Chicago Dermatological Society

December 2015
Monthly Educational Conference

Program Information
Continuing Medical Education Certification
and
Case Presentations

Wednesday, December 2, 2015
Gleacher Conference Center

Conference Host:
Section of Dermatology
University of Chicago Hospitals
Chicago, Illinois

Program

Host: University of Chicago
Wednesday, December 2, 2015
Gleacher Conference Center
450 N. Cityfront Plaza Dr., Chicago

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

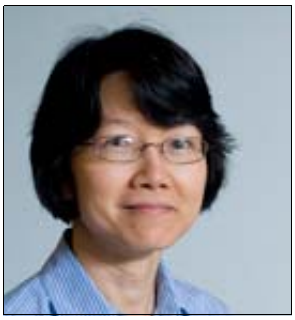
8:00 a.m.	**Registration & Continental Breakfast with Exhibitors** *6th Floor Lobby, Room 600 & North Foyer*
8:30 a.m. - 10:00 a.m.	**Clinical Rounds** Posters – *Room 600 (available throughout the morning)* Slide viewing – *Room 602 (available throughout the morning)*
9:00 a.m. - 10:00 a.m.	**Resident/Basic Science Lecture** – *Room 621* *MEDENICA LECTURE:* "Role of Immunohistochemistry in the Screening of Genetic Syndromes and Detection of Targetable Tumor Mutations" *Mai P. Hoang, MD*
10:00 a.m. - 10:30 a.m.	**Break and Visit with Exhibitors** – *Room 600/North Foyer*
10:30 a.m. - 12:00 p.m.	**Resident Case Presentations & Discussion** *Room 621*
12:00 p.m. - 12:15 p.m.	**MOC Self-Assessment Questions** *Room 621*
12:15 p.m. - 12:45 p.m.	**Box Lunches & visit with exhibitors** *Room 600/North Foyer*
12:45 p.m. - 1:00 p.m.	**CDS Business Meeting** *Room 621*
1:00 p.m. - 2:00 p.m.	**General Session** – *Room 621* *LORINCZ LECTURE*: "Current Issues in Vulvar Pathology" *Mai P. Hoang, MD*
2:00 p.m.	**Meeting adjourns**

Mark the Dates!
Next CDS monthly meeting – President's Conference and Annual Awards Luncheon
Wednesday, February 24, 2016 at the Stephens Conference Center in Rosemont

IDS Practice Management Conference – Wednesday, January 27, 2016 in Rosemont

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

Guest Speaker



MAI P. HOANG, MD

Dermatopathologist, Pathology,
Massachusetts General Hospital
Associate Professor, Pathology,
Harvard Medical School
Boston, MA

Delivering the Medenica & Lorincz Lectures

Dr. Hoang completed her medical and dermatological training at the University of Texas Southwestern Medical School, followed by fellowship at Duke University Medical Center and the University of Texas - MD Anderson Cancer Center. She is a Board-certified Anatomic/Clinical Pathologist and Dermatopathologist.

Dr. Hoang is an anatomic pathologist specializing in dermatopathology. She is the medical director of the Pathology immunohistochemistry laboratory at Mass General Hospital. She is the course director of the Harvard Combined Dermatopathology Update course held annually in Boston. She lectures at the annual American Society of Dermatopathology meetings, annual International Society of Dermatopathology meetings, and annual United States and Canadian Academy of Pathology meetings. She has published more than 120 articles, and has authored and edited three books: "Melanocytic Lesions: a Case Based Approach," "Vulvar Pathology," and "Cutaneous Hematopathology."

CONTINUING MEDICAL EDUCATION CREDITS

Chicago Dermatological Society

Presents

"Chicago Dermatological Society Monthly Meeting Series"

December 2, 2015 *Chicago, IL*

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

JOINT SPONSOR STATEMENT

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

GOAL / PURPOSE

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

EDUCATIONAL OBJECTIVES

Upon completion of this series of activities, participants will be able to:

1. Discuss key factors in the diagnosis and treatment for viral diseases and bacterial infections.
2. Describe the manifestation of skin cancers and the efficacy of treatments available to the dermatologist.
3. List the therapeutic options available to the dermatologist for a variety of skin diseases, both medical and surgical, and discuss how new emerging treatments can be successfully incorporated into a dermatology practice.

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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.

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

Resident Case Presentations

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CASE #1

PRESENTERS

Alex Means, MD; Christopher R Shea, MD; Aisha Sethi, MD

HISTORY OF PRESENT ILLNESS

A 38 year-old Mexican-American male presented to his primary care physician's office complaining of a new abrupt-onset facial rash of 3 days' duration, associated with worsening swelling, fever, chills, fatigue, malaise, dyspnea, dysphagia, sore gums, and nausea. His wife and 2 children, aged 5 and 7, were not affected with similar. He was seen 1 week prior for low back pain thought to be muscle spasm from heavy lifting and started on cyclobenzaprine, which he took for 4 days with resolution of symptoms and discontinued on the day of rash onset. He also received an inactivated influenza vaccine at his initial low back pain appointment.

PAST MEDICAL HISTORY

Autoimmune pancreatitis complicated by splenic vein thrombosis and splenectomy leading to gastric varices and chronic upper gastrointestinal bleeds with resulting iron deficiency anemia; controlled type 2 diabetes mellitus. He denied tobacco or other drug use aside from rare alcohol ingestion, and worked primarily as a landscaper and occasional repossession agent.

FAMILY HISTORY

Diabetes mellitus in mother, solid organ cancers and strokes in maternal extended relatives, alcoholism in paternal extended relatives.

MEDICATIONS

Metformin 1000mg daily, rare naproxen as needed (last use > 6 months prior to presentation)

ALLERGIES

No known drug; pine oil and feline/equine products per allergic prick testing

PHYSICAL EXAMINATION

A toxic, actively rigoring 38 year-old male with numerous edematous well-demarcated pink-red plaques studded with pustules throughout his face and scalp, with diffuse non-pitting 2-3+ edema on his bilateral cheeks and desquamative gingivitis
Vitals: Tmax of 103.8F, HR 110's, BP 129/77, SpO2 98% (RA), accucheck blood glucose 106.

DERMATOPATHOLOGY

A perivascular and perifollicular inflammatory infiltrate was present in the dermis, with intrafollicular collections of neutrophils. The gram stain highlighted gram-positive organisms within the follicular pustule.

LABORATORY DATA

Complete blood count: leukocytes 27.4 K/μL (3.5-11), hemoglobin 14.5g/dL (13.5-17.5), MCV 83.3fL (81-99), platelets 368 K/μL (150-450).
Differential: Neutrophils 73% (39-75), lymphocytes 17% (16-47), monocytes 10% (4-12), eosinophils 0% (0-7), basophils 0% (0-2).

