Chicago Dermatological Society

October 2017 Educational Conference

Program & Speaker Information
CME Certification
Case Presentations
David Fretzin Lecture

Wednesday, October 4, 2017
Gleacher Center
Chicago, IL

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

Program

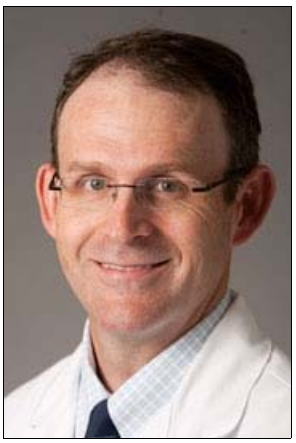
Host: University of Illinois at Chicago
Wednesday, October 4, 2017
Gleacher Center, Chicago

8:00 a.m.	**Registration & Continental Breakfast with Exhibitors** *All activities will take place on the 6th Floor of the Gleacher Center*
9:00 a.m. - 10:30 a.m.	**Clinical Rounds** Slide viewing/posters Patient viewing
9:00 a.m. - 10:00 a.m.	**Basic Science/Residents Lecture** "Game of Unknowns" *Shane Chapman MD*
10:00 a.m. - 10:30 a.m.	**Break and Visit with Exhibitors**
10:30 a.m. - 12:15 p.m.	**Resident Case Presentations & Discussion;** **MOC Self-Assessment Questions**
12:15 p.m. - 12:45 p.m.	**Box Lunches & visit with exhibitors**
12:55 p.m. - 1:00 p.m.	**CDS Business Meeting**
1:00 p.m. - 2:00 p.m.	**General Session** FRETZIN LECTURE - "Non-surgical Treatment of Melanoma" *Shane Chapman MD*
2:00 p.m.	**Meeting adjourns**

Mark the Date!
Next CDS monthly meeting – Hosted by Northwestern University
Wednesday, November 8th; Gleacher Center, Chicago

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

Guest Speaker



SHANE CHAPMAN MD

Associate Professor of Medicine
Geisel School of Medicine Dartmouth
Lebanon, NH

Dr. Shane Chapman is a Section Chief for Dermatology and Associate Professor of Medicine at the Geisel School of Medicine at Dartmouth College. Dr. Chapman received his medical degree from the University of Texas at Houston Medical School in Texas in 1995, where he also completed his internship in internal medicine. Dr. Chapman's residency in dermatology was at Dartmouth-Hitchcock Medical Center in Lebanon, NH (1999).

Dr. Chapman is board certified in dermatology and his clinical interests include psoriasis, atopic dermatitis, laser therapy and immunotherapy of melanoma.

CME Information

This educational activity is jointly provided by the Chicago Dermatological Society in partnership with the Indiana Academy of Ophthalmology

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 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 2017/18 series of meetings, the participant should be able to:

1. Discuss key factors in the diagnosis and treatment for allergic conditions, 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 Indiana Academy of Ophthalmology (IAO) and the Chicago Dermatological Society. IAO is accredited by the Indiana State Medical Association to provide continuing education for physicians.

Credit Designation for Physicians – IAO designates this live activity for a maximum of 4.5 *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 evaluation form, at the registration table when you leave the meeting. Thank you for your attention to this important item. Note - You may complete the paper version of the evaluation form or submit your evaluation online.

Disclosure of Conflicts of Interest

The IAO and CDS require 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 IAO and CDS 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 asked to follow the "first slide" rule to repeat their conflict of interest disclosures during their talk.

Today's guest speaker, Shane Chapman, MD,, has disclosed the following potential conflicts of interest: Grants/Research Support - Genentech, Celgene, Dusa. None of the planning committee members have any conflicts of interest to disclose.

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Contact Information

For information about the physician accreditation of this program please contact the CDS administrative office at: 847-680-1666; email: Rich@RichardPaulAssociates.com

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, contact CDS at: Rich@RichardPaulAssociates.com

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.

Chicago Dermatological Society
Patient Privacy and HIPAA Compliance

APPROVED
June 3, 2015

Purpose

The purpose of this policy is to reaffirm the intent of the Chicago Dermatological Society (CDS) to appropriately safeguard patient privacy with respect to CDS conferences, publications and its website, and also to adhere to HIPAA requirements. All CDS members are expected to be aware of and conform to all regulations concerning patient privacy when attending a conference or utilizing any materials produced by CDS which contains any form of patient information which could be considered to be Protected Health Information.

Background

The Health Insurance Portability and Accountability Act of 1996 (HIPAA) and its implementing regulations restrict health care providers and others to use and disclose protected health information (PHI). Protected health information means information that is created or received by an entity and relates to the past, present, or future physical or mental health condition of a patient; the provision of health care to a patient; or the past, present, or future payment for the provision of health care to a patient; and that identifies the patient or for which there is a reasonable basis to believe the information can be used to identify the patient. Protected health information includes information of persons living or deceased.

Some examples of PHI are:
- Patient's medical record number
- Patient's demographic information (e.g. address, telephone number)
- Information doctors, nurses and other health care providers put in a patient's medical record
- Identifiable images of the patient
- Conversations a provider has about a patient's care or treatment with nurses and others
- Information about a patient in a provider's computer system or a health insurer's computer system
- Billing information about a patient at a clinic
- Any health information that can lead to the identity of an individual or the contents of the information can be used to make a reasonable assumption as to the identity of the individual

Policy

The CDS takes seriously compliance with HIPAA regulations and safeguards concerning protected health information. Accordingly, the Chicago Dermatological Society has adopted the following provisions:

1. Case descriptions included in clinical conference "protocol books" and posters may not include information that could potentially identify a particular patient.

2. Photos of patients will not be published in clinical conference handout materials, including the protocol book.

3. To the extent possible, posters, slide presentations and videos displayed at CDS clinical conferences should avoid using photos that display a patient's full face or other features that could identify a particular patient. When a full-face photo must be used for clinical/educational reasons, the photo must be altered as much as possible to disguise the identity of the patient.

4. At all times, all attendees of CDS clinical conferences must adhere to appropriate behavior that respects the patient's right to privacy. Taking personal photos of posters or other displays, images included in general session lectures/presentations, and live patients at CDS conferences is strictly prohibited. Making audio recordings of any session at a CDS conference also is prohibited.

