Chicago Dermatological Society

June 2015
Monthly Educational Conference

Program Information
Continuing Medical Education Certification
Case Presentations

Wednesday, June 3, 2015
Stephens Convention Center – Rosemont, IL

Conference Host:
Division of Dermatology
Loyola University Medical Center

LOYOLA MEDICINE
We also treat the human spirit.®

Program

Stephens Convention Center
5555 N. River Rd., Rosemont, IL
Level 2, Conference Center

Registration Area – Foyer between Ballroom 41 and 42

Program Events

8:00 a.m.	**Registration begins for all attendees** Continental breakfast & visit with exhibitors *Ballroom 41*
9:00 a.m. - 10:00 a.m.	**Resident Lecture** *Ballroom 42* "Surgical CPT Coding" *Brett M.Coldiron, MD*
9:30 a.m. - 10:45 a.m.	**Clinical Rounds** Patient Viewing – *Rooms 57, 59 & 60* Slide & Poster Viewing; Visit with Exhibitors *Ballroom 41 and Foyer area*
11:00 a.m. - 12:00 p.m.	**General Session** *Ballroom 42* GUEST LECTURE - "The Skin Cancer Epidemic" *Brett M.Coldiron, MD*
12:00 p.m. - 12:40 p.m.	**Lunch & Visit with Exhibitors** *Ballroom 42*
12:40 p.m. - 12:50 p.m.	**CDS Business Meeting** *Ballroom 42*
12:50 p.m. - 2:30 p.m.	**Case Discussions** *Ballroom 42*
2:30 p.m. - 3:00 p.m.	**Maintenance of Certification** - Self-Assessment Questions *Ballroom 42*
3:00 p.m.	**Meeting adjourns**

Mark the Date!

Next CDS monthly meeting – Wednesday, October 14, 2015; hosted by the University of Illinois at Chicago – at the Gleacher Center in downtown Chicago.

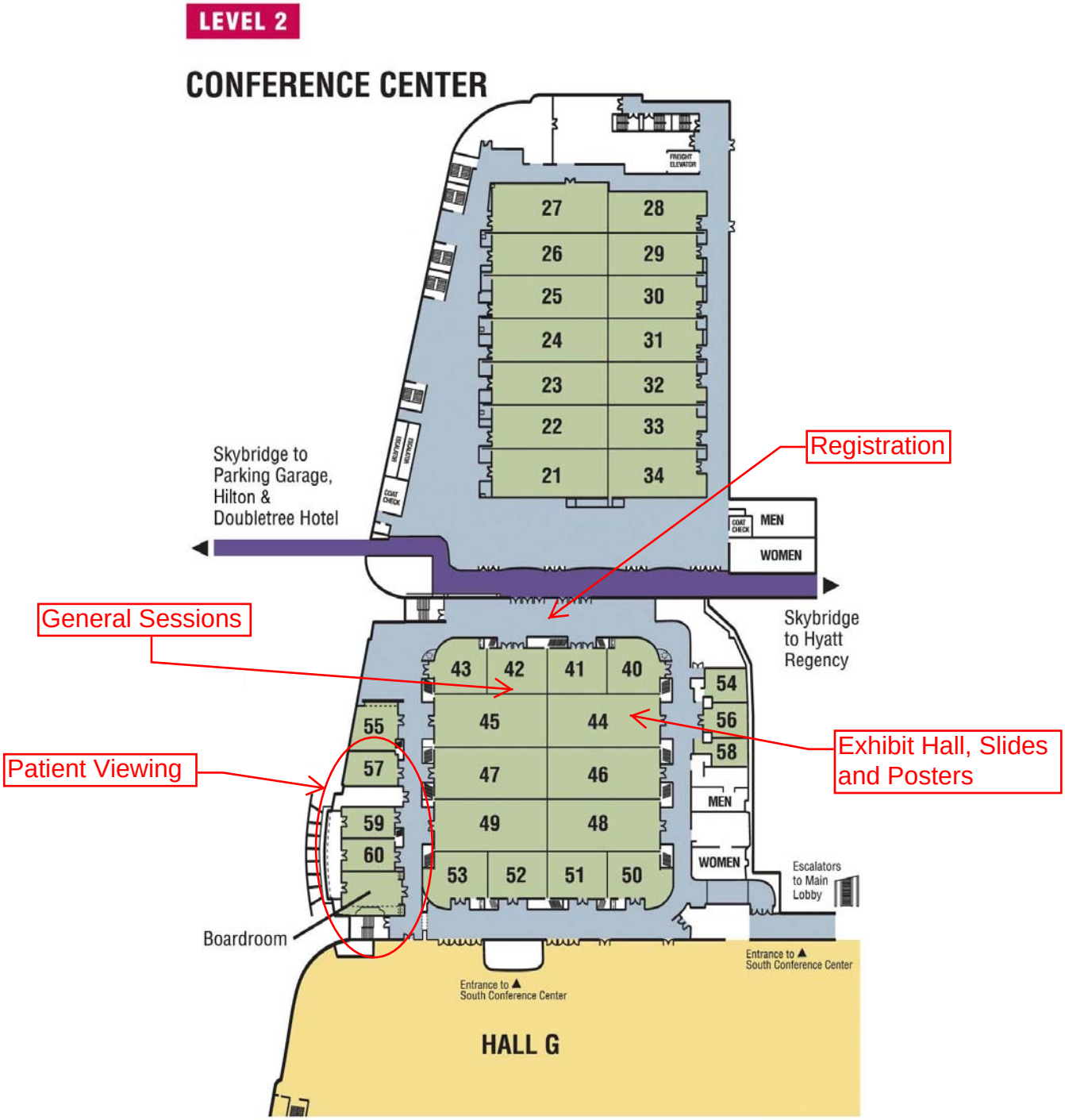
Guest Speaker

BRETT M. COLDIRON, MD
The Skin Cancer Center;
Assistant Professor of Dermatology and Otolaryngology
University of Cincinnati
Cincinnati, OH

Dr. Coldiron attended medical school at the University of Kentucky in Lexington. While there, he won the Lange Book Award for outstanding achievement as a medical student, and was on the curriculum committee, chairman of the president's council, and was class president. Dr. Coldiron returned home to Cincinnati University Hospital to complete his first residency in internal medicine. Following three years of practice in internal medicine, Dr. Coldiron became interested in dermatology and then completed a dermatology residency at the University of Texas Health Science Center at Dallas, (Parkland Memorial Hospital). While a resident there, he won numerous honors, including a national scientific manuscript contest, and had several research papers Published. He then developed a keen interest in dermatologic surgery. Dr. Coldiron completed his residency training and went on to Chicago where he completed a Mohs Micrographic surgery and dermatologic surgery fellowship at Northwestern University and the University of Illinois at Chicago. After completing his fellowship training in July 1989, Dr. Coldiron returned to his hometown of Cincinnati. He was Assistant Professor of Dermatology and Otolaryngology at the University of Cincinnati Medical Center from 1989 - 1992. He currently is in private practice but maintains a Clinical Assistant Professorship at the University of Cincinnati. He currently takes care of patients, teaches medical students, residents, and fellows and has several active clinical research projects. Dr. Coldiron is the founder of The Skin Cancer Center. The Skin Cancer Center is the only JACHO accredited office in Ohio dedicated to the prevention, diagnosis, and treatment of Skin cancer.

CME Conflict of Interest Disclosure: Dr. Coldiron has no conflicts of interest to disclose.

Donald E. Stphens Convention Center
Conference Center - Level 2



CONTINUING MEDICAL EDUCATION CREDITS

Chicago Dermatological Society

Presents

"Chicago Dermatological Society Monthly Meeting Series"

June 3, 2015 *Rosemont, 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 activity, participants will be able to:

1. Discuss key factors in the diagnosis and treatment for a variety of dermatologic diseases and conditions, including psoriasis, hair disorders, and dermatological symptoms of systemic diseases.

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.

None of the faculty, planner and/or content managers have nothing to disclose nor do they have any vested interests or affiliations.

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

Case

Presentations

TABLE OF CONTENTS

NOTES

NOTES

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 1**

Presented by Jennifer Eyler MD, George Garib MD, Madhu Dahiya MD, and James Swan MD
Section of Dermatology, Edward Hines Jr. Veterans Affairs Hospital

HISTORY OF PRESENT ILLNESS

A 69-year-old African American male presented to the dermatology clinic with a 4-month history of a mildly pruritic rash on the trunk, extremities, groin, and scalp. He described round pink lesions on his skin that initially developed several months ago in the groin and recently spread to involve his trunk and extremities. He denied any new medications, recent illnesses, or sick contacts. He had previously been treated unsuccessfully with ketoconazole cream and clotrimazole cream.

