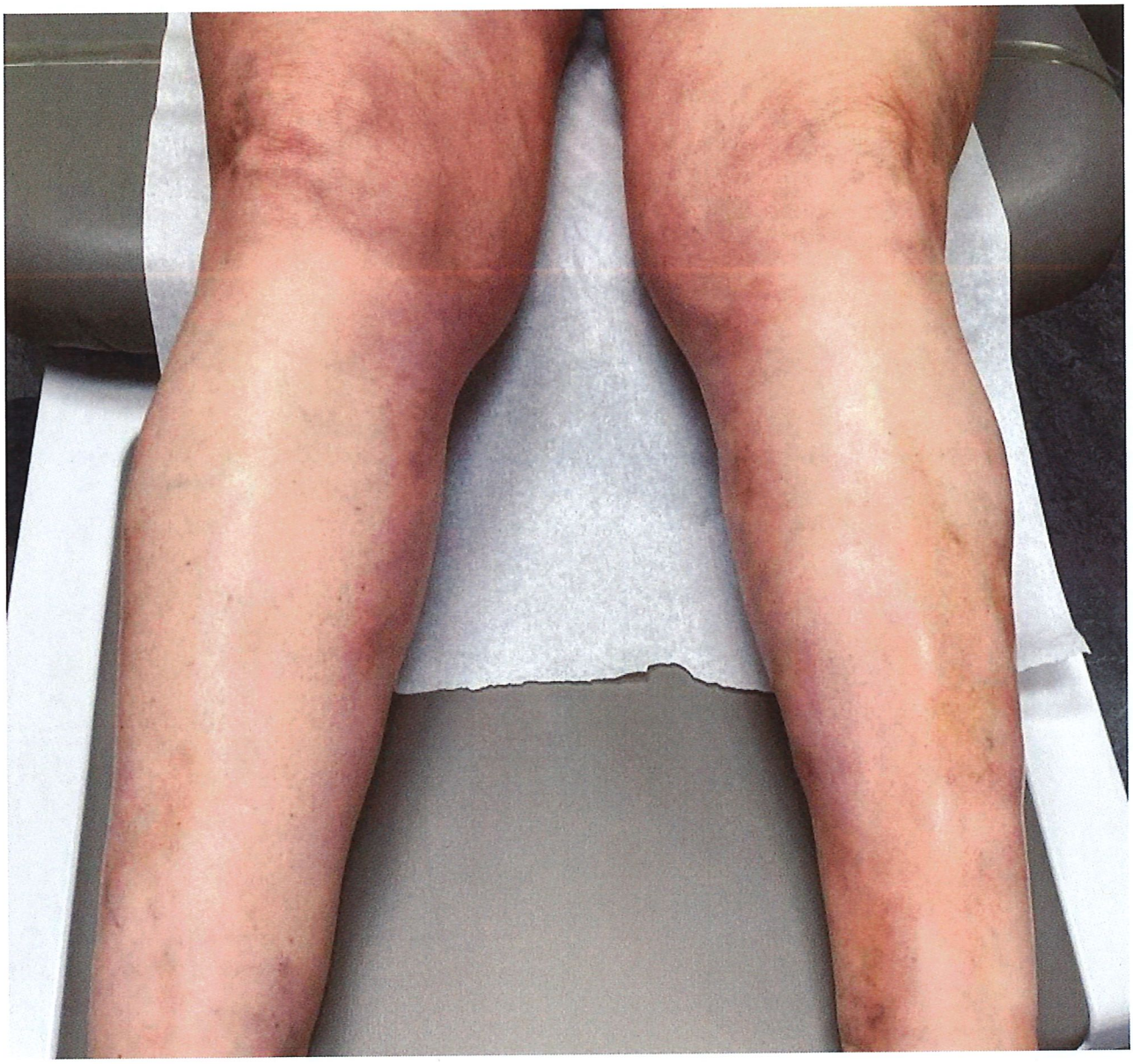
Body: Zilucoplan, a novel subcutaneous complement component 5 (C5) inhibitor, is second line to acetylcholinesterase inhibitors and corticosteroids for myasthenia gravis (MG). While the prescribing information insert noted morphea in 5% in participants, none have been reported in the literature. A woman in her 60s with a history of MG presented after having inadequate response and treatment burden to pyridostigmine (mestinon), prednisone, and eculizumab. She was enrolled in a new clinical trial with zilucoplan. Despite having improvement in her MG symptoms and tapering her prednisone, she reported development of new skin lesions. Skin examination showed large oval violaceous to brown plaques on the bilateral legs, sparing the feet. Punch biopsies confirmed morphea. The patient began topical corticosteroids with mild improvement and zilucoplan was eventually discontinued due to worsening MG and morphea. Within a week of stopping, her morphea improved. Mycophenolate mofetil was started due to her worsening MG and was subsequently tapered and overlapped with methotrexate initiation. Additionally, she was started on light therapy (UVA1) and topical tacrolimus with significant skin improvements after 3 months. Drug-induced morphea has typically been reported with bleomycin and immunotherapy drugs including pembrolizumab and nivolumab. C5 inhibitors work by preventing cleavage of C5 into C5a and C5b. Additionally, the C5a-C5a receptor interaction is integral to fibrosis development. Given this role, we hypothesize that the patient's C5 inhibitor-induced morphea is likely related in this aspect. Furthermore, subcutaneous delivery of zilucoplan may provide a local pathway for fibrotic dysfunction in the skin, compared to the absence of morphea associated with previous intravenous complement inhibitors reported in the literature. **Teaching point**: With increasing use of complement inhibitors for MG, this case highlights an unusual event due to zilucoplan, allowing dermatologists to be aware of its possible cutaneous effects.

Clinical picture:



RECURRENT PULMONARY CAVITARY INFILTRATES AND NEW PAINFUL ULCERATIVE LESION: PYODERMA GANGRNEOSUM PRESENTING WITH PULMONARY MANIFESTATIONS

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Pyoderma gangrenosum (PG) is a rare neutrophilic dermatosis typically presenting as a rapidly progressing painful lesion with violaceous borders most commonly on the lower extremity. Extracutaneous manifestations of PG are rare but preferentially affect the lungs. Pulmonary PG preceding cutaneous manifestations is exceptionally rare, with limited reports in the literature. A 53-year-old woman with a seven-year history of recurrent granulomatous cavitary lung lesions was hospitalized with severe pain due to a rapidly progressive ulcer on the lower extremity (Figure 1). Her pulmonary symptoms, including chest pain, dyspnea, and low-grade hemoptysis, had been managed during previous hospitalizations with a mild response to antibiotics. Multiple chest CTs showed multilobular consolidations with multifocal discoid scarring, ground glass opacities, and cicatricial bronchiectasis at sites of prior consolidations (Figure 2). Tissue cultures from the lung were negative for fungus, bacteria, and acid-fast bacillus. Serial lung biopsies revealed a neutrophil-predominant inflammatory process without vasculitis (Figure 3). Serologic workup with anti-neutrophil cytoplasmic antibodies (ANCA) was negative, as was subsequent workup for histoplasmosis and other infections. Her cavitary masses improved with steroids, only to recur off therapy. Serum protein electrophoresis revealed two IgA kappa monoclonal proteins, mildly elevated IgA and IgM, and normal IgG, without evidence of hematologic malignancy. A papulopustule on the leg was biopsied and rapidly progressed to ulceration; skin histopathological findings were not suggestive of vasculitis (Figures 3 and 4). While hospitalized for her ulcer, the patient received intravenous steroids and immunoglobulin (IVIG). She was discharged on oral tacrolimus and prednisone with improvement in both pulmonary and cutaneous symptoms. Pulmonary manifestations preceding cutaneous PG are exceptionally uncommon and present a diagnostic and therapeutic challenge. This case underscores the importance of maintaining a high index of suspicion for PG with pulmonary involvement in patients presenting with overlapping cutaneous and pulmonary symptoms not suggestive of another etiology.

Category: Clinical Case

Teaching point: Pyoderma gangrenosum should be considered in patients with overlapping pulmonary and dermatologic symptoms not otherwise explained by vasculitis, infection, or malignancy.

Figure 1. Ulcerated plaque with purple undermined borders on lateral aspect of lower extremity consistent with pyoderma gangrenosum.

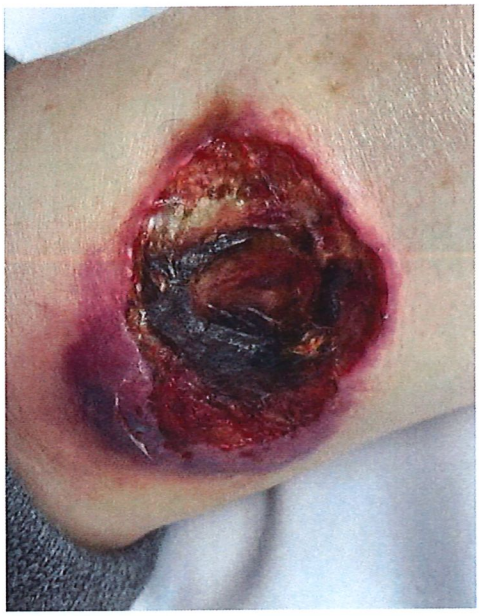


Figure 2. Computed tomography (CT) chest showing left upper lobe consolidation with ground glass opacities and biapical small centrilobular nodules.

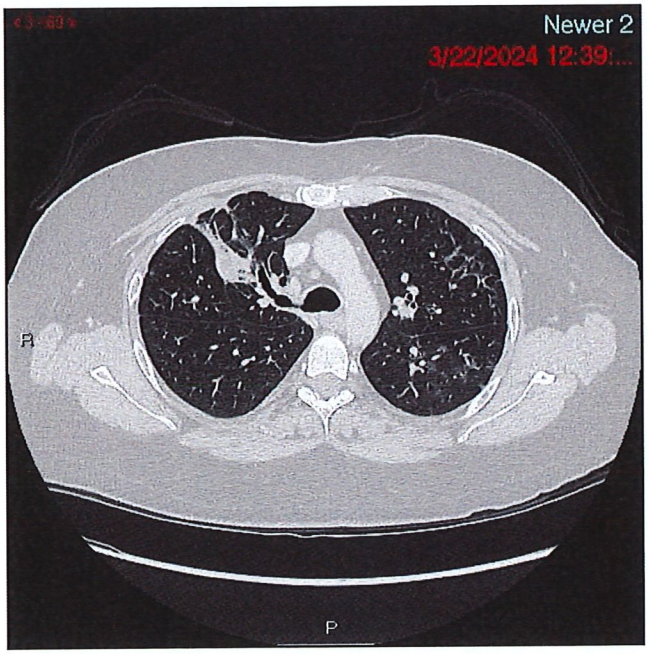


Figure 3. Punch biopsy of papulopustule on lower extremity showing neutrophil predominant dermal inflammation without evidence of vasculitis.

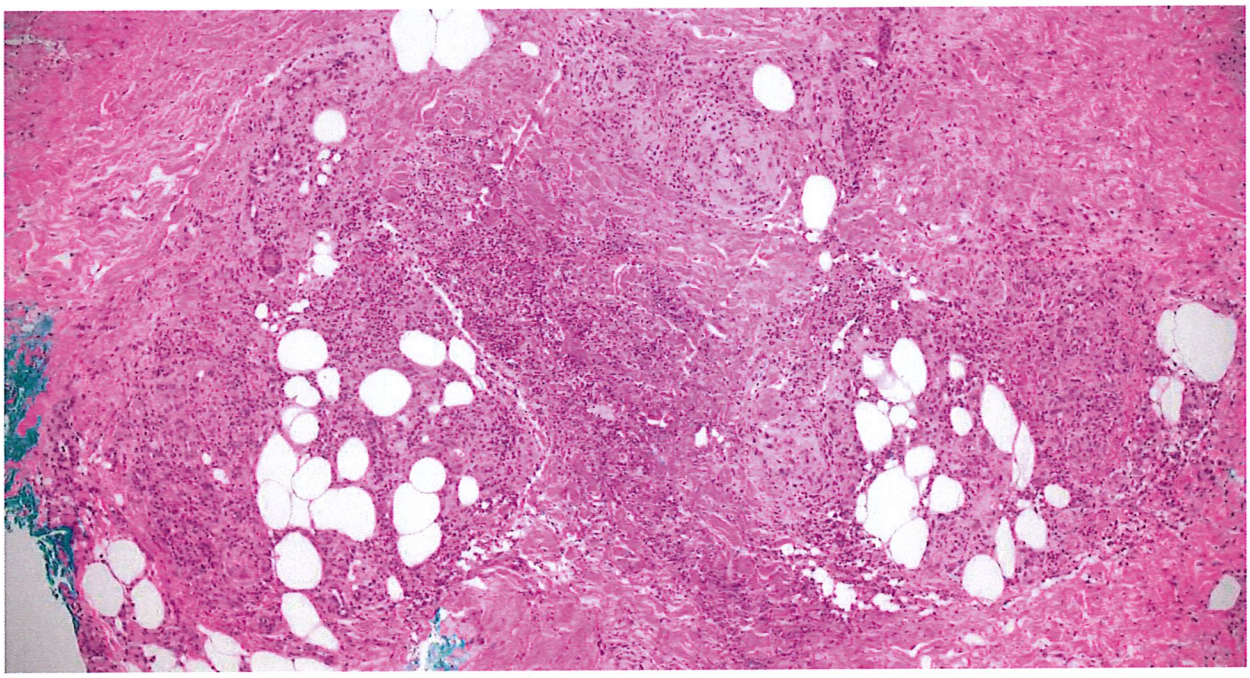
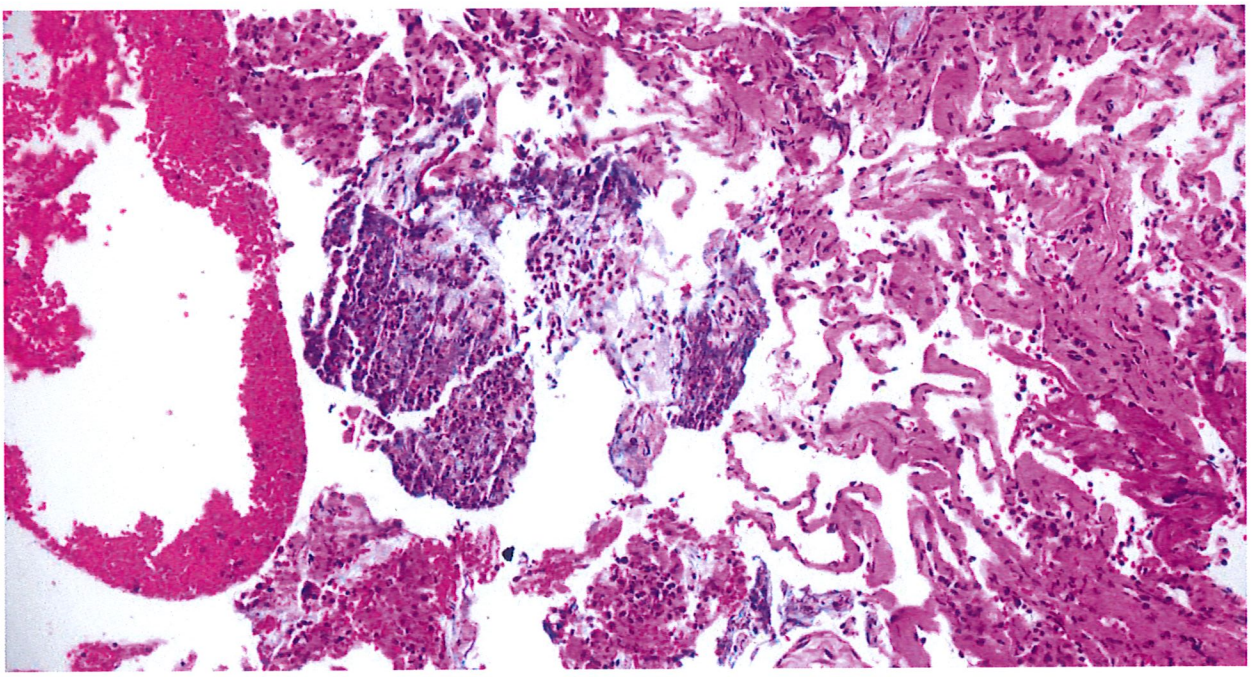


Figure 4. H&E Stain 100x Lung biopsy with neutrophil-predominant inflammation.



NON-UREMIC CALCIPHYLAXIS OF THE LOWER LEGS ASSOCIATED WITH FUTIBATINIB, A NOVEL FGFR INHIBITOR

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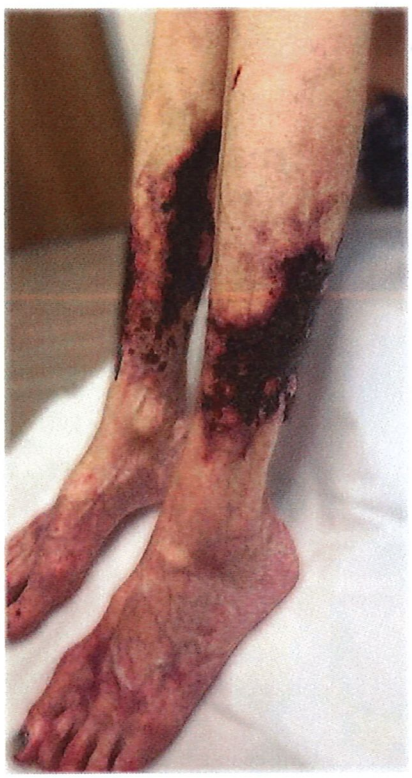
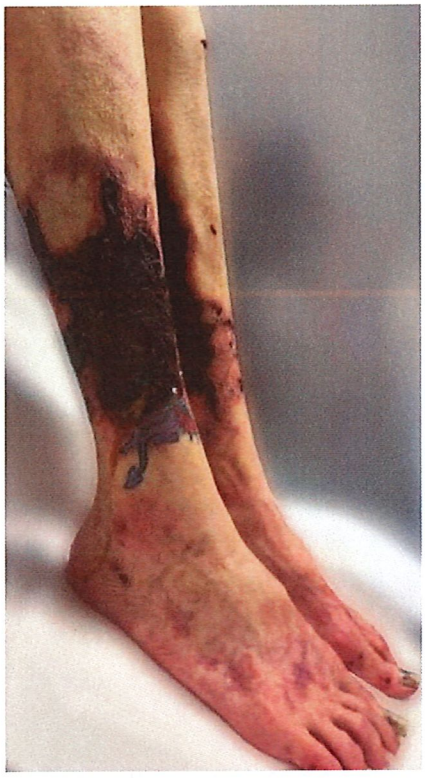
Abstract:
A 61-year-old woman with stage IIIc intrahepatic cholangiocarcinoma presented with exquisitely tender, black, necrotic, and stellate plaques with surrounding livedoid and violaceous erythema of bilateral lower legs. At an outside hospital, vasculitis had initially been suspected, and high-dose corticosteroids were prescribed. Notably, two weeks prior, futibatinib, a fibroblast growth factor receptor (FGFR) inhibitor, had been initiated. Calcium levels peaked at 11.1 mg/dL and phosphorous levels at 7.4 mg/dL, in the setting of tumor lysis syndrome and calcium supplementation. Though atypical for calciphylaxis to occur on the lower legs, this diagnosis was clinically favored in dermatology clinic. X-ray showed faint vascular calcifications in bilateral calves, consistent with calciphylaxis. Given the degree of pain, skin biopsy was initially deferred. IV sodium thiosulfate three times weekly and sevelamer were started. Futibatinib was eventually held. One month later, calciphylaxis worsened. Therefore, aspirin, pentoxifylline, and sildenafil were initiated. Five weeks later, she received a xenograft, wound care, and pain management in the hospital. Unfortunately, sodium thiosulfate and sevelamer had to be held due to refractory hypocalcemia and hypophosphatemia. Upon follow-up two weeks later, fixed livedoid racemosa extended to the thighs with new calcified subcutaneous nodules. Skin biopsy at that time showed lobular fat necrosis with calcification of vessels, confirming calciphylaxis. Given limited therapeutic options, epoprostenol, a prostacyclin analog with potent vasodilatory properties administered for severe Raynaud's crisis, was administered over 72 hours. Thereafter, livedoid changes rapidly receded from the thighs and ulcer progression halted. We present this case to help raise awareness of calciphylaxis as a serious adverse event of futibatinib, a novel FGFR2 inhibitor. In this setting, calciphylaxis presents unconventionally: renal disease can be absent, lower legs are involved, and typical therapies might not be suitable. We report a novel use of a prostacyclin analog, epoprostenol, for those with refractory disease.

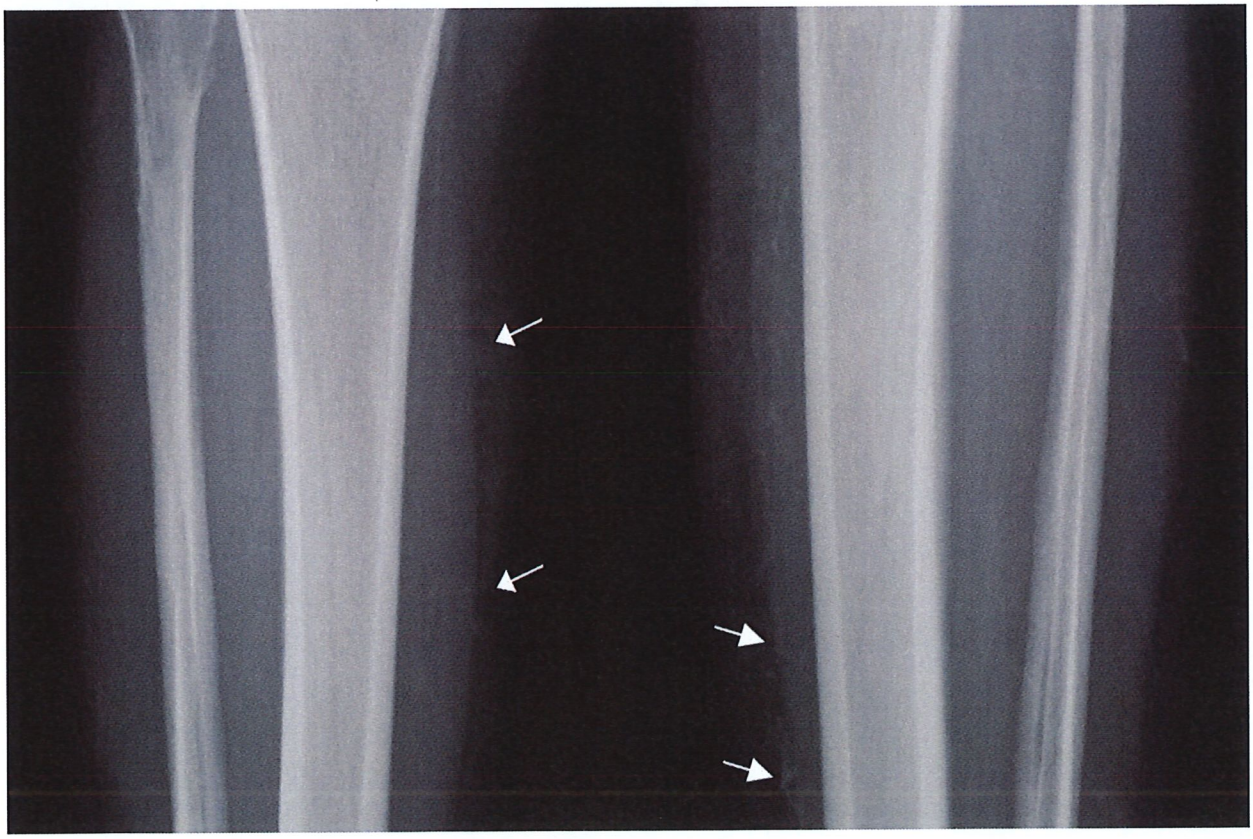
Teaching Point: Non-uremic calciphylaxis is a potential severe adverse effect of FGFR inhibitors like futibatinib, and epoprostenol may be considered for treatment in refractory cases.

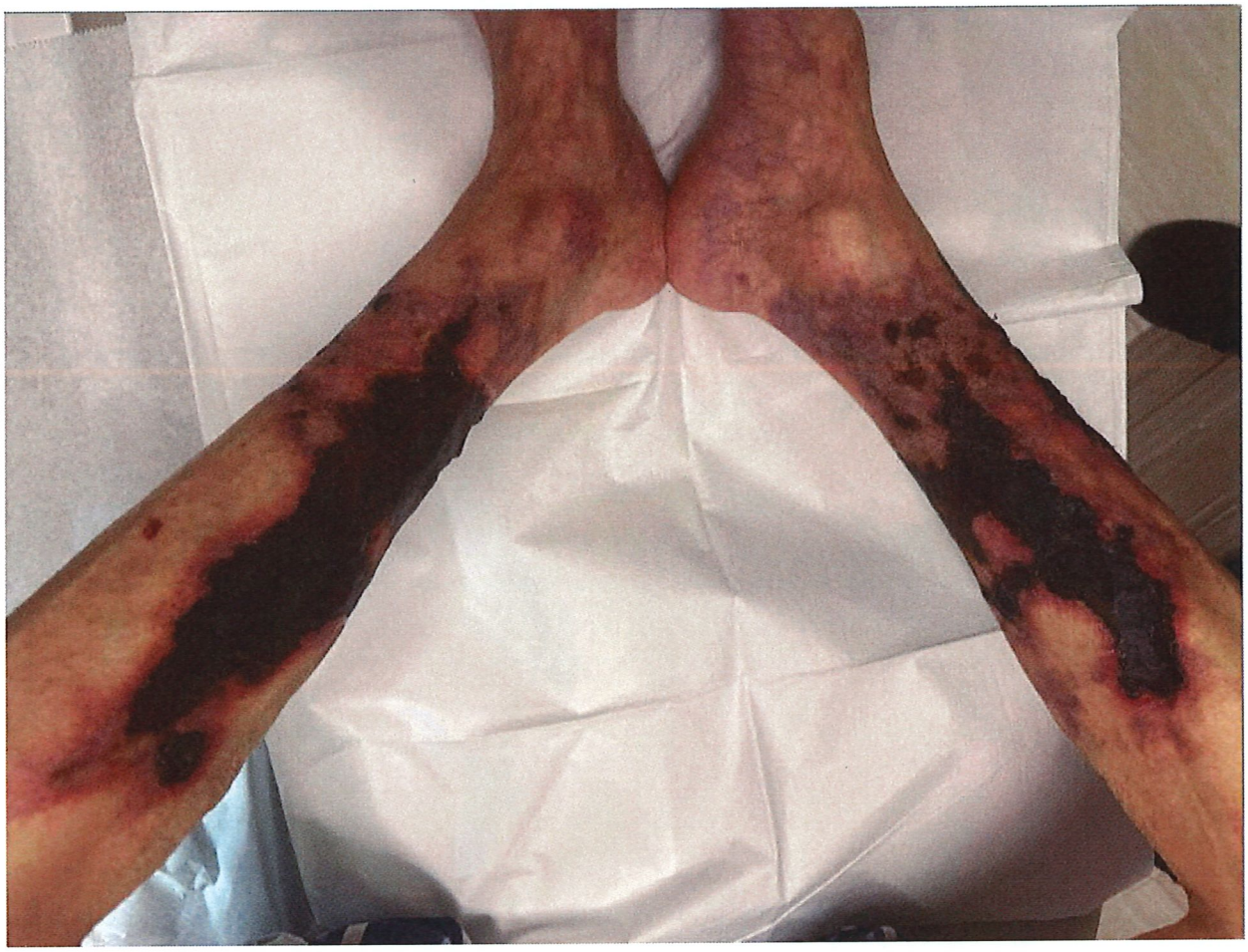
Abstract Category: Clinical Case

Images:









Don't get vexed: critical role of dermatologists in diagnosis of VEXAS syndrome

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Word count: 295

Catergory: Miscellaneous rheumatic skin disease

VEXAS (vacuoles, E1 enzyme, x-linked, autoinflammatory, somatic) syndrome is a recently described systemic autoinflammatory disease with overlapping dermatologic, rheumatologic, pulmonary, and hematologic manifestations, most commonly affecting elderly males. Cutaneous manifestations are often the earliest presenting feature of VEXAS, positioning dermatologists uniquely to facilitate timely diagnosis.

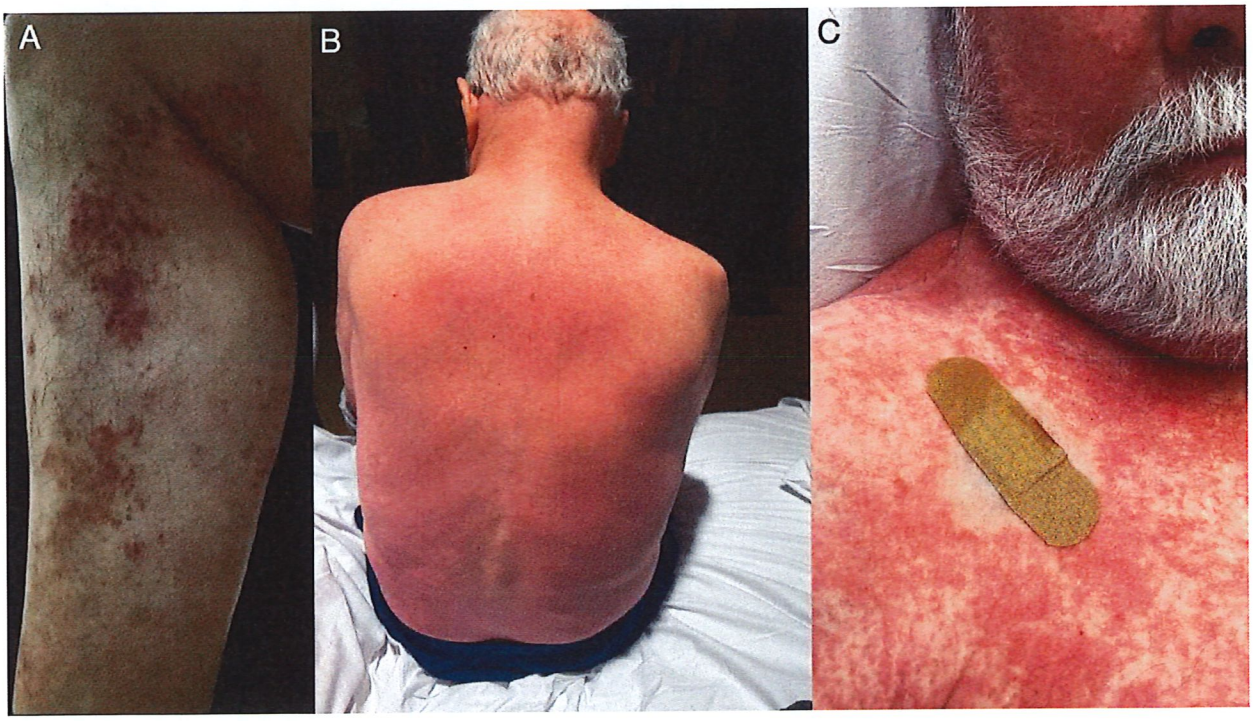
A 74-year-old Caucasian man with a past medical history of childhood asthma, hypertension, steroid-responsive interstitial pneumonitis, and recent cutaneous vasculitis was examined in the inpatient setting for a diffuse morbilliform eruption. (Figure 1) Punch biopsy of morbilliform lesions revealed neutrophil-rich superficial and mid-dermal inflammatory cells infiltrate with rare eosinophils and no evidence of vasculitis. Bone marrow testing or UBA1 mutation was positive for p.M41V mutation. He was started on a

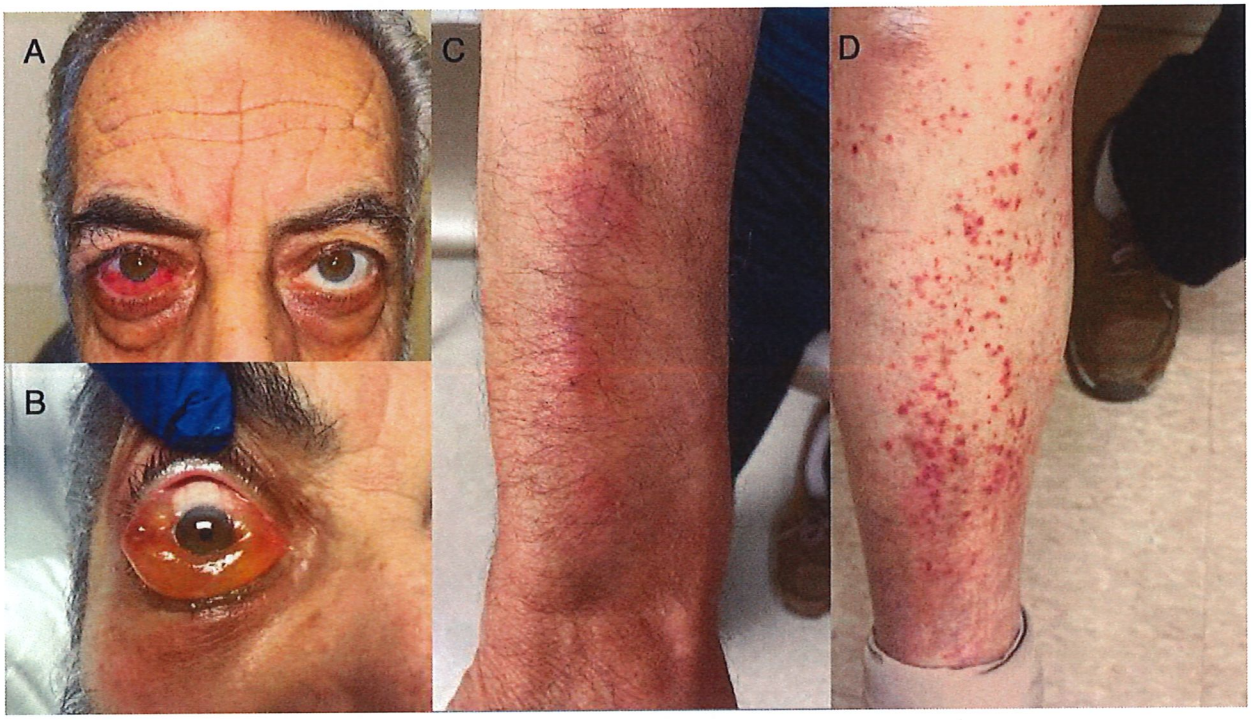
tapering dose of prednisone, tocilizumab, and colchicine, with complete control of his skin and systemic symptoms and improvement of laboratory parameters

A 73-year-old Algerian man with past medical history of monoclonal gammopathy of undetermined significance (MGUS), chronic kidney disease, rheumatoid arthritis, and previous deep vein thrombosis (on rivaroxaban) had a 5-month history of recurrent episodes of left periorbital swelling and pain (Figure 2A, B) that resolved with IV methylprednisone and antibiotics. Positron Emission Tomography (PET) scan revealed diffusely increased bone marrow with increased bone marrow uptake, prominent mediastinal lymph nodes, and similar splenic/liver uptake, significant for a lymphoproliferative process. Bone marrow testing for UBA1 mutation was positive for p.M41V mutation. He was started on 60 mg of prednisone, but his eyelid swelling relapsed on tapering steroids and was accompanied by palpable, non-blanchable purpura on extremities. (Figure 2 C, D) He was not amenable to a skin biopsy. He was subsequently started on ruxolitinib with resolution of symptoms.

Teaching point:

Given the high morbidity and mortality associated with VEXAS syndrome, appropriate detection of cutaneous symptoms and interdisciplinary management can play a vital role in the early diagnosis and subsequent management of patients.




A
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Poster Presentations

EFFICACY AND SAFETY OF METHOTREXATE AND PULSE MYTHYLPREDNISOLONE IN MORPHEA: A COHORT STUDY

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Morphea is an inflammatory skin disorder characterized by fibrosis primarily affecting the dermis. While multiple treatment modalities exist, supporting evidence remains limited. We sought to evaluate the effectiveness and safety of methotrexate and methylprednisolone, both as monotherapy and in combination, in treating morphea patients, and to identify predictors of treatment response. In this retrospective study, we reviewed medical records of 421 morphea patients treated at Tel-Aviv Sourasky Medical Center (2000-2022), with detailed analysis of 41 patients who received methotrexate and/or methylprednisolone. Among the 41 methotrexate-treated patients (78.1% female), 25 (60.9%) received concurrent pulse therapy with methylprednisolone. Complete response was observed in 31 patients (75.6%), partial response in 3 (7.3%), and no response in 7 (17.1%). Patients treated with MTX in combination with pulse methylprednisolone achieved a higher rate of CR (21 of 25 patients, 84%), while CR rate for MTX monotherapy was lower (10/16, 62.5%). Lesions in the head and neck, namely coup de sabre, were significantly associated with treatment success (P = 0.008). Adverse effects occurred in 15 patients (36.6%), 12 of whom were concurrently treated with methylprednisolone. Gastrointestinal symptoms were most common. Treatment discontinuation was primarily due to completion of the treatment regimen (39.0%), non-adherence (36.6%), and adverse effects (22.0%). Methotrexate, particularly in combination with methylprednisolone, is an effective and well-tolerated treatment for morphea, with a high proportion of patients achieving CR. Certain factors, such as lesion location or disease subtype, may influence treatment success. Our results reinforce existing therapeutic strategies but highlight the burden of adverse effects and the need for more targeted interventions in severe cases.

Category: Sclerotic skin disease

NODULAR SCLERODERMA: A CASE SERIES AND CLINICOPATHOLOGIC CHARACTERIZATION OF A RARE SCLEROTIC SKIN DISEASE VARIANT

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Nodular scleroderma (NS) is a rare cutaneous manifestation of systemic sclerosis (SSc) presenting as indurated, circumscribed papules or plaques, clinically resembling keloids or hypertrophic scars[1]. Approximately 66 cases have been reported, and its occurrence in connective tissue overlap syndromes (OS) remains poorly characterized[1]. We conducted a chart review of seven patients (skin types III–VI) with NS; six had skin biopsies. All met the 2013 ACR/EULAR SSc classification criteria. Three patients had OS: one with mixed connective tissue disease, one with systemic lupus erythematous, and one with subacute lupus erythematous. Mean age at nodular lesion onset was 53 years; six were female. In six cases, nodular lesions appeared an average of three years after connective tissue disease diagnosis; one case presented concurrently. Lesions were hyperpigmented, firm dermal plaques on the trunk or upper extremities, without preceding trauma. Six patients had pulmonary involvement, including interstitial lung disease and pulmonary hypertension. Histopathology demonstrated thickened dermal collagen bundles, perivascular lymphoplasmacytic infiltrates, and preservation of subcutaneous fat, distinguishing these lesions from keloids and hypertrophic scars. Topical or systemic corticosteroids, mycophenolate mofetil, and other immunomodulators tried yielded limited improvement. One patient improved after systemic chemotherapy for lung cancer. NS appears more often in middle-aged women and ethnic skin, potentially reflecting genetic predilection to hypertrophic scarring[2,3]. Histopathologic findings for NS are not well characterized and can have overlapping features with morphea, keloid, and hypertrophic scar[4]. Our series highlight NS's diagnostic challenge, which can mimic other fibrosing dermatoses and abnormal scarring processes, and the value of clinicopathologic correlation in patients with OS. Given its rarity, further studies are needed to clarify pathogenesis, prognosis and treatment options in patients with NS.

Teaching Point: Nodular scleroderma is a cutaneous manifestation of systemic sclerosis that clinically mimics keloids and hypertrophic scars, and clinicopathologic correlation can help distinguish this entity from other fibrosing disorders.

Abstract Category: Sclerotic skin disease

References:

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4. Richarz NA, Olivé A, García-Patos V, Quirant B, Fernández Figueras MT, Bielsa I. A review of the clinically distinguishing features of nodular or keloidal scleroderma in systemic sclerosis. Australas J Dermatol. 2020;61(2):e269-e273. doi:10.1111/ajd.13239

ASSOCIATION BETWEEN CELIAC DISEASE AND MORPHEA: A NATIONAL DATABASE ANALYSIS

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Your abstract body here: (300 words maximum)

Celiac disease (CD) is a gluten-induced, immune-mediated enteropathy that can present with a wide spectrum of extraintestinal manifestations including dermatologic manifestations, most commonly dermatitis herpetiformis. Morphea is an inflammatory cutaneous disorder that leads to fibrosis of the skin and underlying soft tissues. While multiple case reports have presented an association between CD and morphea, the relationship is not well studied. This study uses a national database representative of the United States adult population to further delineate the association between CD and morphea. The All of Us Database was used to query patients with celiac disease as well as patients with morphea. Patients were required to have at least two instances of the given diagnosis to be considered within that cohort. Multivariate regression was performed to assess the odds ratio (OR) of patients with celiac disease having morphea, while adjusting for race, gender, ethnicity, and age. Patients with CD were at a significantly increased risk of having morphea (OR=3.08; 95% CI = 2.02 - 4.68; $p<0.001$) compared to the individuals without CD. Expected signals within the general population for morphea were also observed including a significant increase among females (OR=9.92; 95% CI = 7.79 - 12.64; $p<0.001$) and across all non-White populations, including the Asian (OR=0.53; 95% CI = 0.30 - 0.91; $p=0.022$), Black or African American (OR=0.40; 95% CI = 0.31 - 0.51; $p<0.001$), and Other Race (OR=0.68; 95% CI = 0.57 - 0.81; $p<0.001$) cohorts. Based on our findings, patients with CD have over threefold greater odds for morphea compared to individuals without CD. Given the well-known association between CD and dermatologic conditions, morphea may be a presenting manifestation of CD. Recognition of this association may lead to earlier diagnosis and better outcomes for patients.

Category: Sclerotic skin disease (Morphea)

residency and perpetuate chronic fibrosis. Our findings suggest that inhibition of TREM2 and necroptosis represent rational targeted therapeutic strategies in morphea and EF.

Teaching point: Morphea and EF pathogenesis involves an immune circuit in which cytotoxic T cells drive tissue injury and $TREM2^{+}$ macrophages contribute to fibroblast activation.

Category: Sclerotic skin disease

DEMOGRAPHIC PREDICTORS OF GANGRENE AND HOSPITALIZATION IN SYSTEMIC SCLEROSIS PATIENTS WITH RAYNAUD'S PHENOMENON: AN ANALYSIS USING NIS AND NEDS DATABASES

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Systemic sclerosis (SSc) is an autoimmune connective tissue disease commonly presenting with Raynaud's phenomenon (RP), with the concerning progression to gangrene. We analyzed demographics, insurance, healthcare utilization, and other variables in 1) 2015-2019 National Inpatient Sample (NIS) patients with ICD-10 diagnoses of SSc and RP to assess factors associated with gangrene; and 2) 2006-2012 Nationwide Emergency Department (NEDS) patients with ICD-9 diagnoses of SSc seen in the ED for RP to assess factors associated with inpatient admittance. Statistical analysis was performed using SAS 9.4. Using NIS, we identified 8,362 patients with SSc and RP, of which 388 had RP with gangrene (mean age=58.2 years; female=87.6%; White=57.8%, Black=20.5%, Hispanic=17.0%, Asian=2.7%, Other=1.9%) vs 7,974 had RP without gangrene (mean age=61.1 years; female=85.2%; White=68.8%, Black=15.6%, Hispanic=10.4%, Asian=2.0%, Other=3.2%). Statistically significant factors associated with RP with gangrene were age (t-test, $p<0.0001$), race (Fisher's exact $p<0.0001$; notably, 7.3% Hispanic, 5.9% Black, 3.9% White RP patients had RP with gangrene), LOS (mean 9.4 vs 5.9 days, $p<0.0001$), and total charges (mean \$108,760 vs \$71,401, $p<0.0001$). Within NEDS, we identified 7,407 patients with SSc and RP (mean age=58.3 years; female 88.0%), of which 129 had RP as their primary ED diagnosis (mean age=51.0 years; female 89.9%). Statistically significant variables associated with inpatient admittance from the ED (n=87) vs ED discharge (n=42) were primary payer (81.2% of Medicare, 63.8% private, 60.0% Medicaid, 42.9% other/self-pay patients were admitted, Fisher's exact $p=0.0293$) and number of comorbidities [mean 11.2 (SD=2.3) vs 4.5 (SD=4.7), t-test $p<0.0001$]. While not statistically significant given sample size, 92.3% of males vs 64.7% of females were admitted. In patients with SSc and RP, vigilant management, particularly in patients who are younger, Hispanic or Black, with specific insurance, and with higher comorbidities, is warranted to reduce the risk of gangrene and improve healthcare utilization.

Teaching Point: Demographics can be helpful in identifying systemic sclerosis patients with Raynaud's phenomenon at risk of progression to gangrene or severe disease requiring inpatient admittance.

Abstract Category: Sclerotic skin disease (e.g. morphea, systemic sclerosis, etc.)

RESOLUTION OF MORPHEA FOLLOWING TREATMENT WITH DUPILUMAB

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Morphea (localized scleroderma) is a chronic inflammatory skin disorder characterized by excessive collagen deposition in the dermis and subcutaneous tissue. The pathogenesis involves immune dysregulation, endothelial injury, and fibroblast activation. The disease evolves from an initial inflammatory phase, driven by Th1-mediated cytokines, to a Th2-mediated fibrotic phase where IL-4 and IL-13 play key roles in promoting fibrosis. Dupilumab, an IL-4/IL-13 inhibitor, has shown promise in treating Th2-mediated fibrotic diseases. By targeting these cytokines, dupilumab inhibits fibroblast activation and collagen deposition. In treatment-refractory morphea, dupilumab offers a novel therapeutic approach by modulating the immune response and preventing further fibrosis. We present a patient with generalized morphea and concomitant atopic dermatitis who experienced improvement of both conditions when treated with dupilumab. This case highlights the use of dupilumab as a therapeutic option to halt disease progression and potentially reverse established morphea lesions in patients with refractory disease. Additional studies are needed to further evaluate the efficacy of dupilumab in the treatment of morphea.

Teaching point: Dupilumab may represent a targeted therapeutic approach to halt disease progression and potentially reverse existing lesions in patients with refractory morphea.