5. Attendees may not share materials distributed by CDS as part of the clinical conference or on its website with others who are not participating in the conference or who are not members of the CDS.

6. It is the responsibility of the "host" department partnering with CDS for a clinical conference to obtain all appropriate patient waivers and/or informed consent regarding the patient's participation in the CDS conference, including presentation of their case and display of posters or photos.

7. CDS will include a copy of its patient privacy policy in every meeting packet, and it will display a poster reiterating this policy at the entrance to live patient and poster viewing areas.

University of Illinois at Chicago
Department of Dermatology

FACULTY

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Artem Sergeyenko, MD
Stephanie Wang, MD

First Year

Kurt Ashack, MD, MHS
Jeremiah Au, MD
Olivia Lai, MD
Regina O'Brien, MD

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Case Presented by Lisa Blackwood, MD
and Michelle Bain, MD

History of Present Illness:
A five month old boy presented for evaluation of recurrent blisters on the hands and feet, first noted at two weeks of age. He was born at 39 weeks gestation to a healthy mother and had no cutaneous findings at birth. The mother stated that the patient did not appear distressed by the lesions. On disruption, the blisters drained a clear fluid, occasionally admixed with blood. He was otherwise healthy and meeting all development milestones.

Past Medical and Surgical History:
None

Medications:
None

Allergies:
No known drug allergies

Family history:
No known family history of skin diseases or genetic disorders

Physical Examination:
On the dorsal hands and feet, especially over joints, there are multiple pink to white papules ranging in size from 1 to 4 mm. The papules extend proximally to the wrists and ankles with a single tense bulla noted on the right anterior lower leg. Hemorrhagic erosions are also noted on the left first toe and fifth toe. Several pink macules are scattered on the forearms and lower legs. There is mild dystrophy of several toenails, found to be more pronounced at follow-up visit six months later.

Histopathology:
Right leg, skin: The specimen shows a subepidermal blister lacking inflammatory cells. The papillary dermis contains mild perivascular lymphocytic infiltrate. A PAS stain does not show thickening of the walls of small vessels in the papillary dermis.

Left hand, skin: The specimen shows multiple small squamous-lined keratin-filled cysts with a granular cell layer within the dermis. There is fibroplasia around these small cysts.

Diagnostic Procedures and Tests:
Immunofluorescence antigenic mapping, skin left leg (Stanford Dermatopathology Consultants):
Immunofluorescence tissue is stained with antibodies to basement membrane antigens including LAD-1 (123, collagen XVII), B4D5 (laminin 332, gamma 2 chain), collagen IV, and collagen VII. There is normal intensity staining with collagen IV, collagen XVII, and laminin 332, and these antigens all localize to the roof of the blister. Interestingly, the COLVII is present in normal intensity and localized to the roof of the blister in a highly irregular and serrated pattern. Findings suspicious for dystrophic epidermolysis bullosa

Epidermolysis Bullosa Panel Comprehensive Analysis Report:
Mutation/Alteration

1. COL7A1 Exon 74-Heterozygous
2. DSP Exon 23-Heterozygous

Nucleotide

1. C.6190G>A
2. C.3293A>G

Protein

1. Gly2064Arg
2. Asp I098Gly

Results: Consistent with a clinical diagnosis of Epidermolysis bullosa

Diagnosis:

Dominant dystrophic epidermolysis bullosa

Treatment and Course:

The patient and his family were referred to pediatric genetics for genetic testing and counseling. Mutational analysis revealed two heterozygous mutations, as shown above. The first mutation in the COL7A1 gene (c.6190G>A) was known to be a pathogenic variant associated with autosomal dominant dystrophic epidermylosis bullosa (DDEB). Genetic testing of the parents was recommended to determine if the mutation arose de novo or if it was familial.

Unfortunately, there is no cure for dominant dystrophic epidermolysis bullosa or any form of inherited epidermolysis bullosa. Management primarily consists of prevention of skin trauma, gentle and appropriate wound care, maintenance of proper nutrition, prevention of infection, and surveillance for extracutaneous complications. Psychosocial support is also of utmost importance, and all patients and their families should be referred to the Dystrophic Epidermolysis Bullosa Research Association (DEBRA), which provides a wealth of educational information and support.

Discussion:

Epidermolysis Bullosa (EB) is a clinically heterogeneous group of rare genetic disorders that are characterized by varying degrees of skin fragility and recurrent blister formation following minor trauma. There are four major types of EB, based on the level of skin cleavage: EB simplex (intraepidermal), junctional EB (intra-lamina lucida), dystrophic EB (sublamina densa), and Kindler syndrome (multiple levels within and/or beneath the basement membrane zone). These four major subtypes can be further subdivided into more than 30 different subtypes based on distribution and severity of skin findings, mode of inheritance, and molecular testing.

Clinical manifestations range widely from mild, localized cutaneous involvement to severe, generalized cutaneous involvement with extracutaneous manifestations. All forms of dystrophic EB are the result of mutations in the COL7A1 gene encoding type VII collagen, the major constituent of anchoring fibrils in the sublamina densa. In general, patients with DDEB have reduced or altered expression of type VII collagen and have a good prognosis, whereas the recessive form of EB is often associated with complete loss of type VII collagen and a much more severe phenotype and poor prognosis. In DDEB, blistering is often mild and limited to sites of trauma, typically resulting in scarring and milia formation. Additionally, dystrophic nails, especially toenails, are common and may be the only manifestation of DDEB. Conversely, the more severe type of RDEB is characterized by widespread blistering of the skin and mucosa with subsequent mutilating scarring and extracutaneous manifestations leading to shortened lifespan.

Clinical and routine light microscopy features of different EB subtypes may overlap, especially in infancy. Therefore, correct diagnosis requires additional testing, such as immunofluorescence antigenic mapping (IFM), transmission electron microscopy (TEM), or mutation analysis. IFM on a freshly induced blister has replaced TEM as the preferred initial diagnostic test as it has a higher degree of sensitivity and specificity, is more cost effective, and is more widely available. In IFM, antibodies (directed against specific basement membrane antigens) conjugated with fluorochromes are applied to skin sections and examined using fluroescence microscopy. This allows visualization of the specific layer of tissue separation in addition to the relative expression and distribution of various basement membrane zone antigens.