PAST MEDICAL HISTORY

Hepatitis C Virus
Hypertension
Hyperlipidemia
Hypothyroidism

MEDICATION

Levothyroxine
Amlodipine
Hydrochlorothiazide
Lisinopril
Mirtazapine
Sertraline
Trazodone
Losartan
Rosuvastatin

ALLERGIES

No Known Drug Allergies

FAMILY HISTORY

None pertinent

SOCIAL HISTORY

Negative for tobacco, alcohol, or illicit drug use

PHYSICAL EXAM

Physical examination demonstrated well demarcated violaceous papules and plaques on the upper back, anterior chest, extensor forearms, and posterior thighs involving 10% body surface area. The lesions were annular with elevated borders and central depigmented atrophic scarring. The groin and buttocks demonstrated many well demarcated hypopigmented atrophic patches and scattered thin plaques. The temporal scalp had a single small atrophic pink plaque. There was no mucous membrane involvement.

HISTOPATHOLOGY

Initial punch biopsy was consistent with a nonspecific lichenoid dermatitis. Repeat punch biopsies of lesional and perilesional uninvolved skin from the trunk were later obtained for histopathologic confirmation and special stains. A lichenoid dermatitis was again present on the lesional biopsy

while no significant histopathologic changes were observed on the perilesional biopsy. Verhoeff-van Gieson stain for elastic fibers was performed on both biopsies. It revealed destruction of elastic fibers in the central papillary dermis and upper reticular dermis of the lesional biopsy, while the elastic fibers on the perilesional biopsy were preserved.

DIAGNOSIS

Annular Atrophic Lichen Planus

TREATMENT AND COURSE

The patient was initially prescribed triamcinolone 0.1% ointment to the trunk and extremities and tacrolimus 0.1% ointment to the groin. He continued to develop new annular atrophic skin lesions over the next several months. Repeat punch biopsies of lesional and perilesional uninvolved skin were obtained and confirmed a diagnosis of annular atrophic lichen planus. The patient was prescribed a short taper of oral prednisone which halted further progression of disease. He was subsequently started on pentoxifylline and continued on tacrolimus 0.1% ointment with no improvement in existing lesions. Hydroxychloroquine 400mg daily was initiated which initially resulted in some thinning of the plaques on his trunk, however further progression of the disease was noted after several months of treatment. Most recently, acitretin 25mg daily has been added to his treatment regimen.

DISCUSSION

Lichen planus is a common pruritic inflammatory disease of the skin, mucous membranes, hair follicles, and nails typically affecting the adult population with a highly variable clinical pattern and disease course. There are many clinical variants of lichen planus which all show a band-like lymphoid infiltrate, hypergranulosis, destruction of the basal layer, and numerous Civatte bodies histologically. Annular lichen planus is an uncommon variant most commonly seen in men with asymptomatic lesions involving the axillae and groin. Atrophic lichen planus is another variant demonstrating atrophic papules and plaques on the trunk and extremities. Annular atrophic lichen planus (AALP) is the rarest variant of lichen planus incorporating features of both annular and atrophic lichen planus.

AALP was first reported by Friedman and Hashimoto in 1991 with the case of a 56-year-old African American male with a 25-year history of annular atrophic papules and plaques on the trunk and extremities. The second case reported by Requena *et al.* in 1994 described a 65-year-old female with characteristic lesions on her right elbow and left knee. Lipsker *et al.* reported the third case of a 41-year-old male with a history of Sneddon's syndrome with lesions typical for AALP of 20-year duration. In all cases, the histopathologic examination revealed a lichenoid infiltrate with thinning of epidermis and loss of elastic fibers in the center of the active lesions.

More recent cases include Mseddi *et al.* in 2003, Morales-Callaghan *et al.* in 2005, Ponce-Olivera *et al*. in 2007, Kim *et al.* in 2008, and Li *et al.* in 2010. The characteristic findings of AALP in the above mentioned cases primarily occurred on the trunk and extremities. Most patients failed treatment with topical corticosteroids and noted some improvement with tacrolimus 0.1% ointment. Sugashima *et al.* reported the most recent case in 2012 which was unique in that it presented on the lower lip in a 32-year-old female. This patient had notable improvement with tacrolimus 0.1% ointment after 6 months.

All reported cases of AALP to date have occurred in adults, both male and female, presenting with a limited number of annular plaques with slightly elevated borders and depressed atrophic centers. Disease duration has ranged from 2 months to 25 years. Histopathologic findings characteristically demonstrate a lichenoid dermatitis of the raised lesional border with a flattened epidermis, loss of

rete ridges, and fibrosis of dermal papillae in the lesion center. The elastic fibers are destroyed in the papillary dermis of the lesion center presumably due to elastolytic activity of inflammatory cells. Macrophages present in the lichenoid infiltrate of acute lesions release elastases contributing to this destruction. Furthermore, elastic fibers appear fragmented by electron microscopy.

The course of AALP has proven to be chronic in most cases and frequently resistant to treatment with topical corticosteroids, phototherapy, and immunosuppressive agents. Earlier treatment in disease course may favor a more beneficial outcome. Existing lesions in our patient did not improve with topical corticosteroids and showed minimal improvement with tacrolimus 0.1% ointment. Our case displays more extensive involvement than previously reported. Our use of oral pentoxifylline, hydroxychloroquine, and acitretin has not been previously reported in the treatment of AALP. Acitretin is the only systemic agent for lichen planus that has achieved level A evidence, as it has previously been shown highly effective in a placebo-controlled double-blind study in 65 patients. To our knowledge there are no studies to date regarding the efficacy of systemic therapy in treatment of AALP. We report a twelfth case of AALP which is histologically indistinguishable from reported cases and highlights the chronicity of the disease and resistance to treatment. We encourage reporting additional cases of AALP to further characterize its clinical presentation and response to treatments.

REFERENCES

1. Friedman DB, Hashimoto K. Annular atrophic lichen planus. *J Am Acad Dermatol*. 1991;25:392-394.
2. James WD, Berger TG, Elston DM. Lichen planus and related conditions. In: James WD, Berger TG, Elston DM, eds, Andrews' Diseases of the Skin: Clinical Dermatology, 11th edn. Saunders Elsevier; 2011:213-215.
3. Kim BS, Seo SH, Jang BS, Kim MB, Oh CK, Kwon YW, Kwon KS. A case of annular atrophic lichen planus. *J Eur Acad Dermatol Venereol*. 2007;21:989-990.
4. Requena L, Olivares M, Pique E, Farina M, Martin L. Annular atrophic lichen planus. *Dermatology*. 1994;189:95-98.
5. Lipsker D, Piette JC, Laporte JL, Maunoury L, Frances C. Annular atrophic lichen planus and Sneddon's syndrome. *Dermatology*. 1997;105:402-403.
6. Mseddi M, Bouassadi S, Marrakchi S, Khemakhem M, Turki H, Zahaf A. Annular atrophic lichen planus. *Dermatology*. 2003;207:208-209.
7. Morales-Callaghan A Jr, Martinez G, Aragoneses H, Miranda-Romero A. Annular atrophic lichen planus. *J Am Acad Dermatol*. 2005;52:906-908.
8. Ponce-Olivera RM, Tirado-Sanchez A, Montes-de-Oca-Sanchez G, Leon-Dorantes G, Mercadillo-Perez P. Annular atrophic lichen planus. *Int J Dermatol*. 2007;46:490-491.
9. Kim JS, Kang MS, Sagong C, Yu HJ. Annular atrophic lichen planus associated with hypertrophic lichen planus. *Clin Exp Dermatol*. 2008;33:195-197.
10. Li B, Li JH, Xiao T, He CD, Gao XH, Chen HD. Annular atrophic lichen planus. *Eur J Dermatol*. 2010;20:842-843.
11. Sugashima Y, Yamamoto T. Annular atrophic lichen planus of the lip. *Dermatol Online J*. 2012;18:14.
12. Manousaridis I, Manousaridis K, Peitsch WK, Schneider SW. Individualizing treatment and choice of medication in lichen planus: a step by step approach. *J Dtsch Dermatol Ges*. 2013;981-991.

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 2**

Presented by Monika Kaniszewska, MD, Lily Uihlein, MD, and Wendy Schumacher-Kim, DO
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

A 3-year-old Caucasian male presented with progressive muscle weakness, frequent falls, and erythematous rash on the eyelids, ears, hands, and trunk. Patient had a history of frequent falls since he began walking at 9 months of age. Six months prior, patient was admitted to an outside hospital with muscle weakness, elevated creatinine kinase (CK) levels, and transaminitis. Work-up including MRI of the brain and spine were normal and the patient started physical therapy. He continued to have progressive weakness and became more irritable and easily fatigued with minimal exertion. He had difficulty standing, sitting, and often needed to be carried to the bathroom. He had weakness of the muscles in the neck causing his head to be unstable. His appetite was normal, but he had difficulty swallowing food. He walked on his toes and his mother noticed contractures of the ankles. Patient also developed asymptomatic skin changes including patches of hair thinning, redness on the ears, pink bumps on the fingers, and redness on the flanks and elbows. He was treated topically with permethrin for suspected scabies without improvement. There was no history of sun sensitivity, neurologic symptoms, muscle twitching, or joint pain. Patient had been meeting all of his developmental milestones.

