Chicago Dermatological Society

April 2013
Monthly Educational Conference

Program Information
Continuing Medical Education Certification
and
Case Presentations

Wednesday, April 10, 2013
Stroger/Cook County Hospital
Sidney Barsky Lecture

Conference Host:
Division of Dermatology
Stroger Cook County Hospital
Chicago, Illinois

Program

Conference Locations

Stroger Cook County Hospital; 1900 W. Polk, Chicago
Main hospital – entrance corner of Ogden & Damen or through the CCH parking garage
Hektoen Institute – 627 S. Wood St., 1st Floor Lobby & Auditorium
Parking: Cook County Hospital Garage: entrance on Polk St.
Alternate Parking: Rush Medical Center - Harrison just west of Ashland

Registration Area – beginning at 8:00 a.m. (sign-in sheets, name badges, exhibitors)
Lobby area of the Hektoen Institute; 627 S. Wood

Please note: Protocol books will be distributed in the Dermatology Clinic, Main Hospital until 10:30 a.m., and then at the registration area. Registration will be located at the Hektoen Institute only. You may proceed directly to the clinic and register later, if you prefer.

Program Events

8:00 a.m.	Registration begins for all attendees Continental breakfast & visit with exhibitors *Hektoen Institute, 1st floor lobby*
9:00 a.m. - 10:00 a.m.	**Special Lecture** – *Hektoen Auditorium* "Nazi Medical Policy and How It Helped American Dermatology" *Walter Burgdorf, MD* Following decades in America in education, patient care, and as a chairman of dermatology, Dr. Burgdorf has spent the last several years in Germany as a medical editor and author, and has devoted considerable time and effort to understanding what happened to physicians during the Nazi era, and in particular the impact on American dermatologic education and research. We invite all members to take advantage of this special presentation.
9:30 a.m. - 11:00 a.m.	**Clinical Rounds** Patients; Slide & Poster Viewing *Dermatology Clinic "G", 2nd floor, main hospital; use elevator #1*
11:15 a.m. - 12:15 p.m.	**General Session** - *Hektoen Auditorium* BARSKY LECTURE: "The Histiocytoses" *Walter Burgdorf, MD*
12:15 p.m. - 12:45 p.m.	Box Lunches & visit with exhibitors *Hektoen Institute Lobby*
12:45 p.m. - 1:00 p.m.	**CDS Business meeting** – *Hektoen Auditorium*
1:00 p.m. - 2:30 p.m.	**Case Discussions** – *Hektoen Auditorium*
2:30 p.m.	**Meeting adjourns**

Mark the Date!

Next CDS monthly meeting – Wednesday, May 15, 2013 at the Rush University

Guest Speaker

WALTER BURGDORF, MD

Clinical Lecturer, Department of Dermatology; Ludwig Maximilian University Munich, Germany

Delivering the Sidney Barsky Lecture

Walter Burgdorf, MD is a 1969 graduate of the University of Wisconsin School of Medicine and Public Health in Madison. He is a world expert on a variety of dermatological conditions and skin cancer. Dr. Burgdorf is widely respected for his work in a variety of practices covering both clinical and pathological services related to skin. He is an academic writer, editor and translator of prodigious productivity. And he is co-author of one of the world's most widely used dermatology textbooks. Following dermatology residency at University of Minnesota Hospitals, he completed a dermatopathology fellowship, and was fortunate to work with Juan Rosai, MD, one of the world's foremost surgical pathologists, to pioneer a tumor detection process involving labeled monoclonal antibody stains. After a stint on the faculty at the University of Oklahoma, Dr. Burgdorf took a position as professor of pathology at the University of New Mexico School of Medicine, and in 1984, he was named chair of the department. In the mid 1990s, he moved back to his native Germany to take an academic position at Ludwig-Maximilian University in Munich, where he also worked in private practice. More recently, Dr. Burgdorf edited the 1,500-page Braun-Falco's Dermatology, started by Otto Braun-Falco, MD, a friend and earlier chairman at LMU. Using his expertise in translation, Dr. Burgdorf edited the fifth German edition as well as the third English edition. The sixth edition was published in 2012, as will the historical tome The Pantheon of Dermatology, which Burgdorf edited and translated.

CME Financial Disclosure: Dr. Burgdorf has no significant financial relationships to disclose.

Continuing Education Credit

Chicago Dermatological Society

"Chicago Dermatological Society Monthly Conference"

April 10, 2013 *Chicago, IL*

Participants must attend entire session to receive full credit. Please sign-in at the CDS registration table before you leave the conference. A certificate will be sent to you following the meeting. Also, we ask that you complete the evaluation form and return to the CDS registration table. The information collected as part of this process represents an important part of the CME planning process.

The Colorado Foundation of Medical Care (CFMC) will retain a record of attendance on file for six years. CFMC contact information: 303-695-3300, ext. 3372

JOINT SPONSOR STATEMENT

This Continuing Educational activity is Joint-sponsored by the **Colorado Foundation for Medical Care, Office of Continuing Education** and the **Chicago Dermatological Society**. **CFMC is accredited by the ACCME to provide continuing medical education for physicians.**

GOAL/PURPOSE

To broaden the clinical knowledge of dermatologists with respect to diagnostic and therapeutic options.

SESSION OBJECTIVES

Upon completion of sessions, participants will be able to apply new knowledge and skills in the area of physician learning.

1. Discuss the clinical manifestations of Langerhans cell disease and how knowledge in denidritic cell biology helps the doctor to understand histiocytic disorders.
2. Explain the spectrum of xanthogranulomatous disease of the skin.
3. Describe how to treat the cutaneious lesions of Langerhans cell disease.
4. Describe what eugenics, is, how it was misused by the Nazis.
5. Discuss the German medical profession during the Nazi era and how dermatologists who left Germany during this time influenced the development of dermatology in the United States.

.CREDIT STATEMENTS

CME CREDIT

This activity has been planned and implemented in accordance with the Essential Areas and policies of the Accreditation Council for Continuing Medical Education (ACCME) through joint sponsorship of the Colorado Foundation for Medical Care, Office of Continuing Education (CFMC OCE) and Chicago Dermatological Society. CFMC is accredited by the ACCME to provide continuing medical education for physicians.

The Colorado Foundation for Medical Care designates this Live Activity for a maximum of 4.5 *AMA PRA Category 1 Credit(s) ™*. Physicians should claim only the credit commensurate with the extent of their participation in the activity.

OTHER HEALTH CARE PROFESSIONALS

This educational activity has been planned and implemented following the administrative and educational design criteria required for certification of health care professions continuing education credits. Registrants attending this activity may submit their certificate along with a copy of the course content to their professional organizations or state licensing agencies for recognition for 4.5 hours.