Category: Sclerotic skin disease

A CLOSER LOOK AT MORPHEA AND MALIGNANCY

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Patients with systemic sclerosis are at a higher risk of malignancy, including hematologic malignancies. However, these studies often exclude patients with morphea, a cutaneous form of sclerosis characterized by patches or plaques of skin that become thickened, firm, and discolored due to excess collagen deposition. There have been case reports of patients with morphea and hematological malignancies, including MALT lymphoma and cutaneous T-cell lymphoma. Data on the association between morphea and hematological malignancies remains limited. In this study, we aim to explore this association using the All of Us (AoU) database, a US-based multi-modal research dataset of participants collected from surveys, electronic health records (EHR), and physical measurements. Using the AoU Registered Tier Dataset v7, we identified patients with EHR data and performed a matched, cross-sectional analysis with nearest neighbor propensity score matching by age, sex, race/ethnicity, and average EHR observation period. Covariates were well-balanced between groups, with standardized mean differences of <0.1. Participants with systemic sclerosis or missing covariates were excluded. In our analysis, we found no difference in the incidence of hematological malignancies within five and ten years of diagnosis of morphea, including chronic myeloproliferative disease, Hodgkin lymphoma, multiple myeloma, leukemia, non-Hodgkin lymphoma, and monoclonal gammopathy between morphea and control patients. Of note, patients with morphea were more likely to have also been diagnosed with atopic dermatitis (8.2% vs 4.9%, $p=0.004$) and an autoimmune disease (45.9% vs 31.0%, $p<0.001$). The autoimmune variable was defined as the presence of at least one of the following conditions: alopecia areata, autoimmune thyroiditis, celiac disease, dermatomyositis, inflammatory bowel disease, lichen planus, psoriasis, rheumatoid arthritis, systemic lupus erythematosus, type 1 diabetes, vitiligo, ankylosing spondylitis, and Sjögren's syndrome. This report demonstrates that unlike systemic sclerosis, patients with morphea may not be at increased risk of malignancy, despite frequent co-incidence of systemic autoimmune conditions.

Teaching Point: Unlike systemic sclerosis, morphea, a sclerotic condition limited to the skin, may not be associated with hematologic malignancy. Consistent with prior literature, patients with morphea may be more likely to develop other autoimmune diseases.

Category: Sclerotic skin disease

A CASE OF EOSINOPHILIC FASCIITIS REFRACTORY TO CORTICOSTEROIDS

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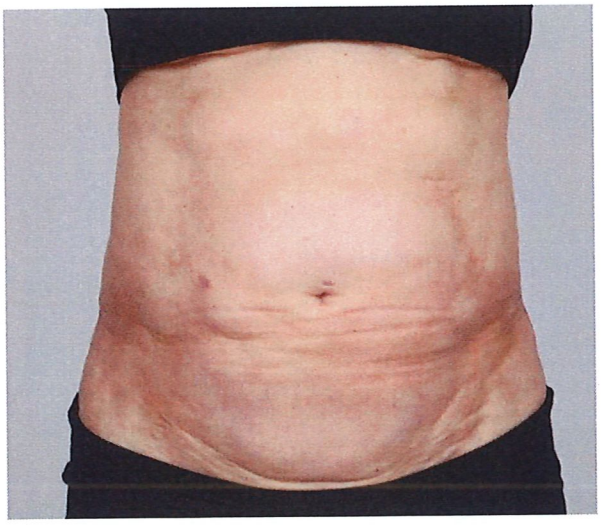
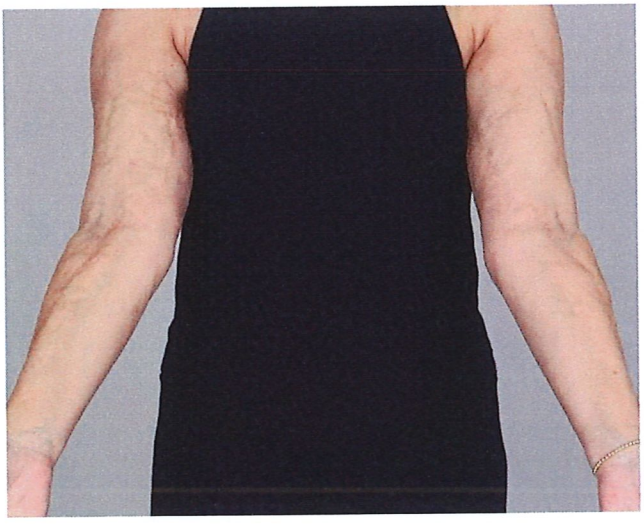
Eosinophilic fasciitis (EF) is a rare connective tissue disorder characterized by inflammation and thickening of the fascia leading to skin induration. It is often accompanied by peripheral eosinophilia and elevated erythrocyte sedimentation rate. We report the case of a 56-year-old female who presented with progressive stiffness, swelling, and pain of the upper and lower extremities, trunk, and buttocks over a 3-month period. Physical examination revealed woody induration of the skin with a "groove sign" along superficial veins. Laboratory evaluation was largely unremarkable except for a mild elevation in C-reactive protein. A full-thickness incisional biopsy confirmed the diagnosis, revealing skin and soft tissue with fascial thickening, plasma cells, and rare eosinophils which supported a diagnosis of eosinophilic fasciitis. Her disease was refractory to high dose corticosteroids and methotrexate. The patient responded favorably to intravenous immunoglobulin (IVIG) and cyclosporine, with marked clinical improvement. This case underscores the importance of early recognition and histopathologic confirmation of eosinophilic fasciitis, which can mimic other inflammatory or infectious conditions, and highlights the role of immunosuppressive therapy beyond corticosteroids in disease management.

Teaching Point: In cases of eosinophilic fasciitis refractory to corticosteroids, treatment with IVIG and cyclosporine may be effective in reducing the clinical burden of this disease.

Category: Sclerotic skin disease





COMPARATIVE EVALUATION OF MORPHEA TREATMENT RESPONSE USING MORPHEA ACTIVITY MEASURE VS. LOSCAT

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Word count: 230

Morphea, also known as localized scleroderma, is a chronic inflammatory skin disorder that requires accurate assessment of disease activity to guide treatment, but current tools have limitations. The Morphea Activity Measure (MAM), initially validated in pediatric populations, offers a potential alternative. To evaluate the correlation between MAM and Localized Scleroderma Cutaneous Assessment Tool (LoSCAT) in assessing treatment response in both adult and pediatric morphea patients and to determine the inter-rater reliability of MAM over a six-month period. This prospective study aims to evaluate the correlation of MAM with LoSCAT, as well as assess the inter-rater reliability of MAM scores during a prolonged follow-up period in both adult and pediatric patients. In this single-center, prospective observational study, 33 patients underwent evaluation at baseline and six months post-treatment. MAM was scored independently by two dermatologists, while LoSCAT was assessed by one evaluator. MAM scores significantly decreased post-treatment (Evaluator 1: $p < 0.001$; Evaluator 2: $p = 0.015$), reflecting improved disease activity. MAM showed moderate correlation with LoSCAT ($\rho = 0.688–0.841$, $p < 0.001$) and excellent inter-rater reliability (ICC > 0.90). Some MAM components, such as violaceous rim and warmth, had poor agreement. Small sample size in subgroups and short follow-up period limit generalizability. In conclusion, MAM score is a responsive and reliable tool for assessing morphea activity, with moderate correlation to LoSCAT. Refinement of certain components may enhance its clinical utility.

Abstract category: Sclerotic skin disease (Morphea)

DRUG, VACCINE, VITAMIN, AND CHEMICALS-INDUCED MORPHEA A SYSTEMATIC REVIEW OF 92 CASES

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Word count: 262

Morphea is a rare autoimmune disease primarily affecting the skin, with a complex pathogenesis involving genetic, environmental, and immune-related factors. There are several reports of morphea progression, relapse, or incidence after using specific chemicals, vaccines, or drugs. We aim to identify chemical agents, vaccines, and drugs associated with morphea, as well as the disease course and outcome of drug-induced cases. We conducted a systematic search through PubMed/Medline, Scopus, Web of Science, and Embase using MeSH terms/keywords Morphea or Localized Scleroderma and drug-induced or drug-associated or vaccine-associated or vaccine-induced or induced until September 27, 2024. Our study encompasses 92 cases of through 78 reports. The study highlights a female predominance (73.9%) with an average patient age of 50.1 years. The onset of morphea varied widely, occurring from 2 days to 10 years post-agent exposure. Four medication categories were identified: biologics (21.8%), immune checkpoint inhibitors (21.8%), vaccines (24.3%), and miscellaneous drugs (32%). Among biologics, interferons and TNF-α inhibitors, with ustekinumab and dupilumab reported as the most common ones. Immune checkpoint inhibitors such as nivolumab and pembrolizumab were the most frequently associated. Vaccines, primarily Pfizer-BioNTech COVID-19, contributed to generalized and deep morphea cases. Also, Balicatib and vitamin K1 injections were showed to be the most common one in last group. Treatments varied, including immunosuppressants, phototherapy, and topical agents. Clinical resolution occurred in many cases, though some progressed or remained uncontrolled. This study highlights the complex relationship between morphea and various triggering factors, particularly immune-modulating therapies and vaccines. Our findings emphasize the predominance of female patients and the significant role of autoimmune mechanisms in disease pathogenesis.

Abstract category: Sclerotic skin disease (Morphea)

SKIN PAIN SEVERITY AND FREQUENCY IN CUTANEOUS LUPUS ERYTHEMATOSUS PATIENTS CORRELATE WITH CLASI ACTIVITY SCORES

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Skin pain and itch are two frequently reported symptoms that can contribute significantly to reduced quality of life (QoL) in cutaneous lupus erythematosus (CLE). Though clinicians observe and evaluate signs of skin disease activity with measures such as CLASI-A, they cannot assess the experience with and symptom burden of CLE as perceived by patients. Earlier studies have assessed health-related QoL in CLE using patient-reported measures (PROMs) like the Dermatology Life Quality Index; however, these tools lack dedicated items for pain and itch. Measures like Skindex-29+3 and patient global assessments (PtGAs) of skin itch and pain provide a focused measurement of symptom frequency and severity, but have not been compared extensively with clinician-assessed skin activity measures. This study investigates the correlation between the frequency and severity of patient-reported skin itch and pain, as measured by Skindex-29+3 and PtGAs respectively, and CLASI-A scores. A cross-sectional analysis of CLE patients diagnosed with discoid lupus erythematosus (DLE) and/or subacute cutaneous lupus erythematosus (SCLE) was conducted at outpatient dermatology clinics at University of Texas Southwestern Medical Center and Parkland Health between January 2009 and June 2024. CLASI-A scores and PROMs including PtGA skin pain and itch and Skindex-29+3 were collected. Correlation analyses were conducted using Spearman's coefficients (r), and Kruskal-Wallis test was used to compare CLASI-A scores across severity groups. Overall, correlations were weak; Skindex pain ($r=0.32$, $n=291$) and PtGA pain ($r=0.34$, $n=94$) showed stronger correlations with CLASI-A than Skindex itch ($r=0.25$, $n=286$) and PtGA itch ($r=0.30$, $n=94$). CLASI-A scores differed significantly across Skin Pain PtGA severity groups ($H=13.31$; $p<0.001$), with patients reporting moderate or severe pain tending to have higher CLASI-A scores compared to those with mild symptoms. In conclusion, frequency and severity of skin pain correlate with CLASI-A scores. Our findings support the integration of skin pain focused PROMs into routine CLE assessments.

Category: Lupus

EVALUATION OF THE APPLICABILITY AND EASE OF USE OF THE CUTANEOUS LUPUS ERYTHEMATOSUS AREA AND SEVERITY INDEX (CLASI) IN FILIPINO CUTANEOUS LUPUS ERYTHEMSTOSUS PATIENTS

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Category: Lupus

ABSTRACT

Background: Skin involvement is common in systemic lupus erythematosus and is essential for diagnosis and monitoring. The Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) is validated in adults, and its use in Asian patients (skin phototypes III -V) is an emerging area of interest. This study evaluates its applicability in Filipino cutaneous lupus erythematosus (CLE) patients, which can aid in standardizing care, expanding clinical trial use, and guiding future therapies across diverse populations.

Methods: Fourteen (14) Filipino CLE patients were evaluated by two board-certified dermatologists, one with CLASI experience across all skin phototypes, and two board-certified rheumatologists. All physicians underwent standardized virtual training in the CLASI and the Physician Global Assessment (PGA) before evaluations. Each independently rated all patients using both tools and re-assessed two randomly selected patients. Reliability was analyzed using Intraclass Correlation Coefficients (ICC) within physician groups. Exit interviews were conducted post-assessment phase.

Results: CLASI activity and damage scores demonstrated excellent inter- and intra-rater reliability (ICC = 0.87–0.98) across all physicians, regardless of prior experience. PGA scores showed excellent inter-rater reliability for activity (ICC = 0.84) and good reliability for damage (ICC = 0.70), with excellent intra-rater reliability for both (ICC = 0.98). By specialty, CLASI reliability remained excellent for dermatologists (ICC = 0.92–1.00) and rheumatologists (ICC = 0.84–1.00). For the PGA, dermatologists had good inter-rater reliability for activity (ICC = 0.76) and moderate for damage (ICC = 0.60), while rheumatologists showed good reliability for activity (ICC = 0.77) and excellent for damage (ICC = 0.89). Intra-rater reliability for PGA activity and damage was excellent across specialties (ICC = 0.98-1.00). All physicians favored CLASI over PGA for CLE evaluation.

Conclusion: CLASI scoring demonstrated superior reliability compared to the PGA, supporting its practicality, consistency, and ease of use for assessing Filipino CLE patients.

Keywords: Cutaneous lupus erythematosus, CLE, Cutaneous Lupus Erythematosus Disease Area and Severity Index, CLASI, Systemic lupus erythematosus

SKIN PAIN SEVERITY AND FREQUENCY IN CUTANEOUS LUPUS ERYTHEMATOSUS PATIENTS CORRELATE WITH CLASI ACTIVITY SCORES

Rohan Ahuja, BA[1], Curtis Liu, BA[1], Grace Lu, BA[1], Jialiang Liu, PhD,[2] Pui Man Chan, MPH,[2] Jan Feifel,[3] Yulia Dyachkova,[3] Paul Kamudoni,[3] Josephine Park[4], Sanjeev Roy,[3] Caroline Foch,[3] Benjamin F. Chong, MD, MSCS[1]

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Category: Lupus

Urban vs Rural Disparities in Cutaneous Lupus Erythematosus

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Abstract:

Cutaneous lupus erythematosus (CLE) is a heterogeneous autoimmune disease subset primarily affecting the skin, causing significant morbidity and quality-of-life impairment. While systemic lupus erythematosus (SLE) has been extensively studied, particularly regarding urban and rural disparities, focused investigations on CLE stratified by geographic residence remain scarce. This review synthesizes existing literature on urban versus rural differences in CLE epidemiology, clinical presentation, environmental exposures, healthcare access, and outcomes. Evidence from systemic lupus cohorts suggests that urban patients often experience earlier diagnosis but face longer specialist wait times, whereas rural patients encounter significant travel burdens and limited local expertise, contributing to diagnostic delays. Environmental factors such as ultraviolet exposure, pesticide contact, and pollutant exposure vary by residence and may differentially influence CLE manifestations. Socioeconomic status and healthcare infrastructure disparities further compound these differences, affecting disease severity and management adherence. The lack of CLE-specific prospective studies and clinical trials evaluating geographic disparities represents a critical research gap. Addressing this gap through multicenter, prospective investigations and innovative care delivery models, including teledermatology, may improve equity in CLE outcomes. Understanding urban–rural distinctions is essential for tailored diagnostic, therapeutic, and public health strategies to optimize care for all CLE patients regardless of geographic location.

Category: Lupus

PRESERVING RESPONSIVENESS OF THE CLASI-A: IMPACT OF ALOPECIA AND MUCOUS MEMBRANE COMPONENTS IN LOCALIZED DISEASE AND FOCAL ALOPECIA

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Alopecia and mucous membrane lesions impair quality of life in cutaneous lupus erythematosus (CLE) and correlate with disease activity/damage. Recent debate has questioned whether these domains should remain in Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI). We assessed the impact of removing alopecia and mucosal components from CLASI-Activity score (CLASI-A) in a registry including subacute CLE (SCLE), localized discoid lupus (DLE-L), and generalized discoid lupus (DLE-G). Patients were selected based on two consecutive visits, baseline CLASI-A $\geq$8, and alopecia or mucosal involvement. The cohort included 149 patients (128 female, 21 male), median age 47.2 years, and average 323 days between visits. Racial distribution: 53% White, 40% Black or African American, and 5% Asian. Alopecia was categorized hierarchically: Category 1 = self-reported alopecia or mucosal lesions without observed alopecia; Category 2 = diffuse, non-scarring alopecia; Category 3 = focal or patchy alopecia. We calculated percent change between visits for total CLASI-A and a modified CLASI-A excluding alopecia/mucosal components. Paired difference (%Δ Total – %Δ Modified) was analyzed using Wilcoxon signed-rank tests. Removing alopecia/mucosal points had the largest and only statistically significant impact in Category 3, baseline CLASI-A 8–10 (n = 38; median –12.5%, IQR –25.7, 0.0; P = 0.004), indicating improvement would be underestimated by 12.5% if these components were excluded. This effect was driven by the DLE-L subgroup (n = 25; P = 0.007). SCLE and DLE-G trended similarly but did not reach significance, likely due to smaller subgroup sizes. No

significant impact was seen in Categories 1–2 or among baseline CLASI-A >10. These findings suggest focal alopecia retains responsiveness and mucosal lesions contribute meaningfully to change at low disease activity. Omitting these domains risks underestimating disease activity and deprioritizing high-impact features. Our data support retaining alopecia and mucous membrane components in CLASI-A scoring for CLE.

Category: Lupus

Keratinocyte-derived Extracellular Vesicles Mediate UVB-induced inflammation through Innate Immune Activation and Type I Interferon Production

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Extracellular vesicles (EVs) are cell-secreted lipid particles linked to photosensitive autoimmune diseases pathogenesis e.g., Dermatomyositis (DM) and SLE, where UVB can trigger disease flares. Because UVB penetrates only superficial skin layers, we hypothesized that keratinocyte-derived EVs (KEVs) mediate UVB-photodamage by transmitting inflammatory signals from keratinocytes (KCs) to dermal immune cells. Herein, we identify immune cells responding to KEVs. KCs were irradiated with 75 mJ/cm² UVB or sham ± IFNβ for KEVs production. Healthy PBMCs were incubated with KEVs and analyzed by flow cytometry. EVs from UVB-irradiated KCs (UVB-KEVs) stimulated more IFNβ production than sham-KEVs in myeloid DCs (mDCs), monocyte-derived DCs (MoDCs), macrophages, T cells ($CD4^+$ & $CD8^+$), and B cells. Notably, only EVs from IFNβ-pretreated irradiated KCs (IFNβ-KEVs) significantly increased IFNβ in pDCs and NKs. UVB-KEVs also increased IFNγ in MoDCs, macrophages, T cells, and B cells. Only IFNβ-KEVs significantly stimulated IFNγ in pDCs. UVB-KEVs increased p-NFκB in NKs, T cells, and B cells. UVB-KEVs significantly activated p-STING in pDCs, NKs, MoDCs, macrophages, and T cells. In mDCs, however, significant pSTING activation occurred only with IFN-β-KEVs compared to UVB-KEVs. p-STING activation was greatest in macrophages and MoDCs. IFNγ was highest in pDCs (by IFNβ-KEVs); IFNβ was most pronounced in mDCs and pDCs (by IFNβ-KEVs). IFNβ-KEVs uniquely co-induced IFNβ and IFNγ in pDCs. Notably, previous work found mDCs enriched in antimalarial non-responder DM skin, expressing high IFNβ and p-STING. Blood pDCs and mDCs in DM secreted more IFNβ than controls, and pDCs were enriched in DM skin, producing high IFNβ and IFNγ. In summary, UVB-KEVs produce type-I IFNs and activate pSTING—both upregulated in SLE and DM— especially innate cells. IFNβ-KEVs distinctively activated mDC p-STING and drove pDC IFNβ and IFNγ, both critical in DM/SLE pathogenesis. This suggests a mechanism of UVB-triggered systemic inflammation via KEVs. Future directions include defining KEV cargo and blocking KEV release.

Abstract category: Lupus

PRESERVING RESPONSIVENESS OF THE CLASI-A: IMPACT OF ALOPECIA AND MUCOUS MEMBRANE COMPONENTS IN LOCALIZED DISEASE AND FOCAL ALOPECIA

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Category: Lupus

TOWARD CONSENSUS DEFINITIONS OF LOW DISEASE ACTIVITY AND REMISSION IN CUTANEOUS LUPUS ERYTHEMATOSUS: THE CASE FOR AN INVESTIGATOR GLOBAL ASSESSMENT INSTRUMENT

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Cutaneous lupus erythematosus (CLE) lacks standardized definitions of low disease activity (LDA), very low disease activity (VLDA), and remission, clinical milestones that are increasingly important in trials and practice. In atopic dermatitis and psoriasis, regulatory endpoints often incorporate composite scores such as the Eczema Area and Severity Index (EASI) or Psoriasis Area and Severity Index (PASI) alongside an Investigator Global Assessment (IGA), with thresholds such as clear (0) or almost clear (1) widely used due to their intuitive face validity and ease of clinical implementation.[1] These frameworks have been endorsed by the National Psoriasis Foundation (NPF), including recent guidance from the NPF Medical Board.[2] The International Eczema Council (IEC) is advancing a similar framework. In CLE, the CLA-IGA-R (Cutaneous Lupus Activity–Investigator Global Assessment–Revised) has undergone validation, demonstrating strong inter- and intra-rater reliability, and is easily deployable in clinical settings (feasibility). Static measures such as IGA also add value in clinical settings and treat-to-target studies, where prior disease activity may not be known or readily available. Ongoing studies are assessing the instrument's clinical meaningfulness through anchoring to patient-reported outcomes. We believe the CLA-IGA-R is especially useful for classifying low disease activity states and remission. Future consensus definitions could help define endpoints such as CLA-IGA-R thresholds alongside CLASI (Cutaneous Lupus Erythematosus Disease Area and Severity Index) scores (especially absolute CLASI cutoffs), as previously proposed.[3,4] In the IEC work, atopic dermatitis low disease activity is being defined by IGA and/or EASI cutoffs, by analogy. A formal Delphi process may play an important role in defining these states through expert consensus. Standardized IGA-based definitions of LDA, VLDA, and remission could help unify CLE outcome measures and ensure therapeutic targets reflect both clinical control and patient priorities. IDEOM, the International Dermatology Outcome Measures group has commenced work to evaluate low disease activity metrics across a variety of inflammatory dermatoses including CLE.

Abstract Category: Lupus

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THE ASSOCIATION BETWEEN SYSTEMIC LUPUS ERYTHEMATOSUS AND HIDRADENITIS SUPPURATIVA: A CROSS-SECTIONAL ANALYSIS IN THE ALL OF US RESEARCH PROGRAM

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Systemic lupus erythematosus (SLE) is a chronic, multisystemic autoimmune disease marked by autoantibody production, immune complex depositions, and inflammation affecting multiple organ systems. Hidradenitis suppurativa (HS) is a chronic, immune-mediated inflammatory skin disorder characterized by recurrent nodules, abscesses, and draining sinus tracts in intertriginous regions, with increasing recognition of systemic involvement. Both conditions disproportionately affect women, particularly those of racial and ethnic minority backgrounds, and share immunologic features, including dysregulation of innate and adaptive immunity and overlapping cytokine pathways. We aimed to investigate the relationship between SLE and HS in a large, diverse population using the All of Us Research Program database. We conducted a cross-sectional analysis identifying adult participants with SLE, HS, or both, using OMOP-standardized diagnostic codes. Demographics and clinical characteristics were compared using descriptive statistics and appropriate inferential tests. Among 628,590 participants, 4,947 had SLE, 2,513 had HS, and 98 were diagnosed with both. Patients with both conditions were more likely to be female, black, and have a higher BMI. The prevalence of HS in SLE patients was higher than in the general cohort, suggesting a potential association. Our findings support a possible immunologic link between HS and SLE, and highlight the importance of screening for systemic autoimmune disease in patients presenting with chronic inflammatory dermatoses. Further studies are warranted to clarify shared pathogenic mechanisms and explore common therapeutic strategies.

Category: Lupus

SINGLE CELL RNA SEQUENCING IN SMOKERS WITH CUTANEOUS LUPUS DEMONSTRATE INCREASED TYPE I INTERFERON RELATED GENE SIGNATURE

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Cutaneous lupus erythematosus (CLE) is an autoimmune skin disorder that can be seen both in conjunction with systemic lupus erythematosus (SLE) or independently. An association between smoking and CLE has been established, with patients who smoke experiencing increased disease prevalence, a worse disease course, and greater refractoriness to treatments such as antimalarials and immunosuppressives. The molecular underpinnings that underly these clinical differences are minimally explored. In this study we sought to characterize molecular differences and drivers of worse disease between CLE smokers and nonsmokers using AMP AIM single cell RNA-sequencing. Lesional and non-lesional scalp biopsies were taken from patients with discoid lupus erythematosus. Five patients were never smokers, and three patients were current smokers. Between lesional skin of smokers and nonsmokers, keratinocytes, fibroblasts, endothelial cells, follicle cells, myeloid cells, and T-cells exhibited the most differentially expressed genes (DEGs). Further Reactome analysis of lesional DEGs revealed that smokers exhibited upregulation in interferon signaling and related pathways across all examined cell types. Further examination of transcription factors showed that IRF-1 and IRF-8 were consistently upregulated in multiple cell types in smokers, while TP63 was downregulated. These transcription factors may play a role in driving clinical differences. Interestingly, nonlesional skin in smokers also showed elevated IFN related gene signature compared to nonlesional nonsmoker CLE patient

biopsies, suggesting that nonlesional skin in smokers with CLE may be comparatively "primed" compared to nonsmokers. Our data suggests that worse disease course in CLE smokers may be driven by smoking specific interactions that drive increased type I interferon responsiveness or signature.

Category: Lupus

TOXIC EPIDERMAL NECROLYSIS (TEN)-LIKE LUPUS OR TEN IN LUPUS: A CASE SERIES

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Background: Toxic Epidermal Necrolysis (TEN)-like lupus is a rare manifestation of lupus erythematosus (LE) characterized by widespread sloughing of the epidermal layer and mucosal ulceration.[1,2,3] Due to the rarity of the condition, little is known about disease triggers and differences in treatment outcomes. This study aims to identify characteristics of TEN-like lupus, evaluate their clinical course, investigate clinical history and potential inciting factors, and increase literature representations of darker skin types.
Methods: We identified patients 18 years old or older with TEN-like lupus seen by UNC Dermatology during UNC Health Medical Center hospitalization from 2013 to 2023 using the North Carolina Translational and Clinical Sciences Institute's i2b2 web application and Carolina Data Warehouse for Health. Several additional patients were found through additional chart review, yielding a total of ten patients.
Results: 80% of identified patients were women and 80% were African American (AA). Histology frequently showed interface dermatitis and labs typically showed high antinuclear titers, pancytopenia, and hypocomplementemia. Common identified inciting factors included medication exposure and immunosuppressive medication holiday. Treatments included hydroxychloroquine, steroids, IVIG, anakinra, IV cyclophosphamide, and mycophenolate mofetil.
Conclusion: The majority of patients in our study were AA, and we suspect that darker skin types are underrepresented in the current literature. Medications, including antibiotics, antiseizure, and NSAIDs, were a common trigger for TEN-like lupus in our study and has also been reported in prior literature.[4] Early identification of TEN-like lupus is important because treatment path may differ slightly from that of TEN, including the use of antimalarials and other immunosuppressants. In our case series, the most significant improvements resulted from treatment with steroids and IVIG.

Category: Lupus

Teaching Point: Medications and immunosuppression holiday may be common inciting factors for TEN-like lupus. Promising treatment options may include steroids and IVIG, though prospective studies are needed to identify optimal treatments.

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INVESTIGATING CXCR6 AND ITS LIGAND CXCL16 IN LUPUS: CLINICAL AND TRANSLATIONAL SIGNIFICANCE FOR SKIN AND SYSTEMIC ORGAN MANIFESTATIONS

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Cutaneous lupus erythematosus (CLE) is an autoimmune skin disorder that can occur independently or with systemic lupus erythematosus (SLE). Chemokines, particularly the CXCR6–CXCL16 axis, play a critical role in immune cell recruitment to inflammatory sites, with evidence linking them to lupus skin and kidney involvement. CXCL16, produced by keratinocytes and induced by TLR7 activation and UV light, is associated with photosensitivity and cutaneous symptoms such as alopecia and malar rash. Elevated CXCL16 has been reported in juvenile SLE and correlates with nephritis and Th1/Th2 imbalance. CXCR6, expressed on tissue-resident memory T cells (Trm), may contribute to chronic skin inflammation. We performed a systematic review that identified 11 primary studies assessing CXCL16/CXCR6 in lupus. Most human cohort studies found elevated CXCL16 in serum, urine, or cerebrospinal fluid of active disease, particularly lupus nephritis (LN). Urinary CXCL16 was a non-invasive marker of LN severity in multiple studies, while serum CXCL16 correlated with both renal and skin manifestations. CXCR6 enrichment was noted in lupus skin across human, mouse, and canine datasets. Meta-analysis of archival CLE biopsies showed CXCR6 upregulation with high diagnostic sensitivity and specificity, while CXCL16 elevation was species- and context-dependent, possibly influenced by UV exposure or treatment. Functionally, the CXCR6/CXCL16 axis recruits $CXCR6^{+}$ effector T cells to inflamed tissues, activating pathways (PI3K/AKT, MAPK/ERK, NF-κB) that promote survival, migration, and cytokine production. This supports its role in sustaining tissue-specific inflammation in lupus. Limitations include variability in study designs, incomplete demographic data, and lack of standardized measurement methods. Overall, evidence supports CXCL16 as a biomarker of lupus nephritis and systemic activity, and CXCR6 as a marker of cutaneous lupus. The conserved expression across species and mechanistic links to Trm biology suggest this axis as a promising diagnostic and therapeutic target for lupus skin and kidney disease.

Category: Lupus

LONG-TERM CARDIOVASCULAR AND MALINGNANT SEQUELAE OF PEDIATRIC-ONSET SYSTEMIC LUPUS ERYTHEMATOUS: A PROPENSITY-MATCHED INPATIENT ANALYSIS

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Abstract

Pediatric-onset systemic lupus erythematosus (pSLE) emerges during critical growth windows and is characterized by greater cumulative disease burden, early high-dose glucocorticoid exposure, and persistent type 1 interferon-driven endothelial activation. Together, these features predispose to prothrombotic, cardiomyopathy, and potentially oncogenic milieu extending from adolescence into adulthood. Using national multi-institutional records, we conducted a retrospective investigation using 1:1 propensity matched cohorts with a caliper of 0.1 (pSLE n=4,132; non-lupus controls n=4,133), adjusting for age, sex, race, and ethnicity. This study aimed to examine the effect of pSLE (age $\leq$ 18 years, ICD-10-CM: M32) on key thrombotic, cardiovascular, and oncogenic outcomes. Logistic regression models were built to calculate risk difference and Wald-based 95% confidence intervals (CI), with Bonferroni correction was applied to account for multiple comparisons. pSLE was associated with increased risk of thrombophlebitis (RD 1.435%, 95% CI 1.012-1.858, p<0.0001), pulmonary embolism (0.438%, 0.236-0.640, p<0.0001), cardiomyopathy (0.511%, 0.207-0.815, p=0.0015), lymphoid/hematopoietic cancers composite (0.414%, 0.218-0.610, p<0.0001), and multi solid-organ cancers composite (RD 0.342%, 0.064-0.619, p=0.0157). All-cause mortality was also increased (0.484%, 0.185-0.783, p=0.0015). Collectively these findings demonstrate the high-risk state of pSLE, namely for venous thrombosis, pulmonary embolism, cardiomyopathy, lymphoid/hematopoietic malignancy, solid-organ malignancy and mortality. Across these domains, proactive risk assessment, prevention, and longitudinal monitoring is warranted and further interventional studies examining the effects of early hydroxychloroquine use to attenuate long-term harm.

Abstract Category: Lupus

UNILATERAL SCARRING ALOPECIA WITH CUTIS VERTICIS GYRATA-LIKE CHANGES IN PRIMARY SJOGRENS SYNDROME

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A 40-year-old woman was referred to the dermatology department for hair loss, which began two years prior. Her past medical history included syringomyelia, scoliosis, ovarian cancer treated with chemotherapy, and migraines. On examination, she had scarring alopecia localized to the right scalp with underlying thickening of the scalp with prominent grooves running from anterior to posterior resembling cutis verticis gyrata (CVG) (Fig 1). CVG is characterized by redundant scalp folds with deep furrows and convolutions, typically oriented in the anterior–posterior direction. Primary CVG usually presents in men before the age of 30, is symmetric, and is often associated with developmental delays.[1] In contrast, secondary CVG tends to be asymmetric and arises from structural changes within the scalp due to inflammatory, neoplastic, or endocrine disorders. Reported causes of secondary CVG include pachydermoperiostosis, acromegaly, amyloidosis, scleromyxedema, and other inflammatory dermatoses.[2,3,4,5] A scalp punch biopsy demonstrated lymphocyte-predominant scarring alopecia with favoring a diagnosis of Central Centrifugal Cicatricial Alopecia (CCCA) vs Discoid Lupus. Laboratory studies were notable for ANA >1:1280 and anti-SSA antibody >8.0 U/mL. Based on clinicopathologic correlation, a diagnosis of unilateral CCCA with CVG-like changes was made. The patient was simultaneously diagnosed with primary Sjogren's by rheumatology. Studies have shown that CCCA patients had a higher prevalence of ANA positivity and systemic lupus erythematosus, supporting potential autoimmune involvement.[6]

Teaching Point: This case highlights a rare presentation of CCCA as unilateral scarring alopecia with CVG-like changes, in the setting of a new diagnosis of Sjogren's Syndrome. A thorough history and examination should be performed in patients presenting with CCCA to rule out associated autoimmune diseases.

Category: Lupus or Clinical Case

Figure:

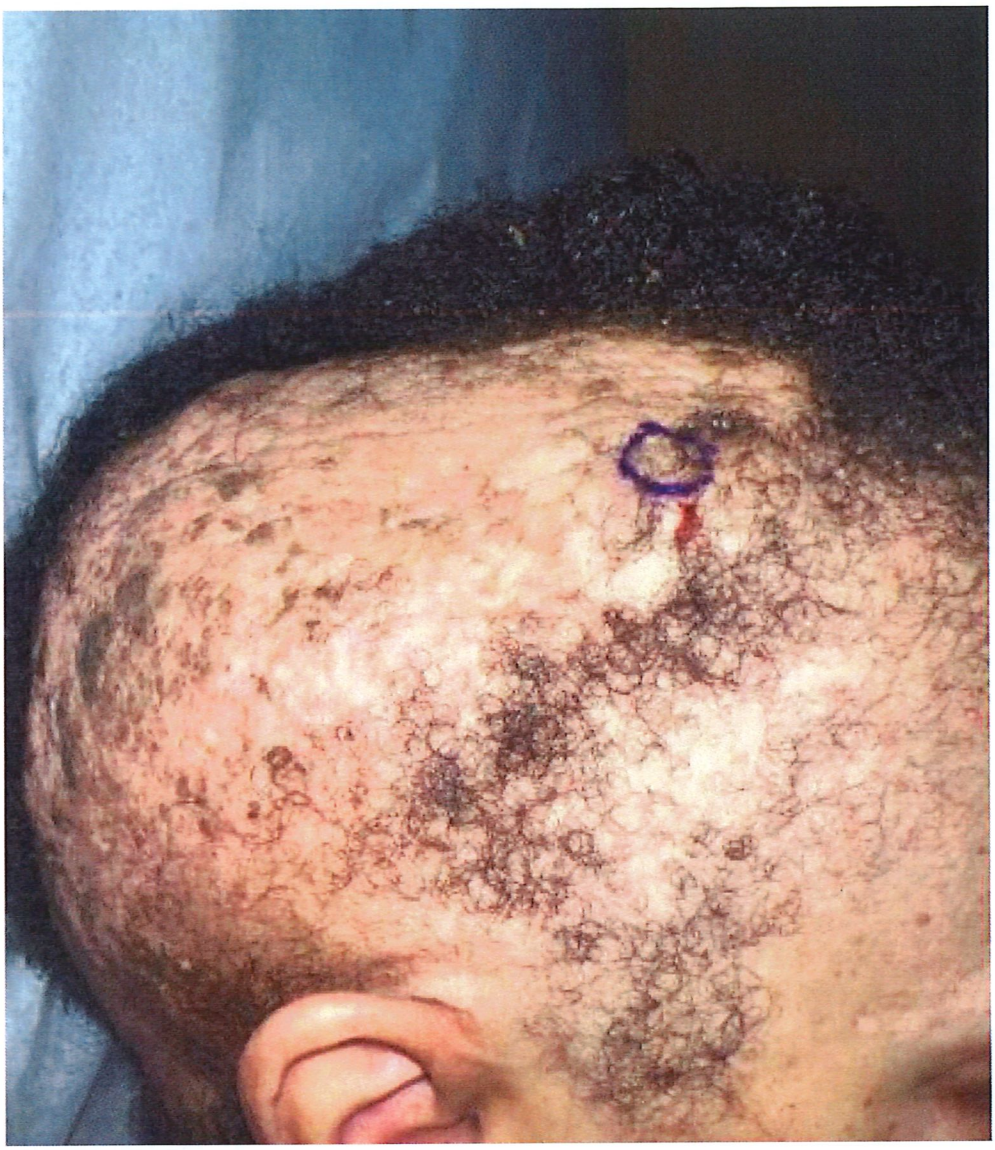

Fig 1. Right scalp scarring alopecia with cutis verticis gyrata-like changes.

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Beyond the Skin: Bridging the Care Gap with Integrated Behavioral Health in a Multidisciplinary Lupus Clinic

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Abstract

Background: Systemic lupus erythematosus (SLE) is frequently accompanied by mental health comorbidities, which are often undertreated due to significant patient and system-level barriers. This study aimed to address this critical gap by integrating behavioral health services directly into our existing multidisciplinary lupus clinic. Methods: Over a two-year period, patients were screened using PHQ-8 and GAD-7. A score of 10 or greater for either survey was deemed a positive screen. The patients with positive screens were initially offered a referral to psychiatry vs referral back to PCP for management. This workflow was subsequently changed to an integrated behavioral health (IBH) clinician embedded in the lupus clinic to increase access. A retrospective chart review evaluated the prevalence of mental health needs and the outcomes of this intervention. Results: Of the 181 patients screened, 98 (54.1%) met the criteria for further evaluation due to GAD-7 or PHQ-8 score ≥10. Fifty-two patients (29%) had a GAD-7 score ≥10, and 82 patients (45%) had a PHQ-8 score ≥10. Mean age was 44.9 years and 93.9% were females. Patients self-identified as Black (39.80%), Other (32.65%), White (37%), and Asian (9.2%). Primary diagnosis was SLE (84.7%) followed by discoid lupus erythematosus (8.2%) and mixed connective tissue disease (4.1%). Most patients had Medicare/Medicaid (55.1%) followed by Commercial (32.6%) and 8.2% were uninsured. Mean referral scores were PHQ-8 14.29 and GAD-7 11.26. 63.27% (n=62) of positive-screened patients accepted a behavioral health referral, and 39.8% completed at least one visit. Initial mental health diagnoses included adjustment disorder, depression, and anxiety. The majority chose to follow up with our IBH clinician (66.4%); whereas 11.2% were referred to Psychiatry for additional management. 38% of patients were already on psychopharmacological treatment managed by a psychiatrist or PCP.Conclusion: This screening intervention identified a 54% prevalence of mental health needs in the lupus patients seen at a safety-net hospital. This intervention led to the successful

integration of a behavioral health clinician into the multidisciplinary lupus clinic, significantly improving access and engagement with essential behavioral health services.

Teaching Point: For patients with lupus, there is a substantial, often hidden, mental health burden and a significant need for improving access to behavioral health services.

Category: Lupus, Mental Health

Keywords: Lupus, Mental Health, Depression, Anxiety, Quality Improvement (QI), Integrated Behavioral Health, Multidisciplinary Clinic, PHQ-8, GAD-7, Comorbidity, Health Services Access.

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Lupus Vaccination Passport: A Quality Improvement Initiative for Optimizing Vaccine Uptake in a Multidisciplinary Lupus Clinic

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Abstract: Systemic lupus erythematosus (SLE) patients are frequently treated with immunosuppressive therapies thus creating inherent infection risk. Hospitalizations for patients with lupus due to infections, particularly sepsis, have risen over time, contributing to increased infection-related mortality. Despite this susceptibility, nationally vaccination rates remain suboptimal. This quality improvement initiative aimed to increase pneumococcal and shingles vaccination rates by 50% over two years among eligible lupus patients in a multidisciplinary clinic. Phase 1 was a retrospective chart review of 400 lupus patients from our lupus registry, revealing baseline pneumococcal vaccination rate of 24% and shingles of 7%. Phase 2 implemented a physician-led, multidisciplinary intervention using an electronic medical record (EMR) tool, "Lupus Vaccination Passport", to identify patients with vaccination indications. Eligible immunosuppressed lupus patients were identified prior to their office visit and offered pneumococcal and shingles vaccines during their visit. Prior authorizations for shingles vaccine were obtained prior to the visit as needed. Nurses administered in-clinic pneumococcal vaccine, while patients obtained the shingles vaccination at the hospital or local pharmacy. Vaccination data was obtained from the Massachusetts Immunization Information System. Data analysis was performed using STATA. Post-intervention, pneumococcal vaccination increased to 57.1% ($p<0.001$) and shingles vaccination (any dose) to 33.8% ($p<0.001$), with 21.93% completing the series. No significant differences were observed by race or gender. Pneumococcal vaccination rates increased in all age groups, but the highest improvement was observed in patients aged 40-49 (48.7%), with a similar trend being noted for shingles series completion. The "Lupus Vaccination Passport" intervention within a multidisciplinary lupus clinic involved support from EMR IT, pharmacist, nursing, and patient navigator in addition to adoption and implementation by clinicians. This intervention significantly improved vaccination rates, demonstrating a successful model for enhancing preventative health maintenance in a vulnerable immunocompromised population.

Teaching Point: This multidisciplinary intervention led to improving vaccine uptake in a multidisciplinary lupus clinic.

Category: Lupus

Keywords: Systemic Lupus Erythematosus (SLE); Immunocompromised host; Vaccination rates; Quality Improvement (QI); Multidisciplinary care; Pneumococcal vaccine; Herpes zoster vaccine; Electronic Medical Record (EMR); Health maintenance

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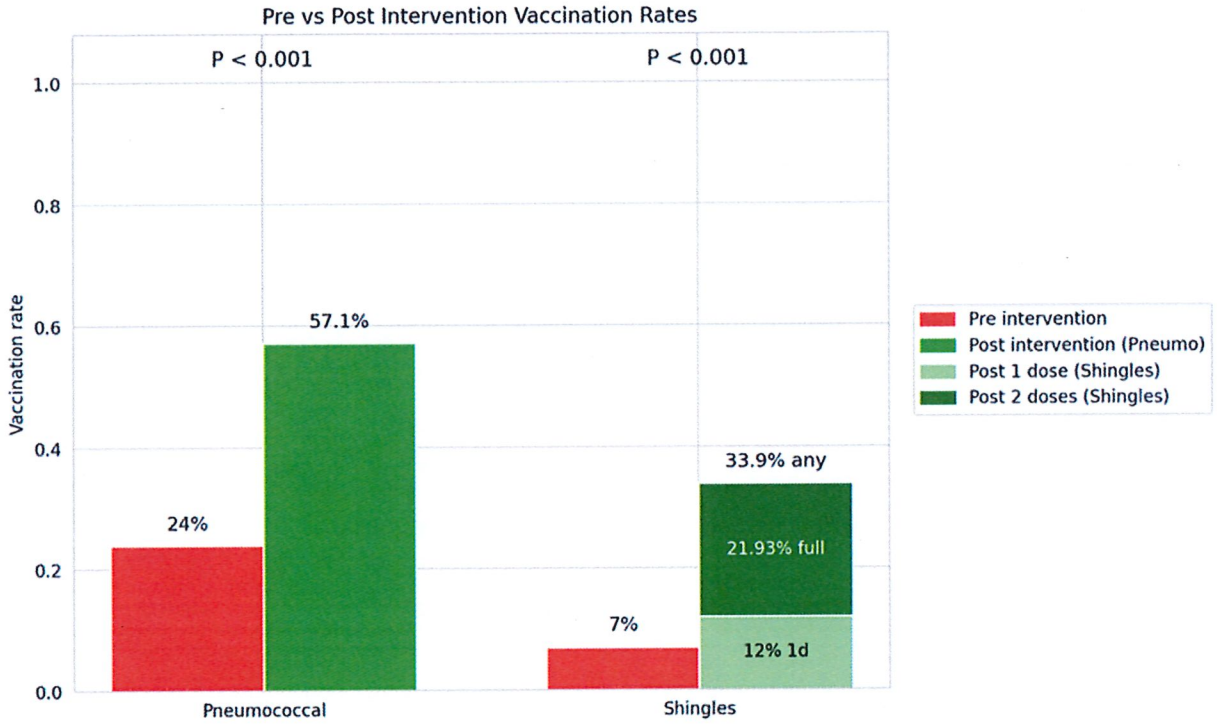
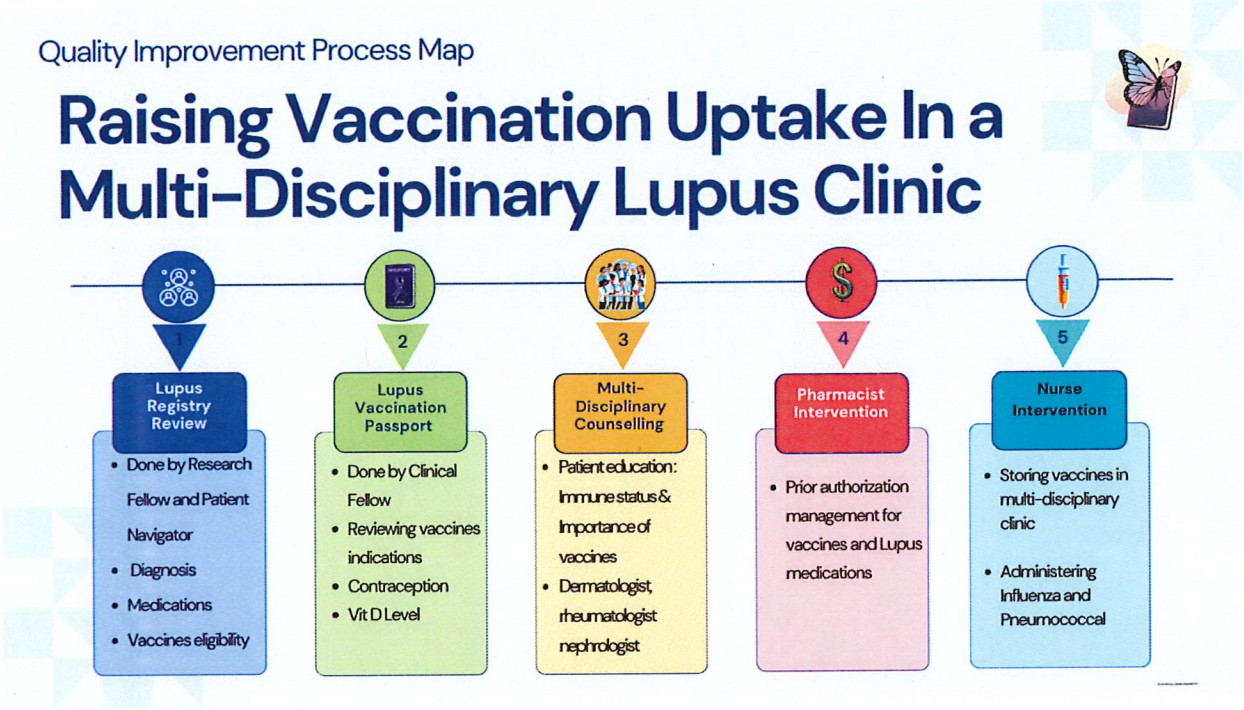
Appendix with Graphs:

Graph 1: Pre vs Post Intervention Vaccination Rates Table



Graph 2: Quality Improvement Process Map



The Cutaneous Lupus Erythematosus Quality of Life Index (CLEQoL) is a multicenter validated and reliable patient-reported outcome measure amongst patients with CLE

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The Cutaneous Lupus Erythematosus Quality of Life Index (CLEQoL) is a patient-reported outcome measure (PROM), which grades quality–of life impacts of cutaneous lupus erythematosus (CLE) amongst five domains: functioning, emotions, symptoms, body image, and photosensitivity. It was validated in a single-center cohort study of 101 CLE patients. For further validation, we performed a larger cross-sectional multi-center study to examine the validity and reliability of the CLEQoL amongst patients with CLE. In total, 130 CLE patients were recruited at outpatient dermatology clinics at the University of Texas Southwestern Medical Center, Parkland Health, and the University of Pennsylvania between July 2018 and July 2025. Demographics, clinical characteristics, CLASI scores, CLEQoL scores were collected. McDonald's Omega analysis was conducted to assess internal consistency of the CLEQoL and its domains. Overall, the CLEQoL demonstrated good internal consistency with an Omega total of 0.98 and Omega Hierarchical of 0.74. In total the model represented 0.61 of the common variance with a root mean square error of approximate (RMSEA) equal to 0.089. The functioning, emotions, and symptom domains demonstrated good internal consistency with Omega total >0.9 and Omega hierarchical >0.7. However, the body image and photosensitivity domains did not display a strong correlation to the overall QoL, likely limited due to a small number of factors in each domain. To assess construct validity, a principal factor analysis with oblimin rotation was conducted (RMSEA = 0.078). Overall, the analysis identified 4 key domains (symptoms, emotions, functioning, and a combined sensitivity and visibility domains). These results highlight that the CLEQoL is a valid and reliable PROM amongst patients with CLE at multiple institutions and can be compacted from five to four domains. Overall, our analysis supports the further integration of the CLEQoL amongst clinical practice and studies.