Mutation analysis is recommended to identify the specific genetic mutation and to determine the mode of inheritance, if testing available and if cost is not prohibitory. It is particularly important when evaluating patients with suspected Kindler syndrome and those who are the first members in the family to be affected, as is the case for our patient. This case illustrates the importance of obtaining both IFM and genetic analysis, if possible, for the diagnosis of EB. A precise diagnosis is critical to evaluate the patient's prognosis and to counsel family members regarding personal risk as well as risk to their offspring.

On mutational analysis, our patient was found to have two heterozygous mutations. The first mutation in the COL7A1 gene (c.6190G>A) was known to be pathogenic and associated with autosomal dominant dystrophic epidermylosis bullosa (DDEB).

The second mutation, c.3293A>G, was found in the gene which codes for desmoplakin (DSP). This specific mutation has not been previously reported as a disease causing mutation and was therefore a variant of uncertain clinical significance. However, mutations in the DSP gene have been associated with several autosomal recessive disorders, including but not limited to dilated cardiomyopathy with woolly hair and keratoderma (Carvajal syndrome) and lethal acantholytic epidermolysis bullosa. Though our patient was not noted to have woolly hair, further workup was recommended to rule out associated findings, such as dilated cardiomyopathy. Baseline echocardiogram, parental genetic testing, and continued follow-up are critical for appropriate treatment of the patient as well as for a better understanding of the clinical significance, if any, of this additional mutation.

Essential Lesson:

- Dominant dystrophic epidermolysis bullosa (EB) is a rare genetic disorder caused by a heterozygous mutation in the type VII collagen gene, COL7A1.
- Immunofluorescence mapping (IFM) on a freshly induced blister is recommended as the primary method for the diagnosis of EB.
- Mutation analysis is recommended for confirmation of the EB subtype and is critical in patients with suspected Kindler syndrome or those with an unclear inheritance pattern
- All patients with EB should be referred to the Dystrophic Epidermolysis Bullosa Research Association (DEBRA)

References:

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Case #2

Case Presented by Stephanie Wang, MD
and Maria Tsoukas, MD, PhD

UNKNOWN

This 32 year old female presented with painful, crusted ulcerations on the abdomen and thighs.

Case Presented by Jeremiah Au, MD
and Iris Aronson, MD

History of Present Illiness:
A 19 year old healthy Caucasian female presented with a seven month history of a spontaneously enlarging red plaque on her right shin. The plaque was mildly painful to touch and had occasional yellow and bloody drainage. Prior to evaluation by UIC Dermatology, the patient's work-up included two biopsies suggesting lobular panniculitis, and negative bacterial and fungal cultures. Additionally, the patient failed trials of both oral clindamycin and doxycycline. Finally, she was treated with dapsone which was discontinued due to a drop in hemoglobin level.

Past Medical History:
None

Medications:
None

Allergies:
Penicillin

Family History:
Mother – Heterozygous MZ alpha-1-antitrypsin deficiency
Three Maternal Aunts – homozygous ZZ alpha-1-antitrypsin deficiency

Social History:
Patient is a college student and teaches children swimming classes in the summer. She does not drink alcohol, smoke, or use illicit drugs.

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

Physical Examination:
The patient has a 6x7 cm erythematous plaque on her right mid-pretibial leg. One section of the lesion is pink and atrophic while another portion is crusted with faint drainage of serosanguinous fluid.

Diagnostic Procedures and Laboratory Data:
Initial laboratory data:
Monospot positive (12/12/14)
Epstein-Barr Virus early antigen 32.4U/mL (>10.9 U/mL positive) (12/19/14)
Epstein-Barr Virus antibody VCA IgG 134 U/mL (>21.9 U/mL positive) (12/19/14)
Epstein-Barr Virus Nuclear Antigen antibody IgG 126 U/mL (>21.9 U/mL positive) (12/19/14)
Erythrocyte sedimentation rate 64 mm/hr (0-29 mm/hr) (12/19/14)
C-reactive protein 40.5 mg/L (<1.0 mg/L) (12/19/14)
Hemoglobin 10.1 g/dL (11.7g/dl – 16 g/dL) (4/9/15)
Alpha 1 Anti-trypsin 99mg/dL (reference 90-200mg/dL) (5/18/15)

The following were negative or within normal limits:
Quantiferon Gold, Gliadin IgA, Gliadin IgG, tissue transglutaminase IgA, tissue transglutaminase IgG, endomysial antibody, C-Anti-neutrophil cytoplasmic antibody, P-Anti-neutrophil cytoplasmic antibody, Hepatitis C antibody IgG, Epstein-Barr Virus antibody VCA IgM, fungal culture, bacterial culture, acid fast bacilli cultures

Histopathology:
On hematoxylin and eosin staining the subcutaneous fat displayed mixed septal and lobular panniculitis with a dense neutrophilic infiltrate. There was evidence of fibrinolysis of the septae. These septae stained negative for elastin and the trichrome stain showed a loss of the normally condensed collagenous matrix within the fibrous septae. MPX and CD68 stained positive. A1AT stain exhibited the presence of cellular uptake.

Diagnosis:
Alpha-1-antitrypsin heterozygous deficiency (MZ) panniculitis

Treatment and Course:
Due to extensive family history of alpha-1-antitrypsin mutations, the patient was sent for evaluation by Genetics. She was found to carry the Z mutation (MZ). Over the course of two years, the patient experienced multiple cycles of flares and improvement while on a topical over-the-counter silver-based antibacterial gel, topical metronidazole and oral medications including doxycyline, dapsone and colchicine. These flares were mostly secondary to minor trauma to the area. Since May of 2017, in addition to Colchicine 0.6mg daily, the patient began weekly augumentation therapy with alpha-1-proteinase inhibitor (Zemaira®) infusions (60mg/kg) and noted significant improvement of the lesions. No new lesions have appeared since that time. The patient continues to have regular follow-ups with her primary care physician and pulmonologist.

Discussion:
Alpha-1-antitrypsin deficiency (ATAD) is a rare genetic disorder that is associated with chronic obstructive pulmonary disease, liver cirrhosis, and panniculitis. ATAD related panniculitis can develop in all age groups but most commonly appears between ages 30 to 60 years. Both genders are equally affected. The ZZ genotype has been linked to most cases of panniculitis, however, homozygous genotype SS, and heterozygous MZ, SZ, and MS genotypes related panniculitis have also been reported.