Comprehensive metabolic panel: glucose 118mg/dL (60-109), sodium 136mEq/L (134-149), potassium 4.1mEq/L (3.5-5.0), chloride 96mEq/L (95-108), carbon dioxide 24mEq/L (23-30), anion gap 16mmol/L (6-15), BUN 11mg/dL (7-20), creatinine 1.2mg/dL (0.5-1.4), calcium 9.1mg/dL (8.4-10.2), total bilirubin 0.8mg/dL (0.1-1.0), total protein 7.5g/dL (6.0-8.3), albumin 3.9g/dL (3.5-5.0), alk phos 102U/L (30-120), AST 28U/L (8-37), ALT 44U/L (8-35)
Urinarlysis: pale yellow, clear, 1.013 specific gravity (1.016-1.022), pH 5.5 (5-9), negative leukocyte esterase/nitrites/protein/blood/glucose/bilirubin, 2+ ketones (normal negative), 0.2g/dL urobilinogen (0.1-1.1)
Wound culture and stain: moderate gram positive bacilli, speciated to *Propionibacterium acnes*
Blood cultures x2: no growth, final
HIV1/2 Antibodies: negative
Anti-nuclear and double-stranded DNA antibodies: negative
Erythrocyte sedimentation rate: 34
Electrocardiogram: normal sinus, normal axis, normal intervals, normal T waves
Rhoenterogram: no evidence of pneumothorax, pleural effusion, or infection; cardiomediastinal silhouette appears normal, with overall low lung volumes. An apparent splenic venous stent is present in the left quadrant.

DIAGNOSIS

Rosacea fulminans, possibly precipitated by influenza vaccination

TREATMENT AND COURSE

The patient was started on pulsed 1g methylprednisolone for 3 days transitioned to a slow 60mg prednisone taper as well as isotretinoin 0.5mg/kg inpatient. Endocrinology was consulted for steroid-induced diabetes and started the patient on appropriate metformin and insulin therapy. He completed 50mg/kg cumulative isotretinoin before being lost to follow-up, though has remained rash-free according to his primary care physician 1 year out.

DISCUSSION

Pyoderma faciale[1, 2], more commonly known today as rosacea fulminans, was first described in 1940 at the Minnesota Dermatologic Society by O'Leary and Kierland of the Mayo Clinic, though similar cases had been presented previously as unknowns at other international meetings[3]. While not considered a formal variant of rosacea like granulomatous rosacea[4], it has a dramatic and relatively specific clinical presentation. Dermatologists should be familiar with this condition in order to initiate aggressive, prompt treatment and minimize the risk of scarring.

There have been less than 100 cases of rosacea fulminans published in the literature to date, almost exclusively in otherwise healthy women, generally between 20-40 years of age[5]. Patients often have a history of easy flushing but not of acne. Edematous well-demarcated asymptomatic papules, pustules, nodules, and sinus tracts appear rapidly over hours to days usually without associated systemic or prodromal symptoms[6]. Mucosal involvement is rare but reported[7]. In addition to the usual rosacea triggers, fulminant rosacea may be precipitated by hormone fluctuations, as some cases were reported both during pregnancy and in patients taking oral contraceptives[8, 9]. High-dose B-vitamins[10] and antiviral stimulation with ribavirin and pegylated interferon alpha-2b have also been reported [11, 12]; it is possible the

antiviral response to the influenza vaccine triggered our patient's eruption. Biopsies and microbiology cultures are rarely obtained and nonspecific.

Treatment consists of high-potency topical or usually systemic steroids initiated concomitantly with oral isotretinoin; it is the only indication for topical or systemic steroid use in the management of rosacea[13]. Antibiotics are usually ineffective, though isolated responses have been reported[14]. The optimal dose of isotretinoin is not known; one author suggested a cumulative dose of 150mg/kg[15], but most case reports describe treatment from 4-6 months at a dose of 0.2-0.5mg/kg. Patients can be reassured the condition does not recur. Only 4 reports have been ascribed to men[16-19]: 1 had significant extrafacial involvement and a complete response to subantimicrobial-dose doxycycline[19], both highly atypical for rosacea fulminans, while another noted isolated nasal involvement with minimal response to isotretinoin or prednisolone and a partial response to dapsone[18].

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CASE #2

PRESENTERS
Rebecca S. Kaiser, MD; Sarah L. Stein, MD

HISTORY OF PRESENT ILLNESS
A seventeen-year-old African American female, well known to dermatology, was referred by gynecology for evaluation of a papillomatous growth of the right labia minora, present for at least 9 months. The patient also noted a papillomatous growth of the right, upper vermilion lip, present for at least 6 months, growing gradually, and occasionally painful.

PAST MEDICAL HISTORY
Several previously excised perioral papillomas; Right fallopian tube torsion and 9 centimeter right paratubal cyst s/p right salpingectomy; Cleft palate repair; developmental delay

FAMILY HISTORY
No family history of inherited skin disease.

MEDICATIONS
Norgestimate/Ethinyl estradiol 0.25mg/0.035mg PO daily

ALLERGIES
No Known Drug Allergies

PHYSICAL EXAMINATION
A 1.5 cm pink, papillomatous plaque was present on the right lateral aspect of upper vermilion lip. Involving the caudal aspect of the right labia minora was a 3-4 cm linear pink, exophytic papillomatous plaque. Extensive hyper- and hypopigmented, atrophic, linear plaques in a Blaschkoid distribution were present on the face, trunk, and extremities. Within these plaques were discrete dark brown macules and grouped follicular, keratotic papules. Several plaques extended the length of select digits with associated nail dystrophy. Syndactyly of the second and third digits of the right foot was present. A well-healed scar of right upper lip from cleft palate repair and associated right alar hypoplasia were also present.

DERMATOPATHOLOGY
A wide excision of the papillomatous lesion of the right labia minora was performed. A papilliform squamous proliferation with marked acute and chronic inflammation was noted on pathology. The dermis was noted to show focal hypoplasia with adipocyte infiltration.

LABORATORY DATA
Complete blood count: Leukocytes 11.1 K/µL (3.5-11.0), hemoglobin 14.0 g/dL (9.8-15.5), platelets 256 K/µL (150-450)
CT abdomen and pelvis with IV contrast: Multiple pelvic cysts noted with mass effect on urinary bladder and rectum.

DIAGNOSIS
Focal Dermal Hypoplasia (Goltz syndrome)

TREATMENT AND COURSE
The perioral papilloma was treated with a shave excision. The patient additionally requested removal of the perivulvar papilloma due to irritation and discomfort. She was referred back to gynecology for surgical excision of the lesion under general anesthesia.

DISCUSSION

Focal dermal hypoplasia (FDH), also known as Goltz syndrome, is a rare, X-linked dominant condition, with less than 300 cases reported worldwide. It is caused by mutations in the PORCN gene (1,2). The vast majority (95%) of FDH cases are sporadic. FDH is antenatally lethal in males with non-mosaic hemizygous mutations. As a result, males make up only 10% of the population affected by FDH (2).

The PORCN gene encodes a protein found in the endoplasmic reticulum. There it functions as a O-acyltranferase, responsible for palmitoylation and secretion of Wnt, a factor crucial in ectomesodermal tissue development (1). Mutations in PORCN result in defects of ectodermal and mesodermal structures, including the skin, bones, teeth, and eyes.

The clinical presentation of FDH is based on the proportion and distribution of cells expressing the mutated X chromosome (1). Cutaneous manifestations are nearly universal, often presenting at birth as semi-translucent dermal atrophy in a Blaschkoid distribution. Later, abnormalities in pigmentation, fat herniation, nail dystrophy, alopecia, and raspberry-like papillomas, with preferential involvement of lips, anogenital skin, larynx, and acral sites, develop. Additional manifestations include: limb malformations, especially split hand/foot deformity (ectrodactyly), microphthalmia and other ophthalmic disorders, enamel hypoplasia, mental retardation, and genitourinary anomalies (1,2).