DISCLOSURE STATEMENTS

All other members of the faculty and planning team have nothing to disclose nor do they have any vested interests or affiliations. **It is the policy of the Chicago Dermatological Society and Colorado Foundation for Medical Care (CFMC) that the faculty discloses real or apparent conflicts of interest relating to the topics of the educational activity.**

Case Presentations

We extend our sincere thanks to
Dr. Jordan Carqueville, Dr. Jesse Jiang, and Dr. Marin Sekosan
for their review of the histopathology.

Key location(s): Diffuse involvement **CASE 1**

Julia Kasprzak MD, David Reid MD, and Warren Piette MD

History

21 year-old African American man with a history of ulcerative colitis and pyoderma gangrenosum who was first seen regularly at Cook County Hospital in April 2012. Over the past year, he has been admitted eight times for the following stereotypic findings: sore throat, malaise, rash, fever, tachycardia, hypotension, and leukocytosis.

The first episodes occurred in February-March 2012. The patient was admitted to outside hospitals during that time. During one admission, he was reportedly diagnosed with DRESS secondary to doxycycline, which was being used as treatment for pyoderma gangrenosum. All hospitalizations at Cook County are outlined in Table 1 (page 6).

On August 16, 2012, he was admitted to our hospital with sore throat, malaise, rash, fever, tachycardia, hypotension, and leukocytosis. He was treated for suspected septic shock by the admitting team with IV fluid boluses and IV antibiotics. The rash began as facial and lower extremity edema, which progressed on hospital day three to pruritic, slightly tender vesicles/bullae in the periocular, perioral, and groin areas. Biopsy showed a necrotic epidermis overlying re-epithelialized epidermis with focal vacuolar change, papillary dermal edema, colloid bodies, and a mild superficial and deep perivascular lymphocytic infiltrate with a few, scattered eosinophils. It was felt that this presentation could be consistent with a generalized bullous fixed drug eruption secondary to the ibuprofen he took for fevers. The lesions resolved within one week without sequelae. Blood cultures from admission were negative.

On September 1, 2012, he was re-admitted for sore throat, malaise, fever, tachycardia, hypotension, and leukocytosis. He again was treated for suspected septic shock with antibiotics and fluids. He had no rash at that visit and was admitted for only 3 days. Blood cultures were again negative.

On September 18, 2012, the patient again developed sore throat and general malaise, with associated fever, tachycardia, hypotension, and leukocytosis. As before, his symptoms were treated as suspected septic shock. In contrast to previous presentations, his skin findings were immediately apparent, with diffuse edema and tender cutaneous erosions. The erosions, which started as bullae and vesicles, continued to progress to a Stevens Johnson/Toxic epidermal necrolysis (SJS/TEN) picture. The only mucosal sites involved were his lips. Skin biopsy showed full epidermal necrosis with superficial dermal lymphocytic infiltrate, sparse eosinophils, and pigment incontinence.

He was admitted to the Burn ICU for wound care and treated only with IV methylprednisolone. The decision was made to withhold antibiotics from his treatment regimen, as there was no evidence for a septic etiology of shock during any of his admissions. His symptoms resolved with steroids and wound care alone.

A thorough medication history investigation was performed during the September 18th hospitalization. He was questioned repeatedly, by multiple team members, on prescribed, over the counter, and illicit drug use. The patient consistently stated he was only taking dapsone (since June 2012), mesalamine (since 2006) and as needed diphenydramine for insomnia along with as needed ibuprofen for his ulcer pain. Toxicology screens were negative for illicit substances. Given the appearance of his

severe TEN-like cutaneous lesions, all medications were stopped due to concern for a drug-induced process.

On October 19, 2012, however, he again presented to the emergency room with sore throat, malaise, mild fever, hypotension, diffuse cutaneous erythema, mucosal lip erosions, and perorificial and lower extremity edema. The symptoms continued to progress after taking 60 mg oral prednisone; therefore, he was placed on IV methylprednisolone 500 mg for a three day pulse. The symptoms then resolved without progression.

On November 6, 2012, he presented with a low-grade fever, borderline hypotension, and mild tacychardia. He had facial edema and eye injection. These symptoms resolved with 90 mg of oral prednisone alone.

Since November, the patient has not experienced any further episodes. He is now taking the following medications: dapsone 150 mg daily, colchicine 0.6 mg BID, cyclosporine 150 mg BID, and azathioprine 150 mg daily. He has been slowly tapered off prednisone.

Other pertinent laboratory data
The following studies were negative/unremarkable:
All blood cultures, CMP, HIV, CMV, EBV, RPR, C3, C4, ANA
CT scan of abdomen/pelvis, toxicology screen

Diagnosis
For discussion

Discussion
This patient's constellation of findings have been difficult to unite under one diagnosis. A thorough literature review has produced no publications describing a stereotypic syndrome consisting of fever, hypotension, leukocytosis, and this bullous eruption with epidermal sloughing.

Nevertheless, we have thus far come to the following conclusions/hypotheses regarding his condition:

1. Though the patient develops a systemic inflammatory response syndrome (SIRS), it is not due to sepsis. His overall clinical course is less prolonged than septic shock, all cultures have been negative, and his clinical findings resolve with systemic steroids alone.

2. His long-term medications (dapsone/mesalamine/diphenydramine) do not cause the clinical presentation, as the symptoms continue to occur while he is off these treatments.

3. His presentation is not attributable to over-the-counter medications, supplements, or illicit drugs. Of note, there have been reports of generalized bullous fixed drug eruptions (FDE) that resemble TEN in patients taking naproxen. The patient's eosinophils were significantly elevated, and he did report taking naproxen. Consequently, it was originally thought that his cutaneous presentation could either be a widespread bullous fixed drug eruption or a manifestation of true TEN due to medication. However, the patient's symptoms continued to occur while he was taking only prednisone alone.

Our present hypothesis to explain the constellation of systemic and cutaneous findings involves an auto-inflammatory etiology possibly related to his ulcerative colitis/pyoderma gangrenosum.

Autoinflammatory diseases (AIDs) are characterized by unprovoked, recurrent inflammation in the absence of established circulating autoantibodies or antigen-specific T cell response. AIDs represent a wide disease spectrum ranging from Mendelian disorders to diseases like inflammatory bowel disease (IBD), which have a more polygenic mode of inheritance.

The immunopathogenesis of ulcerative colitis, a chronic inflammatory disorder, is currently thought to involve a genetic component. NOD2 (CARD15) is the first susceptibility gene identified in inflammatory bowel disease. NOD2 mutations have been described with a disease entity consisting of at least three of the following: periodic fevers skin disease (urticarial or granulomatous rash), inflammatory arthritis, serositis, and Sicca-like symptoms in the absence of autoimmune disease. In addition, other studies have shown the presence of MEFV gene variations in patients with IBD. MEFV, the gene responsible for Familial Mediterranean fever (FMF), encodes pyrin, which interacts with CARD. Both pyrin and NOD-2 play an important role in IL-1 activation. TNF-alpha along with IL-6 can also play a role in autoinflammatory diseases and are usually elevated in each acute inflammatory episode.