Ulcerated nodules and plaques with drainage of serosanguinous and oily material are the typical cutaneous morphology described in many case reports. Lesions are most frequently found on proximal extremities and the lower trunk and may be preceded by trauma. Secondary infection can be life-threatening. Regarding pathogenesis, the following mechanisms have been proposed: (1) decreased activity of A1AT leading to unopposed neutrophil elastase action and subsequent tissue breakdown; (2) neutrophil release of myeloperoxidase, which may inactivate the already poorly functioning A1AT protein; (3) decreased inhibition of membrane-bound serine proteases, which may then activate macrophages and lymphocytes.

There has been no standard of treatment for ATAD panniculitis. However, several treatment options have been utilized with variable levels of effectiveness. Dapsone, with its myeloperoxidase inhibitory effect, has successfully treated many patients with dosage range of 50 to 150mg daily. Doxycycline 100-200 mg per day works by inhibiting lipase and reducing reactive oxygen species produced by neutrophils. Augmentation therapy with A1AT replacement is a promising but expensive treatment option. Since starting Zemaira®

(completed 4 out of 12 weekly infusions at the time of writing), our patient noticed significant improvement in her existing lesion and has not noticed development of new lesions. The length of remission produced by A1AT infusions in MZ panniculitis is not yet known since there are only rare case reports on successful treatments in patients with ZZ genotypes. Other treatment options such as colchicine, cyclophosphamide, fenoprofen, prednisone, and plasma exchange have shown variable effectiveness in case reports.

Our patient initially presented with positive EBV laboratory testing, which may be an independent cause of panniculitis. Infections must be looked for as the etiology of panniculitis with ATAD appearing only as a cofactor. A case was reported detailing an ATAD patient with panniculitis and concomitant histoplasmosis infection whose panniculitis improved with ketoconazole treatment. In addition, EBV infections may be associated with subcutaneous lymphomas, therefore follow-up with periodic biopsies may be indicated. Thus, it is important to be cognizant about other potential causes of panniculitis even in ATAD patients.

Essential Lesson:

- Panniculitis may be the only manifestation of ATAD at the time of diagnosis.
- Dapsone, doxycycline, and colchicine have been shown to be effective treatment options.
- Augmentation therapy with A1AT replacement can potentially lead to resolution of the disease in refractory cases, albeit at a significantly higher cost with unknown length of remission.

References:

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Case #4

Case Presented by Kurt Ashack, MD
and Iris Aronson, MD

History of Present Illness:
A 61 year old female with a history of a recently diagnosed pancreatic neuroendocrine tumor with metastasis to the liver presented with a 10 month history of pruritic and painful erythematous macules, patches, and plaques limited to the back and lower extremities. She had been initially treated for a presumed fungal infection with topical antifungals without improvement and topical steroids, which mildly improved her eruption. The UIC oncology department started the patient on monthly octreotide injections with suspicion that the rash was related to the carcinoid tumor. She received two injections with minimal improvement of the skin eruption. She was referred to dermatology for further management.

Past Medical History:
Metastatic pancreatic neuroendocrine tumor, chronic venous stasis, post-traumatic stress disorder, and recently treated Helicobacter Pylori infection

Medications:
Lansoprazole, lorazepam, venlafaxine, econazole 1% cream, clobetasol 0.05% ointment, hydrocortisone 1% cream, and octreotide injections

Allergies:
No known drug allergies

Family History:
No history of malignancy or skin disorders

Social History:
The patient has a 20 pack-year history of tobacco use, but has since quit smoking. She is married and has three children.

Review of systems:
The patient reported a 15 pound weight loss over the past year, as well as fatigue, nausea, vomiting, tongue pain, sore throat, severe pruritus, and pain with palpation of the lesions. The patient denied any burning sensations, fevers, chills, night sweats, joint pains, flushing, wheezing, palpitations, or diarrhea.

Physical Examination:
The patient has erythematous macules and patches with peripheral scale, rare erosions, and some erythematous and edematous plaques, admixed with a few erythematous pustules on her buttocks, lateral and inner thighs, lower legs, and dorsal feet. The lesions are more concentrated on the upper thighs and buttocks. The lesions on the inner thigh near the pubic area have peripheral erosions. The periungual regions of the left second and third digit have erythema and scaling. There is mild scaling and lichenification of the palms, particularly at the fingertips. The patient's tongue is erythematous and smooth with several erosions on the posterior third aspect. Oropharyngeal erythema is present. There is no regional lymphadenopathy.

Laboratory Data:
The following were positive or abnormal:
Hemoglobin: 10.6 g/dL (11.7-16.0)
Glucose: 222 mg/dL (65-110)
Chromogranin A: 526 ng/mL (0 - 95)
Glucagon: 1930 ng/L (<= 208)
CA 19-9: 65 U/mL (0-37)

The following were negative or within normal limits:
24-hour urine 5-hydroxyindoleacetic acid, remainder of the complete blood count and comprehensive metabolic panel, zinc, niacin, TSH

Diagnostic Procedures and Tests:
Computed Tomography, Chest, Abdomen, and Pelvis: Numerous hepatic lesions, some small, some with peripheral enhancement, and some with more central necrosis. The tail of the pancreas has a bulky globular appearance and is suspicious for a primary lesion. The head and body of the pancreas are atrophic. No intrathoracic or pelvic lesions are noted.

Histopathology:
Right lateral thigh, skin: There is a confluent parakeratosis of the stratum corneum, pallor of the upper third of the epidermis, intracorneal clefting, neutrophilic spongiosis, and rare dyskeratotic cells. There is a superficial and a mid-dermal perivascular lymphohistiocytic infiltrate.

Diagnosis:
Necrolytic migratory erythema secondary to a metastatic pancreatic neuroendocrine tumor

Treatment and Course:
The patient was initially given clobetasol 0.05% ointment and nicotinamide 250 mg PO daily. She continued with octreotide injections 30 mg intramuscularly every month. Some improvement was observed on this regimen. At her two-month follow-up, the patient developed new lesions, on the legs, buttocks, palms, and periorally. Doxycycline 100 mg twice daily for 2 weeks was prescribed due to concern for secondary impetiginization. During this treatment regimen, the patient was found to have complete clearance of skin lesions. However, upon completion of her antibiotic regimen, she developed a significant flare. The patient was restarted on doxycycline 100 mg twice daily and has maintained good control of her skin disease. During this time, her metastatic pancreatic neuroendocrine tumor was being treated with octreotide injections. Everloimus was subsequently added, but then switched to sunitinib secondary to side effects. Her most recent glucagon level was 1,680 ng/L.

