Chicago Dermatological Society

October 2020 - *Online* Educational Conference

Program & Speaker Information
CME Certification
Case Presentations

Wednesday, October 14, 2020
Online

UI Health | UIC

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

Program

Host: University of Illinois at Chicago
Wednesday, October 14, 2020
Online Conference

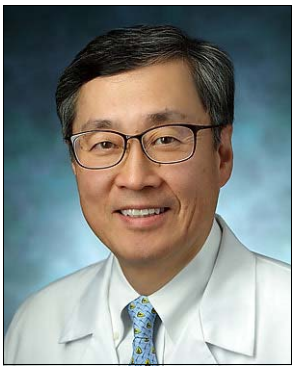
8:30 a.m.	**Sign-in and Member Visitation Time**
9:00 a.m.	**Welcome & Introduction** *David Mann, MD - CDS President*
9:05 a.m. - 9:30 a.m.	**Guest Lecture - Part I** "Understanding Photoaging Pathophysiology: Part 1 - Wrinkles" *Sewon Kang, MD*
9:30 a.m. - 9:40 a.m.	**Questions & Answers**
9:40 a.m. - 10:15 a.m.	**Resident Case Presentations & Discussion** *UIC Residents*
10:15 a.m. - 10:40 a.m.	**Guest Lecture - Part II** "Understanding Photoaging Pathophysiology: Part 2 - Lentigines" *Sewon Kang, MD*
10:40 a.m. - 10:45 a.m.	**Questions & Answers**
10:45 a.m. - 10:50 a.m.	**Closing Remarks and Introduction of Discussion Breakout Rooms** *David Mann, MD*
11:00 a.m. - 12:00 p.m.	**Breakout Rooms*** • Medical Students • Case Discussions • Practice Challenges • Residents Forum
12:00 p.m.	**Meeting adjourns**

* Four breakout sessions will commence at the conclusion of the second guest lecture. They are scheduled for approximately one hour and are intended to be open discussion with a moderator to facilitate the conversation. Meeting attendees were asked to indicate their choice of breakout session when first registering for the Zoom meeting.

Mark the Date!

Next CDS virtual meeting will be on Wednesday, November 11th – Co-hosted by Northwestern
Watch for details on the CDS website: www.ChicagoDerm.org

Guest Speaker



SEWON KANG, MD

Professor and Chair, Department of Dermatology;
Johns Hopkins University
Baltimore, MD

Sewon Kang, MD, is the Noxell Endowed Professor of Dermatology, Dermatologist-in-Chief at Johns Hopkins Hospital and chair of the Department of Dermatology.

A recipient of the Dermatology Foundation's Career Development Award, Dr. Kang has received other research awards and grants from the American Dermatological Association, the National Psoriasis Foundation and the National Institutes of Health. His research focus has been in the areas of skin pharmacology and photomedicine, and he is the past president of the Photomedicine Society and the American Acne and Rosacea Society. Dr. Kang is or has been on the boards of multiple organizations including the Society for Investigative Dermatology, the Association of Professors of Dermatology, the Dermatology Foundation, and the Skin of Color Society. An author on more than 240 publications and book chapters, Dr. Kang also is the Editor-in-Chief of the 9th edition of the Fitzpatrick's Dermatology texbook. He is an inventor/co-inventor of 17 patents and has given more than 300 presentations globally.

Dr. Kang earned his medical degree at the University of Michigan Medical School in 1987. He completed his residency in dermatology in 1992 at Massachusetts General Hospital, Boston.

CME Information

October 14, 2020

Overview

The Chicago Dermatological Society was established in 1901 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. Two lectures are given by the guest speaker, and the residents of the host institution present cases which are offered for audience discussion. During the coronavirus pandemic, CDS has continued to organize our regular educational conferences, but in a half-day "virtual" online live setting.

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 2019/20 series of meetings, the participant should be able to:
1. Discuss the causes, results and the pathophysiology of photoaging.
2. Describe how UV light and exposure to the sun impact upon the development of wrinkles and lentigines.
3. Discuss the pathogenesis, prevention, and treatment for photoaging, especially wrinkles and lentigines.

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 2 *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 an online CME claim form after the completion of the conference. A link to this form along with the online evaluation form will be sent to each conference attendee after the meeting. Thank you for your attention to this important item.

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. None of the participants in this conference have disclosed relevant potential conflicts of interest.

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

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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.