Category: Lupus

ZOSTER RISK AND VACCINATION STATUS IN LUPUS AND DERMATOMYOSITIS PATIENTS ON ANIFROLUMAB: REAL-WORLD DATA FROM A RETROSPECTIVE COHORT STUDY

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Category: lupus, dermatomyositis

Anifrolumab is a type-I interferon receptor antagonist FDA-approved for the treatment of systemic lupus erythematosus (SLE), with preliminary data demonstrating promising results in cutaneous lupus erythematosus (CLE) and dermatomyositis (DM).[1,2] Increased risk of herpes zoster (HZ) in patients with CLE and DM has been previously shown,[3] and anifrolumab is independently associated with an increased HZ risk.[4,5] Despite this, there are currently no guidelines for HZ prevention or prophylaxis amongst those receiving anifrolumab.[6] Our study sought to examine real-world anifrolumab use and HZ risk in CLE and DM patients. We present a cohort of 80 patients treated with anifrolumab at two U.S.-based academic medical centers between January 2022 and April 2025 whose HZ vaccination, prophylaxis, and infectious status were retrospectively collected. Our cohort were 89% female and 56% White, with a mean age of 44 years (SD 14) at the time of anifrolumab initiation. Among this cohort, 40 (50%) never received HZ vaccination, 35 (44%) received at least one HZ vaccine dose prior to initiation of anifrolumab, and 5 (6%) received their first HZ vaccine after initiation. Of the 80 patients, 18 (23%) had a history of HZ prior to anifrolumab, two (11%) of whom were on prophylactic valacyclovir. 51 (64%) received concomitant immunosuppressants during anifrolumab treatment. Three patients (4%) developed HZ, two of whom had a history of it. One had been fully vaccinated prior to initiation, while one received a single dose of recombinant HZ vaccine after initiating anifrolumab. All three were receiving anifrolumab for CLE. One patient was receiving no concomitant immunosuppressants, one was on systemic steroids, and one was receiving both mycophenolate and low dose steroids at the time of infection. Our study contributes additional reassuring real-world data suggesting that even with relatively low rates of vaccination and prophylaxis, the rate of HZ remained low in those treated with anifrolumab.

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CLINICAL PREDICTORS OF QUALITY OF LIFE IN PATIENTS WITH CUTANEOUS LUPUS ERYTHEMATOSUS: A CROSS-SECTIONAL STUDY

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Cutaneous lupus erythematosus (CLE) is a chronic inflammatory skin disease with a substantial quality of life (QoL) burden. The CLE-QoL, a validated disease-specific instrument developed to capture the unique psychosocial and functional impacts of CLE, has been shown to correlate with disease activity and damage indices (CLASI-A and CLASI-D). However, the specific clinical features driving this burden remain poorly defined. We conducted a cross-sectional study of CLE patients seen at a single academic center (Johns Hopkins Hospital, 2023–2025). All patients were enrolled in a prospective database and completed the CLE-QoL at enrollment; CLASI scores and clinical features were collected concurrently. Pearson's correlation tested associations between quantitative variables, t-tests for categorical comparisons, and multivariable linear regression identified independent predictors of QoL. Among 70 patients, most were female (90.0%) and Black (57.1%) with a mean age of 42.2 years (SD 13.9); 64.3% had SLE and 25.7% had lupus nephritis. The majority had chronic CLE (90.0%), with smaller proportions of subacute CLE (5.7%) and acute CLE (4.3%). Mean CLE-QoL was 97.9 (SD 34.4), CLASI-A 8.2 (SD 7.4), and CLASI-D 10.7 (SD 9.8). In univariate analyses, female sex, photosensitivity, scarring alopecia damage (≥3 scalp quadrants), arthritis, and SLE were associated with worse QoL, while hydroxychloroquine responsiveness was associated with improved QoL ($p<0.05$ for all). In multivariable analysis, female sex ($\beta=26.8$, $p=0.028$), scarring alopecia damage ($\beta=21.5$, $p<.001$), photosensitivity ($\beta=16.9$, $p=0.020$), and SLE ($\beta=16.7$, $p=0.028$) remained significant predictors. QoL weakly correlated with CLASI-A ($r=0.306$, $p=0.001$) but not CLASI-D; CLASI-A only correlated with QoL in patients without SLE ($r=0.572$, $p=0.003$), not with SLE ($r=0.196$, $p=0.198$). Overall, female sex, scarring alopecia, photosensitivity, and co-existing SLE independently predicted worse CLE-QoL, and while CLASI-A showed a weak correlation with CLE-QoL overall, this association was notably stronger in patients without SLE.

Category: Lupus

GLUCAGON-LIKE PEPTIDE-1 RECEPTOR AGONIST USE AND CARDIOVASCULAR OUTCOMES IN CUTANEOUS LUPUS ERYTHEMATOSUS WITH AND WITHOUT SYSTEMIC LUPUS ERYTHEMATOSUS

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Patients with cutaneous lupus erythematosus (CLE), particularly with systemic lupus erythematosus (SLE), have increased cardiovascular risk. Glucagon-like peptide-1 receptor agonists (GLP1-RAs) improve cardiovascular outcomes in diabetes and exhibit anti-inflammatory effects in immune-mediated skin diseases, but their impact in patients with CLE remains unclear. We aimed to assess the association of GLP1-RA use with five-year cardiovascular outcomes and mortality in patients with CLE and CLE + SLE. We conducted a retrospective cohort study using TriNetX to evaluate cardiovascular outcomes with GLP1-RA use in adults with CLE and type 2 diabetes mellitus (T2DM), with or without SLE. Patients required ≥2 CLE or SLE diagnoses ≥6 months apart. GLP1-RA users (≥2 prescriptions) and non-users were propensity matched (1:1) separately for CLE and CLE + SLE cohorts based on demographics, cardiometabolic comorbidities, lupus medications, and antidiabetic therapies. Lupus nephritis was included in matching for SLE cohorts as a proxy for disease severity. The index date was the first GLP1-RA prescription for users and a pseudo-index date for non-users, both 1–3 years after CLE diagnosis. Outcomes included incident cardiovascular events and mortality within five years. Matched cohorts included 462 CLE and 903 CLE + SLE pairs. In isolated CLE, GLP1-RA use was associated with reduced risk of major adverse cardiovascular events (MACE) (RR 0.57, 95% CI 0.35–0.94, p=0.024) and death (RR 0.42, 95% CI 0.23–0.78, p=0.004). Among CLE + SLE patients, GLP1-RA use was linked to reduced risk of atherosclerosis (RR 0.63, 95% CI 0.44–0.89, p=0.009), myocardial infarction (RR 0.60, 95% CI 0.39–0.91, p=0.016), ischemic heart disease (RR 0.74, 95% CI 0.57–0.97, p=0.029), cerebral infarction (RR 0.39, 95% CI 0.23–0.64, p<0.001), MACE (RR 0.39, 95% CI 0.29–0.52, p<0.001), and mortality (RR 0.28, 95% CI 0.19–0.41, p<0.001). GLP1-RA use is linked to lower MACE and mortality in CLE, with greater benefits in CLE + SLE.

Category: Lupus

CARDIOVASCULAR COMORBIDITIES OF CUTANEOUS LUPUS ERYTHEMATOSUS: A NESTED CASE-CONTROL STUDY OF THE ALL OF US RESEARCH PROGRAM

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Cutaneous lupus erythematosus (CLE) is an autoimmune disease marked by skin inflammation and may increase cardiovascular disease (CVD) risk, potentially through chronic pro-inflammatory cytokine elevation. Previous studies evaluating CVD in CLE have been racially homogenous, lacked US-specific data, and did not assess CVD subtypes. To address this, we evaluated CVD risk in US-based CLE patients using data from the NIH All of Us Research Program (Curated Data Repository v8). We conducted a case-control study of CLE patients without systemic lupus erythematosus (SLE), matched 1:40 to non-CLE/SLE controls by age, sex, and race/ethnicity using nearest-neighbor propensity scores. SLE-only and CLE with SLE patients were included as comparator groups. Patients were identified using SNOMED codes. Logistic regression estimated odds ratios (ORs) and 95% confidence intervals (CI) for CVD outcomes. Multivariable models adjusted for age, sex, race/ethnicity, income, and obesity. CLE patients had significantly increased odds of chronic kidney disease (CKD, OR;95%CI, 5.91;2.66-13.13), congestive heart failure (CHF, 11.64;3.38-40.09), coronary artery disease (CAD, 3.53;1.21-10.30), hyperlipidemia (HLD, 3.82;2.44-6.00), hypertension (HTN, 3.16;2.02-4.93), stroke/transient ischemic attack (stroke/TIA, 4.34;1.24-15.21]), type II diabetes (T2DM, 3.51;1.71-7.22), valvular stenosis/regurgitation (VSR, 5.35;2.15-13.28), and venous thromboembolism (VTE, 13.15;5.04-34.29). SLE-only patients showed smaller increases in CKD (1.56;1.25-1.95), CHF (1.80;1.17-2.75), stroke/TIA (1.51;1.10-2.07), and VTE (1.81;1.29-2.54). CLE with SLE showed moderate increases across most outcomes: atrial fibrillation (AFib, 1.50;1.19-1.89), CKD (1.62;1.37-1.93), CHF (2.26;1.70-3.01]), CAD (1.52;1.25-1.85), HLD (1.21;1.09-1.33), HTN (1.32;1.21-1.44), stroke/TIA (1.85;1.48-2.31), VSR (1.61;1.33-1.95), and VTE (2.15;1.70-2.71). All associations remained significant after adjustment. Additionally, SLE became associated with HTN (1.13;1.02-1.26), and CLE with SLE with myocardial infarction (MI, 1.50;1.10-2.05). Notably, for every shared CVD outcome, CLE-alone patients had

numerically higher ORs compared to those with both CLE and SLE. This trend points to CLE-specific inflammation as a potential contributor to elevated CVD risk, highlighting the need for targeted CVD screening in CLE, independent of SLE diagnosis.

Category: Lupus

DECREASED ASSOCIATION OF AUTOANTIBODIES WITH SMOKING IN PATIENTS WITH CLE

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Smoking has a known association with severity of cutaneous lupus (CLE), with higher CLASI scores and more refractory disease. It is suggested that smokers with CLE have increased levels of type I interferon in the skin, though it is unclear and never reported whether smoking is linked to differences in autoantibody production. In a population of 415 patients at a single site with CLE that was not drug-induced, 80% were female (n=335) and 38% were black (n=154). 237 (57%) patients had discoid LE, 89 (21%) had subacute CLE, 40 (10%) had acute CLE, 49 (12%) had other chronic CLE. 211 (48%) were never smokers, 111 (25%) were former smokers, and 85 (19%) were current smokers. 230 out of 385 (60%) antinuclear antigen tests were positive, 103/308 (33%) double-stranded DNA, 66/205 (32%) anti-Smith, 84/188 (44%) anti-ribonucleoprotein, 120/303 (40%) Ro/SSA, 30/285 (10%) La/SSB, 17/75 (23%) Russell's viper venom, 42/162 (26%) anti-cardiolipin, and 17/118 (14%) beta2-glycoprotein1 tests were positive. Chi-squared test with logistic regression and post hoc test with Tukey correction showed a significantly higher percentage of positive ANA and dsDNA tests in never smokers compared to former smokers and current smokers. There were 129 positive ANA in never smokers compared to 35 in former smokers ($p=0.020$; OR = 0.47 (95% CI: 0.27-0.81)) and 48 in current smokers ($p=0.045$; OR = 0.54 (95% CI: 0.32-0.89)). There were 65 positive dsDNA in never smokers compared to 11 in former smokers ($p=0.025$; OR = 0.37 (95% CI: 0.17-0.76)) and 13 in current smokers ($p=0.003$; OR = 0.32 (95% CI: 0.16-0.62)). Other associations between smoking status and other autoantibodies were statistically insignificant. Identifying the decreased association of autoantibodies with smoking will help inform the biological mechanism by which smoking impacts CLE disease and tailor individualized patient care.

Category: Lupus

ALL SUBCOMPONENTS OF THE CUTANEOUS LUPUS ERYTHEMATOSUS DISEASE AREA AND SEVERITY INDEX–ACTIVITY (CLASI-A) ARE RELEVANT TO IDENTIFY AND DETECT CHANGES IN SKIN ACTIVITY

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Word count body: max 300 words (300/300)

Part B of the Phase 2 LILAC study (NCT02847598) demonstrated litifilimab efficacy versus placebo, with significant decrease in percent change from baseline in CLASI-A score at Week 16 in active cutaneous lupus erythematosus (CLE) with or without systemic manifestations.[1] The CLASI-A measures CLE disease activity across several anatomical locations based on five subcomponents: Erythema, Scale/Hypertrophy, Mucous Membrane Lesions, Recent Hair Loss, and Non-scarring Alopecia. This exploratory analysis examined the contribution of all subcomponents or anatomical locations in total scoring change and explored association between sunlight-exposed body areas and symptom severity. Study design and participants' baseline characteristics for LILAC Part B were previously reported.[1] CLASI-A subcomponent distribution for the pooled Part B population (all litifilimab doses and placebo; N=132) was analyzed by anatomical location at baseline and Week 16. Changes in subcomponent scores at Week 16 were evaluated using point improvement/worsening from baseline at each location. The data are reported as observed, without imputation of missing data. At baseline, more participants had 'red' (score 2) and 'dark red' (score 3) erythema in more sunlight-exposed areas (frontal V-neck area, ears, arms, nose, rest of face) than in less-exposed areas (feet, legs, abdomen) (**Table 1**). At Week 16, changes in CLASI-A subcomponents scores were observed at all locations, with the greatest improvements in more sunlight-exposed areas (**Table 2**). One- and two-point improvements in erythema scores were reported at all locations in up to 33.3% and 12.4% of participants per location, respectively. Three-point improvements occurred at nearly all locations, in up to 2.9% of participants per location. Similar findings were observed for scale/hypertrophy. No single subcomponent drives the CLASI-A score or changes. CLASI-A effectively captures overall CLE skin activity and changes in highly visible, photosensitive areas, supporting the relevance of all five subcomponents.
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First presented: LUPUS 2025; encored: EADV, APLAR, JDA 2025.

Abstract Category: Lupus

Table 1: Distribution of CLASI-A subcomponent scores for each anatomical location at baseline

Anatomical location, n (%)	Erythema (score)				Scale/hypertrophy (score)		
	Absent (0)	Pink (1)	Red (2)	Dark red (3)	Absent (0)	Scale (1)	Verrucous/ hypertrophic (2)
Rest of the face	33 (25.0)	32 (24.2)	42 (31.8)	25 (18.9)	64 (48.5)	54 (40.9)	14 (10.6)
Nose (including malar area)	48 (36.4)	27 (20.5)	45 (34.1)	12 (9.1)	85 (64.4)	37 (28.0)	10 (7.6)
Arms	53 (40.2)	29 (22.0)	35 (26.5)	15 (11.4)	79 (59.8)	42 (31.8)	11 (8.3)
Ears	54 (40.9)	35 (26.5)	33 (25.0)	10 (7.6)	73 (55.3)	51 (38.6)	8 (6.1)
Front V-neck area	67 (50.8)	29 (22.0)	33 (25.0)	3 (2.3)	101 (76.5)	30 (22.7)	1 (0.8)
Hands	80 (60.6)	24 (18.2)	22 (16.7)	6 (4.5)	101 (76.5)	24 (18.2)	7 (5.3)
Posterior neck and/or shoulders	81 (61.4)	18 (13.6)	24 (18.2)	9 (6.8)	98 (74.2)	32 (24.2)	2 (1.5)
Back, buttocks	85 (64.4)	15 (11.4)	19 (14.4)	13 (9.8)	99 (75.0)	27 (20.5)	6 (4.5)
Chest	86 (65.2)	16 (12.1)	24 (18.2)	6 (4.5)	110 (83.3)	21 (15.9)	1 (0.8)
Legs	108 (81.8)	11 (8.3)	8 (6.1)	5 (3.8)	113 (85.6)	16 (12.1)	3 (2.3)
Abdomen	118 (89.4)	5 (3.8)	8 (6.1)	1 (0.8)	121 (91.7)	11 (8.3)	0
Feet	119 (90.2)	5 (3.8)	6 (4.5)	2 (1.5)	122 (92.4)	9 (6.8)	1 (0.8)

	Mucous membrane lesions (score)		Recent hair loss (preceding 30 days) (score)		Alopecia (score)			
	Absent (0)	Lesion/ ulceration (1)	No (0)	Yes (1)	Absent (0)	Diffuse (1)	Focal or patchy in one quadrant (2)	Focal or patchy in more than one quadrant (3)
n (%	105 (79.5)	27 (20.5)	62 (47.0)	70 (53.0)	46 (34.8)	26 (19.7)	9 (6.8)	51 (38.6)

Participants from all treatment arms in LILAC Part B are included (N = 132). Results are based on observed data; no imputation for missing data was conducted for this analysis.

Table 2: Distribution of change from baseline in CLASI-A subcomponent scores for each anatomical location at Week 16

Anatomical location, n (%)	Erythema (score)							Scale/hypertrophy (score)				
	−3	−2	−1	0	+1	+2	+3	−2	−1	0	+1	+2
Rest of the face	2 (1.9)	13 (12.4)	35 (33.3)	50 (47.6)	3 (2.9)	1 (1.0)	1 (1.0)	3 (2.9)	26 (24.8)	69 (65.7)	7 (6.7)	0
Nose (including malar area)	1 (1.0)	12 (11.4)	35 (33.3)	52 (49.5)	5 (4.8)	0	0	3 (2.9)	23 (21.9)	77 (73.3)	2 (1.9)	0
Arms	3 (2.9)	11 (10.5)	30 (28.6)	55 (52.4)	4 (3.8)	2 (1.9)	0	1 (1.0)	17 (16.2)	84 (80.0)	3 (2.9)	0
Ears	1 (1.0)	12 (11.4)	29 (27.6)	58 (55.2)	5 (4.8)	0	0	1 (1.0)	26 (24.8)	73 (69.5)	5 (4.8)	0
Front V-neck area	0	10 (9.5)	20 (19.0)	71 (67.6)	3 (2.9)	1 (1.0)	0	1 (1.0)	18 (17.1)	84 (80.0)	2 (1.9)	0
Hands	1 (1.0)	5 (4.8)	20 (19.0)	75 (71.4)	3 (2.9)	1 (1.0)	0	2 (1.9)	10 (9.5)	89 (84.8)	4 (3.8)	0
Posterior neck and/or shoulders	1 (1.0)	10 (9.5)	16 (15.2)	71 (67.6)	5 (4.8)	2 (1.9)	0	2 (1.9)	14 (13.3)	82 (78.1)	7 (6.7)	0
Back, buttocks	3 (2.9)	10 (9.5)	14 (13.3)	76 (72.4)	2 (1.9)	0	0	3 (2.9)	10 (9.5)	89 (84.8)	3 (2.9)	0
Chest	2 (1.9)	12 (11.4)	11 (10.5)	76 (72.4)	4 (3.8)	0	0	0	13 (12.4)	90 (85.7)	2 (1.9)	0
Legs	3 (2.9)	3 (2.9)	7 (6.7)	92 (87.6)	0	0	0	0	9 (8.6)	96 (91.4)	0	0
Abdomen	0	4 (3.8)	5 (4.8)	93 (88.6)	3 (2.9)	0	0	0	3 (2.9)	102 (97.1)	0	0
Feet	0	3 (2.9)	4 (3.8)	97 (92.4)	1 (1.0)	0	0	0	5 (4.8)	100 (95.2)	0	0

	Mucous membrane lesions (score)			Recent hair loss (preceding 30 days) (score)			Alopecia (score)						
	−1	0	+1	−1	0	+1	−3	−2	−1	0	+1	+2	+3
n (%)	18 (17.1)	85 (81.0)	2 (1.9)	33 (31.4)	66 (62.9)	6 (5.7)	6 (5.7)	2 (1.9)	11 (10.5)	82 (78.1)	4 (3.8)	0	0

Participants from all treatment arms in LILAC Part B are included (N = 132). Reported changes in CLASI-A subcomponent point scores could fall into the following ranges: erythema (from −3 to +3), scale/hypertrophy (from −2 to +2), mucous membrane lesions and recent hair loss (from −1 to +1), and non-scarring alopecia (from −3 to +3). Results are based on observed data; no imputation for missing data was conducted for this analysis.

References:

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PREVALENCE OF ANTIPHOSPHOLIPID ANTIBODIES IN CUTANEOUS LUPUS ERYTHEMATOSUS WITH AND WITHOUT SYSTEMIC DISEASE

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The prevalence of antiphospholipid antibodies (APLAs) in systemic lupus erythematosus (SLE) has been extensively investigated. However, only a few studies have assessed their prevalence in cutaneous LE (CLE) with discordant findings. This retrospective cross-sectional study analyzed 288 patients in our institutional review board-approved CLE database at the University of Pennsylvania who received APLA testing between January 2007 and June 2025. CLE and SLE were diagnosed according to clinical and pathological criteria. We evaluated the presence of the immunoglobulin G (IgG) isotype of lupus anticoagulant (LA), anticardiolipin antibody (ACA), and anti-β2-glycoprotein I antibody (anti-β2GPI). The APLA positivity rates in CLE patients with and without SLE were compared using chi-square and Fisher's exact tests with $\alpha = 0.05$. One or more of the APLAs were found in 9% (10/116) of the patients with CLE only and in 25% (43/172) of those with both CLE and SLE, $p = 0.0004$. LA and ACA were significantly less common in the CLE-only group (LA 4/84 [5%], ACA 5/113 [4%]) than in the CLE/SLE group (LA 23/151 [15%], ACA 27/171 [16%]), $p = 0.02$ and 0.003 respectively. No significant difference in APLA prevalence was found between CLE-only patients with and without antinuclear antibodies (ANA) (5/68 [7%] vs 5/46 [11%], $p = 0.5$). Among the CLE-only patients, 8/10 (80%) of those with APLA(s) had discoid LE (DLE) whereas 68/116 (59%) of the entire group had DLE, $p = 0.3$. Out of the 10 CLE-only patients with APLA(s), 1 had antiphospholipid syndrome with a history of thrombotic events. The prevalence of APLAs was significantly lower in our CLE-only patients compared to our CLE patients with SLE, regardless of the presence of ANA. Therefore, it may be of limited value to routinely screen for APLAs in CLE-only patients in the absence of compelling clinical indications.

Category: Lupus

PRESENTATION OF MIXED CONNECTIVE TISSUE DISEASE AS AMYOPATHIC DERMATOMYOSITIS

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Mixed Connective Tissue Disease (MCTD) is a challenging diagnosis given its overlap with several autoimmune disorders including systemic lupus erythematosus, scleroderma, myositis, and rheumatoid arthritis. The lack of universally accepted diagnostic criteria means clinicians must rely on a combination of clinical features and laboratory tests to make a diagnosis. The presence of anti-U1-RNP antibodies remains a hallmark in MCTD. The U1-RNP complex is an intranuclear protein that converts pre-messenger RNA to mature RNA. Consequently, the proposed etiology involves the interaction of a genetic predisposition with an environmental insult such as infections, drugs, toxins, ultraviolet radiation, or chemicals. It is important for physicians to maintain a high index of suspicion in the presence of non-specific or atypical findings, particularly if involving multiple organ systems. In this case report we discuss a 65 year old male patient who presents to a dermatology clinic with biopsy-confirmed diagnosis of dermatomyositis. Clinically, he exhibited scaling of the distal fingertips, pinpoint eroded papules on the palms and soles, and pink nodules over the interphalangeal joints suggestive of Gottron's papules. He also had a diffuse erythematous rash across his mid-back but lacked other classic findings such as heliotrope rash, shawl sign, or proximal muscle weakness. His medication history was notable only for rosuvastatin, and he had occupational exposure as a contractor and building inspector. He was treated with topical tacrolimus and triamcinolone, along with a low-dose prednisone taper, and was later transitioned to mycophenolate for maintenance. Although MCTD is known to be an overlap of autoimmune disorders, there is evidence that suggests that these conditions can occur sequentially over time. While there is a paucity of data on the targeted management of MCTD, the recommended treatment is based on the specific autoimmune disease at presentation. This case highlights the importance of a comprehensive, multi-disciplinary approach as we work to better understand and improve the diagnostic criteria for MCTD.

Abstract Category: Dermatomyositis and Clinical Case

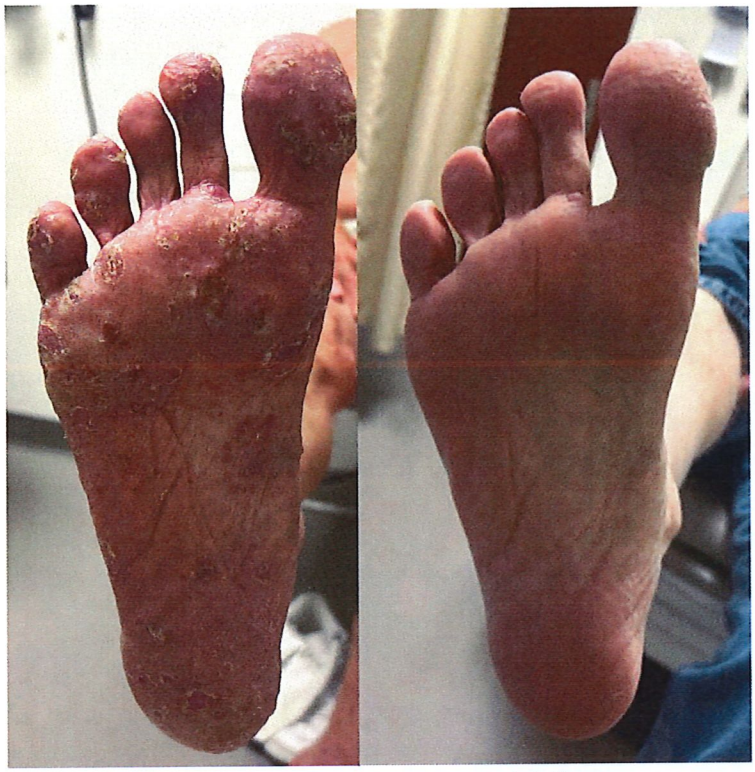

Image 1: Right foot at initial encounter and follow up

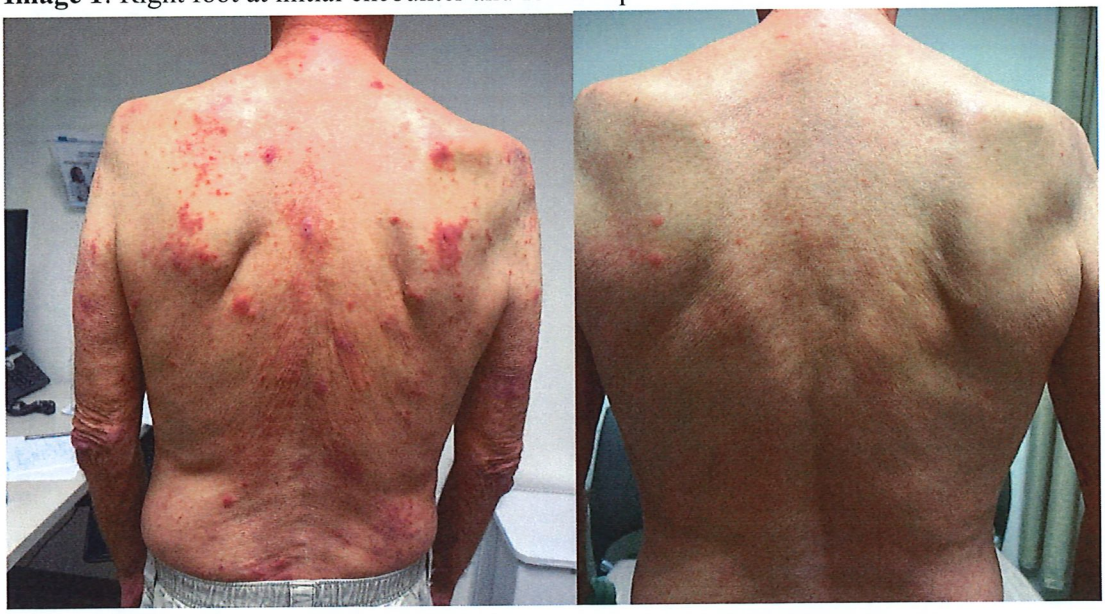

Image 2: Erythematous patches on back at initial encounter and follow up

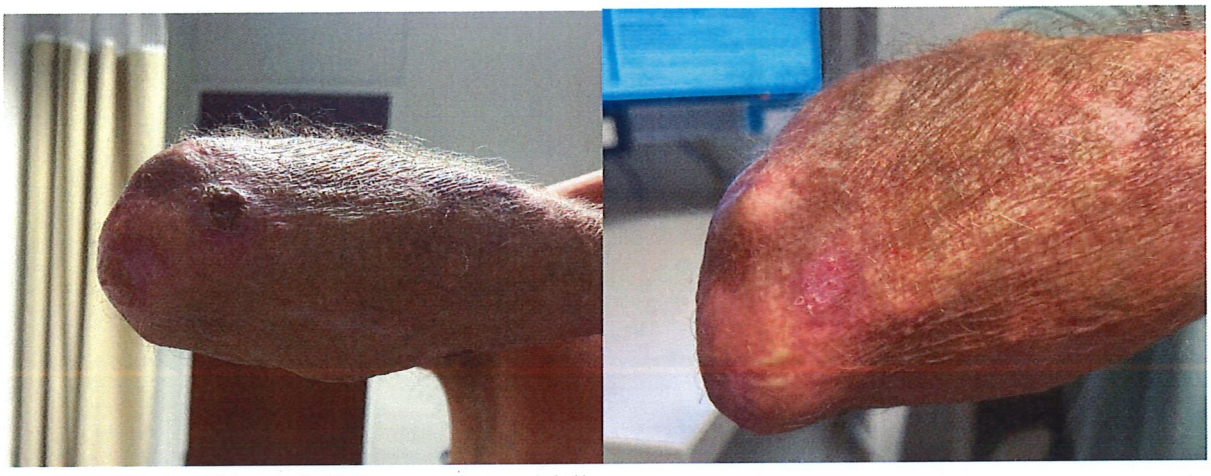

Image 3: Right elbow at initial encounter and follow up

DIAGNOSTIC DELAY IN IDIOPATHIC INFLAMMATORY MYOPATHIES: IMPACT ON DISEASE ACTIVITY, DAMAGE, AND DISPARITIES IN A HISPANIC COHORT

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Idiopathic inflammatory myopathies (IIM) are rare autoimmune disorders characterized by muscle inflammation and extramuscular involvement. Their clinical heterogeneity often leads to diagnostic delays, which are linked to worse outcomes. In individuals with darker skin phototypes, cutaneous signs may be less apparent, further complicating recognition. However, few studies have examined diagnostic delay in Hispanic populations or the impact of skin phototype on disease identification. We conducted a cross-sectional, comparative study of adult IIM patients at a tertiary care hospital in Mexico (Nov 2024–Jul 2025). Patients were classified into IIM subtypes based on established criteria and clinician assessment. Disease activity was assessed using MDAAT (MYOACT + MITAX), and damage using the Myositis Damage Index (MDI). Skin phototype was evaluated using the Fitzpatrick scale. Patients were divided into two groups based on the cohort's median diagnostic delay (≤5 months vs. >5 months). Statistical analyses were conducted using SPSS v27. Among 53 patients (mean age 45.3; 84.6% female), dermatomyositis was the most frequent subtype (62.2%). Median diagnostic delay was 5 months (IQR 3–18). Patients with delay >5 months had significantly higher disease activity (MYOACT: 22.1 vs. 10.1, $p<0.001$; MITAX: 17.7 vs. 9.5, $p=0.005$) and damage (MDI extent: 5.8 vs. 4.2, $p=0.03$). Longer delays were more common in patients with skin phototypes IV–V (80.7% vs. 19.2%, $p<0.05$) and in younger individuals ($\rho = -0.296$, $p = 0.03$). Three patients died, two with significant diagnostic delays and lung involvement. Diagnostic delay >5 months in IIM is associated with higher disease activity, functional impairment, and worse outcomes. Delays were more frequent in younger patients and those with darker skin tones, highlighting potential disparities in recognition. Early diagnosis and increased clinical awareness—especially in underrepresented populations—are crucial to enhancing IIM outcomes.

Abstract category: Dermatomyositis

Table 1. Comparison of clinical and laboratory characteristics according to diagnostic delay time

	Diagnosis <5 months (n=27)	Diagnosis >5 months (n=26)	p-value
Women, n (%)	24 (88.8)	21 (80.7)	0.64
Age, mean (±SD)	50.4 ± 17.1	40 ± 15.3	**0.02**
BMI, kg/m^2, mean (±SD)	26.8 ± 3.8	27.1 ± 5.5	0.84
Prednisone, n (%)	18 (66.6)	23 (88.4)	0.058
Cumulative dose of prednisone, mean (±SD)	3607.0 ± 3725.8	4076.1 ± 4045.8	0.70
Disease duration, months, mean (±SD)	56.9 ± 69.8	52.9 ± 41.0	0.80
SRP, n (%)	5 (18.5)	0 (0)	**0.01**
PL12, n (%)	0 (0)	5 (19.2)	**0.02**
Idiopathic Inflammatory Myopathy			
Dermatomyositis, n (%)	19 (70.3)	14 (53.8)	0.21
Juvenile Dermatomyositis, n (%)	2 (7.4)	1 (3.8)	0.57
Antisynthetase Syndrome, n (%)	3 (11.11)	7 (26.9)	0.14
Polymyositis, n (%)	2 (7.4)	2 (7.6)	0.96
Immune-mediated necrotizing myopathy, n (%)	1 (3.7)	0 (0)	0.32
Amyopathic dermatomyositis, n (%)	0 (0)	2 (7.6)	0.14
Fitzpatrick			
Type II - III, n (%)	13 (48.1)	5 (19.2)	**0.02**
Type IV - V, n (%)	14 (51.8)	21 (80.7)	**0.02**
HAQ, mean (±SD)	0.7 ± 0.6	1.2 ± 0.9	**0.02**
Myositis Damage Index			
Extent of damage, mean (±SD)	4.2 ± 2.6	5.8 ± 2.7	**0.03**
Severity of damage, mean (±SD)	12.9 ± 10.4	18.0 ± 10.0	0.07
Extended damage, median (IQR)	1.0 (0.0 – 1.0)	1.0 (0.2 – 2.0)	0.12
Myositis Disease Activity Assessment Tool			
MYOACT, mean (±SD)	10.1 ± 8.7	22.1 ± 9.7	**<0.001**
MITAX, mean (±SD)	9.5 ± 7.7	17.7 ± 11.9	**0.005**
Laboratories			
CPK, mg/dl, median (IQR)	68 (52.0 – 103.0)	132 (50 – 274)	0.11
Aldolase, mg/dl, mean (±SD)	3.8 ± 0.4	-	-
AST, mg/dl, median (IQR)	24.5 (19.0 – 46.0)	35.0 (22.5 – 50.0)	0.21
ALT, mg/dl, median (IQR)	27.5 (20.5 – 36.7)	30.0 (21.0 – 51.0)	0.44
Albumin, mg/dl mean (±SD)	3.9 ± 0.4	3.8 ± 0.5	0.47
Creatinine, mg/dl, mean (±SD)	0.6 ± 0.1	0.5 ± 0.1	0.09

SD: Standard deviation; HAQ: Health Assessment Questionnaire; MYOACT: Myositis Disease Activity Assessment Visual Analogue Scale; MITAX: Myositis Intention-to-Treat Activity Index; CPK: Creatine Phosphokinase; ALT: Alanine Aminotransferase; AST: Aspartate Aminotransferase.

Thank you very much to all,
Valeria

GENE EXPRESSION COMPARISONS BETWEEN ADULT AND JUVENILE DM SKIN PUNCH BIOPSIES REVEAL POTENTIAL MOLECULAR DISTINCTIONS IN THE DERMAL MICROENVIRONMENT

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Dermatomyositis (DM) is a rare, chronic autoimmune disease that causes inflammation in the muscles and skin. Despite increased prevalence in adults (ADM), children are also susceptible to a similar condition referred to as juvenile DM (JDM). Other than their comparable clinical features and common type I interferon signature, these DM subtypes are relatively distinct as ADM patients are at increased risk for cancer, while JDM patients are more likely to develop complications like calcinosis. We analyzed publicly available bulk RNA-sequencing data of full thickness skin punch biopsies from ADM (n = 7), JDM (n = 4), and healthy control (adult: n = 9; juvenile: n = 5) donors to identify gene signatures in the dermal microenvironment. In this pilot study, differential gene expression (DGE) and Gene Set Enrichment Analysis of Gene Ontology (gseaGO) were performed. Of the top 10 significantly enriched terms for both DM subtypes, only "*negative regulation of viral process*" ($p \leq 0.05$; **Fig. 1A**) was enriched in both ADM and JDM skin. Terms enriched in ADM skin included B-cell and other leukocyte mediated inflammatory processes, while those for JDM focused on antiviral mechanisms, interferon signaling, and keratinization (**Fig. 1A**), highlighting the possibility of distinct pathological mechanisms in these disorders. Notable DEGs in the ADM group were *KIR2DL4*, *CD36*, *THBS1*, and *F5* (**Fig. 1B, 1D**) suggesting and altered wound healing response with enhanced cellular cytotoxicity and resolving inflammation. In JDM samples, increased expression of *CDSN*, *KRT10*, *LORICRIN*, and *SLURP1* (**Fig. 1B, 1D**) were identified, and these genes are overexpressed in other inflammatory skin conditions. Together our findings highlight distinct molecular signatures for ADM and JDM, suggesting potential divergent pathomechanisms. Beyond this pilot, additional studies with larger numbers of patients and controls matched for age, sex, and body site are warranted to fully understand the potential of these findings and direct relevance to patient care.

Category: Dermatomyositis

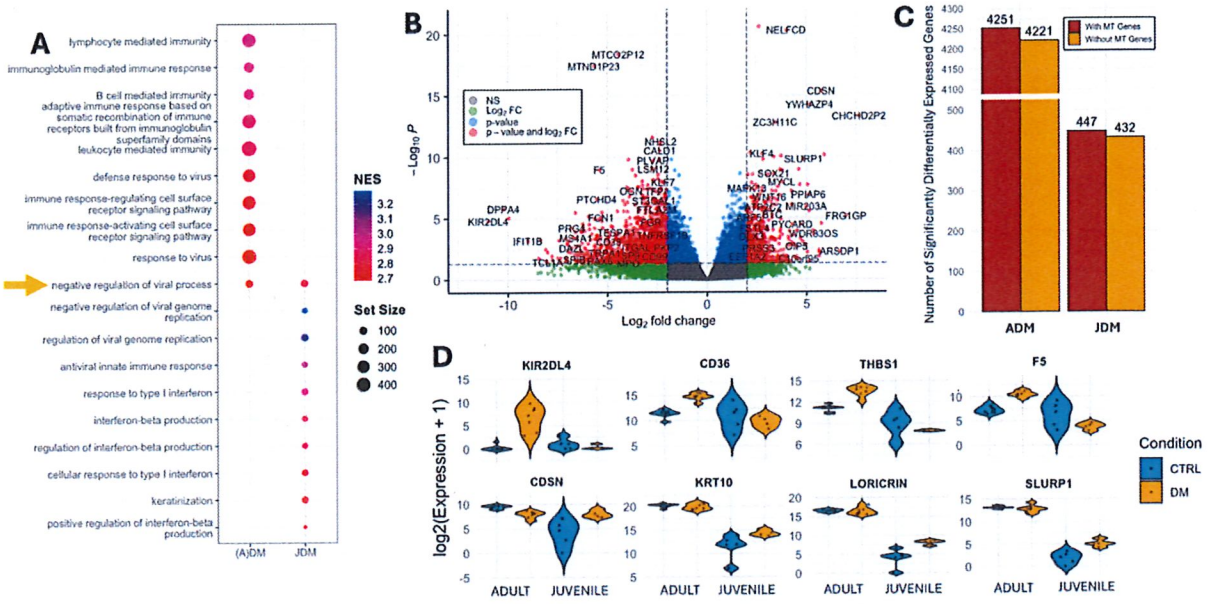

Figure 1. Comparison of DGE between ADM and JDM skin punch biopsies highlights unique underlying molecular signatures. A. Faceted dot plot of the top 10 significantly enriched goGSE terms for both DM subgroups ($p \leq 0.05$). Single overlapping term is indicated by arrow. **B.** Volcano plot of significant DEGs after removal of protein-coding mitochondrial genes (padj ≤ 0.05 and $\log_2FC \geq 2$). Negative $\log_2FC$ indicates stronger effect in ADM samples, while positive $\log_2FC$ indicates stronger effect in JDM samples. **C.** Bar graph of significant DEGs for both ADM and JDM samples before and after removal of protein-coding mitochondrial genes. **D.** Violin plots showing individual log-normalized sample expression values for various top DEGs related to inflammation, wound healing, and keratinization: *KIR2DL4*, *CD36*, *THBS1*, *F5*, *CDSN*, *KRT10*, *LORICRIN*, and *SLURP1*.

THE ASSOCIATION BETWEEN FAMILY HISTORY OF AUTOIMMUNITY, HERBAL SUPPLEMENT USE AND DISEASE ACTIVITY IN DERMATOMYOSITIS

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Herbal supplement usage has been rising in the United States, with 2020 heralding a record-breaking 17.3% increase in sales. In 2024, the prevalence of herbal usage among 673 patients with dermatomyositis (DM) or cutaneous lupus erythematosus was 32%. However, growing evidence suggests that herbal supplements, often immunostimulatory, are harmful to patients with autoimmune conditions. In vitro studies with Spirulina, a common herbal supplement, demonstrated significantly increased activation of inflammatory pathways in the blood of DM patients compared to healthy controls, suggesting that immune cells are predisposed to disease onset or exacerbation following herbal exposure. Genetic factors, potentially associated with positive family history of autoimmunity (FHAI), may contribute to heightened disease activity with herbal usage. To assess the association between positive FHAI, herbal use, and disease activity in DM, we performed a retrospective cross-sectional review and screened 570 patients in the University of Pennsylvania DM database. After excluding 143 patients with missing data, 133 (31.1%) of the remaining 427 reported herbal use, with 66 (49.6%) reporting a positive FHAI. Among the 133 patients with herbal consumption, 19 (14 with positive FHAI) were suspected of DM onset and 26 (14 with positive FHAI) with DM exacerbation associated with herbal usage. Among herbal users, those with disease onset had significantly greater odds of a positive FHAI (Fisher's Exact Test: p=0.027; OR=3.31; 95% CI:1.04-12.54). There was no significant association between positive FHAI and disease exacerbation (Fisher's Exact Test: p=0.67; OR=1.23; 95% CI:0.48-3.22). When disease onset and exacerbation were combined, the group exhibited a borderline but potentially significant association with positive FHAI (Fisher's Exact Test: p=0.042; OR=2.19; 95% CI:0.98-5.01). The findings suggest a trend toward positive FHAI among DM patients whose disease activity is linked to herbal supplement use, highlighting the need to explore genetic determinants of susceptibility to herbal exposures.

Category: Dermatomyositis

DISPARITIES IN DERMATOMYOSITIS MANAGEMENT: A CLOSER LOOK AT RACE, ETHNICITY, GENDER, AND RURAL STATUS

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Abstract:

Dermatomyositis (DM) is a presumed autoimmune inflammatory dermatomyopathy with both cutaneous and systemic manifestations. DM itself has multiple clinical subtypes affecting different demographic groups more than others. This study uses a national database representative of the United States adult population to explore disparities in management of DM based on race, gender, ethnicity, age, and rural status. The All of Us Database was used to query patients with dermatomyositis and dermatomyositis sine myositis (OMOP Codes 80182 and 4081250, respectively) as well as prescriptions of systemic corticosteroids, topical steroids, IVIG, rituximab, azathioprine, methotrexate, and mycophenolate mofetil. Multivariate regression was performed to assess the odds ratio (OR) of treatment with the aforementioned medications based on race, gender, ethnicity, age, and rural status. Age was significantly associated with a decreased odds ratio for treatment with methotrexate (OR=0.98; 95% CI 0.96 - 1.00; p=0.016), azathioprine (OR=0.97; 95% CI 0.94 - 0.99; p=0.014), and IVIG (OR=0.91; 95% CI 0.84 - 0.99; p=0.024). Additionally, females had an increased ratio for treatment with methotrexate (OR=4.17; 95% CI 1.78 - 9.73; p=.00098). There were no differences in treatment patterns across racial, ethnic, or rural/non-rural cohorts. Based on our findings, non-steroidal systemic medications had the largest discrepancy in treatment patterns, primarily among different age cohorts. Specifically, individuals of an older age were less likely to have been prescribed non-steroidal systemic medications. Additionally, women were significant more likely to have been prescribed methotrexate to a clinically significant degree. These patterns require further investigation to elucidate the basis of these discrepancies and – more importantly – determine if these differences had any bearing on outcomes.

