Ablative fractional laser resurfacing for treatment of scars and contractures in chronic GVHD

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Introduction: Chronic cutaneous graft versus host disease (GVHD) can be characterized by thickening and tightening of the skin, so-called “sclerotic” skin changes. Advanced sclerosis can lead to joint contractures and reduced range of motion (ROM), which can be severely limiting. There are no therapies proven to reverse these skin changes. In this study, we used the ablative fractional CO2 laser to treat areas of sclerosis affecting joints, an approach which has been reported to be successful in patients with joint contractures due to linear morphea or burn scars. Methods: Patients underwent three treatment sessions with the ablative fractional CO2 laser spaced one month apart. Range of motion measurements, photographs, and high-resolution ultrasound were performed at baseline, one week after each laser session, and three months following the final laser session. Skin biopsy and patient and provider functional assessment were performed at baseline and three months following the final laser session. Results: Eight patients enrolled, and 6 completed the study. The treatment was generally well-tolerated, with one patient dropping out due to transient exacerbation of pre-existing lower extremity neuropathy. Range of motion measurements, particularly supination and pronation, improved in all patients. High resolution ultrasound showed increased echogenicity of the dermis three months after the final treatment, suggesting remodeling of collagen. Immunohistochemical analysis of skin biopsies showed breaking up of thick and elongated collagen bundles into smaller sizes, decreased intensity of trichrome collagen staining, and greater abundance of fetal-type collagen. Patients reported subjective functional improvement and appearance of treated areas. Discussion: Based on the results of this pilot study, we conclude that the fractional ablative CO2 laser is generally well-tolerated in this group of medically-complex patients. It appears to modify or remodel scar-like collagen, resulting in improved range of motion in these patients, particularly with respect to twisting motion (supination and pronation). Because we treated only small areas limited to the immediate vicinity of involved joints, and motion involves recruitment of tissue from surrounding proximal and distal areas, we speculate that treatment of larger areas, such as whole or partial limbs, may lead to greater response. We further speculate this technique could be combined with post-treatment application of topical therapies, potentially with improved efficacy.

A longitudinal study of cutaneous dermatomyositis

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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 of 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 criteria combining 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 skin improvement without a flare), polyphasic (significant skin improvement with at least one flare), or chronic (significant skin worsening without a significant improvement) based on the literature. Subjects were divided into mild and moderate-severe disease severity at baseline. Outcome measures were 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. More patients had moderate-severe disease activity at baseline (N=24, 60%) compared to mild disease activity at baseline (N=16, 40%). Average disease activity over time was significantly different between the mild and moderate-severe groups (9.1 vs. 14.96; P = 0.004). The majority of DM patients experienced an improvement in disease activity (N=23, 57.5%) compared to a worsening (N=8, 20%) and stable (N=9, 22.5%) progression. Within the mild subgroup, a majority of the patients’ disease activity remained stable (N=8, 50%) while in the moderate-severe group a majority showed improvement in disease activity (N=20, 83%). The majority of DM patients had a polyphasic disease course (N=33, 82.5%) compared to monophasic (N=5, 12.5%) and chronic (N=2, 5%) courses. Variability in disease activity over time, evaluated by calculating the average flares/yr, was independent of baseline disease activity. Conclusion: The majority of our patients had moderate-severe disease activity at baseline that tended to improve with a polyphasic course while those with mild baseline activity scores tended to remain stable with a polyphasic course. Baseline CDASI activity score is associated with particular patterns of disease progression and disease course in patients with cutaneous DM.
Dermoscopic patterns of acral melanocytic nevi and acral melanoma requiring biopsy versus close clinical follow up

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The dermoscopic patterns for acral melanocytic nevi (AMN) and acral melanoma (AM) are well characterized. This study aims to characterize the dermoscopic patterns of plantar acral lesions that were either followed clinically or biopsied. A retrospective pathology review of AMN and AM with dermoscopic photographs biopsied between 2000 and 2016 at Mayo Clinic Arizona was performed. An additional photographic search for AMN followed clinically during the same time frame was performed. 130 plantar AM and AMN with dermoscopy were reviewed. A total of 76 acral lesions were not biopsied and 54 lesions were biopsied (41 benign nevi, 5 atypical nevi, and 8 AM). The most common dermoscopic patterns were lattice-like (22.3% total, 15 biopsied, 14 followed), parallel furrow (20.0% total, 5 biopsied, 21 followed), and non-typical (20.0% total, 7 biopsied, 19 followed). Less common benign patterns were globular (7.7%, 4 biopsied, 8 followed) and homogeneous (9.2%, 2 biopsied, 8 followed). The parallel ridge (4.6% total, 6 biopsied, 0 followed) and multi-component (1.5% total, 2 biopsied, 0 followed) patterns accounted for the majority AM (75%). The parallel furrow pattern was more likely to be followed clinically (p<0.05); while, the non-typical, globular, and homogeneous patterns trended towards less frequent biopsies. All multi-component and parallel ridge patterns were biopsied. Interestingly, the lattice-like and fibrillar patterns were biopsied in 52% and 69% of cases respectively. In conclusion, although the parallel furrow, lattice-like and fibrillar are the most common benign patterns for AMN, the biopsy rates of lattice-like and fibrillar were relatively high. The parallel furrow, lattice-like, and regular fibrillar patterns are benign patterns that do not require a biopsy. Due to mechanical alterations of fibrillar pattern nevi, they can be difficult to interpret. Examination at an oblique angle, parallel to the tension vector provides easier viewing and more clearly shows the furrow origin of pigmentation. Dermoscopic patterns that do not fit a benign pattern, are large (>7mm), or show a parallel ridge pattern should be biopsied. Additional education on the benign nature of lattice-like and how to properly view fibrillar may help reduce unnecessary biopsies. The parallel ridge and multi-component patterns were biopsied 100% of the time and accounted for 6/8 AM, further demonstrating these patterns are indicative of malignancy.