PAST MEDICAL HISTORY

Febrile Seizures
Reactive airway disease

MEDICATION

Multivitamin
Albuterol

ALLERGIES

Amoxicillin

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

The patient lives with his mother and sister. Attends daycare.

PHYSICAL EXAM

Vital signs: T_{max} 98.1°F, P 124, BP 128/73, RR 24/min
Patient sitting up with head tilted to the right. Notable head lag when pulled from supine position.
There was axial and limb weakness with bilateral ankle contractures in plantar flexion position.
Cutaneous examination was notable for thin scaly patches with mildly decreased hair density over the bilateral temporal scalp. The bilateral helices had reticulated erythematous to violaceous patches. Over the bilateral eyelids, most prominent at the margin, there was mild swelling and subtle erythema. The bilateral flanks and axillary vaults had poikilodermatous changes and several atrophic plaques, which were more prominent on the right side. The distal and proximal interphalangeal joints had overlying pink well-defined scaly papules. The proximal nails folds had dilated capillary loops, swelling and ragged cuticles. There were subtle erythematous thin plaques over the bilateral elbows.

HISTOPATHOLOGY

Muscle biopsy of the left lateral thigh reveled CD20 positive B-lymphocytes located predominately around perimysial blood vessels. CD3 positive T cell and CD68 positive macrophages were also present in small numbers. There were atrophic types I and II muscle fibers. These findings are consistent with inflammatory myopathy.

LABORATORY RESULTS

The following labs were remarkable/abnormal:

Potassium	3.2	[3.3 – 5.1MM/L]
AST	52	[6 - 30 UI/L]
ESR	25	[0 – 15 MM]
ANA	Positive (1:160)	Negative
CK	1426 (117 on admission)	[50 – 320 IU/L]

The following laboratory studies were negative/normal:
LDH, aldolase, Von Willebrand Factor, thyroglobulin, Anti-TPO antibodies, TSH, ferritin
No deletion/duplication detected in the *DMD* Gene
Immunoglobulins, SPEP, and immunofixation

RADIOLOGY AND CARDIAC TESTING

Abdominal ultrasound showed hepatomegaly, but otherwise was within normal limits.
Renal ultrasound was normal.
EMG showed mildly reduced amplitude in the right and left peroneal compound muscle action potential with, normal conduction; otherwise normal (consistent with possible mild bilateral peroneal neuropathies).
EKG showed normal sinus rhythm.
Cardiac echocardiogram was normal

DIAGNOSIS

Juvenile Dermatomyositis

TREATMENT AND COURSE

During his hospital admission, patient was initially started on intravenous methylprednisolone 30mg/kg for three days and transitioned to prednisolone 15mg po q 12 hours and methotrexate 10mg po weekly with daily folic acid supplementation. He began to show improvement in his strength. His hospital course was complicated by intermittent episodes of hypertension. The patient was treated with hydralazine 7.5mg q 4 hrs, enalapril 5mg daily, and hydrochlorothiazide 12.5mg with good response. Work-up for underlying etiology of hypertension was negative. The patient is followed by a multi-disciplinary team including rheumatology, cardiology, gastroenterology, and continues to receive physical therapy. He was discharged to a rehabilitation center. His skin changes have remained stable and he was started on triamcinolone 0.1% ointment as well as strict sun protection.

DISCUSSION

We present a case of juvenile dermatomyosistis (JDM) in a 3-year-old child. JDM is a rare autoimmune myopathy with characteristic cutaneous and systemic findings. The underlying pathogenesis of JDM is the result of a capillary vasculopathy. Presentation usually occurs between age 2 and 15 (average 7.7 years) with a female predilection of 1.7-2:1. The criteria for diagnosis of JDM was first described by Bohan and Peter in 1975, which included characteristic rash with the addition of 3 out of 4 of the following: symmetric proximal muscle weakness, increased serum muscle enzymes, characteristic histologic findings, and EMG changes consistent with inflammatory myopathy. Cutaneous findings typically precede muscle weakness by up to 2 years; however, our patient developed weakness prior to the onset of rash. A distinct subset of patients with amyopathic JDM has also been reported.

Cutaneous manifestations of JDM can vary; however, classic findings including Gottron's papules, heliotrope erythema, and proximal nail fold capillary changes are the most common features. Erythematous plaques on extensor surfaces, malar erythema, and pruritic psoriasiform scalp dermatitis may also be present. Reduction in nail-fold capillary density has been found to be not only a sensitive marker for JDM but also an indicator of disease activity. Muscular symptoms usually manifest as difficulty with daily tasks (e.g. brushing hair, walking up stairs). Decreased strength in the quadriceps and deltoid was most often observed on physical examination.

Although the diagnosis of JDM is primarily clinical, laboratory studies may help support the diagnosis. The most common laboratory abnormalities are non-specific, including elevated erythrocyte sedimentation rate (ESR), lactate dehydrogenase (LDH), and aspartate aminotransferase (AST). Muscle enzymes, creatinine kinase (CK) and aldolase, may be initially normal in JDM patients. One study reported that elevated CK and aldolase were seen in 19% of patients on initial presentation, but that 44% of patients had an elevation in one of the tests within the following 6 months period. In addition, muscle enzyme levels can be normal despite clinical evidence of disease. As a result, serial measurements of these values are generally recommended. Up to 60-70% of patients with JDM will have an elevated ANA titer. Myositis-specific autoantibodies (up to 63%) may be found in children, and have been shown to be correlated with distinct clinical presentations.

As in the adult form, certain auto-antibodies may be associated with specific disease phenotypes in JDM. Anti-Jo-1 is an antibody directed against histidyl-tRNA synthetase and is associated with anti-synthetase syndrome (myositis, interstitial pneumonia, arthritis, fever, and mechanic's hands). Studies show that anti-Jo-1 antibodies are uncommonly seen in children (2.6%), unlike adults. Anti-Mi-2 antibodies, directed against a DNA helicase, are associated with a positive prognosis, classic cutaneous findings, good response to treatment, and lower risk of lung disease. As in adults, anti-Mi2 antibodies are detected in about 4-10% of children with dermatomyositis. Anti-p155/140 antibodies, associated with more severe cutaneous involvement with generalized lipodystrophy, is seen in about 23-32% of children. Anti-p140, which is found in up to 23% of children with JDM, is associated with increased risk of calcinosis cutis.

Other studies may also assist in the diagnosis of JDM by providing evidence of muscle inflammation. Muscle biopsy will demonstrate perifascicular atrophy and inflammatory infiltrates with evidence of vasculitis. Electromyography will show inflammatory myopathy. More recently, non-invasive modalities such as magnetic resonance imaging (MRI) have been used more frequently. Typical MRI findings include increased T2-weighted signal intensity of affected muscle due to inflammation and edema. An enhanced chemical-shift artifact is also seen. This is a normal finding highlighting the muscle-fat interfaces but is enhanced in patients with active disease.

Late sequelae of JDM include flexion contractures, calcinosis cutis, dysphagia, cutaneous infarcts, and ulcers. Patients are at risk for development of gastrointestinal vasculitis resulting in sepsis as well as repeated respiratory infections secondary to muscle weakness. Vigilant monitoring for abdominal pain, melena, and fevers is imperative as bowel perforation can be life threatening. Although outcomes are variable, the prognosis for JDM is better than for adult dermatomyositis. In contrast to the adult form, JDM is not reported to be associated with underlying malignancy. There is also no significant association with development of other connective tissue disorders.

Systemic treatment is usually required for JDM; oral or intravenous steroids are the most common initial therapy. Steroid-sparing agents are often necessary, the most common of which are methotrexate, intravenous gammaglobulin (IVIG), cyclosporine, azathioprine, and hydroxychloroquine. Newer targeted therapies such as rituximab have also been used in refractory cases. Physical therapy can improve muscle strength. Cutaneous manifestations can be treated with topical steroids and strict sun protection.

Even with early therapy, inflammatory myositis may continue for up to 2 to 3 years. The average time to remission is 27 months; however, a subset of patients will experience several flares. Prognosis is improved when treatment is aggressive and initiated early in the disease course. A chronic course is predictive of poor outcomes (including muscle damage and weakness). We present this case to illustrate the characteristic cutaneous and muscular manifestations of JDM and to underscore the importance of early and aggressive treatment to avoid these long-term sequelae.