Abstract Category: Dermatomyositis

CLASSIFYING CLINICALLY AMYOPATHIC DERMATOMYOSITIS PATIENTS ON THE CONTINUUM OF CLASSICAL DERMATOMYOSITIS

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Category: Dermatomyositis

Dermatomyositis (DM) is an autoimmune condition that primarily affects the skin and muscle, but can also involve the lungs, joints and gastrointestinal tract, and is associated with internal malignancies. Approximately 20% of patients with serological and cutaneous features of DM never experience muscle weakness and are considered to have *clinically amyopathic dermatomyositis* (CADM). However, it is not well understood if CADM is a fundamentally different disease from classical DM (CDM) or if it is part of the disease's clinical spectrum. We performed a retrospective chart review of patients with clinically diagnosed adult-onset DM who were seen in the Stanford rheumatology-dermatology clinic between December 1998 and July 2025 to characterize CADM. CADM was defined as a lack of historical weakness plus weakness on manual muscle testing in our clinic for at least 6 months following onset of first disease symptom and for the duration of follow-up. Of the 257 patients who met inclusion criteria, 18% (n=45) had CADM. Compared to CDM, CADM patients were mostly female (82% vs. 76%), White (73% vs. 54%), non-Hispanic (11% vs. 72%), and had a median age of disease onset of 51 (vs. 51). The prevalence of ulcers, calcinosis, and alopecia (11%,4%,36%) did not differ significantly from CDM. Maximum CDASI-a scores were also similar with a median of 22 (95% CI 18-26) for CADM versus 20 (95% CI 19-23) for CDM. A significant number of CADM patients reported symptoms typically associated with muscle involvement, including dysphagia (11%), dysphonia (13%), and myalgia (16%). CADM patients also had signs of systemic involvement, including arthritis (7%), interstitial lung disease (13%), and associated malignancy (20%). Our data suggest that clinically amyopathic patients have similar extra-muscular presentation as that seen in classical DM, supporting that they may be considered part of a single disease spectrum.

STEROID INDUCED MYOPATHY AMONG DERMATOMYOSITIS PATIENTS

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Glucocorticoids remain the cornerstone of treatment for dermatomyositis (DM). However, steroid induced myopathy is a well-recognised complication which can clinically mimic disease progression. Distinguishing between these two pathologies is critical to reduce treatment-associated morbidity and prevent unnecessary escalation of immunosuppression. Steroid-induced myopathy among DM patients remains under reported in the literature. A retrospective review was conducted of DM patients diagnosed between 2015 and 2024 at Brigham and Women's Hospital. We identified 13 suspected cases of steroid-induced myopathy and analysed the demographic data, clinical features, diagnostic workup and steroid exposure. The median age was 57 years, and the average BMI was 30.3 kg/m^2. Muscle weakness was primarily proximal and symmetric. The mean cumulative prednisone dose at symptom onset was 21,774 mg, with an earlier onset observed among those who received intravenous pulse steroids. From baseline to the three-month follow-up visit, majority of patients (69.23%) had a reduction in prednisone dose. Despite dose reduction, four patients continued to experience worsening glucocorticoid toxicity compared to baseline. The Glucocorticoid Toxicity Index (GTI) was used to quantify overall steroid burden. The average cumulative worsening score was 59.44, which surpassed the minimal clinically important difference score of 10 points. All patients with steroid myopathy experienced at least one additional steroid-related toxicity. This study therefore highlights the high steroid burden experienced by DM patients, and the importance of recognizing steroid-induced myopathy early, to minimize treatment morbidity and ensure appropriate management.

Teaching point: In dermatomyositis patients, it is vitally important to distinguish between disease progression and steroid-induced myopathy to prevent unnecessary escalation of immunosuppression and reduce significant treatment-related morbidity.

Abstract Category: Dermatomyositis

LINKING THE CDASI AND PGA VISUAL ANALOG SCALE: IDENTIFYING THRESHOLDS OF DISEASE SEVERITY IN ALIGNMENT WITH PATIENT PERSPECTIVES ON QUALITY OF LIFE

Sarah Jun[1,2], Hammad Ali[1,2], Touraj Khosravi-Hafshejani[1,2], Lais Lopes Almeida Gomes[1,2], Sarini Saksena[1,2], Elena Wei[1,2], Victoria P. Werth[1,2]

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Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by muscle and skin involvement. Validated assessments such as the Cutaneous Dermatomyositis Disease Area and Severity Index Activity Score (CDASI-A, scored 0-100) and Physician Global Assessment Visual Analog Scale (PGA-VAS, scored 0-10cm) have been developed to standardize the evaluation of disease activity in DM. Moreover, tools such as the Skindex-29 have been developed to capture the impact of cutaneous disease on the quality of patients' lives in domains of Symptoms, Emotions, and Functioning. We assessed the relationship between the CDASI-A and PGA-VAS, reporting thresholds of disease severity that may guide the classification and monitoring of DM among individual patients and for clinical trial purposes. Data from 565 patients with DM across 2,161 visits were analyzed. Of the 565 patients, we then evaluated the relationships between these thresholds and quality of life assessments via the Skindex-29 among 419 patients across 1,279 visits. ROC curve analyses for PGA-VAS cutoffs of 2.5cm, 5cm, and 7.5cm revealed corresponding CDASI-A scores of 10 (AUC 0.89, sensitivity: 93.2%, specificity 67.5%, correctly identified 78.3%), 16 (AUC 0.94, sensitivity: 95.9%, specificity: 82.5%, correctly identified 84.6%), and 21 (AUC 0.95, sensitivity: 98.2%, specificity: 84.2%, correctly identified 84.5%), respectively. Mean Skindex-29 scores in the domains of Symptoms, Emotions, and Functioning were calculated for each range of CDASI-A scores (<10, 10 to <16, 16 to <21, and ≥21). Kruskal-Wallis testing demonstrated significant differences in Skindex-29 scores across all CDASI-A ranges ($p<2.2e-16$). Post-hoc Dunns testing demonstrated significant pairwise differences across all four ranges except 10 to <16 and 16 to <21, suggesting a broader moderate disease severity range of 10-21. These findings propose severity thresholds aligned with patient-reported quality of life metrics that can enhance the use of the CDASI in the classification and monitoring of cutaneous disease in patients with DM.

Category: Dermatomyositis

HIGH BURDEN OF PSYCHIATRIC COMORBIDITY IN DERMATOMYOSITIS AND ITS CLINICAL PREDICTORS

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Dermatomyositis (DM) is a multisystem autoimmune disease, yet the prevalence and risk factors for psychiatric illness in this population remain poorly defined. This study aimed to determine the prevalence of psychiatric illness and identify associated risk factors among DM patients. We conducted a retrospective cross-sectional study of DM patients with at least one outpatient visit at Johns Hopkins Medicine between June 4, 2014, and June 4, 2019. Patients with a diagnosis of any other autoimmune disorder were excluded, yielding 528 patients. Mean age at DM diagnosis was 56.5 years (SD=14.0), with 65.3% female and 52.1% White. Comorbid conditions included type 2 diabetes mellitus (T2DM, 23.3%), obesity (17.4%), dyslipidemia (30.1%), hypertension (34.8%), cardiovascular disease (51.1%), chronic kidney disease (CKD, 12.9%), chronic obstructive pulmonary disease (COPD, 4.7%), asthma (6.6%), smoking history (41.5%), and malignant neoplasm (15.3%). The all-time prevalence of any psychiatric illness was 34.8%. The most prevalent diagnoses included depressive episode (13.4%), major depressive disorder (7.0%), generalized anxiety disorder (3.0%), bipolar disorder (2.7%), post-traumatic stress disorder (1.1%), and schizophrenia (0.9%). Logistic regression analysis revealed that younger age at DM diagnosis was significantly associated with increased risk of psychiatric illness (estimate=-0.02866, p=0.001), with each one-year increase in age at diagnosis associated with 2.9% decrease in odds of psychiatric illness. Additionally, T2DM (adjusted odds ratio [AOR]=2.20, 95% confidence interval [CI]=1.26–3.86, p=0.005), dyslipidemia (AOR=2.19, 95% CI=1.34–3.58, p=0.002), and COPD (AOR=2.87, 95% CI=1.08–7.67, p=0.03) were independently associated with increased odds of psychiatric illness. Sex, race, hypertension, CKD, cardiovascular disease, asthma, smoking history, and malignant neoplasm history were not significantly associated. Psychiatric illness affects over one-third of DM patients. Younger age at diagnosis, T2DM, dyslipidemia, and COPD are significant risk factors. These findings highlight the need to screen for psychiatric comorbidities in DM patients, particularly in high-risk subgroups.

Abstract Category: Dermatomyositis

Cancer risk in dermatomyositis: a multi-institutional federated database analysis

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While dermatomyositis has reported cancer associations, comprehensive data stratifying malignancy risk by autoantibody profile remain limited. We assessed cancer risk in dermatomyositis patients versus matched controls, stratified by autoantibody profile (anti-Jo1, anti-MDA5, anti-Mi2, anti-NXP2, anti-TIF1γ). Using electronic medical records in TriNetX from 74 healthcare organizations (2016–2025), we identified 11,132 dermatomyositis patients via a validated ICD-10 algorithm (PPV≥91% for idiopathic inflammatory myopathies). We evaluated 3-year cancer incidences versus matched seborrheic keratosis controls, excluding prevalent cancers >6 months before the index-diagnosis. After 1:1 propensity score matching for demographics, comorbidities, and medication use, cohorts included 11,015 patients. Hazard ratios (HRs) with 95% confidence intervals (CIs) were calculated using Cox proportional hazards models. Dermatomyositis patients had higher 3-year risks of hematologic malignancies, such as Hodgkin lymphoma (HR 5.02; 95% CI 1.13–22.43), and solid organ malignancies, including lung (HR 1.42; 95% CI 1.07–1.88), esophageal (HR 3.50; 95% CI 1.48–8.27), lip/oral cavity/pharynx (HR 2.61; 95% CI 1.44–4.73), thyroid (HR 4.56; 95% CI 2.22–9.38), ovarian (HR 1.84; 95% CI 1.10–3.09), and digestive tract (HR 2.13; 95% CI 1.63–2.79) cancers. In antibody-specific cohorts, anti-Jo1 (n=1,646) showed elevated cancer risk (HR 1.30, 95% CI 0.97–1.76), driven by solid-organ cancers (HR 1.54, 95% CI 1.07–2.23). Anti-TIF1γ (n=200) showed elevated cancer risk (HR 1.81, 95% CI 1.03–3.18), driven by solid-organ cancers (HR 2.02, 95% CI 1.06–3.85). Anti-NXP2 (n=163) showed elevated cancer risk (HR 2.80, 95% CI 1.17–6.70). Anti-MDA5 (n=136) showed no increased cancer risk. Anti-Mi-2 (n=308) showed elevated cancer risk (HR 2.76, 95% CI 1.35–5.67), driven by solid-organ cancers (HR 2.27, 95% CI 1.08–4.74). Negative controls (varicose vein, trauma/laceration) showed no elevated risk across cohorts. Our results show dermatomyositis patients have distinct hematologic and solid organ malignancy patterns versus controls, highlighting cancer surveillance and monitoring importance.

Category: Dermatomyositis

RDS 2025 Abstract

TRENDS IN DERMATOMYOSITIS DIAGNOSIS: A RETROSPECTIVE ANALYSIS USING EPIC COSMOS DATA

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Abstract category:
Dermatomyositis

Dermatomyositis (DM) is an autoimmune disease that has evolving diagnostic criteria along with variable geographic/demographic presentations. Epic Cosmos is a large-scale electronic health record platform that aggregates U.S. healthcare systems data in order to provide an overview of national trends. A retrospective study using Cosmos was performed to evaluate trends in DM incidence and prevalence over an 8-year period to explore diagnostic changes, regional variation, and population demographics. Patients diagnosed with DM between 01/01/2017 and 05/01/2025 were identified. Summary statistics were extracted through secure, de-identified queries. Of 53,706 DM patients, 73.8% were female and 26.2% male. Racial breakdown included White (75.6%), Black (14.6%), Asian (4.0%), and others. Most patients (83%) were in metropolitan areas. States with greatest DM rates compared to population size were Connecticut, North Dakota, Ohio, Vermont, and West Virginia; the lowest were Alabama, Arizona, New Mexico, Rhode Island, and Washington. This is the first known epidemiological study of DM using Cosmos. Cosmos provides a powerful tool since data has shown it to be representative of the national population. Notably, findings on racial distribution contrast with prior literature noting a higher prevalence of DM in African American populations. This finding may reflect systemic barriers in diagnosis, specialty care access disparities, healthcare-seeking behavior differences, or a previous lack of systematic collection of DM data. Geographic findings showed that states with large populations had high total number of DM diagnoses, but states such as Connecticut, North Dakota, and Vermont showed higher DM rates relative to population size. Regional variations in environmental exposures, healthcare access, diagnostic recognition, or reporting practices may be factors that play a role. States with lower relative rates may underdiagnose DM due to decreased access to rheumatologic/dermatologic care. Overall, these patterns provide insight into the evolving landscape of DM diagnosis and identify disparities for guiding future research.

PREVALENCE OF DERMATOMYOSITIS IN THE UNITED STATES: A CROSS-SECTIONAL STUDY OF THE *ALL OF US* RESEARCH PROGRAM

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Dermatomyositis (DM) is an inflammatory myopathy characterized by skeletal muscle weakness and distinctive skin manifestations. The current prevalence and incidence data surrounding DM are based on studies from specific population groups, such as US Army Veterans, patients who received healthcare in Olmstead County, and a European cohort. There is a paucity of dermatomyositis epidemiological studies in diverse US cohorts. We aimed to explore the prevalence of dermatomyositis in the United States using the National Institutes of Health *All of Us* Research Program, which includes survey and electronic health record (EHR) data from a socioeconomically diverse cohort of > 800,000 US Americans. A cross-sectional analysis was performed using the *International Classification of Diseases, Tenth Revision, Clinical Modification* codes M33.1, M33.13, M33.10, M33.12, M33.11, M33.19 and *SNOMED* codes 396230008, 239901009, 402425006. Prevalence was calculated using the Wald test with a 95% confidence interval. From 371,398 patients with EHR data, 422 cases of DM were identified (0.11%; 95% CI, 0.1 - 0.12), with a 3:1 female predominance (73.22%). The mean and median age of DM patients were 51.7 and 54, respectively. Dermatomyositis was most prevalent in patients ages 55-64 years (.15%; 95% CI, .12-.18), specifically in females of this age group (79.8%). The prevalence of DM also varied by race/ethnicity and was higher in Black and Hispanic participants with survey data (.13%, 95% CI, .10-.15 and .13%, 95% CI, .10-.15, respectively) compared to White participants (.10%, 95% CI, .09-.12). Our data suggest that DM affects 1100 out of every million individuals in the United States. To our knowledge, this is the first study to examine prevalence data in the general US population using a large population-based dataset. These findings suggest that misdiagnosis or underdiagnosis is probable due to the rarity of this disease.

Abstract Category: Dermatomyositis

SKIN-PREDOMINANT JUVENILE DERMATOMYOSITIS: AN UNDERRECOGNIZED PHENOTYPE WITH SIGNIFICANT MORBIDITY

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Juvenile dermatomyositis (JDM) is a rare myopathy affecting the pediatric population characterized by a range of cutaneous manifestations and proximal muscle inflammation. Cutaneous findings are often the presenting sign of JDM, and can serve as markers for disease activity, chronicity, or systemic complications. Skin-predominant JDM is a clinically significant but underrecognized phenotype characterized by persistent and often severe cutaneous involvement with absent or minimal myositis. Recognizing the importance of an early diagnosis, we conducted a review of the current literature focusing on clinical features, pathophysiology, and management considerations of skin-predominant JDM. Myositis-specific autoantibodies, including anti-NXP2, anti-TIF1γ, and anti-MDA5, are associated with calcinosis, ulceration, and vasculopathy, further stratifying risk in cutaneous-dominant disease. However, key skin findings, such as Gottron's papules, heliotrope rash, ulceration, calcinosis, and periungual telangiectasias, are paramount in the clinical diagnosis and may precede systemic symptoms, persisting long after muscle enzyme normalization. Nailfold capillaroscopy reveals microvascular abnormalities that correlate with disease activity and predict flares, while histology often shows interface dermatitis or perivascular infiltrates. Patients with skin-predominant JDM may experience greater morbidity due to disfigurement, photosensitivity, and psychosocial distress, yet remain undertreated due to muscle biomarker-guided therapy. Prolonged corticosteroid use is common, but early initiation of steroid-sparing agents such as methotrexate, mycophenolate, or IVIG is recommended. Biologics targeting type I interferon pathways, including JAK inhibitors, are emerging as promising options for refractory disease. Early and aggressive immunosuppressive treatment can mitigate long-term cutaneous complications, underscoring the prognostic value of initial skin findings. Dermatologists play a central role in the diagnosis, surveillance, and therapeutic escalation in skin-predominant JDM. Persistent skin disease in JDM warrants aggressive and sustained therapy, as it reflects ongoing systemic immune activation. Early recognition and targeted treatment of this phenotype are crucial for improving long-term outcomes and enhancing the quality of life for affected children.

Category: Dermatomyositis

MORTALITY IN ASIAN PATIENTS WITH DERMATOMYOSITIS: A CASE SERIES

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Recent analysis from our cohort of 153 patients meeting the American College of Rheumatology and the European League Against Rheumatism (ACR/EULAR) criteria for dermatomyositis (DM) revealed significantly higher mortality rates among Asian patients compared to White patients (20.59% [7/34 Asian patients] vs. 3.07% [2/65 White patients], p=0.0035). To further investigate this disparity, we present a case series of the seven deceased Asian patients to characterize their clinical presentation and disease progression. Time from first DM-associated symptom to DM diagnosis for all patients was ≤6 months, with 4/7 (57%) diagnosed in ≤1 month. 6/7 patients (86%) died within 16 months of diagnosis; 2/7 (42%) died within 2 months. Most (5/7, 71%) passed in a hospital setting. 5/7 (71%) had a malignancy. Four (57%) patients were TIF1-γ positive, of which 4/4 (100%) had associated malignancies (2/4 were diagnosed before and 2/4 diagnosed after DM). 2/7 patients (29%) were MDA5 positive; both died ≤1 month of DM diagnosis. Neither of the MDA5 positive patients had known associated malignancy or interstitial lung disease (ILD). Rather, they died from colon perforation or acute hypoxic respiratory failure (AHRF) without ILD. The only patient without a positive myositis-specific antibody was also the sole case of ILD in our cohort, with AHRF as the cause of death. Overall, in this case series, we observed a predominance of TIF1-γ-associated malignancy contributing to death. Though there were cases of MDA5-associated mortality, interestingly, none were associated with ILD in our cohort. Early mortality despite short diagnostic timelines suggests aggressive phenotypes are major contributors to poor outcomes in these patients. The high proportion of hospital-based deaths also suggests acute deterioration. Though larger studies are warranted, these preliminary findings highlight the need for proactive initial management and urgent malignancy screening in Asian patients with newly diagnosed DM, especially in the setting of TIF1-γ positivity.

Teaching point: Our case series of seven deceased Asian patients with dermatomyositis primarily exhibited TIF1-γ positivity, rapid disease progression despite short diagnostic delays, and frequent in-hospital deaths, highlighting the importance of urgent malignancy workup and aggressive initial management.

Category: Dermatomyositis

INCREASED RISK OF RAPID ONSET OF MORTALITY IN ASIAN AMERICAN PATIENTS WITH DERMATOMYOSITIS: A RETROSPECTIVE STUDY

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Myositis specific antibodies (MSAs) associated with dermatomyositis (DM) are linked to unique clinical manifestations. Presentation may vary based on ethnicity. For example, anti-MDA5+ DM is associated with rapidly progressive interstitial lung disease (ILD), particularly amongst Asian populations. Other MSAs have higher rates of malignancy. To evaluate differences in mortality based on ethnicity and MSAs, we conducted a retrospective review of adult-onset DM patients seen between 2016 and 2025 at UCI Medical Center, which serves a diverse population in Southern California. A total of 153 patients met ACR/EULAR criteria for DM: 65/153 (42%) White, 49/153 (32%) Hispanic, 34/153 (22%) Asian, 5/153 (3.2%) Black. Documented deaths were noted in 13/153 (8.5%) records. Of these, 7/13 (53.8%) were Asian, 4/13 (30.8%) were Hispanic, and 2/13 (15.4%) were White. Asian patients experienced higher rates of mortality relative to White patients (20.59% vs. 3.07%, p=0.0035). There were no statistically significant differences in mortality between Hispanic patients and White patients (8.16% vs. 3.07%, p=0.230). Of the six patients who died within one year of diagnosis, 4/6 were Asian (2 TIF1-γ+, 2 MDA5+), 1/6 was Hispanic (MDA5+), and 1/6 was White (TIF1-γ+). Myositis was found in 11/13 (85%) of cases, malignancy in 7/13 (54%), and ILD in 3/13 (23%). Causes of death included infection (4/13, 31%), acute hypoxic respiratory failure (AHRF) in the context of malignancy (2/13, 15%), AHRF due to ILD (2/13, 15%, both in Hispanic patients), and AHRF with multiorgan failure (1/13, 8%). Cause of death was unspecified in 3/13 (23%). Overall, our findings suggest Asian Americans may be at relatively higher risk of rapid onset of mortality, with prominent causes of death extending beyond ILD. Previous literature has emphasized MDA5's role in increased mortality in this demographic, but our data suggests that TIF1-γ positive patients should also be closely monitored.

Teaching Point: Asian Americans may be at higher risk of rapid onset of mortality compared to other populations, especially in the setting of known positivity to TIF1-γ or MDA5.

Category: Dermatomyositis

NON-DIABETIC NECROBIOSIS LIPOIDICA: RHEUMATOLOGIC ASSOCIATIONS, CLINICAL SPECTRUM, CURRENT THERAPEUTICS AND BARRIERS IN A GLOBAL CASE SERIES

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Necrobiosis Lipoidica (NL) is a chronic granulomatous skin disease most frequently associated with diabetes mellitus (DM), although a minority of patients develop NL in the absence of hyperglycemia. These non-diabetic NL cases often present with concurrent autoimmune, rheumatologic, and inflammatory features that may indicate a stronger identification of NL as a rheumatologic vasculopathy. We systematically reviewed 26 published non-diabetic NL case reports and series from 15 countries spanning 2000-2025, extracting demographic data, histopathology, comorbidities, treatment regimen, and treatment response in 49 cases. Most patients were female (76.3%), of mean age 52.8 years. Lesions most frequently involved lower extremities and face. Atypical anatomic sites such as glans penis and upper extremities were more commonly found in non-diabetic NL cases than classic diabetic cases, in which lower extremity anterior tibial region predominates. In addition, 31.6% of cases had known vasculopathy, and 42.1% of cases had concurrent granulomatous skin disease. Furthermore, 24.5% of cases also presented with known autoimmune and rheumatologic disease such as morphea, rheumatoid arthritis, and sarcoidosis. With the lack of a gold standard treatment for NL, treatment modalities varied widely, with 49% of cases being treated with various forms of corticosteroids and 24.5% being treated with biologics, the most popular being adalimumab and secukinumab. 12.2% of cases were treated with methotrexate and cyclophosphamide, primarily in cases with concurrent rheumatoid arthritis. 36.7% of cases showed complete resolution of NL, with the most successful treatment modality being a combination of corticosteroids and biologics. Non-diabetic NL is frequently associated with autoimmune and rheumatologic comorbidities, with systemic involvement reinforcing its position within the rheumatology-dermatology spectrum. Key features of granulomatous inflammation and vascular pathology further support this inclusion. Consequently, treatment should prioritize advances in rheumatologic therapeutic strategies, exploration of novel therapies for NL, and the establishment of a standardized treatment approach.

Category: Miscellaneous rheumatic skin disease

THE IMPACT OF ALOPECIA ON SELF-ESTEEM AND QUALITY OF LIFE IN AUTOIMMUNE CONNECTIVE TISSUE DISEASE: A CROSS-SECTIONAL SURVEY STUDY

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Alopecia is a distressing manifestation of many autoimmune connective tissue diseases (ACTDs), affecting an estimated 36% of those with systemic lupus erythematosus (SLE), 9.5% of dermatomyositis, and 9% of systemic sclerosis. Although often associated with a significant psychosocial burden, alopecia is frequently under-evaluated in this population, likely due to other systemic symptoms being prioritized in management. This study aims to assess patient perspectives on alopecia and its impact on self-esteem and health-related quality of life in those with ACTDs. An anonymous survey was distributed to participants through ResearchMatch, Facebook support groups, and outpatient Dermatology and Rheumatology clinics to 1) assess the importance of alopecia to patients relative to other autoimmune symptoms, 2) characterize management strategies, and 3) study the impact of alopecia on self-esteem and health-related quality of life. English-speaking adults (≥18 years) with a self-reported ACTD were eligible. One-hundred and four individuals participated, with diagnoses of SLE, systemic sclerosis, and morphea, among others. About 64.4% (n=67) of patients reported experiencing hair loss, while only 23.9% (n=16) of these patients received treatment. Oral Minoxidil (56.3%), Spironolactone (31.3%), and corticosteroid topicals (31.3%) were the most utilized treatments. When asked to rate on a scale of 1 to 5 the importance of hair loss relative to their other symptoms, participants with hair loss reported a mean of 3.4 +/- 1.0. Furthermore, those with hair loss were more likely to report lower self-esteem ($p < 0.01$) and worse health-related quality of life globally ($p < 0.01$), and on scales of symptom ($p < 0.01$), function ($p < 0.01$), and emotion ($p < 0.01$). This study provides insight into the psychosocial impact of alopecia in ACTDs, suggesting those with hair loss experience lower self-esteem and reduced quality of life. While often under-addressed by clinicians, alopecia is distressing to patients and should prioritized in management.

Category: Miscellaneous rheumatic skin disease

RISK FACTORS UNDERLYING SECONDARY ERYTHROMELALGIA

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Category: Miscellaneous rheumatic skin disease

Abstract:
Erythromelalgia is a rare disorder characterized by episodes of pain, warmth, and erythema, primarily in the extremities. Secondary erythromelalgia, defined as erythromelalgia associated with risk factors such as myeloproliferative conditions, autoimmune diseases, and infections, remains poorly characterized. We aimed to elucidate the clinical presentations and underlying risk factors in patients with secondary erythromelalgia by conducting a retrospective chart review of adult patients with erythromelalgia who presented to the University of Pennsylvania's dermatology clinic from 2022 to 2025. Of 63 patients identified, 46.0% (n=29) were presumed to have secondary erythromelalgia. Most were female (79.3%), White (86.2%), and non-Hispanic (93.1%). Mean symptom onset was 43.7±15.3 and age at diagnosis 46.5±16.4 years. Nearly all (96.6%) reported erythema, burning pain (93.1%), and warmth (79.3%). 65.5% reported symptoms were triggered by heat, exercise or dependency and relieved by cooling. Body parts most commonly affected were feet (62.1%), hands (69.0%), and face (51.7%). Comorbidities included autoimmune/ connective tissue disorders (82.8%), Raynaud's disease (79.3%), migraines (65.5%), thyroid disease (51.7%), rosacea (48.3%), chillblains (20.6%), myeloproliferative disease (17.2%), diabetes (13.8%), and postural tachycardia syndrome (13.8%). 31.0% patients had prior exposure to iodinated contrast; no patients had exposures to bromocriptine or cyclosporine. Genetic testing was conducted in 7.9% and was negative in all cases. Overall, secondary erythromelalgia was predominantly affected White, non-Hispanic women and was strongly linked to autoimmune comorbidities and Raynaud's disease. The high frequency of facial involvement may be an underrecognized feature of secondary erythromelalgia. Clinicians should consider screening for autoimmune disorders in patients with suspected secondary erythromelalgia. Future studies should investigate causal relationships and evaluate whether targeted treatment of underlying comorbidities improves outcomes.

CHARACTERIZATION OF LABORATORY STUDIES AND TREATMENTS OF INTERSTITIAL GRANULOMATOUS DERMTATITIS WITH ARTHRITIS: A CASE REPORT AND REVIEW OF THE LITERATURE

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Interstitial granulomatous dermatitis with arthritis (IGDA) is a rare entity that manifests with violaceous or erythematous plaques or papules with nonerosive arthritis and is histopathologically described as a dense inflammatory infiltrate composed of histiocytes in the reticular dermis. Risk factors for IGDA are not well-known and the prognosis can be highly variable. There are currently no clear guidelines for treatment of IGDA. We present a case of a 47-year-old female with IGDA who experienced partial symptomatic improvement with prednisone, methotrexate and adalimumab and was recently switched to infliximab for better control of arthralgias and cutaneous symptoms. A narrative literature review was performed to determine trends in laboratory studies and treatments for IGDA patients. A total of 33 articles were analyzed, with a cumulative total of 50 IGDA cases therein. In 46% of cases, patients had a past medical history of rheumatoid arthritis (RA). Regarding laboratory studies, anti-nuclear antibody (ANA) was positive in 42% of cases and negative in 24% of cases; rheumatoid factor was positive in 32% of cases and negative in 34% of cases; C-reactive protein (CRP) was elevated in 28% of cases and normal in 8% of cases; and erythrocyte sedimentation rate (ESR) was elevated in 28% of cases and normal in 12% of cases. Oral prednisone and topical corticosteroids were the most frequently utilized treatments and were each used in 30% of cases. Methotrexate and hydroxychloroquine were each used in 20% of cases, and biologics were used in 8% of cases. The biologics utilized include adalimumab, infliximab, ustekinumab, and tocilizumab. Response to biologics was overall favorable, with cutaneous symptoms improving to a greater degree than arthralgias.

Teaching Point: IGDA is commonly associated with elevated CRP or ESR, ANA positivity, and past medical history of RA; corticosteroids and disease-modifying antirheumatic drugs are likely effective first-line treatments, and biologics may be effective for severe refractory cases.

Category: Miscellaneous Rheumatic Skin Disease

12-Month Risk of Serious Infection: Biologic vs Non-Biologic Psoriasis Treatment Analysis

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Psoriasis is a chronic inflammatory disease, with biologic and non-biologic systemic therapies increasingly utilized. Limited contemporary data compare infection risks between these therapy classes. We conducted a propensity-matched cohort study using the TriNetX Global federated network to assess 12-month risk of serious infection (hospitalization-requiring) after initiation of biologic or non-biologic systemic therapy in adults (≥18 years) with psoriasis (ICD-10-CM L40.0-L40.9). Patients initiating biologic therapies (tumor necrosis factor alpha [TNF-α], interleukin [IL]-12/23, IL-17, IL-23 inhibitors) or non-biologic systemics (methotrexate, cyclosporine, acitretin, apremilast, deucravacitinib) were propensity score-matched (1:1) based on demographics, comorbidities, use patterns, and severity proxies. Exclusions included prior HIV infection, active cancer, pregnancy, or tuberculosis. The primary outcome was serious infection (sepsis, pneumonia, cellulitis/erysipelas, pyelonephritis/urinary tract infection [UTI], septic arthritis/osteomyelitis, bacteremia) within 365 days, defined by inpatient admission and ICD-10 diagnosis. Among 116,664 matched patients (n=58,332 per group; mean age 47.4 years, 51.7% female), biologic therapy was associated with significantly lower rates for pyelonephritis/UTI (1.16% vs 1.53%, risk ratio [RR] 0.763, p<0.0001), pneumonia (0.91% vs 1.12%, RR 0.811, p=0.0003), bacteremia (0.23% vs 0.32%, RR 0.725, p=0.0042), and sepsis (0.62% vs 0.75%, RR 0.827, p=0.0071). Rates of cellulitis/erysipelas and septic arthritis/osteomyelitis were not statistically different between the two groups. Time-to-event analysis corroborated these findings. This analysis indicates biologic therapies confer a lower

risk of several serious infections compared to non-biologic systemics, supporting their safety profile in psoriasis management.

Category: Miscellaneous rheumatic skin disease

Hydalazine-Induced Autoimmune Syndromes: A Case Series

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We describe four female patients, aged between 50-75 years, with a minimum exposure to Hydralazine of 1 year (**Table 1).** The constellation of clinical presentations associated with hydralazine can mimic a broad range of autoimmune and inflammatory disorders. The patients had serologies tested, which are shown in **Table 2**. All treatment regimens involved systemic corticosteroids, though additional therapies of IVIG, rituximab, and/or methotrexate were also used. All patients responded favorably, yet vary from complete resolution to continued ulcerations.

The first two patients (P1and P2) had concomitant exposure to filgrastim and both experienced peri-orbital hemorrhagic bullae. P1 presented with abrupt-onset vesiculobullous lesions with predominant periorbital and oral distribution, while P2 experienced neutropenic fever along with annular pseudo-bullous lesions across the scalp and face. Additionally, P2 developed acral lesions that was initially considered to be ischemic in nature, prompting the initiation of anticoagulation. Only after biopsy, showing neutrophilic dermatosis consistent with Sweet syndrome, and initiation of systemic steroids did her lesions regress. P3 presented with ulcers mimicking pyoderma gangrenosum, even exhibiting its classic violaceous raised borders, that developed into vesiculobullous lesions along her extremities; skin biopsy showed components of leukocytoclastic vasculitis, and the patient was then treated for hydralazine-induced vasculitis. P4 had severe oral ulcerations, annular plaques on the scalp similar to those in P2, palpable purpura, and vesiculobullous ulcerations on her extremities. Skin biopsies revealed most patients having Sweet syndrome (P1, P2, P4), including one with a histiocytoid variant (P4), and multiple showing vasculitis (P1, P3, P4). Notably, P1 showed dense polymorphonuclear (PMN) infiltrate, vasculitis and cryptococcoid neutrophils consistent with reports[1] of Hydralazine induced vasculitis, lupus and Sweet syndrome occurring all together, hence a triumvirate.

Teaching Point: Hydralazine induced autoimmune syndromes can present with a variety of clinical morphologies mimicking infection, neutrophilic dermatoses and vasculitis; It is important to keep a high index of suspicion and order appropriate serologies to diagnose hydralazine induced autoimmune syndromes, even for patients who have been on hydralazine therapy for years.

Abstract Category Miscellaneous Rheumatic Skin Disease

Age/Sex	Hydralazine Exposure Length (years)	Pathological Description	Treatment	Outcome
54 F	1	Dense PMN infiltrate, Vasculitis, Cryptococcoid Neutrophils	Solumedrol 500mg IV taper, Clobetasol 0.05% ointment, IVIG	Resolved, complicated by Large GI bleed
75 F	2	Acute Neutrophilic infiltrate	Methylprednisolone 1g IV taper	Resolved, complicated by Large GI bleed
70 F	10	Dense PMN infiltrate, Vasculitis	Prednisone 10mg, IVIG, Rituximab, Debridement	Slow Improvement over 8 months
59 F	4.5	Mixed Inflammatory infiltrate, Vasculitis, Interstitial Lymphohistiocytic infiltrate	Solumedrol 250mg IV taper, prednisone, Methotrexate, IVIG, timolol, gentamicin	Healing on maintenance therapy for 4 months

Table 1: Association of Patient Case with Respective Exposure to Hydralazine, Pathology, Treatment Course, and Clinical Outcome.
Abbreviations: PMN (Polymorphonuclear Neutrophil), IVIG (Intravenous Immunoglobulin G), GI (Gastrointestinal)

Patient	ANA	ESR	CRP	RF	p-ANCA	PR3	MPO	anti-Histone
P1	**>1:1280**	**29**	**131**	**14.4**	**>1:320**	**8**	**4.7**	**6.4**
P2	**>1:1280**	**24**	**180**	<10	**>1:2560**	0.2	**4.1**	**5.1**
P3	Negative	**92**	**23**	**67**	**>1:1280**	**4.4**	**2.7**	<1.0
P4	**>1:640**	**101**	**75.2**	<10	**>1:640**	<0.2	**3.1**	**6.5**

Table 2: Autoimmune and Inflammatory Antibody Titers in Each Patient
Abbreviations: ANA (antinuclear antibodies), CRP (C-Reactive protein), RF (rheumatoid factor), p-ACNA (peri-nuclear antineutrophil cytoplasmic antibodies), PR3 (Proteinase 3 antibody), MPO (Myeloperoxidase antibody)

References:

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LYMPHOCYTE SIGNAL TRANSDUCTION DEFECTS AND EMERGING THERAPEUTIC TARGETS IN CUTANEOUS SARCOIDOSIS, A NARRATIVE REVIEW

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Abstract category: Miscellaneous rheumatic skin disease

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Cutaneous sarcoidosis is an autoimmune granulomatous disease characterized by an exaggerated inflammatory response of both the innate and adaptive immune systems. Highly refractory to various conventional immunomodulators, cutaneous sarcoidosis represents a therapeutic challenge. Recent studies have described the role of lymphocyte signaling defects in the pathogenesis of sarcoidosis; this review synthesizes recent advances in the immunopathogenesis of cutaneous sarcoidosis and highlights the therapeutic implications of these discoveries. CD4+ T cell dysregulation, toll-like receptor (TLR) activation, JAK/STAT signaling, and B-cell-mediated pathways all contribute to granuloma formation in cutaneous sarcoidosis. Granuloma formation is driven by Th1 and Th17 cell subsets, with recent studies identifying Th17.1 cells as predominant producers of IFN-γ, challenging classical paradigms. Th2 polarization and elevated thymus and activation-regulated chemokine (TARC) levels have also been associated with disease severity. Regulatory T cell dysfunction and PD-1 overexpression further contribute to chronic inflammation and fibrosis. The JAK/STAT pathway is notably upregulated in sarcoid granulomas, and JAK inhibitors (e.g., tofacitinib) have demonstrated promising efficacy in case series. Additional mechanisms include B-cell dysfunction, with elevated BAFF levels and reduced memory B cells, suggesting a role for B-cell-targeted therapies such as rituximab or belimumab. mTOR dysregulation has been implicated in granuloma formation, with sirolimus showing benefit in glucocorticoid-refractory disease. Infectious triggers (e.g., *Mycobacterium* spp.) and genetic susceptibility (e.g., IL-23R, HLA polymorphisms) further support the multifactorial nature of the disease. While prednisone and corticotropin gel remain the only FDA-approved therapies, improved understanding of sarcoidosis signaling has led to promising new therapeutic options. Further clinical trials are needed to validate these treatments and their role in multidisciplinary management.

IMPACT OF A COMMUNITY-BASED RHEUMATOLOGY-DERMATOLOGY CLINIC IN SERVING ASIAN AMERICAN PATIENTS

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Combined rheum-derm clinics are rare in the United States and even fewer are located in Asian American neighborhoods. In these settings, rheum-derm clinics face unique diagnostic and management challenges. Cutaneous lupus and psoriasis may present with subtle pigmentary changes, diminished erythema, or textural alterations, increasing misdiagnosis risk. Other barriers include limited treatment options due to HMO, underinsurance, lack of primary care providers awareness of rheum-derm diseases, and absence of established academic referral pathways. To evaluate the clinical impact of a community-based rheum-derm clinic serving a majority Asian American population, focusing on early diagnosis, access to advanced therapeutics, and overcoming structural barriers to care, a retrospective review was conducted of 351 patients evaluated between January 2021 and June 2025, including psoriasis/psoriatic arthritis (n=249), vasculitis (n=45), cutaneous lupus (n=30), systemic sclerosis (n=22), and dermatomyositis (n=5). Data collected included demographics, diagnosis, time from symptom onset to diagnosis, therapies initiated, use of patient assistance programs, and responses from post-visit surveys of patients and referring PCPs. Referral volume trends were also analyzed. Integrated rheum-derm evaluation reduced average time to diagnosis by 4 months compared to prior referral patterns. Earlier recognition and new treatment options were most impactful for Asian American patients. Access to guselkumab, secukinumab, and belimumab was expanded through patient assistance programs. Post-visit surveys showed high satisfaction from 90% of patients and 72% of PCPs. Referral numbers increased by 66% over the four-year period. A community-based rheum-derm clinic in an Asian American neighborhood is feasible and can accelerate diagnosis, reduce misdiagnosis, and expand treatment access for skin of color patients. Addressing payer limitations, enhancing PCP education, and developing academic referral linkages are essential to sustain and replicate this model.

Abstract Category: Miscellaneous rheumatic skin disease

CHARACTERIZING PERNIO: CLINICAL FEATURES, WORKUP, AND OUTCOMES IN A LARGE U.S. COHORT

Authors: Zasca-aisha Ristianto BS[1,2]; Kaitlin Martins, MS[1]; Sophia Manduca, BS[1,2]; Camille Scandurro, BS[2]; Alisa N. Femia, MD[1]

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Pernio is a rare inflammatory condition affecting acral skin, often triggered by cold, damp environments. Its pathogenesis and clinical patterns remain poorly characterized. This retrospective chart review of 463 patients at a large academic center (median age: 41 years; 70.2% female) explores presentation, comorbidities, and treatment outcomes of pernio. Most cases (80%) occurred during cold, wet months. Pernio affected only the feet in 49%, only the hands in 23.3%, and both in the remainder. Discoloration (80.1%) and tenderness (76.4%) were the most common symptoms; 84.4% had multiple symptoms. Among 325 patients evaluated for autoimmune, hematologic, or viral etiologies, 73% had idiopathic pernio. Immune-mediated disease was found in 16.4%, infection in 9.3%, with 41 of 43 infectious cases attributed to COVID-19, and 1% had drug-induced pernio. Of 241 patients tested for ANA, 107 were positive, although only 76 had connective tissue disease. Among patients with follow-up (mean: 13 months), 190 achieved full resolution. Of those, 65.8% responded to conservative measures alone (e.g., dressing warmly, avoiding cold, quitting smoking). Pharmacologic treatment was required in the remainder, most commonly with corticosteroids, topical tacrolimus, calcium channel blockers, and/or hydroxychloroquine. The average number of treatments needed to reach complete response was 1.27 ($p = 1.33e-10$). This largest single-center analysis of pernio confirms seasonal prevalence, female predominance, and favorable outcomes with non-pharmacologic treatment. While most cases were idiopathic, a subset was associated with autoimmune or post-viral syndromes, particularly COVID-19. The distinct seasonal pattern observed in COVID-19-associated cases, nearly half of which presented during warmer months, suggests a unique clinical subset consistent with the previously documented "COVID toes" phenotype. These cases were also more likely to involve the feet alone, further supporting a distinct presentation compared to idiopathic pernio. This large cohort offers valuable insight into patterns of pernio presentation, comorbidity, and management. Further research should identify predictors of systemic disease to guide individualized testing and management.

Category: Miscellaneous rheumatic skin disease

Clinical Course in Unspecified Panniculitis: A Retrospective Cohort Study

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Panniculitis comprises a diverse group of inflammatory conditions affecting subcutaneous fat. When histopathologic and clinical features do not clearly indicate a defined subtype, clinicians often label the condition as panniculitis, unspecified (UP). Despite the frequency with which UP is used as a diagnostic placeholder, little is known about the clinical trajectories of patients who receive this label and whether it ultimately leads to diagnostic resolution. We conducted a retrospective chart review of adult patients (ages 18–89) treated at a large academic medical center between January 1, 2000, and November 30, 2021, who were assigned ICD-9/10 codes for panniculitis (729.3/M79.3) or its subtypes. Patients with a clear diagnosis of a defined panniculitis subtype (i.e., erythema nodosum) without a preceding UP label were excluded. After manual chart review, 100 patients met inclusion criteria. Of these, 64% were ultimately assigned a specific final diagnosis. Biopsies were performed in 78 patients. Among those, 22 patients (28.2%) received an immediate diagnosis based on the initial pathology results (with 4 weeks of biopsy date). An additional 23 patients (29.5% of patients with biopsies) received a final diagnosis at a later point following biopsy, with a mean time to diagnosis of 714 days and a median of 296 days. Among the 22 patients who did not undergo biopsy, 15 (68.2%) were ultimately assigned a final diagnosis, with a significantly shorter time to diagnosis (mean 100 days, median 17 days; $p = 0.017$ compared to delayed-diagnosis biopsy group). The most common presenting symptoms across the cohort were subcutaneous nodules (61.4%), tenderness (50.5%), and erythema (48.5%). Final diagnoses included erythema nodosum (15%), lipodermatosclerosis (7%), lupus panniculitis (5%), and cold panniculitis (2%). Notably, 39 patients underwent biopsy but never received a definitive diagnosis. These findings suggest that the UP label may reinforce diagnostic inertia and that the presence of a biopsy alone does not appear to improve the likelihood of diagnostic resolution. Improved diagnostic frameworks, algorithms, and follow-up strategies are needed for patients with unclear panniculitis presentations.

Category: Miscellaneous rheumatic skin disease

HYPERPIGMENTATION AS AN ADVERSE EFFECT OF ANTIMALARIAL USE AND THE ASSOCIATION WITH RETINOPATHY AND PREDISPOSING FACTORS: A SYSTEMATIC REVIEW.

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We conducted a systematic review to examine the association between antimalarials and mucocutaneous hyperpigmentation, particularly potential correlations with retinopathy and prior trauma or ecchymosis. 63 studies including 317 patients with antimalarial-induced hyperpigmentation were identified. Most were female (89%), with an average onset age of 25.6 years. Pigmentation typically developed after a median of 854 treatment days and a mean cumulative dose of 204.4 g. Most patients had systemic lupus erythematosus (74.06%), followed by rheumatoid arthritis, Sjögren's syndrome, cutaneous lupus, and other connective tissue diseases. Hydroxychloroquine was used in 85.2% of cases, chloroquine in 8.88%, and other antimalarials in 5.92%. The pigmentation colour was mostly not stated. When reported, blue gray was most common. Frequently affected sites included the face, legs, hands, arms, and hard palate. In 707 patients on antimalarials from 33 studies, 94 had hyperpigmentation; among them, 8.5% had retinopathy. A weak statistically significant association between hyperpigmentation and retinopathy was found (Cramér's $V = 0.2169$, $p < 0.000000005$). In the chloroquine group (n=15), all had hyperpigmentation, and 20% also had retinopathy, suggesting a possible link. In hydroxychloroquine users (n = 562), a weak association was found (Cramér's $V = 0.08$, $p = 0.026$), although based on very few cases. Among 1,104 pooled patients, 228 developed hyperpigmentation. 59.54% reported preceding trauma or ecchymosis. A significant association was confirmed (Cramér's $V = 0.35$, $p < 2.2 \times 10^{-16}$). Histopathology of antimalarial-induced hyperpigmentation showed non-specific patterns. Hyperpigmentation of basal keratinocytes without increased melanocyte number was common. Non-refractile yellow-brown granules composed of melanin, hemosiderin, or both were noted in the dermis. Pigment-laden macrophages were a frequent finding. Oral lesions showed marked subepithelial pigment in the lamina propria, within macrophages and between collagen bundles. No pathognomonic features distinguish it from other causes of pigment incontinence.