Resident
Case
Presentations

University of Illinois at Chicago
Department of Dermatology

Table of Contents

Case #1

Case Presented by Isabelle Sanchez, MD

Elizabeth Kream, MD; Jacob Charny, MD;

Marylee Braniecki, MD; Maria Tsoukas, MD, PhD; and Iris Aronson, MD

Case:

A 25-year-old white female attorney with no pertinent past medical history presented to our clinic complaining of a recurrent facial rash following exposure to sunlight. The first occurrence was 5 years ago when the patient was vacationing to a beach. She described the rash as tender blotchy redness and swelling starting 1-2 days after sun exposure with crops of pustules erupting on the cheeks, nose, and forehead by day 3 after exposure. The pustules begin to crust and exfoliate with clearance of all lesions by 1 week. Upon clearance, the patient denied any dyspigmentation or scarring. This cycle occurs after every prolonged exposure to sunlight.

The patient denied a history of acne or rosacea. Prior to presentation, she had been treated with doxycycline, metronidazole cream, and clindamycin lotion with no improvement. The outbreaks were not prevented with sunscreen use. Labs were notable for an indeterminate pemphigus panel, positive HSV-1 antibody (IgG), and negative comprehensive ANA panel. She was started on valacyclovir, but continued to experience flares. Punch biopsy showed an intraepidermal bulla containing neutrophils and involving the hair follicle. Direct immunofluorescence was non-specific. Based on clinicopathology correlation, a diagnosis of actinic folliculitis was made.

Treatment and Course:

The patient was started on topical adapalene 0.1% gel and strict photoprotection however she still continued to experience flares. Additional treatment options will be discussed with patient including high potency topical retinoid therapy or isotretinoin.

Discussion:

Actinic folliculitis (AF) is a rarely reported photodermatosis, first described in 1985 to encompass both acne aestivalis and actinic superficial folliculitis. Since 1985 only 6 cases have been reported. AF is most prevalent in white females and is characterized by a monomorphic pustular non pruritic eruption on the face and chest 6-36 hours after intense sun exposure, with complete resolution in 7-10 days and without scarring. Importantly, comedones are absent and there is no improvement with oral or topical antibiotics. Pathogenesis is not well understood, but likely involves ultraviolet A radiation induced reaction around the hair follicle infundibulum, probably secondary to keratinocytes & Langerhans cells involvment in the immunomodulary actions to UV radiation. Effective treatment involves both sun protection along with topical or oral retinoids. It is thought that adapalene may be the best topical retinoid for this condition given that it is photo-stable. For cases refractory to topical adapalene, systemic retinoids can be tried. There have been very few reported cases of actinic folliculitis making evidence based treatment difficult. We present this case for clinical interest.

References:

1. Bolognia JL, Schaffer JV, Cerroni L. Dermatology, 4th edition. Chapter 38: Folliculitis and other follicular disorders. Philadelphia, Elsevier. 2018.
2. Jaeger C, Hartschuh W, Jappe U. Actinic superficial folliculitis. J Eur Acad Dermatol Venereol 2003;17:562-5.
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Case #2

Case Presented by Christy Waterman, MD
Cameron Trodello, MD; Owen Kramer, MD;
Nathan Jetter, MD; and Michelle Bain, MD

Case:

A 5-week-old male with no pertinent past medical history was referred from pediatrics for a rash on the chest that first appeared at three weeks of age. The rash spread to involve the scalp and abdomen and was asymptomatic. The pregnancy course and birth were uneventful. The patient was in the 10-20th percentile for height and weight. Review of systems was negative for fever or chills. Family history was negative for known cutaneous or autoimmune diseases. Physical exam of the abdomen revealed approximately fifteen light pink to red annular macules and papules. On the temples, forehead, and neck there were many subtle coalescing annular pink macules and papules that flushed red when the patient cried.

The patient's labs were notable for positive ANA with a titer of 1:1280 and a speckled pattern. Anti-Ro/SSA and anti-La/SSB autoantibodies were also present. The mother's labs were notable for similar ANA positivity with a titer of 1:2560 and a speckled pattern; anti-Ro/SSA and anti-La/SSB autoantibodies were also present. A biopsy was not performed. Based on clinical and laboratory findings, a diagnosis of neonatal lupus erythematosus was made.

Treatment and Course:

Additional workup of the patient with an EKG was unremarkable. A complete blood count was unremarkable. Liver function testing revealed mild transaminitis, which normalized several weeks later. Without any treatments, the patient's rash resolved about six weeks after appearance and has not returned after one year. The mother felt well, denying rash, photosensitivity, dry mouth or eyes, fatigue, or Raynaud's phenomenon during the patient's visits. She did have occasional joint pain while carrying packages at work.