Improvement in Quality of Life with the Goeckerman Regimen in Patients with Moderate-to-Severe Psoriasis: An Interim Report

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Introduction: Psoriasis is an inflammatory skin disease affecting 2-3% of the population that can have significant negative impact on quality-of-life (QoL). Measuring patient-reported outcomes of QoL is an important part of assessing therapeutic efficacy of psoriasis treatments. Skin-specific and nonskin-specific QoL measures in Goeckerman therapy have not been studied. Objective: To assess improvement in QoL of patients undergoing Goeckerman therapy using validated dermatologic and non-dermatologic psychometric instruments in patients with moderate-to-severe psoriasis. Methods: At the University of California San Francisco Psoriasis Center, patients with moderate-to-severe psoriasis were treated with the Goeckerman regimen 5 days a week for 6 weeks. The Goeckerman regimen involves daily phototherapy, followed by application of coal tar under occlusion for 4-5 hours. QoL was measured at weeks 0, 2, 4, and 6, and after treatment at week 12. The validated tools used to measure QoL were Psychological General Well-Being scale (PGWB), Psoriasis Quality of Life – 12 items (PQOL-12), and Dermatology Life Quality Index (DLQI). Improvement in psoriasis was measured by the Psoriasis Area and Severity Index (PASI) and Physician Global Assessment (PGA) at each time point. Results: Currently, 3 patients have completed this study. We anticipate we will have complete data from 5 patients at the time of this presentation. From the data to date, average PGWB score improved from 55.3 at baseline (week 0) to 88.3 at week 6 (p=0.0469) and to 99.0 at week 12 (p=0.0494). The average PQOL-12 score improved from 94.3 at baseline to 31.3 at week 6 (p=0.0144) and to 33.0 at week 12 (p=0.1014). The average DLQI score improved from 15.0 at baseline to 7.0 at 6 weeks (p=0.0202) and to 4.0 at 12 weeks (p=0.0315). At baseline, the average PASI was 28.2. PASI-75 was reached by the 3 patients at week 6 and maintained at week 12. Average IGA was 4 at baseline and 1.3 at week 6, which was also maintained at week 12. The only adverse events were mild phototoxic reaction as well as mild skin irritation. Conclusion: Improvement in QoL as measured by skin-specific and nonspecific tools have shown the positive effect of Goeckerman treatment on psychological well-being. Similar to many psoriasis therapies that have shown to improve QoL, the Goeckerman regimen remains a viable treatment option for patients with moderate-to-severe psoriasis. The study will continue to enroll a target of 30 patients.

Patch Testing with an extensive metal allergen series: Results from 2130 patients at Massachusetts General Hospital between: 2006-2016

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Background: Metal-containing devices are increasingly used in medicine. The standard series for diagnosing metal allergy currently does not include many metals that are commonly used and found in the environment. Objective: Describe results of patch testing in a large patient cohort to an extended series of 45 metal haptens. Methods: Patch testing with Chemotechnique allergens was performed on patients sequentially referred to a tertiary patch testing clinic from July 2006 to September 2016. Data was collected retrospectively in Microsoft Access from chart review. Results: Over a ten-year period, the metals with the highest rates of sensitization were gold sodium thiosulfate 0.5 pet (30.4%), nickel sulfate 2.5% (24.6%), palladium chloride (21.3%), nickel sulfate 5% (18.0), gold sodium thiosulfate 2.0 pet (15.7%), and manganese chloride 2.0 pet (11.3%). Metals with no reactions were: aluminum, ammonium hexachlororidate, ammonium hexachloroplattinate, ammonium molybdatehydrate, ammonium tetrachloroplattinate, indium chloride, indium sulphate, lead chloride, silver nitrate, titanium nitride, and tungsten. Metals with low rates of allergic patch test reactions (<1%) included cadmium chloride, calcium titanate, lead acetate trichydrate, stannous oxalate, zinc, molybdenum, titanium, and tin. Conclusions: Currently, the metals that are commonly assessed for allergic reactions include: nickel, cobalt, gold, and potassium dichromate. Little is known about the sensitivity rates of other metals found in the environment. These study results may help to guide future diagnostic testing for patients undergoing metallic device implantation surgeries, as no consensus regarding implant metal test series currently exists.
Intravenous immunoglobulin as adjunct therapy for refractory pyoderma gangrenosum: Case series and systematic review

