Chicago Dermatological Society

Monthly Educational Conference

Program Information
CME Certification
and
Case Presentations

Wednesday, November 4, 2015
Gleacher Center – Chicago, IL

Conference Host:
Department of Dermatology
Feinberg School of Medicine
Northwestern University
Chicago, Illinois

Program

Host: Northwestern University

Conference Location

Gleacher Conference Center
450 N. Cityfront Plaza Dr., Chicago

All meeting activities take place on the 6th Floor of the Gleacher Center.

8:00 a.m.	**Registration & Continental Breakfast with Exhibitors** *6th Floor Lobby and Room 600*
8:30 a.m. - 10:00 a.m.	**Clinical Rounds** Patient Viewing – *Rooms 602 and 604* Posters – *North Foyer (available throughout the morning)* Slide viewing – *Room 608 (available throughout the morning)*
9:00 a.m. - 10:00 a.m.	**Resident/Basic Science Lecture** – *Room 621* "Immunotherapeutic Strategies in CL" *Youn H. Kim, MD*
10:00 a.m. - 10:30 a.m.	**Break and Visit with Exhibitors** – *Room 600*
10:30 a.m. - 12:00 p.m.	**Resident Case Presentations & Discussion** *Room 621*
12:00 p.m. - 12:15 p.m.	**MOC Self-Assessment Questions** *Room 621*
12:15 p.m. - 12:45 p.m.	**Box Lunches & visit with exhibitors** *Room 600*
12:45 p.m. - 1:00 p.m.	**CDS Business Meeting** *Room 621*
1:00 p.m. - 2:00 p.m.	**General Session** – *Room 621* *BLUEFARB LECTURE*: "Therapeutic Advances in CTCL" *Youn H. Kim, MD*
2:00 p.m.	**Meeting adjourns**

Mark the Date!

Next CDS monthly meeting – Wednesday, December 2, 2015 at the Gleacher Center
Hosted by the University of Chicago; Guest speaker: Mai P. Hoang, MD; Department of Pathology, Massachusetts General Hospital

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

Guest Speaker

YOUN H. KIM, MD

The Joanne and Peter Haas, Jr., Professor for Cutaneous Lymphoma Research and Professor, by courtesy, of Medicine (Oncology) at the Stanford University Medical Center Palo Alto, CA

Delivering the Samuel Bluefarb Lecture

Dr. Kim is a Member of Cutaneous T-Cell Lymphoma Medical Advisory Board at Soligenix, Inc. She serves as a Professor for Cutaneous Lymphoma at Stanford University, Director of the Multi-disciplinary Cutaneous Lymphoma Program and Medical Director of the Photopheresis Unit at Stanford Medical Center, and Co-Director of the Lymphoma Research Program at the Stanford Cancer Institute. She is an internationally renowned expert in cutaneous lymphomas and is Co-Director of the Lymphoma Research Program at the Stanford Cancer Institute. Dr. Kim earned her medical degree Stanford University School of Medicine (1984) where she also completed a dermatology residency (1989). She is Board Certified in Dermatology.

CONTINUING MEDICAL EDUCATION CREDITS

Chicago Dermatological Society

Presents

"Chicago Dermatological Society Monthly Meeting Series"

November 4, 2015 *Chicago, IL*

Please be sure to complete and return the attendance/CME claim form and the evaluation form before you leave the meeting. The information collected in the evaluation form represents an important part of the CME planning process. A certificate of credit will be mailed to you following the conference. Participants must attend entire session to receive full credit. SynAptiv will retain a record of attendance on file for six years.

JOINT SPONSOR STATEMENT

This educational activity is jointly provided by SynAptiv and the Chicago Dermatological Society.

GOAL / PURPOSE

To broaden the clinical knowledge of dermatologists in a number of areas relating to management of patients and new therapy options.

EDUCATIONAL OBJECTIVES

Upon completion of this series of activities, participants will 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.

Continued next page

CREDIT STATEMENTS

This activity has been planned and implemented in accordance with the accreditation requirements and policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint providership of SynAptiv and Chicago Dermatological Society. SynAptiv is accredited by the ACCME to provide continuing medical education for physicians.

SynAptiv designates this live activity for a maximum of 4.5 *AMA PRA Category 1 Credits*™. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

DISCLAIMER STATEMENTS

The content, views and opinions presented in this educational activity are those of the authors and do not necessarily reflect those of SynAptiv and Chicago Dermatological Society. The authors have disclosed if there is any discussion of published and/or investigational uses of agents that are not indicated by the FDA in their presentations. Before prescribing any medicine, primary references and full prescribing information should be consulted. Any procedures, medications, or other courses of diagnosis or treatment discussed or suggested in this activity should not be used by clinicians without evaluation of their patient's conditions and possible contraindications on dangers in use, review of any applicable manufacturer's product information, and comparison with recommendations of other authorities. The information presented in this activity is not meant to serve as a guideline for patient management.