PROTECTIVE EFFECTS OF AUTOIMMUNE DERMATOLOGIC CONDITIONS ON THYROID EYE DISEASE DEVELOPMENT: A LARGE-SCALE RETROSPECTIVE COHORT STUDY

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Background: Thyroid eye disease (TED) is a serious autoimmune orbital inflammatory condition affecting up to 50% of patients with Graves' disease. While autoimmune conditions often cluster together, the relationship between autoimmune dermatologic diseases and TED development remains unclear. Understanding these associations could inform screening practices and reveal protective mechanisms. Our objective is o determine the risk of TED development in patients with systemic lupus erythematosus (SLE) and dermatomyositis compared to matched controls without autoimmune skin diseases. We conducted a retrospective cohort study using the TriNetX Research Network (70 healthcare organizations) from 2005-2024. Patients ≥18 years with SLE or dermatomyositis were compared to propensity score-matched controls without autoimmune skin diseases. Primary outcome was incident TED (ICD-10: H05.2x) occurring ≥1 day after autoimmune disease diagnosis. Cohorts were balanced on age, sex, thyroid disorders, and glucocorticoid use. After propensity matching, we analyzed 578,085 patients: 267,095 with SLE, 44,895 with dermatomyositis, and matched controls. Contrary to expectations of autoimmune clustering, both conditions showed protective effects against TED development. SLE patients had 542 TED cases versus 702 in controls (0.20% vs 0.26% risk; Risk Ratio 0.77, $p<0.001$). Dermatomyositis patients had 109 cases versus 151 in controls (0.24% vs 0.34% risk; Risk Ratio 0.72, $p=0.009$). Autoimmune dermatologic conditions appear protective against TED development, challenging conventional assumptions about autoimmune disease clustering. Potential mechanisms include immunosuppressive treatment effects, immune tolerance, enhanced surveillance, or competitive autoimmune responses. These findings suggest novel approaches to TED risk stratification and highlight complex interactions between autoimmune conditions warranting mechanistic investigation.

Table 1. Baseline Characteristics and TED Outcomes by Autoimmune Dermatologic Condition

Characteristic	SLE (n=267,095)	SLE Controls (n=267,095)	Dermatomyositis (n=44,895)	Dermatomyositis Controls (n=44,895)
Demographics				
Age, mean ± SD (years)	48.4 ± 17.0	48.8 ± 17.3	54.3 ± 17.9	54.3 ± 17.9

Female, n (%)	219,115 (82.0)	219,307 (82.1)	29,958 (66.7)	29,925 (66.7)
White, n (%)	151,539 (56.7)	152,582 (57.1)	28,057 (62.5)	28,117 (62.6)
Comorbidities				
Thyrotoxicosis, n (%)	1,242 (0.5)	1,343 (0.5)	300 (0.7)	254 (0.6)
Hypothyroidism, n (%)	12,057 (4.5)	12,386 (4.6)	2,885 (6.4)	2,879 (6.4)
Thyroiditis, n (%)	1,254 (0.5)	1,175 (0.4)	298 (0.7)	257 (0.6)
Medications				
Glucocorticoids, n (%)	39,915 (14.9)	39,438 (14.8)	16,959 (37.8)	17,003 (37.9)
Primary Outcome				
TED cases, n	542	702	109	151
TED risk, %	0.20	0.26	0.24	0.34
Risk Ratio (95% CI)	0.77 (0.69-0.86)	Reference	0.72 (0.57-0.91)	Reference
P-value	<0.001	--	0.009	--

Abbreviations: SLE, systemic lupus erythematosus; TED, thyroid eye disease; SD, standard deviation; CI, confidence interval

All comparisons made after propensity score matching on age, sex, race, thyroid disorders, and glucocorticoid use

Category: Miscellaneous rheumatic skin disease

ANEMIA RISK IN PATIENTS WITH HIDRADENITIS SUPPURATIVA: A MULTI-INSTITUTIONAL COHORT STUDY

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Hidradenitis suppurativa (HS) is a chronic autoinflammatory skin disorder associated with systemic comorbidities, including anemia. Although single-center studies report a substantial anemia burden in HS, multi-institutional evidence is limited. We conducted a retrospective cohort analysis using TriNetX to compare the cumulative incidence of anemia (ICD-10 D50–D64) after initial HS diagnosis (ICD-10 L73.2) with matched controls. Adults without prior anemia diagnosis were matched 1:1 on age, sex, race, ethnicity, and index date, yielding two balanced cohorts (n = 33,355 each). Median follow-up was 194 days (IQR 230) for HS patients and 104 days (IQR 236) for controls. Follow-up continued until the end of available records. Relative risk (RR) with 95% confidence intervals (CI) was calculated; $p < 0.05$ was considered significant after Benjamini–Hochberg correction. The cumulative incidence of all anemia subtypes was 2.0% in HS versus 1.7% in controls (RR = 1.2, CI: 1.02–1.3). Iron deficiency anemia showed the strongest association (1.0% vs. 0.7%; RR = 1.4, CI: 1.2–1.7). Nutritional anemias were also common (1.2% vs. 0.8%; RR = 1.5, CI: 1.2–1.7), as were other nutritional subtypes and hereditary hemolytic anemias. Other subtypes were not significantly different. Smaller effect sizes than prior reports likely reflect differences in measuring incident anemia, excluding preexisting cases, and underestimation from diagnostic code capture. Shorter median follow-up in controls may attenuate cumulative incidence and bias RR upward; this should be considered in interpretation. HS was associated with a modestly increased risk of incident anemia, most often coded as "other anemias," likely including anemia of chronic disease. Iron deficiency and nutritional deficiencies were also more frequent, potentially related to chronic inflammation, blood loss, or nutritional factors. Although absolute incidence was low, periodic anemia screening in HS may be warranted, and prospective studies with laboratory confirmation are needed.

Category: Miscellaneous rheumatic skin disease

JAK INHIBITORS FOR TREATMENT OF SAPHO SYNDROME: A SYSTEMATIC REVIEW OF 72 CASES

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Synovitis, acne, pustulosis, hyperostosis, and osteitis (SAPHO) syndrome usually involves bone, joints, and skin. In the lack of known pathogenesis and clinical trials, there is no standard treatment for SAPHO patients. Janus kinase inhibitors (JAK-I) are a group of small-molecule drugs with a

wide range of effects on inflammatory and autoimmune pathways. A systematic search was conducted using MeSH terms/keywords related to JAK-I and SAPHO Syndrome through PubMed/Medline, Scopus, Web of Science, and Embase until September 8th, 2024. The inclusion criteria were a diagnosed SAPHO syndrome patient who received at least one JAK-I. We excluded reviews and animal studies. Out of 287 initially researched articles, we included 34 articles. These 34 articles involved 72 patients with a mean age of 39.36 years and a female (78%) predominance. All patients had bone or joint involvement, most commonly in the anterior chest wall (62.5%) and vertebrae (45.8%). Skin involvement was seen in 64 patients, with 52 (72.2%) presenting palmoplantar pustulosis (PPP). Nearly all (97.2%) had prior treatments, with nonsteroidal anti-inflammatory drugs (NSAIDs), corticosteroids, and methotrexate (MTX) being the most common. Tofacitinib was the most used JAK-I (54/72, 75%), followed by baricitinib (15/72, 20.8%) and upadacitinib (3/72, 4.2%). 18 (25%) patients received concomitant treatment with another medication; of these, 7 received NSAIDs and 6 received MTX. Different variants were used as responses to treatment in these studies, which were as follows: alleviating or clearing symptoms, significant changes in imaging, a decrease in laboratory inflammatory markers, or a Visual Analogue Scale (VAS). Almost all patients (71/72, 98.6%) showed a good to complete response. Adverse effects occurred in ten patients (13.9%), all on tofacitinib; only one led to drug discontinuation. Lab abnormalities were seen in 6.9% without clinical symptoms. In conclusion, JAK-I seems to be a promising treatment for SAPHO syndrome with manageable adverse effects.

Abstract category: Miscellaneous rheumatic skin disease

Rapidly Progressive Systemic Sclerosis Following COVID-19 Infection

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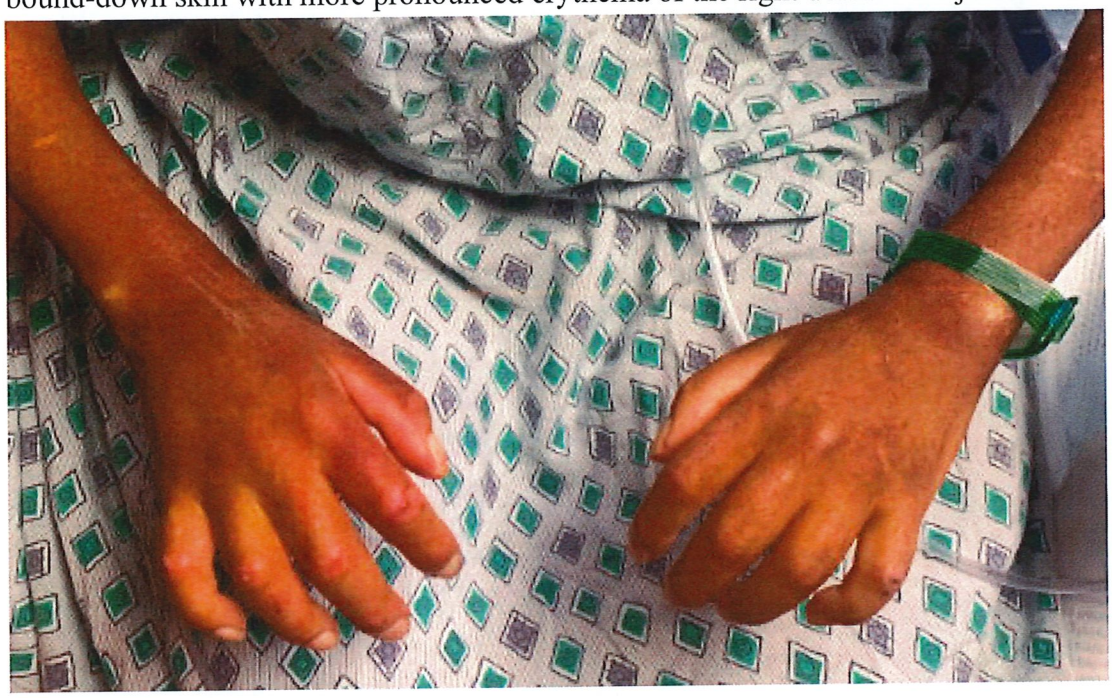
Systemic sclerosis (SSc) is a chronic, multisystem connective tissue disease characterized by vascular dysfunction and progressive fibrosis. While COVID-19 has been associated with several autoimmune conditions, there have only been three published cases of post-COVID SSc. We present the case of a 68-year-old female with diabetes, gastroesophageal reflux, and hypertension who developed lower extremity edema and hand skin tightening nine days after a self-limited COVID-19 infection. Rheumatologic evaluation revealed sclerodactyly, telangiectasias, and anti-U1 RNP positivity, leading to a diagnosis of SSc (**Figure 1**). Initial methotrexate therapy was ineffective, prompting a switch to mycophenolate mofetil (1,500 mg twice daily). Within six months, she developed acute pericarditis with inflammatory pericardial effusion requiring drainage. Three months later, she was hospitalized for euglycemic diabetic ketoacidosis, complicated by acute kidney injury (creatinine 4.42 mg/dL). Over the following three months, she developed acute systolic heart failure (EF 45%) with pulmonary hypertension, large pleural effusion, and bilateral ground-glass opacities. Thoracentesis showed a transudative effusion, attributed to SSc. Her renal function deteriorated to scleroderma renal crisis, requiring hemodialysis. Subsequent months were marked by recurrent Serratia bacteremia from permacath infections, severe oropharyngeal dysphagia, and progressive decline. Despite aggressive immunosuppression and supportive care, she expired less than two years after SSc diagnosis, having developed multiorgan involvement (skin, renal, pulmonary, cardiac) within one year of onset. This case highlights a fulminant, multisystem SSc presentation closely following COVID-19 infection. While causality remains unproven, the rapid onset and progression suggest COVID-19 may have accelerated the disease course.

Teaching Point: Post-COVID-19 SSc presentations may follow an unusually aggressive trajectory, warranting earlier recognition and potentially intensified treatment strategies to slow progression and mitigate organ damage.

Category: Clinical Case

Figure 1: Evidence of sclerodactyly affecting bilateral hands with edematous changes and bound-down skin with more pronounced erythema of the right dorsal PCP joints



REFRACTORY SCLE RESOLVED BY IDENTIFYING AN UNCONVENTIONAL TRIGGER: A CASE OF CANNABIS-INDUCED EXACERBATION

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Subacute Cutaneous Lupus Erythematosus (SCLE) is an autoimmune disorder characterized by non-scaring, photosensitive, ring-shaped lesions on sun-exposed areas. Cigarette smoking is well known to exacerbate cutaneous lupus activity through pro-inflammatory and immunomodulatory pathways and contribute to treatment failure. We report a case of refractory SCLE in which transition from combusted cannabis to edible and vaporized preparations facilitated lasting remission. A 45-year-old female presented with SLE and SCLE. The patient had been refractory to hydroxychloroquine, quinacrine hydrochloride, intravenous immunoglobulin (IVIg), rituximab, belimumab, lenalidomide, dapsone, and colchicine over the course of a year of management. Physical examination revealed red annular and circinate plaques with areas of depigmentation involving the face, trunk, neck, arms, and distal legs, along with diffuse non-scarring alopecia and oral ulcerations. Although the patient denied cigarette smoking, the physician failed to ask about other types of smoking. After months of additional treatment failure, further discussion revealed prolific cannabis smoking. Following transition to non-smoked cannabis, her skin lesions improved significantly and entered lasting remission. Although the direct role of cannabis smoke in SCLE is not well-studied, existing literature illustrates a strong association between tobacco smoking and increased Cutaneous Lupus Erythematosus Disease Area and Severity Index (CLASI) scores and risk of cutaneous manifestations. Given that there may be an overlap in toxins, such as aromatic amines, carbon monoxide, and polycyclic aromatic hydrocarbons, similar effects on disease activity may be observed between tobacco and cannabis smoke. This case highlights the importance of facilitating open communication about social and smoking history, as well as educating patients on how these factors can influence disease activity. Incorporating harm reduction strategies, such as recommending non-smoked cannabis formulations, offers a practical, patient-centered approach to managing SCLE flares, especially in cases unresponsive to standard treatments.

Teaching Point: Inhalation of cannabis smoke may act as a modifiable trigger for cutaneous lupus flares, particularly in cases refractory to standard therapy.

Category: Clinical Case

Figures

Figure 1: Initial SCLE presentation with the use of smoked cannabis.

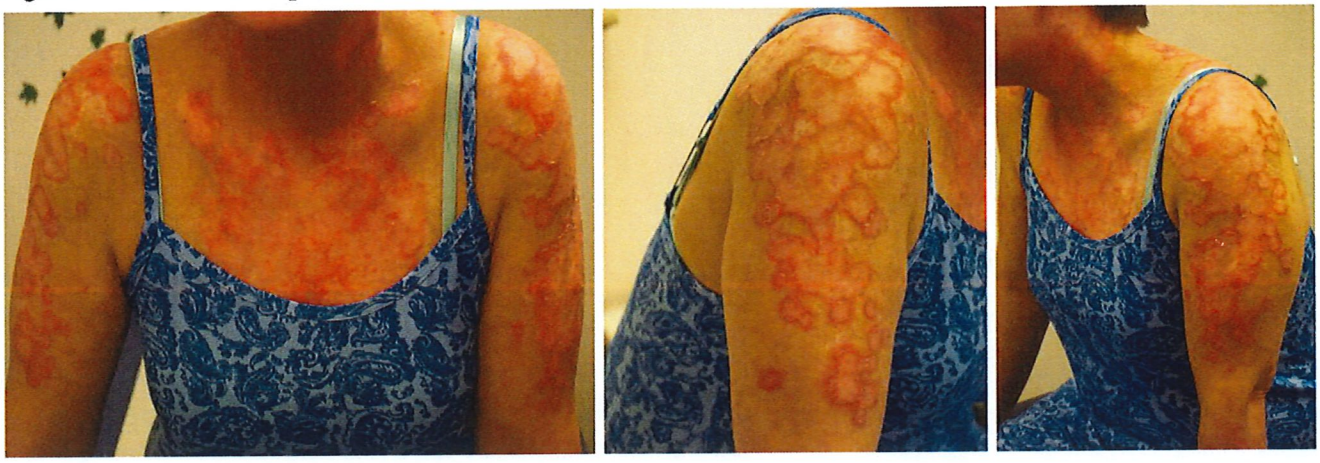
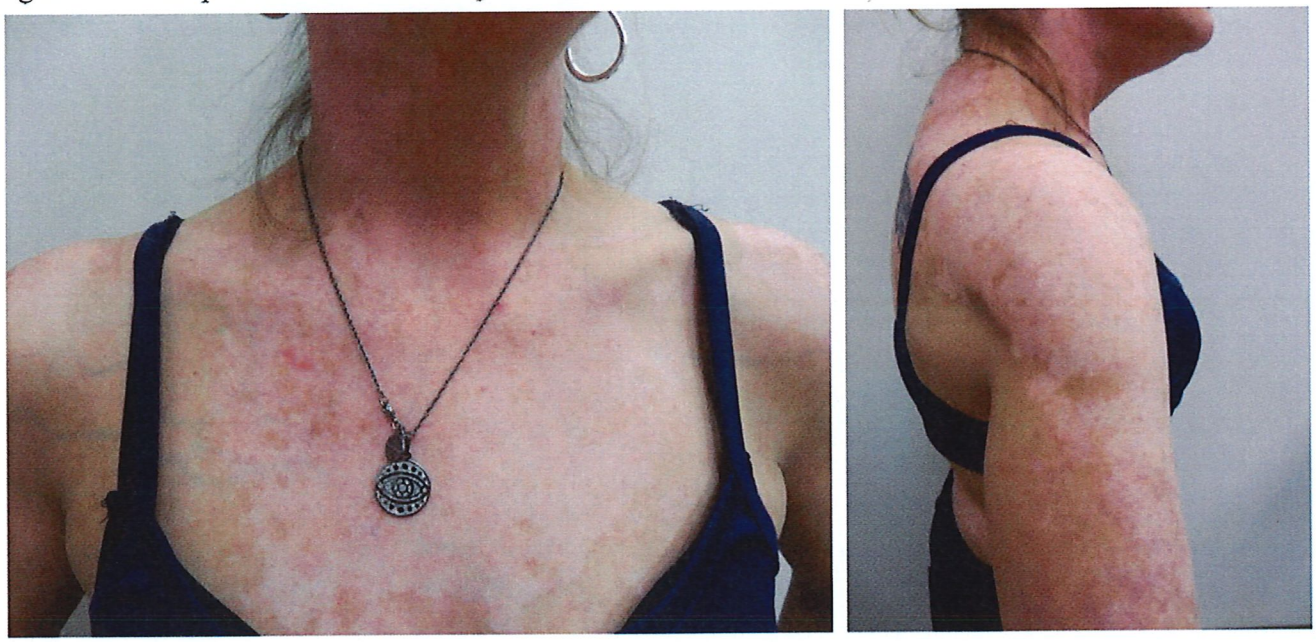


Figure 2: SCLE presentation following transition to non-smoked cannabis.



HEAT-INDUCED DISCOID LUPUS ERYTHEMATOSUS

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We present a 54-year-old male presented with a six-week history of a pruritic rash on his chest, back, and knees. Past medical history includes hypertension, hyperlipidemia, and a five-year history of discoid lupus erythematosus (DLE), for which he has been on oral hydroxychloroquine, and topical ruxolitinib and clobetasol cream. He reported nightly use of a heating blanket. Dermatologic examination revealed hyperpigmented, scaly linear and papular plaques on the left chest, flanks, lower back, right lower abdomen, and knees. Anifrolumab infusions were initiated, leading to improvement in the rash except for persistent lesions on the abdomen and back. Punch biopsies were performed on the right periumbilical region and lower back. Histopathology of the right periumbilical region revealed lichenified interface dermatitis with increased dermal mucin, consistent with DLE and lichen simplex chronicus (LSC). PAS staining showed a focally thickened basement membrane, and colloidal iron staining indicated increased dermal mucin throughout the reticular dermis. The lower back biopsy was consistent with erythema ab igne with LSC changes. Antinuclear antibody was positive with a titer of 1:1280 in a speckled pattern. Complete blood count, metabolic panel, C4, C3, and urinalysis were normal. Double-stranded DNA antibody was negative. This case represents a rare instance of heat-induced discoid lupus erythematosus, due to the Koebner phenomenon. While ultraviolet radiation is a known trigger for lupus, heat as a trigger is uncommon. The patient discontinued the heating pad and continued oral hydroxychloroquine, anifrolumab infusions, and topical clobetasol and ruxolitinib. The abdominal lesions gradually improved, and the itching subsided.

Teaching Point: Although trauma can trigger or worsen symptoms in lupus patients, it is often overlooked in patient education; this case highlights the role of skin injury in DLE pathogenesis and the need to avoid prolonged, low-grade heat exposure to reduce the risk of flares.

Category: Clinical Case

Figure 1. 54-year-old man with hyperpigmented, scaly linear and popular plaques on the left chest, flanks, lower back, right lower abdomen, and knees.

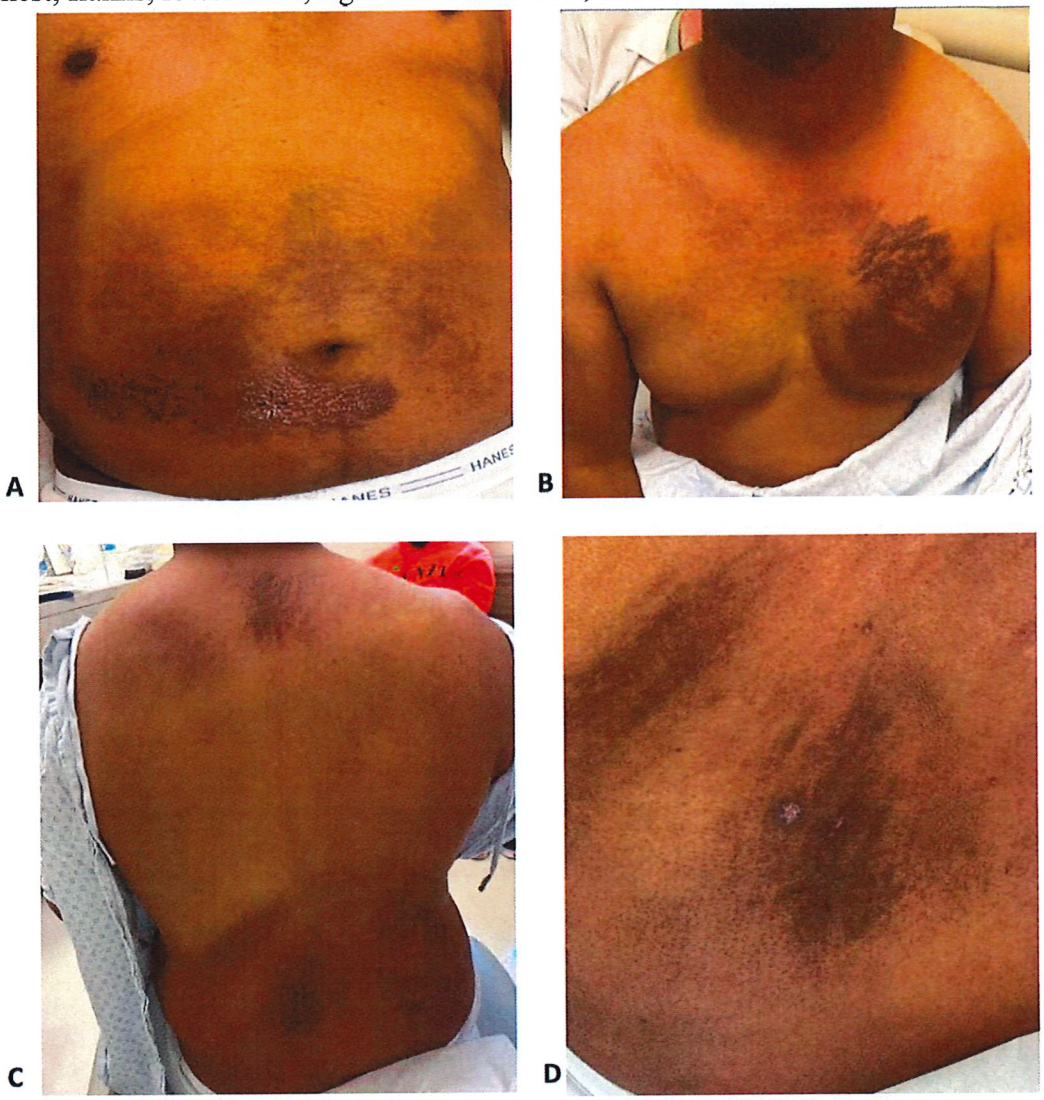
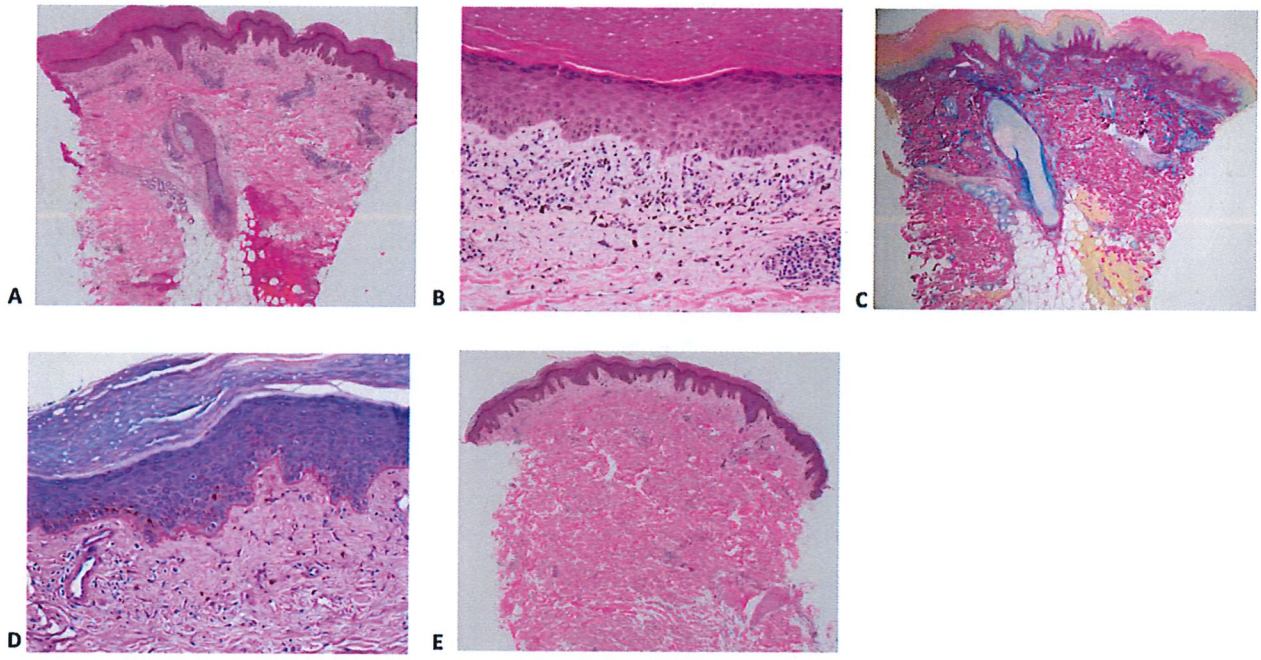


Figure 2. Hematoxylin and eosin: left lower back, 4x (A); and right periumbilical, 4x (B) and 20x (E). Right periumbilical, 4x (C), colloidal iron stain. Right periumbilical, 20x (D), Periodic Acid-Schiff stain.



CUTANEOUS SCLEROSIS DUE TO STAT3 GAIN-OF-FUNCTION MUTATION

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Signal transducer and activator of transcription 3 (STAT3) regulates cellular survival, proliferation, differentiation, and immune activation by promoting adaptive immunity and T-cell survival. Gain-of-function (GOF) mutations are rare, with only several hundred reported. Common cutaneous findings include eczema (60%), psoriasis (12%), and alopecia (10%).[1] However, STAT3 also appears central to cutaneous fibrosis: profibrotic signals such as TGFβ in systemic sclerosis (SSc) converge on STAT3, phosphoSTAT3 is elevated in SSc skin, and STAT3 knockout ameliorates fibrosis in murine models.[2,3] We report a 42-year-old man with seronegative RA, type 1 diabetes, vitiligo, hypothyroidism, bilateral carpal tunnel release, and persistent lymphadenopathy, presenting with a decade of skin tightening on forearms and thigh. He also reported fatigue and morning stiffness but denied Raynaud's, photosensitivity, muscle symptoms, or dyspnea. Exam revealed depigmented patches, sclerosis over ankles and wrists, waxy thickening of dorsal digits with limited extension but without nailfold capillary changes or digital pits, and a reticulated hyperpigmented plaque on the right buttock/thigh. CBC, quantiferon, hepatitis serologies, aldolase, ESR, CRP, CK, ANA, and ENA were negative; myositis panel was positive for TIF1γ. Chest CT was unremarkable. Thigh punch biopsy showed dermal sclerosis with deep dermal–subcutaneous lymphoplasmacellular infiltrate and retained adnexa to the eccrine coils. Genetic testing revealed a p.R278C STAT3 GOF mutation, previously unreported. In the absence of dermatomyositis features, TIF1γ positivity was attributed to interferon dysregulation, and hand changes to diabetic cheiroarthropathy. His morpheaform plaque and ankle/wrist sclerosis were felt to be STAT3-driven. Tofacitinib treatment was chosen for modulation of JAK/STAT signaling and historical success in treating STAT3 GOF,[4] and has led to stabilization of symptoms at 11 mg daily. This case demonstrates the possible relationship between STAT3 signaling and cutaneous sclerosis, which may provide a future avenue for disease-modifying therapy.

Teaching point: Gain-of-function mutations in STAT3 can predispose to autoimmune phenomena, and janus kinase inhibitor therapy is a reasonable strategy in such cases.

Clinical photographs:

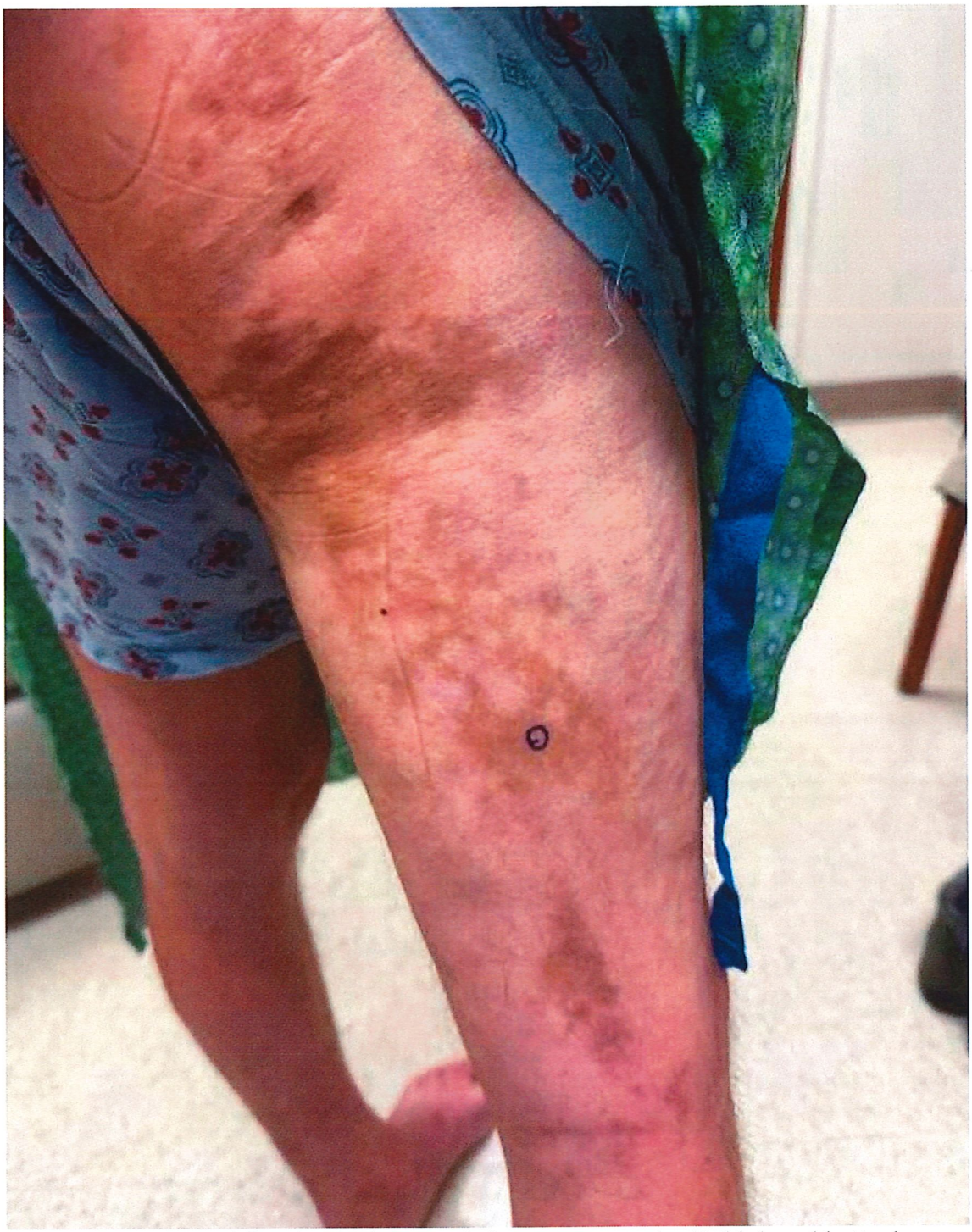

Figure 1: Reticulated firm hyperpigmented plaque on posterior R thigh and buttock.

Abstract category: Clinical case

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RELAPSING SYSTEMIC LUPUS ERYTHEMATOSUS TREATED WITH DUAL RITUXIMAB AND ANIFROLUMAB THERAPY

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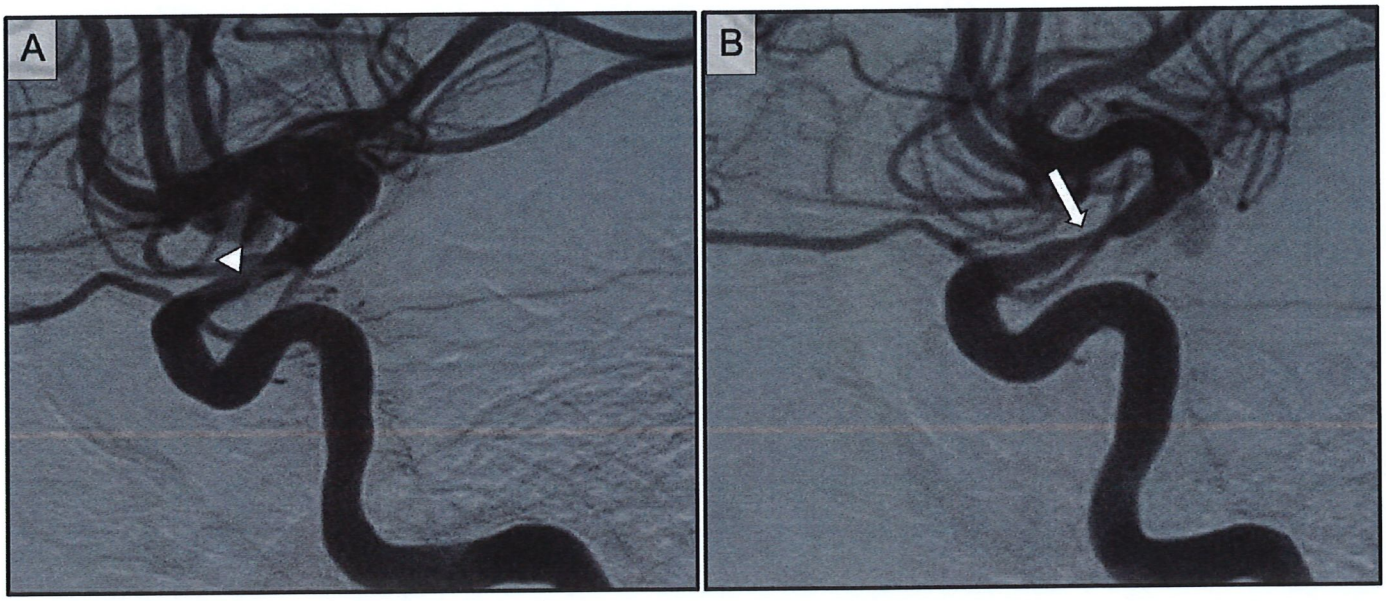
Biologic monotherapy is often used in the management of moderate-to-severe systemic lupus erythematosus (SLE), particularly in patients who exhibit an inadequate response to standard immunosuppressive agents. A few case reports, including the present case, have demonstrated that anifrolumab can effectively treat cutaneous manifestations of Rowell Syndrome (RS) that is refractory to conventional therapies. However, little is known about the efficacy and safety of dual biologic therapy in SLE, and there have been no reported cases of combined rituximab and anifrolumab therapy to treat both cutaneous and CNS manifestations. Herein, we present a 42-year-old woman with refractory SLE with moyamoya syndrome and RS. She presented to the hospital with acute onset left leg weakness. Laboratory data showed elevated anti-double-stranded DNA (63 U/mL), low C3 (44 mg/dL), and low C4 levels (6 mg/dL). Digital Subtraction Angiography revealed severe stenosis of the right supraclinoid internal carotid artery (Fig. 1) and Magnetic Resonance Angiography showed increased collaterals arising from the posterior cerebral arteries (Fig. 2), findings compatible with moyamoya syndrome. She was initiated on pulse intravenous (IV) methylprednisolone 1 g daily for three days and one dose of IV rituximab 1 g, resulting in marked neurologic improvement. Two months later, the patient experienced a flare of her RS (Figs. 3, 4) and anifrolumab 300 mg monthly was initiated while she remained on rituximab 1g every 6 months. At five-month follow-up, she remained in clinical remission with normalization of her serologies. This case highlights the potential limitations of biologic monotherapy in the treatment of complex SLE manifestations caused by impairments in multiple pathways. Specifically, anifrolumab alone may be insufficient for treating CNS lupus while rituximab alone may not adequately control cutaneous lupus. This is the first case to our knowledge demonstrating the efficacy and safety of dual therapy with rituximab and anifrolumab in refractory SLE.

Teaching Point: Rituximab and anifrolumab combination therapy may be effective in SLE to target both CNS and cutaneous manifestations.
Category: Clinical Case

Figure 1: Digital subtraction angiography (DSA) images demonstrate progression of narrowing of the right supraclinoid internal carotid artery from approximately five months earlier (A, arrowhead) to the time of current admission (B, arrow).
Figure 2: Maximum intensity projection magnetic resonance angiography images of the head demonstrate increased collaterals arising from the posterior cerebral arteries (arrow).
Figure 3: Clinical images of Rowell Syndrome on the face before (A) and five months post-concomittant rituximab and anifrolumab therapy (B).
Figure 4: Clinical images of Rowell Syndrome on the palms before (A, B) and five months post-concomittant rituximab and anifrolumab therapy (C).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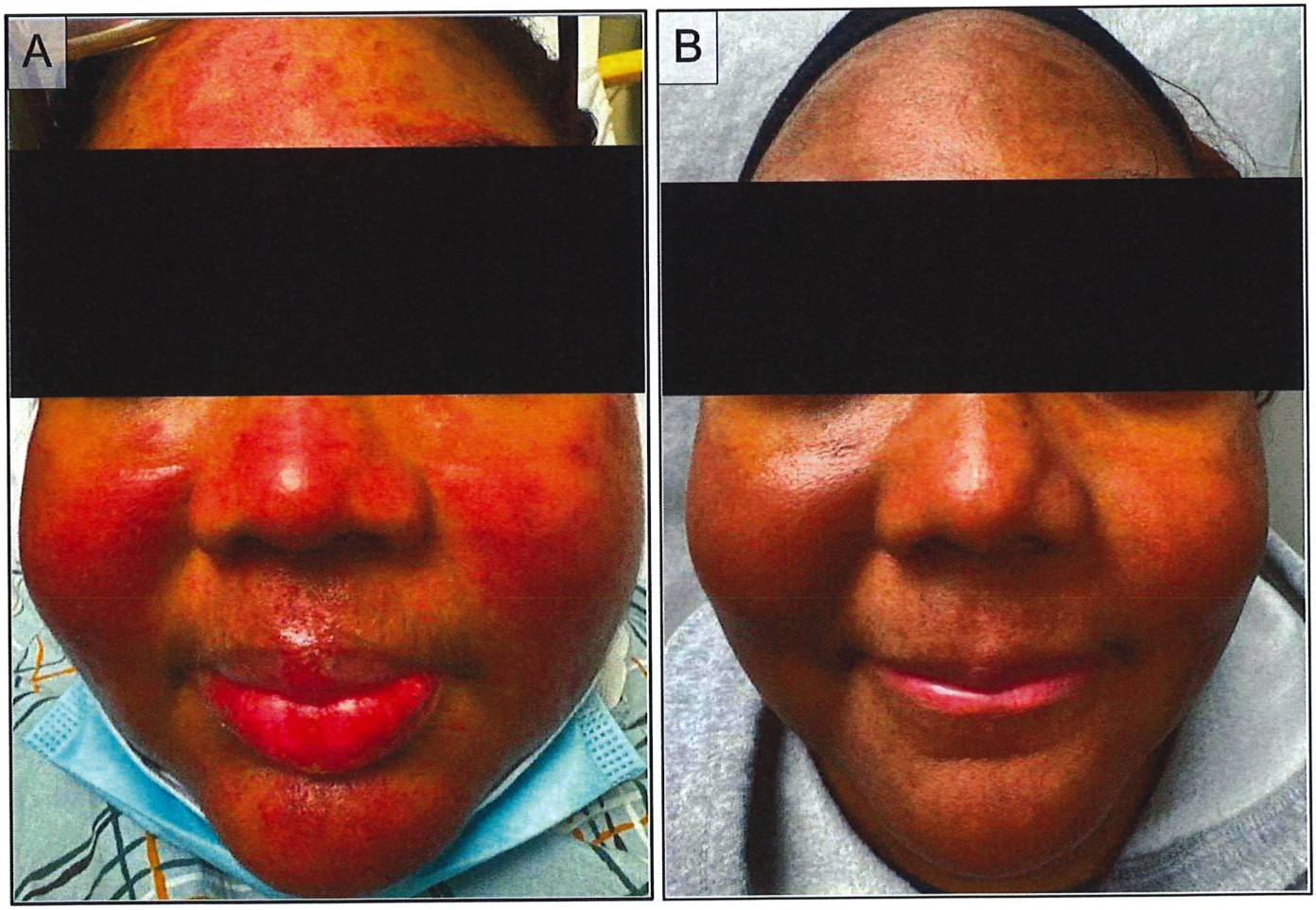
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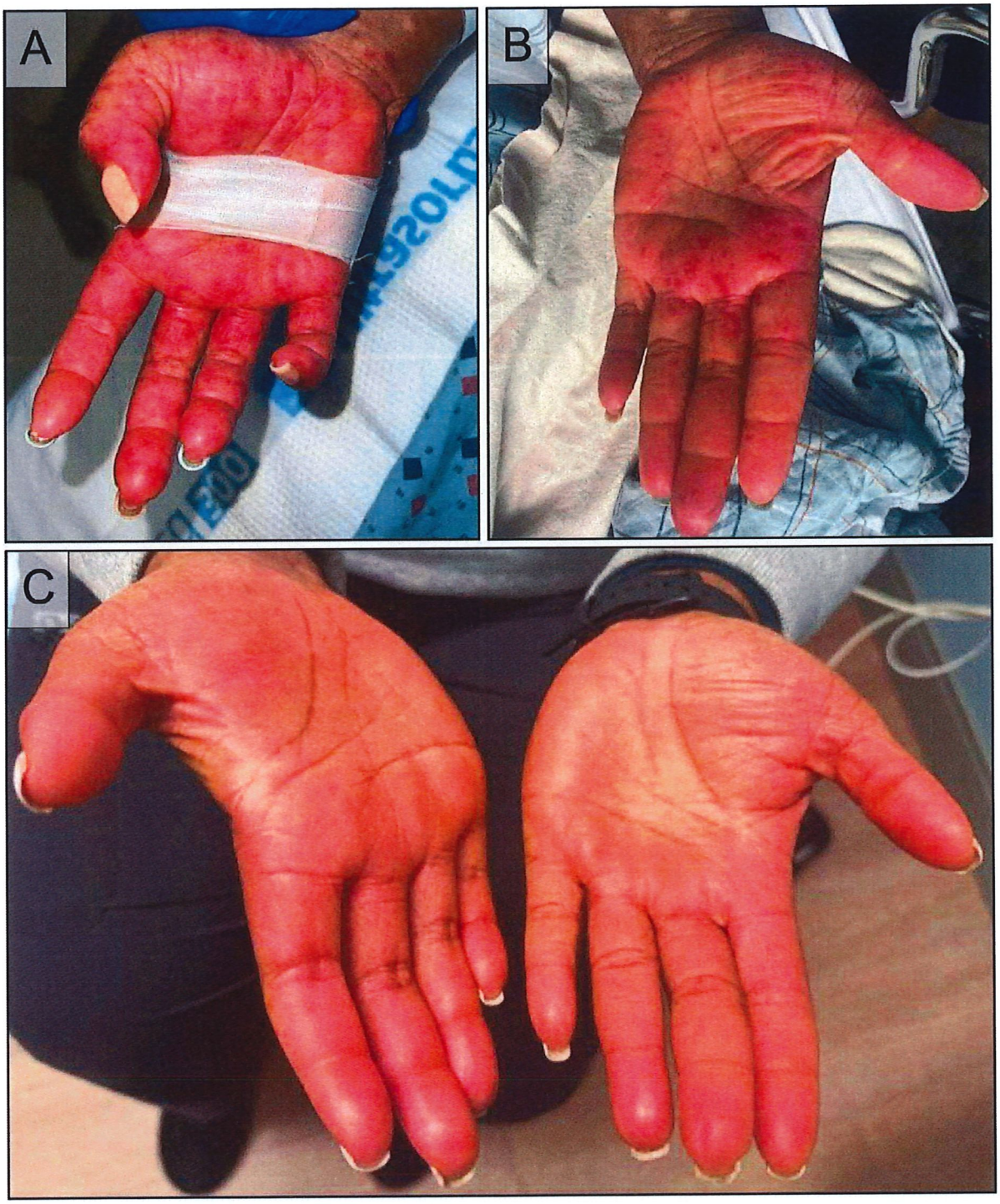
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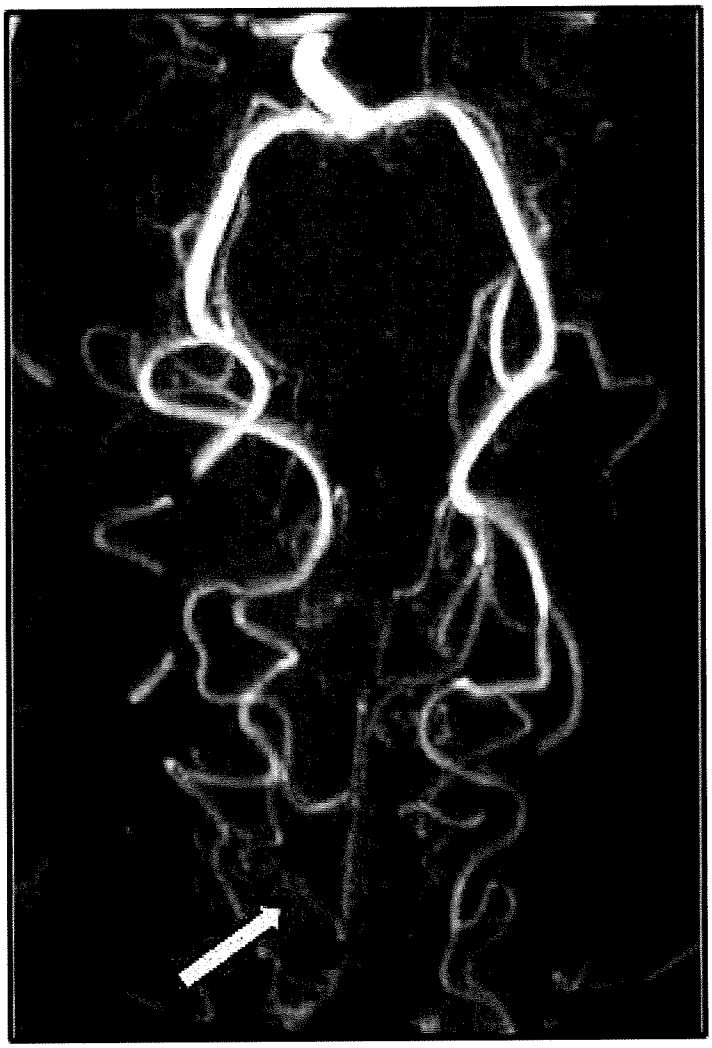

NECROBIOTIC XANTHOGRANULOMA PRESENTING ALONGSIDE CUTANEOUS VASCULITIS IN A PATIENT WITH LYMPHOPLASMACYTIC LYMPHOMA

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Clinical Case:

Necrobiotic xanthogranuloma (NXG) is a non-Langerhans cell histiocytosis associated with paraproteinemia from plasma-cell dyscrasia or lymphoproliferative disorders. We present the case of a 59-year-old female with lymphoplasmacytic lymphoma (LPL) who developed painful plaques on her lower extremities following a three-year history of cutaneous vasculitis. Initially, her vasculitis was treated for six months with colchicine. There was no improvement with this therapy; the patient reported worsening pain that increasingly hindered her ability to walk and work. She subsequently developed scattered indurated oval, brown/pink plaques on her inner thighs and shins. Punch biopsy from the left thigh for further workup revealed aggregates of epithelioid and bizarre multinucleate histiocytes with intermixed lymphocytes and eosinophils associated with necrobiosis. Though her lesions were not in the classic periorbital area, these histologic findings are consistent with NXG. Concern over possibility of an autoimmune neutropenia and low disease burden limited previous administration of bendamustine and rituximab or Bruton tyrosine kinase inhibitors (acalabrutinib) for her LPL. However, as the mainstay of management of NXG is treatment of underlying malignancy, the patient began treatment with rituximab 4x, weekly, alongside intralesional injections of triamcinolone. NXG can pose a challenge to treat as recurrence is likely, but prompt management of symptoms and surveillance for worsening hematologic malignancy is essential. Currently, 1 month after beginning treatment, the patient is responding well. Her lesions are smaller, less painful, and she has experienced minimal side effects. Though NXG is rare and this presentation is uncommon, this case underscores the need to include NXG in the differential diagnosis when new granulomatous skin lesions arise in patients with known hematologic malignancies or paraproteinemia.

