CHICAGO DERMATOLOGICAL SOCIETY CASE # 1

Presented by James Swan, M.D., Mary Martini, M.D. and Keren Horn, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 45 year-old Caucasian male presented for a problem-focused exam of a right upper extremity lesion. However, complete physical exam revealed peri-orbital slate-gray hyperpigmentation as well as generalized bronze hyperpigmentation in sun-exposed areas, prompting questions regarding these abnormalities. Other abnormalities in pigment included approximately 10 café au lait macules, nearly all greater than 1.5cm in size. Axillary freckling was also noted. Review of systems was positive for migraine headaches, arthritis, decreased libido and fatigue (the patient sleeps greater than 12 hours each night without feeling refreshed).

PAST MEDICAL HISTORY

Right orbital tumor status post excision 2001
Depression

MEDICATIONS

Depakote
Remeron

ALLERGIES

Amoxicillin (angioedema)

FAMILY HISTORY

Brother with the same pigmentation anomalies.
Both his father and his sister died of liver cancer; his sister passed away at 40 years of age.

SOCIAL HISTORY

The patient does not drink alcohol and has smoked one pack of cigarettes per day for the past 30 years. He is on disability due to "depression" and exhaustion which make it unable for him to sustain an eight hour work day.

PHYSICAL EXAM

There is peri-orbital slate-gray hyperpigmentation as well as generalized bronze hyperpigmentation in sun-exposed areas. In addition, the patient has approximately 10 café au lait macules, nearly all greater than 1.5cm in size. Axillary freckling is also noted, and there are protuberances of the left elbow and right knee.

LABORATORY RESULTS

The following were negative or within normal limits:
Complete blood count with differential, acute hepatitis panel, glucose, comprehensive metabolic panel, and urine and plasma porphyrins.

The following were abnormal:
Ferritin = 3,331.1 (24-336), iron = 236 (40-170), transferrin saturation = 101.3%, alanine aminotransferase (ALT) = 105, aspartate aminotransferase (AST) = 60. Hereditary hemochromatosis gene analysis was homozygous positive for the C282Y mutation.

HISTOPATHOLOGY
01B-59 L1 (right orbital tumor) – Consistent with neurofibroma
DP6136-03 (left abdomen) – Neurofibromatosis-associated café au lait macule with giant melanosomes

DIAGNOSIS
Hereditary Hemochromatosis and Neurofibromatosis Type I

TREATMENT AND COURSE
The patient was referred to hepatology, hematology and general medicine after confirmation of the hemochromatosis mutation. He has since undergone a liver ultrasound, and has begun bimonthly phlebotomy treatments. He will undergo a liver biopsy in the next month.

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 2

Presented by Joan Guitart, M.D. and Prashant Singri, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 81 year old Asian male presented with a six-month history of a generalized erythematous rash with pruritus. After several months of the dermatitis, the patient noticed that his skin was becoming "loose and rubbery." Review of systems was positive for a 20-pound weight loss, fatigue and night sweats. An initial elevated white blood cell count led to a full oncological workup which included bone marrow aspiration, lymph node biopsy and metastatic workup.

PAST MEDICAL HISTORY

Hypertension
Hypothyroid
Cerebrovascular accident (1996)

MEDICATIONS

Interferon alpha three million units
Atenolol
Aspirin

ALLERGIES

No known drug allergies.

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

Noncontributory

PHYSICAL EXAMINATION

There is generalized erythematous patches with ichthyotic type scaling and an increase in skin laxity. Palpable lymphadenopathy was noted in the bilateral submandibular, axillary and inguinal regions.

LABORATORY RESULTS

The following were negative or within normal limits:
Basic metabolic panel, liver function tests, lactate dehydrogenase, metastatic bone survey, human T-lymphotropic virus types I and II (HTLV), Epstein Barr virus.

The following were abnormal:
White blood cell (52.6), hemoglobin (11.3), platelets (445), neutrophils, absolute (46.8), albumin (2.4), protein (8.7), alkaline phospatase (609), calcium (8.2).

Quantitative IgG -3860, IgA – 642, Kappa light chain – 3590, Lamda light chain 1510.

Serum Protein electrophoresis – Two restricted bands in the gamma region
Serum immunofixation electrophoresis – Monoclonal IgG Kappa
Urine immunofixation electrophoresis – Triclonal IgG Lamda

Bone Marrow aspiration – Hypercellular bone marrow with plasmacytosis and myeloid predominance and no evidence of clonality.

Chest radiography – Chronic interstitial changes in lower lobes consistent with bronchiectasis.

HISTOPATHOLOGY
DP000664-03 (right arm) – Dense dermal infiltrate of atypical lymphocytes, histiocytes and multinucleated giant cells. Focal elastic fibers are noted within the cytoplasm of histiocytes.

S02-9474,11/1/02 (lymph node) – Extensive plasmacytosis, capsular fibrosis and neutrophilic infiltrate.

DIAGNOSIS
Cutis laxa associated with plasma cell dyscrasia

TREATMENT AND COURSE
The patient has been treated with interferon alpha and narrow band UVB phototherapy for eight months with improvement of his erythema and pruritus and reduction of his lymphadenopathy. The phototherapy was discontinued two months ago as his condition improved and pruritus resolved.