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Introduction: Pyoderma gangrenosum is an ulcerative cutaneous disease commonly treated with steroids. The use of IVIG for refractory disease has been reported in the literature. In this study, we present 5 patients with IVIG treated at our institution and perform a systematic review of published cases, with an aim of understanding the efficacy and appropriate dosing of this medication. Methods: A systematic review was performed using PubMed with the search terms: “pyoderma intravenous immunoglobulin”, “pyoderma immunoglobulin”, “pyoderma gangrenosum treatment”, and “cutaneous ulcer intravenous immunoglobulin”. MESH terms for “pyoderma gangrenosum” and “ivig”, “intravenous immunoglobulin”, and “IV immunoglobulin” were also searched. All English-language studies and case reports were reviewed. Pediatric cases and cases that did not include treatment dose, response, or patient information were excluded. Concurrently, all patients with PG treated at Massachusetts General Hospital or Brigham & Women’s Hospital between 2000 and 2015 were manually reviewed for identification of patients with PG treated with IVIG. Results: Our study review identified 37 published cases and 5 clinical cases from our institution. All patients had refractory PG and received IVIG in addition to systemic steroids. 4 patients were reported to have no response. The complete response rate was 64% (CI % 40% - 78%), and the combined complete and partial response rate was 90% (CI 81% - 99%). Fisher exact comparison of response (complete or partial) to 2g/kg dose vs lower doses was not statistically significant (p=0.107). All comparisons between doses above 0.4mg/kg did not produce statistically significant results. In most publications, response to treatment was observed within the first month. Some studies reported initial reduction in pain and halting lesion progression before gradual healing over several weeks to months. Duration of treatment ranged between 1 dose and 4 years. On average, IVIG was continued for 6 months with a median of 3 months. Discussion: Our systematic review suggests a potential role for IVIG as an adjuvant therapy for refractory PG. Our literature review and clinical experience, although promising, is largely biased by publication because of the limited data, lack of randomized trials, and propensity to publish positive studies. Prospective clinical trials testing the response of IVIG for refractory PG are needed to validate these findings and guide treatment.

Long-term improvement in three cases of recalcitrant Darier’s Disease with photon and electron beam radiotherapy

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Darier’s Disease is a rare autosomal dominant genodermatosis characterized by acantholysis and dyskeratosis of the skin, nails and mucosa. It is characterized by hyperkeratotic greasy papules that coalesce into large malodorous vegetative masses on seborrheic sites, particularly in the flexures. Dermatology life quality index can be as high as 19.6% and mortality can result secondary to recurrent severe infections. Management is difficult and treatments are unsatisfactory. The first case that used radiotherapy (RT) dates back to the 1950’s, but was reported to have only a temporary anti-inflammatory effect with x-rays and grenz rays. The longest known length of remission with RT was 18 months. The only other report was incidental, lasting 9 months after thoracic radiation in a cancer patient. Remission with erbium YAG lasers, another promising alternative, in one report, lasted 8 to 20 months. We describe three cases of severe, recalcitrant Darier’s treated with intensity-modulated photon and electron beam RT that resulted in remission for at least 1 year and up to 4 years 2 months. Individualized dose intensity patterns conforming to lesions were developed using CT and MRI to minimize irradiation of critical structures. The adverse effects were limited to pain, erythema, itching, flaking, hyperpigmentation, and cellulitis. Acute side effects subsided on follow-up and were replaced with skin sloughing and healthy skin growth. In support of this data, we have also used RT to treat a patient with Hailey-hailey’s disease (HHD), demonstrating decreased intensity of lesions without requiring retreatment for more than 4 years. Studies on HHD suggest that effects of RT may be attributed to electrons causing direct ionization, resulting in intracellular function disturbance, inhibition of epidermal cell proliferation, and immunosuppression. Based on these results, we conclude that RT provides a beneficial prolonged remission to irradiated areas and is an effective and safe option for patients with severe, recalcitrant Darier’s and Hailey-hailey’s. We will continue to follow-up these patients to monitor the long-term efficacy and side effects of RT, as well as continue to improve the technique of RT for treatment of these patients.

