Chicago Dermatological Society

October 2016 Educational Conference

Program & Speaker Information
CME Certification
Case Presentations

David Fretzin Lecture

Wednesday, October 26, 2016

University of Illinois at Chicago - Student Center West
Chicago, IL

Conference Host
Department of Dermatology
University of Illinois at Chicago
Chicago, Illinois

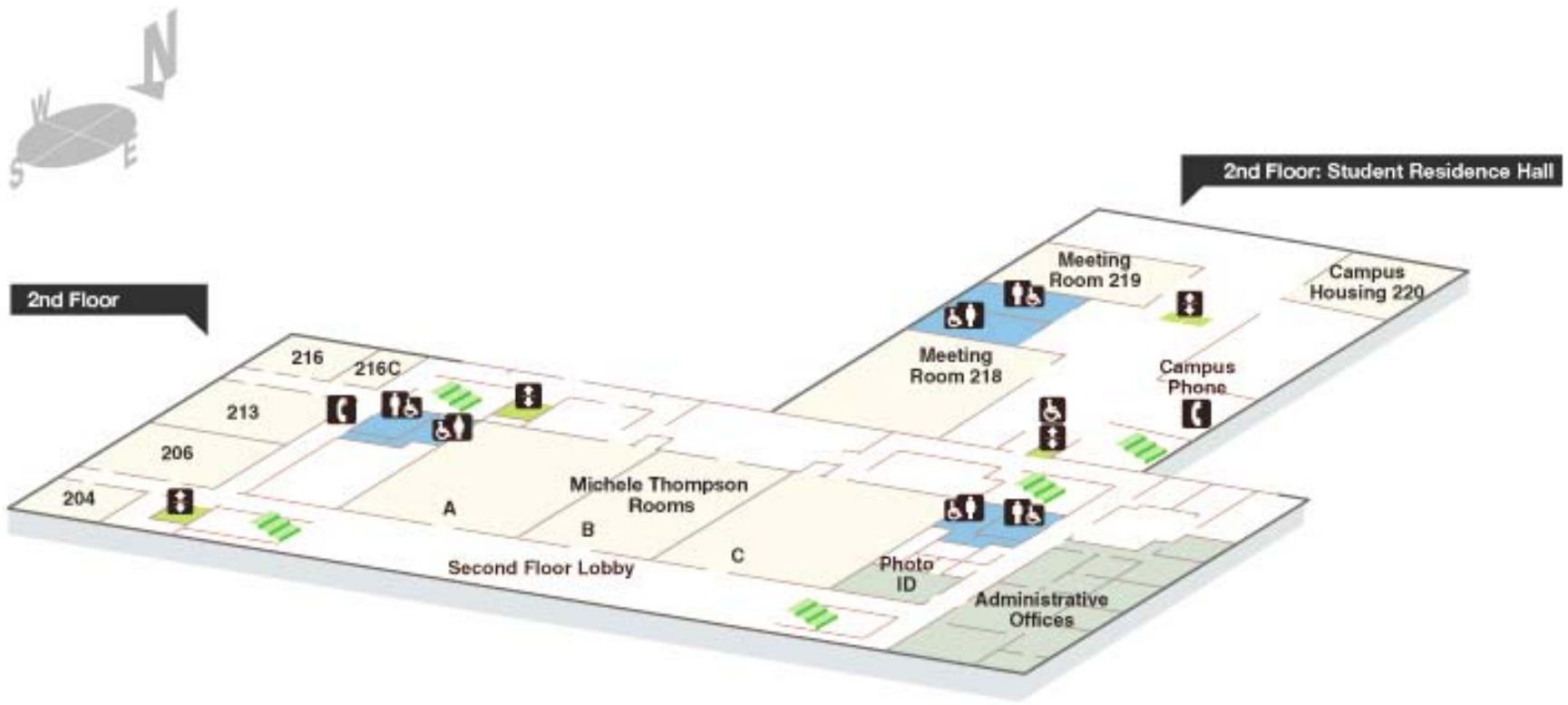
Program

Host: University of Illinois at Chicago
Wednesday, October 26, 2016
UIC Student Center West - Chicago, IL

8:00 a.m.	**Registration & Continental Breakfast with Exhibitors** *2nd Floor Foyer*
8:30 a.m. - 10:30 a.m.	**Clinical Rounds** Slide viewing – *Room 204* Patient viewing – *Rooms 216A, 216B, 218 SRH, 219 SRH* Posters – *2nd Floor Foyer*
9:00 a.m. - 10:00 a.m.	**Basic Science Lecture** – *Thompson Room ABC* "Hemangiomas and Vascular Malformations for the Dermatologist" *Julie Powell, MD*
10:00 a.m. - 10:30 a.m.	**Break and Visit with Exhibitors** – *2nd Floor Foyer*
10:30 a.m. - 12:00 p.m.	**Resident Case Presentations & Discussion –** *Thompson Room ABC*
12:00 p.m. - 12:15 p.m.	**MOC Self-Assessment Questions** – *Thompson Room ABC*
12:15 p.m. - 12:45 p.m.	**Box Lunches & visit with exhibitors** – *2nd Floor Foyer*
12:55 p.m. - 1:00 p.m.	**CDS Business Meeting** – *Thompson Room ABC*
1:00 p.m. - 2:15 p.m.	**General Session** – *Thompson Room ABC*
1:00 p.m.	"Report from the Illinois State Medical Society" *Thomas M. Anderson, MD; President, ISMS*
1:15 p.m.	"Lessons Learned from a Multidsicplinary Vascular Anomalies Clinic" *Julie Powell, MD*
2:15 p.m.	**Meeting adjourns**

Mark the Date in 2 weeks!
Next CDS monthly meeting – Hosted by Northwestern University
Wednesday, November 9, 2016; Gleacher Center

Watch for details on the CDS website: www.ChicagoDerm.org
Save time and money – consider registering online!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Guest Speaker

JULIE POWELL, MD

Director of the Pediatric Dermatology Division
CHU Sainte-Justine
Montréal, Québec

Dr. Julie Powell is a pediatrician-dermatologist, and she is director of the Department of Dermatology at Sainte-Justine University Hospital (CHU) in Montréal. She also is a full professor, clinical, at the University of Montréal.

Dr. Powell received her medical degree from the University of Sherbrooke in Québec, followed by a residency in pediatrics at the same institution. She undertook her residency in dermatology at the University of Montréal and the University of Iowa Hospitals and Clinics.

Dr. Powell is co-founder of the Vascular Anomalies Team at CHU. She is a member of the scientific committee of the International Society for the Study of Vascular Anomalies (ISSVA). She also is a former Vice-President of the Canadian Dermatology Association (CDA) and currently is the President-Elect of the CDA.