REFERENCES
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CHICAGO DERMATOLOGICAL SOCIETY **CASE # 3**

Presented by Amy Paller, M.D. and Peter Bachmann, M.D.
Division of Dermatology, Children's Memorial Hospital, an affiliate of the Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
This 13 year-old girl presented initially in 1994 with asymptomatic, slowly growing nodules on her nose that were first noticed at three years of age. These nodules progressed and, to a lesser extent, were noted to involve the lip and right periorbital area. Intralesional steroid injections, serial surgical resections and laser treatments during the following year resulted in no significant change. Pathologic evaluation of the removed tissue revealed "adenofibroma" or "angiofibroma."

PAST MEDICAL HISTORY
Noncontributory

MEDICATIONS
None

ALLERGIES
No known drug allergy

FAMILY HISTORY
Diabetes in maternal great-grandmother

SOCIAL HISTORY
No siblings

PHYSICAL EXAMINATION
Multiple flesh-colored, non-tender, firm coalescing nodules on the face (nose, lip, right periorbital), most prominent at the perinasal area. Oral exam showed gingival hyperplasia.

LABORATORY RESULTS
Mutation in the capillary morphogenesis gene (CMG)-2: replacement of a leucine residue by arginine (L329R).

HISTOPATHOLOGY
S94-3391 (nose) – Consistent with juvenile hyaline fibromatosis

DIAGNOSIS
Juvenile hyaline fibromatosis

TREATMENT AND COURSE
The condition has gradually progressed over the years with disfigurement of the face. Treatment options are open for discussion.

REFERENCES

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 4

Presented by Joan Guitart, M.D. and Lisa Rhodes, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 44 year-old man presented with a three-month history of slightly tender nodules on his bilateral shins. He was diagnosed with erythema nodosum after biopsy at an outside hospital, and new lesions developed during treatment with prednisone. One month prior to his presentation at Northwestern, he was hospitalized for staphylococcus epidermidis bacteremia and an escherichia coli urinary tract infection. He continued to develop new lesions during treatment with cefazolin during and after his hospitalization. He reported persistent fevers and lower extremity swelling. He denied abdominal pain, diarrhea, anorexia, weight loss and shortness of breath. Vasculitis was suspected, and two punch biopsies were performed.

At an outside hospital, he was noted to have lymphopenia on complete blood count. A bone marrow biopsy was performed which was negative. He was evaluated by Rheumatology for vague joint complaints and paresthesias of his bilateral lower extremities, and was found to have lower extremity sensorimotor polyneuropathy by electromyogram (EMG). Additionally, a three-centimeter mass behind his left knee was biopsied which revealed findings consistent with his previous skin biopsies.

PAST MEDICAL HISTORY

Noncontributory

MEDICATIONS

Calcium with vitamin D
Prednisone

ALLERGIES

None

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

Occasional alcohol use. Patient works as a hospital manager at a veterinary office.

PHYSICAL EXAMINATION

His bilateral lower extremities have multiple reddish-brown papules and plaques, some with superficial scale. Many lesions are slightly indurated, and scattered lesions are eroded with necrotic centers. His bilateral medial upper extremities have slightly hyperpigmented macules and thin erythematous thin papules.

LABORATORY AND RADIOLOGY RESULTS

The following were negative or within normal limits:

Rapid plasma reagin (RPR), antineutrophil cytoplasmic antibodies (P-ANCA, C-ANCA), human immunodeficiency virus (HIV), anti-nuclear antibody (ANA), cytomegalovirus IgM and IgG, Epstein Barr virus antibody and IgM, basic chemistry panel, C-reactive panel, complement C4, hepatitis C polymerase chain reaction, hepatitis B surface antigen, coagulation panel, thyroid stimulating hormone, stool studies, rheumatoid factor, bone marrow biopsy including cultures, bone marrow flow cytometry, chest radiograph, chest computerized tomography scan, bilateral lower extremity dopplers, renal ultrasound transesophageal echocardiogram.

The following were abnormal:

Complete blood count with isolated low white cell count, angiotensin-converting enzyme (ACE), erythrocyte sedimentation rate, Epstein Barr virus IgG, liver transaminases, CH50, complement C3, hepatitis C antibody, urinalysis with trace protein, whole body positron emission tomography showing abnormal subcutaneous activity in bilateral lower extremities.

HISTOPATHOLOGY

03-005780-1 (outside hospital): Suggestive of erythema nodosum.

DP001354-03-I (left foot): Consistent with granulomatous vasculitis. Fungal and acid fast bacilli stains were negative.

DP001354-03-II (right calf): Granulomatous vasculitis. Immunohistochemistry – the perivascular infiltrate was positive for CD68 and negative for CD20 and CD3. Gram stain was negative.

CD000064-03 (consult biopsy): Granulomatous dermatitis and panniculitis. Focal entrapment of small nerve bundles was noted. Fite stain was negative.

DIAGNOSIS

Idiopathic granulomatous process with systemic symptoms and neuropathy; suspect sarcoidosis

TREATMENT AND COURSE

After biopsy, he was initially treated with low-dose prednisone and trimethoprim/sulfamethoxazole with minimal improvement. He reported the development of night sweats and leg cramps with any decrease of prednisone below 7.5mg a day. Within the past two months, however, he reports no new lesions have appeared and his lower extremitiy swelling has improved. Trimethoprim/ sulfamethaxazole was discontinued approximately six weeks ago after a mild rise in the patient's liver transaminases, and the patient continues to note improvement on only prednisone 7.5mg every other day, alternating with 5mg every other day. His liver transaminases are trending downward.

REFERENCES

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 5

Presented by Joaquin Brieva, M.D. and Stacy McClure, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
This 23 year-old woman initially presented in February 2003 for evaluation and treatment of "scars" on her chest, present since 1998. The lesions have progressed in size, number and distribution, spreading to involve the upper back and shoulders. There was no preceding inflammation, and the lesions were asymptomatic. Review of systems was positive for morning stiffness, swollen, tender joints and mild to moderate fatigue.

