RDS Rheumatologic Dermatology Society North America

ORAL SESSION I

(8:35 AM)

LENALIDOMIDE TREATMENT OF CUTANEOUS LUPUS ERYTHEMATOSUS: THE MAYO CLINIC EXPERIENCE

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Background: Published case series describe lenalidomide as an effective treatment of refractory cutaneous lupus erythematosus (CLE).
Objectives: The present study aimed to further characterize lenalidomide use in the treatment of CLE.
Methods: A retrospective review of patients treated with lenalidomide for CLE from January 1, 2000, to December 17, 2014, was conducted.
Results: Eight of the nine patients (89%) were women. Their median age at initiation of lenalidomide was 62 years (range: 41–86 years). Subtypes of CLE included discoid lupus erythematosus (DLE) (n = 6), lupus panniculitis (n = 2), and subacute CLE (n = 1). Before the initiation of lenalidomide, all patients had been previously treated unsuccessfully or were intolerant to at least one antimalarial and one immunosuppressive agent. With lenalidomide, five patients achieved a complete response (CR), two a partial response, and two had no response (lupus panniculitis). Time to initial response (dose range: 2.5– 10.0 mg/d) varied from 2 weeks to 3 months; the median time to CR in five patients was 3 months (range: 3–6 months). The median duration of lenalidomide therapy was 12 months (range: 2–67 months). The median duration of follow-up was 48 months (range: 20–103 months). Adverse effects included mild leukopenia; one patient had deep vein thrombosis of unclear etiology during a hospitalization. No patients developed or showed progression of systemic LE while receiving lenalidomide.
Conclusions: Lenalidomide was effective for the treatment of CLE (particularly DLE) but not for the treatment of lupus panniculitis in this series.

(8:43 AM)

LOW DOSE UVA1 RADIATION IN PATIENTS WITH CUTANEOUS LUPUS

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UVA1 radiation has been proposed as a treatment modality for patients with cutaneous lupus erythematosus (CLE). Previous clinical trials of patients with systemic lupus treated with UVA1 radiation showed improvement in disease activity but included small numbers of patients with CLE. The objective of this study is to investigate the safety and efficacy of low dose UVA1 radiation in patients with CLE. Participants received 20 J/cm^2 of UVA1 radiation on active CLE

lesions three times per week for ten weeks followed by an eight-week wash-out period. Clinical assessments including Cutaneous Lupus Activity and Severity Index, Systemic Lupus Erythematous Disease Activity Index, physician global assessment scores, and blood and skin samples were collected at four study visits. ELISAs measuring serum antibody levels (e.g. ANA, ssDNA, dsDNA, RNP), and flow cytometry analyses of peripheral blood mononuclear cells (e.g. $CD3^+$ T cells, $CD14^+$ monocytes, $CD19^+$ B cells) were performed. Immunostainings of lesional CLE biopsies evaluated the presence of T cells, B cells, plasmacytoid dendritic cells, macrophages, CXCR3, and CXCL10. Five subjects with CLE completed UVA1 radiation treatments, with no disease flares. Pruritus was the most commonly reported side effect. Clinical assessment scores, autoantibody levels, and peripheral blood immune cell composition remained stable. $CD8^+$ T cells and $CD20^+$ B cells decreased in treated CLE skin, but did not reach significance. Results demonstrated that low dose UVA1 radiation in patients with CLE was well tolerated. Future clinical trials will test moderate doses of UVA1 radiation to assess safety and efficacy in patients with CLE.

(8:51 AM)
PREDICTORS OF CLINICAL RESPONSE IN CUTANEOUS LUPUS: A LONGITUDINAL STUDY FROM THE UNIVERSITY OF TEXAS SOUTHWESTERN CUTANEOUS LUPUS REGISTRY
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While a broad range of therapies are available for patients with cutaneous lupus erythematosus (CLE), little is known on which factors distinguish patients most likely to respond to treatment. To identify patient factors associated with clinical response, we analyzed longitudinal observational data from patients with CLE enrolled in the University of Texas Southwestern Cutaneous Lupus Registry. These data include demographics, medication history, smoking history, and disease severity scores such as Cutaneous Lupus Activity and Severity Index (CLASI). Patients with baseline CLASI activity score ≥5 and ≥2 study visits were included in the analysis. We designated two clinical response endpoints based on relative decrease in CLASI activity score of ≥50% and ≥75% on consecutive visits. Univariate and multivariate logistic regression models identified patient factors associated with clinical response. 63 patients with 114 visit-pairs between 7/2009 and 9/2016 were analyzed. At the ≥50% CLASI activity reduction endpoint, higher initial CLASI activity (OR 1.12/CLASI point) and older age of CLE development (1.04/year) were associated with higher response rates, while smoking (0.35) and baseline use of steroid-sparing immunosuppressants (0.21) were associated with lower response rates. At the ≥75% CLASI activity reduction endpoint, older age of CLE development (1.06/year) and predominance of subacute CLE lesions (5.25) were associated with higher response rates, while smoking (0.11) was associated with lower response rates. More study visits are necessary to perform sub-analyses of different treatment modalities. This will help formulate evidence-based guidelines for CLE treatment ladders and establish patient inclusion criteria for clinical trials.