Although the genes associated with the periodic fever syndromes and NOD2 have been associated with cutaneous eruptions, none have been reported with a bullous eruption of this nature. Our patient may have genetic polymorphism causing his stereotypic symptoms. We are in the process of testing for cytokine and gene abnormalities related to the periodic fever syndromes and AIDs.

References

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	8/16/2013	9/1/2013	9/18/2013	10/19/2013	11/6/2013
Temperature (F)	101.5	102.8	101.2	99.2	99.8
Blood Pressure	80/40	90/50	80/40	90/50	105/55
Heart Rate	132	134	110	112	102
WBC [4.4 – 10.6 k/μL]	22.1	20.3	19.5	25	15.2
Eosinophil Count [0.0 - 0.4 k/μL]	0.6	2.2	0.2	0.2	0
Rash	Pruritus, facial edema at admission; day 3-- periorificial and inguinal vesicles/bullae Lesions resolved without sequelae.	No eruption	Facial edema, erosions, vesicles, present at time of admission. Progression to widespread epidermal detachment.	Facial edema, diffuse erythema, erosions on lips. (Presumably aborted due to treatment)	Facial edema, pruritus. (Presumably aborted due to treatment)
ROS	Sore throat/malaise	Sore throat/malaise	Sore throat/malaise	Sore throat/malaise	Malaise/no sore throat
Treatment	IV fluid and antibiotics	IV fluid and antibiotics	Methylprednisolone IV, dopamine, IV antibiotics, IV Fluid boluses, FFP	Methylprednisolone IV	90 mg prednisone

Table 1. Summary of symptoms at all hospitalizations at Cook County Hospital.

CHICAGO DERMATOLOGIC SOCIETY

Key locations: Upper and lower extremities **CASE 2**

Stephanie St. Pierre MD, Michelle Chevalier MD, and Warren Piette MD

History of Present Illness

51 year-old Hispanic man with a history of hemorrhoids was admitted with one week of rectal bleeding and an eruption on the extremities. The skin changes were initially asymptomatic. They started as a faint redness of the left ankle, which then progressed to involve both legs. The right wrist later became erythematous, stiff and painful.

Past Medical History

Hemorrhoids with hemorrhoidectomy
Low testosterone

Medications/Allergies

None/NKDA

Social History

No history of tobacco use or alcohol abuse. Uses marijuana but no other illicit drugs. Unemployed janitor. Lives with his mother.

Review of Systems

Positive for right eye redness for two weeks, intermittent bilateral wrist pain for one month, recurrent oral ulcers for one month, and ten-pound weight loss over one month. No fever, chills, night sweats, malaise, or genital ulcerations.

Physical Exam

Vital signs:	Afebrile, normotensive, normal heart rate
Eyes:	Medial right eye with scleral injection
Oral mucosa:	Numerous shallow grey ulcers on an erythematous base
Skin:	Ankles, anterior lower legs, knees and right forearm with non-tender, minimally indurated, erythematous plaques
Musculoskeletal:	Wrists with mild edema and tenderness to palpation; no synovitis

Laboratory Data

The following labs were remarkable/abnormal:

WBC	12.6 k/µL	[4.4 – 10.6 k/µL]
Neutrophils	9.2 k/µL	[2.2 – 6.9 k/µL]
Lymphocytes	0.6 k/µL	[1.2 – 3.4 k/µL]
Monocytes	1.1 k/µL	[0.2 – 0.8 k/µL]
Hemoglobin	12.1 g/dL	[12.9 – 16.8 g/dL]
ANA	>1:160, speckled	[<1:160]
Urinalysis	Protein 50, WBC 4, RBC 5	[0]
CRP	14.89 mg/dL	[0 – 0.5 mg/dL]
ESR	72 mm/hr	[0 – 26 mm/hr]

Histopathology

RIGHT WRIST, PUNCH BIOPSY:
Granulomatous dermatitis with nodular neutrophilic infiltrates extending to subcutis in a lobular pattern. No evidence of true vasculitis. Special stains for fungus (GMS) and acid-fast bacilli are negative.

Radiology
CT Abdomen/Pelvis: findings suggestive of ankylosing spondylitis

Colonoscopy
Multiple shallow ulcers throughout colon. Opening of ileocecal valve inflamed and ulcerated. Biopsy showed fragments of granulation tissue and fragments of colonic mucosa with marked chronic inflammation. Stains for fungal organisms and CMV were negative. No viral inclusions seen.

Diagnosis
Neutrophilic lobular panniculitis and noncaseating granulomas associated with Crohn's disease

Treatment and Course
The patient was treated with prednisone 40 mg daily and mesalamine 800 mg TID. Ophthalmology recommended steroid drops for nodular scleritis of the right eye. Two weeks after discharge from the hospital, azathioprine 50mg daily was added and mesalamine was increased to 1600 mg BID. A prednisone taper was planned, but the patient self-discontinued after two weeks on the 40 mg dose because the skin lesions had cleared. Three weeks after discharge, he noted improvement in joint symptoms and decreased amount of blood in the stool. He reported a recurrence of several nodules on the legs about two months after hospital discharge that resolved spontaneously after one week.

Discussion
Neutrophilic lobular panniculitis (NLP) has been associated with myelodysplasia, rheumatoid arthritis, alpha-1-antitrypsin deficiency, and inflammatory bowel disease. It may also occur in the initial phase of early panniculitis of many types, including Behcet's syndrome, pancreatic panniculitis, and factitial panniculitis. NLP with noncaseating granuloma formation has been reported as an id reaction to infection.

Crohn's disease has several associated skin findings including pyoderma gangrenosum, erythema nodosum, acrodermatitis enteropathica, Sweet's syndrome, erythema multiforme, epidermolysis bullosa acquisita, and metastatic Crohn's disease (MCD). Our patient's clinical findings initially seemed most suggestive of Sweet's syndrome (subcutaneous type), while the histopathologic findings suggested MCD.

The subcutaneous variant of Sweet's syndrome is a rare disease consisting of erythematous plaques and nodules in a symmetric distribution accompanied by fever and malaise. Histopathology shows a lobular and/or septal neutrophilic subcutaneous infiltrate without vasculitis. It is typically associated with myelodysplastic conditions.