PAST MEDICAL HISTORY
None

MEDICATIONS
Oral contraceptives

ALLERGIES
No known drug allergies

FAMILY HISTORY
Maternal aunt with lupus erythematosus

SOCIAL HISTORY
Graduate student of the University of Chicago

PHYSICAL EXAMINATION
There are multiple scattered soft, depressed, flesh-colored atrophic macules scattered over the V-area of the neck, upper arms and back. These lesions are focal round fat-like herniations with button-hole softness and cigarette paper wrinkling. They are non-tender and non-inflammatory.

LABORATORY RESULTS
The following were negative or within normal limits:
Complete blood count with differential, comprehensive chemistry panel, anti-Smith, anti-SSA(Ro), anti-SSB(La), anti-Jo-1, anti-RNP/Sm, anti-Scl-70, C3 complement, rheumatoid factor, flourescent treponemal antibody.

The following were abnormal:
Antinuclear antibody (1:320, speckled pattern), anti-double-stranded DNA, anti-histone 2.9 (<1), C4 complement 11 (14-43), rapid plasma reagin, erythrocyte sedimentation rate(46), urinalysis - trace protein.

Pending results:
Antiphospholipid antibodies, human immunodeficiency virus and Lyme disease antibodies.

HISTOPATHOLOGY
DP1366-03-I lesional (left chest) – Consistent with anetoderma. Elastic stain negative.

DP1366-03-II perilesional (left chest) - Normal skin

DIAGNOSIS
Primary Anetoderma

TREATMENT AND COURSE
Tretinoin microgel 0.04% was started in March 2003 with no noticeable improvement to date. She was referred to Rheumatology clinic for the evaluation of possible systemic lupus erythematosus and concluded that she did not yet meet criteria for a diagnosis of systemic lupus erythematosus. She was started on hydroxycholoquine in September 2003.

REFERENCES
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CHICAGO DERMATOLOGICAL SOCIETY CASE # 6

Presented by Amy Paller, M.D. and Naomi Donnelley, M.D.
Division of Dermatology, Children's Memorial Hospital, an affiliate of the Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 4-1/2-year-old Caucasian boy presented with a low-grade fever, mild malaise and an eruption of pruritic, erythematous papules and edematous plaques on his trunk and extremities. Four of the plaques were topped with bullae. He was initially thought to have a viral exanthem with an urticarial component. Within the next 12 hours the patient developed a more extensive bullous eruption and was admitted from the emergency room.

PAST MEDICAL HISTORY

No blistering at birth

MEDICATIONS

Acetaminophen
Hydroxyzine

ALLERGIES

NKDA

FAMILY HISTORY

Mother and maternal grandmother with presumed ichthyosis vulgaris
Mother had one episode of a widespread blistering eruption during college for which she was hospitalized

SOCIAL HISTORY

The patient lives with his parents. His mother was several months pregnant at the time of his admission.

PHYSICAL EXAMINATION

Examination 24 hours after his initial examination showed diffuse erythematous, edematous plaques on his trunk and extremities, with more than 20 large, tense bullae superimposed on these plaques. His face was relatively spared with no periorificial vesicles, crusting or erythema. Nikolsky sign was negative. Several of the bullae had erupted, leaving superficial erosions. His palms revealed scattered erythematous macules, and his soles were unaffected. He was scratching throughout the examination. All mucous membranes were spared. Mild hyperkeratosis and lichenification over flexural areas was noted.

LABORATORY RESULTS

The following were negative or within normal limits:
Complete blood count, comprehensive chemistry panel, urinalysis; nasopharyngeal, tissue and blood cultures

HISTOPATHOLOGY

S03-565 (abdomen) – Superficial subcorneal bullous dermatitis. Direct immunoflourescence from peri-lesional skin showed negative staining for IgG, IgA, IgM and C3.

S03-699 (right buttock) – Hyperkeratosis with superficial epidermal vacuolization, acanthosis and focal superficial hypergranulosis.

DIAGNOSIS

Ichthyosis bullosa of Siemens

TREATMENT AND COURSE

The initial concern of the infectious disease service was staphylococcal scalded skin syndrome, although the picture was not consistent with this. For this reason he was started on IV clindamycin and topical wound care. He rapidly re-epithelialized the very superficial erosions present when the bullae ruptured. Given the clinical appearance of scaling at flexural areas, typical of epidermal hyperkeratosis, and the autosomal dominant inheritance in this family, we considered a mild form of epidermal hyperkeratosis with an atypical presentation. The mother was then biopsied which showed findings classic for icthyosis bullosa of Siemens.

Ichthyosis bullosa of Siemens is a superficial form of epidermolytic hyperkeratosis with autosomal recessive inheritance and a mutation in the gene encoding keratin 2e. Our patient improved rapidly over several days and was discharged with wound care instructions. He completed a 10-day course of clindamycin, despite negative cultures, due to the unknown trigger of his initial blistering. No scarring resulted.

REFERENCES

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 7

Presented by Annette Wagner, M.D. and William Posten, M.D.
Division of Dermatology, Children's Memorial Hospital, an affiliate of the Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 15 month-old female initially presented to our clinic as a three day-old newborn with a vascular lesion on the right side of the scalp noted at birth. A skin biopsy was performed at two weeks of age. In subsequent visits, the lesion has appeared softer, less violaceous and less indurated in appearance. The lesion has been asymptomatic and there is no history of bleeding. With time, the lesion has appeared to atrophy and regress spontaneously.