As demonstrated in our case, some patients with FDH have been noted to develop dark-brown lentigo-like lesions within areas of dermal atrophy. A histopathologic study of such lesions revealed lentigo-like pathology with an acanthotic epidermis and elongated, occasionally club-shaped rete pegs (3). The basal epidermal layer contained increased amounts of melanin and the underlying dermis was rich in melanophages. Of interest, immunostaining with HMB-45 revealed several basal and suprabasal epidermal melanocytes. The authors postulate that FDH lesions may be progressive, with active inflammation causing stimulation of local melanocyte populations.

Treatment for FDH is supportive in nature, often requiring an interdisciplinary team approach.

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CASE #3

PRESENTERS
Ashley Jenkins, MD, Olga Radkevich-Brown, MD, PhD, Farah Abdulla, MD

HISTORY OF PRESENT ILLNESS
A 65-year-old African-American female presented with a 2-year history of progressive pruritic rash that started on her chest and spread to her face, scalp, back, and extremities, including palms and soles. Her soles were painful, but the remainder of her rash was pruritic. She noted patchy hair loss and scalp scaling. The patient's lesions failed to respond to topical triamcinolone or multiple month-long courses of 10 mg daily oral prednisone incidentally prescribed for acute gout. She denied any new medications or change in diet prior to the onset of rash. Review of systems was significant for occasional joint pain, Raynaud's phenomenon, and new-onset occasional muscle weakness including difficulty standing up and lifting her coffee cup. She reported being up-to-date on age-appropriate colon and breast cancer screening, but had not had cervical cancer screening in 5 years. Biopsy of the rash obtained at an outside institution in 2013 was interpreted as pityriasis rubra pilaris (PRP) versus hypertrophic lichen planus.

PAST MEDICAL HISTORY
Her breast cancer was successfully treated with right-sided mastectomy. Other medical conditions include diabetes mellitus, hypertension, gastroesophageal reflux disease, and a remote 8-pack-years smoking history.

FAMILY HISTORY
Non-contributory

MEDICATIONS
Triamcinolone 0.1% ointment, desonide 0.05% ointment, metformin, glimepiride, losartan, metoprolol, pantoprazole, ibuprofen as needed, amitriptyline.

ALLERGIES
No known drug allergies

PHYSICAL EXAMINATION
On the face and neck, there are violaceous and erythematous ill-demarcated non-scaly patches involving the forehead, cheeks, chin and sparing periorbital skin. On the chest, back, and shoulders, there are diffuse violaceous-brown papules coalescing into reticulated plaques. Lesions on upper and lower extremities have similar morphology, but increased degree of hyperkeratosis, particularly bony prominences including dorsal hands and interphalangeal joints. Prominent islands of flexural sparing are noted in both the antecubital and popliteal fossae. The plantar soles have reticulated thick hyperkeratotic plaques with wide pits.

DERMATOPATHOLOGY
A punch biopsy from the left arm demonstrated patchy parakeratosis and follicular plugging. The epidermis showed irregular acanthosis and focal basal vacuolization. Prominent melanophages were noted within the superficial dermis. Superficial perivascular infiltrate of lymphocytes and histiocytes was present. Periodic acid-Schiff stain highlighted significant focal thickening of basement membrane. Colloidal iron stain demonstrated mildly increased amount of dermal mucin in the superficial dermis. A second biopsy from the left thigh demonstrated similar histopathologic findings.

LABORATORY DATA
Complete blood count with differential: within normal limits except absolute lymphocyte number 4.35

K/μL (normal range 0.9-3.3 K/μL)
Comprehensive metabolic panel: within normal limit except elevated glucose 171 mg/dL (normal range 60-109 mg/dL)
Creatinine kinase: 285 U/L (normal range 9-185 U/L)
Aldolase: 11.5 U/L (normal range 2-8 U/L)
Complement C3 and C4: within normal limits
Connective tissue disease autoantibodies: Anti-nuclear antibody: positive 1:160, homogenous and cytoplasmic antibodies present; Negative for Anti-dsDNA, anti-histone, Jo-1, SCL-70/topoisomerase-1, SS-A (Ro), SS-B (La), RNA Polymerase III IgG, RNP, and Smith antibodies
Myositis-specific autoantibodies: Negative for MI-2, PL-7, PL-12, EJ, OJ, KU, U2-RNP, TIF1-gamma autoantibodies
Immunoglobulin subclasses: within normal limits
Protein Electrophoresis: no monoclonal gammopathy

NEURODIAGNOSTIC TESTING
Electromyography: No evidence of myopathy

DIAGNOSIS
Wong-type dermatomyositis

TREATMENT AND COURSE
Our patient was started on azathioprine 50 mg as well as prednisone 30 mg per day with taper by 10 mg per week to 5 mg per day. Patient reported subjective improvement in the pliability of her skin at 3-month follow-up. Her azathioprine was later increased to 100 mg per day. However this was not tolerated due to severe nausea, vomiting and diarrhea despite subsequent dose decrease. She was recently transitioned to methotrexate 20 mg per week and continued on low-dose prednisone 5 mg daily.

To evaluate for systemic disease, a computed tomography (CT) of the chest was ordered which demonstrated multiple prominent axillary lymph nodes bilaterally but no evidence of hilar or mediastinal lymphadenopathy. No evidence of interstitial fibrosis was seen. CT of the abdomen and pelvis revealed enlarged, enhancing bilateral inguinal lymph nodes of uncertain etiology. Papanicolau smear and mammogram were negative for malignancy. Colonoscopy is scheduled in the near future.

DISCUSSION
Dermatomyositis (DM) is a chronic inflammatory disorder thought to be autoimmune in nature with cutaneous and systemic features, including proximal muscle weakness, interstitial lung disease, cardiac involvement, inflammatory arthritis, and association with internal malignancy. DM is increasingly recognized as a heterogeneous entity with a variety of clinical phenotypes and autoantibody profiles. Classic cutaneous features of DM include heliotrope swelling of the eyelids, a violaceous poikilodermatous skin eruption, most prominent on sun-exposed skin, Gottron's papules over bony prominences, and periungual telangiectasias. The histologic diagnosis of dermatomyositis is based upon epidermal atrophy, liquefaction degeneration of the basal layer, and vascular dilatation; other findings include PAS-positive basement membrane thickening and increased mucin deposition. Histopathology of a Gottron's papule is also diagnostic of DM, showing basal layer vacuolization, PAS-positive basement membrane thickening, and upper dermal mucin deposition in addition to orthokeratosis and varying degrees of acanthosis/papillomatosis, rather than epidermal atrophy.

Wong-type DM refers to a small subgroup of patients with dermatomyositis who have additional clinical features of pityriasis rubra pilaris (PRP). The association of PRP-like lesions and DM was originally described by O'Leary in 1953 in a patient with DM, widespread erythroderma and thick keratinization of the soles; histopathology of the erythroderma showed features of PRP. In 1969, Wong reported the largest

case series to date of this distinctive presentation of DM. He described 23 patients with DM, 11 of whom demonstrated a distinctive PRP-like eruption consisting of follicular, erythematous and hyperkeratotic papules located on the dorsal hands/feet (usually in a linear array over bony prominences), posterior neck, forehead, trunk, and limbs. These papules coalesced into plaques especially over trunk and limbs resembling PRP. These patients did not, however, have other features common in PRP, including yellow-orange palmoplantar discoloration or desquamating hyperkeratosis. Twenty-five patients with Wong-type DM have been reported in the literature to date.