REFERENCES

1. Batthish M, Feldman B. Juvenile Dermatomyositis. *Curr Rheumatol Rep*. 2011;13:216-224.
2. Peloro T, Miller F, Han T, Newman E. Juvenile dermatomyositis: A retrospective review of a 30-year experience. *J AM ACAD DERMATOL.* 2001;45:28-34.
3. Stockton D, Doherty VR, Brewster DH. Risk of cancer in patients with dermatomyositis or polymyositis, and follow-up implications: a Scottish population-based cohort study. *British Journal of Cancer.* 2001;85(1):41-45.
4. McCann LJ, Juggins AD, Maillard SM, *et al*. The juvenile Dermatomyostitis national registry and repository (UK and Ireland) – clinical characteristics of children recruited within the first 5 yr. *Rheumatology* 2006;45:1255-1260.
5. Rider LG, Shah M, Mayrova G, Huber A, *et al*. The myositis autoantibody phenotypes of the juvenile idiopathic inflammatory myopathies. *Medicine (Baltimore).* 2013;92(4):223-243.
6. Gunawardena H, Betteridge ZE, McHugh N. Myositis-specific autoantibodies: their clinical and pathogenic significance in disease expression. *Rheumatology* 2009;48:607-612.
7. Schmeling H, Stephens S, Goia C *et al*. Nailfold capillary density is importantly associated over time with muscle and skin disease activity in juvenile dermatomyositis. *Rheumatology* 2011;50:885-893.
8. Malttia C, Damasio M, Madeo A *et al*. Whole-body MRI in assessment of disease activity in juvenile dermatomyostiis. *Ann Rheum Dis* 2014;73:1083-1090.
9. Brown VE, Pilkington CA, Feldman BM *et al*. An international consensus survey of diagnostic criteria for juvenile dermatomyositis. *Rheumatology.* 2006;45:990-993.
10. Ravelli A, Trail L, Ferrari C *et al*. Juvenile Dermatomyositis: A multinational, multicenter study of 490 patients. *Arthritis Care & Research.* 2010;62(1):63-72.

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 3**

Presented by Anne Marie Mahoney MD, George Garib MD, Jodi Speiser MD, and Lily Uihlein MD
Division of Dermatology, Loyola University Medical Center

UNKNOWN

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 4**

Presented by Kelly K. Park MD, MSL, Madhu Dahiya MD, James Swan MD and David Eilers MD
Section of Dermatology, Edward Hines Jr. Veterans Affairs Hospital

HISTORY OF PRESENT ILLNESS

A 70-year-old Caucasian male presented for evaluation of a 6-year history of a bright red, scaly rash involving his forehead, cheeks, and central face. It was waxing and waning in nature and thought to be exacerbated by sunlight. His skin burned and itched intermittently, in particular when lesions were scaly. He had been evaluated by outside dermatologists and had 2 biopsies that showed non-specific parakeratosis and spongiosis. He was presented as an "unknown" to the Chicago Dermatological Society in 2014 and work-up for possible cutaneous lymphoma was initiated.

PAST MEDICAL HISTORY

Hyperlipidemia, hypertension, atrial fibrillation with history of ablation, type II diabetes mellitus, history of malaria, fatty liver disease, Gilbert's disease, testicular dysfunction, allergic rhinitis, coronary artery disease with history of stent placement, post-traumatic stress disorder, mucocutaneous HSV, sciatica, sleep apnea on CPAP

MEDICATIONS

Insulin, metoprolol, testosterone gel, digoxin, furosemide, lisinopril, loratadine, magnesium oxide, simvastatin, metformin, folic acid, multivitamin, warfarin, aspirin, omega-3-acid ethyl esters, valacyclovir, exenatide, hydrocortisone valerate cream

ALLERGIES

Cephalexin, testosterone transdermal patch

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Married with two children, Vietnam veteran, retired

PHYSICAL EXAM

Previous:
Forehead, glabella, nose, cheeks, temples, and upper eyelids with well-demarcated, brightly erythematous confluent plaques with areas of overlying fine white scaling. There were several areas of sparing on the forehead.

Current:
Left upper forehead with atrophic hyperpigmented patch. Right and left temporal scalp with thin pink plaques.

DERMATOPATHOLOGY

9/2014
Right cheek – Dense dermal lymphohistiocytic infiltrate with neutrophils and numerous eosinophils. Lymphocytes ranged from small and mature to larger, more transformed types. Immunohistochemical staining showed that the majority of the lymphocytes were T cells, highlighted by CD3 and CD5. Fewer B cells were seen, highlighted with CD20. The features were

those of a reactive infiltrate. Evidence of lymphoma was lacking. While some of the inflammatory cells were certain to be those of CLL/SLL, they were not the source of this lesion.

4/2013
Right forehead – Superficial, deep, and perifollicular lymphoplasmacytic infiltrate consistent with the impression of rosacea. Mounds of scale and neutrophilic debris near the follicular ostia were noted. B-cell PCR gene rearrangement studies performed showed positivity of immunoglobulin heavy and kappa chains (IGH, IGK), indicating the presence of a clonal B cell expansion. Sections of tissue showed a dense and monotonous inflammatory infiltrate composed of small sized lymphocytes with hyperchromatic nuclei and scant cytoplasm. Immunohistochemical stains showed that the lymphocytes were positive for CD20, CD5, CD23, and Bcl-2. The B cells were negative for CD10 and Bcl-6. Overall, the findings were suggestive of CLL/SLL, given the correct clinical setting.

11/2011
Right forehead – Acute/subacute spongiotic dermatitis with perivascular lymphohistiocytic infiltrate containing eosinophils. PAS stain was negative for fungal organisms.

11/2010 (outside dermatologist)
Parakeratosis and crust overlying an acantholytic and spongiotic epidermis, pustular folliculitis, reactive lymphoid hyperplasia.

3/2010 (outside dermatologist)
Parakeratosis and crust overlying a spongiotic epidermal hyperplasia. Moderately dense infiltrate in the superficial dermis.

LABORATORY DATA
No absolute lymphocytosis.
Peripheral blood eosinophilia.
LDH, beta 2 microglobulin, SPEP within normal limits.

BONE MARROW BIOPSY
Bone marrow was normocellular (40%).
Mildly increased erythrocytogenes with normoblastic maturation.
Lymphocytes comprised 13% of cellularity and were small and round with a small lymphoid aggregate.

Flow cytometry, bone marrow:
Small monoclonal B-cell population, consistent with CLL/SLL of B-cell origin with partial expression of CD38 suggesting a less favorable prognosis.

Cytogenetic study (FISH), bone marrow:
Trisomy 12 was observed.

IMAGING
CT: Mildly enlarged lymph nodes, maximum size 1.6 cm.

PREVIOUS TREATMENTS
Hydrocortisone valerate 0.2% cream, tacrolimus 0.1% ointment, metronidazole 1% gel, acitretin, prednisone, hydroxychloroquine

DIAGNOSIS

Leukemia Cutis in the Setting of Chronic Lymphocytic Leukemia/Small Lymphocytic Lymphoma (CLL/SLL)

TREATMENT AND COURSE

On initial presentation, the patient was thought to have seborrheic dermatitis with a possible allergic contact dermatitis. He was treated with hydrocortisone valerate 0.2% cream and underwent patch testing. He showed a 1+ reaction to Kathon CG and began practicing allergen avoidance. After 11 months of non-responsiveness to this management, repeat biopsy was performed which showed features of a spongiotic dermatitis and rosacea. Hydrocortisone valerate was discontinued and he started tacrolimus 0.1% ointment and metronidazole 1% gel, both of which worsened his symptoms. Approximately 18 months afterwards, he was biopsied again. The histopathology was suggestive of rosacea and seborrheic dermatitis. He was started on acitretin 10 mg daily and initially showed significant improvement. He was increased to acitretin 25 mg daily but the rash progressed and he had side effects from the medication, including sticky skin. He was decreased to acitretin 10 mg daily. He then had flaring of his disease as well as superinfection of facial lesions while he was on vacation in Florida. An urgent care facility treated him as an allergic contact dermatitis and gave him a prednisone taper that resulted in transient improvement of the rash. Upon his return from vacation and having been a few weeks after discontinuing prednisone, he was started on hydroxychloroquine 200 mg by mouth daily in addition to continuing acitretin 10 mg daily. He also switched his CPAP mask to a nasal cannula device, but this did not impact the rash. He eventually discontinued acitretin due to lack of improvement and side effects, including recurrence of sticky skin symptoms. He also had a brief trial of hydroxychloroquine but self-discontinued due to symptoms including subjective lower eyelid edema.