MCD is a rare disease with cutaneous findings consisting of foci of noncaseating granulomatous inflammation that are non-contiguous with gastrointestinal disease. It may precede gastrointestinal involvement by years. The etiology is poorly understood with proposed mechanisms including circulating immune complexes or a T-lymphocyte-mediated type IV hypersensitivity reaction. The disease typically manifests as red-brown papules or plaques, erythematous nodules, or ulcers found most commonly on the legs and intertriginous areas but also described on the genitals, face, trunk, and upper extremities. Children may present with genital swelling. Characteristic histopathologic features of metastatic Crohn's disease include sarcoid-like noncaseating granulomas associated with multinucleated giant cells in the upper and lower dermis and sometimes in the fat. Other findings may include necrobiosis, leukocytoclasis and vasculitis, and large numbers of lymphocytes, plasma cells, and eosinophils.

NLP associated with Crohn's disease has been rarely reported in the literature. Yosipovitch et al reported a 69 year-old woman with Crohn's disease whose red shiny leg nodules demonstrated loosely formed noncaseating granulomas and lobular panniculitis with many neutrophils. The dermis

showed a sparse perivascular lymphohistiocytic infiltrate. These lesions resolved spontaneously without treatment. Ogawa et al reported similar clinical and histopathological findings in a 38 year-old woman whose gastrointestinal Crohn's disease and skin responded dramatically to infliximab. Haczell-Bradley et al reported tender, red papules and nodules on the legs of a 67 year-old woman with Crohn's disease. Histopathology demonstrated granulomatous lobular and septal panniculitis with a predominant neutrophilic infiltrate and leukocytoclastic vasculitis. There was no granulomatous reaction in the dermis. The lesions responded to prednisone and metronidazole but recurred several times. The authors of the Yosipovitch and Haczell-Bradley articles proposed that these presentations may represent cutaneous Crohn's disease.

References

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CHICAGO DERMATOLOGIC SOCIETY

Key location(s): Trunk **CASE 3**

Donna Sadowski MD, Julia Kasprzak MD, and David Reid MD

History of Present Illness

63 year-old Cantonese man with a history of non-small cell lung cancer, currently in remission, presented with six months of a diffuse, intensely pruritic rash. The eruption began on his legs and subsequently spread to involve his torso and upper extremities. For the past three years, he had been taking lingzhi mushroom supplement (a medicinal mushroom in traditional Chinese medicine thought to have anti-neoplastic and immunomodulatory effects). He had no other new medications other than hydroxyzine, prescribed by his primary care physician, which provided minimal symptomatic relief. He had no new topical exposures and no recent history of travel. He denied fever, chills, dyspnea, weight loss, malaise, gastrointestinal or upper respiratory illness.

At initial presentation, his rash consisted of edematous, mildly indurated, erythematous papules coalescing into plaques. He was instructed to stop taking lingzhi mushroom supplements, given a suspected drug eruption. Although he initially improved with triamcinolone 0.1% ointment topically to affected areas, the eruption recurred three months later.

Past Medical History

Non-small cell lung adenocarcinoma (T2N0M0, s/p left lower lobectomy in 2008, s/p 4 cycles of paclitaxel and carboplatin in 2008)

Medications/Allergies

Lingzhi mushroom supplement
Hydroxyzine
NKDA

Social History

50 pack-year history of tobacco use, no alcohol or drug abuse. Immigrated from China in 2003. Married. Works as a cook.

Physical Exam

Skin: Arms, legs, and trunk: edematous, reddish-brown, flat-topped papules coalescing into plaques, some with overlying excoriation.
Abdomen: red-brown, flat-topped papules with cobblestone-like appearance and well-defined linear sparing of folds.

Laboratory Data

The following labs were remarkable:

Eosinophils	0.4 k/μL	[0.0 - 0.4 k/μL]

CBC, CMP, HIV, and hepatitis panel were normal.

Histopathology

RIGHT UPPER BACK, SKIN, PUNCH BIOPSY:
Focal subacute spongiotic dermatitis with hyperkeratosis and a superficial perivascular and interstitial lymphocytic infiltrate with numerous eosinophils and dermal edema. Scattered lymphocytes present at the dermal-epidermal junction without dyskeratosis. The differential diagnosis includes contact dermatitis, eczematous dermatitis and drug eruption.

Radiology
CT chest/abdomen/pelvis: left sided lobectomy changes; no adenopathy, bony lesions, or visceral masses

Diagnosis
Papuloerythroderma of Ofuji

Treatment and Course
CT, as detailed above, was negative for local adenocarcinoma recurrence or metastatic disease. He was prescribed clobetasol 0.05% ointment, and then transitioned to triamcinolone 0.1% ointment. The lesions ultimately resolved, approximately one year after initial presentation.

Discussion
Papuloerythroderma of Ofuji (PEO) was originally described by Professor Shigeo Ofuji in 1984. Since then, approximately 170 cases have been reported in the literature. The majority of these are in Asian men over 55 years old, with a male-to-female ratio of 4:1. While the etiology is unclear, medications have been proposed as causative agents, with drug-reactive Th2 cells playing a major role in pathogenesis. Only 5% of PEO cases, however, have been reported to be drug-induced. Additionally, PEO has been associated with solid and hematological malignancies, the most common being cutaneous T-cell lymphoma and gastric carcinoma. Other associations include atopy and infections.

PEO clinically presents as widespread, erythematous-to-brown, flat-topped papules coalescing into plaques. These plaques often have a cobblestone-like appearance and are symmetrically distributed. Consistent features in all reported cases are pruritus and the presence of the "deck-chair" sign, in which the rash strikingly spares the skin folds. The eruption also spares the face, scalp, and mucous membranes. Additional findings consist of palmoplantar keratoderma (20%) and dermatopathic lymphadenopathy (25%). Laboratory abnormalities may also occur, including peripheral eosinophilia (85%), lymphocytopenia (29%), and elevated serum IgE (50%).

Histolopathologic findings, which are nonspecific, include epidermal spongiosis, acanthosis, and parakeratosis. There is often a mixed dermal infiltrate composed of lymphocytes and eosinophils, and occasionally scarce histiocytes and plasma cells.

Identification of PEO can be challenging. In a review by Torchia et al, the median duration of the cutaneous eruption at the time of diagnosis was seven months. Due to lack of recognition by clinicians, PEO is likely underreported. Treatment of PEO has yielded mixed results and includes topical and systemic corticosteroids, UVB, PUVA, cyclosporine, acitretin, and azathioprine.

References
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CHICAGO DERMATOLOGIC SOCIETY

Key location(s): Diffuse involvement **CASE 4a**

Michelle Chevalier MD and Warren Piette MD

History of Present Illness
17 year-old African American male presented with a four-day history of a diffuse, pruritic eruption. He noted initial progression of the eruption with stabilization over a two-day period. Two days prior to the eruption, he experienced diffuse itching followed by an episode of nausea and vomiting. The patient took diphenhydramine frequently for chronic sinus symptoms, but denied taking any new medications.

Past Medical History
Asthma
Childhood episodic fevers

Medications/Allergies
Diphenhydramine PRN
Albuterol inhaler PRN
NKDA

Social History
No alcohol, tobacco, or illicit drug use.