Histopathologic features of Wong-type DM vary depending on the type of lesion biopsied. Biopsies of lesions clinically resembling classic DM show typical vacuolar interface change with few dyskeratotic keratinocytes and increased interstitial mucin. Biopsies of skin resembling PRP with follicular hyperkeratosis show irregular psoriasiform hyperplasia, alternating parakeratosis and orthokeratosis, and follicular plugging. Many biopsies show combined histologic features of both PRP and DM, which was seen in our case.

The diagnosis of Wong-type DM in our patient was based on clinical and histopathologic findings of both DM and PRP. Our patient had a violaceous hue to the lesions on her face and trunk and Gottron's papules in addition to diffuse follicular hyperkeratotic papules with islands of sparing and palmoplantar hyperkeratosis. She had elevated muscle enzymes and subjective proximal muscle weakness. Biopsy showed a hybrid histopathology of PRP and DM, with basal layer vacuolization, increased mucin and basement membrane thickening in addition to parakeratosis, irregular psoriasiform acanthosis, and follicular plugging. Although acanthosis can be seen in biopsies of Gottron's papules in classic DM, the findings of follicular plugging and parakeratosis have not been described in classic DM and are rather findings unique to the Wong-type DM. To our knowledge, thickening of basement membrane has not been previously reported in Wong-type DM, and this could represent another connective tissue disease feature of this entity.

The degree of muscle involvement in our patient placed her into the category of hypomyopathic dermatomyositis. This category is defined as DM-specific skin disease without clinical evidence of muscle disease but with subclinical evidence found on either laboratory, electrophysiologic, or radiographic studies. Our patient had no evidence of clinical muscle weakness on exam or elecromyography, however did have elevated muscle enzymes and subjective muscle weakness. Wong-type DM has been described in patients with or without myositis.

After the diagnosis of Wong-type DM has been made, an evaluation for systemic disease is warranted. Similar to classic DM, Wong-type DM appears to carry a risk for myositis and interstitial lung disease. A CT chest and/or pulmonary function tests as well as evaluation for myositis by physical examination and enzymes should be completed. Because the onset of myositis and lung disease can lag behind the cutaneous involvement, prolonged monitoring is warranted. Wong-type DM appears to lack association with internal malignancy.

Oral prednisone, methotrexate, and the anti-malarials, chloroquine and hydroxychloroquine, have been reported to be successful in single-patient case reports. Other treatment options that were reportedly ineffective, include azathioprine, cyclosporine, high-dose immunoglobulins. The efficacy of methotrexate in our patient with extensive long-standing disease remains to be determined.

In summary, Wong-type DM is a hybrid disease with features of both DM and PRP clinically and histopathologically, and could present a diagnostic challenge to clinicians.

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CASE #4

PRESENTERS
Carly Roman, MD; Christopher R. Shea, MD

HISTORY OF PRESENT ILLNESS
A 73-year-old African-American man presented for dermatologic evaluation of a lesion of the left parietal scalp. The lesion had been present since early childhood and had been slowly increasing in size over the last year. It was otherwise asymptomatic. Prior to evaluation, he had a magnetic resonance imaging (MRI) study of the head performed, which showed a "heterogeneous soft tissue mass of the left parietal scalp with evidence of restriction of diffusion. The lesion measures 3.5 x 1.1 x 3 cm. Findings are suspicious for malignancy. There is a subtle abnormality of the underlying skull worrisome for invasive disease."

PAST MEDICAL HISTORY
Coronary artery disease, status post coronary artery bypass grafting, diabetes mellitus, hypertension and hypothyroidism

FAMILY HISTORY
Non-contributory

MEDICATIONS
Aspirin, clopidogrel, furosemide, isosorbide mononitrate, levothyroxine, insulin

ALLERGIES
None

PHYSICAL EXAMINATION
A 5 x 3 cm firm blue nodule of the left parietal scalp with a black reticular patch at the edge was noted. Depigmented hairs outlined the periphery of the lesion.

DERMATOPATHOLOGY
A 4 mm punch biopsy was taken at the periphery of the lesion for sampling. Histopathologic analysis showed a fibrotic dermis with pigmented spindle cells. There was no significant atypia or mitotic figures. Plasma cells were numerous throughout the specimen. The final pathologic diagnosis was signed out as a common blue nevus.

TREATMENT AND COURSE
Although the histopathology did not reveal atypia, a complete excision was recommended to rule out a concurrent malignancy, specifically melanoma. Despite this recommendation, the patient was lost to follow-up.

He presented one year later to the otolaryngology clinic for evaluation of a left-sided neck mass. Fine needle sampling was done and showed findings consistent with metastatic melanoma. The patient is now status post wide local excision with left cervical node dissection. Breslow thickness is not applicable as there was no intraepidermal component, but the greatest depth was measured at 21 mm. There were 4 mitotic figures per mm^2 and both vasculo-lymphatic and perinerual invasion were present. There was no ulceration, regression, or satellitosis. Positron emission tomography (PET) scan showed widely metastatic disease involving the liver, lungs, and ischium. He is followed by oncology and has recently been started on pembrolizumab.

DIAGNOSIS

Melanoma Arising from a Blue Nevus

DISCUSSION

Melanoma arising from a blue nevus is an exceedingly rare variant of melanoma, with only 109 cases reported in the literature since it was first described in 1953.[1,2] The most common anatomic site is the head and neck, particularly the scalp, followed by the trunk and lower extremities.[1,2] Overall, there is a slight male predominance and a median age at diagnosis of 44 years.[1]

The histopathologic diagnosis can be difficult.[4] The most commonly reported pathologic findings are as follows: a malignant melanocytic proliferation mimicking a cellular blue nevus but lacking a benign component; melanoma arising from a cellular blue nevus; and melanoma arising from a common blue nevus.[1,3]

Although melanomas arising from blue nevi exhibit similar outcomes to conventional melanomas, it is controversial as to which prognostic factors are applicable to this type of melanoma.[1] Many of these tumors arise from the dermis or subcutaneous tissue without an associated intraepidermal component, so tumor thickness is defined as the largest tumor dimension in lieu of measuring a true Breslow depth.[1] A recent case series of 24 cases of melanoma arising from a blue nevus did demonstrate a significant association between tumor thickness and reduced recurrence-free survival and reduced time to distant metastasis. Other prognostic indicators predictive in conventional cutaneous melanomas (age, gender, mitotic figures, lymphovascular invasion, perineural invasion and ulceration) did not correlate with outcomes in this small series.[1]

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CASE #5

PRESENTERS
Eduardo K Moioli, MD, PhD; Christopher R Shea, MD; and Arlene M Ruiz de Luzuriaga, MD

HISTORY OF PRESENT ILLNESS
A 79 year old female presented with multiple small projecting skin lesions on the bilateral palmar hands. The lesions had been present for many months and were occasionally pruritic. Patient endorsed "plucking off" some of these lesions. There was no involvement of the plantar feet and no associated hair or nail changes. Patient had tried various emollients without improvement.

PAST MEDICAL HISTORY
Diabetes mellitus
End stage renal disease
Hypertension

MEDICATIONS
Carbidopa-Levodopa 25-100mg PO bid
Hydralazine 50 mg PO qid
Insulin glargine SQ daily
Metoprolol-XL 25mg PO daily
Nifedical XL 60mg PO daily
Sevelamer carbonate 800mg PO daily
Simvastatin 40mg PO qhs

ALLERGIES
No known drug allergies

PHYSICAL EXAMINATION
Multiple non-tender, 1mm, firmly attached, keratotic digitate projections were noted on the bilateral palms and flexor digits. The dorsal hands and feet were spared. No hair or nail changes.