Dysregulation of the innate immune system in affected and non-affected skin of patients with pyoderma gangrenosum

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Pyoderma gangrenosum (PG) is a neutrophilic, inflammatory skin condition that often presents as painful, chronic ulcers. Due to the lack of definitive diagnostic tests, PG is a clinical diagnosis of exclusion, which frequently leads to misdiagnoses. A delay in the proper diagnosis often increases a patient’s exposure to improper and risky management. A better molecular understanding of PG is essential to improve diagnostic accuracy and targeted therapy. Recent reports suggest that neutrophilic dermatoses may be within the spectrum of autoinflammatory disorders; as such, innate immunity may have a role in PG. We decided to evaluate and compare cytokine expression in affected and non-affected skin of patients with PG. Paired skin biopsies were collected from affected and non-affected (control) skin in five patients with PG. Gene expression from the matched skin samples was analyzed via Real-Time Quantitative Reverse Transcription PCR (qRTPCR) array of 84 genes (Innate and Adaptive Immune responses RT² Profiler PCR array; SABiosciences). The array analysis revealed different patterns of gene expression between affected skin and non-affected skin. The effect size (delta [δ]) between the Ct* values of affected and non-affected skin was calculated: (mean in non-affected skin) - (mean in affected skin) / (standard deviation in affected skin). A multidimensional scaling plot of matched affected and non-affected samples was used to assess similarity across the 49 inflammatory genes selected based on AUC>0.90, and demonstrated significant discrimination between affected and non-affected skin based on delta Ct values. Based on the Qiagen pathway annotation, we used a hypergeometric test to identify the pathways enriched (beyond chance) in the 49 genes, and found the strongest enrichment in the innate immunity pathways: Pattern Recognition Receptors (PRR) (p=0.05) and Other Innate Immunity Genes (p=0.05). In summary, we discovered, in this first study assessing affected versus non-affected skin in patients with PG, an alteration in innate immunity pathways. These findings suggest that focal upregulation of innate immunity...
might somehow affect the evolution of PG. However, these results need to be confirmed in a large cohort of patients and their clinical relevance further explored. *Ct = Threshold cycle (quantification of gene expression).

The quinacrine experience in a population of cutaneous lupus erythematosus and dermatomyositis patients

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Importance: Quinacrine, a compounded antimalarial, is crucial for those intolerant or unresponsive to hydroxychloroquine. At the March 2016 PCAC (Pharmacy Compounding Advisory Committee) meeting, members of the FDA raised concerns about the drug’s safety. As a result, quinacrine’s future availability is uncertain.

Objective: To determine the extent of quinacrine usage at the Hospital of the University of Pennsylvania (HUP) in the last year and in two databases of cutaneous lupus erythematosus (CLE) and dermatomyositis (DM).

Design: 1) We quantified the patients on quinacrine in the past year at the institution level and 2) retrospectively reviewed records in two prospectively collected longitudinal databases of CLE and DM.

Setting: Outpatient dermatology clinic at the University of Pennsylvania.

Participants: Of the 636 database patients (66.2% CLE, 33.8% DM), those without antimalarial treatment history (n = 73) or incomplete records (n = 25) were excluded. 538 patients were included in the final analysis (mean age = 52.3, 85.1% female, 14.9% male, 68.8% CLE, 31.2% DM).

Results: At the entire institution, 241 patients were prescribed quinacrine from June 1, 2015-May 31, 2016. HUP pharmacy records for 111 patients receiving quinacrine indicate 63.1% (n = 316) used quinacrine.

Conclusions and Relevance: 1) Quinacrine was prescribed predominantly for rheumatic skin disease at HUP. 2) Over half of CLE and DM database patients used quinacrine largely due to hydroxychloroquine refractoriness. 3) Quinacrine was discontinued more often than the other antimalarials due to access barriers as opposed to side effects, and 4), following side effects, quinacrine treatment was frequently resumed.

A New Mortality Prediction Tool for Stevens-Johnson syndrome / Toxic Epidermal Necrolysis: An update to SCORTEN derived from a large, multicenter cohort in the United States

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Background: Stevens-Johnson Syndrome/toxic epidermal necrolysis (SJS/TEN) is a spectrum of severe mucocutaneous drug reaction associated with significant morbidity and mortality. SCORTEN is a SJS/TEN-specific severity of illness score to predict in-hospital mortality, originally developed in a cohort of 165 patients from a single institution in France. The objective of this study is to determine the accuracy of SCORTEN in our multi-institutional cohort from the United States and to examine additional risk factors for death in this population, with the goal of creating an updated severity of illness score to predict in-hospital mortality from SJS/TEN.