PAST MEDICAL HISTORY

Ventricular septal defect – surgical correction has not been performed
Mild atopic dermatitis

ALLERGIES

None

MEDICATIONS

Elidel cream

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

Noncontributory

PHYSICAL EXAMINATION

Examination reveals a healthy appearing, well-developed, well-nourished infant. On the right temple, there is a 4x3cm pink to violaceous to erythematous plaque, demonstrating signs of atrophy and regression. The center appears darker with more grayish coloration at the periphery. There is no evidence of ulceration.

LABORATORY RESULTS

The following were negative or within normal limits:
Complete blood count with platelets.

HISTOPATHOLOGY

DP-6865-02 (scalp) – Upper dermis shows dilated capillary vessels. Mid to lower dermis shows rounded patches of basophilic cells, which have irregular lumina lined by plump endothelial cells. No cellular atypia noted. GLUT1 staining done at Boston Children's Hospital is negative.

DIAGNOSIS
Tufted hemangioma

TREATMENT AND COURSE
The patient has been monitored closely for signs of Kasabach-Merritt syndrome, with followup approximately every one to two months. No evidence of coagulopathy has been noted, and the lesion has appeared to regress spontaneously with time.

REFERENCES
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Powell J. Update on hemangiomas and vascular malformations. *Current Opinion Ped* 1999;11(5):457-63.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 8

Presented by Joan Guitart, M.D. and Kim Nussbaum, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 57 year-old woman presented with a 30-year history of recurrent erythematous, pruritic papules on her right medial ankle. The lesions resolved with topical corticosteroids, then recurred in the same location 20 years later. The new skin eruption was recalcitrant to the same therapy and spread to involve her bilateral shins, buttocks, abdomen and posterior neck. Upon presentation, the patient had tense bullae on her bilateral lower extremities in addition to violaceous papules and plaques, despite a new treatment regimen of oral and topical corticosteroids and azathrioprine. Her review of systems was negative for oral erosions or constitutional symptoms. She has been menopausal for the past seven months.

PAST MEDICAL HISTORY

Cholycysectomy
Diabetes
Hypertension
Kidney stones

MEDICATIONS

Allopurinol
Fenofibrate
Fluocinonide cream
Fosinopril
Furosemide
Glipizide
Metoprolol
Potassium citrate
Prednisone

ALLERGIES

Intravenous contrast dye

FAMILY HISTORY

Mother, brother, maternal grandmother and maternal aunt with lichen planus

SOCIAL HISTORY

The patient is single and works as an executive administrator.

PHYSICAL EXAM

There are erythematous-to-violaceous macules, papules and plaques with erosions and crusting on the bilateral anterior and posterior shins. There is edema of both lower extremities. On the bilateral forearms there are violaceous polygonal papules.

LABORATORY RESULTS

The following were negative or within normal limits:
Complete blood count with differential, liver function, electrolytes, creatinine and hepatitis C serologies.

LABORATORY RESULTS
The following were abnormal:
Glucose (176), blood urea nitrogen (23).

HISTOPATHOLOGY
DP010505-02 (right shin) – Acute vesiculobullous dermatitis, suggestive of acute lichenoid process. Direct immunoflourescence showed colloid bodies staining with IgG, IgA and IgM with C3 and fibrinogen on the roof of the blister, suggestive of bullous lichen planus.

DIAGNOSIS
Bullous lichen planus

TREATMENT AND COURSE
The patient subsequently failed combination treatment with topical and oral corticosteroids, azathioprine and oral metronidazole. Later, a trial of oral tacrolimus was discontinued due to recurrent infections. Most recently, acitretin was discontinued due to hyperlipidemia.
Future options are limited by the patient's susceptibility to infection, hyperlipidemia and potential nephrotoxicity of selected agents. Thalidomide is a potential therapeutic option.

REFERENCES
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Cribier B, Frances C, Chosidow O. Treatment of lichen planus: an evidence-based medicine analysis of efficacy. *Arch Dermatol* 1998; 134: 1521-1530.

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 9

Presented by Joaquin Brieva, M.D. and Jill Weinstein, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 30 year-old woman presented with a 19-year history of recurrent upper lip swelling which had become persistent over the past year. The patient's initial episode of lip swelling occurred at age 11 after a period of left-sided facial paralysis which was diagnosed as Bell's palsy. The facial paralysis resolved over several weeks and has never recurred. The lip swelling resolved spontaneously over several days and recurred one to two times per year until approximately two years prior to presentation when the patient began to have almost monthly recurrences. Some of these episodes were accompanied by periorbital swelling, mild tongue swelling or perioral herpes simplex eruptions. The patient was treated successfully with corticosteroid-dose packs for some episodes, and the untreated episodes resolved spontaneously.

One year prior to presentation the patient's upper lip swelling became more permanent. She presented to the Allergy clinic for evaluation, at which time she was thought to have idiopathic angioedema and was treated with a prednisone taper over four weeks, along with fexofenadine and cetirizine. The swelling slightly worsened despite this treatment and subsequent courses of prednisone, and she was ultimately referred to the Dermatology clinic for further evaluation. At presentation, the patient's review of systems was negative for recent tongue or eyelid swelling, trouble breathing, voice changes, sore throat and gastrointestinal upset.