Discussion:

Neonatal lupus erythematosus (NLE) is a rare autoimmune disease that is characterized by the development of a typical rash or heart block in the neonate of a mother with anti-Ro/SSA and/or anti-La/SSB antibodies. Cardiac NLE occurs in 2% of births of mothers with anti-Ro/SSA and/or anti-La/SSB antibodies, while cutaneous NLE occurs in 4% to 16% of births of affected mothers. Pathogenesis involves transplacental passage of maternal SSA and/or SSB antibodies to the fetus. The typical rash of NLE is a papular, annular eruption affecting the scalp, face, or chest of a newborn that can be present at birth or develop several weeks thereafter, usually resolving within five to six months of appearance. Heart block is often present at birth but may develop afterwards. NLE occurrence is independent of maternal disease; oftentimes asymptomatic women find they are SSA/SSB positive only when their child develops NLE. Fifty percent of asymptomatic mothers will subsequently develop an autoimmune process, typically Sjögren's syndrome. An EKG should be performed on all neonates born to mothers with anti-Ro/SSA and/or anti-La/SSB antibodies to evaluate for heart block. In women with these antibodies who also gave birth to a child with cardiac NLE, preemptive treatment with hydroxychloroquine during future pregnancies is recommended. We present this case for clinical interest.

References:

1. Bolognia JL, Schaffer JV, Cerroni L. Dermatology, 4th edition. Chapter 41: Lupus Erythematosus. Philadelphia, Elsevier. 2018.
2. Brucato A, Cimaz R, Caporali R, Ramoni V, Buyon J. Pregnancy outcomes in patients with autoimmune diseases and anti-Ro/SSA antibodies. Clin Rev Allergy Immunol. 2011;40(1):27.
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Case #3

Case Presented by Priyanka Patel MD

Taryn Murray, MD; Thomas Klis, MD, Payal Patel, MD;

Marylee Braniecki, MD; Kyle Amber, MD

Case:

A 20-year-old white female with history of herpes labialis presented with a 7-month history of a recurrent pruritic, blistering rash predominantly on the bilateral lower extremities. The first lesion presented as a small blister on the right knee, which subsequently progressed to numerous blisters on the medial thighs and lower legs. The patient had previously been treated with topical corticosteroids, mupirocin, doxycycline, and valacyclovir without improvement. Treatment with 10-40 mg of prednisone daily resulted in improvement, but upon discontinuation of prednisone the blisters recurred at the original sites. There was no mucosal involvement. Biopsies were previously performed for hematoxylin and eosin (H&E) and direct immunofluorescence (DIF). H&E demonstrated a sub-epidermal bulla with a mixed eosinophilic and neutrophilic infiltrate, without interface changes. Both initial and repeat DIF were negative.

On presentation, the history and morphology of the blisters were suspicious for linear IgA bullous dermatosis (LABD). Indirect immunofluorescence (IIF) on monkey esophagus demonstrated intercellular IgG staining with a titer of 1:160, negative IgA intercellular staining, and negative IgG and IgA reactivity to the basement membrane zone (BMZ). Enzyme-linked immunosorbent assay (ELISA) revealed the presence of IgG autoantibodies to desmoglein 3 (27 units/mL; Ref <9 units) and BP180 (24 units/mL; Ref <9 units).

The combination of intercellular and BMZ autoantibodies raised concern for paraneoplastic autoimmune multiorgan syndrome (PAMS). Repeat IIF on rat bladder revealed IgG cell surface reactivity with a titer of 1:40. A CT scan of the chest, abdomen and pelvis was negative for malignancy.

Another biopsy was obtained for repeat DIF, which revealed linear deposition of IgA along the dermo-epidermal junction. Based on clinicopathologic correlation, a diagnosis of LABD was made.

Treatment and Course:

The patient was started on 50 mg of dapsone daily. Within 1 week of initiating dapsone her blisters resolved. The patient is presently well controlled on this regimen.

Discussion:

Linear IgA bullous dermatosis (LABD) is a rare dermatosis characterized by IgA deposition on various antigens along the basement membrane zone. Due to the variable antigen subset, the condition frequently demonstrates a phenomenon known as epitope spreading. This phenomenon is a result of the initial autoimmune insult damaging tissue and exposing previously sequestered antigens, leading to B or T cell response diversification from the initial epitope to others. DIF is the diagnostic gold standard for autoimmune blistering disease. In our patient, serologic findings were concerning for PAMS. DIF, however, did not support this diagnosis; rather, it was consistent with LABD. This case emphasizes the importance of repeating DIF to increase diagnostic sensitivity when there is strong suspicion for an autoimmune blistering dermatosis.