After re-biopsy and resulting studies suggestive of lymphoma, the patient was referred to the hematology/oncology service. Imaging, laboratory data, and biopsies were reviewed, and the case was discussed at the Loyola Hematology Conference. Consensus was to refer to radiation oncology if the patient desired radiation treatment for rash, and that there was no indication for chemotherapy. Subsequently, the patient was amenable to radiation treatment. Focal external beam radiation treatment (EBRT) was performed by radiation oncology as follows: total scalp received 30 gray (Gy) in 15 fractions, and the total face and shoulders received 20 Gy in 10 fractions from November 17, 2014 to December 9, 2014. His course was complicated by self-limited mild fatigue, moderate erythema, and mucositis. At 5-week follow-up, patient reported significant improvement of all areas of previously involved skin, in particular, of the forehead. Currently, the patient sustains this improvement without recurrence and continues to follow-up with radiation oncology, hematology/oncology, and dermatology.

DISCUSSION

CLL/SLL are lymphoid neoplasms of the elderly that are considered to be the same disease at different stages; CLL refers to disease that is characterized by hematological involvement while SLL refers to involvement limited to the lymph nodes. These are monoclonal lymphocytic neoplasms that share the same histopathologic and immunophenotypic findings. B cell markers including CD19, CD20 (weak), and CD23 are seen, as well as the T cell marker, CD5. Immunoglobulin (IgM or IgM and IgD) expression may be seen and immunoglobulin light chain expression is a marker of the clonal nature of the disease. Abnormal karyotypes can be seen in approximately 50% of patients; trisomy 12 may be seen as in our patient, and is associated with atypical histology and is a somewhat less favorable finding with an intermediate prognosis (median survival 114 months).

Cutaneous lesions may occur in 4-44% of CLL, including leukemia cutis, cutaneous malignancies, manifestations and complications of malignancy and treatment including hematology and infectious findings, and autoimmune diseases. Atypical clinical and histopathological presentations of these cutaneous findings are not uncommon. Leukemia cutis occurs in 4-20% of cases of CLL. These infiltrates commonly involve the face, and vary in morphology and can ulcerate or develop bullae. Facial lesions varying from erythematous plaques limited to the eyebrows and rosacealike and rhinophymatous eruptions of the face have been reported as presenting signs of CLL. Biopsy of these lesions may be helpful in certain cases to determine prognosis of CLL.

There is no specific treatment for leukemia cutis but lesions generally resolve with treatment of the underlying malignancy. Asymptomatic patients may be observed closely. For patients with localized (stage I) SLL, monotherapy utilizing localized radiation therapy of the involved area may be a consideration rather than chemotherapy or observation. Patients with higher stage SLL and symptomatic CLL are generally treated with chemotherapy. Radiation, allogeneic hematopoietic cell transplantation, and new novel agents are also options in certain cases.

CONCLUSION

We report the follow-up to an "unknown" case from a previous Chicago Dermatological Society meeting. This patient presented with an erythematous facial dermatosis that is now known to represent leukemia cutis in the setting of previously undiagnosed CLL/SLL. The presence of a protracted erythematous eruption of the head and neck refractory to multiple topical and systemic agents may prompt evaluation for underlying systemic disease or malignancy, particularly one that may be indolent. Appropriate work-up, referral to hematology oncology, and follow-up in a multidisciplinary fashion may be appropriate.

REFERENCES

1. Müller-Hermelink HK, Montserrat E, Catovsky D, *et al*. Chronic lymphocytic leukaemia/small lymphocytic lymphoma. In: Swerdlow SH, Campo E, Harris NL, *et al*. WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. Lyon, France: IARC Press; 2008. p 180-182.
2. Rai KR, Stilgenbauer S. Overview of the treatment of chronic lymphocytic leukemia. In: UpToDate, Larson RA (Ed), UpToDate, Waltham, MA. (Accessed on April 30, 2015.)
3. Freedman AS, Aster JC. Clinical manifestations, pathologic features, and diagnosis of small lymphocytic lymphoma. In: UpToDate, Lister A (Ed), UpToDate, Waltham, MA. (Accessed on April 30, 2015.)
4. Döhner H, Stilgenbauer S, Benner A, *et al*. Genomic aberrations and survival in chronic lymphocytic leukemia. *N Engl J Med*. 2000 Dec 28;343(26):1910-6.
5. Bonato M, Pittaluga S, Tierens A, *et al*. Lymph node histology in typical and atypical chronic lymphocytic leukemia. *Am J Surg Pathol.* 1998; 22:49.
6. Knuutila S, Elonen E, Teerenhovi L, *et al*. Trisomy 12 in B cells of patients with B cell chronic lymphocytic leukemia. *N Engl J Med*. 1986; 314:865.
7. Juliusson G, Merup M. Cytogenetics in chronic lymphocytic leukemia. *Semin Oncol*. 1998; 25:19.
8. Bonvalet D, Foldes C, Civatte J. Cutaneous manifestations in chronic lymphocytic leukemia. *J Dermatol Surg Oncol*. 1984; 10 (4): 278-282.
9. Robak E, Robak T. Skin lesions in chronic lymphocytic leukemia. *Leukemia and Lymphoma*. 2007; 48(5):855-65.
10. Smoller BR, Warnke RA. Cutaneous infiltrate of chronic lymphocytic leukemia and relationship to primary cutaneous epithelial neoplasms. *J Cutan Pathol. 1998*; 25: 160–4.
11. Greenwood R, Barker DJ, Tring FC, *et al*. Clinical and immunohistological characterization of cutaneous lesions in chronic lymphocytic leukaemia. *Br J Dermatol.*

1985; 113: 447–53.
12. Schmid-Wendtner MH, Sander C, Volkenandt M, *et al*. Case 2. Chronic lymphocytic leukemia presenting with cutaneous lesions. *Diag in Oncol.* 1999; 1084-85.
13. Barzilai A, Feuerman H, Quaglino P, *et al*. Cutaneous B-cell neoplasms mimicking granulomatous rosacea or rhinophyma. *Arch Dermatol.* 2012;148(7):824-831.
14. Kaddu S, Smolle J, Cerroni L, *et al*. Prognostic evaluation of specific cutaneous infiltrates in B-chronic lymphocytic leukemia. *J Cutan Pathol.* 1996; 23: 487–94.
15. Winfield HL, Smoller BR. Other lymphoproliferative and myeloproliferative diseases. In: Bolognia JL, Jorizzo JL, Rapini RP, eds. Dermatology. 3rd ed. Mosby: London. 2012.
16. Morrison WH, Hoppe RT, Weiss LM, *et al*. Small lymphocytic lymphoma. *J Clin Oncol.* 1989; 7:598.

FAST BREAK

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 5**

Presented by Patricia Todd MD, George Garib MD, Jodi Speiser MD, Wendy Schumacher-Kim DO
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

A 9-year-old Caucasian male presented to our clinic with a diffuse pustular eruption. His parents noted that these bumps had appeared quickly over a period of days and were bothersome to the patient but not particularly symptomatic. He had been diagnosed with Crohn's disease 2 months prior to presentation and was taking prednisone, allopurinol, and 6-mercaptopurine.

PAST MEDICAL HISTORY

Crohn's Disease
Recurrent staphylococcal furunculosis

MEDICATIONS

Prednisone
6-Mercaptopurine
Allopurinol

ALLERGIES

No Known Drug Allergies

FAMILY HISTORY

Ulcerative colitis – Grandmother
Crohn's disease – Mother

SOCIAL HISTORY

Lives at home with both parents, brother, and sister.

PHYSICAL EXAMINATION

The patient was thin, but well-appearing. There were scattered 3-4 mm pustules on erythematous bases on the cheeks, upper and lower extremities. There were also several indurated erythematous papules with collarettes of scale. A few of these lesions had central erosion with overlying heme crust.

DERMATOPATHOLOGY

There was subcorneal and intraepidermal neutrophilic pustulosis. There was also an underlying predominantly perifollicular mixed infiltrate with neutrophils and some granulomas. Infectious stains were negative (Gram, PAS, GMS, Fite AFB).

LABORATORY RESULTS

Complete blood count (CBC) showed anemia with hemoglobin of 9.7 g/dL (baseline 12). ESR and CRP were elevated at 40 mm and 1.8 mg/dL, respectively.

ADDITIONAL TESTS

Surface culture was negative.

DIAGNOSIS

Pustular Pyoderma Gangrenosum in a patient with Crohn's Disease

TREATMENT AND COURSE

The patient was started on dapsone 50 mg daily in addition to continuation of his Crohn's Disease regimen. He was also given tacrolimus 0.1% ointment to apply topically twice daily to the lesions. CBC was monitored closely while on this therapy and only a mild drop in hemoglobin was seen. His skin improved within one month of dapsone therapy which was tapered over two weeks. His lesions healed with subtle, hypopigmented and atrophic scars.