DISCLOSURE STATEMENTS

SynAptiv insures balance, independence, objectivity, and scientific rigor in all our educational activities. In accordance with this policy, SynAptiv identifies conflicts of interest with its instructors, planners, content managers, and other individuals who are in a position to control the content of an activity.

The guest speaker, Youn H. Kim, MD, has made available the following disclosure of potential conflicts of interest: Grants/Research Support - Eisai, Innate, Kyowa, Merck, Millennium, Seattle Genetics, Tetralogic; Consulting fees - Actelion, Celgene, Eisai, Galderma, Kyowa, Millennium, miRagen, Seattle Genetics.

No other faculty, planners and/or content managers have conflicts of interest to disclose nor do they have any vested interests or affiliations.

Fee Information - There is no fee for this educational activity.

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.

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CHICAGO DERMATOLOGICAL SOCIETY **Case #1**

Presented by Lara E. Rosenbaum, MD, MHS and Maria Colavincenzo, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
This 47-year-old male, with past medical history notable for hepatitis C (genotype 1B), presented with a 4-month history of rapid hair loss. In April 2014, the patient was initiated on treatment for his hepatitis C with triple therapy (pegylated interferon α-2a, ribavirin and sofosbuvir). The therapy was well-tolerated and his viral load became undetectable. In the last week of his 12-week treatment course, he had rapid complete hair loss including the scalp, eyebrows, eyelashes and body hair. He also noted associated mild pruritus and xerosis.

Of note, the patient had previously undergone unsuccessful treatment of hepatitis C in 1996 with interferon. He recalled having patchy hair loss on his scalp during this therapy but it rapidly improved off treatment.

PAST MEDICAL HISTORY:
Hepatitis C s/p treatment in 1996 and 4/2015-7/2015, now with undetectable viral load
Hypothyroidism – diagnosed 10/2015

MEDICATIONS:
Levothyroxine

PHYSICAL EXAM:
On total body skin exam, the patient was noted to have few 1-2 mm terminal hairs on the occiput but otherwise with total alopecia of the scalp. The follicular ostia were intact and there was no erythema or scale. He had complete loss of his eyebrows and few fine white eyelashes. He was also noted to have few fine terminal brown hairs on the trunk and extremities (which per patient report were far finer and fewer in number compared to previous).

LABS:
Abnormal:
TSH: 5.83 uIU/ml (0.4 – 4.00 uIU/ml)

Normal/Negative:
Free T4: 0.7 ng/dL
Hepatitis C quantitative PCR: <15 IU/ml (not detected)

DIAGNOSIS:
Alopecia universalis in the setting of immunotherapy for hepatitis C

TREATMENT AND COURSE:
In the setting of the patient's recent hepatitis C treatment, possible prior episode of alopecia areata after receiving interferon, and review of the literature, the patient was diagnosed with alopecia universalis precipitated by his recent course of hepatitis C therapy. The patient was quite distressed by his rapid hair loss and resulting change in appearance. Although the patient's viral load was undetectable, systemic immunomodulatory agents were not thought to be in the patient's best interest. He was started on minoxidil 5% foam nightly to the scalp, clobetasol 0.05% cream daily to the scalp, and bimatoprost 0.03% solution daily to the eyebrows and eyelashes. The patient also underwent injection of intralesional triamcinolone 3.3 mg/ml to

the eyebrows on three separate occasions, at least one month apart. Over the course of 10 months, the patient had significant improvement in hair growth.

DISCUSSION:

Alopecia universalis is an uncommon, extensive presentation of alopecia areata that is often challenging to treat. Here we present the case of a patient who developed alopecia universalis after undergoing 12 weeks of triple therapy for hepatitis C.

Hepatitis C is increasingly being treated by hepatologists due to the development of new, effective medications such as sofosbuvir and simeprevir, which were FDA-approved in 2013. These medications are often combined with pegylated interferon α-2a (IFN) and ribavirin (RBV), which were the previous standard of care. It has been recognized that IFN can lead to many autoimmune side effects, including type 1 diabetes mellitus, autoimmune hemolytic anemia and thyroiditis.