CME Information

This educational activity is jointly provided by AXIS Medical Education, Refine Me CE and the Chicago Dermatological Society

Overview

The Chicago Dermatological Society was established in 1903 and has strived to provide meaningful educational opportunities to dermatologists in the Chicago area for more than a century. Guest speakers from across the country share their expertise with CDS members, as well as residents in training medical students doing their dermatology rotation. CDS schedules six day-long meetings each year which are "hosted" by one of the dermatology residency programs in the city. In addition to two lectures given by the guest speaker, the residents of the host institution present cases which are offered for audience discussion. In addition, live patients, posters and microscopic slides prepared by the residents are made available during the "clinical rounds" portion of the meeting. CDS also offers a 15-minute session that qualifies for "Maintenance of Certification" self-assessment questions under the auspices of the American Board of Dermatology.

Target Audience

This activity has been designed to meet the educational needs of dermatologists. CDS members, residents in training and medical students engaged in their dermatology rotation are invited to attend.

Learning Objectives

At the conclusion of the 2016/17 series of meetings, the participant should be able to:

1. Discuss key factors in the diagnosis and treatment for viral diseases and bacterial infections.
2. Describe the manifestation of skin cancers and the efficacy of treatments available to the dermatologist.
3. List the therapeutic options available to the dermatologist for a variety of skin diseases, both medical and surgical, and discuss how new emerging treatments can be successfully incorporated into a dermatology practice.

Physician Accreditation Statement

This activity is planned and implemented by AXIS Medical Education, Refine Me CE and the Chicago Dermatological Society. AXIS Medical Education is accredited by the Accreditation Council for Continuing Medical Education (ACCME), the Accreditation Council for Pharmacy Education (ACPE), and the American Nurses Credentialing Center (ANCC) to provide continuing education for the healthcare team.

Credit Designation for Physicians – AXIS Medical Education designates this live activity for a maximum of 4.75 *AMA PRA Category 1 Credit(s)*™. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

Attendees are required to submit a CME claim form upon departure from the conference. Please leave your form, along with the attached evaluation form, at the registration table when you leave the meeting. Thank you for your attention to this important item.

Disclosure of Conflicts of Interest

AXIS Medical Education requires instructors, planners, managers and other individuals and their spouse/life partner who are in a position to control the content of this activity to disclose any real or apparent conflict of interest they may have as related to the content of this activity. All identified conflicts of interest are thoroughly vetted by AXIS for fair balance, scientific objectivity of studies mentioned in the materials or used as the basis for content, and appropriateness of patient care recommendations. All speakers are expected to follow the "first slide" rule to repeat their conflict of interest disclosures during their talk.

Today's guest speaker, Julie Powell, MD, has disclosed the following relationships: Speaker or member of a speakers bureau - Pierre Babre, Galderma, Johnson & Johnson; Contracted research - Pierre Fabre. The following have no conflicts of interest to disclose: Residents presenting cases at this meeting; planning committee members - Alix Charles, MD, program chair and CDS president; Julie Moore, MD, CDS past-president; Richard Paul, CDS Executive Director; Ronald Viggiani, MD and Dee Morgillo, MEd, MT(ASCP), CHCP, AXIS Medical Education.

AXIS Contact Information

For information about the physician accreditation of this program please contact AXIS at 954-281-7524 or info@axismeded.org.

Americans with Disabilities Act

In compliance with the Americans with Disabilities Act, we will make every reasonable effort to accommodate your request. For any special requests, please contact the CDS at: Rich@ChicagoDerm.org.

Disclaimer

Participants have an implied responsibility to use the newly acquired information to enhance patient outcomes and their own professional development. The information presented in this activity is not meant to serve as a guideline for patient management. Any procedures, medications, or other courses of diagnosis or treatment discussed in this activity should not be used by clinicians without evaluation of patient conditions and possible contraindications on dangers in use, review of any applicable manufacturer's product information, and comparison with recommendations of other authorities.

Dislosure of Unlabeled Use

This educational activity may contain discussion of published and/or investigational uses of agents that are not indicated by the FDA. The planners of this activity do not recommend the use of any agent outside of the labeled indications. The opinions expressed in the educational activity are those of the faculty and do not necessarily represent the views of the planners. Please refer to the official prescribing information for each product for discussion of approved indications, contraindications, and warnings.

University of Illinois at Chicago
Department of Dermatology

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Second Year

Lisa Blackwood, MD, MS
Benjamin Garden, MD
Michael Sotiriou, MD
Huayi Zhang, MD, MS

First Year

Lorelei E. DiTommaso, MD, MPH
Mark Juhl, MD
Artem Sergeyenko, MD
Stephanie Wang, MD

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Case 1

Case Presented by Leigh Stone, MD, Ramya Tripuraneni, MD and Michelle Bain, MD

History of Present Illness:
A two year old male with multiple developmental disabilities was referred to dermatology clinic for diffuse congenital skin findings. His mother reported that he was born after an uncomplicated pregnancy and that she noted discoloration of most of his skin on his first day of life. She denied any change in appearance or progression of the lesions since his birth.

Past Medical History:
Global developmental delay, epilepsy, hypotonia, and recurrent aspiration pneumonia

Medications:
Levetiracetam

Allergies:
No known drug allergies

Family History:
No one in the patient's family had a history of similar skin findings, including two siblings. His maternal aunt had epilepsy and developmental delay. His maternal uncle died at age 38 due to stroke; he also had a history of epilepsy and developmental delay.

Social History:
The child lived at home with his parents and siblings. The family was noted to have poor compliance with medical appointments.

Review of Systems:
The mother reported speech delay and difficulty walking.

Physical Examination:
The patient has Blaschkoid hypo- and hyperpigmented linear patches on his trunk as well as the upper and lower extremities, a high forehead, wide-set eyes, broad nasal root, low-set ears, invasion of philtral skin onto the vermillion of the upper lip, and decreased scalp hair density.

Diagnostic Procedures and Tests:
02/14 Microarray: normal male microarray. Microarray analysis using a whole genome oligonucleotide array detected no clinically significant abnormalities.

08/15 Chromosome Analysis of Skin: abnormal mosaic male karyotype. 55% of the cells examined contained an isochromosome 12p.

Diagnosis:
Pallister-Killian syndrome

Treatment Course:
The patient is responding well to physical, speech, and behavioral therapy as well as follow up with multiple medical specialties including genetics, neurology, and ophthalmology. However, compliance has been a continued issue.