DISCUSSION

Pyoderma gangrenosum (PG) has several clinical presentations including ulcerative (classic), bullous, pustular and vegetative. Only 4% of cases occur in children. The pustular variant is more common in children and is more commonly associated with bowel disease rather than other underlying diagnoses. All forms of PG demonstrate pathergy and most lesions are tender. In adults, the most common location is on the legs and peri-stomal skin, whereas lesions on the face, buttocks, genital, and perianal skin are more common in children.

PG in children is associated with an underlying systemic disorder in roughly 75% of cases. Frequent associations include inflammatory bowel disease, lymphoma/leukemia, and arthritis (listed in descending order of frequency). Within the realm of inflammatory bowel disease, ulcerative colitis is more common than Crohn's disease as the underlying condition. PG is considered a reactive lesion in inflammatory bowel disease.

Crohn's disease has many cutaneous manifestations including metastatic Crohn's disease, gastro-cutaneous fistulas, aphthous ulcers, and erythema nodosum. PG is uncommon, occurring in less than 3% of Crohn's patients. Unlike erythema nodosum, the presence and severity of PG does not parallel bowel activity. It may precede Crohn's diagnosis by months to years, and diagnosis in an otherwise healthy patient should increase the physician's awareness of other systemic symptoms.

Treatment of PG is difficult. Controlling the underlying systemic disease is important but is usually not adequate for resolution of PG. Local wound care and pain control are key. For mild, shallow disease, topical or intralesional corticosteroids may be sufficient. However, in more severe cases, systemic therapy is required. Prednisone is first-line therapy. Several other systemic medications have also been used to manage PG including TNF-alpha inhibitors and anti-neutrophil agents. Recently, topical calcineurin inhibitors have been reported to be successful in helping clear PG lesions.

REFERENCES

1. Powell FC, Su WPD, Perry HO. Pyoderma gangrenosum: classification and management. *J Am Acad Dermatol.* 1996 Mar; 34(3): 395-405.
2. Graham JA, Hansen KK, Rabinowitz LG, Esterly N. Pyoderma gangrenosum in infants and children. *Pediatr Dermatol.* 1994 Mar; 11(1): 10-7.
3. Paller A. Cutaneous changes associated with inflammatory bowel disease. *Pediatr Dermatol Dec*. 1986; 3(6): 439-45.
4. Thrash B, Patel M, Shah KR, Boland CR, Mener A. Cutaneous manifestations of gastrointestinal disease Part II. *J Am Acad Dermatol.* 2013 Feb; 68(2): 211e.1-33.
5. Miller J, Yenzer BA, Clark A, Jorizzo JL, Feldman SR. Pyoderma gangrenosum: a review and update on new therapies. *J Am Acad Dermatol.* 2010 Apr; 62(4): 646-54.
6. Rice SA, Woo PN, El-Omar E, Keenan RA, Ormerod AD. Topical tacrolimus 0.1% ointment for the treatment of cutaneous Crohn's disease. *BMC Res Notes.* 2013; 6: 19.

CHICAGO DERMATOLOGICAL CASE SOCIETY **CASE # 6**

Presented by Michael Dreifke MD and Laura Winterfield MD
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

A 35-year-old male presented with a 3-year history of a recurrent, non-healing ulcer over his left knee extending onto his left pretibial leg. He had associated tenderness in his left knee but was able to ambulate freely. He also complained of generalized joint pains and frequent non-traumatic shoulder dislocations beginning in his mid-twenties. In July 2011, he developed red nodules on both lower extremities that quickly ulcerated and expanded. He was diagnosed with necrotizing fasciitis and underwent extensive debridement and removal of his left bursa. Shortly after, he was referred to the National Institutes of Health (NIH) and diagnosed with PAPA syndrome. A treatment regimen of prednisone 80 mg daily and adalimumab yielded moderate improvement. Due to breakthrough joint pains and recurrent leg ulcers, adalimumab was discontinued, prednisone was decreased, and golimumab was initiated.

PAST MEDICAL HISTORY

Moderate inflammatory and cystic acne in adolescence
Frequent non-traumatic shoulder dislocations beginning in his 20s
Diagnosed with PAPA syndrome via genetic testing at the NIH in 2011

MEDICATIONS

Amlodipine
Golimumab
Pantoprazole
Prednisone
Oxycodone
Clobetasol 0.05% ointment

ALLERGIES

No Known Drug Allergies

FAMILY HISTORY

Father: Acne and arthritis, diagnosed with PAPA syndrome

SOCIAL HISTORY

1 PPD cigarette smoker x 20 years
Active in wrestling and football throughout high school

REVIEW OF SYSTEMS

Positive for generalized joint pains

PHYSICAL EXAM

The left anterior leg extending from the knee to the pretibia had a 22.5 cm x 13.0 cm ulcer with granulation tissue at the base. The superior, superior-lateral, and medial margins of the ulcer had focally undermined and inflamed erythematous borders. The back and chest had several scattered inflammatory papules and pustules. There was extensive scarring over the right lower extremity.

DIAGNOSIS

Pyoderma gangrenosum in setting of Pyogenic Arthritis, Pyoderma gangrenosum, and Acne (PAPA) syndrome

TREATMENT AND COURSE

At the initial visit in our clinic a trial of intralesional kenalog (ILK) 10 mg/ml was injected to one test area at the superior lateral portion of the left anterior leg ulcer. A 15-day prednisone taper, timolol 0.25% drops twice daily to the inferior granulated portion of the ulcer base, and clobetasol 0.05% ointment to the inflamed border were initiated. Golimumab was continued. At one-month follow-up, there was no improvement, and there was evidence of possible worsening at the previously injected site. ILK and clobetasol were discontinued and tacrolimus 0.1% ointment and prednisone 10 mg daily were started. At the patient's three-month follow-up visit, the ulcer edges were no longer actively inflamed. A bilayered tissue-engineered graft (Apligraf®) was placed on the superior, inferior, and medial/superior portion of the ulcer, covered with a non-adherent dressing, followed by absorptive bacteriostatic foam dressing (Hydrofera Blue®) and compression wrap. The graft remained in place for 4 days prior to falling off due to excessive drainage. Local wound care was continued and two additional units of the bilayered tissue-engineered graft were applied one month later. One month after the second application of the bilayered tissue-engineered graft, the ulcer was noted to be 18 cm x 10.5 cm (previously 22.5 cm x 13.0 cm) with re-epithelialization noted over superior and medial aspects of ulcer. The wound edges remained non-flamed.

DISCUSSION

Pyogenic Arthritis, Pyoderma gangrenosum, and Acne (PAPA) syndrome is a disorder first described in the literature in 1997 by Lindor *et al*. The disease is an autosomal dominant condition that leads to the dysregulated production of IL-1β. It frequently presents in the first decade of life as recurrent sterile arthritis. The cutaneous manifestations of the syndrome include pathergy, cystic acne, and recurrent ulcers resembling pyoderma gangrenosum. The arthritic symptoms are typically provoked by minimal trauma and tend to precede the cutaneous expressions of the disorder.

The syndrome is an auto-inflammatory disorder associated with two known heterozygous mutations within the PSTPIP1 gene, also known as CD2 antigen-binding protein 1 on chromosome 15q. PSTPIP1 encodes for proline-serine threonine phosphatase interacting protein 1, a cytoskeletal adaptor protein which functions to regulate the production of inflammasomes. Mutations lead to an aberrant and continual production of IL-1β, a potent inducer of TNF-alpha.

There have been variations of PAPA syndrome reported in recent years. In 2012, Braun-Falco *et al*. described two patients presenting with pyoderma gangrenosum, acne, and suppurative hidradenitis without evidence of pyogenic arthritis. There were no specific mutations described in these cases, however an increased number of microsatellite repeats within the PSTPIP1 gene were observed. A number of sporadic cases of PAPA syndrome have also been described in patients with rosacea and psoriasis, which had non-traditional mutations identified within the PSTPIP1 gene.

While systemic corticosteroids, cyclosporine, and other systemic immunosuppressive agents are considered first line therapies for pyoderma gangrenosum (PG), there are numerous case series within the literature highlighting the successful treatment of ulcers including PG with bioengineered tissue substitutes. Tissue-engineered grafts are particularly appealing in the treatment of PG due to the avoidance of potential pathergy that may occur at the donor site of an autologous skin graft. Bilayered tissue-engineered grafts such as Apligraf® may enhance the healing process by

inducing secondary intention wound repair through the release of growth factors and cytokines locally within the wound.