PAST MEDICAL HISTORY

Allergic rhinitis
Bell's palsy
Herpes labialis

MEDICATIONS

Fexofenadine
Cetirizine

ALLERGIES

No known drug allergies

FAMILY HISTORY

Half brother (same father) with intermittent lip swelling
Father with fissured tongue but no history of facial swelling or paralysis

SOCIAL HISTORY

The patient is an architecture student and is married with two children

PHYSICAL EXAM

This is a well-appearing woman with significant edema of the upper lip. Oral examination reveals a fissured tongue but no tongue edema or mucosal lesions. She has no periorbital edema, no involvement of the lower lip and no voice changes or respiratory distress.

LABORATORY RESULTS

The following were negative or within normal limits:

Complete blood count with differential, liver function, C3, C4, CH50.

HISTOPATHOLOGY

DP007413-03 (upper lip) – Non-caseating granulomas consistent with cheilitis granulomatosa

DIAGNOSIS

Melkersson-Rosenthal Syndrome

TREATMENT AND COURSE

The patient received two series of intralesional injections of triamcinolone, first 5mg/cc and then 10mg/cc, with only temporary improvement. She also was given a five-week course of minocycline without significant improvement.

REFERENCES

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 10

Presented by Joaquin Brieva, M.D. and Eva R. Parker, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 31 year-old woman presented with a history of systemic lupus erythematosus and lupus anti-phospholipid antibody syndrome diagnosed at age 20. At age 28, she began to develop painful, pruritic blisters involving her scalp, face, trunk and extremities. Based on a work-up that included biopsies, immunofluorescence and serum antibody titers, she was diagnosed with pemphigus foliaceus and started topical corticosteroids, prednisone and azathioprine. The patient initially improved for several months but then began to worsen on this regimen. Noncompliance issues and failure to keep clinic appointments further complicated her course. She was changed to mycophenolate mofetil but did not tolerate the medication. She subsequently failed dapsone, a medication that was prescribed by a physician in Mexico, and we then began cyclophosphamide. Despite prolonged therapy with cyclophosphamide, she showed no improvement. Due to the severe and refractory nature of her disease, she was considered for stem cell transplant.

PAST MEDICAL HISTORY

Systemic lupus erythematosus manifest by photosensitivity, malar rash and arthritis
Antiphospholipid antibody syndrome with recurrent deep venous thromboses, pulmonary emboli, and miscarriages

MEDICATIONS

Warfarin

ALLERGIES

Metronidazole

FAMILY HISTORY

No history of autoimmune disease.

SOCIAL HISTORY

Noncontributory

PHYSICAL EXAM

Prior to transplant, she had diffuse erythema and scale of her scalp and erythematous, desquamating annular plaques on the nose and perioral and periorbital regions. There were polycyclic, annular patches and plaques with an erythematous rim and peripheral scale scattered diffusely over her trunk and extremities. Currently, she has no active lesions.

LABORATORY RESULTS

The following were negative or within normal limits:
White blood cell count, hemoglobin, hematocrit, platelets, electrolytes, blood urea nitrogen, creatinine, liver function tests, antinuclear antibody (ANA) 8 panel, complement C3, complement C4 and urinalysis.

The following were abnormal:
Desmoglein 1 by ELISA, ANA (1:160, homogeneous pattern), lupus anticoagulant, anticardiolipin IgG (119), anticardiolipin IgM (28).

HISTOPATHOLOGY

ELN-00-14281 (punch biopsy from abdomen) – Superficial vesicular dermatitis with features suggestive of a superficial form of pemphigus such as pemphigus foliaceus. Direct immunofluorescent study demonstrated granular deposits of IgG and C3 in the intercellular regions between keratinocytes in the upper layers of the epidermis, consistent with pemphigus foliaceus. Indirect immunofluorescent study demonstrated positive pemphigus antibodies to a dilution of 1:320.

DIAGNOSIS

Refractory pemphigus foliaceus in the setting of systemic lupus erythematosus

TREATMENT AND COURSE

Due to the refractory nature of this patient's pemphigus foliaceus, she underwent near complete immunoablation with high-dose cyclophosphamide (200mg/kg) with rabbit anti-thymocyte globulin (5mg/kg) followed by T-cell depleted autologous hematopoietic stem cell transplant (HSCT) in August of 2002. Following transplant, the patient experienced neutropenia of seven days duration and developed culture-negative neutropenic fever for one day. She also suffered from grade I nausea and grade II anorexia by the National Cancer Institute Toxicity Criteria. Otherwise, her transplantation course was uncomplicated and without unexpected toxicity. No late opportunistic infections have occurred to this point.

Following HSCT, her skin lesions gradually resolved over the following six months and corticosteroids were tapered off. Serum autoantibody titers became negative six months following HSCT; however, repeat titers were again positive at 1:160 in May 2003. Other laboratory values performed at 11 months post-HSCT include a positive ANA of 8 IU/mL with a homogeneous pattern (reference range <7.5), negative ANA-8 panel and normal complement C3 and C4 levels. Additionally, lupus anticoagulant antibody and anticardiolipin antibodies continued to be positive with anticardiolipin IgG 30, IgM 42 and IgA 33. Despite the recurrence of autoantibodies, the patient continues to be in medication-free, clinical remission from pemphigus foliaceus.

REFERENCES

Cohen MA, Cohen JJ, Kerdel FA. Immunoablative high-dose cyclophosphamide without stem cell rescue in pemphigus foliaceus. *Int J Dermatol* 2002;41(6):340-4.