One retrospective case series of 152 patients on IFN/RBV revealed that only one patient developed alopecia areata during the study period, which reversed a month after completing treatment. However, there have been several case reports of alopecia totalis or universalis occurring in this setting. There are two case reports of alopecia totalis, both reported as "irreversible" (though follow-up time was not clearly stated). There are seven reported cases of alopecia universalis occurring during hepatitis C treatment. Regrowth in five of these patients occurred between 3-12 months following therapy with IFN (± RBV) and in two patients was "irreversible." Patients with regrowth were treated with topical steroids, oral steroids, PUVA or received no treatment. Of note, four patients who were reported to have alopecia universalis were also newly diagnosed with hypothyroidism or anti-thyroid peroxidase antibodies (anti-TPO), as was the case in our patient.

This case illustrates a rare and distressing side effect of hepatitis C treatment. Dermatologists are uniquely suited to both counsel and treat these patients. Although there are few case reports in the literature, many of these patients seem to improve following completion of therapy.

KEY POINTS:

1. Pegylated interferon α-2a (IFN) is a commonly used medication to treat hepatitis C and carries the risk of autoimmune and dermatologic side effects.
2. IFN may rarely result in alopecia areata, including alopecia totalis or alopecia universalis.
3. Topical and intralesional treatments can be safely offered to patients to enhance hair growth, though many of these patients may spontaneously improve.

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CHICAGO DERMATOLOGICAL SOCIETY **Case #2**

Presented by Ainah Tan, MD, Sapna Amin, MD and Joan Guitart, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
This 65-year-old Jamaican male presented with a 4-month history of pruritic papules and plaques that started on his chest and spread to his extremities, face, and scalp. He was initially diagnosed with seborrheic dermatitis and was given selenium sulfide shampoo. One month later, a biopsy of a lesion on his neck showed atypical epidermotropic T-cell infiltrate with clonal T-cell gene rearrangement, consistent with mycosis fungoides. He was started on narrowband-UVB and topical steroids with subsequent clearance of most lesions. Two months later, he had recurrence of lesions despite the above therapies.

PAST MEDICAL HISTORY:
Hypertension, hyperlipidemia

MEDICATIONS:
Lisinopril-hydrochlorothiazide, atorvastatin, triamcinolone 0.1% ointment to face, clobetasol 0.05% ointment to trunk and extremities

PHYSICAL EXAM:
Erythematous confluent papules on trunk and extremities. More indurated, erythematous papules and plaques on bilateral cheeks, perioral region, and neck. Bilateral soles with hyperpigmented and erythematous scaly plaques. A total of 25% BSA involvement.

LABS/IMAGING:
Abnormal:
Sézary count: 56
Human T-cell lymphotrophic virus type-1 (HTLV-1) IgG: reactive
Bone marrow biopsy and flow cytometry: Immunophenotypically abnormal T-cell population with evidence of TCR gene rearrangement

Normal/negative:
CBC with differential, CMP, SPEP, UPEP, HIV, RPR and CT of the chest, abdomen and pelvis

HISTOPATHOLOGY:
Consistent with adult T-cell leukemia/lymphoma, with infiltrate composed primarily of T-cells (CD3/CD7+) with strong CD25 and FOXP3 expression.

DIAGNOSIS:
HTLV type-1 associated adult T-cell leukemia/lymphoma (ATLL), chronic/smoldering type

TREATMENT AND COURSE:
The patient was started on interferon-alfa-2B TIW and continued on topical steroids and NB-UVB TIW with improvement. Given good treatment response and the fact that patient had to pay out of pocket, the decision was made to withhold zidovudine.

DISCUSSION
Adult T-cell leukemia/lymphoma (ATLL) is a rare peripheral T-lymphocytic malignancy associated with human T-cell lymphotrophic virus type-1 (HTLV-1). HTLV-1 was the first virus implicated in causing malignancy in humans. The virus is endemic in the Caribbean, Japan, South America,

and Africa. ATLL is an emerging public health problem in metropolitan areas in the United States. Although the US age-adjusted incidence is low at 0.4 per 100,000 cases diagnosed annually, the incidence is rising, especially in areas with large Caribbean migrant populations. As a result, clinicians may increasingly encounter patients with ATLL.

ATLL is divided according to the Shimoyama classification into acute, chronic, lymphoma, and smoldering subtypes based on clinical presentation, which also correlate with prognosis and survival. Chronic and smoldering subtypes may have a more indolent and protracted course, whereas the acute and lymphoma subtypes tend to be aggressive with median survival of less than 12 months. Skin manifestations in ATLL are seen in 39-57% of cases and can be the first clinical sign of ATLL. The clinical presentation can vary significantly and often mimic other skin conditions, such as seborrheic dermatitis, eczematous dermatitis, and cutaneous T-cell lymphoma. Similar to mycosis fungoides, type of skin lesions correlate with prognosis; patches and plaques are associated with better survival whereas erythrodermic and nodulotumoral lesions are associated with the poorest prognosis. The varied cutaneous presentation of ATLL makes diagnosis especially challenging. However, recognition of cutaneous findings may lead to earlier diagnosis and treatment.