DERMATOPATHOLOGY
Compact parakeratotic columns were seen in the stratum corneum arising from the inter-adnexal epidermis. Deep to the column, the underlying epidermis showed an attenuated granular layer compared to the surrounding epidermis. No dyskeratosis or vacuolar changes were noted.

DIAGNOSIS
Acquired Spiny Keratoderma

TREATMENT AND COURSE
Upon clinical diagnosis of acquired spiny keratoderma and histopathological confirmation, the association of this diagnosis with systemic diseases and malignancies were discussed with the patient and her family. Malignancy screening was recommended. The patient subsequently presented to primary care with new onset bloody stools. Colonoscopy demonstrated a large, nearly obstructive mass in the transverse colon near the hepatic flexure with biopsy consistent with tubulovillous adenoma. The patient was recommended to have a subtotal colectomy with end ileostomy. Given her complicated medical history and high risk for post-operative complications, the patient and family opted to defer surgical treatment.

Lesional treatment options were also reviewed with the patient including topical therapy with 5-fluoruracil and ammonium lactate as well as systemic therapy with acitretin. Given minimal symptoms

and patient comorbidities, the patient and family opted for local treatment with ammonium lactate.

DISCUSSION

Spiny keratoderma is a rare disorder of keratinization. It has been described in the past with many alternative names including punctate porokeratotic keratoderma, music box spine dermatosis, multiple minute palmar-plantar digitate hyperkeratosis, and filiform hyperkeratosis. The term spiny keratoderma is currently the preferred nomenclature[1,2]. The disorder is characterized by multiple firm, keratotic, digitate projections on the bilateral palms, soles, or both. The dorsal hands and feet are spared. Lesions are typically asymptomatic, however pain and pruritus have been described. Histopathological findings are characterized by columns of parakeratosis with attenuation of the granular layer in the epidermis immediately deep to the column [3]. No dyskeratosis or vacuolar changes are seen.

Although the pathophysiology of spiny keratoderma has not been well described, immunohistochemical studies have demonstrated that the hair-specific antikeratin antibody AE13 stained the lower part of the keratotic column and was variably positive in the viable epidermis, whereas AE14 staining was negative [4]. Moreover, accumulation of keratin filaments without production of keratohyalin or trichohyalin granules was observed with electron microscopy, demonstrating features of hair cortex keratinization. Given these results, it has been postulated that spiny keratoderma may represent a disorder of ectopic hair formation. Additional studies have also shown increased expression of keratins 6 and 16, which are markers of hyperproliferative cells [5].

Both hereditary and acquired forms of the disorder exist. Approximately 20% of the cases reported in the literature were hereditary, typically presenting between the 2nd and 5th decades of life, and were inherited in an autosomal dominant pattern [6]. No association with systemic disease or malignancy has been described in hereditary cases [7]. On the other hand, both have been associated with the acquired form of the disorder. To our knowledge, there are no prior reports of association with significant occult benign tumors such as in the present case. Acquired spiny keratoderma presents after the age of 50. In one previous review article, 9 of 29 cases were associated with a malignancy[8]. Associated malignancies include rectal, sigmoid colon, lung, esophageal, renal, and breast carcinomas as well as chronic lymphocytic leukemia and myelofibrosis[1,8]. Cutaneous malignancies including squamous cell carcinoma and melanoma have also been associated. The cutaneous findings of spiny keratoderma do not tend to resolve after treatment of the associated malignancy [1]. Systemic disorders described in association with acquired spiny keratoderma have included Darier's disease, type IV hyperlipoproteinemia, renal disease, and pulmonary tuberculosis [1,9,10].

Skin directed treatment options are not well established and are limited. Clinical improvement has been reported in the literature after treatment with 5-fluoruracil[11], topical tacalcitol 0.002% ointment[12], as well as systemic therapy with acitretin[13]. In general, adequate initial management of patients presenting with the acquired form of spiny keratoderma include a thorough history of presenting illness, full review of systems, basic serology, and malignancy screening. Additional testing such as lipid panel, quantiferon gold test, antinuclear antibodies, or others should be pursued based on clinical suspicion. Considerations for malignancy screening include skin exam, fecal occult blood test, colonoscopy, computed tomography imaging, mammogram, Pap test, prostate-specific antigen, carcinoembryonic antigen (CEA), and CA-125 together with a transvaginal ultrasound. Skin treatment may be geared toward the relief of symptoms when present and continued follow up is imperative with a low threshold for testing if a malignancy or systemic disorder is suspected.

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CASE # 6

PRESENTERS
Haider K Bangash, MD; Farah R Abdulla, MD

UNKNOWN

CASE #7

PRESENTERS
Juliana Gao, MD; Aisha Sethi, MD; Diana Bolotin, MD PhD

PATIENT A

HISTORY OF PRESENT ILLNESS
A 22-year-old African American male was admitted for fevers, migrating polyarthritis and a rash of 5 days duration. The rash started first, and was described as small painless red-purple lesions of the hands and feet. Two days following onset of the rash, he developed pain and swelling of the shoulders, knees, hips and ankles. The patient is the son of a pastor and initially denied any preceding sexual encounters but later endorsed unprotected sex with two female partners in the past few months.

PAST MEDICAL HISTORY
Hypertension, seasonal allergies and paronychia of the R great toe

FAMILY HISTORY
Non-contributory

MEDICATIONS
Cetirizine, mometasone nasal spray, naproxen, hydrocodone-acetaminophen

ALLERGIES
Codeine

PHYSICAL EXAMINATION
Few small 2mm non-blanching violaceous macules scattered on the dorsal hands and feet were noted. Also, there were two areas of retiform purpura about 1 cm in diameter located on the medial right 5th toe and right 5th fingertip.

DERMATOPATHOLOGY
Two 3 mm punch biopsies from the R finger were obtained for histopathologic analysis and direct immunofluorescence. On histopathology, the epidermis revealed parakeratosis, acanthosis and intercellular edema. A superficial perivascular infiltrate composed of lymphocyte with few eosinophils was noted. Extravasated erythrocytes were seen focally in the papillary dermis and within the epidermis. Periodic acid-Schiff (PAS) stain was negative for fungi or significant basement membrane thickening. Immunofluorescence was negative except for some immunoglobulin and fibrinogen in the superficial layers of the epidermis. There was no evidence of vasculitis.

LABORATORY DATA
Complete blood count: Leukocytes 9.2 K/μL (3.5-11.0), hemoglobin 14.2 g/dL (9.8-17.6), platelets 175 K/μL (150-450).
Differential: Neutrophils 81% (39-75%), lymphocytes 12% (16-47%), monocytes 7% (4-12%), eosinophils 0% (0-7%).
Immunology/serology: ANA titer 160 speckled (0-80), anti-dsDNA titer <10 (<10), ANCA titer < 20 (<20), C3 159 mg/dL (83-188), C4 37 (mg/dL), cryoglobulin < 0.5 % volume (0-0.5%).
Erythrocyte sedimentation rate: 40 mm/h (0-20).
C-reactive protein: 198 mg/L (<5).
Infectious: Blood culture – negative X 2; joint fluid culture – negative X 1; urine gonococcus and chlamydia probe – negative; **oropharyngeal gonococcus and chlamydia probe – positive for**

gonococcus.
Echocardiogram: No vegetation or apical thrombus noted.