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 11

Presented by Amy Paller, M.D. and Jon Dyer, M.D.
Division of Dermatology, Children's Memorial Hospital, an affiliate of the Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 6 year-old boy presented with a two-month history of a rash that began on his abdomen and rapidly disseminated. Initially diagnosed as varicella, the eruption did not resolve with time. Clinical and histopathologic evaluation at an outside hospital suggested pemphigus foliaceus, but direct immunofluorescence was negative. Topical steroid and oral prednisone therapy were initiated which did not improve his eruption. At his two-week followup he was referred further evaluation. Given the extent of his rash, he was relatively asymptomatic complaining only of some "burning."

PAST MEDICAL HISTORY

Noncontributory

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

Noncontributory

PHYSICAL EXAMINATION

Well-developed, well-nourished boy with an erythrodermic eruption and pronounced generalized desquamation of thick, yellow scale. Focal areas on the trunk, extremities, and cheeks show thicker, darker scale. Palms are involved and soles to a lesser extent. Oral cavity, eyes, hair and nails are all normal. Minimal lymphadenopathy noted.

LABORATORY RESULTS

The following were negative or within normal limits:
Bone marrow biopsy with T-cell receptor gene rearrangement, peripheral blood T-cell receptor gene rearrangement, chest computerized tomography, Epstein Barr virus and human T-lymphotropic virus types I and II (HTLV).

The following were abnormal:
Flow cytometry (skin): 60% of isolated cells are one immunotype: CD3+ (dim); CD4-; CD8+(strongly); CD16/56 +; CD5-.

Flow cytometry (peripheral blood) initially revealed a profound NK cell deficiency. Repeat studies show improvement but persistence of this deficiency.

T-cell receptor gene rearrangement (skin): Infiltrate is clonal.

HISTOPATHOLOGY

DP009914-02 (left arm, left back) – Atypical lymphocytic infiltrate of upper dermis with pronounced exocytosis/epidermotropism and folliculotropism. The atypical cells are intermediate in size with a hyperconvoluted nuclear contour and some hyperchromasia. The atypical lymphocytes are strongly positive for CD3, CD8, TIA-1 and CD30, and negative for CD4. There was a decreased number of Langerhans cells with CD1a staining. Direct immunofluorescence was negative. Fungal stain showed rare yeast forms in the infundibulum.

DIAGNOSIS

CD8+/ CD56+ cutaneous T-cell lymphoma

TREATMENT AND COURSE

The patient's skin disease evolved rapidly with a decrease in his erythroderma and thickening of individual lesions, suggesting progression towards tumors. Aggressive workup found no evidence of systemic disease. After discussion with many cutaneous T-cell lymphoma experts, pediatric dermatologists and pediatric oncologists, narrow-band ultraviolet light B was begun three times per week, with a plan to recheck viscera every three months and add bexarotene if response was not excellent. During the next three months, all skin lesions cleared with light therapy, and repeat studies to date have shown no involvement beyond the skin. More than nine months from diagnosis, he remains free of worrisome skin lesions with no signs of systemic involvement. A peripheral CD56+ cell deficiency persists on repeat flow cytometry.

REFERENCES

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Kerl H, Shabrawi-Caelen L, Cerroni L. *Seminars Cut Med Surg* 2000;19(2):118-123.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 12

Presented by Joan Guitart, M.D., James Swan, M.D. and Keren Horn, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 63 year-old man with scleromyxedema was first presented at the October 2002 Chicago Dermatologic Society meeting with a two-month history of progressive periocular swelling and redness followed by a rapidly progressing, generalized eruption. He was treated over a six-month period with oral pulse-dose dexamethasone 40mg once daily for four days (160mg/week), three out of four weeks per month. In addition to having an excellent clinical response with resolution of cutaneous manifestations of disease, he also exhibited a decline in paraprotein levels from 0.6 to 0.1g/dL. Adverse effects were not noted. The patient is currently on a maintenance regimen of oral dexamethasone 40mg each Friday of every month. He remains in complete clinical remission with no paraprotein detected upon repeat electrophoresis, now one year after presentation. He has also undergone collection of stem cells for an autologous stem cell transplant should the disease recur in the future.

PAST MEDICAL HISTORY

Noncontributory

MEDICATIONS

Dexamethasone
Sulfamethoxazole/trimethoprim
Lansoprazole

ALLERGIES

No known drug allergies

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

Patient works as a manager at a manufacturing company, is married and has one child.

PHYSICAL EXAM

There are no physical exam findings. There is complete resolution of the previously noted widespread, symmetric waxy, firm, flesh-colored to erythematous 1-3 mm papules. There is no erythema or deep longitudinal furrows of the glabella. There is no evidence of indurated plaques, sclerosis or thickening of skin.

LABORATORY RESULTS

The following were negative or within normal limits:
Serum and urine plasma electrophoresis.

HISTOPATHOLOGY

DP006554-02 (left forearm): Scleromyxedema

DIAGNOSIS

Scleromyxedema

TREATMENT AND COURSE

The patient remains on a maintenance regimen of oral dexamethasone 40mg each Friday of every month and is in complete clinical remission with no paraprotein detected upon repeat electrophoresis, now one year after presentation. He has also undergone collection of stem cells for an autologous stem cell transplant should the disease recur in the future.

REFERENCES

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Jackson EM, English JC. Diffuse cutaneous mucinoses. *Dermatologic Clinics* 2002; 20(3):493-501.

Rayson D, Lust JA, Duncan A, Su WP. Scleromyxedema: a complete response to prednisone. *Mayo Clin Proc* 1999;74(5):481-4.