Discussion:
Pallister-Killian syndrome (PKS) is a rare, sporadic, multisystem disorder caused by tissue-limited mosaic tetrasomy of 12p. In PKS, tetrasomy is produced by the presence of an isochromosome, which is comprised of two extra copies of 12p arranged in mirror image. The isochromosome is created by a non-disjunction event, thought to occur most often during maternal meiosis II.

Nearly half of PKS patients exhibit cutaneous findings, specifically linear patches of hyperpigmentation or hypopigmentation and can appear as a whorled pattern following the lines of Blaschko. PKS represents one of the many chromosome mosaicisms that can present with Blaschkoid dyschromia, historically referred to by the descriptive rather than diagnostic term Hypomelanosis of Ito.

Distinct craniofacial features are associated with PKS and include fronto-parietal alopecia, sparse eyebrows, philtral skin projecting onto the upper lip vermillion, depressed nasal bridge, large mandible, bifid uvula, and a short neck. Systemic features that can be associated with PKS include decreased vision, structural brain malformations, epilepsy, structural cardiac defects, lung hypoplasia secondary to a diaphragmatic hernia, intestinal malrotation, displacement of the anus, and a growth pattern unique to PKS which consists of an accelerated prenatal growth period followed by a decelerated postnatal growth period. Like many mosaic conditions, PKS has a vast spectrum of disease severity ranging from intrauterine death to very mild forms. The overall neurologic prognosis is poor with significant mental and motor retardation being common.

For diagnosis, the genetic changes of PKS may be detected by karyotype of cultured skin fibroblasts as was done in this case. Alternatively, fluorescent in situ hybridization (FISH) can be employed with chromosome 12 specific DNA probes in order to identify isochromosome 12p. It is typically not detected in rapidly dividing cells such as those found in the peripheral blood. However, there are limited reports of cases being identified in peripheral lymphocytes. The diagnosis is highest among amniocytes and bone marrow cells with a detection rate of 100%, followed by a detection rate of 50-100% in fibroblasts and 0-2% in lymphocytes. Prenatal detection by chorionic villous sampling, amniocentesis, and cordocentesis is also possible.

At this time, treatment of PKS is supportive and requires a multidisciplinary approach.

Essential Lesson:
- Pallister-Killian syndrome is caused by mosaic tetrasomy of 12p resulting from an isochromosome.
- Almost half of Pallister-Killian patients have cutaneous findings, which fit under the descriptive rather than diagnostic term Hypomelanosis of Ito.
- Additionally, Pallister–Killian syndrome is characterized by facial dysmorphism, heart defects, congenital diaphragmatic hernia, hypotonia, intellectual disability, and epilepsy.
- In all patients presenting with Blaschkoid pigmentary changes, consider obtaining a skin biopsy with subsequent karyotyping of cultured fibroblasts.

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Case Presented by Iona Chapman, MD
and Milena J. Lyon, MD

UNKNOWN

This 49 year old female presented with multiple stellate necrotic plaques and overlying hemorrhagic bullae.

Case 3

Case Presented by Lorelei E. DiTommaso, MD
Benjamin Garden, MD, and Iris K. Aronson, MD

History of Present Illness:
A 46 year old cognitively impaired female was referred by rheumatology due to concern for vitiligo. The patient's mother stated that over the past year the skin on the patient's chest started to lighten, and in recent months had progressed to include the arms, face, back, and legs. There had been no improvement with hydrocortisone cream.

Past Medical History:
Cognitive impairment, recent pneumonia, inflammatory arthritis (undergoing work-up with orthopedics and rheumatology)

Medications:
Montelukast, omeprazole, and cetirizine

Allergies:
No known drug allergies

Family History:
Niece: Systemic lupus erythematosus

Review of Systems:
The patient's mother reported weakness, chronic cough in recent months, fatigue, dysphagia, and joint pain. She denied fevers, chills, shortness of breath, constipation, or diarrhea.

Physical Examination:
The patient has diffuse depigmented patches with uniform peri-follicular pigmentary retention on the scalp, face, upper chest, upper back, and upper and lower extremities. There is periungual loss of pigment as well as mild sclerodactyly of both hands. Diffuse skin tightening is observed on the face, torso, upper and lower extremities proximal to the elbows and knees. On the volar tip of the right fourth digit, there is a three millimeter atrophic macule, consistent with a scar.

Laboratory Data:
The following were positive or abnormal:
Antinuclear antibody: dual homogenous and anti-centromere patterns, both >1:10,240 (≥1:160 clinically significant titer) on indirect fluorescence assay
Anti-topoisomerase I (Scl-70) antibody: 351 AU/mL (≥41 positive)
Erythrocyte sedimentation rate: 58 mm/hr (1 – 10)
Hemoglobin 10.1 g/dl (13.2 – 18)
Albumin 2.9 g/dl (3.4-5)
Urine analysis: protein 30 g/dL
Brain natriuretic peptide 558 pg/mL (>100 high)

The following were negative or within normal limits:
Complete metabolic panel, ferritin, Vitamin B12, angiotensin-converting enzyme level, complement 3 level, complement 4 level, anti-neutrophil cytoplasmic antibody, C-reactive protein, creatinine kinase, aldolase, quantiferon gold, as well as antibodies to dsDNA, Smith, cyclic citrullinated peptide, and ribonucleoprotein.

Diagnostic Procedures and Tests:

06/15 Computed Tomography, Chest: Mild interstitial lung disease, axillary and mediastinal adenopathy, cardiomegaly, and pulmonary arterial hypertension.

08/15 Transthoracic Echocardiogram: Severely enlarged right ventricle, reduced right ventricular function, dilated right atrium, tricuspid regurgitation, and elevated pulmonary artery systolic pressure.

Diagnosis:

Diffuse cutaneous systemic sclerosis

Treatment Course:

The patient was started on pentoxyphylline, prednisone, ambrisentan, and mycophenolate mofetil. However, there was disease progression involving the viscera, with worsening of pulmonary fibrosis, pulmonary hypertension, and subsequent cor pulmonale. Two months later, the patient died from septic shock secondary to a gastrointestinal infection.

Discussion:

Systemic sclerosis (SSc) is a progressive and debilitating disease that includes a wide spectrum of diverse cutaneous findings, typified by skin thickening, as well as varying degrees of multisystem involvement. It is important for the physician to be aware of this disease spectrum as the diagnosis of SSc rests largely on clinical findings. In an effort to increase diagnostic sensitivity, particularly for patients with early and limited disease, the American College of Rheumatology and European League Against Rheumatism updated the 1980 classification criteria in 2013. Classification criteria may be met with 91% sensitivity and 92% specificity. Skin manifestations included in the criteria are skin thickening, telangiectasias, Raynaud's phenomenon, abnormal nailfold capillaries, as well as digital edema, sclerodactyly, ulcers, and pitting scars.