Treatment of PAPA syndrome presents unique challenges. The disease often responds poorly to systemic glucocorticoids and the considerable side effect profiles associated with chronic use of immunosuppressive therapies like cyclosporine and methotrexate make alternative therapies desirable. Today, therapy primarily involves TNF-alpha antagonists including infliximab and adalimumab. However, due to the persistent elevation of IL-1β, alternative treatment options focusing on IL-1 receptor antagonists like anakinra have shown promise. With costs relatively comparable to TNF-alpha inhibitors and similar side-effect profiles, IL-1 receptor antagonists may provide a promising new treatment option.

REFERENCES

1. Lindor NM, Arsenault TM, Solomon H *et al.* A new autosomal dominant disorder of pyogenic sterile arthritis, pyoderma gangrenosum, and acne: PAPA syndrome. *Mayo Clin Proc*. 1997;72:611–6152.
2. Veves A, Falanga V, Armstrong DG, Sabolinski ML. Apligraf Diabetic Foot Ulcer Study. Graftskin, a human skin equivalent, is effective in management of non-infected neuropathic diabetic foot ulcers. *Diabetes Care*. 2001;24:290-5.
3. Dierselhuis MP, Frenkel J, Wulffraat NM, Boelens JJ. Anakinra for flares of pyogenic arthritis in PAPA syndrome. *Rheumatology (Oxford)*. 2005; 44:406–408.
4. Renn CN, Helmer A, Megahed M. Pyogenic arthritis, pyoderma gangrenosum and acne syndrome (PAPA syndrome). *Hautarzt*. 2007;58:383–384
5. Jacobs JC, Goetzl EJ. “Streaking leukocytes factor,” arthritis and pyoderma gangrenosum. *Paediatrics*. 1975;56:570–578.
6. Ah-Weng JAA, Langtry S, Velangi CD, *et al*. Pyoderma gangrenosum associated with hidradenitis suppurativa. *Clin Exp Dermatol*. 2005;30:669–671.
7. Ruocco E, Sangiuliano S, Gravina AG, *et al.* Pyoderma gangrenosum: an updated review. *Eur J Acad Dermatol Venereol*. 2009;23:1008–1017.

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 7**

Presented by Dana Griffin MD, George Garib MD, Jodi Speiser MD, Swati Mehrotra MD and Lily Uihlein, MD
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

A 14-year-old boy presented for evaluation of an enlarging right-sided neck mass. The lesion was initially noted one year ago, with subsequent resolution. Six months later, the patient was involved in a motor vehicle accident resulting in minor trauma to his neck. Following this incident, the mass recurred and increased in size. The patient reported occasional mild tenderness to palpation and numbness of his right arm when pressure was applied to the mass. He had no history of fever, night sweats, or recent foreign travel.

PAST MEDICAL HISTORY

Otherwise healthy

MEDICATION

No medications

ALLERGIES

No known drug allergies

FAMILY HISTORY

No family history of tuberculosis.

SOCIAL HISTORY

The patient's family emigrated from Mexico, but the patient was born in the United States.

PHYSICAL EXAM

Physical exam revealed a 6 cm firm, non-tender, skin-colored to slightly bluish subcutaneous mass of the right mid-neck, underlying the sternocleidomastoid muscle. There was no regional lymphadenopathy.

LABORATORY RESULTS

Negative PPD and quantiferon gold.

RADIOLOGY

Ultrasound of the neck showed a heterogeneous and bilobed vascular mass that was separate from the sternocleidomastoid muscle. A CT scan of the neck revealed an enhancing well-circumscribed mass isodense to adjacent skeletal muscle.

HISTOPATHOLOGY

A fine needle aspiration was performed and was non-diagnostic. The lesion was subsequently excised. Histopathologic examination revealed a spindle cell lesion of neural origin. There were hypocellular areas and hypercellular areas with palisading nuclei. Thickened and hyalinized vessels were present throughout.

DIAGNOSIS

Clinical and histological features confirmed a diagnosis of a non-vestibular, extracranial schwannoma, likely arising from the brachial plexus.

TREATMENT AND COURSE

After excision, there has been no evidence of recurrence of the lesion.

DISCUSSION

Schwannomas are benign, slow-growing neural sheath tumors rarely seen in children. More common etiologies of a unilateral pediatric neck mass include reactive or infectious lymphadenitis, epidermal cyst, thyroglossal duct cyst, branchial cleft cyst, and dermoid cyst. Histologically, schwannomas are encapsulated tumors composed of two distinct cellular architectures: hypercellular Antoni A and hypocellular Antoni B areas.

Diagnosis of a solitary schwannoma is often challenging. Magnetic resonance imaging (MRI) is now felt to be the imaging modality of choice due to superior visualization of the lesion and surrounding anatomic structures. In many cases MRI can determine the nerve of origin, which is helpful in planning surgical removal. Complete excision without neurologic compromise is the therapeutic goal, and recurrence is extremely rare. Although neurogenic tumors in children are uncommon, they should be included in the differential diagnosis of an enlarging, asymptomatic neck mass in a child.

REFERENCES

1. Bondi S *et al*. Non-vestibular Head and Neck Schwannomas: A 10 Year Experience. *Eur Arch Otorhinolaryngol.* 2013;270(8):2365-9.
2. Kim DH *et al*. Operative Outcomes of 546 Louisiana State University Health Sciences Center Peripheral Nerve Tumors. *Neurosurg Clin N Am.* 2004;15:177.
3. McGuirt FW. Differential Diagnosis of Neck Masses. In: Cumming C *et al*., eds. Otolaryngology Head Neck Surgery. St. Louis:Mosby-YearBook 1993:1543-1553.
4. Bhagat S at al. Pediatric Cervical Sympathetic Chain Schwannoma with Horner Syndrome: A Rare Case Presentation. *Ear Nose Throat J.* 2014;93(3):E1-3.

FAST BREAK

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 8**

Presented by Smita Aggarwal MD, Patricia Todd MD, Lily Uihlein MD, Kelli Hutchens MD and Wendy Schumacher-Kim DO
Division of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

This 5-month-old female infant presented with a six week history of a rash which began as white draining bumps on her neck. The rash subsequently erupted into widespread blisters involving her scalp and trunk, and her skin was extremely itchy. She would also experience intermittent flushing, which would resolve in less than one hour, and mild diarrhea. Mom denied any fevers at home. Prior treatments included ketoconazole cream, oral amoxicillin and IV cephalexin for a presumed diagnoses of bullous impetigo, all with only mild improvement.

PAST MEDICAL HISTORY

Otherwise healthy. Delivered at 40wga via normal spontaneous vaginal delivery.

MEDICATIONS

Ketoconazole 2% cream
Amoxicillin

ALLERGIES

No known drug allergies

FAMILY HISTORY

None pertinent

SOCIAL HISTORY

Lives with both parents and two healthy older siblings.

PHYSICAL EXAM

Well appearing infant female. No hepatosplenomegaly. Numerous tense vesicles and bullae were present over the scalp, upper back and upper arms. Several large bullae with underlying erythematous edematous plaques were present over the chest and abdomen.

LABS

WBC: 20,900K/uL
Serum Tryptase: 27ng/mL

HISTOPATHOLOGY

A punch biopsy from the right abdomen demonstrated a perivascular and interstitial infiltrate in the papillary dermis. The infiltrate was predominantly composed of cells with a round central hyperchromatic nucleus with granular cytoplasm, cytologically consistent with mast cells. This was confirmed with a mast cell tryptase immunohistochemical stain which demonstrated an overall increase in mast cell quantity.

DIAGNOSIS

Bullous Mastocytosis

TREATMENT AND COURSE

The patient continued to develop several large new bullae every week and was very uncomfortable due to severe generalized pruritus. A regimen of systemic antihistamines was initiated including cetirizine, hydroxyzine, and cimetidine. She was also counseled on mast cell triggers and indications for epinephrine pen use. In spite of being compliant with this regimen, she continued to develop several new bullae every week on her scalp and trunk. Her flushing episodes persisted during which she would be extremely itchy. Topical cromolyn sodium 4% compounded in petrolatum was added to her regimen and clinical improvement has been noted at subsequent follow up visits.

DISCUSSION

Mastocytosis refers to a heterogeneous group of clinical disorders, all of which are characterized by the accumulation of mast cells in one or more organs including the skin. Up to 55% of patients present before 2 years of age, and gene analysis has revealed activating c-kit mutations in both adult and pediatric cases. Mastocytosis is divided into two main subgroups: systemic and cutaneous mastocytosis, with particular variants more common in children. In an extensive review by Meni *et al*., 75% of pediatric cases presented as urticaria pigmentosa, 20% as mastocytoma, and only 5% as diffuse cutaneous mastocytosis. Darier's sign is present in 90% of patients with cutaneous mastocytosis. However, a negative Darier's sign does not preclude the diagnosis, especially in patients with entirely flat lesions at baseline. All forms can become vesicular or bullous, due to histamine or other chemical mediator induced transudation of fluid – a presentation termed bullous mastocytosis. These blisters may become secondarily infected but generally heal without scarring. Bullous changes are unusual after 2 years of age.