Discussion:
Becker *et al.* first described necrolytic migratory erythema (NME) in 1942 as being part of a triad of symptoms including NME, stomatitis, and weight loss, in association with a glucagon-secreting alpha-cell pancreatic tumor. This triad is known as glucagonoma syndrome. Since 1942, NME has also been described in the presence of an extra-pancreatic glucagon-secreting tumor, or pseudoglucagonoma syndrome, and in the absence of glucagonemia, such as in conditions with intestinal malabsorption issues, cirrhosis, and nutritional deficiencies including zinc, essential amino acids, and fatty acids.

Clinically, NME is considered the hallmark of the glucagonoma and pseudoglucagonoma syndromes. Therefore, prompt recognition is essential to favorable patient outcomes. Typically, this skin eruption arises as a painful and pruritic bullous dermatosis. Lesions consist of

erythematous patches and plaques with irregular borders, crusting, erosions, and scaling. Central clearing may give an annular appearance. Eruptions can be generalized, but often are localized to the perioral region, lower abdomen, groin, distal extremities, and intertriginous areas. Koebnerization is also a common finding.

The pathogenesis of NME is likely multifactorial. Hyperglucagonemia is the primary contributing factor as removal of the glucagon secreting tumor or control of glucagon levels with medication results in resolution of cutaneous disease. Elevated glucagon levels cause hypoaminoacidemia and elevated levels of arachidonic acid within the epidermis, resulting in inflammation, epidermal necrolysis, and development of NME. Malnutrition or deficiency of zinc, amino acids, and essential fatty acids are also thought to contribute to the disease process, with several case reports demonstrating resolution of cutaneous disease through supplementation of these nutritional deficiencies.

Histology often shows a nonspecific subacute dermatitis. Biopsy of an early advancing edge should demonstrate superficial epithelial necrosis. Vacuolated, pale, swollen epidermal cells are also characteristic. Scattered dyskeratotic cells within the epidermis may also provide a clue for diagnosis. Additionally, moderate hyperkeratosis with slight acanthosis and diffuse parakeratosis is more often the histological pattern observed. Nutritional deficiencies such as pellagra, zinc deficiency, and necrolytic acral erythema may also resemble NME histologically with characteristic pallor and ballooning of the upper epidermis.

Treatment for NME primarily revolves around removing the glucagon-producing tumor, if possible, in addition to reducing glucagon levels through somatostatin analogues (i.e. octreotide). Amino acid supplementation has also been demonstrated in resolution of cutaneous disease in patients with pancreatic glucagon secreting tumors. Unfortunately, by the time the diagnosis is made, many tumors have already metastasized, most often to the liver. Therefore, surgical removal is no longer an option. Thus, alternative treatment options are necessary for these patients. Tumor response to chemotherapy is often weak and requires supplemental treatments. Topical and systemic steroids, ultraviolet light, dapsone, methotrexate, tar preparations, and vitamin supplementation including nicotinamide have shown limited success in existing literature. Octreotide endogenously suppresses glucagon release and often aids in reduction of cutaneous eruptions in some, but not all patients.

Currently, streptozocin is the only antibiotic mentioned in the literature to have been used for patients with glucagonoma syndrome, with only limited therapeutic success. Therefore, our use of doxycycline demonstrates a novel adjunctive therapy for patients with NME. In the case of our patient, doxycycline 100 mg twice daily in addition to monthly octreotide injections and sunitinib achieved significant control of her skin disease. Thus, use of antibiotics with anti-inflammatory properties may be of use in conjunction with octreotide in patients with NME.

Essential Lesson:

- Necrolytic Migratory Erythema (NME) is often the primary manifestation of glucagonoma and pseudoglucagonoma syndrome
- Lesions are often painful and pruritic, localizing to the perioral region, lower abdomen, groin, distal extremities, and intertriginous areas
- Histology may resemble nutritional deficiencies with characteristic pallor and ballooning of the upper epidermis; however, many biopsies may be nonspecific
- Due to its late diagnosis and rarity, few treatment options exist in cases of metastatic glucagonoma or in pseudoglucagonoma syndrome
- Antibiotics with anti-inflammatory properties may be of use for patients with NME

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Case #5

Case Presented by Huayi Zhang, MD,
Michelle Bain, MD and James Feinberg, MD, JD, MPH

Telemedicine has become an important part of health care in the United States. Various technological advancements in the last two decades have made it an invaluable tool in medicine - especially in dermatology where its visual character lends itself well and is rapidly being adopted to satisfy an ever-increasing demand. Teledermatology is regarded as an efficient and cost effective method to deliver care to patients in remote areas with limited access to dermatologists. Currently, there are 30 physician centered teledermatology programs in the US - including the Kaiser Permanente Healthcare System and the Veteran's Affairs Medical Centers. Generally, there are two types of teledermatology consult modalities; store and forward (SAF), in which patient information such as photos and medical histories are sent online as digital files to teledermatology physicians, and live interactive (LI), in which video conference connects the patient and physician from separate locations.

As the consultation volume for teledermatology continues to rise, there is a significant opportunity to include it as an essential part of the resident's training curriculum and practical experience. A survey conducted in 2016 by the Department of Dermatology at the Harvard Medical School found that although 67% of US accredited residency programs utilized teledermatology. Within these programs only 21% of residents reported participation in its use and from these, only 10% had greater than 20 sessions in the three years of training. The survey data revealed a need to expand resident exposure to teledermatology in order to provide further practice based learning and medical knowledge. To better prepare residents for a future in dermatology, programs should consider incorporating teledermatology clinics as part of their standard curricula.

At the Jesse Brown Veterans Affairs Medical Center (JBVA), teledermatology is read by both the University of Illinois (UIC) and Northwestern residents. Each UIC resident participates in four months of teledermatology rotations over the three year training period, with 15 half day sessions each month. The UIC resident manages on average 200 teledermatology consultations every month and reviews all cases with a faculty preceptor who is responsible for all consults. The cases are presented with a brief clinical history of the patient's condition followed by images. The VA medical system provides training sessions for imagers so that the written consultations and clinical photos are standardized. The resident and attending physician review the case and respond by either giving management recommendations to the general practitioner or suggesting face to face follow-up with dermatology. Residents use information sources other than immediate faculty feedback when formulating the differential diagnosis and treatment plans. This contributes to both increased medical knowledge and improvement in practice-based learning. In addition, making a diagnosis based solely on a clinical image simulates the test environment of in-service and board certification examinations, thus better preparing the resident.