DIAGNOSIS
Disseminated gonococcal infection

TREATMENT AND COURSE
The patient was treated with a seven-day course of ceftriaxone while inpatient and discharged to rehab with a three-day course of cefdinir for a total of a ten-day course. At two week follow up, he reported marked improvement of arthralgia and resolution of skin findings.

PATIENT B

HISTORY OF PRESENT ILLNESS
A 45-year-old Caucasian female with history of acute lymphoid leukemia status post stem cell transplant complicated by graft-versus-host disease (GVHD) and two relapses of her leukemia, undergoing experimental therapies presented from oncology clinic as urgent consult for a new rash. Patient was seen in infectious disease clinic several days earlier, and blood culture from that visit grew yeast, later speciated as *Fusarium.*

PAST MEDICAL HISTORY
Acute lymphoid leukemia status post stem cell transplant with relapse, graft-versus-host disease, *Staphylococcus* and vancomycin resistant *Enterococcus* bacteremia, chronic sinusitis, and cirrhosis.

FAMILY HISTORY
Non-contributory

MEDICATIONS
Chemotherapy: GS-9973 (selective spleen tyrosine kinase inhibitor), vincristine, dexamethasone, and intra-thecal cytarabine
Antimicrobial prophylaxis: levofloxacin, acyclovir and posaconazole
Others: acetaminophen, furosemide, gabapentin, lorazepam, oxycodone, peri-colace, potassium chloride, prochloperazine, tramadol, ursodiol

ALLERGIES
Meropenem, piperacillin-tazobactam, sulfa, and silver

PHYSICAL EXAMINATION
Multiple erythematous to purpuric and slightly indurated papules and plaques scattered on face, arms and legs and several tender papulonodules with occasional dusky centers on the lower legs.

DERMATOPATHOLOGY
Punch biopsy of the right lower extremities was obtained for histopathologic analysis which showed cross-sections of numerous hyphal elements as well as septated hyphae occluding blood vessel lumina and within the vessel walls. Periodic acid-Schiff (PAS) and Grocott's methenamine silver (GMS) stains were positive. The Gram and Fite stains were negative for microorganisms. There was abundant dermal hemorrhage and hemosiderin. These findings were consistent with vaso-occlusive and angioinvasive fungal infection.

LABORATORY DATA
Complete blood count: **Leukocytes 1.9 K/µL (3.5-11.0)**, hemoglobin 9.3 g/dL (9.8-17.6), platelets 10

K/µL (150-450).
Absolute neutrophil count: 0.15 K/µL (1.12-6.72)
Infectious: blood cultures positive for *Fusarium*

DIAGNOSIS
Disseminated *Fusarium* infection

TREATMENT AND COURSE
Patient was admitted to the hospital given positive blood culture of *Fusarium.* Otolaryngology (ENT) was consulted and performed endoscopic examination which showed invasive yeast growth in the sinuses with necrotic mucosa. Computed tomography (CT) showed multiple nodular scattered airspace opacities suspicious for hematogenous spread of the infection. Patient was started on amphotericin B and underwent two endoscopic debridements by ENT. Despite these aggressive treatment measures, the patient continued to worsen clinically with repeated spiking fevers , epistaxis and drainage. She eventually opted for home hospice and was discharged home.

DISCUSSION
Gonorrhea is one of the most commonly reported communicable diseases, with approximately 820,000 new infections occurring each year and the majority between ages 15 and 24 [1]. Genital infections are the most common, including cervicitis, urethritis in women and urethritis and epididymitis in men. Extragenital infections such as that of the rectum and pharynx have also been reported, and are typically asymptomatic. The prevalence of pharyngeal gonorrhea is approximately 2-8% depending on study populations [2].

Disseminated gonococcal infection occurs in 0.5-3% of all reported cases, and the most common symptoms are arthritis/arthralgia and skin findings (pustules and purpura) – therefore referred to as arthritis-dermatosis syndrome. If biopsied, pathology demonstrates epidermal necrosis with pustule(s) as well as neutrophilic vasculitis with extravasated erythrocytes and thrombi indicative of septic embolic [3]. The predisposing factors to disseminated disease include preceding asymptomatic mucosal infection, the virulence factor displayed on the outer membrane of the gonococci, with some glycoprotein promoting adhesion and invasion to host cells as well as host immune system though most host are not immunocompromised. Due to the fastidious growth requirement of *N. gonorrhea*, blood and synovial culture are often negative. Hence, PCR probe for gonococcal DNA is the preferred method of screening and negative culture in the setting of high suspicious does not rule out disease.

According to CDC 2010 guidelines, the recommended treatment for disseminated gonococcal infection is ceftriaxone 1g IM or IV Q24H for 7 days plus azithromycin 1g PO as a single dose. Dual therapy is intended for treatment of potential concomitant *C. trachomatis* infection even if PCR negative, and helps reduce emergence of bacterial resistance. Oral step-down agents can be used to complete a seven-day course pending local susceptibility once patient improves with parenteral therapy. Doxycycline can be used in place of azithromycin in the case of medication allergies. Unfortunately, there is no good data regarding alternative treatment in patients with cephalosporin/penicillin allergies. In the past, fluoroquinolone has been used, but data from 2007 suggest emerging resistance [2].

Fusaria are ubiquitous fungal species in the environment, which can be found in soil, plants as well as biofilms in water structure. They grow easily and rapidly in most media that does not contain cycloheximide, which inhibit its growth. Classically, *Fusarium* species are described as fusoid or banana-shaped macroconidia with or without microconidia in culture. On histology, the hyphae are narrow and septated with dichotomous branching at acute or right angle. Pathogenesis of *Fusariosis* involves direct tissue destruction in localized disease, and angioinvasion in disseminated infection [4].

In immunocompetent host, *Fusarium* can cause keratitis, onychomycosis, superficial and deep cutaneous infections that include intertrigo, tinea, cellulitis, ulcers and abscess. In immunocompromised hosts, this angioinvasive fungal species often causes sinusitis, which can progress to periorbital and paranasal cellulitis and necrosis of involved mucosa, pneumonia and disseminated infection. With disseminated skin infection, typical findings are painful red to violaceous nodules, with or without ulcerations and eschar.

Risk factors of disseminated infection include prolonged neutropenia and severe T-cell immunodeficiency, especially in patients who are status post stem cell transplant with severe graft-versus-host disease. According to one study, the incidences of *Fusarium* infection status post stem cell transplant ranges from 4.21 to 20.19 cases per 1000 cases, with higher incidence in patients who underwent HLA-mismatched transplant than those with HLA-matched related transplant [5].

The most common presentation of disseminated fusariosis is a combination of characteristic cutaneous lesions and positive blood culture. Hence, histologic examination of skin tissue can be extremely useful in arriving at the diagnosis. Treatments for disseminated Fusarium infection include systemic antifungal therapy in conjunction with surgical debridement of infected tissues when possible. Different formulations of amphotericin B have been reported with various success, and azole antifungals such as voriconazole and posaconazole have also been used. Unfortunately, the prognosis of disseminated *Fusarium* infection is extremely poor, with survival rate of 13% overall and death rate approaching 100% in patients who are persistently neutropenic [4, 5, 6].

Disseminated infections are relatively uncommon but often-feared complications of hospitalized patients. Here, we present two cases of disseminated infections for clinical interest.