(8:59 AM)
AN EXAMINATION OF QUINACRINE USE AT THE HOSPITAL OF THE UNIVERSITY OF PENNSYLVANIA AND IN A POPULATION OF CLE AND DERMATOMYOSITIS PATIENTS
Lavanya Mittal, BA and Victoria P. Werth, MD

Background: Quinacrine is a compounded antimalarial that has demonstrated therapeutic promise in the treatment of rheumatic skin disease since the 1950's. This orphan drug is critically important for a subset of patients that are either intolerant or unresponsive to the first-line antimalarial, hydroxychloroquine. In light of the March 2016 PCAC meeting, however, the FDA may either prohibit compounding of quinacrine or tremendously restrict its access by requiring that the drug be maintained under an IND.
Objectives: These proceedings are a consequence of unfounded concerns regarding quinacrine safety as well as cursory perceptions that the population of patients who rely on quinacrine is a sparse minority. The number of individuals on quinacrine, however, is unclear because patients frequently pay for the drug entirely out-of-pocket. In this study, we aim to better define the patients on quinacrine at the Hospital of the University of Pennsylvania (HUP).
Methods: We retrospectively quantified the patients on quinacrine in the past year at the institution level. We also reviewed EPIC charts of patients in two prospectively collected longitudinal databases of CLE and dermatomyositis patients.
Results: At the entire hospital, 241 patients were prescribed quinacrine between June 2015-2016. We have HUP pharmacy records for 111 of these patients, of which quinacrine was prescribed to 63.1% (n=70) by dermatology, 34.2% (n=38) by rheumatology, and the remaining 2.7% (n=3) by internal medicine. Of the patients in the CLE (n=421) and dermatomyositis (n= 215) databases combined, 53.3% (n=317) have a history of quinacrine use out of which 49.8% (n=158) discontinued treatment after initially starting. 26.8% (n=42) of these discontinuations occurred due to problems with cost and access to the drug. In comparison, of the 558 database patients with a history of hydroxychloroquine, 43.7% (n=244) discontinued treatment after initiation. Of these cases, only 3.7% (n=12) occurred as a result of cost or impediments to access. Additionally, 95% (n=150) of patients that discontinued initial quinacrine treatment later restarted this medication, whereas only 46.7% (n=114) of those initially on hydroxychloroquine resumed treatment following discontinuation.
Conclusions: These results suggest that 1) quinacrine is being discontinued disproportionately more often than comparable drugs due to cost and access problems and 2) these discontinuations are occurring even when quinacrine may be benefiting patients because treatment is being largely resumed after discontinuation. Our study is limited by the retrospective nature of the data collection. However, based on these figures, it is evident that while the numbers of patients on quinacrine may not be immense, this is an important group whose needs must be better understood and addressed before a decision about the future of quinacrine should be made.

(9:07 AM)
USING THE AMERICAN COLLEGE OF RHEUMATOLOGY AND SYSTEMIC LUPUS INTERNATIONAL COLLABORATING CLINICS CRITERIA TO MEASURE DISEASE SEVERITY IN DISCOID LUPUS ERYTHEMATOSUS.
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Background: Discoid lupus erythematosus (DLE) progresses to systemic lupus erythematosus (SLE) in up to 28% of cases. The Systemic Lupus International Collaborating Clinics (SLICC) SLE criteria were developed to improve the American College of Rheumatology (ACR) criteria, but have not been assessed in DLE patients.
Methods: This was a retrospective study of 172 DLE patients enrolled in a cutaneous lupus database at the University of Pennsylvania. Patients were assessed for the presence of ACR and SLICC criteria using the database and their respective electronic medical records. The Fischer's

exact test was used for each criterion in the ACR and SLICC to determine if there was a difference between patients with DLE/SLE and DLE-only disease.
Results: Using the ACR criteria, 74 patients (52%) were classified as DLE/SLE and 68 (48%) as DLE-only, compared with 66 (46%) DLE/SLE and 76 (54%) DLE-only patients using the SLICC criteria (p=0.08). This net increase of eight patients meeting ACR criteria was due to the presence of the photosensitivity criterion and fewer immunologic criteria under ACR. Due to the immunologic criteria requirement under SLICC, it can be challenging to determine an SLE diagnosis retrospectively. Overall, DLE/SLE patients were more likely than DLE-only patients to exhibit significant systemic symptoms with regard to arthritis (ACR 73% *vs.* 9%, p<0.0001; SLICC 70% *vs.* 18%, p<0.0001), serositis (ACR 22% *vs.* 0%, p<0.0001; SLICC 22% *vs.* 3%, p<0.0001), and renal disorder (ACR 27 % *vs.* 2%, p<0.0001; SLICC 33% *vs.* 0%, p<0.0001) using both criteria. SLE was more common in generalized DLE than in localized DLE using ACR (p=0.0001) and SLICC (p=0.0068) criteria. DLE/SLE patients were more likely to have worse skin disease compared to DLE-only patients when classified according to ACR criteria, with 40.5% of DLE/SLE patients having CLASI activity ≥ 10 and 25.0% of DLE-only patients having CLASI ≥ 10 (Table 1).
Conclusions: DLE/SLE patients appear to have more significant internal disease than DLE patients who do not meet SLE criteria. Our findings trended toward the ACR criteria classifying more DLE patients with SLE than the SLICC criteria, particularly in patients without extensive laboratory testing. DLE-only patients may have significant skin disease with approximately 25% of DLE-only patients having moderate to severe skin disease.

	DLE with SLE n (%)	DLE without SLE n (%)	P-value
CLASI^{TM} ≥ 10	30 (40.5)	17 (25.0)	0.0493*
CLASI^{TM} < 10	44 (59.5)	51 (75.0)	

Table 1A. Skin activity in DLE/SLE vs DLE-only patients using ACR criteria. DLE/SLE patients are more likely to have worse skin disease compared to DLE-only patients when classified according to the ACR criteria.