Methods: Data were collected from 18 academic medical centers in the United States between January 1, 2000 and June 1, 2015. Individuals were included if they were 18 years of age or older and had a diagnosis of SJS/TEN confirmed by a dermatologist. Results: 370 patients had information available for analysis. 54 patients (15.14%) did not survive to hospital discharge. In our cohort, the SCORTEN model predicted a mortality rate of 74 (20.0%) (Standardized Mortality Ratio: 0.73). Examining factors included in the original SCORTEN model as well as additional potential prognostic factors, 6 covariates were found to be independent predictors of in-hospital mortality: age ≥ 50 years (OR: 2.99, 95% CI: 1.42-6.31), BSA ≥ 40% on admission (OR: 2.94, 95% CI: 1.41–6.09), malignancy (OR: 3.91, 95% CI: 1.72–8.88), dialysis (OR: 11.22, 95% CI: 2.72–46.28), serum BUN > 10 mmol/L (OR:3.46, 95% CI: 1.72–6.94) and serum bicarbonate < 20mmol/L (OR: 2.37, 95% CI: 1.19–4.73). This updated severity of illness score assigns a value of 1 for the presence of each predictor, with the exception of dialysis, which receives 2 points for a positive score. Calibration demonstrated excellent agreement between predicted mortality (15.0%) and actual mortality (15.1%) (Brier Score 0.1039). Discrimination was excellent with an ROC area under the curve of 0.8495. The scoring system (Table 1) and predicted mortality for each score (Table 2) are shown below.

Discussion: In conclusion, SCORTEN overestimated mortality in this large cohort of SJS/TEN patients from the United States. Changing the age cutoff to ≥ 50 years, increasing the BSA cutoff to ≥ 40%, and adding information regarding dialysis improved prognostication. Future use of a more accurate prediction model such as this one can provide improved prognostic information to patients and physicians and can help stratify patients more accurately in clinical trials.
Characterizing the burden of skin disease in Hispanic Americans with HIV
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Background: Skin disease is a significant source of morbidity in patients with HIV. Previous studies have shown increased rates of skin cancer as well as infectious and inflammatory dermatoses in HIV patients. However, there is a lack of data on HIV-associated skin diseases in Hispanic populations. Given that Hispanics now outnumber non-Hispanic whites in California according to the US Census, there is a pressing need to study skin disease in Hispanic HIV patients. Objective: We aim to characterize the spectrum and burden of skin disease in a primarily Hispanic population of HIV patients, with particular attention to the demographics of our patients, their categories of skin diseases, and associated CD4 counts and viral loads. Methods: We conducted a retrospective chart review of patients seen at the Los Angeles County-University of Southern California HIV Dermatology clinic over an 18-month period from June 2015-November 2016. Results: A total of 185 patients were included in the study, of whom 133 (71.9%) were Hispanic. 78.2% of the Hispanic patients were male and the mean age was 48.5±10.3. The most common diagnoses among Hispanics were inflammatory dermatoses (25.6%), including diseases such as seborrheic dermatitis and psoriasis. Other common diagnostic categories included benign skin lesions (24.8%), skin infections (19.5%), lipodystrophy (6.9%), pigmentation disorders (6.1%), and skin cancer (4.5%). The most common individual diagnosis overall was warts (10.1%). Notably, the mean CD4 count at the index visit for patients with infections (384.3) was significantly lower than the overall mean CD4 count for all the patients in our sample (499.7) (p-value=0.022). The distribution of skin conditions was similar to that of non-Hispanic patients from the same clinic. Limitations: This study included patients at only one clinic, therefore the results may not be generalizable to the broader population. Conclusions: Inflammatory skin conditions were the greatest source of morbidity in this study, though warts were the overall most common diagnosis. Patients presenting with skin infections had significantly lower CD4 counts than the overall patient population. Hispanics with HIV have diverse cutaneous manifestations and are a population meriting further study to more fully characterize their burden of skin disease.

Rosacea has greater impact in women and younger patients: DLQI and RosAQOL in a broad rosacea population
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Background: Rosacea reduces patients’ quality of life, but the extent of the impact has not been assessed in large patient samples. Objective: To assess the impact of rosacea on patient’s lives and to test whether demographic variables are related to the impact. Methods: 194 patients with a clinical diagnosis of rosacea were recruited from the Wake Forest Dermatology Clinic over three (3) years. Patients completed a survey that included a self-assessed rosacea severity tool, demographic questions, Dermatology Life Quality Index (DLQI) and Rosacea Quality of Life Index (ROSAQOL). DLQI and ROSAQOL scores among demographic groups (gender, age, marital status, education level, income level) were assessed. Results: 183 patients completed the DLQI and 194 completed the ROSAQOL questionnaires. The median DLQI score was 1.0, indicating that for at least half of the patients, the DLQI could not detect significant effect on quality of life. The DLQI score was worse with younger age (p=0.003) and lower income (p=0.03). The median ROSAQOL total score was 2.8 out of a possible 5. The total ROSAQOL score was worse in female gender (p=0.002) and younger age (p=0.0002). The symptom (p=0.003) and function (p=0.0001) constructs of the ROSAQOL score were both worse in females, but the functional impairment scores demonstrated the greatest disparity between males and females. Limitations: All subjects were patients at a single center; this may have an effect on the generalizability of the results and conclusions. Discussion: Younger rosacea patients had diminished quality of life compared to their older counterparts. Patients in the lower-middle class income bracket reported their quality of life was impacted more severely by their disease. Female patients had higher propensity towards worsened quality of life in spite of recent research indicating that men tend to have more severe rosacea symptoms. Our data suggests that certain demographic groups may be more susceptible to quality of life impact due to rosacea. Providers should keep this information in mind as they develop treatment regimens. More aggressive treatment regimens may be indicated for individuals at high risk for quality of life impact. Conclusion: Treatment planning should account for the impact of the disease. Younger, lower income, female patients appear to be at risk for greater quality of life impact.