Definitive diagnosis is usually dependent on serologic and/or molecular markers showing infection with HTLV-1. There are no specific histopathologic or cytogenetic characteristics that aid in diagnosis. Histopathologic patterns vary from an eczematous dermatitis-like to lichenoid to mycosis fungoides-like infiltrate. The infiltrate varies in terms of number of CD8+ and CD4+ cells, and there is variable T-regulatory cell infiltrate (CD25+, FOXP3+) expression depending on subtype of ATLL. Marchetti et al. did a retrospective study of ATLL patients and found that the following were the most common findings leading to clinicians to suspect ATLL in patients presenting with cutaneous findings first: 1) Caribbean origin, 2) unusual clinical presentation of T-cell lymphoma, and 3) HTLV-1 serology work-up of T-cell lymphoma. The authors also suggest that ATLL should be considered when patients have the following features: 1) Caribbean origin or other well-known endemic area (e.g., Japan), 2) clinicopathologic features of mycosis fungoides/Sézary syndrome, 3) nodulotumoral skin lesions appearing early in the disease course.

The best treatment of ATLL is unclear and there are ongoing clinical trials with the Japan Clinical Oncology Group. Treatment options vary from antivirals, chemotherapy regimens, to allogeneic hematopoietic stem cell transplantation.

KEY POINTS

1. HTLV-1 is a virus that is endemic to the Caribbean, Japan, South America, and Africa, and causes hematologic and cutaneous malignancy, more commonly ATLL.
2. ATLL has varied cutaneous manifestations and should be suspected if the patient is of Caribbean or Japanese origin and has an unusual presentation of T-cell lymphoma.
3. Diagnosis of ATLL is dependent on serologic and/or molecular markers showing infection or exposure to HTLV-1.

REFERENCES:

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CHICAGO DERMATOLOGICAL SOCIETY **Case #3**

Presented by Katherine Mercy, MD, Lisa Shen, MD and Sarah Chamlin, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

This is a newborn 31.6 week male with diffuse scarring, atrophic plaques, erosions and nail anomalies.

Unknown

CHICAGO DERMATOLOGICAL SOCIETY **Case #4**

Presented by Sreya Talasila, MD, and Emily Keimig, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
This 73-year-old Caucasian female with a history of psoriasis, type II diabetes mellitus, and breast cancer status-post lumpectomy with radiation presented with a 2-year history of firm brown plaques on the left breast and the left shin that began as enlarging small flesh-colored papules. The lesions were occasionally pruritic. She had been treated with betamethasone valerate 0.1% ointment without improvement. She had also been briefly treated with calcipotriene 0.005% cream. She noted intermittent dysphagia without a history of GERD. She denied a history of Raynaud's phenomenon.

PAST MEDICAL HISTORY
Psoriasis, alopecia totalis, type 2 diabetes mellitus, infiltrating lobular carcinoma of the left breast (ER/PR positive, sentinel lymph node negative)

MEDICATIONS
Amphetamine-dextroamphetamine, metformin, aspirin, exenatide, anastrozole

PHYSICAL EXAM
The left breast was notable for a well-healed surgical scar along the border of the areola. There was contraction and decreased mass of the left breast when compared to the right breast. Superiorly and inferiorly to the areola, there were hyperpigmented, indurated sclerotic plaques with violaceous borders. On the right calf, there was a subtly hyperpigmented, indurated plaque. The left shin had a hyperpigmented, indurated plaque extending onto the calf with a palpable step off.

Labs/Imaging
Mammogram: Heterogeneously dense breast tissue with stable post-treatment changes and benign calcifications. No masses were noted.

Breast Ultrasound: Normal fibroglandular tissue with generalized skin thickening involving the upper inner portion of the left breast. Findings consistent with benign calcifications without mass were noted.

HISTOPATHOLOGY
Surgical pathology was initially interpreted as a dermatofibroma. Further review revealed reticular dermal fibrosis with loss of adipose tissue around eccrine glands and an accompanying lymphoplasmocytic infiltrate. There was no evidence of metastatic carcinoma.

Three skin biopsies from the left leg and the left breast were performed. All specimens were notable for their square-shape. There was marked thickening of the deep dermal collagenous bundles. The dermis showed a deep perivascular and interstitial lymphohistiocytic infiltrate with plasma cells. There was associated mild depletion of the periadnexal fat pad.