	DLE with SLE n (%)	DLE without SLE n (%)	P-value
CLASI^{TM} ≥ 10	27 (40.9)	20 (26.3)	0.0653
CLASI^{TM} < 10	39 (59.1)	56 (73.7)	

Table 1B. Skin activity in DLE/SLE vs DLE-only patients using SLICC criteria. There is a trend of DLE/SLE patients having worse skin disease compared to DLE-only patients when classified according to the SLICC criteria.

(9:15 AM)
PRACTICE-BASED DIFFERENCES BETWEEN PEDIATRIC RHEUMATOLOGISTS AND DERMATOLOGISTS CARING FOR CHILDREN WITH DISCOID LUPUS
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* Both authors contributed equally to this project.

This study compared practice patterns among pediatric dermatologists and rheumatologists caring for children with DLE. A survey, e-mailed to 292 rheumatologists (Childhood Arthritis & Rheumatology Research Alliance [CARRA]) and 200 dermatologists (Pediatric Dermatology Research Alliance [PeDRA]), addressed initial laboratory screening for SLE, risk factors impacting screening strategy, and first/second-line systemic and topical therapies. Consensus was pre-defined as $\geq$70% agreement from both specialties. Fifty-three rheumatologists and 69 dermatologists responded (18% and 35% response rates respectively). There was no consensus on the choice of labs for initial SLE screening, but most respondents (rheum n = 42 [79%] vs derm n = 28 [41%]) chose the following panel: CBC/diff, renal/hepatic function, ESR, CRP, urine studies, complements, autoantibodies, anti-phospholipid antibodies. Of those who selected a partial laboratory work up (rheum n = 9 [17%]; derm n = 33 [48%]), only CBC (rheum n = 9 [100%]; derm n =32 [97%] and urinalysis (rheum n = 7 [78%]; derm n = 24 [73%]) achieved consensus. There was consensus that more thorough SLE screening is warranted in those with 1st degree relative with SLE, positive autoantibodies, arthritis or nephritis. Rheumatologists more often initiated hydroxychloroquine first line (rheum n = 24 [45%]; derm n = 9 [13%]) while dermatologists frequently started with topicals (rheum n = 16 [30%]; derm n = 50 [72%]). There was no consensus on choice of second-line systemic agents. This study demonstrates a lack of consensus between and among dermatologists and rheumatologists caring for children with DLE, underscoring the need for collaborative study.

ORAL SESSION II

(11:38 AM)

CANNABINOID REDUCES INFLAMMATORY CYTOKINES IN DERMATOMYOSITIS *IN VITRO*

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Background: Dermatomyositis is an autoimmune disease that affects the skin and muscles, and severely decreases quality of life. Available treatments for cutaneous dermatomyositis are frequently ineffective and/or have toxic side effects. We hypothesized that ajulemic acid, an experimental therapy, could suppress the production of inflammatory cytokines from immune cells of dermatomyositis patients *in vitro* and thus potentially provide a new therapeutic option for dermatomyositis. Ajulemic acid is a nonpsychoactive analog of tetrahydrocannabinol that selectively binds to a pro-resolving receptor on immune cells.
Methods: Peripheral blood mononuclear cells were isolated from 18 dermatomyositis patients and treated with increasing concentrations of ajulemic acid: 0, 3, 10 and 15 μM. The cells were incubated with lipopolysaccharide and CpG oligonucleotides to quantify the effect of ajulemic acid on the cellular production of tumor necrosis factor alpha (TNFα), interferon alpha (IFN-α), and interferon beta (IFN-b), key pathogenic cytokines in dermatomyositis. Cytokine production was measured by enzyme-linked immunosorbent assay.
Results: The lipopolysaccharide stimulated cells secreted mean (standard error) ln(TNFα) levels of 7.37 (0.38), 7.69 (0.39), 5.56 (0.39) and 4.16 (0.40) pg/ml at 0, 3, 10 and 15 μM concentrations of ajulemic acid, respectively. CpG stimulated cells treated with 0, 3, 10 and 15 μM ajulemic acid secreted mean (standard error) of ln(IFN-α) of 5.33 (0.53), 1.38 (0.57), 1.06 (0.75) and -0.03 (0.86) pg/ml. Compared to untreated cells, ajulemic acid suppressed cellular secretion of TNFα at 10 μM ($p=0.0002$) and 15 μM ($p<0.0001$). Ajulemic acid also decreased IFN-α ($p\leq0.0007$) and INF-b secretion from CpG stimulated cells at all studied ajulemic acid concentrations.
Conclusions: Ajulemic acid, an investigational, nonpsychoactive and pro-resolving cannabinoid suppressed secretion of TNFα, IFN-α, and IFN-b from immune cells of dermatomyositis patients *in vitro*. These cytokines are thought to be key immunostimulatory cytokines causing cutaneous dermatomyositis. Ajulemic acid may offer patients with cutaneous dermatomyositis a new therapeutic option that is potentially more effective and less toxic than the currently available treatments.