Review of Systems
Positive for a recent episode of nausea and vomiting, but denied the following symptoms: diarrhea, cough, throat pain, fever, chills, night sweats, weight loss, or oral/ocular/genital symptoms.

Physical Exam

General:	Well-appearing; alert and oriented. Afebrile.
Skin:	Neck, trunk, extremities (including palms/soles): diffuse minimally elevated, erythematous to purpuric papules and plaques; several lesions with dusky centers
Oral:	Lips: erythema, edema, and mild fissuring Soft/hard palate: two small petechiae
Eyes:	Normal appearing
Lymph:	No cervical or supraclavicular lymphadenopathy

Laboratory Data
The following labs were remarkable/abnormal at initial evaluation:

WBC	1.8 k/μL	[4.4 – 10.6 k/μL]
CRP	8.09 mg/dL	[0.00 – 0.50 mg/dL]

The following labs were remarkable/abnormal during or after admission:

Hemoglobin	11.4 – 12.6 g/dL	[11.5 – 14.8 g/dL for age 12-18]
Eosinophil count	0.6 – 0.7 k/μL	[0.0 - 0.4 k/μL]
Platelet count	478 – 527 k/μL	[161 – 269 k/μL]
CRP	0.70 - 7.01 mg/dL	[0.00 – 0.50 mg/dL]
ESR	63 mm/hr	[0-10 mm/hr]
Urinalysis	30 protein, 1 RBC, 2 WBC	[Negative]
HLA-B27 typing	Positive	[Negative]

Histopathology

LEFT UPPER ARM, PUNCH BIOPSY:
Focal vacuolar interface dermatitis with superficial perivascular and interstitial infiltrate comprised of lymphocytes, neutrophils, and scattered nuclear dust.

Radiology:

Chest radiograph: bilateral central peribronchial thickening

Transthoracic echocardiogram: normal

EKG:

Sinus rhythm with 1st degree AV block (bigeminy with frequent PVCs noted on continuous cardiac monitoring)

Diagnosis

Kawasaki disease

Treatment and Course

The patient was initially treated symptomatically with hydroxyzine and triamcinolone 0.1% ointment. On the third day of admission, he developed a fever of 103ºF and increased malaise. He continued to have intermittent fevers for two days and was discharged two days following defervescence. Three days later in dermatology clinic, his cutaneous lesions were resolving, but he had developed conjunctival and scleral injection along with anterior cervical lymphadenopathy. He continued to report malaise but denied any fevers after discharge. Laboratory examination revealed a normalized eosinophil count, down trending CRP and ESR, and new thrombocytosis. A repeat echocardiogram was unremarkable.

The patient returned four days later with worsening of his eye redness, photophobia, and new onset joint pain. Ophthalmologic examination revealed anterior uveitis. These symptoms prompted admission to the pediatric service for treatment of presumed incomplete (atypical) Kawasaki's disease despite the absence of prolonged fever. The patient refused intravenous immunoglobulin due to religious beliefs; therefore, methylprednisolone IV and high-dose aspirin were initiated. The CRP normalized three days later and a repeat echocardiogram was unremarkable. Diffuse palmoplantar desquamation was noted at a follow-up visit one month after discharge.

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Key location(s): Diffuse involvement **CASE 4b**

Julia Kasprzak MD, Michelle Chevalier MD, and David Reid MD

History of Present Illness
14 year-old Mexican young man presented with a rash that started four days prior to admission. The eruption began on his arms and subsequently extended to the rest of his body. He also reported sore throat, nausea, vomiting, and subjective fever/chills. He could not recall any sick contacts or insect bites and had no recent travel.

Past Medical History
None

Medications/Allergies
None/NKDA

Social History
No alcohol, tobacco, or illicit drug use.

Review of Systems
Intermittent headache, alleviated by acetaminophen. One episode of chest pain after admission. No loss of consciousness, visual changes, or arthritis.

Physical Exam

Vitals:	T 101.1° F, P 90, BP 105/58, RR 20/min
Skin:	Face, extremities, trunk: confluent erythematous and violaceous, nonblanching macules and patches
	Palms, soles: scattered violaceous macules, minimal edema of digits
	Genitalia: small erosion at urethral meatus and diffuse scrotal erythema with scaling
	Perineal region: clear, no erythema
Oral:	Lips: diffuse erythema and fissuring
	Hard palate: mild erythema and scattered petechiae
	Tongue: mildly edematous and erythematous with prominent papillae
Eyes:	Conjunctival injection
Lymph:	Right superior posterior cervical chain: 1.5 cm lymph node. No anterior cervical, supraclavicular, axillary, or inguinal LAD

Laboratory Data
The following labs were remarkable/abnormal:

Platelet count	127 k/μL	[161 – 269 k/μL]
CRP	3.09 mg/dL	[0.00 – 0.50 mg/dL]
ESR	40 mm/hr	[0-10 mm/hr]
Total cholesterol	86 mg/dL	[130-240 mg/dL]

The following laboratory studies were negative/normal:
Blood and throat cultures, urinalysis, hepatitis panel, Cocksackie B1 antibody, Parvovirus B19 antibody, antistreptolysin titer, CK, troponin-I, ANA, and anti-centromere antibody

Other Studies:
Transthoracic echocardiogram: normal

EKG: normal

Treatment and Course:

Although the patient was older than the typical age range reported with Kawasaki disease, he did fulfill the American Heart Association (AHA) 2004 criteria for Kawasaki disease, including 5 days of fever along with the following:

1. bilateral nonsuppurative conjunctivitis
2. classic mucous membrane changes including fissured lips and strawberry tongue
3. a dysmorphic rash
4. edema/erythema of the hands and feet along with desquamation
5. acute, nonpurulent cervical lymphadenopathy

Laboratory findings of thrombocytopenia, elevated CRP/ESR, and hypocholesterolemia also supported a diagnosis of Kawasaki disease.

The decision was made to initiate treatment with IVIG and high dose aspirin. He experienced resolution of fever, sore throat, nausea, and vomiting within 24 hours of starting therapy. Initial echocardiogram was normal, and the patient is scheduled for a follow-up echocardiogram in the near future. One week after discharge, he remained on 81 mg of aspirin daily and his cutaneous symptoms were almost completely resolved, with only residual desquamation.

Discussion

Kawasaki disease is an acute febrile systemic vasculitis with potentially fatal cardiac complications, most often affecting children under the age of five. These two cases highlight presentations of Kawasaki disease in adolescents. Both cases had fever and cutaneous findings, including a polymorphous rash, classically found in Kawasaki disease. However, the patient in case A did not meet the criteria for 5 days of fever. These cases showcase that it is important to consider a diagnosis of Kawasaki disease in patients who are older than five and who do not meet the strict fever criteria.