DIAGNOSIS
Radiation-induced morphea

TREATMENT AND COURSE
Plan to initiate UVA1 phototherapy

DISCUSSION

Radiation-induced morphea (RIM) is a rare and under-recognized complication of radiotherapy. As breast conservation surgery becomes more common in the treatment of breast cancer, adjuvant treatment with radiation therapy has also increased. RIM occurs months to years after treatment and can extend beyond the irradiated area. Sixty-six cases of RIM have been reported in the literature. A case of widespread morphea developing following radiotherapy for breast cancer has also been reported. The development of RIM is independent of the radiotherapy dose, fractionation scheme, use of a boost, patient age, date of irradiation, or the presence of other dermatological or connective tissue diseases. Clinically, RIM presents as sclerotic plaques with violaceous borders forming within the irradiated field often leading to disfigurement. Significant pain is often reported. As previously mentioned, the cutaneous lesions may extend and/or appear in areas not directly irradiated. While the pathogenesis of RIM is unknown, several theories have been proposed. One theory is that an abnormally high secretion of cytokines causes fibroblast activation and fibrosis. Another hypothesis is that post-radiation neoantigen formation triggers autoimmunity via TGF-β. This theory arose from the discovery of a TGF-β dependent pathway linked to the over-expression of connective tissue growth factor.

Diagnosis requires tissue to exclude clinically similar conditions including chronic radiation dermatitis, recurrent malignancy, infectious mastitis, lichen sclerosus, graft-versus-host disease, or radiation dermatitis. In RIM, histology reveals inflammatory and sclerosing phases. Classically, there are thickened collagen bundles in the reticular dermis. Rarely, the subcutaneous fat is involved with a mixed lobular and septal panniculitis. RIM is differentiated from post-radiation fibrosis by the presence of dermal fibrosis accompanied by an inflammatory infiltrate not seen in the latter.

The natural course has not been well studied, but reports of spontaneous resolution exist. There are no established treatment regimens and clinical outcomes are often unsatisfactory. Common therapies include topical and intralesional corticosteroids, topical calcineurin inhibitors, systemic immunosuppression, and ultraviolet light therapy. PUVA likely works via a reduction in cytokines, specifically of TGF-β. Both high and low dose UVA1 therapy have been reported to result in clearance. Improvement has been reported with intralesional and topical corticosteroids, particularly when utilized early in the disease course. Total resection of sclerotic areas may be beneficial for painful lesions. Imatinib has been suggested as a therapeutic option, acting via PDGF receptor pathway blockage and subsequent TGF-β inhibition.

KEY POINTS:

1. RIM of the breast is a rare condition that can have a profoundly negative impact on a patient's quality of life.
2. Early biopsy is critical to establish the diagnosis and rule out mimickers, notably metastatic disease.
3. While the pathogenesis and treatments are not well defined, patients may benefit from both skin-directed and systemic therapies.

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CHICAGO DERMATOLOGICAL SOCIETY **Case #5**

Presented by Victoria Godinez-Puig, MD and Simon Yoo, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
An 80-year-old Hispanic female presented with a 10-year history of a nodule on the left thigh. Although initially stable and asymptomatic, the nodule rapidly enlarged and became exquisitely tender over three months. The patient reported having had a "benign lesion" excised on the same location fifteen years prior in El Salvador. She denied other skin lesions. Her review of systems was otherwise negative.

PAST MEDICAL HISTORY:
Renal cell carcinoma s/p radical nephrectomy and IVC thrombectomy, small bowel obstruction s/p intestinal excision, diabetes mellitus c/b peripheral neuropathy, hypertension, osteoporosis, depression, Parkinson's disease, spinal stenosis, hemorrhoids.

MEDICATIONS:
Insulin, sitagliptin, clonidine, carbidopa-levodopa, enalapril, sertraline.

PHYSICAL EXAM:
Left lateral thigh with a 2.2 cm well-demarcated, sessile, smooth, deep purple, frambesiform nodule. Dermoscopy revealed a septated, exophytic nodule composed of greenish blue pigmented clods (large globules) interspersed by prominent irregular arborizing vessels.

HISTOPATHOLOGY:
Dermal infiltrative nodule composed of large, interconnected lobules and strands of large polygonal cells with focal ductal differentiation. Nuclear atypia and frequent mitotic activity were noted. Geographic zones of tumor necrosis were also identified. Deep dermal infiltrating tumor strands were detected at the base of the specimen.

DIAGNOSIS:
Porocarcinoma

TREATMENT AND COURSE:
The patient underwent Mohs micrographic surgery, and a tumor-free plane was obtained after two stages. We will follow the patient with a skin exam every 3 months for the next year, and every 6-12 months thereafter.