The VA does not have medical licensing restrictions across state lines, granting a larger population access to dermatology. Since implementing teledermatology in 2014, the JBVA has received over 7000 teledermatology consults from 10 different states, and continues to increase the referral base.

Generally, the limitations of SAF teledermatology arise from the lack of the patient-provider relationship, the inability to perform hands-on examinations, and other elements such as

performing bedside diagnostic testing, and asking follow-up questions. Thus, the SAF modality cannot handle emergency cases or blistering diseases, and is best suited for lesional dermatology. Additionally, clinicians in remote locations have a limited set of treatment options to choose from. At the VA, we take into consideration these limitations. For example, rural healthcare clinic often cannot provide photodynamic therapy or narrowband ultraviolet B treatment rooms necessitating alternative recommendations. About 30% of teledermatology patients are then further referred for face-to-face consultation with a dermatologist. As residents become familiar with these limitations, they develop competency and provide strategic treatment options.

Patient satisfaction is paramount to the successful implementation of the teledermatology program. A systematic review published in 2017 showed that from 2000-2016, a high level of patient and provider satisfaction was observed in both the SAF and LI teledermatology modalities. A key driver for this was turnaround time. On average, it can take two to six months for a new patient to see a dermatologist, whereas it may take as little as two weeks to have a diagnosis made via teledermatology consultation. Patients appreciated the swift response to their skin care concerns in addition to avoiding long distance travel to visit dermatology clinics. Physicians enjoyed the change in pace of the work and the added variety to their daily routines.

Additionally, most studies evaluating the economics of teledermatology have concluded that the SAF method is cost effective when used as a triage mechanism to reduce face-to-face appointment requirements. This is especially true when patients are required to travel large distances to access dermatologic services. The quality of patient care is likely not compromised by this treatment modality. Recent literature has consistently demonstrated a comparable accuracy of teledermatology to in person visits for certain skin conditions including actinic keratosis and non-melanoma skin cancers.

Teledermatology serves as an effective medical diagnostic tool that embraces new technology to enhance resident learning while providing quality healthcare.

We provide overviews of three actual teledermatology consultations.

Case 1:

A 65 year old Caucasian male presented with an enlarged blue to purple 1.5 cm nodule on the vertex of the scalp. The lesion had been present for over 10 years; however, it began to grow after the patient underwent radiation therapy to treat his thyroid cancer. The nodule is occasionally tender to touch.

Case 2:

A 31 year old Caucasian male presented with several annular erythematous patches on his chest and back. He explained that the rash appeared shortly after returning from a vacation in Iceland. He denied any symptoms associated with the lesions in addition to using new fragrances, body wash, or laundry detergents. He did complain of mild fatigue.

Case 3:

A 57 year old Caucasian female presented with skin colored nodules on the plantar surfaces of both feet for several years duration. She complains of pain associated with the lesions especially after walking for long distances. Patient explained her mother had a similar condition.

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Case #6

Case Presented by Benjamin Garden, MD
Lawrence Chan, MD and Sophie Worobec, MD

FAST BREAK

A 47 year old male with no significant past medical history presented with two lesions on his right hand that developed over the last 3 months.

Case #7

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

History of Present Illness:
A 41 year old female presented for treatment of red, scaly papules on her skin for over ten years. The lesions were generally asymptomatic. In the past ten years, she had seen multiple physicians and tried many treatments. Notably, she used topical 5-fluorouracil with minimal improvement, cryotherapy with unfavorable cosmetic outcomes, and a variety of other topical therapies which she could not recall. She presented for further management given the extensive distribution of her lesions.

Past Medical and Surgical History:
Diabetes mellitus

Medications:
Metformin

Allergies:
None

Family History:
The patient denies any family history of skin cancer, or other similar lesions

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

Physical Examination:
The anterior legs, thighs, lateral forearms, and upper central chest are symmetrically covered with many red-brown, slightly scaly papules, some annular in shape, ranging from 3 mm to 8 mm. The larger lesions display central atrophy and a well-demarcated border featuring a thin, elevated and furrowed keratotic rim.

Histopathology:
Right leg, skin: At low power, the specimen shows a psoriasiform epidermis with an invagination of the skin at the lateral border. The dermis appears largely unremarkable. At higher power, one appreciates a thin column of tightly packed parakeratotic cells that extends from the invaginated epidermis. The parakeratotic cells appear to be arranged in a 45° angle relative to the underlying epidermis composing a cornoid lamella. There are dyskeratotic keratinocytes underneath the cornoid lamella.

Diagnosis:
Disseminated superficial actinic porokeratosis

Treatment and Course:
After confirming the diagnosis of disseminated superficial actinic porokeratosis, the patient was started on topical calcipotriene 0.005% cream, twice daily, for 1 month to the affected areas. After treatment, the clinical observation revealed only minimal improvement.

Discussion:

Porokeratosis is a chronic disorder characterized by hyperkeratotic papules and plaques surrounded by an elevated border. It is believed to be a disorder of keratinization, but the exact pathogenesis is unclear. A leading hypothesis is that porokeratosis represents an expanding mutant clone of keratinocytes. There are several common forms of porokeratosis, including porokeratosis of Mibelli, linear porokeratosis, disseminated superficial actinic porokeratosis, porokeratosis palmaris et plantaris disseminata, and punctate porokeratosis.

Disseminated superficial actinic porokeratosis (DSAP) is the most common form of porokeratosis, often appearing on the sun-exposed skin of affected individuals, and traditionally spares the palms and soles. Some authors make a distinction between disseminated superficial *actinic* porokeratosis and disseminated superficial porokeratosis (DSP), although this is not widely accepted. The distinction between DSAP and DSP is based on clinical history and age of onset, as DSAP typically appears in the third or fourth decade, whereas DSP usually appears in the first decade and can be found on non-sun-exposed skin, such the groin. Though ultraviolet light exposure and immunosuppression are often cited as eliciting stimuli of DSAP, our patient denied these triggers. The development of squamous cell carcinoma has been described in lesions of porokeratosis, but the DSAP variant is reported to have the lowest risk of malignant transformation.