Rongioletti F, Rebora A. Updated classification of papular mucinosis, lichen myxedematosus and scleromyxedema. *J Am Acad Dermatol* 2001;44(2):273-281.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 13

Presented by Anthony Mancini, M.D. and Stacy McClure, M.D.
Division of Dermatology, Children's Memorial Hospital, an affiliate of the Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 12 month-old female presented at 10 days of age with very dry, cracked, peeling skin present since birth. She was born at term by spontaneous vaginal delivery after an uncomplicated pregnancy. There was no history of a colloidian membrane or blisters. She was otherwise healthy.

PAST MEDICAL HISTORY

Unremarkable

MEDICATIONS

None

ALLERGIES

None

FAMILY HISTORY

Hypertension
Diabetes
No history of ichthyosis or other skin disorders

SOCIAL HISTORY

Lives at home with both parents

PHYSICAL EXAMINATION

This is a well-appearing female with diffuse erythema, fine scale and desquamation. There was very subtle atophoderma within areas of marked ichthyosis on the back and extremities in a Blaschkoian distribution. No scalp alopecia, mucosal lesions, dystrophic nails or dysmorphic features were seen. There was no obvious musculoskeletal deformity.

LABORATORY AND RADIOLOGY RESULTS

The following were negative or within normal limits:
Ophthalmologic exam

The following were abnormal:
Skeletal survey showed punctate calcifications in the periphery of the calcaneus consistent with chondrodysplasia punctata.

DIAGNOSIS

Conradi-Hunermann syndrome (X-linked dominant chondrodysplasia punctata)

TREATMENT AND COURSE

The patient showed rapid improvement with daily short baths and application of Aquaphor. She was fitted for a harness for right hip dysplasia that was not apparent on initial presentation. A repeat skeletal survey showed less prominent punctate calcifications around the margins of the calcaneus.

At six months of age, she was noted to have right foot eversion and a small S-shaped patch of scalp alopecia with some atrophy. On skin exam, she had very faint Blaschkoian follicular atrophoderma over her back and extremities but no ichthyosiform changes. Repeat ophthalmology exam showed no evidence of cataracts. A genetics evaluation was performed, and blood was drawn for molecular diagnosis of Conradi-Hunermann syndrome. The results are pending.

REFERENCES

Ikegawa S, Ohashi H, Ogata T, et al. Novel and recurrent EBP mutations in X-linked dominant chondrodysplasia punctata. *Am J Med Genet* 2000 Oct 2;94(4):300-5. Review.

Kelley RI, Herman GE. Inborn errors of sterol biosynthesis. *Ann Rev Genomics Hum Genet* 2001;2:299-341. Review.

Shuttleworth D, Burns DA. Chondrodysplasia punctata—Conradi-Hunermann type. *Clin Exp Dermatol* 1986 Jan;11(1):73-8.

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 14

Presented by Cheryl Armstrong, M.D. and William Posten, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
This 39 year-old woman was admitted for severe generalized joint pains, which were most severe around the ankles. The patient was noted to have facial cystic lesions which had erupted suddenly four months prior to her hospitalization.

PAST MEDICAL HISTORY
Unremarkable

ALLERGIES
None

MEDICATIONS
Prednisone

FAMILY HISTORY
Noncontributory

SOCIAL HISTORY
Noncontributory

PHYSICAL EXAMINATION
Examination reveals a healthy appearing, well-developed, well-nourished female. On the forehead, nose, cheeks and chin there are confluent cystic nodules, some with purulent discharge. Background hyperpigmentation is prominent. No cysts are noted on the neck, chest or back. There was no evidence of joint swelling.

LABORATORY RESULTS
The following were negative or within normal limits:
Antinuclear antibody, antinuclear cytoplasmic antibodies, testosterone, dehydroepiandrosterone sulfate, angiotensin converting enzyme level, radiographic films of the thoracic, lumbar and sacroiliac spines and both ankles.

The following were abnormal:
Chest radiography demonstrated cardiomegaly (but no bone abnormalities).

DIAGNOSIS
Pyoderma faciale

TREATMENT AND COURSE

The patient was treated with a course of oral prednisone and cefdinir with improvement of her cutaneous and rheumatologic symptoms. As her symptoms improved her oral medications were discontinued, and she was switched to topical clindamycin gel, adapelene gel and tacrolimus ointment with continued improvement of her facial lesions.

REFERENCES

Massa MC, Su WP. Pyoderma faciale: a clinical study of twenty-nine patients. *J Am Acad Dermatol* 1982. 6(1):84-91.

Plewig G, Jansen T, Kligman AM. Pyoderma faciale. A review and report of 20 additional cases: is it rosacea? *Arch Dermatol* 1992. 128(12):1611-7.

Jansen T, Plewig G, Kligman AM. Diagnosis and treatment of rosacea fulminans. *Dermatol* 1994. 188(4):251-4.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 15

Presented by Vikram Khanna, M.D. and Kim Nussbaum, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 63 year-old man presented with a two-week history of erythematous, tender, ulcerated nodules on his bilateral dorsal hands and fingers. The patient denied any recent history of travel, trauma, sick contacts, medication changes or similar skin eruptions in the past. His review of systems was negative for mucosal lesions or constitutional symptoms. Upon further questioning, the patient reported ingestion of four liters of Ruby Red Squirt per day for the past several months.

PAST MEDICAL HISTORY

Cluster headaches
Congestive heart failure
Diabetes
Knee surgery

MEDICATIONS

Amlodipine
Carvedilol
Clonazepam
Diltiazem
Furosemide
Glipizide
Isosorbide mononitrate
Ranitidine
Terazosin
Warfarin
Zolpidem

ALLERGIES

No known drug allergies

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

The patient is married and retired, with three adult children. He worked as a limousine driver.