Though not included in the aforementioned criteria, cutaneous pigmentary changes have long been recognized as an associated feature of SSc. In 1898, Sir William Osler was the first to publish the observation of dyspigmentation, which he noted in three out of the eight patients with SSc for whom he was investigating treatment with thyroid extract. Thereafter, a large case series from the Mayo Clinic of 727 patients with SSc, from 1935 – 1958, reported 222 patients (30.5%) with pigmentary changes. Interestingly, pigmentary changes were found to occur later in the disease, with only eight patients presenting with hyperpigmentation as the initial manifestation of SSc. Decades later, in 1983, a case series reported the first histologic features from skin biopsies of depigmented skin in seven patients with SSc. Haematoxylin and eosin (H&E) stain revealed minimal or absent melanin-laden melanosomes within the papillary dermis, rare melanocytes, and an abundance of Langerhans cells in the lower third of the epidermis. Observing pigmentary retention overlying blood vessels within larger field of depigmentation in three patients with SSc, Jawitz et al. in 1984 used thermography to test, inconclusively, the hypothesis that temperature variations may be causative in this phenomenon. Overall, the pigmentary alterations reported in the literature include the following: vitiligo-like depigmentation with perifollicular pigment retention (leukoderma of scleroderma, "salt and pepper" sign) most commonly on the central face and upper trunk, diffuse hyperpigmentation, localized hyper- or hypopigmentation especially in areas of pressure, and retention of pigment overlying superficial veins within larger patches of depigmentation.

For the dermatologist, knowledge and attention to pigmentary changes, as well as the cutaneous findings beyond fibrosis, may aid in diagnosis of SSc. Some patients may experience acute swelling of the distal upper and lower extremities prior to the onset of fibrosis. Nailfold capillary anomalies, such as enlarged loops, avascular areas, and neoformation are common, and were found in >90% in a case-control study of 75 patients with SSc compared to twenty healthy subjects. Other commonly described cutaneous findings include telangiectasias, most often on lips and palms, which are often described as matted or squared off. Calcinosis cutis most commonly near the joints and the distal extremities, diminished hair growth in areas of fibrosis, Raynaud's phenomenon, and ulcerations of the tips of digits are additional well-described cutaneous manifestations. As opposed to limited cutaneous systemic sclerosis, diffuse cutaneous SSc follows a more rapidly progressive course, owing to early internal organ involvement. Overall, pulmonary disease is the leading cause of death. Autoantibodies associated with a less favorable prognosis include anti- topoisomerase I, anti-U3RNP, and anti-T_h/T_o. Management of SSc is challenging, and therapies may be of limited efficacy due to the debilitating and often progressive nature of the disease. Evidence-based treatment options for cutaneous fibrosis in SSc include methotrexate, cyclophosphamide, mycophenolate mofetil, and even autologous hematopoietic stem cell replacement. Current investigations into cytokine-based therapies, particularly TGF-β, owing to its role in inducing endothelial damage and fibroblast activation, could in theory provide disease modification, especially in patients with early disease.

Essential Lesson:

- Systemic sclerosis is a debilitating and often progressive disease, typified by symmetrical fibrosis and variable organ involvement with a spectrum of cutaneous features.
- Pigmentary changes are a poorly understood but well-established feature of the disease, though not included in the current disease classification criteria.
- Pigmentary features include "salt and pepper" sign, hyperpigmentation particularly in areas of pressure, and pigmentary retention overlying superficial veins within larger patches of depigmentation
- Dermoscopy to assess nailfold capillary anomalies is an important component in screening for systemic sclerosis.

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Case Presented by Lisa Blackwood, MD
and Milena J. Lyon, MD

UNKNOWN

This 31 year old male presented with a one year history of multiple, rapidly-growing papules that began on the right chest and subsequently spread to the right arm.

Case 5

Case Presented by Artem Sergeyenko, MD
and Iris K. Aronson, MD

History of Present Illness:
A 72 year old male presented for evaluation of his left hand. The patient reported that he first noticed a small firm lump on his palm in 2013 that was originally diagnosed as a Dupuytren's contracture. Over the next few months, the mass continued to grow and eventually started to bleed. After inconclusive imaging, biopsies, and aspirations by orthopedic surgery, and continued growth, the patient was referred to dermatology for further evaluation.

Past Medical and Surgical History:
Hypertension and myectomy in 2011

Medications:
Atenolol and lisinopril

Allergies:
Sulfa drugs – rash

Family History:
No history of skin cancer or skin diseases

Review of Systems:
The patient denied fevers, chills, nausea, vomiting, diarrhea, shortness of breath, cough, or weight loss.

Physical Examination:
The left central palm has a three centimeter by two centimeter heterogeneous, exophytic, pink nodule with areas of blue and violaceous discoloration with surrounding hemorrhage. The dorsal left hand between the third and fourth metacarpophalangeal joints has a three centimeter by two centimeter ulceration with active hemorrhagic weeping. The entire left hand appears edematous.

Diagnostic Procedures and Tests:
10/14 Magnetic Resonance Imaging, left hand with and without contrast: Large hematomas of the hand with areas of nodular enhancement raise the possibility of tumor dorsal to the third proximal phalanx abutting the extensor tendon, and between the third and fourth proximal phalanges. This could represent a sarcoma or a giant cell tumor, among other etiologies.

10/14 Computed Tomography Angiogram, left hand with contrast: Two large hematomas about the hand, one at the volar aspect of the third and fourth metacarpals and one at the dorsal aspect of the third proximal phalanx dorsally, extending between the third and fourth proximal phalanges. Vessels appear to course around these collections. The enhancing areas suggesting tumor, visualized on the magnetic resonance imaging, are not well seen on this exam. No vascular malformation.

Histopathology:
Left palmar hand, skin: The specimen shows a large, soft tissue neoplasm with extensive hemorrhage. The tumor consists mostly of irregular slit-shaped vessels lined by atypical epithelioid endothelial cells. At higher power, one appreciates the atypical features of the endothelial cells lining the slit-shaped vessels. Immunostaining of the cells shows focal positivity for CD31. Additional stains demonstrate diffuse nuclear positivity for ERG in the cytologically malignant epithelioid cells. There were also irregularly distributed SMA-positive spindle cells around some of these vessels. HHV-8 stain was negative.