Learning Point: This case highlights the importance of considering NXG as a potential diagnosis in patients with known hematologic malignancies or paraproteinemia, even when lesions are not located in the periorbital region.

Image:

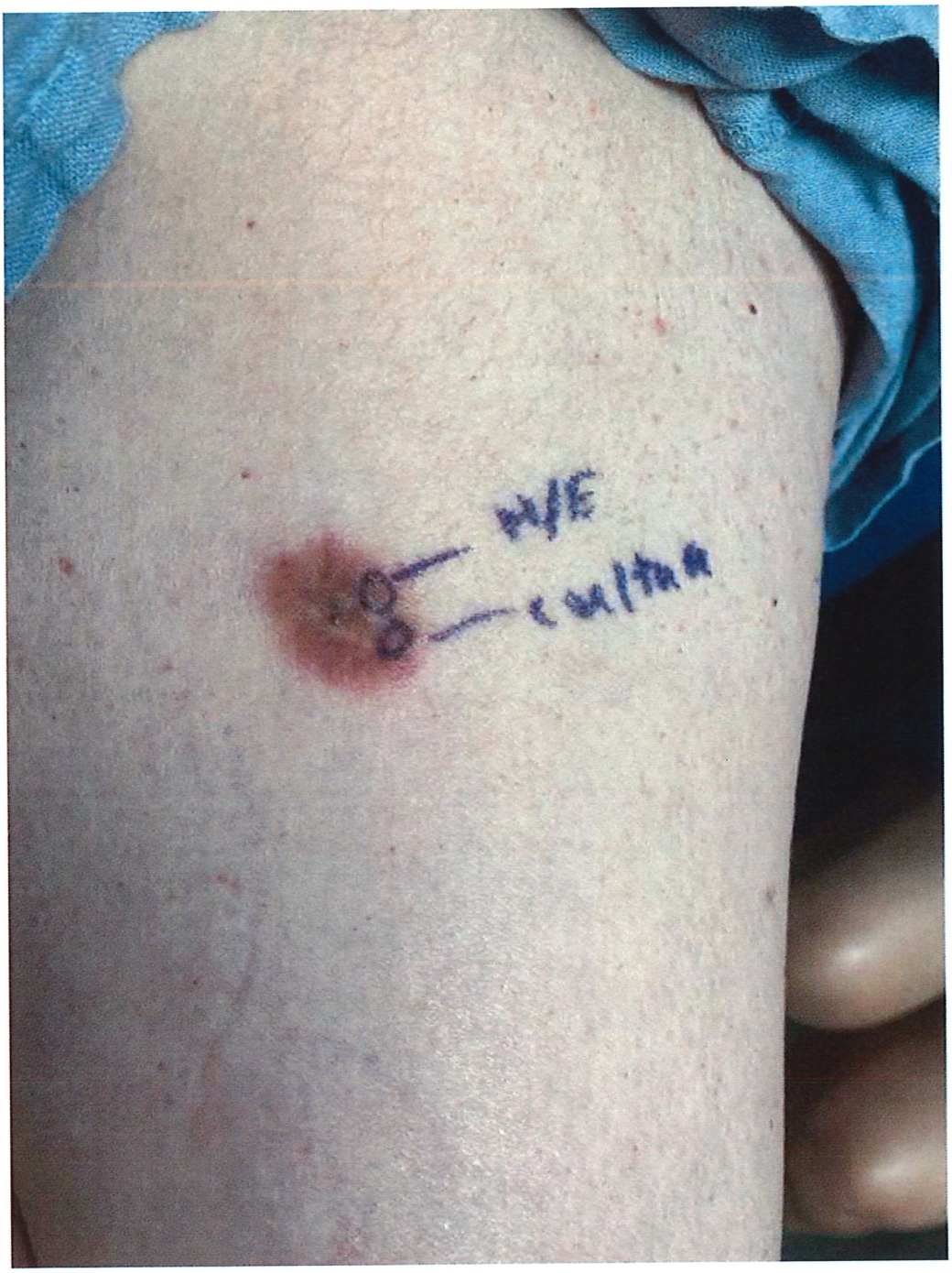

Figure 1. Indurated oval brown/pink papule on left thigh

ALOPECIA AREATA ASSOCIATED WITH BIMEKIZUMAB THERAPY

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Bimekizumab is a humanized monoclonal IgG1 antibody that selectively inhibits IL-17A and IL-17F, approved for psoriasis, psoriatic arthritis (PsA), axial spondylarthritis, and hidradenitis suppurativa. While candidiasis is its most common adverse event, alopecia areata (AA) has been rarely reported with other IL-17 inhibitors, such as secukinumab and ixekizumab. We report a case of diffuse AA in a 51-year-old woman with psoriasis and PsA following bimekizumab therapy. Previous treatments, including prednisone, methotrexate, and certolizumab, had been discontinued due to limited efficacy or adverse events. Certolizumab was withdrawn after 5 months due to an exacerbation of palmoplantar lesions (Figure 1). Bimekizumab was initiated at 320 mg subcutaneously every 4 weeks for 5 months, then spaced to every 8 weeks, leading to rapid improvement in both cutaneous and joint symptoms. Three months after initiating bimekizumab, the patient developed diffuse scalp hair loss (Figure 2A, 2B) and severe generalized pruritus, with no identifiable triggers. Dermatologic evaluation revealed diffuse non-scarring alopecia, a positive hair pull test and dermoscopic findings including black dots, broken hairs, circle hairs, and exclamation mark hairs (Figure 2C). Histopathology revealed a perifollicular lymphocytic infiltrate, follicular miniaturization, and an increased catagen/telogen ratio, consistent with AA (Figure 2D). Given the temporal association with bimekizumab and absence of other triggers, drug-induced AA was diagnosed. In a shared decision with the patient and due to the clinical response, bimekizumab was continued and topical corticosteroids prescribed with partial response. This case underscores the importance of individualized treatment decisions when managing new-onset adverse events like AA during biologic therapy for psoriatic disease. While bimekizumab offers substantial clinical benefits, its dual inhibition of IL-17A and IL-17F may disrupt immune regulation, activating pathways mediated by cytotoxic T cells and IFN-γ, implicated in AA. As the use of IL-17 inhibitors expands, recognizing paradoxical immune-mediated events such as AA remains essential.

Teaching Point: Alopecia areata represents a rare but potentially underrecognized paradoxical effect of IL-17 inhibitors.

Category: Clinical Case

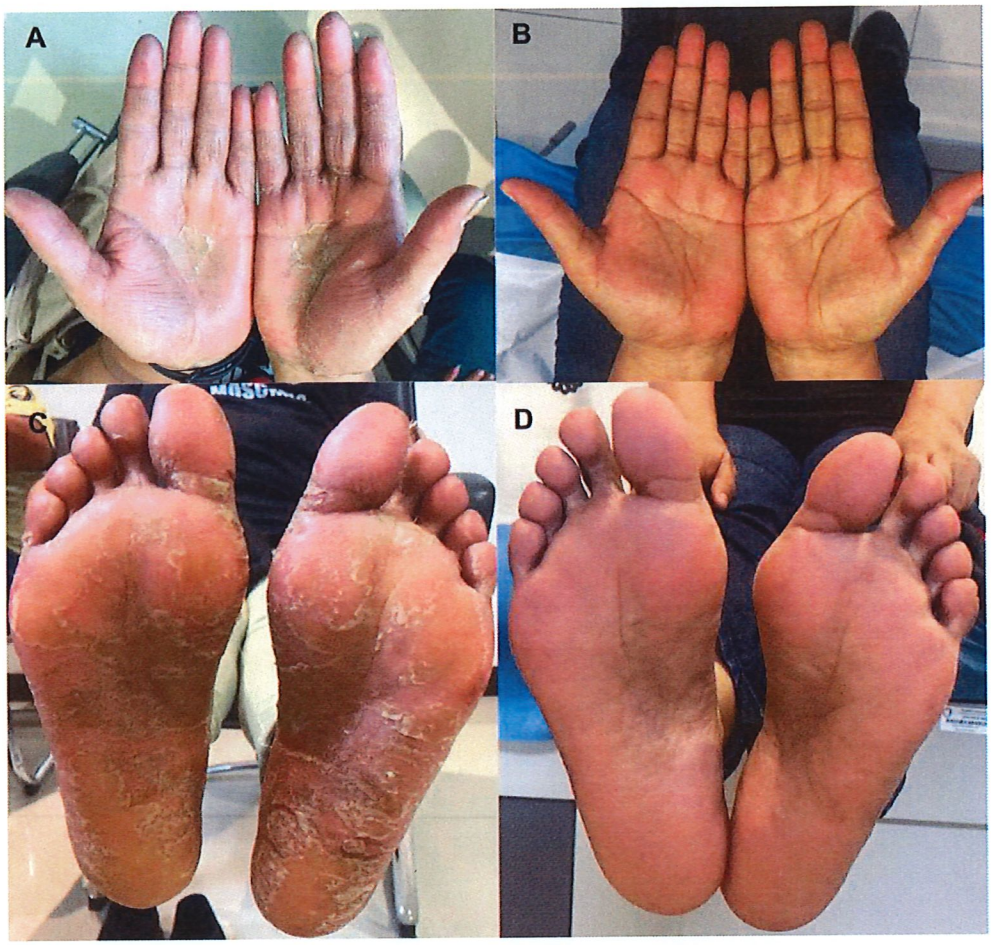

Figure 1. Palmar and plantar psoriasis before and after bimekizumab therapy. (A) and (C) Show palmar and plantar psoriasis at presentation. (B) and (D) Demonstrate resolution of the lesions after four months of bimekizumab treatment.

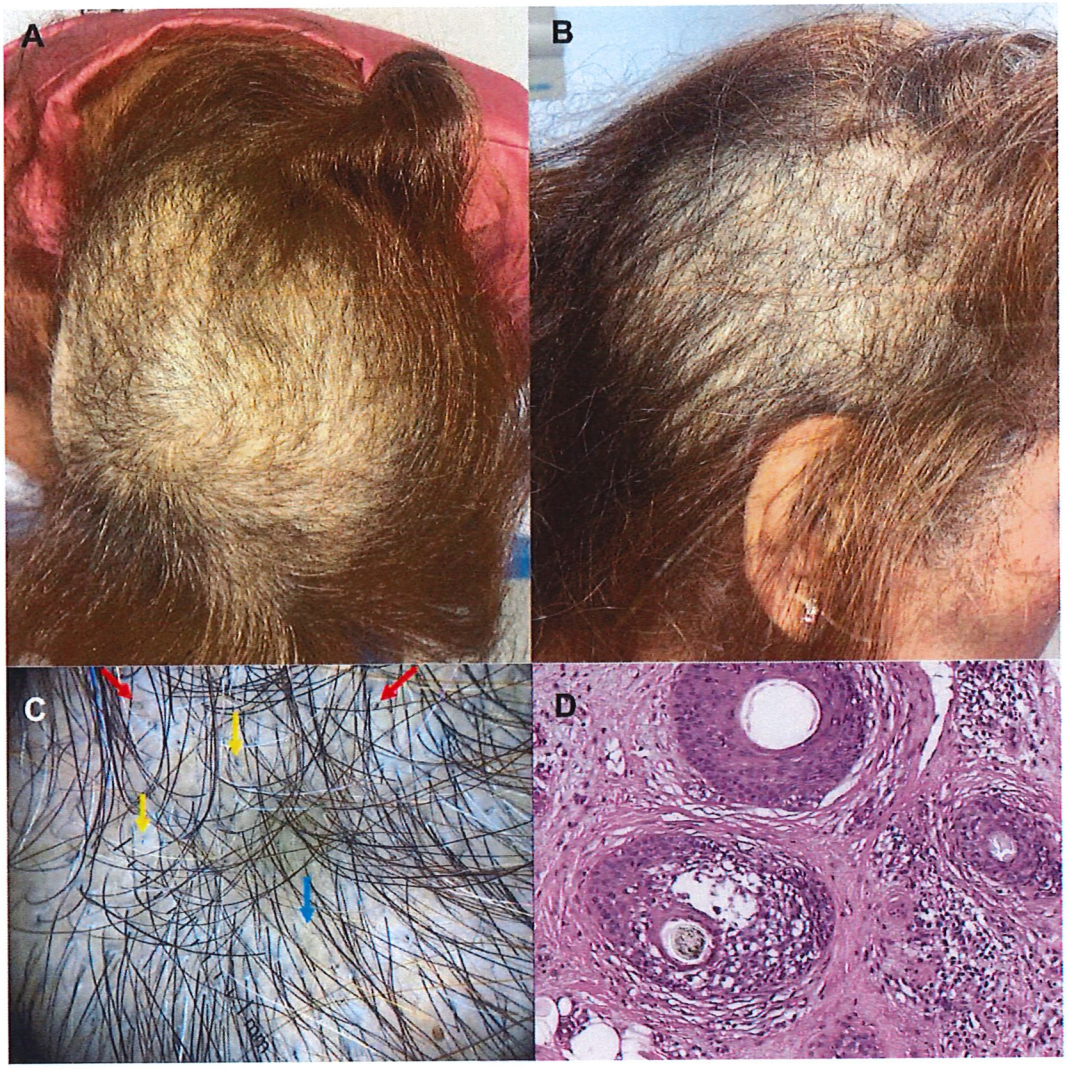

Figure 2. Alopecia areata during bimekizumab therapy Alopecia areata during bimekizumab therapy. (A) and (B) Shows diffuse non-scarring alopecia on the temporoparietal scalp. (C) Dermoscopy shows black dots and broken hairs (yellow arrows), circle hairs (red arrows), and exclamation mark hairs (blue arrows). (D) Histopathology examination revealing a perifollicular lymphocytic infiltrate with a "swarm of bees" appearance surrounding miniaturized hair follicles. (H&E, original magnification ×20).

BIMEKIZUMAB-INDUCED REMISSION IN REFRACTORY HURLEY STAGE III HIDRADENITIS SUPPARATIVA: A CASE REPORT

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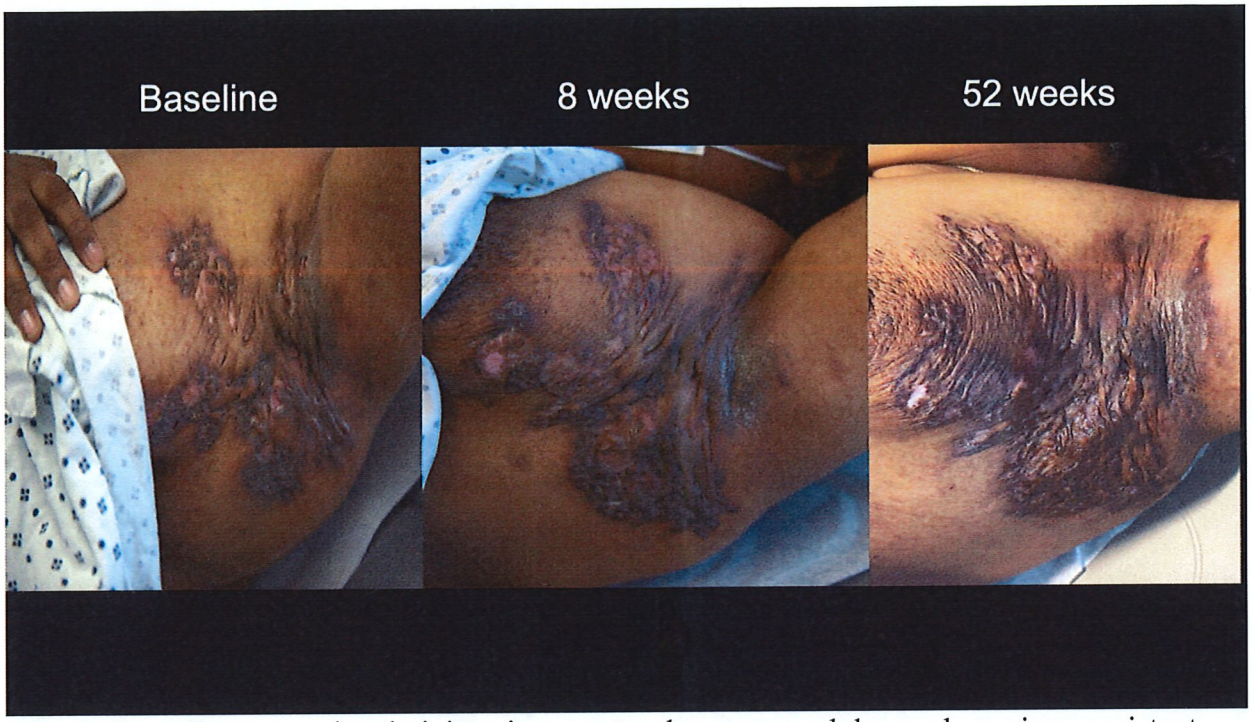
Hidradenitis suppurativa (HS) is a chronic, inflammatory skin disease characterized by recurrent nodules, abscesses, and draining sinus tracts, most often affecting intertriginous regions. It has a global prevalence of approximately 1%, with onset typically in early adulthood. HS pathogenesis is multifactorial, involving genetic predisposition, dysregulation of the immune system, and environmental risk factors such as obesity and smoking. Management is complex, including lifestyle modification, antibiotics, biologics, and surgery in advanced cases. However, therapeutic options remain limited for refractory disease. We present the case of a 24-year-old woman with Hurley Stage III HS who failed multiple systemic and biologic therapies, including adalimumab, infliximab, and secukinumab. She presented with severe pain, malodorous drainage, and extensive sinus tracts and nodules across the axillae, inframammary folds, groin, and thighs. Given her refractory disease, bimekizumab 320 mg every four weeks was initiated in April 2024. By two weeks, she reported decreased pain and drainage, supported by ultrasound evidence of reduced inflammation. At 12 weeks, she achieved clinical remission with no active nodules and sustained improvements in pain and quality of life. This remission was maintained for 52 weeks, with no new lesions and no need for daily wound care, though scarring and sinus tracts persisted. She experienced no adverse effects during treatment. Ultrasound with grayscale and power Doppler imaging provided valuable objective evidence of treatment response, detecting reductions in subclinical inflammation and vascularity not fully appreciable on physical examination. This case highlights the effectiveness of dual IL-17A/F inhibition with bimekizumab in treatment-resistant HS and supports its potential as a promising therapeutic option in severe disease.

Teaching Point: High-frequency ultrasound is a valuable adjunct for tracking therapeutic response to bimekizumab in hidradenitis suppurativa.

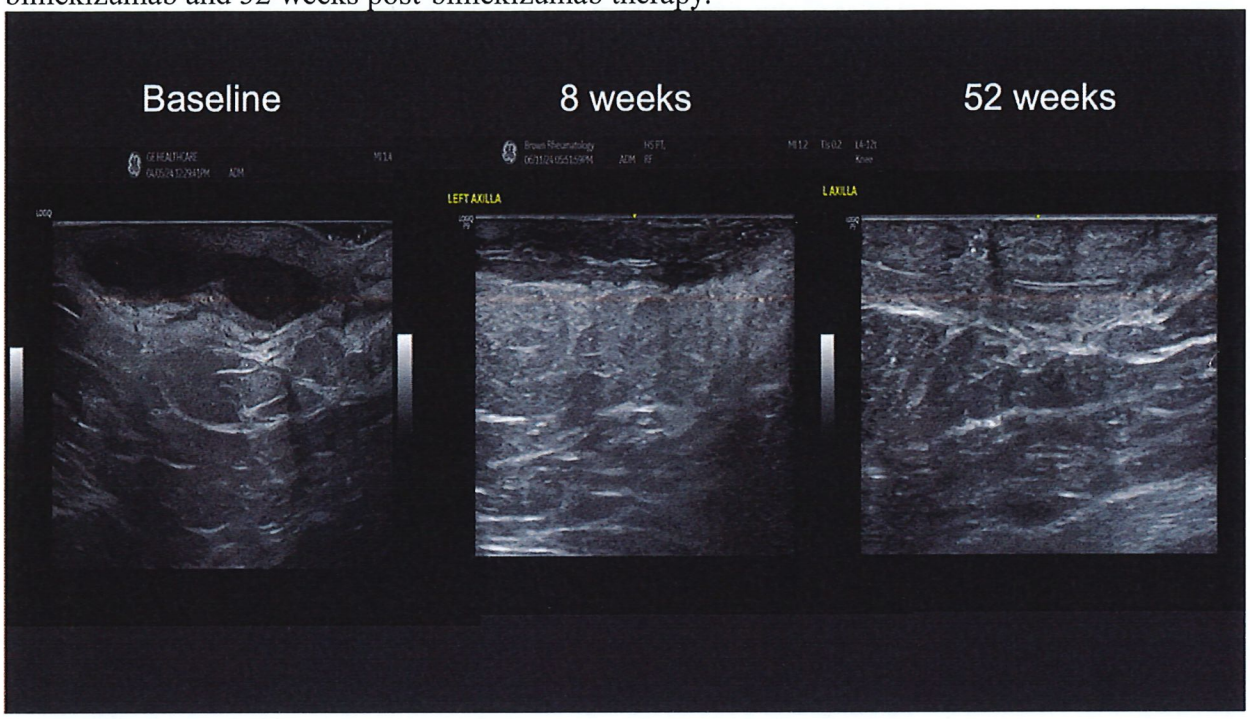
Category: Clinical Case

Figure 1: Clinical images of the left axilla at baseline, 8 weeks post-bimekizumab and 52 weeks post-bimekizumab therapy.



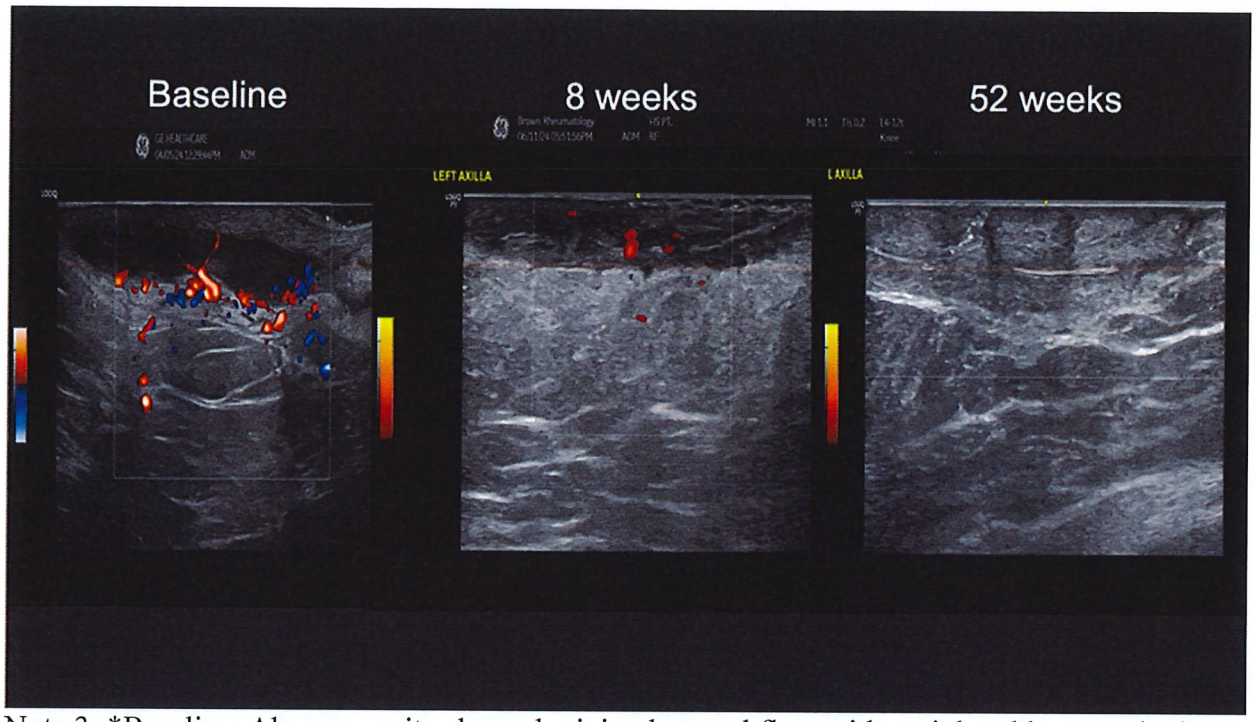
Note 1: *Baseline: extensive draining sinus tracts, abscesses, nodules, and scarring consistent with Hurley Stage III, **8 weeks: reduced inflammation and reduced abscesses/ nodules, ***52 weeks: Stable, inactive disease pattern with resolved abscesses/nodules but persistent scarring.

Figure 2: High frequency grayscale ultrasound images of the left axilla at baseline, 8 weeks post-bimekizumab and 52 weeks post-bimekizumab therapy.



Note 2: *Baseline: hypoechoic oval structures indicate abscesses, **8 weeks: hypoechoic tubular structure representing a sinus tract, ***52 weeks: Clinical remission; inactive nodules and resolved abscesses BUT persistent scarring and sinus tracts (hypoechoic tubular structures with internal echoes due to debris).

Figure 3: High frequency power doppler ultrasound of the left axilla at baseline, 8 weeks post-Bimzelx and 52 weeks most recent.



Note 3: *Baseline: Abscess cavity showed minimal central flow with peripheral hyperemia that indicates active inflammation, **8 weeks: reduced inflammation and minimal peripheral hyperemia, ***52 weeks: Chronic fibrotic tunnels without doppler flow are indicative of stable, inactive disease.

BREAKING THE CYCLE: SUCCESSFUL LONG-TERM REMISSION OF CHRONIC ERYTHEMA MULTIFORME WITH RITUXIMAB THERAPY

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Abstract

Erythema multiforme (EM) is an immune-mediated hypersensitivity reaction characterized by classic target lesions, with severe forms involving mucosal sites that distinguish EM major from minor. Although most cases are self-limiting and resolve within weeks, chronic and refractory EM may require systemic immunosuppression. Infections are implicated in 90% of cases, most commonly herpes simplex virus, with Mycoplasma pneumoniae as another frequent trigger. Management centers on treating the underlying cause, and while antivirals, thalidomide, and dapsone may provide benefit, treatment-resistant EM remains challenging. We present the case of a 37-year-old man with chronic, refractory EM who failed multiple therapies, including prednisone, hydroxychloroquine, doxycycline, methotrexate, dapsone, apremilast, cyclosporine, thalidomide, and IVIG. Despite temporary improvements, he experienced persistent flares over several years, with biopsy and immunofluorescence confirming EM. In June 2019, rituximab infusions (1000 mg IV × 2, two weeks apart) were initiated following negative hepatitis serology and exclusion of hematologic malignancy. Remarkably, the patient achieved complete remission within weeks of treatment, enabling discontinuation of prednisone and thalidomide. At follow-up in December 2019, he remained lesion-free, and this remission was sustained through December 2024, over five years after initiation, with no further systemic therapy required. Laboratory evaluation showed persistently elevated Mycoplasma IgG but negative IgM, suggesting remission was not dependent on active infection. This case underscores rituximab's role as an effective therapy for refractory EM, consistent with limited prior reports documenting its efficacy in patients unresponsive to conventional treatment. By achieving durable remission after multiple therapeutic failures, rituximab highlights the promise of CD20-targeted therapy in EM and supports its consideration in select, treatment-resistant cases.

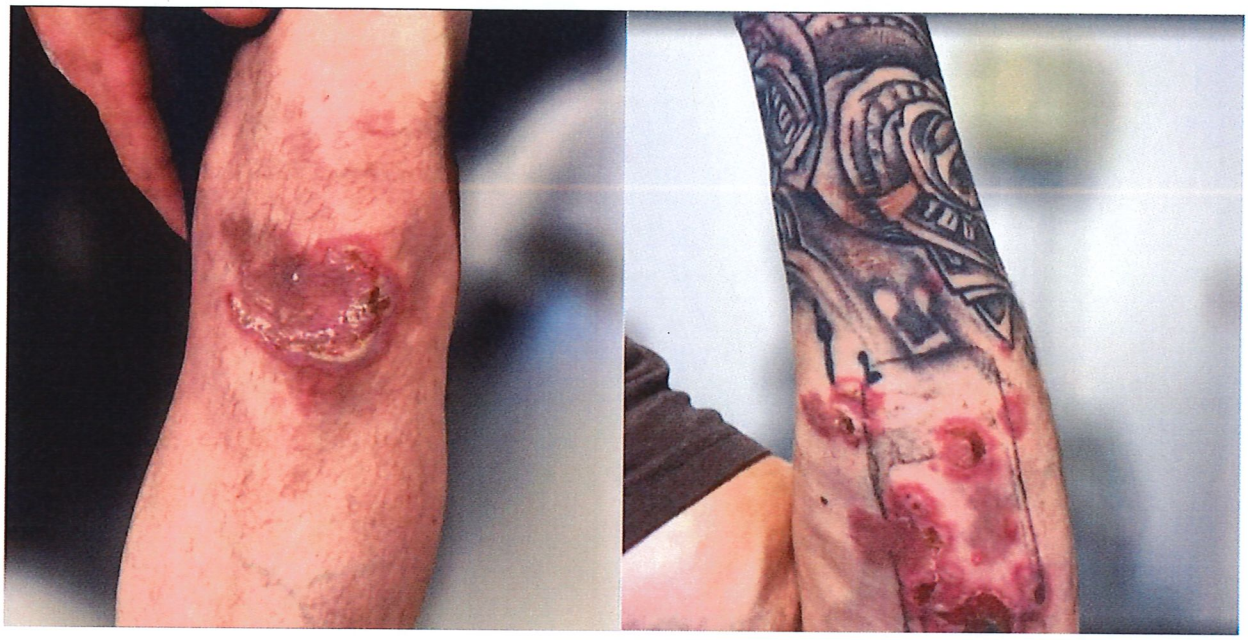
Teaching Point: Erythema multiforme (EM) is typically a self-limited condition, but chronic, refractory cases may require systemic immunosuppression. Rituximab has demonstrated benefit in limited reports, though evidence remains sparse. We describe a case of treatment-resistant EM that achieved sustained remission for over five years following rituximab therapy, despite failure

of multiple prior systemic agents. This case highlights rituximab as a viable option for refractory EM and supports the therapeutic potential of CD20-targeted therapy in select patients.

Category: Clinical Case

Figures 1 and 2 depict images of the patient's upper and lower extremities before and after initiating rituximab therapy.

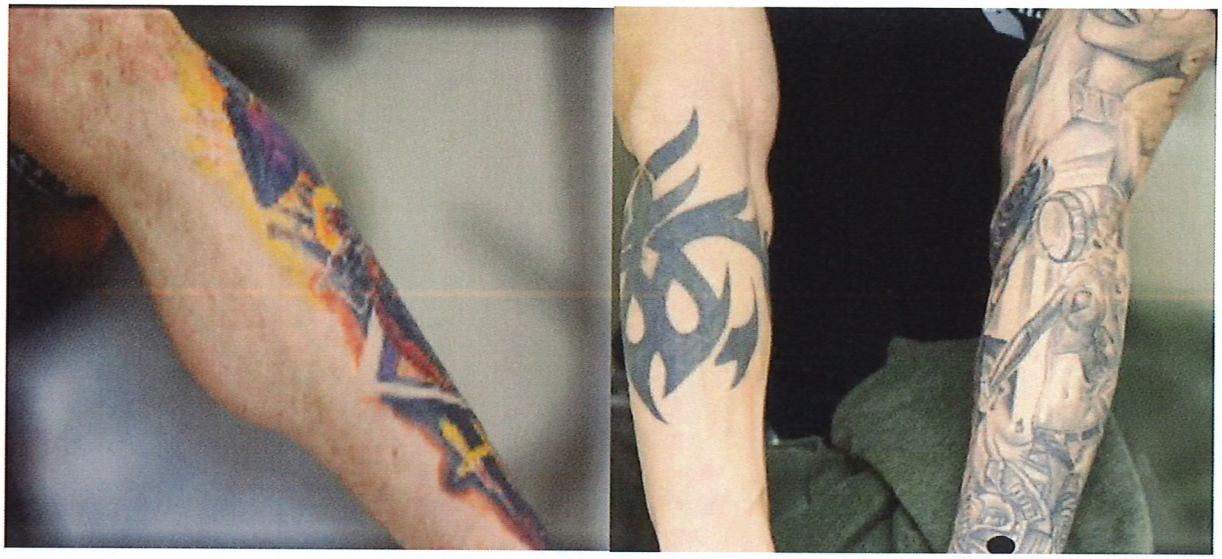
Figure 1.



Top left: Erythematous, well-demarcated, annular plaque with overlying scale and crust on the left knee.

Top right: Multiple erythematous, well-demarcated, annular, and arcuate plaques with overlying scale and hemorrhagic crust on the right forearm.

Figure 2.



Bottom left and right: Resolution of lesions with clear, rash-free skin on the left leg and bilateral upper extremities following

Photodamage or Dermatomyositis? A Case of an Erythematous Rash in a 59-Year-Old Patient

s Maya Hamaker, MPH[2], Rabindra Dhakal, MD[3] , Mohammad Maruf, MD[3] and Gita Vatandoust, MD[3]

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Abstract:
Dermatomyositis (DM) is a subtype of idiopathic inflammatory myopathy (IIM), a rare autoimmune disorder characterized by chronic muscle inflammation and weakness. These conditions significantly impair quality of life due to their multisystem involvement. DM presents with progressive, proximal, symmetrical weakness of the upper or lower extremities. The presence of distinct cutaneous findings, including Gottron papules, periorbital edema, heliotrope rash, and V-sign or shawl sign rashes, distinguishes DM from other IIMs. Laboratory findings often include elevated creatine kinase and liver enzymes, indicative of muscle damage. The incidence of DM is estimated to be 1 to 6 per 100,000 individuals in the U.S., with a higher prevalence in African Americans. Risk factors include female sex, genetic predisposition, ultraviolet radiation, and environmental factors. Autoantibodies play a crucial role in diagnosis, with anti-Mi-2 antibodies found in 5-10% of cases, often correlating with higher creatine kinase levels and more prominent weakness. We present the case of a 59-year-old female with diabetes mellitus and hypertension who was admitted from a nursing home with a three-month history of progressive weakness, bilateral leg swelling, and a rash. She initially presented with palpitations, dyspnea, and lower extremity edema, leading to evaluations by cardiology, pulmonology, and internal medicine. Her symptoms worsened over subsequent admissions and autoimmune workup eventually revealed a positive antinuclear antibody (ANA) with a titer of 1:1280 and a speckled pattern, as well as the presence of Mi-2 antibodies. The patient developed dysphagia, distal cutaneous erythema, and significant weight loss. Treatment with intravenous glucocorticoids and intravenous immune globulin led to clinical improvement, and she was discharged to a subacute rehabilitation facility.

Teaching Point: Early recognition of dermatomyositis in patients with skin of color is critical, as atypical rash presentation may be mistaken for photodamage contributing to diagnostic delays.

Abstract category: Clinical Case

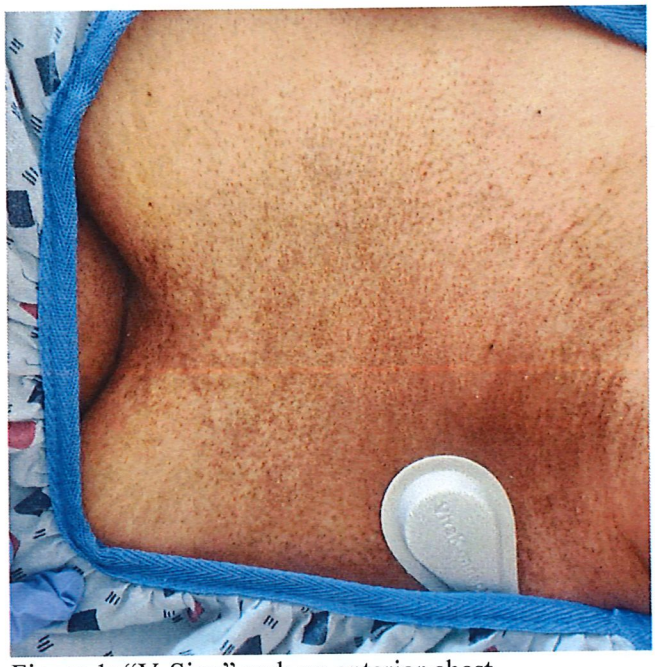

Figure 1: “V-Sign” rash on anterior chest.

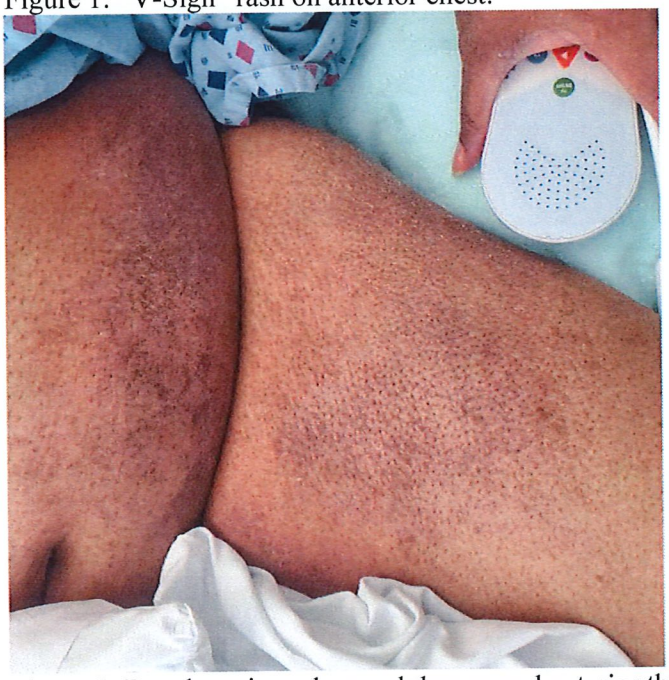

Figure 2: Dyschromia on lower abdomen and anterior thigh

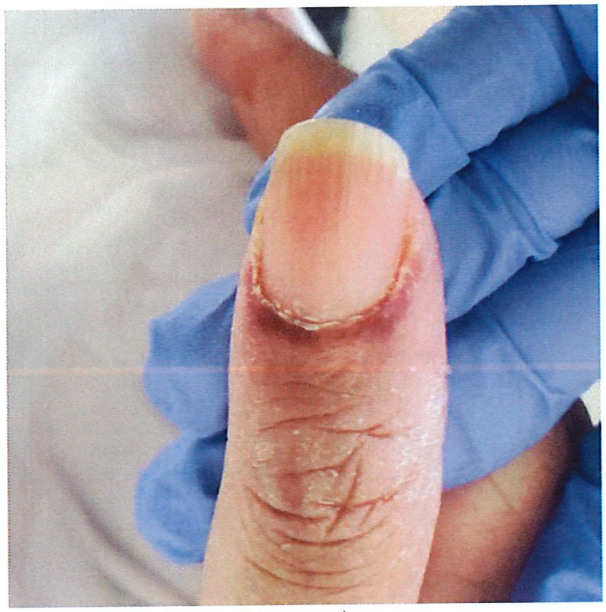

Figure 3. Periungual hemorrhages

SCLEROMYXEDEMA PRESENTING WITH GOTTRON-LIKE PAPULES

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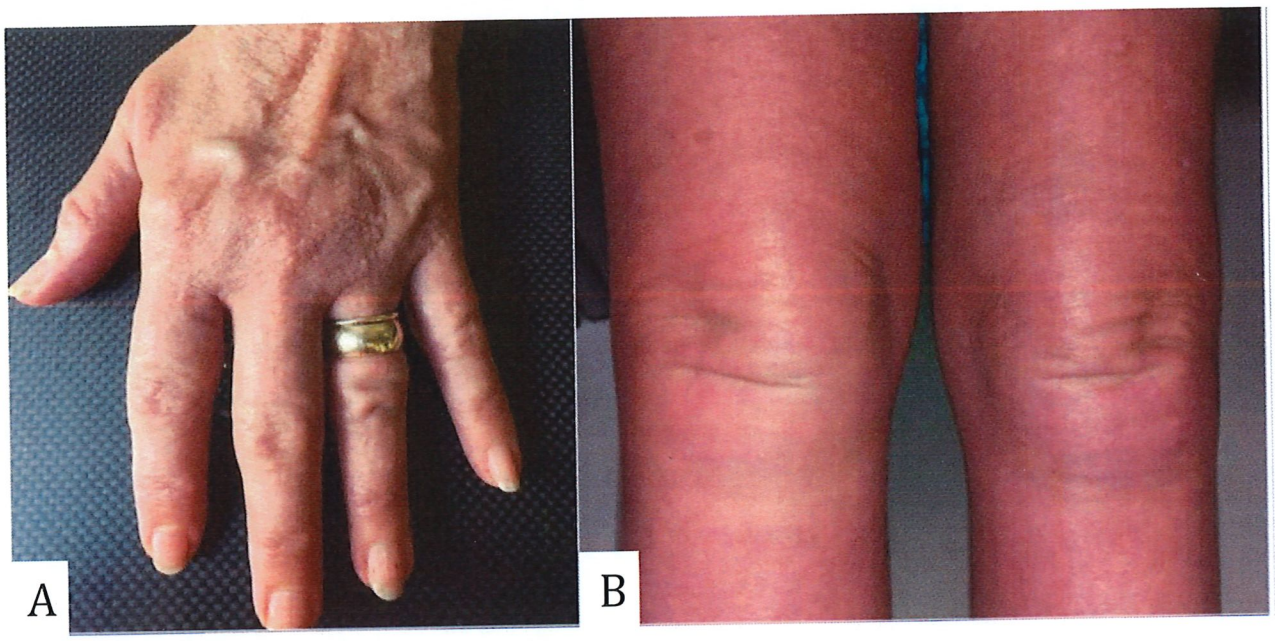
Scleromyxedema is a rare cutaneous disorder of unknown etiology and is considered the generalized form of lichen myxedematosus. It typically presents with generalized papular and sclerodermoid eruptions characterized by mucin deposition in a thickened dermis, increased fibroblast proliferation with fibrosis, and an associated monoclonal gammopathy.[1,2] We present the case of a 72-year-old female with a history of monoclonal gammopathy of undetermined significance (MGUS) who was referred to dermatology for a nine-month history of pain in hands and pruritic papules on hands and forearms with suspicion of dermatomyositis (DM)/DM overlapping with other connective tissue diseases. With suspicion of DM, she had been trialed on hydroxychloroquine, oral steroids, and was on weekly methotrexate at the time of presentation. Prior biopsies at an outside dermatology clinic were interpreted as granuloma annulare. On presentation, firm, flesh-colored to pink papules were noted over the metacarpophalangeal joints, resembling Gottron papules. In addition to these papules, there was thickening of the skin on the dorsum of the hands, as well as scattered papules on the forearms, upper back, and chest, with many areas showing a linear configuration to the papules. Laboratory evaluation revealed an IgG lambda monoclonal spike, ANA 1:80 speckled, and otherwise unremarkable myomarker panel. Punch biopsy of the right chest demonstrated dense dermal spindle cell proliferation with mucin deposition, consistent with scleromyxedema. The patient was started on prednisone 40 mg daily and continued the methotrexate 15 mg weekly, with plans for intravenous immunoglobulin (IVIG) pending insurance approval. This case highlights how scleromyxedema can mimic connective tissue diseases, especially DM in this case with Gottron-like papules. Given its strong association with monoclonal gammopathy and potential for systemic complications, dermatologists should maintain a high index of suspicion for scleromyxedema in papular eruptions over joints and promptly pursue biopsy for diagnostic confirmation.