(11:46 AM)
INTERIM RESULTS OF AN OPEN-LABEL STUDY ASSESSING EFFICACY AND SAFETY OF H.P. ACTHAR GEL FOR THE TREATMENT OF REFRACTORY CUTANEOUS MANIFESTATIONS OF DERMATOMYOSITIS
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Introduction: Cutaneous dermatomyositis can be refractory to multiple medications, suggesting better treatments are needed. H.P. Acthar gel (Acthar) is a repository corticotropin injection that is FDA-approved for dermatomyositis. However, little is known about the safety and efficacy of Acthar for dermatomyositis. We are conducting an open-label study assessing efficacy and safety of Acthar for treatment of refractory cutaneous dermatomyositis. Here, results of the initial patients enrolled in the study are reported. **Methods:** Dermatomyositis patients with ≥ mild active disease despite prior treatment with ≥2 systemic agents were eligible for inclusion. **Results:** Five adult females, all with classic dermatomyositis, have been enrolled (Avg age 54yrs). Patients were treated with an average of 4.4+/-2.1 systemic medications prior to study entry, and were being treated with an average of 3.0+/-1.6 systemic medications at study entry. Average baseline mCDASI activity score was 20.2. Four patients completed ≥3 months of

treatment. At 3 months mCDASI activity scores improved an average of 8.25 +/- 4.14 points. Average baseline mCDASI damage score was 5.75, and there was an average increase of 1.5 +/- 1.32 at 3 months. In terms of patient-perceived effects, there was average improvement in DLQI score of 5.0 +/-4.7 at 3 months. Patients also noted average improvement in a 10-point global skin score of 3.25 +/-2.22, as well as an average improvement in a 10-point global itch score of 3.5+/-1.66 at 3 months. Adverse effects were mild. **Conclusions:** Patients on Acthar thus far have demonstrated improvements in cutaneous dermatomyositis with minimal adverse effects.

(11:52 AM)

COST COMPARISON OF POSITRON EMISSION TOMOGRAPHY VERSUS TYPICAL BROAD MALIGNANCY SURVEILLANCE IN DERMATOMYOSITIS

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Background: About 1 in 5 cases of dermatomyositis occurs as paraneoplastic disease. Often the malignancy occurs in organs which are not subject to routine screenings, such as the ovaries, lungs, or urinary tract. As debate over best practices in malignancy screening for these patients continues, positron emission tomography (PET) has been discussed as a single study alternative to more complex batteries of testing.[1] To our knowledge, cost comparisons between PET and typical malignancy screening tests in dermatomyositis patients have not been performed.
Objective: To compare the cost of PET and its variants to the cost of common practices in malignancy screenings in men and women with dermatomyositis.
Methods: The authors identified tests utilized in malignancy screening in men and women with dermatomyositis based on those used for comparison in Selva-O'Callaghan et al's original publication. In men, computed tomography (CT) of the chest, CT of the abdomen and pelvis, CEA and prostate specific antigen (PSA) testing were chosen. For women, CT of the chest, CT of the abdomen and pelvis, mammography, pelvic exam with cervical and vaginal cytopathology (PAP smear), CEA, CA 125, CA 19.9 and transvaginal/pelvic ultrasound were selected. PET variants included PET whole body, PET skull base to thighs, PET limited area, PET-CT whole body, PET-CT skull base to thighs, and PET-CT limited area for comparisons. The Current Procedural Terminology (CPT) codes for each study were identified. The MarketScan database, a collection of insurance claims data from 53 million Americans with private insurance, was queried for every instance of each test from 2005 until 2014 and the average cost of each was recorded. The mean sum cost of selected screenings in men and in women were compared with the cost of PET variants. Since there are multiple variants of mammography and pelvic exam with cervical/vaginal cytopathology, the mean cost of all variants per incidence were obtained as a representative value for comparison.
Results: The average cost of a broad panel of selected malignancy screening in men was $1,352.22 (across 1,324,913 instances) and $1,682.38 in women (24,792,926 instances). PET-CT, whole body had an average cost of $2,192.30 per incidence (n=45,379), or $509.92 more expensive than our selected panel of testing in women with dermatomyositis, and $840.08 more than in men ($p< 0.0001$). PET-CT skull base to thighs had a mean cost per incidence of $2,274.22, and PET whole body a mean cost of $1,597.07, or $85.31 less expensive than broader testing in women ($p< 0.0001$).
Conclusions: Previously published works have observed similar predictive values between PET-CT and other broad conventional screenings. While this study makes no effort to compare quality of screening, a single PET-CT whole body costs $509.92 more in women and $840.08

more in men than the screening panel described by Selva-O'Callaghan et al. Our study does not capture the indirect costs incurred on the patient, such as travel or time away from work, which might increase the patient's perceived value of a single PET-CT. While large sample size is a strength of this study, it does not include Medicare or Medicaid patients.

(12:00 PM)

THE CUTANEOUS AND SYSTEMIC FINDINGS ASSOCIATED WITH NUCLEAR MATRIX PROTEIN-2 ANTIBODIES IN ADULT DERMATOMYOSITIS PATIENTS

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To characterize the cutaneous and systemic clinical phenotype of dermatomyositis patients with anti-nuclear matrix protein-2 (NXP-2) antibodies, we conducted a retrospective cohort analysis of 178 dermatomyositis patients seen at the Stanford University Clinic. Electronic chart review employing a keyword search strategy was performed to collect clinical and laboratory data. Anti-NXP-2 antibodies were assayed by immunoprecipitation using NXP-2 produced by in vitro transcription/translation. Antibodies to NXP-2 were detected in 20 (11%) of the 178 patients. Anti-NXP-2 antibodies were associated with male gender (50% vs. 25%, p=0.02), dysphagia (74% vs. 39%, p=0.006), myalgia (89% vs. 52%, p=0.002), peripheral edema (35% vs. 11%, p=0.016), and calcinosis (37% vs. 11%, p=0.007). These patients were less likely to be clinically amyopathic (5% vs 23%, p=0.08). Five of the 20 patients with NXP-2 antibodies (25%) had an associated internal malignancy. No other cutaneous characteristics were associated with anti-NXP-2 antibodies except a decreased frequency of Gottron's sign (44% vs. 75%, p=0.012) and the fact that these patients were more likely to have mild skin disease. Dermatomyositis patients with anti-NXP2 antibodies have a distinct and often severe systemic phenotype that includes myalgia, peripheral edema and significant dysphagia despite having milder inflammatory skin disease. Because cancers also frequently occur in this population, the presence of anti-NXP-2 antibodies should alert the clinician to vigilantly screen for malignancy as well as be prepared for severe dysphagia in this subgroup.