Diagnosis:
Epithelioid angiosarcoma

Treatment Course:
After being diagnosed with an epithelioid angiosarcoma the patient underwent a full malignancy work-up that was negative for metastases. In January 2015, he underwent a left upper extremity amputation of the forearm and sentinel lymph node biopsy. The sentinel lymph nodes were negative for metastases. In October 2015, the patient developed a recurrence in the left forearm stump, based on repeat imaging. The patient was referred to Mayo Clinic where he underwent an above left elbow amputation in December 2015. After evaluation by oncology and radiation oncology, no further treatment with chemotherapy or radiation therapy was recommended.

Discussion:
Epithelioid angiosarcomas are rare and aggressive malignancies of endothelial origin. They are more prevalent in men and have a peak incidence in the seventh decade. Tumors most commonly occur in the deep soft tissues of the extremities, but have been reported to form in a variety of primary sites, including the skin, bone, thyroid, and adrenal glands. Tumors tend to be highly aggressive and demonstrate early nodal and solid organ metastases. Within two to three years of diagnosis, 50% of patients die of the disease, and the five-year survival rate is estimated to be 12-20%. The etiology remains unknown, but it has been linked to previous toxic chemicals, irradiation, or Thorotrast contrast media exposure, and may arise in the setting of arteriovenous fistulae or chronic lymphedema. Diagnosis is made with hematoxylin-eosin (H&E) stained sections and immunochemical stains; although, it is often a complex diagnosis and can often be mistaken for a poorly differentiated carcinoma or malignant melanoma. On H&E, one appreciates pleomorphic, polygonal epithelioid cells with eccentric nuclei, prominent nucleoli, abundant eosinophilic cytoplasm, and focal areas of irregularly-anastomosing vessel formation with cellular stratification in a papillary appearance. Additionally, mitotic figures, necrosis, and hemorrhage can also be appreciated. The tumor is often strongly positive for vimentin and CD 31, and can likely be positive for factor VIII, FLI-1, and CD34.

While radiation therapy is often utilized, surgery is the primary treatment modality. Despite wide excision, local recurrence is common. Tumor size is one of the most important prognostic features, with a worse prognosis for tumors greater than five centimeters. Evidence suggests that paclitaxel-based chemotherapeutic regimens may improve survival, and a combination of paclitaxel with sorafenib has been reported to induce remission in metastatic epithelioid angiosarcoma of parietal origin. Currently, no standardized treatment regimen for this condition exists.

Our case demonstrates the classic presentation, in terms of patient age, location, and anatomic location of epithelioid angiosarcoma. The histopathologic diagnosis can be subtle and requires

appropriate use of immunohistochemical stains to confirm the diagnosis. Finally, the recurrent and recalcitrant nature of the disease, despite wide resection, was also apparent in our patient.

Essential Lessons:

- Epithelioid angiosarcoma is an aggressive tumor that requires appropriate use of histochemical stains to facilitate diagnosis.
- Despite wide excision, epithelioid angiosarcoma is a disease with a very high recurrence rate.
- Metastatic epithelioid angiosarcoma treatment with paclitaxel-based chemotherapy has promising results, but standardized therapeutic regimens have not been established for this rare condition

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Case 6

Case Presented by Kimberly Jerdan, MD
and Michelle Bain, MD

History of Present Illness:
Four year old identical triplets were referred from general pediatrics for concern for molluscum contagiosum. These papules were present for four months and were limited to the chest. Per the father, there was no history of trauma, irritation, or manipulation to the affected areas.

Past Medical History:
Prematurity (born at 32 weeks) and congenital dermal melanocytosis.

Medications:
None

Allergies:
No known drug allergies

Family History:
The father reports he had similar papules on his chest during adolescence that resolved with isotretinoin.

Review of Systems:
The patients' father denied fevers, chills, night sweats, or weight loss on their behalf.

Physical Examination:
Erythematous to maroon papules are scattered on the central chest of all three patients. On dermoscopy, homogenous white macules were surrounded by light brown to erythematous halos.

Laboratory Data/Diagnostic Procedures and Tests:
None

Histopathology:
None

Diagnosis:
Eruptive vellus hair cysts in identical triplets

Treatment Course:
The family elected to defer treatment at this time.

Discussion:
Eruptive vellus hair cysts (EVHCs) were first described by Esterly, Fretzin, and Pinkus in 1977. EVHCs are red or brown monomorphous papules overlapping with pilosebaceous and apocrine units. EVHCs are typically found on the chest and extremities, although some have been reported on the face, abdomen, axilla, buttocks or genital area as well.

It has been suggested that most cases of EVHCs are the result of a de novo mutation. However, in the literature, 20 families are affected by autosomal dominant EVHCs based on

phylogeny. In 2015, EVHCs were reported in identical twins further supporting the case for a genetic mutation. Today we augment that by presenting an occurrence of triplets with EVHCs. Interestingly, the patients' father reports similar lesions in his own childhood, further underscoring a genetic basis.

The de novo form is noted to be more common and clinically presents later, with average onset at 16 years old and an average age of diagnosis of 24 years old. This form occurs without preceding trauma or manipulation.

Other variants of EVHCs have been described. Late Onset EVHC occurs age 35 or older, with 57 as the average age of reported lesions, and a female to male predominance of 2.5 to 1. This may be attributed to proliferation of ductal follicular keratinocytes or loss of perifollicular elastic fibers exacerbated by exogenous factors such as manipulation, UV rays, or trauma. Unilesional EVHC is reported with an average age of diagnosis of 27 years old. Some of these lesions may be pedunculated at greater than eight millimeters. There is also a female to male predominance of 2 to 1. EVHCs with steatocystoma multiplex can be seen with an average age of onset 18.7 years old and a female to male predominance of 0.2 to 1. There may be a family history of this subset as reported in three patients with this pattern.

There are two theories to explain the pathogenesis of eruptive vellus hair cysts. The first theory proposes retention of vellus hair and keratin in a cavity formed by an abnormal vellus hair follicle causing infundibular occlusion. The second theory proposes the growth of benign, follicular hamartomas that differentiate to become vellus hairs.

The recommended work up for EVHCs varies by patient and age. EVHCs present an opportunity to employ non-invasive diagnostic procedures, especially for the pediatric population, to avoid scarring and pain from manipulation or biopsy. Although many clinicians may comfortably diagnose EVHCs clinically, one paper suggested six cases with a diagnosis of steatocystoma multiplex, KP or milia prior to histopathology revealing vellus hair cysts.

Dermoscopy presents as a possible diagnostic aid for EVHCs. EVHCs exhibit yellowish-white homogenous circular structures with a maroon or erythematous halo. One may see a central gray-blue color point due to melanin in the pigmented hair shaft. One dermoscopy review of EVHCs also reports radiating capillaries. Occasionally non-follicular homogenous blue pigmentation may be seen due to a connection to atrophic hair follicles in the mid-dermis and no normal hair follicle around the cysts.