Systemic treatment for clinically amyopathic dermatomyositis: A retrospective cohort study at four 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 as most studies have focused on pulmonary disease outcomes. Our study investigated the use of systemic therapy for skin disease in CADM at four 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 2000 and 2016. Inclusion criteria included diagnosis and management of CADM by a board-certified dermatologist or rheumatologist. Data collected included demographics, referral types, laboratories, medications used and adverse effects. 117 patients with CADM (96 amyopathic and 21 hypomyopathic) were identified. The mean age of diagnosis was 49.8 years. Most patients were referred from dermatology (46%) and rheumatology (30.1%) providers. Antimalarial agents were the most commonly used treatment type (77%), but achieved good control of skin disease in only 11% of cases. Consistent with the existing literature, 26.7% of patients developed a cutaneous hypersensitivity reaction to hydroxychloroquine. Of the entire cohort, only 19.7% of patients were treated with antimalarials and topical therapy alone, while 80.3% required at least one immunosuppressive therapy to control their cutaneous disease. Furthermore, patients tried a mean of 3.6 (SD = ±1.9) treatments prior to achieving skin disease control. Among these additional therapies, methotrexate (53.8%) and mycophenolate mofetil (39.3%) were the most commonly used, followed closely by IVIG (29.1%). Notably, 13 patients were followed exclusively by a rheumatology provider, and none of these patients received IVIG (32.7% vs 0%, p=0.02). As existing data supports the concept that IVIG is likely the most effective treatment of skin disease in DM, this difference in treatment approach may impact patient outcomes. In this cohort of CADM patients, which is the largest reported to date, cutaneous disease was refractory to antimalarial agents in the vast majority. 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.
A pilot study characterizing factors in adherence to cutaneous lupus treatment

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Background: Cutaneous lupus erythematous (CLE) is an autoimmune skin disease that manifests as scarring, dyspigmentation, erythema, itching, scaling, disfigurement, and discomfort. Topical corticosteroids are one of the mainstays of treatment. However, local irritation, messiness, inconvenience, and tediousness may deter use. Non-adherence, rather than non-response may be a cause of treatment failure. Aim: To obtain preliminary data on adherence rates to topical corticosteroid therapy for CLE and identify factors that affect adherence, including disease severity, physician trust, and depression. Methods: A 6-month investigator-blinded, prospective study was performed on subjects with active CLE. Subjects were instructed to apply fluocinonide, 0.05% ointment twice a day to active lesions of CLE. Adherence was assessed using Medication Event Monitoring System (MEMS®) caps. Measures of disease severity, physician trust, and depression were obtained and analyzed. Results: Ten participants completed the study. Mean adherence was 56.2% (SD=38.1%) for percent of prescribed doses taken. Mean adherence by month declined from 88.2% (SD= 68.0%) at 1 month to 44.0% (SD=34.0%) at 6 months. Better adherence was associated with worse depression scores (P=0.03), lower physician trust (-0.647, P=0.06), and higher patient-reported disease severity (0.303, P=0.08). Limitations: The small sample size may affect statistical significance and generalizability of results and conclusions. Discussion: Adherence to fluocinonide ointment in patients with CLE is poor and declines over time. Compared to studies that report greater physician trust associated with higher adherence and more severe depression scores associated with poorer adherence, we observed an opposite trend in this pilot study. Measures to improve adherence to existing CLE therapies should be examined to improve patient outcomes.