(12:08 PM)

A LONGITUDINAL STUDY OF CUTANEOUS DERMATOMYOSITIS

Peter Chansky, Victoria Werth

Background: Previous studies have described the course of dermatomyositis (DM) using muscle weakness and enzymes as their primary endpoints. Limited studies have described the course of cutaneous disease in DM.
Objective: To characterize the disease course in cutaneous DM.
Methods: A retrospective cohort study was performed. Patients 18 years or older with clinical or histologic evidence of DM who had the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) activity subscores recorded for at least 2 years from baseline were included. Statistical methods were used to determine average disease activity, overall disease progression, disease course, and variability. Disease progression was classified into improved,

worsened, or stable based on a criteria which combines the net area under the curve per unit time relative to baseline CDASI score and a fitted linear slope. Disease course was classified into monophasic (significant improvement in skin disease without a flare), polyphasic (significant improvement in skin disease with at least one flare), or chronic (significant worsening in skin disease without a significant improvement) based on prior classification in the literature. Subjects were divided into two groups based on disease severity at baseline (mild versus moderate-severe disease activity). Outcome measures were then compared between groups.
Results: Our final cohort consisted of 40 patients with DM. The majority of the patients were female (90%) and Caucasian (95%), with a mean age of 52.9 years at baseline. Disease subtype was classified as classic in 52.5% of patients and skin predominant in 47.5%. Mean follow-up time was 3.50 years. Patients with moderate-severe disease activity at baseline made up a majority of the patients (N=24, or 60% of patients) while those with mild disease activity at baseline made up 40% (N=16) of the patients. Average disease activity over time was different significantly between the mild and moderate-severe groups (9.10 versus 14.96; P = 0.004). The majority of DM patients experienced an improvement in disease activity (N=23, 57.5%). A worsening in disease activity was seen in 8 DM patients (20%) and stable disease activity was seen in 9 DM patients (22.5%). Patients with mild disease activity at baseline whose disease activity remained stable made up a majority of this subgroup (N=8, 50%) while patients with moderate-severe disease activity at baseline whose disease activity improved made up a majority of this subgroup (N=20, 83%). The majority of DM patients had a polyphasic disease course (N=33, 82.5%) with a monophasic disease course seen in 5 DM patients (12.5%) and a chronic disease course seen in 2 DM patients (5%). Variability in disease activity over time, evaluated by calculating the average flares/yr, was independent of baseline disease activity.
Limitations: This is a retrospective study. Our center is a tertiary care academic medical center, and it is possible that our cohort could have had greater disease severity due to referral bias. However, the difference in CDASI we observed between groups shouldn't be affected by such bias. Also our results were not adjusted for potentially confounders as they were not collected in our data. The adjusted analysis should be warranted in future studies.
Conclusion: The majority of our patients had moderate-severe disease activity at baseline that tended to improve with a polyphasic course. Baseline CDASI activity score is associated with particular patterns of disease progression and disease course in patients with cutaneous DM. Most patients with a mild baseline activity score remained stable with a polyphasic course while most patients with a moderate-severe baseline activity score tended to improve with a polyphasic course.

ORAL SESSION III

(1:30 PM)
SYSTEMIC TREATMENT FOR CLINICALLY AMYOPATHIC DERMATOMYOSITIS: A RETROSPECTIVE COHORT STUDY AT THREE TERTIARY CARE CENTERS

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Clinically amyopathic dermatomyositis (CADM), characterized by pathognomonic cutaneous findings without muscle weakness, is an important subset and accounts for 20% of patients with dermatomyositis (DM). In patients with CADM, limited literature exists regarding treatment specifically for cutaneous disease. Our study investigated the use of systemic therapy for skin disease in CADM at three tertiary care centers. Using the Partners Healthcare Research Patient Data Registry and New York University medical record systems, we reviewed the medical records of all patients with CADM treated between 2010 and 2016. Data collected included demographics, referral types, laboratories, medications used and adverse effects. 95 patients with CADM (76 amyopathic and 19 hypomyopathic) were identified. The mean age of diagnosis was 49.7 years. Most patients were referred from dermatology (51%) and rheumatology (31.5%) providers. Only 18% of patients were treated with hydroxychloroquine and topical therapy alone, while 82% required at least one immunosuppressive therapy to control their cutaneous disease. Among these additional therapies, methotrexate (52.6%) and mycophenolate mofetil (41.1%) were the most commonly used. 31.6% of patients required treatment with IVIG for adequate control of skin disease. Consistent with existing literature, 23.2% of patients developed a cutaneous hypersensitivity reaction to hydroxychloroquine. In conclusion, cutaneous disease is refractory to antimalarials in the majority of CADM patients. This study emphasizes the recalcitrant nature of DM skin disease, and highlights that aggressive therapy is often warranted on the basis of cutaneous involvement, even in the absence of muscle disease.