Diffuse cutaneous mastocytosis is the most rare form of pediatric mastocytosis, and portends a risk of systemic disease.

Pediatric mastocytosis is often considered a benign and self-limiting disease, and an analysis of the literature reveals that regression occurs in 2/3 of children after a median follow up of 6 years. However, 1/3 of patients did demonstrate persistent or progressive disease and some have suggested that progression could be correlated with lesion size. Serum tryptase levels are often evaluated and while these correlate with extent of skin involvement, they do not appear to have significant prognostic value. 3% of cases may progress to mast cell sarcoma or mast leukemia with a reported mortality of > 50%, emphasizing the importance of long term follow up for these patients.

Systemic mastocytosis, when it does occur, is far more common in adults than in children. The gastrointestinal and skeletal systems are most commonly affected, but involvement of many other organs can be seen including the bone marrow. Importantly, death can result from massive histamine release causing bronchospasm, flushing, and hypotension.

There is no curative treatment for mastocytosis. Treatment generally consists of drugs for symptomatic relief and avoidance of mast cell triggers. The most commonly used agents are histamine receptor blockers, leukotriene antagonists, oral cromolyn sodium, oral corticosteroids and phototherapy. Treatment of small infants is especially challenging, and there is no established regimen. While oral cromolyn sodium is licensed for the treatment of systemic mastoctyosis, its use in children below 2 years of age is not recommended. As a result, several authors have recently utilized a cutaneous emulsion of 4% cromolyn sodium as an alternative, which is being developed for the treatment of atopic dermatitis in children. Due to the propensity for Darier's sign, it is important for the emulsion to be rubbed in gently. A slight worsening of symptoms may be seen initially, but this will eventually subside. This novel use of an established agent may prove useful

for small infants with not only mastocytosis, but also inflammatory conditions related to excessive histamine release and mast cell activity.

REFERENCES

1. Castells M, Metcalf DD, Escribano L. Guidelines for the diagnosis and treatment of cutaneous mastocytosis in children. *Am J Clin Dermatol*. 2011. Aug; 12(4): 259-70.
2. Edwards AM, Capkova S. Oral and topical sodium cromoglicate in the treatment of diffuse cutaneous mastocytosis in an infant. *BMJ Case Rep*. 2011: bcr0220113910.
3. Paller AS, Mancini AJ. Hurwitz Clinical Pediatric Dermatology: a textbook of skin disorders of Childhood and adolescence, 4th edition. China: Elsevier, 2011.
4. Meni C, *et al*. Paediatric mastocytosis: a systematic review of 1747 cases. *Br J Dermatol*. 2015; 172: 642-51.

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 9**

Presented by Rebecca Rovner, MD, George Garib, MD, Jodi Speiser, MD, Lily Uihlein MD
Department of Dermatology, Loyola University Medical Center

HISTORY OF PRESENT ILLNESS

A 3-year-old boy was referred for evaluation of a generalized itchy eruption. The rash initially appeared on his abdomen two weeks prior and subsequently spread to involve his back, arms, legs, face, and neck. The eruption developed about three weeks after receiving his first infusion of cyclophosphamide for treatment of Evan's syndrome. Review of systems was positive for a three-day history of chills, fever, and cough.

PAST MEDICAL HISTORY

Evan's syndrome (autoimmune hemolytic anemia and thrombocytopenia), previously treated with intravenous immunoglobulin, rituximab and cyclosporine
Asthma
Eczema

MEDICATIONS

Albuterol
Cyclophosphamide x 1 dose (administered 3 weeks prior to rash onset)

ALLERGIES

No known drug allergies

FAMILY HISTORY

No family history of hematologic disease or similar eruptions.

SOCIAL HISTORY

Lives at home with his parents, sister and cousin. No recent travel.

PHYSICAL EXAMINATION

Vital signs: T 98.8 F, P 120
The patient was ill-appearing and crying. Oropharynx and conjunctivae were unremarkable. On the bilateral upper and lower extremities, abdomen, back, cheeks and upper forehead extending into scalp, there were scattered 1 - 2 mm perifollicular erythematous papules and pustules, many of which were excoriated. Palms and soles were clear.

DERMATOPATHOLOGY

A punch biopsy of the right thigh demonstrated predominantly intrafollicular and perifollicular mixed inflammation with primarily eosinophils. No fungal or bacterial organisms were seen on PAS, GMS or special stains.

LABORATORY RESULTS

Laboratory studies were notable for the following:

WBC	8.9	4.0 – 11.0 K/UL
% eosinophils	49%	
Absolute eosinophil count	4.5	0.0 – 0.7 K/MM3
HGB	10.9	11.0 – 14.5 GM/DL
HCT	32.8	32.0 – 43.0 %
Platelets	349	150 – 400 K/UL

Bacterial culture	Few colonies of S. aureus	
Mineral oil preparation	Negative for scabies	

DIAGNOSIS

Eosinophilic pustular folliculitis

TREATMENT AND COURSE

The patient was prescribed mometasone 0.1% ointment twice daily and hydroxyzine as needed. Dilute bleach baths were recommended. He was subsequently lost to follow up.

DISCUSSION

Eosinophilic pustular folliculitis (EPF) is a disorder characterized by noninfectious infiltration of hair follicles by eosinophils. It is predominantly a disease of males and is relatively rare, except in the AIDS population (in which its incidence has been estimated to be as high as 9%). In 1970, Ofuji and colleagues first described EPF in three Japanese young men with recurrent pruritic papulopustules involving the face, trunk, and extremities. EPF subsequently has been reported in infants, in patients with HIV and other infectious diseases, in patients with hematological disorders, and following bone marrow suppression. EPF has also been associated with certain medications, including minocycline, indeloxazine hydrochloride, timepidium bromide, carbamazepine, allopurinol, cyclophosphamide, 5-fluorouracil, and methotrexate.

EPF is classified into three main clinical subtypes. Classic EPF, which is most frequently described in Japanese adults, presents with chronic, recurrent follicular pustules forming circinate plaques in a seborrheic distribution. Immunosuppression-associated EPF typically manifests with severely pruritic papules on the face and upper trunk. While this category of EPF most commonly occurs in association with HIV, it is also seen in patients with hematologic or lymphoproliferative disease (including lymphoma, leukemia, polycythemia vera, and myelodysplastic syndrome), following stem cell and bone marrow transplantation, and in the setting of bacterial, parasitic and fungal infections. EPF of infancy usually presents with recurrent crops of grouped follicular papules and pustules on the scalp.

Topical steroids and calcineurin inhibitors are generally the initial treatment for all types of EPF. Anti-retroviral treatments are also considered first line therapy for HIV-associated EPF. NSAIDs, particularly indomethacin, are often recommended for classic EPF. Other treatments include metronidazole, isotretinoin, minocycline, and phototherapy.

Based on clinical and histopathologic findings, our patient was diagnosed with eosinophilic pustular folliculitis. This eruption developed in a patient with Evan's syndrome during treatment with cyclophosphamide. There is one report of eosinophilic pustular folliculitis arising in a patient receiving cyclophosphamide, methotrexate, and 5-fluorouracil. Our patient demonstrates a unique presentation of a relatively uncommon disorder in the setting of both hematologic disease and chemotherapy.

REFERENCES

1. Norton A, Roberts I. Management of Evans syndrome. *British J Haematology*. 2005; 132: 125-137.
2. Nervi SJ, Schwartz RA, Dmochowski M. Eosinophilic pustular folliculitis: A 40 year retrospect. *J Am Acad Dermatol*. 2006; 55: 285-9.
3. Katoh M, Takashi N, Miyachi Y, Kabashima K. Eosinophilic pustular folliculitis: A review of the Japanese published works. *J Dermatol.* 2013; 40: 15-20.

4. Fujiyama T, Tokura Y. Clinical and histopathological differential diagnosis of eosinophilic pustular folliculitis. *J Dermatol*. 2013; 40: 419-423.
5. Fukamachi S, Kabashima K, Sugita K, Kobayashi M, Tokura Y. Therapeutic effectiveness of various treatments for eosinophilic pustular folliculitis. *Act Derm Venereol.* 2009; 89: 155-159.
6. Laing ME, Laing TA, Mulligan NJ, Keane FM. Eosinophilic pustular folliculitis induced by chemotherapy. *J Am Acad Dermatol.* 2006; 54: 729-730.

CHICAGO DERMATOLOGICAL SOCIETY **CASE # 10**

Presented by Bailey Tayebi MD
Division of Dermatology, Loyola University Medical Center

UNKNOWN