Chronic Granulomatous Mastitis: A Retrospective Cohort Study

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Background: Chronic Granulomatous Mastitis (CGM) is a benign inflammatory breast disease that can mimic breast cancer. It affects women of childbearing age with recent history of pregnancy and lactation. The main etiologies are postulated to be autoimmune, infectious, and hormonal processes. Objective: To identify in our cohort, common presenting signs and symptoms, ideal first line therapy, as well as potential associations and predisposing factors. Methods: A retrospective study of histologically proven chronic granulomatous mastitis patients at Bellevue dermatology clinic between 2013 and 2015. The observed parameters included time to diagnosis, common presenting symptoms, and first and second line treatment. Results: The study population included 19 female patients with a mean follow up time of 4 months (3-21). The mean age at presentation was 35 (29-41). The cohort race: 84% Hispanic, 11% Bangladeshi, 5% Caucasian. Breastfeeding history among the patient cohort was 85%. Pregnancy within the last 5 years was 55%. The infectious disease work up was negative for gram stain, AFB cultures, bacterial cultures, and fungal cultures. Histopathology showed non nécrotizing granulomas, acute on chronic inflammation, fat necrosis. Mean time to diagnosis was 2 months if patient initially presented to Bellevue (n=14) vs 12 months if patient initially presented to outside provider (n=5). Initial treatment was: doxycycline (68%), combined doxycycline and prednisone (11%), prednisone alone (5%). No treatment was needed in 16% of cases. Among patients treated with doxycycline as first line therapy, 64% (n=9) showed initial improvement and 70% (n=7) required second line therapy. Second line therapy: prednisone 62% (n=5) and combined prednisone and methotrexate 38% (n=3). Conclusion: Initial patient presentation to Bellevue resulted in significant shorter duration to diagnosis compared to outside presentations. Doxycycline treatment resulted in initial improvement in high percentage of cases. Study limitations included short duration and small sample size. We are prospectively following this cohort and will examine other treatment options and algorithms.

Silver nitrate sticks for draining sinus tracts in patients with hidradenitis suppurativa (HS): A case series

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Objective: To highlight a previously unreported technique for management of draining sinus tracts of hidradenitis suppurativa. Background: Treatment options for HS are limited and under-studied. Medical treatments including antibiotics, anti-androgens and TNFα-inhibitors help control disease, however some draining sinus tracts can be recalcitrant to medical therapy. Management options in the literature include cryosuflillation, de-roofing and excision. We present a case series of patients with moderate to severe HS with recalcitrant painful and draining sinus tracts treated with insertion of silver nitrate sticks. Methods: Eight patients with moderate to severe HS with a total of 16 draining sinus tract sites were treated with insertion of silver nitrate sticks. Fourteen of the 16 sites were also treated with concomitant intraleisional triamcinolone. A majority (12/16) of the sites were anesthetized prior to the procedure with lidocaine 1% with epinephrine 1:100,000. For most of the patients, we instilled the triamcinolone mixed 1:1 with the local anesthetic to limit the number of injections. Patients were asked about severity of their pain and drainage at the target sites before the procedure and at follow-up visits. Results: Thirty-eight percent (6/16) demonstrated mild to moderate improvement, while 31% (5/16) had marked improvement, with three sites having complete resolution of pain and drainage. One of the patients with complete resolution of pain and drainage had a recurrent flare in the same site several months later. Notably, two sinus tracts developed ulceration at the site, though one did have improvement of pain despite ulcer formation. A total of three sites had no change in symptoms. Of three patients treated without local anesthetic, two had significant pain. Conclusion: Draining sinus tracts recalcitrant to medical management are a common problem in the management of moderate to severe HS. In our practice, silver nitrate can improve pain and drainage from these actively draining inflamed sinus tracts in most patients, however improvement seems to be mostly temporary. We do think it is a worthwhile approach for recalcitrant, symptomatic sinus tracts as an attempt to avoid surgical procedures. We hypothesize that it works via antimicrobial properties, cauterization of granulation tissue, and possibly breakdown biofilms. More studies are needed to investigate the use of silver nitrate for the treatment of active draining sinus tracts in HS.
Sub-minimal blistering dose: a novel plaque-based dosimetry strategy in excimer laser phototherapy

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Introduction: Phototherapy with ultraviolet (UV)-B spectrum light is a safe, effective therapy for psoriasis. Excimer laser phototherapy is a convenient modality, clearing plaques in fewer treatments than traditional booth phototherapy. As higher doses lead to faster clearance, the administered UVB dose is critical for treating patients. Excimer targets only psoriatic lesions and thus allows administration of high dose UVB leading to faster clearance than standard booth phototherapy where dosing is limited by the sensitivity of normal skin. Current standard treatment protocols typically utilize 15-20 treatments for clearance, which can be inconvenient for patients. This study evaluates a novel plaque-based dosing strategy designed to improve therapeutic response time and make excimer treatment more practical for patients. Methods: Subjects with psoriasis on bilateral symmetrical surfaces were enrolled. One plaque was randomized to standard excimer dosing and the contralateral to the more aggressive experimental dosing (SBD). Plaques were tested with a special tool that simultaneously delivered multiple doses to determine the minimal blistering dose. Starting EXP dose was 20% less than the minimal blistering dose with subsequent adherence to standard dose adjustments. Standard sides were treated according to standard protocol guided by Fitzpatrick skin type and plaque induration. The primary outcome was modified Psoriasis Area Severity Index (mPASI) determined by erythema, induration, scaling, and area of target lesion remaining. Treatments were carried out up to twice weekly for 10 treatments with blinded assessments at each treatment. Results: On average, initial treatments were 2.8 times higher on the SBD side. All 6 paired plaques achieved at least mPASI-75 by the end of 10 treatments. By treatment 4, three SBD plaques had achieved mPASI-90, while two standard treatment plaques achieved mPASI-90. Three SBD plaques were treated once weekly while their pairs were treated twice weekly with nearly identical mPASI improvements across paired plaques. Conclusion: A novel excimer dosing regimen starting with a sub-blistering dose 2.8 times higher than standard initial dosing resulted in quicker response and clearance of psoriasis in approximately 4 treatment sessions. Data also suggest that once weekly dosing using the experimental protocol may be equivalent to twice weekly dosing with the standard protocol, providing a more convenient treatment regimen for patients.