(1:42 PM)

CLINICAL CHARACTERIZATION OF ITCH IN DERMATOMYOSITIS AND THE ROLE OF INCREASED SKIN INTERLEUKIN-31

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Background/Purpose: Interleukin-31 (IL-31) has been implicated in pruritus associated with various itchy skin diseases, including atopic dermatitis and cutaneous T cell lymphoma. While pruritus is a prominent feature in dermatomyositis (DM), there are few studies to evaluate clinical characteristics and pathogenesis of itch in DM. We examined the prevalence and severity of pruritus in patients with DM, and investigated the presence of IL-31 and IL-31 receptor in skin tissue to explain the pathomechanism of itch in DM patients.
Methods: Pruritus and disease activity of DM were evaluated by a visual analog scale (VAS) and the Cutaneous Disease and Activity Severity Index (CDASI), respectively. Gene expression of IL-31 and IL-31 receptor alpha (IL-31RA) in lesional DM skin was evaluated by qRT-PCR, and was compared with that of non-lesional DM and healthy control (HC) skin. Immunohistochemical analysis assessed IL-31 expression in skin tissue. The Spearman rank test

was used to evaluate the relationship between itch intensity and disease activity, as well as itch intensity and lesional gene expression of IL-31 and IL-31RA. The Kruskal-Wallis test with Dunn's post hoc test was used to compare the difference of IL-31 and IL-31RA mRNA expression, and immunohistochemical analysis of skin IL-31 expression in DM and HC. **Results:** About half of 164 patients with DM (25 male, 139 female; 61 classic DM and 103 clinically amyopathic DM; mean age ± SD 52.5 ± 14 years) had moderate to severe itch (28.66% moderate, 20.73% severe itch). Pruritus in DM was positively correlated with disease activity, with a correlation coefficient of 0.337 between VAS itch score and CDASI activity score ($p<0.01$). Skin IL-31 and IL-31RA gene expression was significantly up-regulated in DM compared to HC ($p<0.05$). IL-31 mRNA expression was positively correlated with VAS itch score ($r= 0.6748$, $p=0.039$). Immunoreactivity for IL-31 was also stronger in lesional skin of DM ($p=0.0001$).
Conclusion: In conclusion, we confirmed that itch is a prevalent symptom in many patients with DM involving the skin, and that skin IL-31 is significantly higher in DM than in HC. This is the first study to suggest IL-31's crucial role in the skin for pruritus in DM.

(2:06 PM)
IMMUNE DYSREGULATION IN MORPHEA
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Background: Morphea, a disorder that may produce devastating cosmetic and functional impairment, is characterized by active inflammatory lesions followed by sclerosis in the skin and soft tissue. The molecular underpinnings of morphea are poorly understood, limiting development of targeted therapies. **Objective:** To use gene expression profiling to characterize and compare dysregulated immune pathways in active and inactive morphea and to compare morphea gene expression with that in systemic sclerosis. **Methods:** Case control study of whole skin specimens from 17 morphea patients (including x lesional and y site matched non lesional skin taken from inflammatory and sclerotic lesions). Transcriptional profiling was performed on Illumina gene chips using standard methods. DEG were identified at a cut off of FC >1.5 and FDR <0.5. Pathways of interest were identified using Ingenuity Pathway Analysis and modular analysis. ComBat software was utilized to merge microarray data sets and compare morphea gene expression to that in systemic sclerosis (SSc), specifically the 5 SSc subsets described by Whitfield, et al. **Results:** The top upregulated pathways in samples from the inflammatory border were interferon signaling, antigen presenting pathway, and pattern recognition receptors; while in sclerotic samples OX40, leukocyte extravasation, and hepatic fibrosis/hepatic stellate cell activation predominated. All morphea skin samples aligned most closely with the inflammatory subset of SSc, and showed little overlap with proliferative or normal like profiles of SSc. **Conclusions:** Interferon and inflammatory pathways predominate in all morphea lesions, while a proliferative profile TGF beta) is present in a minority of sclerotic lesions.

(2:18 PM)
PERCEPTIONS OF MORPHEA IN CHILDREN COMPARED TO THEIR PARENTS
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The disease burden of morphea may be perceived differently by parents than their affected children. Our group has previously reported that children generally had mild to moderate impact on life quality due to morphea. However, the perception of disease impact by children compared to their parents has not been investigated. The purpose of this study is to investigate the differences in perception of morphea impact in children and their parents. Parents' perspectives represented by Short Form Health Survey-10 (SF-10) were compared to their childrens' represented by the Children's Dermatology Life Quality Index (CDQLI) from participants in the Morphea in Adults and Children (MAC) cohort. Demographical information and clinical data including the Localized Scleroderma Activity and Damage indices (LoSAI, LoSDI), physician global scores for activity and damage (PGA-A, D), and disease modifiers were collected. 75 MAC participants met inclusion criteria for this study. Mean age was 11.52, 60% were female, and 75% were Caucasian. CDQLI and SF-10 physical health summary scores were positively correlated ($r=0.5776$, $p=0.0039$) in age less than 10, and not correlated in age greater than 10 years old. CDQLI and SF-10 psychosocial summary scores were negatively correlated however this did not reach significance. There was a trend towards a discrepancy between parents and their children in their perception of disease impact. In regards to disease activity, an increase in disease activity had greater impact on life quality among children and parents. In contrast, disease damage did not correlate with life quality in the perceptions of child or parent.

(2:30)

HIGH FREQUENCY ULTRASOUND: A NOVEL INSTRUMENT TO QUANTIFY GRANULOMA BURDEN IN CUTANEOUS SARCOIDOSIS.