DISCUSSION
Porocarcinoma (PC) is a rare malignant tumor of the acrosyringeal portion of the eccrine sweat gland. It is thought to represent only 0.005% to 0.01% of cutaneous neoplasms. Elderly patients are most commonly affected, with an average age at diagnosis of 73 years.

Similar to other malignant adnexal neoplasms, the clinical presentation of PC is non-specific. PCs have been reported to present with various morphologies, including reddish ulcerated nodules and plaques, enlarging subcutaneous tumors, brown to black nodules with a whitish halo, and similar to our patient, smooth, multilobulated deep purple nodules. PCs may develop *de novo* or in association with an existent poroma. Thus, recent changes such as bleeding, ulceration or growth in a pre-existing, untreated poroma (which may present as an isolated papule, nodule or plaque) are suggestive of malignant transformation. PCs most commonly develop on the lower extremities (44% of cases), followed by the head, scalp, and upper

extremities.

Little is known about the dermoscopic features of PC. Hairpin, linear irregular and dotted vessels (known as polymorphous vascular pattern) surrounded by white to pink halos and round pink structureless areas have been demonstrated in PCs and benign poromas alike. Of note, the polymorphous vascular pattern is regarded as highly nonspecific, and is also found in various forms of melanoma and non-melanoma skin cancer. To the best of our knowledge, blue structureless areas similar to the ones described in our patient have not been described in PCs to date.

Microscopically, PCs are tumors at least partially composed of basaloid epithelial cells displaying variable degrees of duct formation and thus acrosyringeal differentiation. A wide variety of histologic patterns have been described, including clear, squamous, and spindle cell differentiation, mucus cell metaplasia, and colonization by melanocytes. Cytologic pleomorphism and nuclear atypia are variable. Pagetoid extension of malignant cells is occasionally seen. The largest published clinicopathologic study of 69 cases of porocarcinoma found that lymphovascular invasion, number of mitoses (>14/HPF), and tumor depth >7 mm were independently predictive of death. Furthermore, after adjusting for mitoses and depth, infiltrative borders were strongly predictive of local recurrence.

Given its rarity, the management of PC remains controversial. A review of 105 cases of PC treated with wide local excision showed local recurrence rates of 20%, in addition to regional and metastatic recurrence rates of 20% and 12%, respectively. Mortality surpassed 65% in patients with nodal metastases. In contrast, the single largest series of PCs treated with Mohs micrographic surgery (MMS) published to date, found that only 7% of patients developed regional metastases to lymph nodes, whereas no patients experienced local recurrence, distant metastases or disease-specific death after mean follow-up periods ranging from 19 months to 6 years. Hence, MMS may be an effective treatment for PC.

KEY POINTS

1. Porocarcinoma (PC) is a rare malignant tumor of the eccrine sweat gland that presents as a non-specific papule or nodule.
2. Given its rarity, little is known about the dermoscopic features of PC. Polymorphous vascular patterns have been described in a number of cases.
3. Recently published data suggests that Mohs micrographic surgery may be the best suitable approach for treating PC and limiting recurrence as well as regional and distant metastases.

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CHICAGO DERMATOLOGICAL SOCIETY **Case #6**

Presented by Melanie Clark, MD and Stavonnie Patterson, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

This is a 70-year-old African American male who presented with a 45-year history of pruritic papules and plaques on the neck, trunk, and upper extremities.

Unknown

CHICAGO DERMATOLOGICAL SOCIETY **Case #7**

Presented by Sarah Adams, MD and Lacey Kruse, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS:
A 2-day-old otherwise healthy Hispanic male was born at term with a diffuse *blueberry muffin* eruption. There was no history of maternal infection and prenatal serologies were unremarkable. He was feeding well and urinating and stooling appropriately.

PHYSICAL EXAM:
Vitals: Afebrile, weight 3.420 kg (40%), head circumference 36 cm (90%).
HEENT: Normocephalic, atraumatic, red reflex intact, no corneal opacities, normal ears.
Abdominal: Soft, non-tender, non-distended, no hepatosplenomegaly.
Neurological: Normal tone, grasp, suck and Moro reflexes intact.
Lymph nodes: No cervical, axillary or inguinal lymphadenopathy.
Skin: Innumerable violaceous to blue, non-blanching macules and infiltrative papules over the scalp, face, trunk and extremities.

LABORATORY:
Abnormal: Platelet 134 thou/uL (150-450 thou/uL).
Normal/Negative: WBC, Hg/Hct, BMP, LFTs, CMV by urine PCR, toxoplasma IgM and IgG ab, rubella IgM and IgG ab, RPR, blood cultures.

IMAGING:
Head Ultrasound: No evidence of intracranial abnormalities.
Abdominal Ultrasound: A 0.9 x 0.7 x 1 cm right hepatic lobe hyperechoic lesion with possible internal calcification was visualized.