Teaching Point: Scleromyxedema should be considered in patients with generalized papular eruptions and monoclonal gammopathy, particularly when lesions present as skin-colored papules in a linear arrangement with waxy induration to distinguish it from mimicking autoimmune connective tissue diseases such as dermatomyositis, scleroderma, lupus, and eosinophilic fasciitis, and guide timely diagnosis.

Abstract Category: Clinical Case

Figure 1: (A) Skin-colored papules over the metacarpophalangeal and interphalangeal joints. Careful examination reveals thickening of the background skin, particularly pronounced on the ring finger, creating a "donut sign" over the proximal interphalangeal joint. (B) Confluent violaceous erythema of the lower extremities with deep furrows over the knees due to skin thickening, producing a "Shar-Pei sign."



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1. Cokonis Georgakis, C.-D., Falasca, G., Georgakis, A., & Heymann, W. R. (2006). Scleromyxedema. *Clinics in Dermatology, 24*(6), 493-497. doi:https://doi.org/10.1016/j.clindermatol.2006.07.011
2. Pomann, J. J., & Rudner, E. J. (2003). Scleromyxedema revisited. *International Journal of Dermatology, 42*(1), 31-35. doi:https://doi.org/10.1046/j.1365-4362.2003.01565.x

Underlying Lymphoproliferative Disorder in Schnitzler's Syndrome– Report of One Case Requiring Daratumumab for Underlying Myeloma

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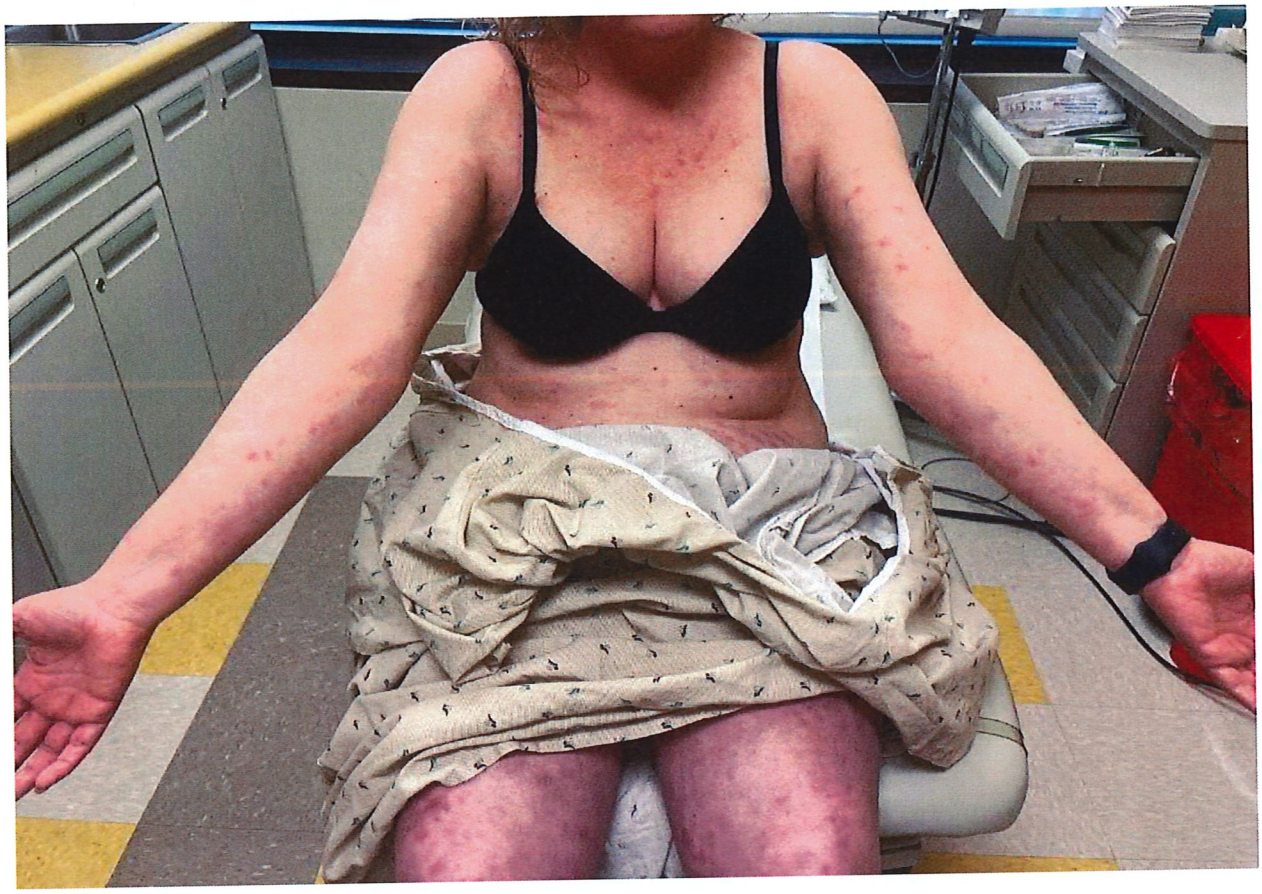
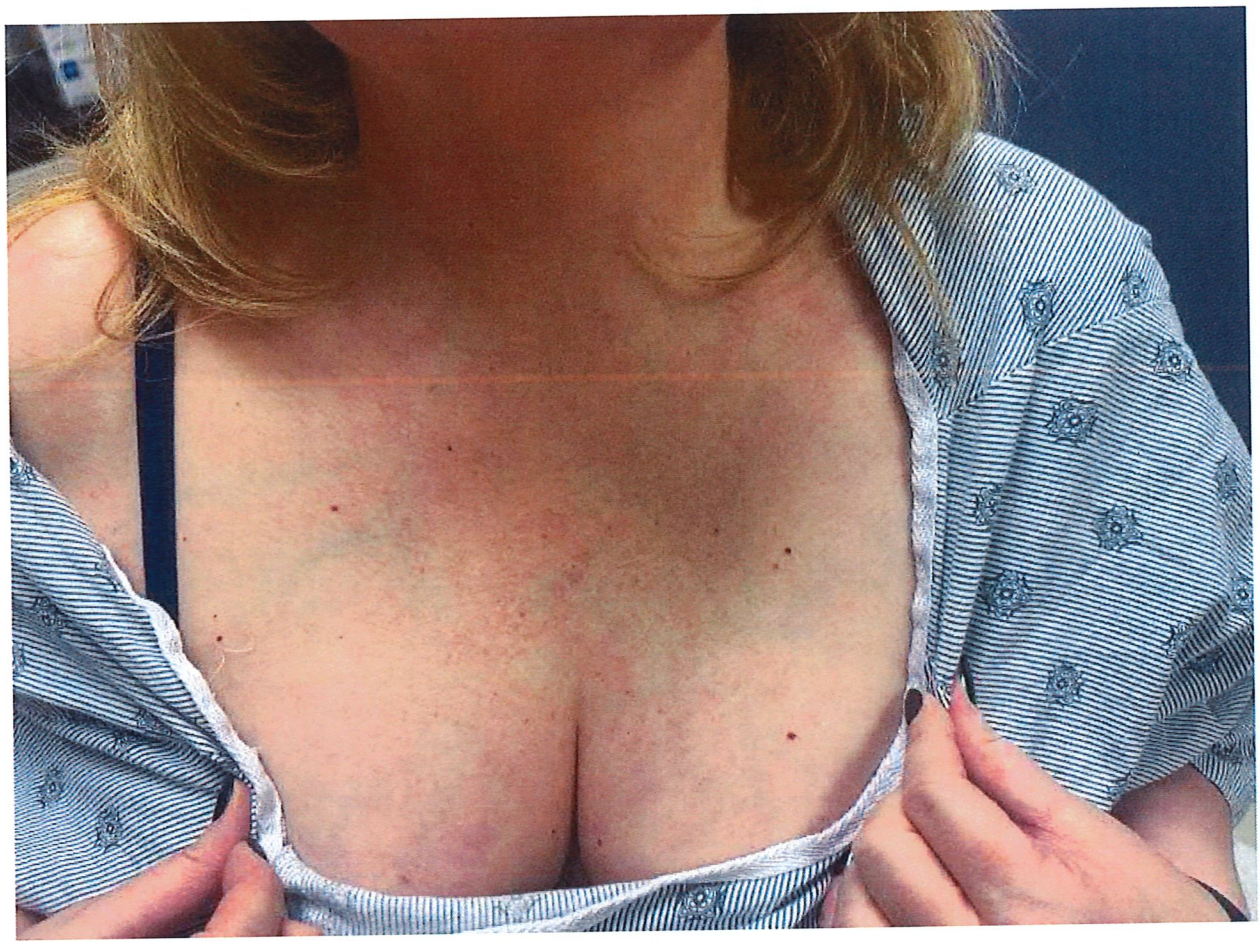
Schnitzler's syndrome (SS) is a rare acquired autoinflammatory syndrome characterized by recurrent, non-pruritic urticarial lesions, intermittent fevers, bone pain, arthralgia, increased ESR, and monoclonal gammopathy. Though its pathogenesis is not well understood, it is theorized that systemic overproduction of IL-1β by dermal mast cells results in loss of anti-inflammatory Th17 cell activity. Additionally, IL-6 levels correlate positively with disease activity. Treatment is typically with IL-1 blockade, such as anakinra, or IL-6 blockade in refractory cases.

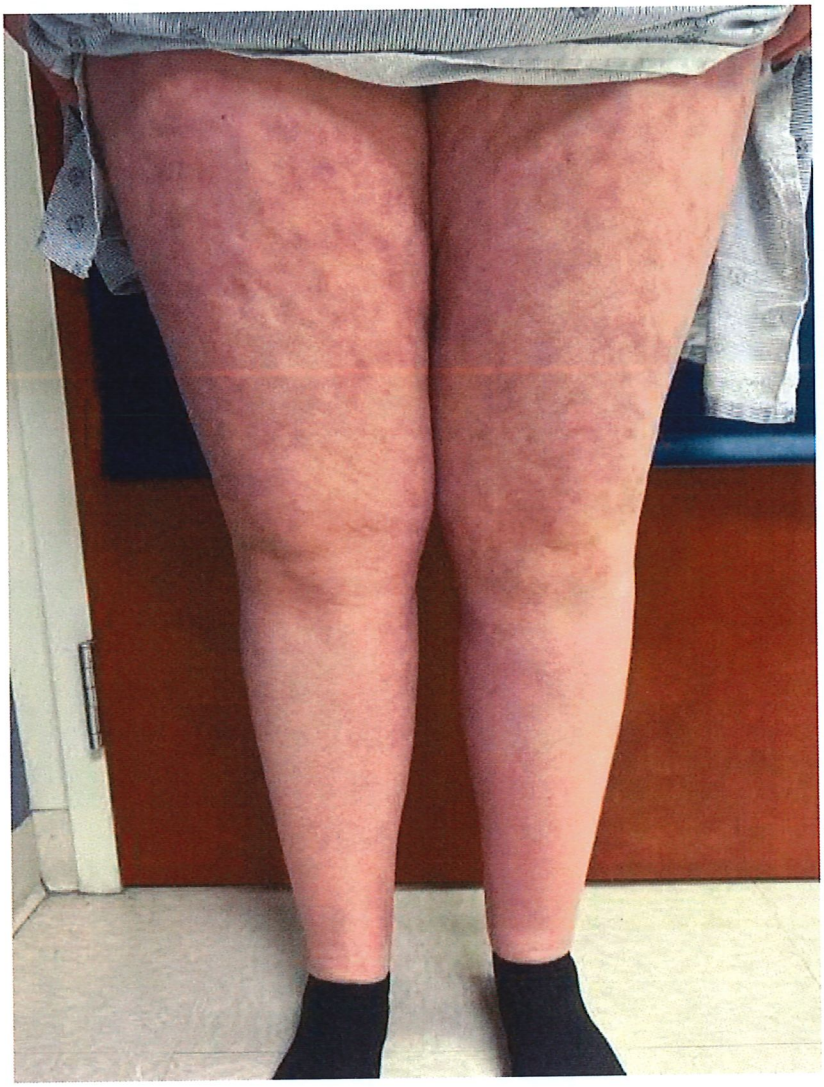
A 44-year-old female presented to dermatology with a 5-year history of refractory urticarial lesions, night sweats, fevers, joint pains, muscle weakness, and fatigue. Physical exam revealed edematous red papules with rim of pallor, red-brown patches, thin plaques, and purpuric patches distributed on the chest, arms, and legs. Past oncologic evaluation was significant for serum protein electrophoresis (SPEP) with M-Spike 0.4 g/dL (not observed g/dL), IgG monoclonal protein with lambda light chain specificity, and normocellular bone marrow biopsy (BMB). Histopathology demonstrated perivascular and interstitial infiltrate composed of numerous neutrophils and histiocytes without significant karyorrhexis or fibrinoid necrosis, most consistent with urticarial dermatosis. With these findings, SS with neutrophilic urticarial dermatosis and monoclonal gammopathy of unknown significance (MGUS) were diagnosed. Despite various treatments (canakinumab 300mg/mL every 4 weeks, colchicine 0.6mg daily, and tocilizumab 162mg/0.9mL weekly), symptoms persisted. At follow-up, elevated IL-6 (123 pg/mL, reference < 5 pg/mL) was found despite tocilizumab treatment. She was referred to a myeloma specialist given high suspicion for concurrent lymphoproliferative disease. Repeat BMB showed 1-2% monotypic lambda-restricted plasma cells (CD38+, CD138+, CD19-) indicative of a plasma cell neoplasm, and the patient was diagnosed with myeloma. She was transitioned to daratumumab 18,000mg/15mL weekly for 4 months, subsequently daratumumab 18,000mg/15mL and bortezomib 2.7mg/1.08mL biweekly for 4 months, and currently requires therapy monthly. She has remained without significant skin flare since initiation of myeloma-targeting therapy.

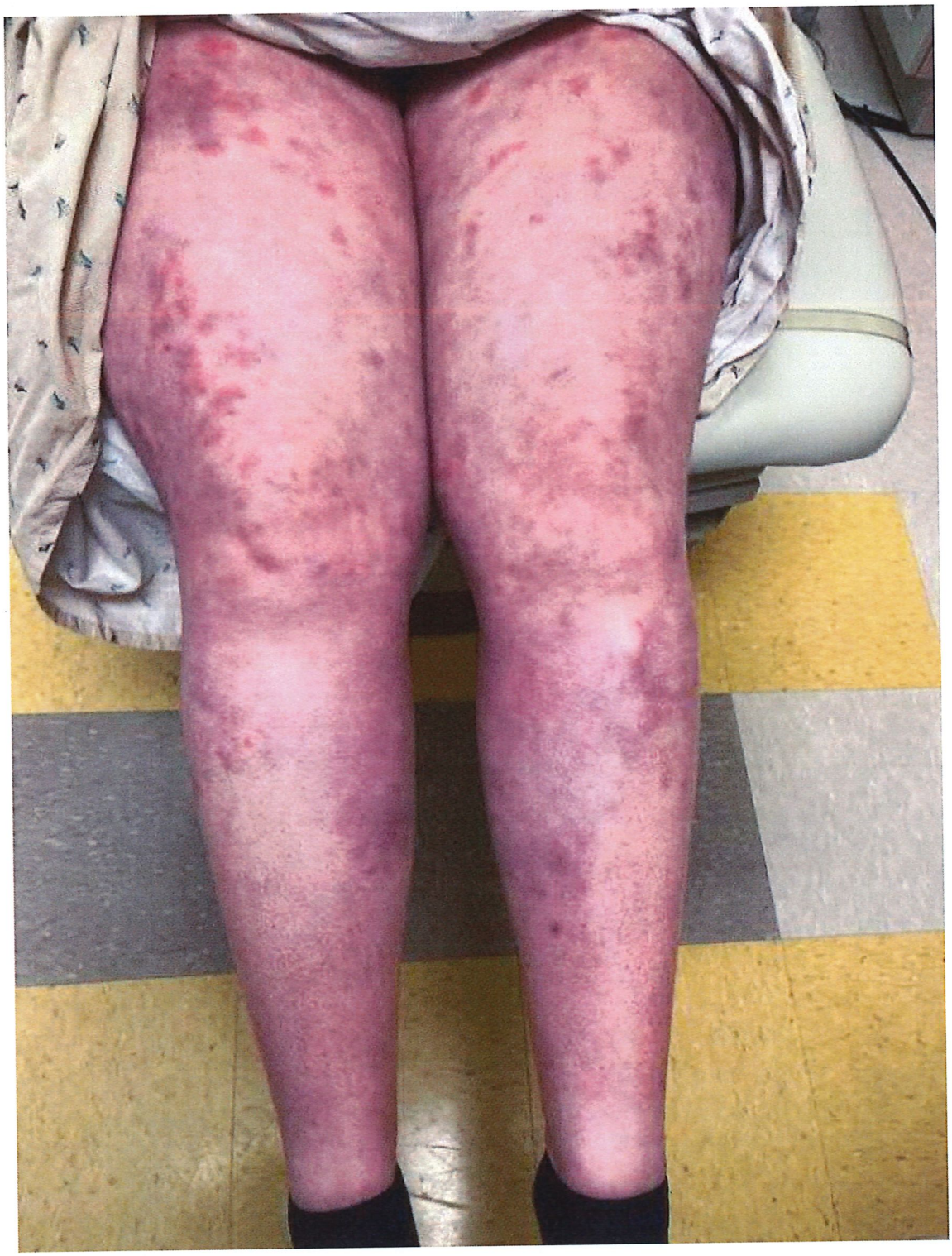
Teaching Point: Patients with Schnitzler's syndrome should undergo long term surveillance for lymphoproliferative disorders, and treatment of these disorders is recommended once identified.

Abstract Category Clinical Case









THE CURIOUS CASE OF ERYTHEMA NODOSUM AND A HORMONAL CULPRIT

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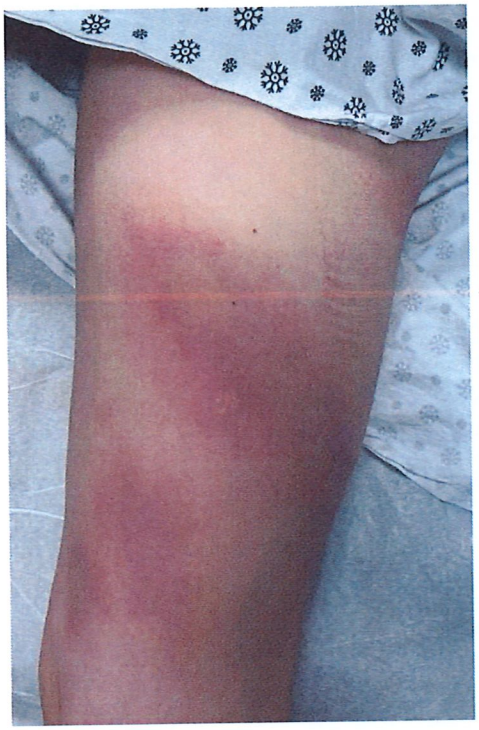
Erythema nodosum (EN), the most common panniculitis, may be trigged by hormonal contraception and menses. Despite this association, hormonal treatment of EN has not been previously reported to our knowledge. We describe the case of a 25-year-old woman with biopsy-consistent chronic EN refractory to traditional therapies (Figure 1). Since age seven, painful nodules developed on her extremities approximately every three months, associated with fatigue and fevers. Flares were triggered by menses, combined oral contraceptives (COCs), stress, and infections, with menses and COCs being the most severe triggers. Past treatments—including non-steroidal anti-inflammatory drugs, prednisone, potassium iodide, colchicine, dapsone, hydroxychloroquine, and methotrexate—resulted in an incomplete response. Upon presentation, the patient was using a levonorgestrel 52 mg intrauterine device (IUD) for contraception. Skin biopsies were repeated and consistent with EN. Further workup was unremarkable, including a normal or negative anti-streptolysin O antibody titer, angiotensin converting enzyme level, QuantiFERON Gold, fecal calprotectin level, peripheral flow cytometry, genetic testing for autoinflammatory syndromes, whole exome sequencing, and chest computed tomography scan. Given the patient's history of flaring with menses and COCs, an association with progesterone, which typically peaks one week before menses and is artificially elevated by COCs (but not IUDs), was suspected. As a result, spironolactone 100 mg/day was initiated and, due to continued flares, increased to 150 mg/day. This dose resulted in EN resolution for ten months despite menses and stress. The patient then flared following an upper respiratory tract infection, which prompted up-titration of spironolactone to 100 mg twice daily. At the time of writing, her EN has remained quiescent for five months on this dose. In conclusion, EN may be induced by elevations in progesterone levels, and spironolactone may inhibit this trigger. This case supports consideration of spironolactone as a potential therapeutic option for patients with progesterone-driven EN.

Teaching Point: Progesterone-driven EN may be effectively treated with spironolactone.

Category: Clinical Case

Figure 1. Right anterior thigh with large, erythematous, indurated, subcutaneous nodules.



ADULT-ONSET STILL'S DISEASE IN A 40-YEAR-OLD FEMALE: A CASE REPORT

Alexa Mier, BS[1]; Jourdan Brandon, MD[2]; Stephen Bearman, MD[2]; Michael Loncharich, MD[3]; Anne Trager, DO[2]

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Abstract

Adult-onset Still's Disease (AOSD) is a rare systemic inflammatory disease with a broad range of clinical manifestations, including high spiking fevers, evanescent rash, arthralgias/arthritis, and multiorgan involvement. Diagnosis depends on Yamaguchi criteria, which include clinical and laboratory components. The etiology of the disease is unknown, but it is recognized mainly as a polygenic autoinflammatory disorder with potential infectious, malignant, and genetic triggers. Early recognition is crucial to prevent life-threatening complications such as macrophage activation syndrome (MAS). We report a 40-year-old female who presented to the emergency department with a two-day history of daily spiking fevers, arthralgias, diffuse urticarial skin rash, and sore throat. She was admitted to the hospital for further evaluation and assessed by infectious disease and dermatology specialists. Her symptoms improved spontaneously while inpatient. No definitive diagnosis was established. She was discharged with cetirizine for presumed urticaria, doxycycline for empiric tick-borne illness coverage, and instructions for close outpatient follow-up. Following discharge, the patient had worsening joint pains, sore throat, and remittent fever in the evenings that coincided with the exacerbation of her rash. The patient was readmitted to the hospital one week later with similar clinical findings, new labs demonstrating uptrending ferritin, liver enzymes, WBC, CRP, ESR, and imaging significant for hepato-splenomegaly. The patient met the Yamaguchi criteria for AOSD and was diagnosed with polycyclic AOSD. Treatment with anakinra and solumedrol was initiated, resulting in rapid improvement. She was later transitioned to canakinumab with sustained remission of the disease. AOSD poses significant diagnostic challenges due to its variable manifestations and overlap with infectious, autoimmune, and malignant conditions. Early recognition of this condition is critical to prevent progression to MAS. Key clinical and laboratory findings outlined in the Yamaguchi criteria can assist in making the diagnosis. However, prompt recognition of the characteristic dermatologic findings can help distinguish AOSD.

Teaching point: In Adult-onset Still's Disease, prompt recognition of its varying rash can aid early diagnosis and prevent life-threatening complications like macrophage activation syndrome.

Category: clinical case

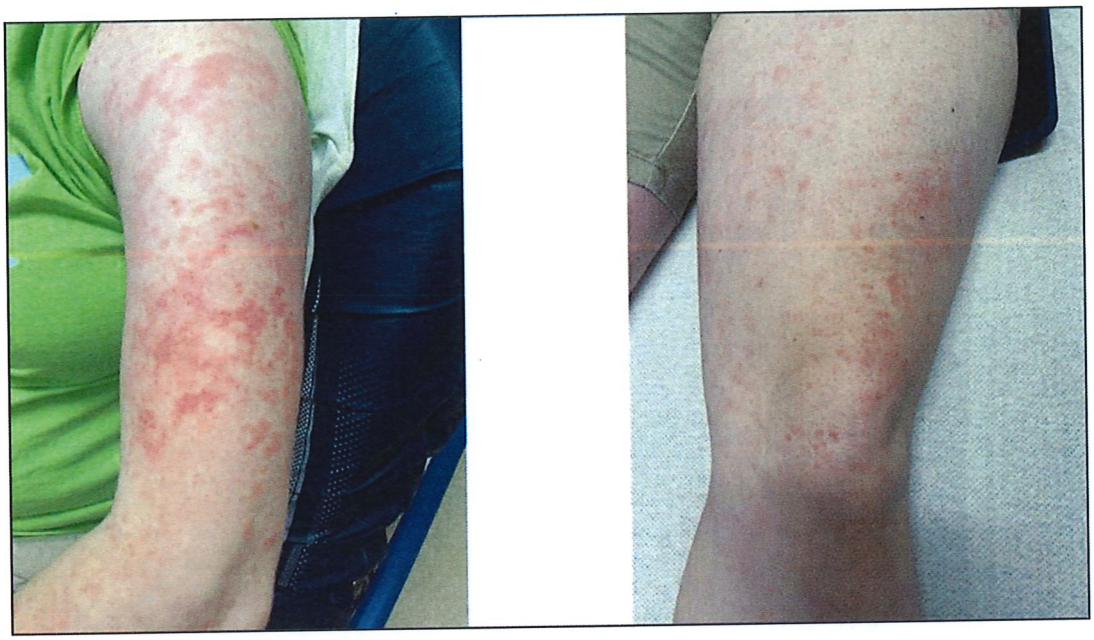

Figure 1. a. Left forearm. b. Left thigh

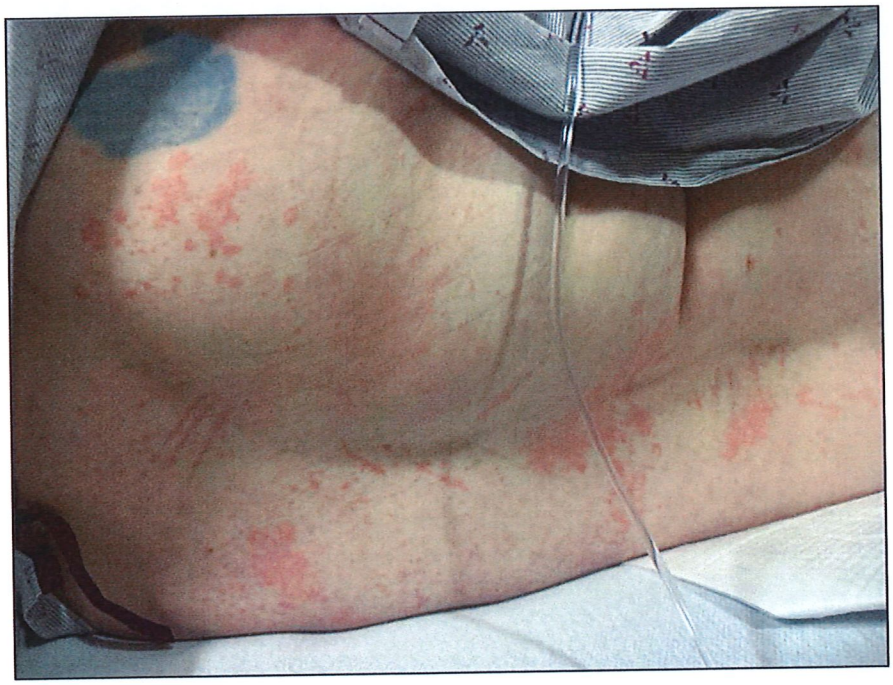

Figure 2. Superior back

MEDICAL AND GENETIC EVALUATION OF A PATIENT WITH AQUIRED CUTIS LAXA

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Cutis laxa is a rare connective tissue disorder, diagnosed in fewer than an estimated 5,000 persons in the United States.[1] Characterized by a loss of elastic fibers, this condition results in redundant skin that can significantly alter appearance and functionality. There are several subtypes exhibiting varying clinical presentations rooted in acquired and genetic causes, but these categorizations may not be mutually exclusive. Here we present the case of a 25-year-old female with cutis laxa who presented for loose, rapidly aging skin after initial onset of earlobe and facial wrinkling three years prior. Biopsies confirmed decreased elastic fibers throughout the dermis. Direct immunofluorescence testing revealed intense staining for IgA and lambda light chain deposition of the lymphatics and papillary dermis in a pattern consistent with immunoglobulin deposition to an autoantigen. Cutis laxa is associated with plasma cell dyscrasia, however her SPEP was negative. Further workup with electron microscopy allowed for enhanced specificity and revealed a lack of elastic fibers. Additional labs including CBC, CMP, and urinalysis returned unremarkable, and the patient reported no constitutional symptoms. Age of onset indicated an acquired form of cutis laxa; however, given reports of genetic predispositions to development, this patient received whole exome sequencing, which was unremarkable for reported risk variants.[2] Depending on clinical manifestations, cutis laxa patients may require monitoring for cardiovascular, pulmonary, musculoskeletal, and renal function. Plastic surgery may be utilized for cosmetic concerns. There is no current cure, but it is imperative to investigate associated comorbidities.

Teaching point: Electron microscopy, used in conjunction with traditional immunofluorescence methods, is a useful technique in the evaluation of cutis laxa.

Category: Clinical Case

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[1] U.S. National Institutes of Health (NIH); the specific page is produced by GARD, part of NCATS. *Cutis Laxa | About the Disease | GARD.*; 2025. https://rarediseases.info.nih.gov/diseases/6227/cutis-laxa

[2] O'Connell KA, Schaefer M, Atzmony L, et al. Clinical features in adults with acquired cutis laxa: a retrospective review. *Br J Dermatol*. 2023;188(6):800-816. doi:10.1093/bjd/ljad043

PERSISTENT THROMBOTIC EVENTS IN A SERONEGATIVE PATIENT WITH ANTIPHOSPHOLIPID SYNDROME

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Antiphospholipid syndrome (APS) is a systemic autoimmune disorder characterized by arterial, venous, or small vessel thromboses in the persistent presence of antiphospholipid antibodies. The current standard of care involves anticoagulation with heparin, warfarin, or other vitamin K antagonists, while use of direct oral anticoagulants remains controversial. Seroconversion to negative antibodies has been reported in the literature with an incidence ranging from 9-59%. The clinical implication of a fluctuating profile remains unclear, including whether anticoagulation can be safely discontinued after achieving seronegativity, as risk for thrombosis continues despite seronegativization. Here we present the case of a patient with APS who experienced persistent thrombotic events despite continuous anticoagulation and seronegative status. A 66-year-old male with a history of peripheral vascular disease, coronary artery disease, atrial fibrillation on apixaban, and a prior history of APS seropositivity (negative for 7 years) presented with a retiform purpuric plaque on the right lateral chest and a purple macule over the left second toe. During hospitalization, new purpura developed over the previously unaffected left breast and nose, with worsening of the primary right chest lesion. A punch biopsy of the right chest lesion revealed micro-occlusive vasculopathy with intravascular PAS-positive material, and epidermal and dermal necrosis. Laboratory data showed elevated D-dimer (19 μg/mL, normal <0.5 μg/mL) and ESR (54 mm/hr). Anti-Beta-2 glycoprotein, anti-cardiolipin, and lupus anticoagulant antibodies were negative. Despite seronegative status, his presentation was attributed to APS based on biopsy findings and clinical history. The patient was transitioned from apixaban to warfarin and treated with prednisone, IVIG, and wound debridement. This case underscores the challenges of managing cutaneous APS in patients with complex comorbidities, while highlighting the risk of DOACs in APS patients. Despite the patient's low serological findings, ongoing thrombotic risk from APS compounded by cardiovascular profiles requiring DOACs emphasizes the need for individualized anticoagulation strategies.

Teaching point: Fulfillment of APS criteria may warrant lifelong anticoagulation with warfarin regardless of seroconversion in patients with continued risk factors.

Category: Clinical Case

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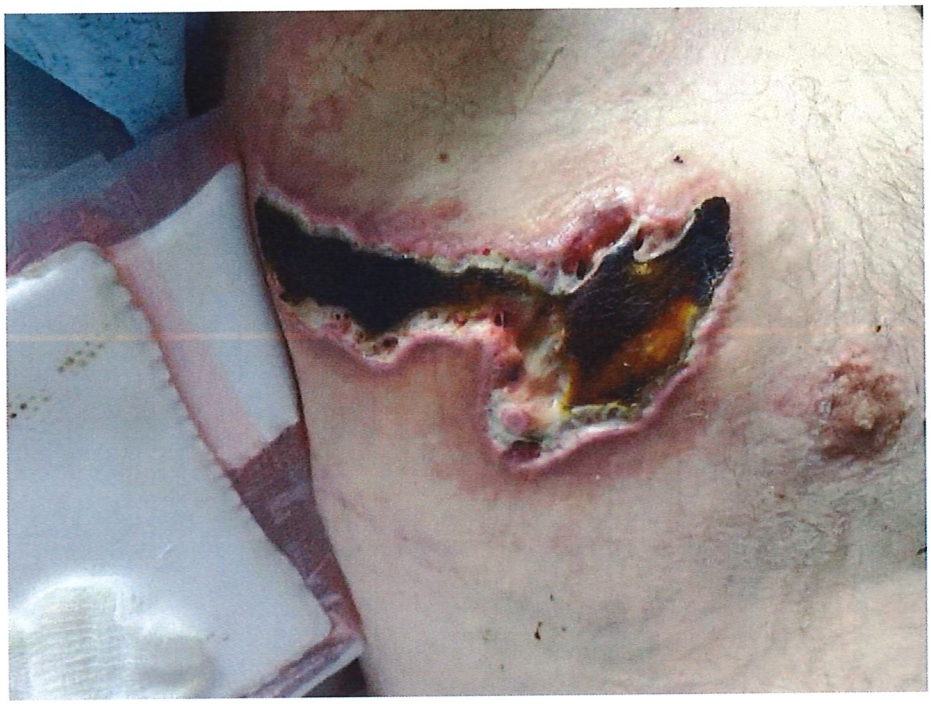

Title: Neonatal Lupus Erythematosus in the Absence of Anti-Ro antibodies

Authors and Affiliations:

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Abstract:

Neonatal lupus erythematosus (NLE) is caused by the transplacental transfer of anti-Ro (SSA), anti-LA (SSB), and/or ribonucleoprotein (RNP) antibodies and can cause rash, blood/liver dysfunction, and/or cardiomyopathy in the first couple of weeks to months of life. [1] Of these symptoms, only cardiac disease and congenital heart block can cause irreversible damage. Most mothers with positive antibodies have known autoimmune connective tissue disease. However, up to 25% of mothers are asymptomatic making the diagnosis of NLE challenging.[1,2]

A five-month year old female presented to clinic with a progressive, pruritic, well-defined rash of the face (Figure 1). Maternal history was significant for systemic lupus erythematosus with positive SSA and RNP antibodies. Delivery was uncomplicated with no abnormalities on prenatal or newborn exams including both cardiac ultrasound and electrocardiogram. A preliminary diagnosis of NLE was made, and additional workup was collected including CBC, CMP, SSA, SSB, and RNP antibodies with recommendations to use desonide ointment twice daily as needed for itch. Work up was remarkable for positive RNP antibodies only.

There are few cases of NLE presenting after 3 months and/or isolated RNP positivity with limited data on clinical significance.[3] From what is published, infants with NLE who have isolated RNP positivity tend to have cutaneous changes only.[4] It is important to note that only 15-25% of infants with NLE develop cutaneous changes making maternal antibody screening crucial to early diagnosis and appropriate monitoring.[5] Mothers should be counseled that their risk of having another child with NLE is increased tenfold making regular maternal/fetal monitoring critical in subsequent pregnancies.[6]

Teaching point: Not all patients with NLE have positive SSA antibodies and/or traditional skin findings making maternal antibody screening crucial to appropriate monitoring and treatment prior to and after birth

Abstract category: clinical case (lupus)

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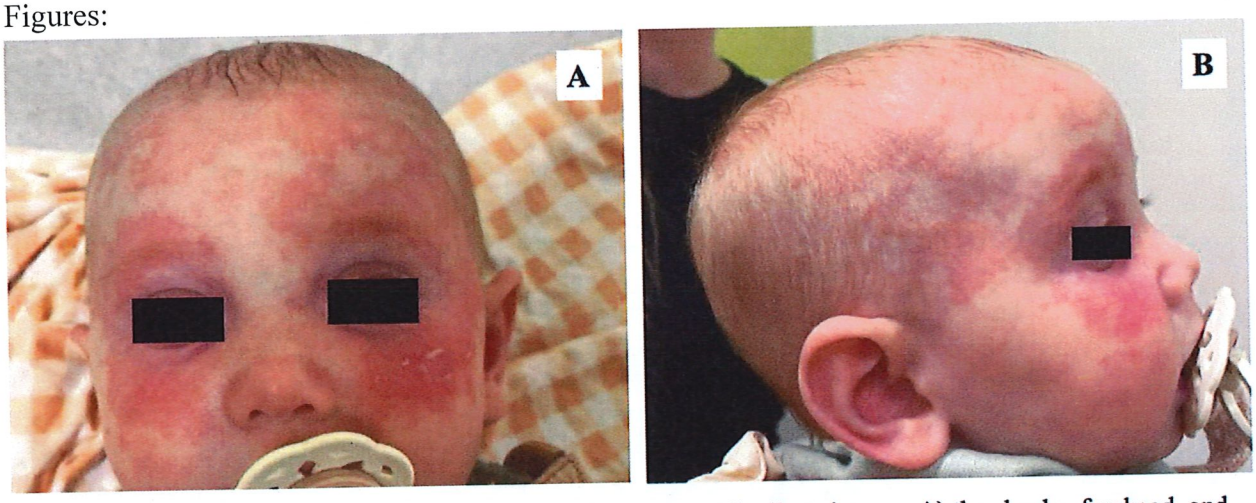

Figure 1. Well-defined erythematous patches and plaques photo-distributed across A) the cheeks, forehead, and periocular region and B) the scalp

Title: Ocular Sarcoidosis Hiding Behind Ocular Cicatricial Pemphigoid: A Unique Case of Dual Diagnosis

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Abstract:

Ocular cicatricial pemphigoid (OCP) is a rare blistering disease caused by autoantibody activity towards basement membrane proteins that can lead to blindness in 30% of affected eyes.[1] Cicatricial conjunctivitis and symblepharon are clinical hallmarks of disease with direct immunofluorescence (DIF) testing being the gold standard for diagnosis.[2] Positive DIF is not required to initiate treatment with sensitivity of DIF reported as low as 50%.[2] Additional hemotoxin and eosin testing (H&E) highlights the presence of inflammatory cells and scarring, but these findings aren't specific to OCP. Given the lack of specificity, there is some question about the value of H&E in diagnostic workup.

In this report, we present a 53-year-old patient with progressive irritation, cicatricial conjunctivitis, and symblepharon of the left eye diagnosed with both OCP and ocular sarcoid confirmed with DIF and H&E (Figures 1-3). Additional systemic workup revealed chest nodules with granulomatous inflammation. Despite treatment with dapsone and local injections of 5 fluorouracil, her ocular disease progressed with new involvement of the right eye. Adjunct therapy with methotrexate was tried with no response. Given disease progression with conventional OCP therapy, methotrexate was discontinued and she transitioned to infliximab-dyyb infusions to target ocular sarcoid. She has received one infusion thus far without adequate time since to assess clinical response.

To the best of our knowledge this is one of the only reported cases of OCP and ocular sarcoid in the same patient.[3] Although rare, ocular sarcoid can present with cicatricial conjunctivitis and symblepharon.[3,4] Examples like this highlight the importance of thorough histologic examination in the work up of autoimmune blistering diseases.

Teaching point: Cicatricial conjunctivitis is a clinical manifestation of OCP and/or ocular sarcoid, and should be considered in patients with cicatricial conjunctivitis that do not respond to traditional OCP therapies.

Abstract Category: clinical case (sarcoid)

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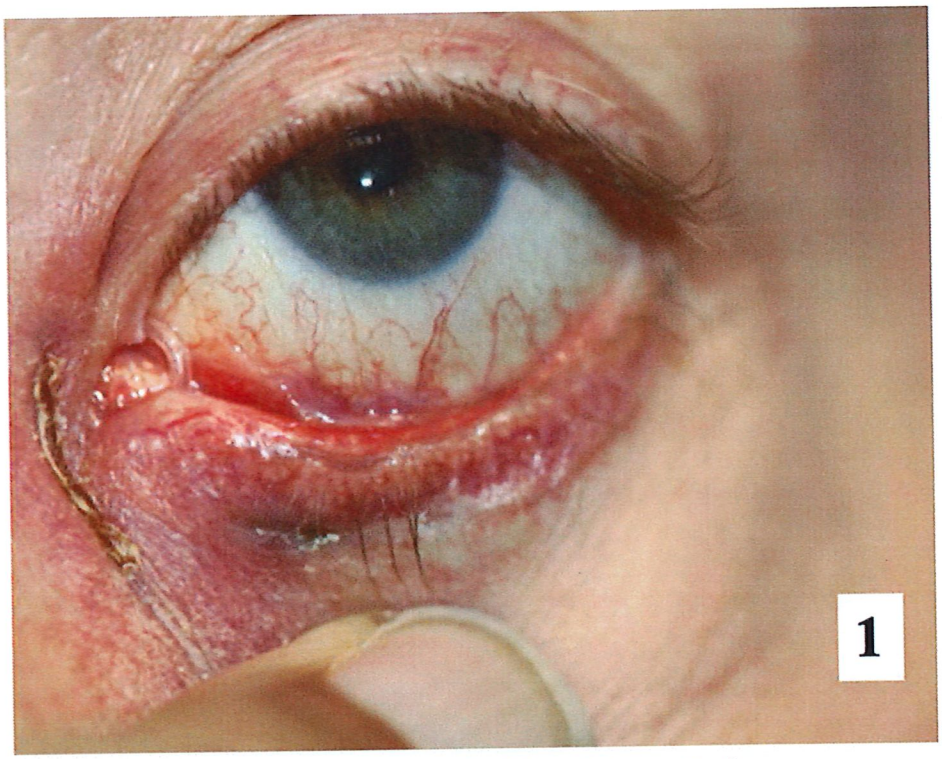

Figure 1. Cicatricial conjunctivitis with central symblepharon of the left eye

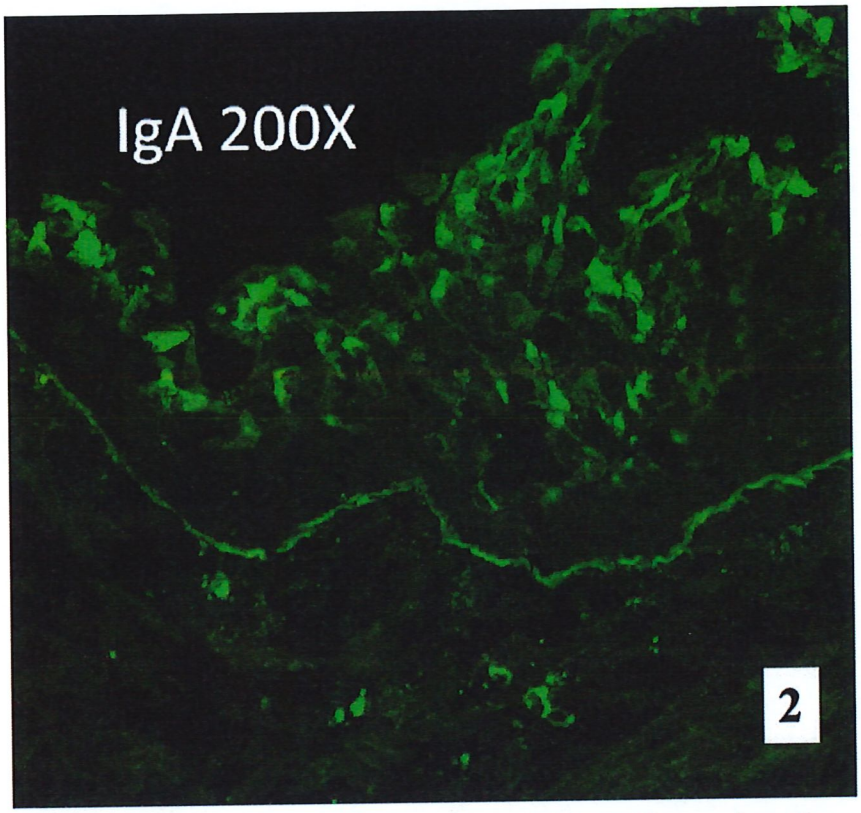

Figure 2. Direct immunofluorescence (DIF) of the conjunctiva with 2+ IgA in a linear pattern within the basement membrane zone.