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Background: As is the case for many skin diseases, cutaneous sarcoidosis does not currently have an objective measure of disease burden to establish disease severity and response to treatment. The disease has traditionally been assessed by visual skin changes, including induration and erythema; however, such assessments may fail to quantify the total skin granuloma burden, as the majority of the granulomatous inflammation may lie deep within the dermis and not be reliably detected by sight or palpation. **Objectives:** The purpose of this pilot study is to evaluate the feasibility of high frequency ultrasound as an objective measure of granuloma burden in cutaneous sarcoidosis and to compare high frequency ultrasound to a previously validated clinical instrument (CSAMI) and histopathology evaluation. **Results:** A strong correlation was observed between the mean brightness of high frequency ultrasound images and both the lesional CSAMI score (Spearman's rho: 0.9710, $p = 0.0012$) and percent of dermis with granulomas histopathology (Spearman's rho: 0.8407 $p = 0.0361$). **Conclusions:** These results suggest high frequency ultrasound is a valid, objective measure of granuloma burden in cutaneous sarcoidosis and represents a novel, non-invasive measure of disease severity that correlates to the previously validated CSAMI clinical severity score and histopathology evaluation.

(2:42 PM)
SCLERODERMA AMONG PATIENTS WITH BREAST CANCER
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Connective tissue disorders such as scleroderma have been reported in association with solid malignancies. However, the exact relationship, risk factors, and features of these patients have not been established.
Objectives: The aim of this study was to characterize the clinical features and described the outcomes among patients with breast cancer, with an associated diagnosis of scleroderma (SSc).
Methodology: We conducted a retrospective study by reviewing the records of patients diagnosed with breast cancer at Duke University Hospital from 1991 to 2011. Clinical features, treatment, and outcomes were reviewed.
Results: Out of a total of 11,764 patients diagnosed with breast cancer within that time period, a total of 24 patients also had an associated diagnosis of systemic sclerosis, whereas only 1 patient had an associated diagnosis of morphea. Fourteen (14) of the 24 patients were diagnosed with SSc at least 1 year or more after their initial breast cancer diagnosis. Seven (7) out of the 24 patients had undergone radiation therapy, and 2/7 patients reported skin associated complications from radiation. Both were mild reactions. Median years of mortality is 8 years. 11/24 patients were deceased. Median mortality was 8 years from the diagnosis of breast cancer.
Conclusion: The incidence of scleroderma among our patients with breast cancer is markedly higher compared to what was would be expected in the general population. In contrast to previously published data, majority of the patients in this study were diagnosed with SSc after, and not prior to, the diagnosis of breast cancer. Further studies to elucidate the causes behind this relationship are required.

(2:52PM)
DIAGNOSTIC EVALUATION AND MANAGEMENT OF CLASSIC ULCERATIVE PYODERMA GANGRENOSUM: A SURVEY OF U.S. DERMATOLOGISTS
Ladan Afifi MS[1], Alex G. Ortega-Loayza MD[2], Kanade Shinkai MD, PhD[1] on behalf of the Society of Dermatology Hospitalists
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(WITHDRAWN)

CASE PRESTATIONS

(3:30 PM)
"BONITA" LEPROSY WITH LUCIO'S PHENOMENON MISDIAGNOSED AS UNDIFFERENTIATED CONNECTIVE TISSUE DISEASE
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The patient is an 18-year-old woman originally from Cuba with 5-year history of "undifferentiated connective tissue disease" and "fibromyalgia" on periodic pulse steroids. The patient had visited multiple rheumatologists for symptoms of fatigue, alopecia of the frontal

scalp, eyebrows and body, painful swelling of face, hands and feet, and intermittent purpuric plaques on the bilateral legs. Serologic workup included positive ANA, anticardiolipin IgM, Lupus anticoagulant, β2 glycoprotein IgM, Rheumatoid Factor, pANCA, and cryoglobulins. The patient presented to the ED for worsening rash on her lower extremities, now associated with ulceration. On physical examination, she was noted to have prominent cheeks and earlobes, madarosis, and dysesthesia of the face and extremities, as well as purpuric and ulcerated plaques on the lower legs. Biopsies were performed from the eyebrow and leg, and revealed confluent acid-fast bacilli; biopsy of lower leg additionally showed heavy infiltration of the acid-fast bacilli in the vessels and multiple fibrin thrombi. The patient was diagnosed with "Bonita" leprosy with Lucio's phenomenon, and started on antimicrobial therapy with rapid improvement in her symptoms. "Bonita" leprosy is a rare manifestation of early, diffuse lepromatous leprosy, characterized by deep tissue infiltration of acid fast bacilli that causes swelling and softening of facial lines and wrinkles. Infiltration of joint space can also lead to stiffness, pain, and swelling, mimicking inflammatory arthropathies. Serology for autoantibodies may be positive as in other multibacillary forms of leprosy. Treatment with immunosuppression without antimicrobials can precipitate in potentially life-threatening Lucio's phenomenon, as in this patient.

(3:36 PM)

HAIRY CELL LEUKEMIA PRESENTING AS "INCOMPLETE SYSTEMIC LUPUS"

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Background: Multiple eruptive dermtofibromas is a rare clinical variant of dermatofibroma that can be associated with an underlying immune mediated disease such as systemic lupus erythematosus (SLE), Sjogrens syndrome, HIV, myasthenia gravis and Hashimoto's thyroiditis.
Case report: We describe a 34 years old healthy Asian Indian man who presented to our dermatology outpatient clinic with 6 months history of rapidly developing multiple, firm, brown papules distributed on chest, arm and legs, histopathologically proven as dermatofibroma. He was found to have bicytopenia with positive autoimmune markers, suggestive of "incomplete systemic lupus". The bone marrow biopsy later revealed hairy cell leukemia as a cause of his bicytopenia and paraneoplastic autoimmunity.
Conclusion: "Incomplete systemic lupus" can be due to conditions other than incipient SLE, such as medications, chronic infections or an underlying malignancy. Our case highlights the potential cognitive bias for attributing a patient to having SLE (Sontheimer's Corollary), as well as the new association between eruptive dermatofibromas, paraneoplastic autoimmunity and hairy cell leukemia.