HISTOPATHOLOGY:
Within the superficial and deep dermis is a nodular and diffuse pleomorphic infiltrate composed of histiocytes with reniform nuclei and abundant eosinophilic cytoplasm with scattered eosinophils and neutrophils. Immunostaining for CD1a and S-100 are positive.

DIAGNOSIS:
Congenital Langerhans cell histiocytosis

TREATMENT AND COURSE:
The patient was referred to oncology for further evaluation. A skeletal survey was performed and did not show evidence of bone involvement. A repeat abdominal ultrasound demonstrated resolution of the previously noted hyperechoic liver mass and appearance of a new, solitary, ill-defined 1.1 x 0.7 x 1 cm hypoechoic lesion at a distinct location in the right hepatic lobe. Liver function testing remains normal. His imaging findings are of unclear clinical significance and he will continue to be monitored closely.

DISCUSSION:
The term *blueberry muffin baby* was initially coined in the 1960s to describe the cutaneous findings found in newborns with congenital rubella infections. Clinically, patients present with diffuse violaceous to blue, non-blanching macules, papules and nodules. This eruption has classically been described in association with neonatal TORCH infections, and today, it is most commonly seen with maternal cytomegalovirus infection. These cases show evidence of dermal extramedullary hematopoiesis (EMH) on histopathology. Less commonly, *blueberry muffin* lesions

can be seen in primary disorders of red blood cells including erythroblastosis fetalis due to ABO or Rhesus incompatibility, inherited hemolytic disorders, hereditary spherocytosis, or in the donor twin in cases of twin-twin transfusion syndrome. In these disorders, it is hypothesized that anemia serves as a trigger for sustained expression of dermal EMH. The *blueberry muffin* rash can also be seen in association with neoplastic disorders such as metastatic neuroblastoma, alveolar rhabdomyosarcoma, leukemia, and as in this case, Langerhans cell histiocytosis (LCH).

Langerhans cells (LCs) are derived from the bone marrow and normally reside in the epidermis where they act as antigen presenting cells. LCH is characterized by infiltration of the skin and other tissues with LCs, which are identified histologically with positive CD1a, Langerin and S-100 staining. Electron microscopy demonstrates the characteristic intracytoplasmic, racquet-shaped Birbeck granules. The precise etiology of LCH remains unclear, and hypotheses include somatic mutations, viral infection, immune or cytokine dysregulation, and altered apoptosis. Several investigators have demonstrated clonality in LCH tissue, suggesting that it may be best regarded as a neoplastic disorder.

LCH can affect multiple organ systems, with skin and bone being the most common sites of involvement, though lesions can present in lymph nodes, liver, spleen, gastrointestinal tract, lungs, thymus, bone marrow, oral mucosa, kidney, endocrine glands, and the central nervous system. It most commonly affects children ages 1 to 4 years but can present at anytime from birth into adulthood. Prognosis in LCH is highly variable and depends on the extent of organ involvement. Those with limited disease tend to have a good overall prognosis but multisystem disease with hematopoietic, liver, lung or splenic lesions has a mortality approaching 55%, even with aggressive treatment.

Cutaneous involvement causing a persistent skin eruption is often the presenting complaint and has classically been described as a severe, treatment-resistant seborrheic dermatitis-like eruption in the scalp and flexural areas. LCH occurring in the neonatal period more commonly presents as a crusted, red-brown, vesiculopustular eruption. Congenital LCH presenting as a *blueberry muffin* rash is a rare entity with only isolated cases reported in the literature. Most of these patients were noted to have skin-limited disease at birth, with the majority of lesions resolving spontaneously over the course of several months. However, some patients did progress to develop extra-cutaneous involvement, most commonly bone lesions, months to years later.

KEY POINTS:

1) Langerhans cell histiocytosis should be considered in the differential diagnosis of a *blueberry muffin* eruption.
2) Langerhans cell histiocytosis is a multi-organ system disease and close clinical monitoring is imperative for early identification of disseminated involvement.

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Fast Break

Case #8

Presented by Steve Xu MD, and Joaquin C. Brieva MD

Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This is a 46-year-old female who presented with multiple violaceous nodules on her legs and arms that were noted to be exquisitely tender. The lesions appeared shortly after sustaining injuries in a motor vehicle accident.

What is the etiology of this patient's symptoms?

CHICAGO DERMATOLOGICAL SOCIETY **CASE #9**

Presented by Michael W. Pelster, MD and Joan Guitart, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
A 69-year-old African-American male presented with skin changes and mild pruritus of the axillae, buttocks, elbows, hands, thighs, legs, and knees for 6-7 years. He had been treated with topical corticosteroids and anti-fungals without significant benefit. He was otherwise asymptomatic.