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CASE #8

PATIENT A

PRESENTERS
Duri Yun, MD, Sarah L. Stein, MD

HISTORY OF PRESENT ILLNESS
A 6-week-old full term healthy infant girl presented for evaluation of a birthmark. The patient's mother reported that a red patch on the left shoulder had been present at delivery. The patch has become slightly more red over time, but has remained flat. The lesion has not bled and does not appear to cause discomfort for the patient.

PAST MEDICAL HISTORY
Full term, uncomplicated pregnancy and delivery. Normal growth and development.

FAMILY HISTORY
Non-contributory

MEDICATIONS
None

ALLERGIES
NKDA

PHYSICAL EXAMINATION
Well appearing, well developed 6-week-old female. On the left anterior shoulder there is a segmental 3x 4 cm telangiectatic red vascular patch with brighter red slightly elevated papules at the periphery.

DERMATOPATHOLOGY
None

LABORATORY DATA
None

DIAGNOSIS
Infantile hemangioma – minimal or arrested growth type (IH-MAG)

CLINICAL COURSE
Clinical monitoring without intervention was recommended. By 8 months of age the lesion had grown proportionately with the child with more prominent bright red reticulated papules at the periphery. No motor deficits of the involved left arm and no limb length or girth discrepancy was appreciated. The lesion continued to be asymptomatic without bleeding or apparent discomfort.

PATIENT B

HISTORY OF PRESENT ILLNESS
A 4-month-old full term healthy girl presented for evaluation of a bluish patch on the left arm that has been present since birth. The patch has increased in size over time with more red macules appearing at the periphery, but has remained mostly flat. It has not bled and does not appear to cause discomfort for

the patient.

PAST MEDICAL HISTORY
Full term, uncomplicated pregnancy and delivery. Normal growth and development.

FAMILY HISTORY
Non-contributory

MEDICATIONS
None

ALLERGIES
NKDA

PHYSICAL EXAMINATION
Well appearing, well developed 4-month-old female. On the left upper inner arm, extending into the antecubital fossa, there is a 9 x 5 cm blue purple patch with prominent telangiectasias and a blanched halo. Bright red papules and macules are noted at the periphery. The girth and contour of the arm is symmetric with the other arm.

DERMATOPATHOLOGY
None

LABORATORY DATA
None

DIAGNOSIS
Infantile hemangioma – minimal or arrested growth type (IH-MAG)

CLINICAL COURSE
Clinical monitoring without intervention was recommended. At 7 months of age the lesion had grown proportionately with the child. No motor deficits of the involved left arm and no limb length or girth discrepancy was appreciated. The hemangioma had developed some mild xerosis and a more diffusely reticulated appearance. The lesion continued to be asymptomatic without bleeding or apparent discomfort.

PATIENT C

HISTORY OF PRESENT ILLNESS
Dermatology was consulted by the neonatal intensive care unit to evaluate a 1-day-old 31-week gestational age girl with prominent vasculature on the distal right arm. The infant was otherwise well. The infant was born via spontaneous vaginal delivery and Apgars at delivery were 8 and 9 at 1 and 5 minutes respectively. The infant was moving the extremity normally and did not demonstrate evidence of pain.

PAST MEDICAL HISTORY
Born at 31 weeks gestation via spontaneous vaginal delivery as a result of preterm labor precipitated by cocaine and heroine intrauterine drug exposure to a G8P10 33-year-old mother with a history of untreated schizophrenia, no prenatal care and perinatal labs significant for hepatitis C.

FAMILY HISTORY
Non-contributory

MEDICATIONS
None

ALLERGIES
NKDA

PHYSICAL EXAMINATION
Well appearing 1-day-old 31-week gestation infant girl breathing room air in an isolette. On the right arm, extending from the right proximal forearm to the distal fingers in a biker-glove distribution, there are coarse telangiectasias and prominent reticular veins with subcutaneous atrophy of the involved region. Grip strength is symmetric. No limb length discrepancy is noted.

DERMATOPATHOLOGY
None

LABORATORY DATA
Complete blood count with differential: within normal limits for age
Comprehensive metabolic panel: within normal limits for age
Hepatitis B IgM antibody: negative
Hepatitis C viral load: none
Hepatitis C antibody: positive
HIV proviral DNA: negative
Urine toxicology screen: positive for cocaine and opiates
Newborn screen: normal
Venous and arterial ultrasound of right upper extremity: vessels with normal arterial and venous waveforms without discrete vascular masses

DIAGNOSIS
Infantile hemangioma – minimal or arrested growth type (IH-MAG) with lipoatrophy

CLINICAL COURSE
Clinical monitoring without intervention was recommended. At three months of age the lesion had grown proportionately with the child. No motor deficits of the involved right arm and no limb length discrepancy was appreciated. The lesion continued to be asymptomatic without bleeding or apparent discomfort. The hemangioma had developed a more diffusely red appearance with more prominent clusters of blanchable red to purple papules on the affected palm and extensor arm. The associated lipoatrophy of the involved region has remained stable.

DISCUSSION
Infantile hemangiomas are the most common benign vascular tumor of infancy with an estimated incidence between 4-10% of all children [1]. These lesions can be distinguished by their clinical course in which lesions are typically noted to appear within the first few weeks of life, followed by subsequent rapid growth. Eighty percent of final hemangioma size is typically reached at a mean age of 3 months [2]. This growth phase is followed by attenuation of growth and subsequent gradual involution. Hemangiomas can present as focal, segmental, solitary or multiple lesions, affecting the skin in a superficial, deep or mixed manner. Identifying segmental lesions is of particular importance due to the higher association of these lesions with complications and syndromic manifestations. [1]. In contrast to

congenital hemangiomas, infantile hemangiomas will exhibit GLUT-1 positivity on immunohistochemical staining [1, 3].

A subset of infantile hemangiomas has been described in which there is minimal growth beyond the premonitory mark [4]. Multiple terms have been used to describe this type of hemangioma in the literature such as "precursor," "arrested" or "reticular," and recent efforts have been made to uniformly categorize these lesions as "infantile hemangioma with minimal or arrested growth (IH-MAG)" [4]. These lesions have been defined to have a proliferative component equaling less than 25% of the total surface area; proliferative is further defined as any component that appears bright red, papular, plaque-like or nodular [4]. Histopathologic examination has revealed ectatic vessels in the papillary dermis or few lobules in the reticular dermis and subcutis with GLUT-1 positivity on immunohistochemical staining, consistent with typical infantile hemangiomas [5]. In one retrospective study involving 42 patients with IH-MAG, 64% were female and in 78% of patients the lesion was present at birth. The most commonly described appearance of the hemangioma was a fine or coarse telangiectatic patch. Several described lesions also exhibited a vasoconstricted halo around the vascular markings. Those hemangiomas with a proliferative component typically exhibited bright red papules at the periphery of the lesion. Sixty-four percent of IH-MAG were localized and 30% were segmental. In contrast to typical infantile hemangiomas, IH-MAG lesions most often involved the lower half of the body (68%) [4].

More recently, a subset of IH-MAG have been described associated with lipoatrophy. In a retrospective chart review of 53 patients with IH-MAG, 7 patients were noted to have associated lipoatrophy in 3 main patterns: focal cutaneous depression; semicircular lipoatrophy with horizontal bandlike depression; and segmental soft-tissue atrophy [6]. Most of these lesions did not require treatment and only one case was complicated by significant ulceration requiring propranolol. Patients that had follow up after 2 years of age were noted to have persistent residual reticulated lesions with persistent mild lipoatrophy, but no significant limb length discrepancy [6].