Treatment for EVHCs is usually for aesthetic discomfort. Twenty-five percent of EVHCS resolve spontaneously with transepidermal hair elimination or a granulomatous reaction. A case report of four siblings with congenital EVHCs also described a mother with similar lesions that resolved spontaneously in early adulthood, just as our patients' father noted. Otherwise, the treatments listed above have been tried with minimal improvement. Of note, one paper demonstrated that 0.1% tazarotene cream yielded better results than erbium:YAG or incision and drainage of EVHCs. One report demonstrated partial improvement with calcipotriene within two months with some lesions completely resolved and others with flattening. This may be attributed to the antiproliferative and prodifferentiating effects on the ductal follicular keratinocytes by calcipotriene. Another report stated that isotretinoin and Vitamin A derivatives were not effective for clearing EVHCs.

Essential Lesson:

- A subset of eruptive vellus hair cysts are genetic, with a likely autosomal dominant inheritance.
- Dermoscopy can aid in the diagnosis of eruptive vellus hair cysts, which is of particular use in the pediatric population.

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Case Presented by Stephanie Wang, MD, Benjamin Garden, MD
Iris K. Aronson, MD and Michelle Bain, MD

FAST BREAK

This 63-year-old Mexican male with a history of renal transplantation presented for evaluation of a non-healing ulcer on the right ear.

Case 8

Case Presented by Huayi Zhang, MD
and Carlotta Hill, MD

History of Present Illness:
A 63 year old Caucasian male with a past medical history of psoriasis was referred for evaluation of grey, hyperpigmented skin patches and numbness of extremities. The patient was initially diagnosed with psoriasis on skin biopsy in 2004, for which he received topical steroids, narrow band ultraviolet B phototherapy and cyclosporine from 2005-2010. The patient recalled visiting southern Florida and Mexico on numerous occasions during those years. As his psoriasis improved in 2010, he began to feel numbness in his feet which gradually spread to his hands and face. In 2013, the patient underwent electromyography and a positron emission tomography scan, and was diagnosed with small fiber neuropathy. Finally, in March 2016, because of progressive hypoesthesia, the patient underwent left superficial peroneal nerve biopsy at the Mayo clinic which showed acid fast bacilli concerning for leprosy. Four skin biopsies were performed and sent to the National Hansen's Disease Clinic for further evaluation.

Past Medical and Surgical History:
1. Psoriasis
2. Coronary artery disease, status post coronary artery bypass grafting in 2013
3. Aortic regurgitation (status post aortic valve replacement)
4. Hypertension
5. Hyperlipidemia
6. History of nicotine abuse, last use 2011

Medications:
Metoprolol, aspirin, gabapentin, tramadol, and acetaminophen

Allergies:
No known drug allergies

Family History:
Father with psoriasis. No family member with Hansen's disease. No family history of malignancy

Social History:
The patient does not use tobacco (last use 2011), drinks alcohol socially and does not use illicit drugs. He is divorced but is currently engaged. He has several grandchildren that he visits often.

Review of Systems:
The patient confirmed numbness of the cheeks, and numbness and tingling of the hands and feet bilaterally. The patient denied nausea, vomiting, shortness of breath, chest pain, depression, oral pain, or difficulty swallowing.

Physical Examination:
The patient's abdomen and trunk show scattered large erythematous plaques with overlying silvery scales. On the upper and lower back are large grey, hyperpigmented annular patches with mild scaling. Scattered erythematous plaques with silvery scale are also noted on the

patient's elbows, and upper and lower legs bilaterally. A neurological exam shows decreased tactile sensitivity in a stocking and glove distribution.

Laboratory Data:
The following were negative or within normal limits:
Complete blood count with differentials, basic metabolic panel and liver function test, serum angiotensin converting enzyme level, Glucose-6-phosphate dehydrogenase level, urinalysis, and Quantiferon gold

Histopathology:
05/16: Left superficial peroneal nerve – National Hansen's Disease Clinic: Hansen's disease, lepromatous (lepromatous leprosy-borderline lepromatous), active. Polymerase chain reaction assay for *Mycobacterium leprae* DNA is positive

06/16: Left scapula skin – National Hansen's Disease Clinic: Chronic inflammatory infiltrates replace approximately 15% of the dermis. These are composed of disorganized aggregates of lymphocytes and histiocytes at all levels of the dermis. Fite stains reveal small numbers of acid fast organisms consistent with tuberculoid leprosy.

06/16: Right trunk skin – University of Illinois at Chicago: Lesion shows psoriasiform dermatitis without granulomas. Fite stain shows no microorganisms, periodic acid–Schiff and Gomori methenamine silver stain do not show definitive fungal elements.

Diagnosis:
Hansen's disease – lepromatous leprosy

Treatment Course:
The patient was started on minocycline 100 milligrams once daily, rifampin 600 milligrams once a month, dapsone 100 milligrams daily, and prednisone 60 milligrams tapered every two weeks by 10 milligrams. He noted improvement of the numbness in his feet bilaterally.

Discussion:
Leprosy, also known as Hansen's disease, is a chronic infection of the skin and peripheral nerves caused by *Mycobacterium leprae*. Although its coexistence with psoriasis is extremely rare, the two diseases shared a similar classification in ancient times. Recent publications suggest that genetic factors, reinforced innate immunity and the role of neuropeptides and apoptosis may have an impact on the rarity of this coexistence. In a global survey conducted by Kumar et al of 145,661 cases of leprosy, only 20 individuals had psoriasis. This corresponds to a psoriasis prevalence of 0.014%, two orders of magnitude lower than the expected for the world population.

Several findings serve to illustrate the seemingly disparate nature of the two diseases. *Mycobacterium leprae* invades nerves, causing nerve damage, which results in neuritis and hypoesthesia of the skin. Psoriasis on the other hand, requires intact nerves, with studies showing the functional role of cutaneous nerves and their neuropeptides in the pathogenesis of psoriasis. Previous nerve damage decreases neurogenic inflammation, which can inhibit psoriatic plaque formation. Genetic studies have shown the association of HLA-DR2 and HLA-DQW1 with leprosy, but HLA-B13 and HLA-B17 with psoriasis. It has also been noted that thick psoriatic plaques may be the result of decreased apoptosis, whereas in leprosy an increase in spontaneous apoptosis is often seen. All above findings support the hypothesis that psoriasis and leprosy are almost mutually exclusive.