Both of the patients were adolescent young men, ages 17 and 14. Later-onset Kawasaki disease is rare, but at least 81 cases have been reported in adulthood. In comparison to small children, older patients present more frequently with cervical adenopathy, hepatitis, and arthralgia, while they are less often affected by meningitis, thrombocytosis, and coronary aneurysms (5% in adults vs. 20% in children). The polymorphous rash of Kawasaki disease is similar in both children and adults; morbilliform, erythema multiforme-like, urticarial, scarlatiniform, and pustular presentations have been reported.

Fever of five days is listed in the AHA guidelines as a requirement for the diagnosis of Kawasaki disease. However, cases of coronary aneurysm in the absence of preceding fever have been described, which calls into question the obligatory inclusion of fever for the diagnosis of Kawasaki disease. In fact, in Japan, criteria are similar, with the important exception that fever is not required to make the diagnosis (Japanese Circulation Society, 2008 criteria).

Although patient A did not have fever of at least five days duration, our clinical suspicion remained high for Kawasaki disease given the following co-existent clinical signs: diffuse skin eruption including palmoplantar involvement, lip edema and fissuring, anterior uveitis, markedly elevated inflammatory markers, thrombocytosis, anemia, and electrocardiogram abnormalities. Peripheral eosinophiliia, as seen in this patient, has also been reported to be relatively common, compared to febrile control patients.

Additionally, an "incomplete" form of Kawasaki disease in older patients has been reported. A diagnosis of incomplete (atypical) Kawasaki disease may be made if the patient demonstrates fever of ≥ five days in association with elevated CRP (≥ 3.0 mg/dL) or ESR (≥ 40 mm/hr), and at least 3 supplemental criteria. These include the following: (1) albumin ≤ 3.0 g/dL, (2) anemia, (3) elevation of alanine aminotransferase, (4) platelets after 7 days ≥ 450,000/mm^3, (5) white blood count ≥ 15,000/mm^3, (6) urine ≥ 10 WBC/high-power field. In suspicious cases lacking elevated

inflammatory markers and falling short of supplemental criteria, an echocardiogram may be ordered to assess for evidence of coronary involvement.

After our experience with these two cases, we suggest two important points to consider during the evaluation of a patient who may have signs and symptoms of Kawasaki disease. First, Kawasaki disease is not limited to young children. Second, current criteria for the diagnosis of Kawasaki disease may be too strict especially with duration of fever, leading to underdiagnosis. By considering Kawasaki disease on the differential of cases that do not meet strict criteria, potentially devastating cardiac sequelae may be avoided.

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CHICAGO DERMATOLOGIC SOCIETY

Vignette A

What is the unifying diagnosis?

Please place your answer in the ballot box.

Vignette B

What is the unifying diagnosis?

Please place your answer in the ballot box.

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Vignette C

What is the unifying diagnosis?

Please place your answer in the ballot box.

CHICAGO DERMATOLOGIC SOCIETY

Key locations: Helical rim of the ear and earlobe **CASE 6**

Sumul Gandhi MD, Joerg Albrecht MD, and George Engel MD

Patient A

History of Present Illness

20 year-old Hispanic man presented to clinic with an enlarging, pruritic mass on the right earlobe. The lesion started two years after having his ear pierced nonprofessionally.

Physical Exam

Right earlobe: Well-defined, firm, nontender 1.9 x 2.4 x 4.0 cm nodule

Diagnosis

Right earlobe keloid

Treatment and Course

The patient opted for excision of his earlobe keloid. After administration of local anesthesia (1% lidocaine with epinephrine), the surgical site was prepped and draped. A wedge excision of the keloid was performed, leaving a triangular defect with sharp opposing edges. A single vertical mattress stitch using 5-0 nylon suture was placed to align the most distal aspect of the lobule. The remaining wound edges were approximated with simple interrupted stitches using 6-0 nylon.

Eleven days later, the patient presented for suture removal and intralesional steroid treatment. The wound edges were well approximated and there was no discomfort or pruritus of the surgical site. At the time of suture removal, 1mL of 40mg/mL triamcinolone acetonide was injected to the wound. The patient returned for monthly intralesional triamcinolone acetonide therapy with no recurrence after 6 months.

Patient B

History of Present Illness

85 year-old African American woman presented to clinic with a 50-year history of a large, non-painful, non-pruritic firm plaque on the left ear with extension onto the preauricular cheek and postauricular skin. Prior treatments included excision and subsequent intralesional triamcinolone acetonide, but the plaque recurred after each treatment.

Physical Examination

Right ear: 10 x 7 cm firm keloidal plaque involving the majority of earlobe and extending to the pre- and postauricular skin

Diagnosis

Recurrent right earlobe keloid extending onto pre- and postauricular surfaces

Treatment and Course

After discussion of the treatment options, the patient opted for surgical excision with post-operative radiation therapy. Local anesthesia was administered (1% lidocaine with epinephrine) and the surgical site was prepped and draped. The keloidal tissue was excised tangentially from the pre- and postauricular surfaces and the anterior lobule. Hemostasis was achieved with electrocautery. Nine 4-0 polypropylene simple interrupted sutures were used to approximate the wound edges at the preauricular cheek and earlobe, while the remainder of wound was allowed to heal by secondary intention.

Immediately after the procedure, the patient was started a five day course of radiation therapy to the surgical site to prevent recurrence of the keloid.

Sutures were removed on post-operative day 12. Three weeks after completion of radiation therapy she was started on monthly intralesional triamcinolone acetonide therapy, which she continued for 5 months. To date, she shows no signs of recurrence.

Patient C

History of Present Illness

31 year-old Hispanic woman presented to clinic with a one-year history of an enlarging mass on the right helix. The lesion developed three months after an ear piercing and was occasionally painful and pruritic.

Physical Exam

Helix of right ear: 1 x 2 cm pink, firm, nontender nodule

Diagnosis

Keloid of the right ear helix

Treatment and Course

The patient initially underwent a trial of intralesional triamcinolone therapy. Eventually, she opted for excision after intralesional triamcinolone acetonide failed to alleviate the symptoms and reduce the size of the keloid. A keloid fillet flap technique was chosen to preserve the integrity of the helical rim. Local anesthesia was administered (1% lidocaine with epinephrine) and the surgical site was prepped and draped. The posteromedial margin of the keloid was incised with a 15 blade and the overlying epidermis and superficial dermis was dissected from the underlying keloidal tissue. The keloid was separated from the normal underlying dermal tissue. The flap was trimmed of excess tissue and sutured into place over the helical rim using superficial interrupted 5-0 polypropylene sutures, recreating the contour of the helical rim.

The patient returned for suture removal seven days later. Monthly intralesional triamcinolone acetonide injections were started two weeks postoperatively. After five monthly injections and no signs of recurrence, the patient was lost to follow-up.