Atypical presentations of hidradenitis suppurativa: A case series

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Background: Hidradenitis suppurativa (HS) is a chronic inflammatory disorder that classically affects apocrine gland-bearing areas of the body, especially the axillae and groin. The prevalence of disease is estimated to be up to 4% of the population, and affects females more than males at a 3:1 ratio, typically in the second and third decades of life. HS is thought to arise from follicular occlusion, mechanical stress, and associated immune dysregulation. It classically affects the intriginous and apocrine gland-bearing areas of the skin, including the axillae, inframammary region, inguinal folds, genitals, and buttocks. However, involvement of atypical regions of the body, including the posterior neck, cheek, beard, and back, has previously been reported. Objective: We present five patients with non-infectious, deep-seated painful nodules and abscesses, which are classic features of severe HS, on the extremities, which are an atypical location for HS. We also refer to the recent literature on atypical presentations of HS. Methods: The medical records of five patients at Brigham and Women’s Hospital were retrospectively reviewed to highlight the clinical presentations and medical management of these patients. We also conducted a literature search to describe the reported literature of atypical presentations of hidradenitis suppurativa. Results: Five patients presented with classic cutaneous findings of hidradenitis suppurativa in the axillae and groin, but also had atypical lesions on the upper and lower extremities. These lesions were confirmed by clinical examination and/or histopathologic correlation as atypical hidradenitis suppurativa. All patients underwent trials of systemic treatment with TNF-inhibitors and additional treatment as indicated. Conclusion: This case series highlights the clinical features and considerations for the diagnosis and management of atypical presentations of hidradenitis suppurativa on the extremities. All of our patients had severe HS; therefore, the atypical lesions may reflect the natural course of HS progression, initially affecting the axillae and groin, and then occasionally spreading to other areas. It is important to recognize that hidradenitis suppurativa can present on the extremities, in order to avoid a delay in diagnosis and treatment.

Utility of tissue microbiology studies in suspected skin infections

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Skin tissue microbiology studies are often performed concurrently with tissue histology when infection is on the differential diagnosis (DDx). However, obtaining microbiology studies requires performing a second biopsy, which may be associated with additional patient discomfort, procedures, time, and cost. Little is known about the frequency in which microbiology studies return positive or influence treatment decisions. We performed a retrospective analysis of patients for whom both pathologic and microbiology studies were sent on skin biopsies performed during in-patient and out-patient dermatology visits at Massachusetts General Hospital and Brigham & Women’s Hospital from 2000-2015. Preliminarily, 100 cases have been identified. Infection was among the leading differential diagnoses in 72% (n=72) of the cases. Bacterial culture 95% (n=95) and gram stain 81% (n=81) were the most common microbiology studies sent. Bacterial, fungal, and AFB cultures were positive in 47.4% (n=45), 6.4% (n=5), and 6.0% (n=4) of cases, respectively; gram, fungal, and AFB stain returned with positive results in 12.3% (n=10), 0% (n=0), and 3.0% (n=2) of cases, respectively. Microbiology studies were 65% (n=65) concordant with pathology results. We defined true positive and true negative test results based on the documented clinical interpretation. Microbiology studies had a sensitivity of 75.6%, specificity of 77.8%, positive predictive value of 58.5%, and negative predictive value of 88.5%. Microbiology studies altered diagnosis in 45.0% (n=45) of cases (showing absence of infection when infection was on the DDx, n=24; demonstrating infection when infection was not on the DDx, n=3) while concordant with the original DDx in the remainder 55.0% (n=55) of cases (confirming infectious DDx, n=31; confirming non-infectious DDx, n=14; showing no infection but were overruled by high clinical suspicion of infectious DDx: n =10). Based on microbiology results, new antimicrobial therapy was started in 26.0% (n=26) of cases, antimicrobial therapy was stopped in 2.0% (n=2) of cases, and triage/disposition recommendations were changed in 1.0% (n=1) of cases. We preliminarily conclude that microbiology studies have important influence on diagnosis and clinical management, with relatively high sensitivity, specificity, but lower positive predictive value. These findings suggest that the utility of microbiology results may justify the additional time, cost, and risks that may be associated.