However, our patient presents with this rare coexistence. Recent studies published by Bassukas et al pose a new hypothesis that psoriasis may have the propensity to protect against the development of clinical leprosy through an overstimulation of the innate immunity and an amplification of the antibacterial defense mechanisms. The Th1 response is heightened in the skin of psoriasis patients, which may be responsible for local control of *M. Leprae.* Even though our patient travelled frequently to areas where leprosy is endemic and may have contracted the disease early on, the signs and symptoms of leprosy were not present while he had more aggressive psoriasis. He manifested with leprosy only after undergoing treatment for psoriasis. However, to completely support the hypothesis, one will need to pursue genetic and additional testing for confirmation.

Essential Lesson:

- Leprosy and psoriasis are almost mutually exclusive diseases due to genetic, immune and cell mediated factors.
- Leprosy is associated with HLA-DR2 and HLA-DQW1, psoriasis is associated with HLA-Cw6, HLA-B13, and HLA-B17.
- Psoriasis may protect against the development of clinical leprosy through overstimulation of the innate immunity, amplification of the antibacterial defense mechanisms, and a heightened Th1 response.

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Case 9

Case Presented by Mark Juhl, MD, Michael Sotiriou, MD and Maria M. Tsoukas, MD, PhD

History of Present Illness:
A 35 year old male presented for a facial rash of six months duration. He described several small papules, initially on the left chin, which progressively enlarged. He subsequently developed similar lesions on the scalp and cheek. The lesions were mildly pruritic but otherwise asymptomatic. He reported shaving, but denied other forms of trauma, sick contacts, or recent travel.

Past Medical History:
Asthma and hypertension

Medications:
Albuterol and furosemide

Allergies:
No known drug allergies

Family History:
No history of skin cancer

Social History:
The patient smoked cigarettes daily with a ten pack-year history. He drank a quart of vodka weekly, smoked marijuana occasionally, and reported unprotected sex with multiple partners, both male and female.

Review of Systems:
The patient denied fevers, chills, weight loss, diarrhea, headaches, stiff neck, abnormal gait, and numbness.

Physical Examination:
The mentum and submentum has a large, crusted, eroded, and indurated verrucous plaque with a raised border and draining purulent material. Similar boggy plaques are noted on the left cheek and left scalp. There is no regional lymphadenopathy.

Laboratory Data:
The following were positive or abnormal:
Human immunodeficiency virus antibody screen: reactive
Human immunodeficiency virus, quantitative: 910 copies/ml
Rapid plasma reagin, qualitative: reactive
Rapid plasma regain, quantitative: 1:128 dilutions
Tissue culture for aerobic bacteria: Methicillin-resistant *Staphylococcus aureus* and *Citrobacter koseri*

The following were negative or normal:
CD4 Count: 607 cells/µl (normal 500-1500 cells/µl)
Quantiferon gold
Tissue culture for anaerobic bacteria, fungal, viral, and atypical mycobacteria

Histopathology:
Chin, skin: Pseudoepitheliomatous hyperplasia with dense dermal acute and chronic inflammation, composed of neutrophils, lymphocytes, and prominent plasma cells. Gomori methenamine silver, Fite, and treponemal immunostaining were negative. A gram stain showed gram-positive cocci in clusters.

Diagnosis:
Blastomycosis-like pyoderma

Treatment Course:
Based on sensitivities, a 14 day course of doxycycline was initiated. Simultaneously, he was referred to infectious disease for treatment of concurrent syphilis with benzathine penicillin G and human immunodeficiency virus with antiretroviral therapy. Skin lesions completely resolved after five weeks.

Discussion:
First described in 1903 as “pseudoepiteliomas cutanés,” blastomycosis-like pyoderma (BLP) is a chronic pyoderma that presents similarly to vegetating deep fungal infections. It typically presents as one or multiple vegetating nodules and/or plaques on the extremities of middle aged to elderly adults. Necrosis, pustules, fistulae, and abscesses may also be present. Minor trauma and sun-damaged skin are thought to increase the likelihood of BLP. This entity potentially represents an exaggerated inflammatory reaction due to immune dysregulation and an underlying, prolonged pyogenic bacterial infection. The most commonly reported pathogen is *Staphylococcus aureus*; additionally, β-hemolytic streptococci, certain gram-negative bacteria including *Pseudomonas aeruginosa,* and members of the enterobacteriaceae family, notably citrobacter species, have been reported. Mixed bacterial infections have rarely been reported in BLP.

Proposed diagnostic criteria in recent literature for the diagnosis of BLP include: (1) large verrucous plaques with multiple pustules and an elevated border, (2) typical histologic findings of psudoepitheliomatous hyperplasia with abscesses on biopsy, (3) growth of one or more pathogenic bacteria, and (4) negative cultures for other infectious etiologies.

Many patients with this entity have decreased immunologic resistance to bacterial infections due to diagnoses including human immunodeficiency virus, as seen in our patient, malnutrition, alcoholism, leukemia, immunosuppressant use, and/or radiation therapy. Monotherapy with systemic antibiotics requires long-term treatment and often fails. Our patient’s immune status was not only compromised by an untreated human immunodeficiency virus (HIV) infection, but also by a concurrent syphilitic infection. Syphilis has been reported to cause immune dysregulation including, but not limited to, altered cell surface markers and increased likelihood of HIV co-infection. Our patient’s rapid and complete response to doxycycline and penicillin is unusual for BLP. This raises the hypothesis that the concurrent syphilitic infection may have contributed to immune evasion by these bacteria. Treatment of the concurrent syphilis may have hastened the resolution of his BLP.

Due to the rarity of this entity, no controlled trials have been completed, but additional treatments with varying results have been reported including curettage, surgical excision, carbon dioxide laser, potassium iodide, permanganate soaks, radiotherapy, disodium chromoglycate, and acitretin. Effective treatment with systemic retinoids, as well as combination of trimethoprim-sulfamethoxazole and cryotherapy, have been reported.

Essential Lesson:

- Blastomycosis-like pyoderma should be included in the differential diagnosis of large verrucous plaques.
- If Blastomycosis-like pyoderma is diagnosed, an emphasis should be placed on discovering and treating any underlying immunodeficiency.

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Case 10

Case Presented By Eden Lake, MD,
Lawrence S. Chan, MD and Maria M. Tsoukas, MD, PhD

History of Present Illness:
This 24 year old female presented with several months of a progressive scalp ulcer. The scalp ulcer began in 2012 as a coin-shaped lesion, and was initially diagnosed as discoid lupus erythematosus based on biopsy results by an outside physician. She was treated with topical triamcinolone cream which was not effective. A few months prior to presentation at our hospital, the patient noted skin lesions on the back, arms and face. She presented to the emergency department due to increased pain and purulence from the scalp lesion.