PAST MEDICAL HISTORY
Type 2 diabetes mellitus, HTN, GERD, prostate cancer

MEDICATIONS
Aspirin, atorvastatin, cyclobenzaprine, hydroxyzine, insulin glargine, pantoprazole

PHYSICAL EXAM
Multiple hyperpigmented, slack-appearing patches involving the axillae, buttocks, elbows, hands, thighs, legs, and knees. A total of 35% body surface area involvement. No cervical, axillary, or supraclavicular lymphadenopathy.

LABS
Abnormal: T-Cell Receptor (TCR) rearrangement (peripheral blood): suspicious for clonal TCR gene rearrangement, WBC: 3.4 K/UL (nl: 3.5-10.5 K/UL), Hgb: 11.7 g/dL (nl: 13-17.5 g/dL), creatinine 1.54 (nl: 0.00-1.30), glucose: 107 (nl: 65-100)

Normal/Negative: Sezary cell count, LDH, WBC differential, platelets, basic metabolic panel except for creatinine and glucose (above), LFTs

HISTOPATHOLOGY
A biopsy taken from the buttocks demonstrated a dense infiltrate in the superficial dermis composed predominantly of atypical lymphocytes and scattered histiocytes between dermal collagen bundles. Focal epidermotropism was noted. Multinucleated giant cells or elastolytic changes were not identified. The DPAS stain was negative for fungi. Immunohistochemistry was positive for CD3, CD4, and CD5 and negative for CD7, CD8, CD30, and TIA-1, and the tissue was also positive for TCR rearrangement.

DIAGNOSIS
Patch-stage interstitial mycosis fungoides presenting as granulomatous slack skin

TREATMENT AND COURSE
Treatment was begun with daily triamcinolone 0.025% ointment and thrice weekly NB-UVB with improvement noted at subsequent appointments.

DISCUSSION
Granulomatous slack skin (GSS) is an extremely rare variant of mycosis fungoides with an estimated 60 cases reported in the literature. Epidemiologically, it has a male predominance and primarily affects adolescents and adults. It is characterized clinically by the indolent progression of papules and plaques evolving into pendulous, lax skin in the intertriginous zones but can involve other sites as well. This variant is not thought to portend any adverse prognostic significance. There is, however, a reported association with secondary lymphoproliferative

neoplasms (particularly Hodgkin's lymphoma) in as many as 30-50% of cases, and deaths have been reported due to these associated malignancies. Given its rarity, there are limited data regarding treatment of granulomatous slack skin, but topical corticosteroids, radiation, NBUVB, PUVA, interferon, and topical nitrogen mustard have been suggested as treatment possibilities. Case reports showing recurrence after attempted surgical resection of the pendulous plaques argue against excision as an effective treatment modality.

Histologically, the granulomatous slack skin variant of MF is typically described as a granulomatous infiltrate of small, atypical T cells with cerebriform nuclei and often shows fragmentation of the elastic fibers (as seen in cutis laxa), elastophagocytosis, and/or emperipolesis. Macrophages and multinucleated giant cells are often admixed within the atypical T cell infiltrate. There are sometimes features of classic MF as well (i.e. epidermotropism of atypical T cells without spongiosis). Immunohistochemistry of the infiltrate is also similar to that of classic MF (CD3+, CD4+, CD8-). There is significant histologic overlap between granulomatous slack skin and granulomatous mycosis fungoides (GMF), and they are distinguished primarily by their clinical features (e.g. GMF does not demonstrate lax-appearing plaques).

To the best of our knowledge, what has not been reported is a link between the interstitial variant of mycosis fungoides and granulomatous slack skin. It is our institution's observation that a subset of patients with interstitial mycosis fungoides seem to exist on a continuum with granulomatous slack skin. With this patient and in other cases as well, we have observed presentations with an initial interstitial histology that later evolved into more classic GSS as well as cases with interstitial histology that demonstrate the clinical features of granulomatous slack skin at initial presentation.

KEY POINTS

1) Granulomatous slack skin is a rare clinical and histologic variant of mycosis fungoides that presents with lax skin, predominantly affecting intertriginous skin.
2) Some cases of Interstitial mycosis fungoides may exist on a continuum with granulomatous slack skin.

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Fast Break

Case #10

Presented by Amin Esfahani, MD, MSc, and Maria Colavincenzo, MD.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This is a 22-year-old female who presented with multiple, persistent, asymptomatic fleshy pink papules on the buccal mucosa, tongue and mucosal lip since early childhood.

What is the etiology of this patient's presentation?