The cosmetic appearance of lesions in DSAP, rather than symptom manifestation or systemic risk, is the primary impetus toward treatment. Thus, risks and benefits must be carefully weighed when choosing a therapeutic regimen. Traditionally, the disorder is difficult to treat and multiple treatment modalities have been proposed, such as topical and systemic retinoids, topical vitamin D analogs, cryotherapy, photodynamic therapy, 5-fluorouracil, and others. The latest review of treatments suggests that topical vitamin D analogs, such as calcipotriene, may be an appropriate first line therapy for treatment of the disease given their efficacy and relatively benign safety profile. It is believed that vitamin D analogs may help modulate inflammation, as well as keratinocyte differentiation and proliferation in DSAP, although the exact mechanism is unclear. We present this case to highlight the extensive distribution of DSAP in a patient and to discuss treatment recommendations for a historically challenging condition to manage.

Essential Lesson:

- Disseminated superficial actinic porokeratosis (DSAP) is the most common type of porokeratosis
- Treatment of DSAP is often difficult and without consistent outcomes
- Topical Vitamin D analogs, such as calcipotriene, are a safe, effective first-line treatment for DSAP, even in extensive cases

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Case #8

Case Presented by Olivia Lai, MD,
Lawrence Chan, MD, and Maria Tsoukas, MD, PhD

History of Present Illness:
A 37 year old male with myelodysplastic syndrome, undergoing workup for sepsis of unknown etiology, was admitted for evaluation of painful nodules and fever. Dermatology was consulted for the evaluation of subcutaneous tender nodules, which appeared over his lower extremities and abdomen.

Past Medical History:
Myelodysplastic syndrome previously treated with intravenous azacytidine 43 days prior to the day of dermatology consult, diffuse pain syndrome, and hypertension

Medications:
Amoxicillin-clavulanate, sulfamethoxazole-trimethoprim, and paroxetine

Allergies:
Intravenous contrast dye, ibuprofen, lisinopril, tramadol

Family History:
No family history of skin disease or malignancy

Review of Systems:
The patient's review of systems was positive for fevers, malaise, weight loss, swelling and pain in the gluteal area, and hematuria. He denied arthralgias, arthritis, acute vision changes, cough, hemoptysis, bloody stool, nausea/vomiting, diarrhea, constipation, or dysuria.

Physical Examination:
On the abdomen and bilateral thighs, there are multiple firm, tender, subcutaneous, flat-topped nodules with sharp edges measuring 3-6cm in diameter and overlying faint hyperpigmentation. Rare hyperpigmented excoriated papules are noted on the arms and hands.

Laboratory Data:
The following were positive or abnormal:
Hemoglobin 5.7 (12.5-17)
Platelets 58 (140-415)
Creatinine 1.92 (0.6-1.3)
Total bilirubin 3.1 (0.3-1.2)
Urinalysis 30 protein (negative)
Urinalysis small amount of blood (negative)
CBC Differential Neutrophil 81.0% (50.0-70.0%)

The following were negative or within normal limits:
White blood cell count 6.8 (4.8-10.8)
Aspartate Aminotransferase 33 (5-40)
Alanine Aminotransferase 30 (7-56)

Diagnostic Procedures and Tests:
Ultrasound, Soft Tissue Abdomen: Ill-defined hyperechoic area in the subcutaneous tissue of the left upper quadrant anterior abdominal wall in the subchondral region and another one in the anterior abdominal wall subcutaneous tissue superior to the umbilicus are noted. This may suggest a thickening of the normal subcutaneous tissue or lipoma.

Histopathology:
Excisional biopsies from lesional skin of the left thigh and left subcostal region revealed an unremarkable epidermis and dermis; subcutaneous tissue with a neutrophilic panniculitis, with findings most consistent with subcutaneous Sweet's Syndrome associated with myelodysplastic cells. Special stains were negative for acid fast bacilli, fungi, and bacteria

Diagnosis:
Subcutaneous Sweet's Syndrome

Treatment and Course:
Shortly after biopsy, the patient was started on 60mg daily of prednisone. This dose was decreased to 40mg daily several weeks later and slowly tapered off. Colchicine 1.2mg daily was started several days after biopsy, but the patient did not tolerate the medication well and the medication was discontinued 4 days later due to signs of acute kidney injury. After a total of 33 days of treatment with prednisone, the patient's fevers and skin findings resolved completely.

Discussion:
Sweet's Syndrome (SS), also known as acute febrile neutrophilic dermatosis, is an inflammatory disorder that is typically characterized by the sudden onset of painful nodules, plaques, and papules of the skin. Often, patients with SS also will present with fever and leukocytosis. The most common lab abnormality found in patients with Sweet's syndrome is peripheral leukocytosis with neutrophilia. Three subtypes of SS have been proposed based on etiology: classic SS, malignancy-associated SS, and drug-induced SS. Additionally, SS can have atypical presentations. For instance, bullous SS, neutrophilic dermatosis of the dorsal hands, and subcutaneous SS (as in the case of our patient) are all possible manifestations. In subcutaneous SS, neutrophilic infiltration occurs in the subcutaneous fat rather than the dermis.

Subcutaneous SS has been reported in the setting of myeloid disorders. A case series and literature review of subcutaneous SS in patients with myeloid disorders showed that patients typically presented with erythematous tender nodules that usually involved the extremities, accompanied by fevers, and symptoms typically resolved either spontaneously or after systemic steroid treatment. The patient presented today similarly had myelodysplastic syndrome, and his symptoms resolved after systemic steroid treatment.

Lastly, certain medications have been associated with the development of SS. Case reports have shown that azacytidine, a chemical analog of cytidine used in the treatment of myelodysplastic syndrome, is one of these medications. Although the pathogenesis of SS is not fully understood, hypersensitivity reactions, cytokine dysregulation, and genetic susceptibility have all been proposed as contributing factors. It is possible that certain patients may experience hypersensitivity reactions to azacytidine. Additionally, patients with myelodysplastic syndrome have a dysregulated proinflammatory cytokine landscape in their bone marrow. It has been shown that administration of azacytidine to myelodysplastic syndrome patients causes further increases in interleukin 8, interleukin 27 and the chemokine monocyte chemoattractant protein-1, therefore leading to further dysregulation. This may in turn contribute to the development of SS in patients with myelodysplastic syndrome who are given azacytidine.