References:

1. Allen J, Wojnarowska F. Linear IgA disease: the IgA and IgG response to the epidermal antigens demonstrates that intermolecular epitope spreading is associated with IgA rather than IgG antibodies, and is more common in adults. Br J Dermatol. 2003 Nov;149(5):977–85.
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Case #4

Case Presented by Stephanie Kuschel, MD;

Nathan Jetter, MD; Krishna Patel, MD;

Marylee Braniecki, MD; Michelle Bain, MD

Case:

A 2-month old otherwise healthy female presented with her mother for persistent and increasing number of red-brown skin lesions. The first lesion was noticed at 1 day of age on the left mid-abdomen. Additional red-brown macules, patches, and occasional firm papules were scattered across the face, posterior auricular, abdomen, back, buttocks, and extremities. Darier sign was negative. Physical exam was otherwise normal with no lymphadenopathy or hepatosplenomegaly. The patient's mother had been applying topical triamcinolone 0.1% ointment to affected areas twice daily for 2 weeks without improvement. Biopsy of a papule on the left mid-back showed a diffuse dermal infiltrate of monomorphic cells with scattered eosinophils. CD117, CD68, and CD163 staining were positive. CD1a and S100 staining were negative. Laboratory analysis including CBC, CMP, CRP, ferritin, lactate dehydrogenase, and coagulation studies were within normal limits. Serum tryptase was also within normal limits at 6.4 mg/L (Reference range: ≤10.9). Genetic testing of the biopsy specimen revealed a KIT p.D816V mutation. Based on the clinical presentation and histopathologic data, a leading diagnosis of a mast cell proliferative disorder with histiocytic features was made.

Treatment and Course:

Due to concern for systemic involvement based on the polymorphic nature of the lesions and positive activating KIT mutation, the patient was referred to hematology-oncology, and from there to the Mastocytosis Center at Brigham and Women's Hospital. Expert assessment as well as pathology consultation at this institution supported a diagnosis of polymorphic maculopapular cutaneous mastocytosis, also known as urticaria pigmentosa. At the recommendation of the mastocytosis clinic, an abdominal ultrasound to evaluate for organomegaly was ordered (and pending at the time of this writing). Additional imaging studies and bone marrow biopsy are deferred at this time due to low concern for systemic disease. The patient will continue to follow in the Brigham and Women's mastocytosis clinic every 2-3 months with serial lab monitoring of serum tryptase level.

Discussion:

Mastocytosis is an umbrella term for a group of disorders characterized by abnormal accumulation of mast cells in the tissues. There are two main subtypes of mastocytosis: cutaneous mastocytosis (CM), or skin-limited disease, and systemic mastocytosis (SM) with multi-organ involvement (including bone marrow, liver, spleen, and/or lymph nodes) with or without skin involvement. It is helpful to stratify mastocytosis by age of onset as clinical presentation and prognosis significantly differs in pediatric-onset and adult-onset populations. Pediatric mastocytosis takes the cutaneous form of disease in about 80% of cases. The majority of cases present prior to the age of two and regress prior to the onset of puberty. In contrast, most adults have the systemic form, which may progress over time. There are three main subtypes of pediatric cutaneous mastocytosis. The most common form is maculopapular cutaneous

mastocytosis, representing 70-90% of cases. The other two subtypes of CM include solitary mastocytomas (10-35%) and diffuse cutaneous mastocytosis (1-3%). The pathogenesis of mastocytosis is poorly understood but gain-of-function mutations in *KIT* have been implicated. Mutations in *KIT* are believed to induce ligand-independent activation, promoting dysregulated mast cell development. Most adult patients with SM have mutations in KIT on exon 17, especially mutations in D816V. Detection of activating mutations at codon 816 of KIT in extracutaneous tissues is one of the World Health Organization's diagnostic minor criteria for SM. Consequently, D816V mutations have both diagnostic and prognostic implications in adults. Many children with CM also have KIT D816V mutations (approximately 34%); however, the prognostic implications have not been well-established. Several studies have failed to correlate D816V mutations with clinical outcomes in children. However, one study found that approximately 96% of children with systemic disease had mutations at codon 816. Available studies suggest most pediatric patients with mastocytosis clinically regress or stabilize (67% and 27%, respectively). Rarely CM may progress (3 %), more commonly in the maculopapular mastocytosis subtype. Progression is characterized by worsening cutaneous symptoms including flushing and itching as well as systemic involvement. Fatalities have been reported (3%) in aggressive forms of systemic disease including mast cell sarcoma and mast cell leukemia. Treatment of cutaneous disease is primarily symptomatic control and monitoring for development of systemic involvement. Due to the belief that most children will regress, non-invasive serial monitoring of serum tryptase, CBC, and CMP has been favored. Consequently, bone marrow biopsy is recommended only if multi-organ involvement is suspected as demonstrated by laboratory abnormalities, persistent skin lesions, and/or imaging studies showing evidence of extracutaneous organ involvement.

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