(3:42)

A CASE OF LIVEDOID VASCULOPATHY WITH ULCERS: SUCCESSFUL RESPONSE TO CLOPIDOGREL

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Treatments of livedoid vasculopathy (LV) range from fibrinolytics, vasodilators, anti-coagulants, and anti-platelet agents. Anti-platelet agents are promising treatments because they treat the vaso-occlusive nature of LV. We present a case of dramatic improvement of LV with clopidogrel,

an anti-platelet agent with little reported use in LV. A 35 year-old woman with a history of interstitial lung disease and undifferentiated connective tissue disease presented to University of Texas Southwestern Medical Center with a 2 month history of a painful, progressive rash on her lower extremities. Physical examination revealed stellate, non-inflammatory retiform purpuric patches with central fibrinoid necrosis on the bilateral medial arches of the plantar foot and heel. Serological studies were positive for PL-12, SS-A antibodies, and low complement C3 and C4. A punch biopsy showed occlusion of small vessels with hyalinized thrombi, consistent with LV. Over the next few weeks the rash progressed into exquisitely tender ulcerations. She was treated with prednisone, aspirin, azathioprine, and dapsone with only slight improvement. Three months after presentation, she was started on clopidogrel 75 mg daily. One month after starting clopidogrel, there was marked improvement of her ulcers and no new lesions. After five months of treatments, all ulcers completely re-epithelialized. We postulate that the anti-platelet aggregation properties of clopidogrel, which hasten re-canalization of the small vessels, and prevent further vaso-occlusion, contributed to the healing of our patient's ulcers. While clopidogrel is a widely available and affordable treatment option, larger studies are necessary to confirm the therapeutic potential of clopidogrel in LV.

(3:48)

A CASE-BASED DISCUSSION OF CUTANEOUS MUCINOSES

Michael Roman, BS[a]; Joanie Pinard, MD[b]; Drew Kurtzman, MD[b]; Keith Morley, MD[b]; Ruth Ann Vleugels, MD, MPH[b*]; Alisa Femia, MD[a*]

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Primary cutaneous mucinoses are a diverse group of disorders characterized by excessive dermal mucin.[1] The cause of the abnormal deposition of mucin is unclear, but may be related to an increase in glycosaminoglycan synthesis. Amongst the many forms of primary cutaneous mucinoses, perhaps the most notable distinction is between the localized form (localized variants of lichen myxedematosus), and the generalized form (scleromyxedema).[1] The localized forms are further subdivided into four main subtypes: 1) discrete papular, 2) nodular, 3) acral persistent papular mucinosis, and 4) papular mucinosis of infancy; some authors consider self-healing popular mucinosis to be a fifth subtype.[2] The diagnostic criteria for these subtypes includes an absence of any systemic disease including gammopathies and/or thyroid disease.[2] On the other hand, scleromyxedema is characterized by a generalized papular eruption in the absence of thyroid disease.[3] Sclerodermoid features may develop, particularly in long-standing disease. An important and distinguishing feature of scleromyxedema is the presence of a monoclonal gammopathy (typically an IgGλ paraproteinemia), observed in more than 80% of patients.[3] Distinguishing this form from localized variants is imperative due to a host of associated extracutaneous manifestations including cardiovascular, gastrointestinal, pulmonary, rheumatologic, and central nervous system involvement. However, despite existing diagnostic criteria and classification systems, the diagnosis of various cutaneous mucinoses remains a challenge, particularly due to the existence of many cases with overlapping and atypical features. Furthermore, treatment of these conditions is challenging, and there is a lack of literature to guide optimal evaluation and management.

(3:52)

A CASE-BASED DISCUSSION OF ANTI-MELANOMA DIFFERENTIATION-ASSOCIATED GENE 5 (MDA-5) DERMATOMYOSITIS

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Dermatomyositis (DM) is an uncommon connective tissue disease characterized by a distinctive cutaneous eruption and proximal inflammatory myopathy. Recent studies have highlighted the importance of autoantibodies in DM, as certain antibodies have been shown to reliably predict specific clinical phenotypes.[1] The presence of anti-melanoma differentiation-associated gene (MDA-5) antibodies in individuals with DM exemplifies this clinical-serologic phenotypic correlation. In addition to typical cutaneous DM findings, patients with anti-MDA-5 DM characteristically display skin ulceration involving Gottron's papules and other sites of trauma including the elbows, digital pulp, and nailfolds, as well as erythematous, painful palmar papules and macules, alopecia, panniculitis, and oral ulcers.[2] Apart from these unique cutaneous features, anti-MDA-5 DM is strongly associated with rapidly progressive interstitial lung disease and poor overall survival related to pulmonary complications.[3] Additionally, anti-MDA-5 DM exhibits a higher propensity for arthritis as well as lower rates of myopathy. Occlusive vasculopathy affecting the microvasculature is thought to be responsible for the cutaneous ulceration observed in anti-MDA-5 DM,[2] and may also contribute to interstitial lung disease. While studies are lacking, our experience with this DM subtype suggests that in addition to the institution of typical immunosuppressive and immunomodulatory therapies, vasodilatory medications are also warranted in order to effectively manage the disorder and likely exert their beneficial effect by targeting the underlying vasculopathy. Recognition of the characteristic clinical features of anti-MDA-5 DM may facilitate an early diagnosis allowing for appropriate management, and may ultimately reduce the morbidity and potential mortality associated with disorder.