Although the etiology of SS in our case cannot be proven, the use of azacytidine may have been an inciting factor in the development of his SS. We present this case to highlight a rare presentation of SS.

Essential Lesson:

- Sweet's Syndrome (SS), also known as acute febrile neutrophilic dermatosis, is an inflammatory disorder typically characterized by painful nodules, plaques, and papules of the skin.
- Bullous SS, neutrophilic dermatosis of the dorsal hands, and subcutaneous SS (as in the case of our patient) are all possible atypical manifestations.
- Azacytidine is an additional medication linked to development of SS in certain specific patient populations.

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Case #9

Case Presented by Michael Sotiriou, MD
James Feinberg, MD, JD, MPH and Milena Lyon, MD

UNKNOWN

A 57 year old male presented with blisters on his foot and ankle.

Case #10

Case Presented by Regina O'Brien, MD
Iris Aronson, MD and Maria Tsoukas, MD, PhD

History of Present Illness:
A 45 year old female with no significant past medical history presented with ongoing hair loss for the past 6 months. Initially, the hair loss began as a small area on the posterior scalp that slowly enlarged. Over the next several weeks, she noted that the scalp became red, lost pigment, and developed blisters at the alopecic site. She denied pain but stated that the affected area was occasionally itchy. She denied a history of scalp trauma. The patient was started on oral griseofulvin and ketoconazole shampoo for suspected tinea capitis by her physician prior to presenting to our clinic. After one month of treatment, she had noted no improvement. The patient underwent two punch biopsies of the scalp for histopathologic evaluation and direct immunofluorescence (DIF).

Past Medical History:
Essential Hypertension

Medications:
Amlodipine, ibuprofen, and polyethylene glycol

Allergies:
No known drug allergies

Social History:
Patient works as a lunch attendant and lives in a house with her two children. She is a current smoker with a 3 pack-year history. She drinks alcohol socially and does not use illicit drugs.

Review of Systems:
Review of systems was otherwise negative.

Physical Examination:
The crown of the patient's scalp shows two depigmented, atrophic 2-8cm pink plaques with extensive lesional and peri-lesional alopecia. Within the plaques are islands of pigment retention. There are peripheral erosions with overlying serosanguinous crusting noted as well.

Laboratory Data:
The following were positive or abnormal:
Antinuclear antibody 1:40 (<1:40)
RNP IgG 43 (0-40)

The following were negative or within normal limits:
Anti-dsDNA, Smith, SSA, SSB, ELISA IgG to BP230.

Histopathology:
Crown of Scalp, skin: Hematoxylin and eosin (H&E) staining showed scarring alopecia, with sections of skin showing parakeratosis and acanthosis. Within the dermis, there were collections of a dense inflammatory cell infiltrate composed of plasma cells, neutrophils, scattered eosinophils, and lymphocytes. The infiltrate was mainly present around the follicular infundibulum. Elastic fiber stain revealed the disrupted elastic fiber network around the hair follicles. A periodic acid-Schiff (PAS) stain was negative for fungi.

Immunopathology Tests:
Direct immunofluorescence (DIF): Linear deposition of IgG, IgA, and C3 at the basement membrane zone, with weaker IgM deposition and predominance of IgG over IgA.

Indirect immunofluorescence (IIF): Positive cell surface staining of IgG2 and IgG3

Enzyme-linked immunosorbent assay (ELISA) IgG to BP180 43 (<10)

Diagnosis:
Brunsting-Perry pemphigoid

Treatment and Course:
The patient was started on doxycycline 100mg PO twice daily and topical fluocinonide 0.05% solution twice daily. On follow-up a few weeks later, the patient noted that her erosions had healed, and the itching had resolved. On further follow-up 5 months later, the patient noted no recurrent episodes of blisters, erosions, or itching.

Discussion:
Brunsting Perry pemphigoid (BPP), first described by Brunsting and Perry in 1957, is a rare variant of mucous membrane pemphigoid (MMP) in which mucosal involvement is rare. It classically presents in 40-70 year old men as recurrent, circumscribed, pruritic blisters localized to the head, neck, and upper trunk. The lesions usually heal with atrophic scarring over months to years. Involvement of the scalp may lead to scarring alopecia.

Diagnosis is made via a combination of clinical findings, histopathologic assessment, and direct immunofluorescence (DIF). In 80-100% of cases, the DIF will demonstrate linear IgG and C3 deposition at the basement membrane zone. IgA or IgM may also be seen at the basement membrane zone. Out of 58 reported cases, only 11 have identified target antigens with the most commonly found antigens being BP180 (5 cases) and Type VII collagen (4 cases). Other identified target antigens include Laminin 332 (laminin 5), BP230, Desmoplakin 1 and 2.

Interestingly, the identification of Type VII collagen as a target antigen as well as reports in the literature of epidermolysis bullosa acquisita (EBA) localized to the face with a classic clinical BPP presentation have caused some authors to call into question the classification of BPP as a variant of MMP. They propose that a significant number of BPP cases may actually represent a localized form of EBA.

Brunsting-Perry pemphigoid generally has a better prognosis than other forms of MMP. Treatment is similar to that of classic MMP, with the aim being to promote healing and minimize scarring. Therapies generally consist of potent intralesional or topical steroids in addition to systemic corticosteroid therapy, dapsone, and immunosuppressive agents such as methotrexate. Excision with full-thickness skin grafts has also be an effective treatment option.

In MMP cases, systemic therapy for mild cases may include a tetracycline antibiotic, such as doxycycline. We demonstrate in our case that doxycycline may also be an effective systemic treatment for BPP.

Essential Lesson:

- BPP is a rare variant of MMP where skin lesions are usually limited to the head and neck and mucosal involvement is minimal or absent. Healing with scarring occurs over months to years, and on the scalp may lead to scarring alopecia.
- BP180, Collagen VII, BP 230, and laminin 332 are thought to be the major targeted autoantigens. However, many BPP cases do not have an identified autoantigen.
- The mainstay of treatment for BPP is intralesional or topical steroids in addition to systemic medications such as corticosteroids, dapsone, or immunosuppressives. Doxycycline, commonly used for MMP, may also be efficacious in its rare variant BPP.

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