Discussion

Keloids represent aberrant proliferation of scar tissue in response to cutaneous injury. The altered wound healing process is characterized by hyperproliferation of fibroblasts with subsequent deposition of disorganized collagen. These lesions characteristically outgrow sites of cutaneous injury and can be severely disfiguring.

Keloids occur 15 times more frequently in patients with skin of color. Earlobe keloids are most commonly a result of ear piercings. One pediatric study showed that keloids are more likely to occur if piercing occurs after eleven years of age. Keloids are usually symptomatic. Nearly 80% of patients report pruritus and almost 50% report pain.

While the optimal approach is prevention and avoidance of unnecessary surgery or trauma in patients who tend to form keloids, a number of treatment modalities exist. Non-excisional treatments are often used in concert with excisional therapy. Examples of non-excisional therapy are intralesional (corticosteroids, fluorouracil, verapamil), topical (imiquimod, calcineurin inhibitors, corticosteroids), radiation, laser, cryosurgery, and pressure earrings.

Surgical excision may be considered with failure of non-excisional therapy or when the keloid is unlikely to respond well to non-excisional approaches. As recurrence rates range from 45 to 100% in patients treated with excision alone, combination therapy is essential and has been shown to reduce recurrence rates to less than 50%.

While posterior earlobe keloids can be simply excised and allowed to heal by secondary intention, special consideration should be given to keloids in areas of high cosmetic concern, like the anterior earlobe or helical rim.

In treating full-thickness earlobe keloids that involve the central lobe, Music and Engel described the wedge excision. With careful excision of all palpable keloid during the procedure and dosing immediate postoperative and monthly treatments of intralesional triamcinolone acetonide, zero of twelve patients had signs of recurrence at follow-up periods ranging from three to sixteen months.

The keloid fillet flap is useful in the excision and repair of keloids on the lobule or helical rim, where maintaining the normal architecture of the ear may otherwise be difficult (refer to Patient C "Treatment and Course" for description of procedure). Benefits of this approach include maintenance of normal architecture, a faster recovery time, and the ability to avoid secondary intention healing, which many suggest is related to contracture at the surgical site and eventual recurrence. Additionally, the flap can be used to close large defects, with less color mismatch and no donor site morbidity as compared to a skin graft.

Postoperative radiation therapy should be considered in the treatment of recurrent keloids. While the potential long-term risk of malignancy associated with radiation therapy for a benign condition limits its widespread use, only six cases of post-radiation malignancy have been reported. Most studies have found radiation alone to be effective in reducing recurrence after keloid excision, with long-term keloid-free rates ranging from 70-90%. In a study of 80 keloids that failed initial excision, intralesional triamcinolone acetonide, and other treatments, immediate postoperative single-fraction radiotherapy resulted in lower recurrence rates to 9% at one year and 16% at five years after excision.

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CHICAGO DERMATOLOGIC SOCIETY

Key location(s): Face, chest, abdomen, arms **CASE 8**

Elizabeth Fahrenbach MD and Warren Piette MD

History of Present Illness

60 year-old Pakistani man presented to the hospital with three days of fever, odynophagia and decreased oral intake as well as increasing pain of skin lesions on his chest, arms and face. He is known to have borderline lepromatous leprosy and erythema nodosum leprosum. He originally presented to the dermatology clinic in 2008. He was not on ideal treatment for leprosy due to medication intolerances and variable compliance with long absences from our clinic.

Past Medical History

Borderline lepromatous leprosy with erythema nodosum leprosum
Hypertension
Gout
Benign prostatic hypertrophy
Asthma

Medications/Allergies

Prednisone 20 mg daily
Rifampin 600 mg daily
Minocycline 100 mg daily
NKDA

Social and Family History

No tobacco, alcohol or illicit drug use. Educated as an engineer, currently unemployed. Brother with history of leprosy.

Review of Systems

Positive for fever, cough, sore throat and malaise. No weakness, tingling, or eye complaints.

Physical Exam

General	Vital signs normal
	Lying in bed, mildly uncomfortable, speaking with difficulty
Skin	Leonine facies
	Erythematous, edematous plaques, some studded with pustules
Oropharynx	Edema and intense erythema of posterior oropharynx
	Bilateral anterior cervical lymphadenopathy
Neurologic	No motor or sensory deficits

Laboratory Data

The following labs were remarkable/abnormal:

Hemoglobin/Hematocrit	8.0 g/dL / 24.6%	[12.9 – 16.8 g/dL / 38.1-49%]
White blood cells	14.1 k/μL	[4.4 – 10.6 k/μL]
Neutrophils	91%	[45-70]
Total protein	4.9 g/dL	[6.5-8.2]
Albumin	2.4 g/dL	[3.9-5.0]

Microbiology

The following studies were normal: culture oropharnyx, Monospot, HIV, ELISA for EBV and CMV, HSV culture

Radiology
Chest radiograph: normal

Histopathology
LEFT UPPER ARM, PUNCH BIOPSY:
Granulomatous dermatitis admixed with lymphocytes and neutrophils with an overlying epidermal pustule. Dermal edema, scattered perivascular extravasated erythrocytes, nuclear dust and vasculopathy suggestive of early leukocytoclastic vasculitis. Fite stain demonstrates scattered acid fast bacilli. AFB stain revealing rare acid fast bacilli. GMS and PAS stains for fungus negative. No polarizable material identified.

Diagnosis
Pustular erythema nodosum leprosum

Treatment and Course
On admission, the patient was given IV fluid rehydration and treated with IM penicillin for suspected streptococcal pharyngitis. Dermatology was consulted. Punch biopsies were taken to confirm the diagnosis of erythema nodosum leprosum, pustular variant. He continued on rifampin and minocycline, and his dose of prednisone was increased to 60 mg daily. He was discharged on hospital day six with plans for close clinic follow-up. However, prior to his clinic appointment he returned to Pakistan and initiated a course of clofazimine. When he returned to the Stroger dermatology clinic three months later, he was doing well with the addition of clofazimine 100 mg daily to his multidrug therapy. This regimen was continued and his care was eventually transferred to the UIC leprosy specialty clinic.

Discussion
Erythema nodosum leprosum is a dermal hypersensitivity reaction seen in lepromatous and borderline lepromatous leprosy, resulting from immune complex deposition with elevated levels of TNF-α within the lesions. It is associated with a high bacterial index with response to treatment. Constitutional symptoms and tender erythematous plaques and nodules on the face and extremities are common. Other manifestations include neuritis, lymphadenopathy, iritis, orchitis, glomerulonephritis, dactylitis and arthritis. While tender plaques are the usual cutaneous manifestation, in rare cases the lesions may take hemorrhagic, vesicular, erythema-multiforme-like, pustular or ulcerated forms. Bullous lesions are more commonly reported among the ENL variants, while reports of pustular lesions are rare.