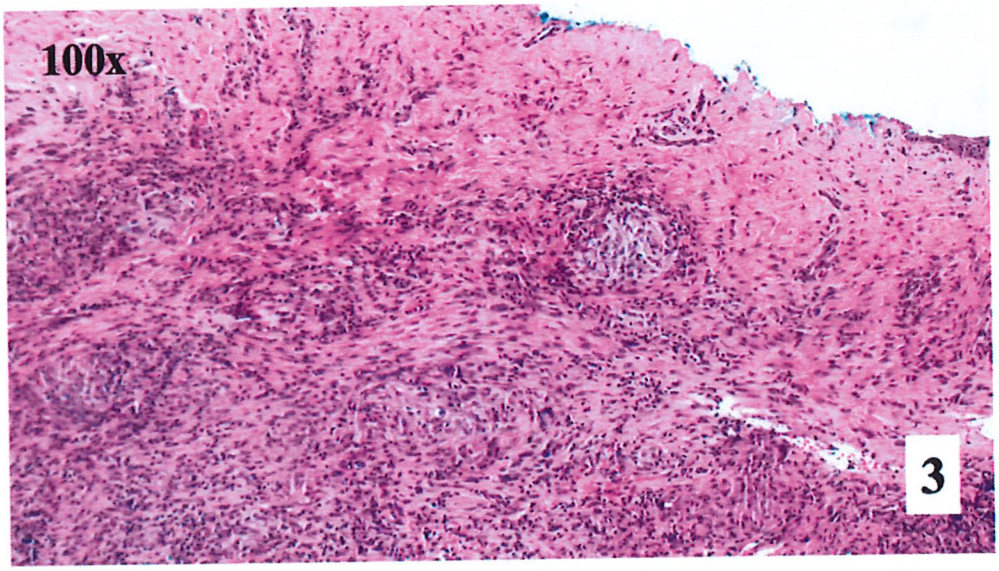

Figure 3. Hemotoxin and eosin (H&E) of the conjunctiva with an inflammatory infiltrate composed of lymphocytes and epithelioid histiocytes arranged in well-defined granulomas

Title: Palm Reading in Practice; Unique Cutaneous Manifestations of Anti-MDA-5 Dermatomyositis

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Abstract:

Dermatomyositis is a chronic inflammatory disease of the skin and muscle that is associated with an increased risk of malignancy and lung disease in adult patients. Autoantibodies can be found in 40-80% of patients with both myositis specific and myositis associated subtypes.[1] Of particular significance is anti-melanoma differentiation-associated gene 5 antibodies (anti-MDA5) subtype that unlike other forms of dermatomyositis has limited to absent myopathy, unique skin findings, and pulmonary lesions that may be rapidly progressive. [2,3]

A 26-year-old female presented with worsening rash of the bilateral hands, progressive fatigue, shortness of breath, and difficulty swallowing. On physical exam flat-topped pink papules overlying the dorsal MCP joints, tender palmar papules, and nailfold erythema were present with scattered erythematous papules and patches on the bilateral cheeks and elbows (Figures 1-3). A clinical diagnosis of dermatomyositis was made, and the patient was started on combination therapy with mycophenolate mofetil, tapered methylprednisolone, and trimethoprim-sulfamethoxazole prophylaxis. Additional systemic workup was significant for evidence of scattered myositis of the bilateral lower extremities with mild nonspecific ground glass opacities on CT chest without evidence of interstitial lung disease. Labs including CBC, CRP, CMP, aldolase, creatinine kinase, and lactate dehydrogenase were all within normal limits. MDA-5 was positive on myositis panel. Since her initial visit, adjunct therapy with hydroxychloroquine and intravenous immunoglobulin infusions were started to help reach treatment goal.

Our patient exhibited tender palmar papules which is a unique and early onset cutaneous finding seen in MDA-5-associated dermatomyositis. [2] This pathognomonic cutaneous finding is essential to recognize due to the associated rapidly-progressive interstitial lung disease seen in a subset of patients.

Teaching point: Dermatomyositis patients with the anti-MDA5 have specific cutaneous manifestations including tender palmar papules and ulcerations typically overlying extensor surfaces that play an important role in early diagnosis and pulmonary screening.

Abstract Category: clinical case (dermatomyositis)

References:

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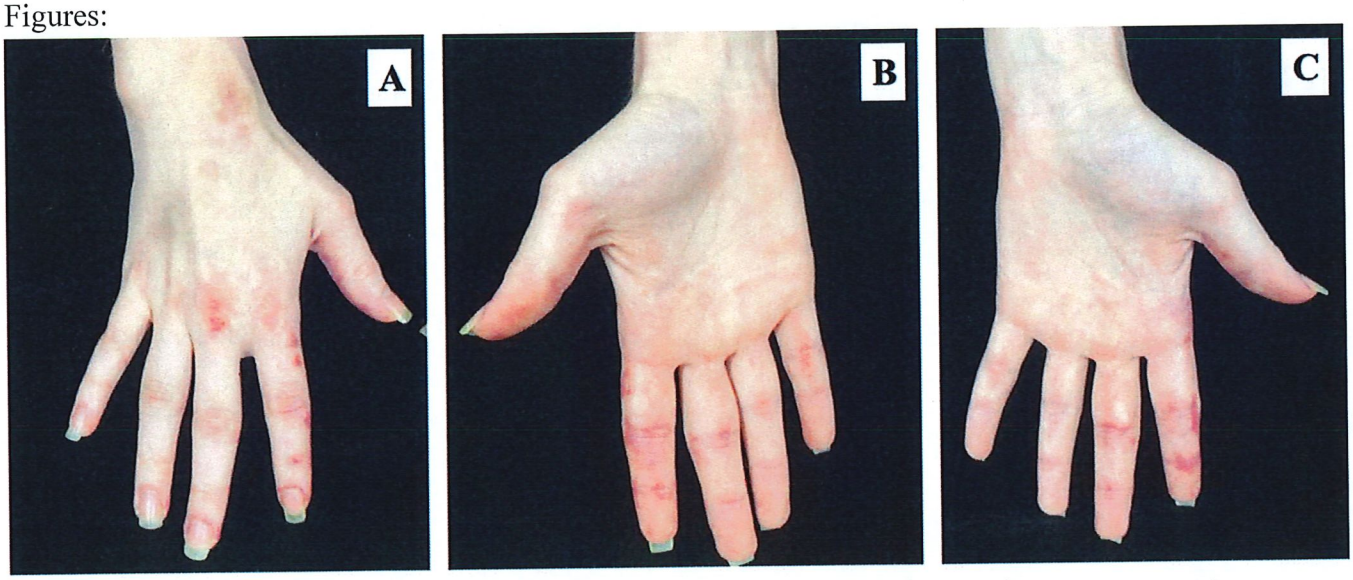

Figure 1. A) Dorsal right hand with flat topped pink papules on the 2nd and 3rd MCP joints with erythematous nailfolds. Tender palmar macules and papules on the B) right and C) left hand

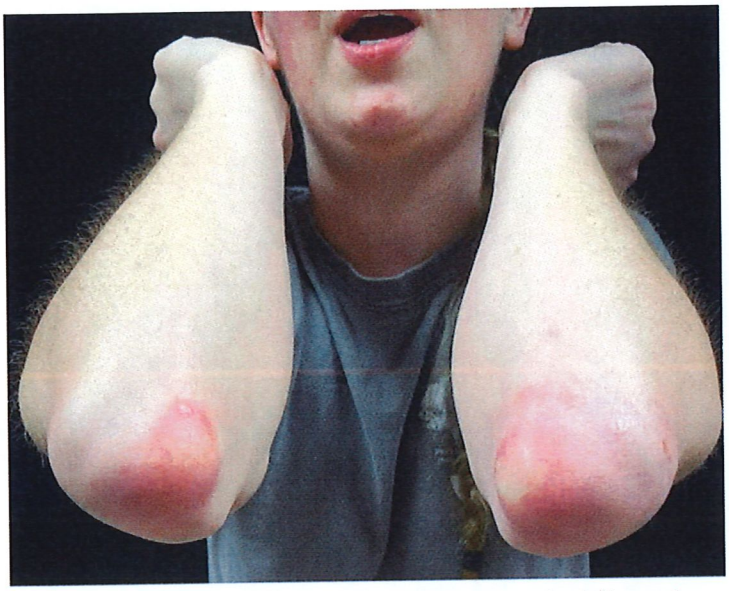

Figure 2. Erythematous tender plaques on the bilateral elbows

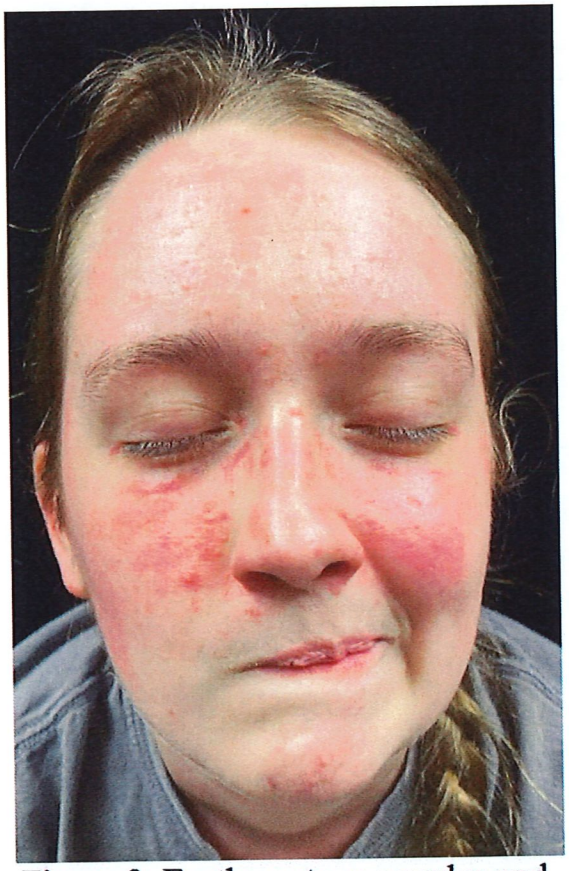

Figure 3. Erythematous papules and patches scattered across the bilateral cheeks

SUCCESSFUL TREATMENT OF DISCOID LUPUS ERYTHEMATOSUS WITH TAPINAROF 1% CREAM MONOTHERAPY: A CASE REPORT

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Abstract:
Discoid lupus erythematosus (DLE) is a chronic cutaneous lupus variant that can cause scarring and dyspigmentation and is commonly treated with ultrapotent topical corticosteroids, topical calcineurin inhibitors, intralesional corticosteroids, and systemic agents; however, some patients remain refractory or decline invasive or systemic therapy. Tapinarof 1% cream is an aryl hydrocarbon receptor (AhR) agonist approved for plaque psoriasis with immunomodulatory and barrier-supportive effects that may be relevant to DLE. We report a 56-year-old woman with three months of painful, ulcerating erythematous plaques on the nasal bridge and frontal scalp. Biopsy demonstrated interface dermatitis with superficial and deep periadnexal lymphocytic infiltrates, consistent with DLE; evaluation did not suggest systemic lupus erythematosus. Sequential trials of tacrolimus 0.1% ointment and clobetasol 0.05% ointment (each twice daily for four weeks) were ineffective. The patient declined intralesional corticosteroids and systemic therapy, and ruxolitinib 1.5% cream was unobtainable. Off-label tapinarof 1% cream monotherapy was initiated once daily. The patient experienced rapid improvement in burning within days and complete clinical resolution of the facial plaque by three weeks without adverse events. Remission was sustained at three-month follow-up off additional therapy. Tapinarof's AhR-mediated modulation of pro-inflammatory cytokines (e.g., interleukin-17 and interleukin-22) and T-cell subsets provides a plausible mechanism for efficacy in DLE and suggests a potential steroid-sparing role. While limited by single-patient design and short follow-up, this case supports further study of tapinarof as a topical option for DLE patients who are refractory to, intolerant of, or unwilling to use corticosteroids or systemic agents.

Teaching Point: Tapinarof 1% cream, an aryl hydrocarbon receptor agonist, may offer a rapid, steroid-sparing topical option for discoid lupus erythematosus refractory to standard therapies.

Abstract Category: Clinical Case

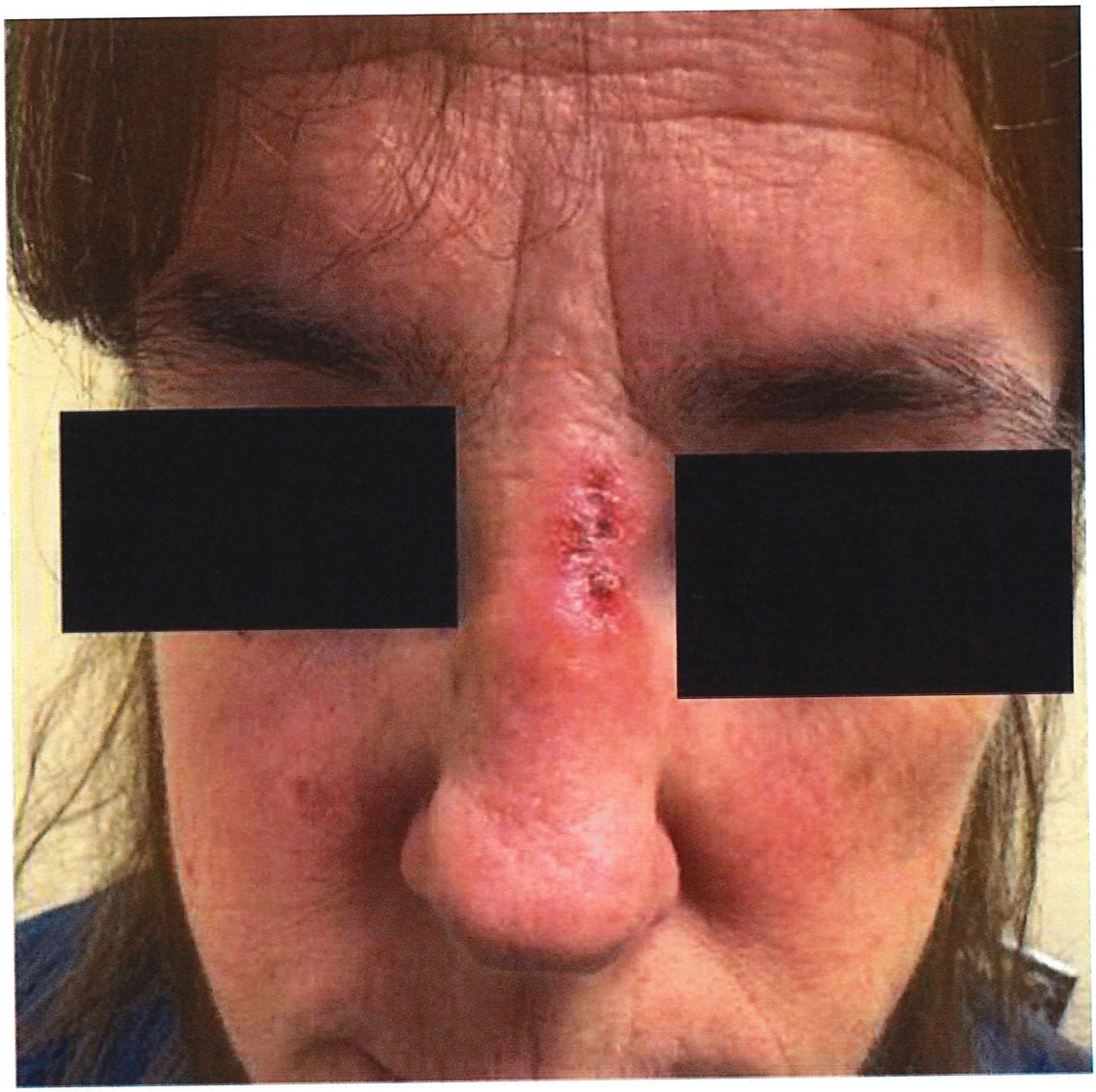

Figure 1: Initial DLE presentation with indurated and ulcerated erythematous plaque on the nasal bridge. *DLE*, Discoid lupus erythematosus.

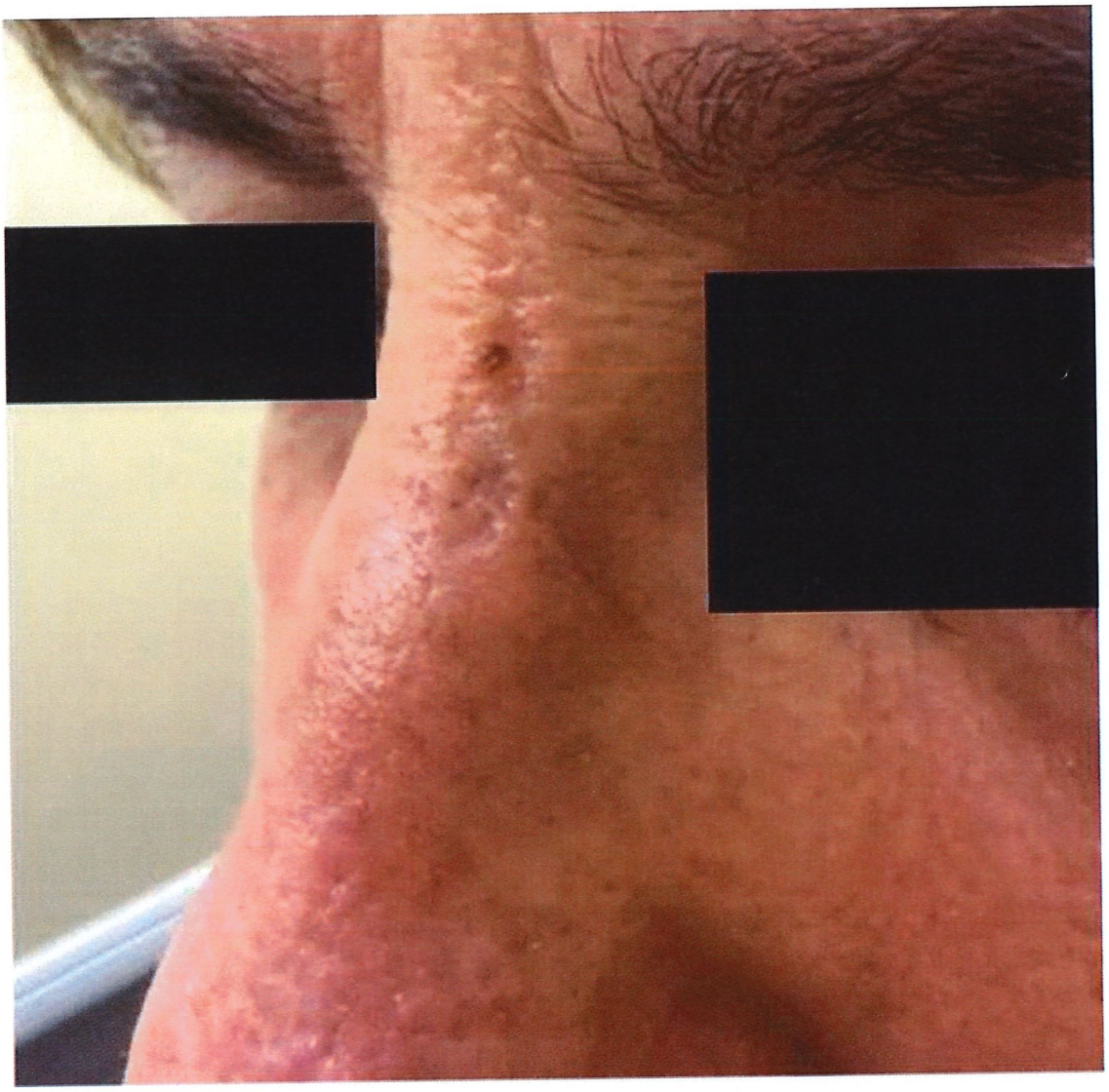

Figure 2: Resolution of DLE following application of once-a-day tapinarof 1% cream for 3 weeks. *DLE*, Discoid lupus erythematosus.

TREATMENT OF RECALCITRANT DISCOID LUPUS ERYTHEMATOSUS WITH ROFLUMILAST 0.3% CREAM FOLLOWING SUBOPTIMAL RESPONSE TO ANIFROLUMAB AND IMPROVEMENT OF HYPOPIGMENTATION

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[3]Department of Dermatology, The George Washington University Medical Faculty Associates, Washington, DC

Abstract:

Discoid lupus erythematosus (DLE) is a chronic cutaneous lupus subtype characterized by erythematous, scaly plaques that may progress to scarring and dyschromia; therapies include ultrapotent topical corticosteroids, topical calcineurin inhibitors, Janus kinase inhibitors, antimalarials, and systemic immunosuppressants. We report a woman in her 60s with severe systemic lupus erythematosus (SLE) and recalcitrant DLE presenting with pruritic, burning, erythematous, atrophic, hypopigmented papules and plaques of the face, scalp, and ears. Biopsy showed interface dermatitis with periadnexal lymphocytic infiltrates, supportive of DLE. Despite hydroxychloroquine 200 mg twice daily, methotrexate 20 mg weekly, sun protection, and clobetasol 0.05% ointment, disease control was poor. Hydroxychloroquine and methotrexate were discontinued and anifrolumab 300 mg infusions every 4 weeks were initiated, producing early improvement after one infusion; however, cutaneous response plateaued by month 5 with persistent erythema and pruritus despite alternating tacrolimus 0.1% ointment and clobetasol 0.05% ointment. Off-label roflumilast 0.3% cream (a phosphodiesterase-4 [PDE4] inhibitor) once daily was then started, and the tacrolimus/clobetasol regimen was discontinued. Over 8 weeks, pruritus gradually improved with complete resolution of erythema and symptoms, accompanied by marked improvement of hypopigmentation; remission was sustained at follow-up and no adverse events were observed. The clinical benefit may relate to PDE4-mediated modulation of cyclic adenosine monophosphate signaling and downstream T helper 17/22 pathways (e.g., interleukin-17), while reported melanocyte effects of roflumilast could explain the observed repigmentation. Although limited by single-patient design and concomitant anifrolumab, this case suggests topical roflumilast 0.3% cream as a useful adjunct for DLE refractory to biologic therapy, including in patients with postinflammatory hypopigmentation, warranting controlled studies to clarify efficacy, durability, and safety.

Teaching Point: Topical roflumilast 0.3% cream may be an effective adjunct for recalcitrant discoid lupus erythematosus and can concurrently improve postinflammatory hypopigmentation.

Abstract Category: Clinical Case

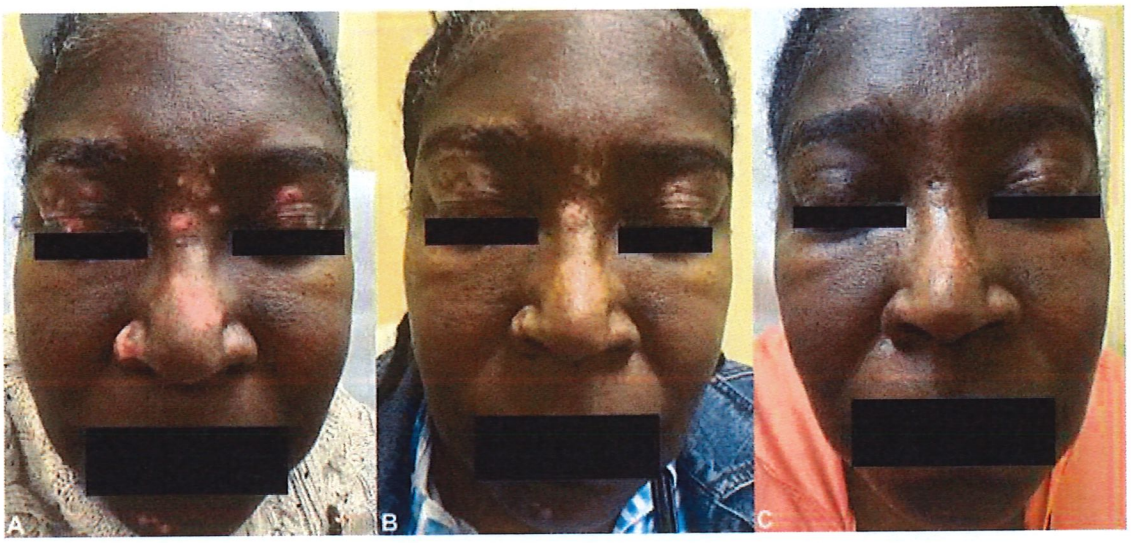

Figure 1: Discoid lupus erythematosus (DLE) that presented as scaly, erythematous, atrophic, hypopigmented papules, and plaques on the face, scalp, and ears (**A**) that improved after 1 month infusion of anifrolumab, but remains significantly pruritic and slightly erythematous despite 5 months of treatment (**B**). Resolution of the DLE and improvement in hypopigmentation after 8 weeks use of roflumilast 0.3% cream (**C**).

REACTIVATION OF LATENT TUBERCULOSIS FOLLOWING TNF-ALPHA INHIBITOR THERAPY IN A PATIENT WITH RHEUMATOID ARTHRITIS: A CASE REPORT

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Abstract:

This clinical case describes the reactivation of latent tuberculosis (TB) in a 42-year-old woman with rheumatoid arthritis following 14 months of adalimumab therapy, a tumor necrosis factor-alpha (TNF-α) inhibitor. The patient had a remote history of successfully treated pulmonary TB nine years prior. She presented with persistent cough, weight loss, anorexia, and night sweats. Chest radiography revealed pulmonary infiltrates, and sputum smear tested positive for acid-fast bacilli, confirming reactivation TB.

Adalimumab was promptly discontinued, and standard antitubercular therapy (ATT) was initiated. TNF-α is essential for granuloma formation and containment of Mycobacterium tuberculosis. Its inhibition compromises granuloma integrity, allowing dormant bacilli to reactivate. Despite prior completion of ATT, immunosuppressive therapy created a permissive environment for reactivation.

This case underscores that patients in TB-endemic regions like India may harbor residual bacilli even after full treatment. Additionally, a negative latent TB screening prior to initiating TNF-α inhibitors does not fully exclude the risk of reactivation. Vigilance, comprehensive TB history, and consideration of prophylactic therapy in high-risk individuals are critical when planning biologic treatment.

Teaching Point:

Even patients with previously treated TB remain at risk of reactivation when exposed to TNF-α inhibitors. In TB-endemic regions, pre-treatment screening protocols should be re-evaluated, and prophylaxis considered more broadly.

Abstract Category: Clinical Case

Filename for Submission: Gunjan_Reactivation_of_Latent_TB

Submission Email Subject Line: RDS2025

LINEAR MORPHEA WITH CONCOMITANT CLINICAL AMYOPATHIC DERMATOMYOSITIS: EXPANDING THE SPECTRUM OF AUTOIMMUNE OVERLAP SYNDROMES

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Linear morphea presents as linear plaques involving the skin, subcutaneous tissue, muscle, and bone.[1] Clinical amyopathic dermatomyositis (CADM) is a dermatomyositis subtype that exhibits cutaneous findings without clinical or laboratory muscle involvement for at least six months.[2] We report a 22-year-old Hispanic male with a 12-year history of linear morphea and recent onset of knuckle rash. Patient has been without therapy for the past three years. Prior treatments, including methotrexate, mycophenolate mofetil, and phototherapy, were discontinued due to adverse effects. Examination revealed erythematous-violaceous linear sclerotic plaque from the right axilla to the elbow, multiple ulcerated, rock-hard yellow papules consistent with calcinosis cutis near both elbows (Figure 1), dilated nailfold capillaries with microhemorrhages, erythematous papules on roughened dorsal hands, and skin pitting along the ulnar aspect of the right palm (Figure 2). Muscle strength was preserved, but right elbow and shoulder range of motion was reduced by pain (Figure 3). Biopsy confirmed Gottron's papules, and laboratory results were unremarkable except for markedly positive anti-TIF1-γ antibodies. A diagnosis of linear morphea with concomitant CADM was established, representing the first reported case of this association. CADM carries an increased risk of interstitial lung disease, and in adults, anti-TIF1-γ positivity is strongly associated with malignancy.[3, 4, 5] The patient's borderline age at dermatomyositis onset complicates malignancy risk assessment, as juvenile-onset cases demonstrate weaker associations.[6] Extensive calcinosis and functional impairment may reflect longstanding uncontrolled disease and delayed treatment, patterns more common among non-White patients.[3] Early, aggressive control of linear morphea and other autoimmune skin diseases is critical, as prolonged inflammation may promote development of additional autoimmune conditions, as seen in our patient. This case raises awareness of rare overlapping autoimmune presentations and underscores the importance of early recognition, malignancy screening, and timely immunosuppression, to prevent irreversible damage, secondary autoimmune disease, and cancer.

Teaching Point: This first reported case of concomitant linear morphea and clinical amyopathic dermatomyositis underscores the importance of maintaining clinical suspicion for autoimmune overlap syndromes in patients presenting with new symptoms, to initiate timely immunosuppression, enhance malignancy screening, and prevent irreversible damage and systemic complications.

Category: Clinical case

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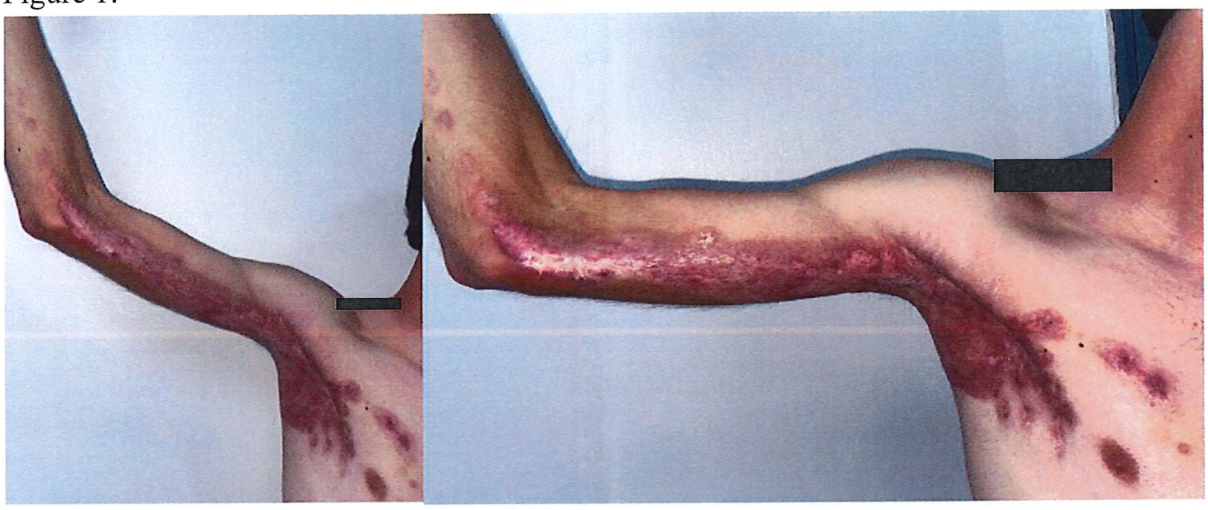
Figure 1:



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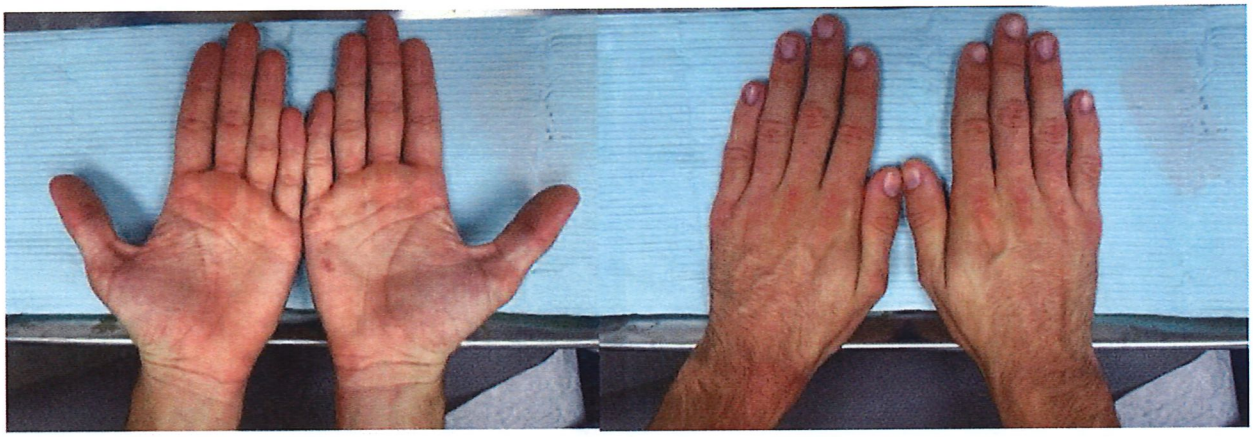

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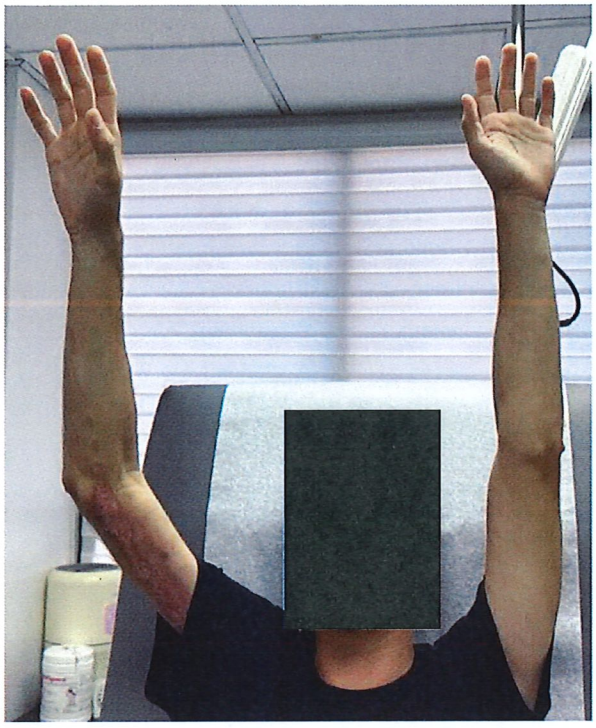

POINT-OF-CARE RISK FACTORS FOR SYSTEMIC DISEASE IN PATIENTS PRESENTING WITH SMALL VESSEL VASCULITIS OF THE SKIN

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Small vessel vasculitis (SVV) presenting in the skin may be skin-limited or a manifestation of systemic disease. However, no evidence-based protocols exist to identify patients at risk for systemic involvement. This study aimed to identify clinical features at initial evaluation that predict systemic disease in patients with cutaneous SVV, to support early risk stratification and management. This multi-center case-control study included 430 adults with biopsy-confirmed cutaneous SVV, identified from pathology databases at Massachusetts General Hospital, Brigham and Women's Hospital, and the Hospital of the University of Pennsylvania. Aligned with the Chapel Hill consensus criteria, cases were defined as systemic if SVV was associated with a systemic vasculitis diagnosis, connective tissue disease, or caused objective organ dysfunction. Demographic, clinical, and laboratory features at presentation were assessed. Univariate and multivariate logistic regression analyses estimated odds ratios (ORs) and 95% confidence intervals (CIs), with variable selection guided by elastic net regression and 10-fold cross-validation. Among 430 patients (mean [SD] age, 55 [17.7] years; 55.5% male), 87 had systemic disease and 343 had skin-limited SVV. Multivariate analysis identified nausea/vomiting (OR 3.92; 95% CI 1.58–9.77), ulcerating or necrotic lesions (OR 3.26; 95% CI 1.24–8.59), abdominal pain/cramping (OR 2.71; 95% CI 1.19–6.18), and fatigue/malaise/lethargy (OR 2.61; 95% CI 1.28–5.32) as significantly associated with systemic disease. In a subset with urinalysis data (n=388), dipstick hematuria (3+) was associated (OR 2.40; 95% CI 1.23–4.68) and recent antibiotic use was inversely associated (OR 0.54; 95% CI 0.29–0.98). Younger age (OR 0.74 per SD increase; 95% CI 0.58–0.93) was associated in exclusively univariate analysis. In this multi-institutional study of SVV, we identified key risk factors–ascertainable at initial presentation–that may aid in identifying high-risk patients requiring further evaluation and closer monitoring, potentially improving outcomes through earlier detection and management of critical complications.

Category: Vasculitis

RDS Annual Meeting 2025 Abstract

Title: Cutaneous manifestations of vasculitis: A cross-sectional analysis from an international cohort

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Body (300 words)

Characterization of frequency, type, and significance of cutaneous manifestations (CM) of vasculitis is incomplete. This cross-sectional study analyzed data from the Diagnostic and Classification Criteria in Vasculitis (DCVAS) study, involving 136 sites in 32 countries, collecting comprehensive data on patients with vasculitis from disease onset to 6 months. We used summary statistics to describe CM and logistic regression to calculate odds ratios for severe systemic manifestations. Data from 4,468 adults were analyzed—270 with IgA vasculitis (IgAV), 201 with Behçet's disease (BD), 57 with cryoglobulinemic vasculitis (CV), 1,023 with granulomatosis with polyangiitis (GPA), 505 with microscopic polyangiitis (MPA), 382 with eosinophilic granulomatosis with polyangiitis (EGPA), 194 with polyarteritis nodosa (PAN), 1,206 with giant cell arteritis (GCA), and 630 with Takayasu arteritis (TAK). CM were common at presentation for all types except large-vessel (GCA and TAK), ranging from 20% with MPA to 97% with IgAV (Table 1). While specific manifestations varied considerably, petechiae/purpura was most common in all types of small-vessel vasculitis (IgAV, CV, GPA, MPA, and EGPA). Skin biopsy was performed in 22-82% of those with small- or medium-vessel vasculitis. When performed, it was usually diagnostic, including 95% (205/216) of IgAV, 73% (16/22) of CV, 73% (67/92) of GPA, 73% (16/22) of MPA, 67% (49/73) of EGPA, and 70% (52/74) of PAN. Skin biopsy was less often useful for diagnosis of BD (35%; 7/20). Univariable logistic regression demonstrated patients with CM had significantly greater odds of severe systemic manifestations of GPA (OR 1.8, 95% CI 1.3, 2.6) and EGPA (OR 1.8, 95% CI 1.1, 2.9); those with CM had lower odds of severe systemic manifestations of PAN (OR 0.5, 95% CI 0.3, 0.99) (Table 2). These findings characterize CM of vasculitis at the time of diagnosis and highlight potential diagnostic and prognostic importance of the skin in these multisystem diseases.

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Table 1. Type and frequency of cutaneous manifestations in different types of vasculitis.

	IgAV	BD	CV	GPA	MPA	EGPA	PAN	GCA	TAK
	N (%)	N (%)	N (%)	N (%)	N (%)	N (%)	N (%)	N (%)	N (%)
Participants, *N*	270	201	57	1023	505	382	194	1206	630
Any skin findings at time of presentation	262 (97%)	118 (59%)	50 (88%)	299 (29%)	102 (20%)	168 (44%)	111 (57%)	34 (3%)	37 (6%)
Petechiae or purpura	243 (90%)	2 (<1%)	38 (67%)	152 (15%)	48 (10%)	78 (20%)	23 (12%)	3 (<1%)	3 (<1%)
Livedo reticularis or racemose	0 (0%)	0 (0%)	1 (2%)	4 (<1%)	7 (1%)	1 (<1%)	10 (5%)	0 (0%)	1 (<1%)
Painful skin lesions	89 (33%)	65 (32%)	15 (26%)	95 (9%)	23 (5%)	40 (11%)	50 (26%)	8 (<1%)	12 (2%)
Maculopapular rash	29 (11%)	44 (22%)	6 (11%)	71 (7%)	25 (5%)	59 (16%)	30 (16%)	7 (<1%)	3 (<1%)
Pruritus	33 (12%)	6 (3%)	5 (9%)	31 (3%)	16 (3%)	44 (12%)	13 (7%)	12 (<1%)	3 (<1%)
Urticaria	2 (<1%)	1 (<1%)	1 (2%)	7 (<1%)	3 (<1%)	30 (8%)	3 (2%)	4 (<1%)	2 (<1%)
Splinter hemorrhage	1 (<1%)	0 (0%)	0 (0%)	17 (2%)	2 (<1%)	7 (2%)	2 (1%)	0 (0%)	2 (<1%)
Cutaneous ulcers	43 (16%)	43 (21%)	12 (21%)	41 (4%)	9 (2%)	14 (4%)	30 (16%)	3 (<1%)	6 (<1%)
Mouth ulcers	11 (4%)	171 (85%)	1 (2%)	66 (7%)	18 (4%)	11 (3%)	9 (5%)	7 (<1%)	12 (2%)
Genital ulcers	0 (0%)	152 (76%)	0 (0%)	2 (<1%)	0 (0%)	0 (0%)	3 (2%)	2 (<1%)	0 (0%)
Gangrene	12 (4%)	2 (<1%)	5 (9%)	22 (2%)	6 (1%)	7 (2%)	22 (11%)	1 (<1%)	7 (1%)
Cutaneous infarct	38 (14%)	1 (<1%)	3 (5%)	16 (2%)	1 (<1%)	5 (1%)	8 (4%)	2 (<1%)	1 (<1%)

Tender skin nodules	3 (1%)	41 (20%)	1 (2%)	22 (2%)	6 (1%)	7 (2%)	27 (14%)	3 (<1%)	8 (1%)
Non-tender skin nodules	2 (<1%)	5 (3%)	1 (2%)	13 (1%)	5 (<1%)	7 (2%)	11 (6%)	3 (<1%)	3 (<1%)
Two or more different skin findings at time of presentation	144 (53%)	66 (33%)	23 (40%)	132 (13%)	31 (6%)	85 (22%)	63 (33%)	9 (<1%)	11 (2%)
Average number of different skin findings at time of presentation, *mean (standard deviation)*	1.8 (1.0)	1.0 (1.1)	1.5 (1.1)	0.5 (0.9)	0.3 (0.7)	0.8 (1.1)	1.2 (1.4)	0.04 (0.3)	0.1 (0.4)

Abbreviations: IgA vasculitis, IgAV; Behçet's disease, BD; cryoglobulinemic vasculitis, CV; granulomatosis with polyangiitis, GPA; microscopic polyangiitis, MPA; eosinophilic granulomatosis with polyangiitis, EGPA; polyarteritis nodosa, PAN; Takayasu arteritis, TAK; giant cell arteritis, GCA

Table 2. Crude Odds of severe systemic findings of vasculitis based on presence of cutaneous manifestations*

	Odds ratio (95% confidence interval)	p-value
IgA vasculitis	1.3 (0.3, 6.8)	0.726
Behçet's disease	1.0 (0.5, 2.3)	0.938
Cryoglobulinemic vasculitis	1.5 (0.2, 9)	0.684
Granulomatosis with polyangiitis	1.8 (1.3, 2.6)	<0.001
Microscopic polyangiitis	0.9 (0.4, 1.7)	0.666
Eosinophilic granulomatosis with polyangiitis	1.8 (1.1, 2.9)	0.019
Polyarteritis nodosa	0.5 (0.3, 0.99)	0.046

* Severe systemic manifestations of vasculitis were defined based on components of the Birmingham Vasculitis Activity Score (BVAS) and include: scleritis, sensorineural deafness, mesenteric ischemia, alveolar hemorrhage, respiratory failure, red blood cell casts in urine, rise in serum creatinine >30% or fall in creatinine clearance >25%, meningitis, spinal cord lesion, stroke, cranial nerve palsy, sensory peripheral neuropathy, or mononeuritis multiplex. Analysis was limited to vasculitis types with >10% prevalence of cutaneous manifestations.

EMERGING BIOLOGIC AND SMALL MOLECULE INHIBTIOR THERAPIES FOR REFRACTORY AND RELAPSING CUTANEOUS POLYARTERITIS NODOSA, A NARRATIVE REVIEW

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Cutaneous polyarteritis nodosa (cPAN) is a rare, small- to medium-vessel vasculitis characterized by painful subcutaneous nodules, livedo reticularis, and ulcerations, frequently accompanied by systemic symptoms such as arthralgia and neuropathy. Conventional treatments, including corticosteroids and immunosuppressants, remain first-line but are often limited by relapse and long-term steroid-dependence. Given the lack of randomized controlled trials, recent attention has turned to targeted biologics and small molecule inhibitors, although data are restricted to case reports and small retrospective series. To evaluate emerging therapies, a PubMed search was conducted and identified 32 cases of cPAN treated with biologic or small molecule agents, excluding cases of systemic PAN. Tumor necrosis factor-alpha (TNF-α) inhibitors represented the most frequently reported class (n=15), inducing rapid ulcer healing and steroid sparing in pediatric and adult patients; however, relapses commonly occurred following discontinuation. Interleukin-6 (IL-6) blockade (n=4) was effective in refractory and ulcerative phenotypes, aligning with evidence that elevated IL-6 correlates with more severe disease. B-cell depletion with rituximab (n=6) demonstrated variable outcomes, suggesting B-cell–driven disease mechanisms are relevant in only a subset of patients, particularly those with neuropathic involvement. Janus kinase (JAK) inhibitors (n=6) were consistently effective, producing durable remission, ulcer resolution, and steroid tapering, with the added advantage of oral administration. A single case of PDE4 inhibition with apremilast demonstrated lesion clearance in a patient with concomitant SAPHO syndrome, though this remains anecdotal. Across all reports, adverse events were rarely described, though limited follow-up precludes firm conclusions on long-term safety. Collectively, these findings suggest that TNF-α inhibitors remain the most commonly utilized, IL-6 blockade, and rituximab may be effective in specific contexts, and JAK inhibitors appear the most consistently efficacious emerging option. Future priorities include multicenter registries, biomarker-guided stratification, and adaptive trial designs to validate these therapies and establish treatment algorithms for refractory and relapsing cPAN.

Abstract category: Vasculitis

DIAGNOSTIC YIELD OF BIOPSY IN CUTANEOUS POLYARTERITIS NODOSA: REAL-WORLD DATA FROM TWO ACADEMIC MEDICAL CENTERS

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Cutaneous polyarteritis nodosa (cPAN) is a rare skin-limited vasculitis that is difficult to diagnose. Given the paucity of data regarding the optimal cutaneous morphology to biopsy, this study evaluated the diagnostic yield of skin biopsies in cPAN. We identified patients with histologically confirmed cPAN diagnosed between 1997-2024 at Mass General Brigham. Those with unavailable pathology reports, non-skin biopsy, or diagnosis of systemic PAN or another vasculitis were excluded. 52 patients met inclusion criteria. Most cases were idiopathic (92%), with few associated with infection (4%) or hematolymphoid malignancy (4%). Legs (96%) and arms (37%) were frequently affected, with subcutaneous nodules (SQNs, 81%), livedo (48%), and ulcers (35%) the most common morphologies. 94 biopsies were analyzed, 57 of which (61%) were diagnostic. 46% of patients required more than one biopsy, with 15% needing three or more. Mean time to diagnosis (TTD) was 28 (±38) months, and mean biopsy count was 1.8 (±1.3). Biopsy of SQN resulted in diagnostic biopsy in 34/44 cases (77%) ($p=0.003$). Sample sizes of other morphologies were too small to see statistical significance. Because expert consensus is to biopsy SQNs over other morphologies if present, we combined biopsies of livedo, ulceration, plaque, retiform purpura and unspecified morphologies and calculated the probability of successful biopsy compared with SQN using Fisher's exact test. Results indicated that the probability of successful biopsy was significantly less than biopsy of SQN (OR 0.25 [0.09–0.67], $p=0.003$). When small diameter punch biopsies (≤3mm) were excluded, we found substantially reduced likelihood of successful biopsy compared with SQNs OR 0.07 [0.007–0.32] ($p<0.0001$). Our findings highlight the prolonged TTD for patients with cPAN and suggest that SQNs have the highest diagnostic yield compared with other morphologies, especially with larger biopsy samples.

Category: Vasculitis