Past Medical History:
Self-reported history of lupus

Medications:
None prior to the current diagnosis

Allergies:
No known drug allergy

Review of Systems:
The patient denied fevers, chills, weight loss, changes in urination, easy bruising, fatigue, but did notice intermittent left upper quadrant pain.

Physical Examination:
The patient's scalp is superficially debrided revealing an erythematous, eroded, boggy scalp that is very tender to palpation. The remaining scalp shows crusting and scale adherent to the residual hair, with yellow to brown debris. Her face has few erythematous papules and hyperpigmented macules. There is no conjunctival injection of the eyes and no erosions or erythema of the oral mucosa. The bilateral extensor arms have erythematous, hyperpigmented macules and patches as well as crusted plaques and few flaccid bullae, one which shows a positive Nikolski sign. The bilateral anterior lower extremities have erythematous crusted plaques. The lower abdomen has one large erythematous erosion as well as hyperpigmented macules and patches and one violaceous plaque with overlying crust. The upper to mid-back has erythematous, violaceous, and hyperpigmented patches and eroded plaques, some with overlying hemorrhagic crust. Some macules and patches are annular in configuration with a hyperpigmented rim and central hypopigmentation. The upper back has few flaccid, slightly erythematous bullae.

Laboratory Data:
The following were positive or abnormal:
Complement 3 elevated to 177 mg/dl (normal 79-152)
Erythrocyte sedimentation rate elevated to 55 mm/hr (normal 0-20)
Enzyme Linked Immunosorbent Assay:
IgG Desmoglein 3 antibodies: elevated to 69 units (normal <20)
IgG Desmoglein 1 antibodies: elevated to 340 units (normal <20)

The following were negative or within normal limits:
Antinuclear antibody, antibodies to dsDNA, ssDNA, Smith, SSA, SSB, RNP.
Complete blood count, basic metabolic profile, urinalysis, HIV antibody, hepatitis acute panel and Quantiferon Gold assay. Blood culture had no growth

Diagnostic Procedures and Tests:
05/16 X-ray, Skull Partial: Negative for osseous destruction to suggest osteomyelitis

Histopathology:
Right scalp, skin (hematoxylin and eosin stain): Suprabasilar and intraepidermal acantholysis with no interface or basal vacuolar changes. Herpes simplex virus and periodic acid–Schiff stains are negative.

Right forearm, skin (direct immunofluorescence): 3+ granular IgG deposition along the dermal epidermal junction; 2+ intraepidermal IgG intercellular deposition. 2+ speckled to granular deposition of C3 along the dermoepidermal junction. 1-2 + fibrinogen is seen around blood vessels.

Diagnosis:
Pemphigus erythematosus (Senear-Usher Syndrome)

Treatment Course:
The patient was treated with systemic steroids during her hospitalization. She received intravenous vancomycin and oral clindamycin for superimposed infection of the scalp with methicillin-resistant *Staphylococcus aureus*. Upon discharge from her first hospitalization the patient was prescribed oral prednisone 60 milligrams daily, which she took for one month. She was asked to return for follow-up but could not due to lack of insurance. Instead, she presented to the emergency department after six weeks, seeking further treatment. She had improved with the prednisone but was observing recurrence. No additional medications could be started also due to lack of insurance, and the patient had not been able to return to clinic for treatment or laboratory monitoring. At her most recent visit, upon obtaining insurance coverage, the workup was initiated to start her on a steroid-sparing immunosuppressant such as azathioprine.

Discussion:
Pemphigus erythematosus (PE) was first described in 1926 by Dr. Senear and Dr. Usher in a case series of 11 patients. The case series demonstrated an overlapping clinical presentation of pemphigus foliaceus (PF) and lupus erythematosus, seen in middle-aged patients with higher prevalence in females. Clinically the patient often has malar involvement that mimics a severe seborrheic dermatitis with well-defined erythematous, scaly, crusted plaques. Non-facial lesions may begin as small, flaccid bullae with a positive Nikolsky sign, favoring the upper trunk and face, although lesions have been reported to extend to the feet. Lesions often resolve with hyperpigmentation. Consistent with the clinical findings in PF, mucosal involvement in PE is rare. Our patient has an unusual presentation with significant scalp involvement and lack of a prominent facial seborrheic or malar dermatitis.

Diagnosis of PE is made with direct immunofluorescence (DIF) demonstrating immunoglobulin G (IgG) and complement deposition both intercellularly and at the dermoepidermal junction (DEJ), along with the clinical and pathological findings of pemphigus foliaceus. This DEJ deposition (defined particularly in non-lesional skin) is occasionally referred to as a lupus band. Complement at the basement membrane may be seen in PF, however the presence of immunoglobulins at the junction is rare. While antinuclear antibody serology is positive in 30-

80% of patients, PE patients rarely meet the diagnostic criteria for systemic lupus erythematosus. Cases have been reported with normal lupus serologies, normal complement studies, and normal inflammatory markers. Enzyme Linked Immunosorbent Assay serology is also helpful, often yielding positivity for both antibodies to desmoglein 1 and desmoglein 3, as seen in our patient. Histopathology alone demonstrates acantholysis within the superficial epidermis, consistent with PF.

There is an academic debate regarding the significance and etiology of the linear deposition of IgG and complement at the DEJ. Both PE and PF have been reported to have severe exacerbations with ultraviolet (UV) exposure. It has been demonstrated that *in vivo* high doses of UV exposure can induce cleavage of the desmoglein 1 ectodomain, and in PF the auto-antibodies to desmoglein 1 can precipitate the cleaved ectodomain along the basement membrane, resulting in DEJ deposition. These findings may be present on only UV-exposed sites in a patient with PE. This same finding can be seen in other forms of cutaneous lupus, with DEJ deposition present on sun-exposed lesional skin but not on sun-exposed non-lesional skin.

PE is often easier to manage than pemphigus vulgaris. Treatments such as systemic prednisone as well as topical corticosteroids, and dapsone may be particularly effective. Other potential treatments include methotrexate, cyclophosphamide and azathioprine. Avoidance of UV exposure is critical for the overall management of both PE and PF.

Essential Lesson:

- Senear-Usher syndrome (pemphigus erythematosus) is a rare variant of pemphigus foliaceus with deposition of IgG and/or complement at the basement membrane seen on direct immunofluorescence.
- The treatment consists of systemic steroids, dapsone, and/or immunosuppressants. Ultraviolet protection is critical to management.

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Presented by Monica Boen, MD
and Maria M. Tsoukas, MD, PhD

UPDATE

We presented a case of Acrodermatitis Continua of Hallopeau to CDS in 2014. This is a brief update on his care.