ENL may be triggered by illness, surgery, pregnancy, parturition, lactation, menstruation, trauma, vaccination, and physical or mental stress. It may be precipitated by drugs such as iodides, bromides, dapsone and chaulmoogra (a traditional Eastern medicine remedy for leprosy). Initiation of therapy or the introduction of new drugs to an already established regimen, such as the highly bactericidal ofloxacin, may lead to large numbers of dead bacilli, thus precipitating an ENL reaction.

Severe erythema nodosum leprosum is defined by the WHO to include numerous nodules with high fever, the presence of nodules and neuritis, ulcerating and pustular variants, recurrent episodes of ENL or the involvement of other organs (i.e. eyes, testes, lymph nodes, joints). In all cases, continuation of multi-drug therapy is recommended. Management with corticosteroids is standard, but the addition of clofazimine is recommended for cases not responding satisfactorily to treatment with steroids alone or where the risk of corticosteroid toxicity is high. In cases of severe ENL where the use of corticosteroids is contraindicated, clofazimine may be the only agent added to the standard multidrug therapy.

A randomized study compared the efficacy of thalidomide to corticosteroids in the treatment of moderate to severe ENL. In that study, patients taking thalidomide showed a quicker clinical response and a lower number of relapses or recurrences than patients taking prednisolone. While a greater frequency of adverse effects was associated with prednisolone, peripheral neuropathy was seen more frequently in patients taking thalidomide, though thalidomide neuropathy can be difficult to distinguish from leprosy neuropathy. Thalidomide is not currently included in the WHO guidelines due to its teratogenicity.

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CHICAGO DERMATOLOGIC SOCIETY

Key location(s): Lower legs **CASE 9**

Shilpa Mehta MD, Kavita Menon MD, and Warren Piette MD

History of Present Illness

57 year-old Pakistani woman with no significant past medical history presented with a two month history of multiple indurated, tender lesions on her legs. She complained of subjective fevers but denied any cough, hemoptysis, weight loss, night sweats, loss of appetite, or gastrointestinal symptoms. She reported no recent illnesses and had not started any new medications. The patient has lived Chicago for the last six years, and she last traveled to Pakistan three years ago.

Medications/Allergies

None/NKDA

Social History

No history of tobacco, alcohol or illicit drug abuse. Works as a homemaker and lives with her husband.

Physical Exam

Skin:	Anterior lower legs and posterior calves: multiple 1 to 2 cm erythematous to brown, tender nodules
Mucosa/ Nails:	Normal appearing

Laboratory Data

The following labs were remarkable/abnormal:

ESR	97 mm/hr	[0-10 mm/hr]
CRP	6.12 mg/dL	[0.00 – 0.50 mg/dL]
Tuberculin skin test (48 hours)	18 mm	< 5mm

Histopathology

RIGHT POSTERIOR LEG, DEEP INCISIONAL BIOPSY:
Necrotizing granulomatous inflammation involving the dermis and subcutaneous adipose tissue. Perivascular mixed infiltrates with giant cells, endothelial swelling of deep vessels suggestive of early vasculitis. Special stains for GMS, PAS and AFB were negative. No polarizable material identified.

Microbiology

RIGHT POSTERIOR LEG, TISSUE CULTURE AND PCR:
Suspected acid fast bacilli were noted, but Ziehl Neelsen stain was negative. PCR- sequencing assay was negative for acid fast bacilli.

Radiology

Chest radiograph: normal

Diagnosis

Erythema induratum of Bazin secondary to prior exposure to tuberculosis

Treatment and Course

The patient developed similar painful nodules on the lower abdomen and right thigh after her initial visit. For treatment of latent tuberculosis, she was started on four-drug antitubercular therapy (rifampin, isoniazid, pyrazinamide, ethambutol) with regular monitoring of liver function tests. She is being closely followed by the direct observation therapy nurse every week and is compliant with her treatments. No side effects have been reported. The nodules subsided with post inflammatory hyperpigmentation within eight weeks of initiating treatment.

Discussion

The term erythema induratum was first coined by Bazin to describe a nodular eruption occurring on the lower legs of young women with tuberculosis (TB). He classified the eruption as an "erythematous benign scrofulid", which was thought to be tuberculous in origin because of its co-existence with pulmonary TB. This causal relationship between erythema induratum and TB was later questioned and several reports emerged of patients with a similar clinical picture without positive tuberculin skin tests or known exposure to TB. In 1945, to address this debate, Montgomery et al introduced the term nodular vasculitis to differentiate the lesions of erythema induratum of non-tuberculous origin from those of tuberculous origin, also known as erythema induratum of Bazin (EIB). Today, many authors classify this disease as a lobular panniculitis with two variants: EIB (TB-related) and nodular vasculitis (TB-unrelated).

Clinically, EIB, like nodular vasculitis, is characterized by tender, erythematous to violaceous nodules and plaques that most often occur on the lower legs, preferentially on the posterior or anterolateral aspects. Lesions have also been reported on the feet, thighs, buttocks, and arms. They are persistent and frequently develop focal ulceration and drainage. They heal with scarring and post inflammatory hyperpigmentation, and they are prone to recurrence.

EIB is associated with *Mycobacterium tuberculosis* (MTB). Therefore, patients may have clinical and radiographic evidence of active tuberculosis or evidence of latent tuberculosis with a positive tuberculin skin test or Quantiferon-TB Gold test. MTB DNA may be isolated by polymerase chain reaction (PCR) from skin lesions, though recovery ranges from 0 to 77 percent. However, the inability to detect MTB DNA by this method does not exclude the diagnosis of EIB. The diagnosis is typically confirmed by clinical presentation, histopathologic findings, a positive tuberculin skin test or Quantiferon TB-Gold test, and response to anti-tubercular therapy.

On histopathology, erythema induratum is described as a lobular or mixed septal panniculitis with a mixed inflammatory infiltrate comprised of neutrophils, lymphocytes, macrophages, and multinucleated giant cells. Vasculitis is present in the vast majority of cases, and most frequently involves veins or arteries of connective tissue septae and small venules of the fat lobules. Caseating necrosis may be present, and the degree of necrosis is greater in those cases that are positive for MTB DNA by PCR.

Management of erythema induratum should be directed at treating the underlying cause, if found. All patients should receive a tuberculin skin test or Quantiferon TB-Gold test. Patients found to have EIB with evidence of active or latent TB should receive multi-drug antitubercular therapy with rifampin, isoniazid, pyrazinamide and ethambutol for two months, followed by rifampin and isoniazid for 4 months under the care of an infectious disease specialist. Other supportive therapies include NSAIDs, rest, elevation, and compression.

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Vignette D

What is the unifying diagnosis?

Please place your answer in the ballot box.