These cases highlight the unique subtype of infantile hemangioma with minimal or arrested growth. Familiarity with this phenotype will assist clinicians when considering the differential diagnosis of vascular birthmarks of infancy.

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CASE # 9

PRESENTERS

Olga Radkevich-Brown MD, PhD; Vesna Petronic-Rosic MD, MSc

UNKNOWN

CASE #10

PRESENTERS
Laura Buford, MD; Alex Means, MD; Aisha Sethi, MD

HISTORY OF PRESENT ILLNESS
Patient is a 55-year-old Caucasian gentleman who presented with a one year history of progressive penile ulceration. Approximately twelve months prior, he noticed a small lesion on his penile shaft and was subsequently diagnosed with Peyronie's disease. Over the next five months, the lesion increased in size and eventually ulcerated, which prompted a skin biopsy. The biopsy site dehisced over the next few weeks, which lead to an attempted phalloplasty. The phalloplasty was similarly complicated by wound dehiscence and progressive worsening ulceration resulting in urethrocutaneous fistula formation. The patient was admitted to the Urology service for wound debridement and percutaneous suprapubic tube placement. Dermatology was consulted for evaluation of the ulceration.

PAST MEDICAL HISTORY
Polyarteritis nodosa, erectile dysfunction, hypertension, hyperlipidemia, GERD, asthma, and seasonal allergies

FAMILY HISTORY
Family history of prostate cancer in father. No family history of skin conditions or skin cancer

MEDICATIONS
Docosahexanoic acid/EPA (Fish Oil), fluticasone nasal, glucosamine-chondroitin, lisinopril, lovastatin, multivitamin, omeprazole, organ concentrates (prostate), aspirin, and tamsulosin

ALLERGIES
Gabapentin (rash)

PHYSICAL EXAMINATION
A deep ulcer with a purulent base and an irregular, undermined, overhanging violaceous border extended centrifugally on the ventral penile shaft to the base of the penis proximally and the glans penis distally. The corpus spongiosum and urethra were exposed from the distal penile shaft to the corona.

DERMATOPATHOLOGY
Penile glans lesion; debridement: hematoxylin and eosin staining showed dermal tissue with neutrophilic and chronic inflammation and microabscess formation.
Penile shaft lesion; debridement: hematoxylin and eosin staining showed fragments of skin with ulceration, dermal neutrophilic and chronic inflammation and microabscess formation. Gram and Grocott-Gomori's methenamine silver stains were negative for bacterial and fungal organisms.
Penile shaft skin; debridement: hematoxylin and eosin staining showed skin with patchy dermal neutrophilic and chronic inflammation.

LABORATORY DATA
Comprehensive Metabolic Panel: Sodium 139 mEq/L (134-149), Potassium 4.6 mEq/L (3.5-5), Chloride 100 mEq/L (95-108), Carbon Dioxide mEq/L (23-30), Anion Gap 11 mmol/L (6-15), BUN 11 mg/dL (7-20), Creatinine 0.9 mg/dL (0.5-1.4), Glucose 95 mg/dL (60-109), Calcium 9.3 mg/dL (8.4-10.2), GFR Estimate 88 mL/min/BSA (>59), Bilirubin, Total 0.3 mg/dL (0.1-1.0), Total Protein 6.9 g/dL (6.0-8.3), Albumin 3.8 g/dL (3.5-5.0), Alk Phos 60 U/L (30-120), AST 19 U/L (8-37), ALT 16 U/L (8-35).
Complete blood count: Leukocytes 8.8 K/μL (3.5-17.7), Hemoglobin 14.2 g/dL (9.8-17.6), Hematocrit 43.5% (41-53), Platelets 200 K/μL (150-450)

Differential: Neutrophils 81%, lymphocytes 10%, monocytes 6%, eosinophils 2%, basophils 1%
CRP: 7 mg/L (<5)
ESR: 16 mm/Hr (0-28)
Infectious Disease: HBV surface antigen negative, HBV surface antibody negative, HCV core antibody negative, RPR negative, quantiferon gold negative, AFB/fungal culture x3 negative, bacterial culture < 20,000 CFU *Staphylococcus epidermidis*
Immunology: ANA 1:160 speckled pattern, anti-dsDNA negative, ANCA IFA <20 titer (<20), Antiphospholipid antibody panel negative, SPEP negative, UPEP negative

DIAGNOSIS
Penile Pyoderma Gangrenosum

TREATMENT AND COURSE
An extensive work up for associated systemic conditions, infection, and other causes of chronic ulceration was negative. The patient was strongly advised to avoid any elective surgeries and sexual activity indefinitely. A suprapubic percutaneous urinary catheter was placed, and he was started on prednisone 60 mg by mouth with topical mupirocin 2% ointment with temporary improvement. He was started on 150mg azathioprine daily by Rheumatology outpatient, but had progression of disease and was admitted again for penile debridement by Plastic Surgery 6 months after initial dermatologic evaluation. Unfortunately, the patient's course has continued to be complicated by multiple interventions and unintended trauma, as well as difficulty coordinating care among his numerous in-house and outside hospital providers including Dermatology, Rheumatology, Infectious Disease, Urology, and Plastic Surgery. He is currently maintained on low-dose prednisone and topical tacrolimus. A perineal urethrostomy is planned for the near future.

DISCUSSION
Pyoderma Gangrenosum (PG) was first described in the 1930s by Drs. Brunsting, O'Leary, and Goekerman and was thought to be caused by infection.[1] PG is an uncommon ulcerative disease characterized by chronic and recurrent cutaneous ulcers.[1-3] It affects more women than men, and typically presents between 20 and 50 years of age.[1] Minor trauma or surgery may initiate or worsen PG, which is a phenomenon known as pathergy.[1-5] Approximately 50-70% of affected patients have an associated systemic condition, most commonly inflammatory bowel disease (Crohn's disease and ulcerative colitis), hematologic malignancy (acute and chronic myelogenous leukemia, myelodysplastic syndrome, and monoclonal gammopathy), and rheumatologic disorders (seronegative arthritis and rheumatoid arthritis).[1-3]

The pathogenesis of this neutrophilic dermatosis remains poorly understood, but both autoimmune and autoinflammatory etiologies have been implicated.[1-3] The histopathology varies depending on the stage of ulcer development and is nonspecific.[1-2, 6] A folliculocentric neutrophilic infiltrate that may or may not have leukocytoclasia is seen in early lesions, and marked necrosis with a mononuclear infiltrate is seen in fully developed lesions.[1,2] Histopathology supports the diagnosis, but it is a diagnosis of exclusion that heavily relies on the clinical presentation.[6]

PG of the penis is rare with less than twenty known case reports, but it should be included in the differential diagnosis of chronic genital ulcerative diseases with negative laboratory and histopathological evaluations.[7] Similar to classic presentations of PG, penile PG may develop in the setting of systemic disease, following local trauma, or be idiopathic. The initial evaluation should focus on both identifying associated systemic disorders and excluding other possibilities.[3,6] In cases of suspected penile PG, all possible causes of genital ulceration must be considered. This widens the differential diagnosis and requires additional diagnostic testing.[7] Treatment is focused on immunosuppression, controlling underlying disease (if any), and avoidance of pathergic stimuli. The condition follows a chronic and often

recurrent course, and there is not a uniformly accepted management algorithm;[1] however, early recognition of PG is helpful in avoiding the unintended exacerbation of ulceration by unnecessary surgical intervention.[5-7]

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