PHYSICAL EXAM

There were multiple, erythematous, boggy, ulcerated nodules with surrounding erythema on the dorsum and fingers of both hands, left greater than right. There was serous fluid drainage and crusting of some nodules.

LABORATORY RESULTS

The following were negative or within normal limits:

Complete blood count with differential, liver function tests, renal function tests and multiple tissue cultures.

The following were abnormal:

Serum bromide (0.96 mg per deciliter; normal <0.5 mg per deciliter).

HISTOPATHOLOGY

D02-006953 (left hand) – Exoendophytic epidermal hyperplasia with keratinocyte atypia and superficial dermal and intraepithelial abscesses. Fungal and mycobacterial stains were negative for microorganisms.

DIAGNOSIS

Bromoderma

TREATMENT AND COURSE

The patient was treated with colchicine and discontinued the brominated soft drink. He showed significant improvement after four months and is clear at this time.

REFERENCES

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Jih D, Khanna V, Somach S. Bromoderma after excessive ingestion of ruby red squirt. *NEJM* 2003; 348 (19): 1932-1934.

Smith SZ, Scheen SR. Bromoderma. *Arch Dermatol* 1978; 114; 458-9.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 16

Presented by Joaquin Brieva, M.D. and Eva R. Parker, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 73 year-old woman presented with a seven-month history of left leg redness, pain and swelling. These symptoms progressively worsened to the point that ambulation was very painful. She also began to develop large, draining abscesses on her leg. Prior to the start of her symptoms the patient reported minor trauma to her left leg from a dishwasher door. She was evaluated at several outside institutions where three biopsies were non-diagnostic. She was empirically treated with nasal muciprocin ointment, terbinafine and several courses of antibiotics including cefadroxil, trimethoprim/sulfamethoxazole, levofloxacin and minocycline without improvement. The patient was in the Chicago area visiting family in July 2003 and was seen for a second opinion of her worsening condition. A review of systems was positive only for low-grade fevers.

PAST MEDICAL HISTORY

Asthma
Hypertension
Hiatal hernia complicated by gastroesophageal reflux disease
Anxiety and depression

MEDICATIONS

Venlafaxine
Fluoxetine
Rofecoxib
Propoxyphene
Indomethacin
Pentazocine
Hydrochlorothiazide
Atorvastatin
Omeprazole
Loratadine
Cetirizine
Prednisone
Zolpidem

ALLERGIES

Codeine
Iodine

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

The patient is married and has several adult children. She is a retired teacher. She denies tobacco and alcohol use.

PHYSICAL EXAM
At the time of presentation, the left leg had violaceous erythema and pitting edema extending from the foot to the knee. Numerous fluctuant, draining sinuses, abscesses and punched-out ulcers were noted. There was no inguinal lymphadenopathy.

LABORATORY RESULTS
The following were negative or within normal limits:
Bacterial cultures and fungal cultures.

The following were abnormal:
Mycobacterial cultures were positive for growth of *Mycobacterium abscessus* on day eight.

HISTOPATHOLOGY
DP007474-03 (deep wedge excisional biopsy from left leg) – Granulomatous and suppurative inflammation. Bacterial, fungal and mycobacterial stains were negative.

DIAGNOSIS
Cutaneous *Mycobacterium abscessus* infection

TREATMENT AND COURSE
She was started on an extended course of oral antibiotics with appropriate mycobacterial coverage: clarithomycin 500 milligrams twice a day and ciprofloxacin 750 milligrams twice a day. She is slowly improving and reports that the draining lesions are healing and the pain, erythema and edema have decreased. She is receiving her follow-up care at the Cleveland Clinic in Ohio where she permanently resides.

REFERENCES
Nelson BR, Rapini RP, Wallace RJ Jr, Tschen JA. Disseminated Mycobacterium chelonae ssp. abscessus in an immunocompetent host and with a known portal of entry. *J Am Acad Dermatol* 1989;20(5):909-12.

Ozluer SM, De'Ambrosis BJ. Mycobacterium abscessus wound infection. *Australas J Dermatol* 2001; 42(1):26-9.

Sniezek PJ, Graham BS, et al. Rapidly growing mycobacterial infections after pedicures. *Arch Dermatol* 2003;139(5):629-34.

Wallace RJ Jr, Brown BA, Griffith DE. Nosocomial outbreaks/pseudo-outbreaks caused by nontuberculous mycobacteria. *Annu Rev Microbiol* 1998;52(2 Pt 2):453-90.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 17

Presented by Kent Krach, M.D. and Prashant Singri, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 22 year-old woman presented with a five-month history of tender, erythematous lesions on her bilateral lower extremities. Review of systems was negative. Social history was significant for having had a pedicure one month prior to the onset of the lesions. After initial biopsy, she was treated with a two-week course of minocycline without improvement. A repeat biopsy was performed.

PAST MEDICAL HISTORY

Ulcerative colitis

MEDICATIONS

Amoxicillin/clavulanate potassium
Levofloxacin
Oral contraceptives

ALLERGIES

No known drug allergies

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

Noncontributory

PHYSICAL EXAMINATION

There were tender erythematous patches and plaques on the bilateral anterior lower extremities, with occasional telangectasias, serous crusting and mild scaling.

LABORATORY RESULTS

The following were negative or within normal limits:
Chest radiography, purified protein derivative.

The following were abnormal:
Tissue culture (left lower leg) – *Mycobacterium fortuitum* grew after 10 days. Sensitivities demonstrated resistance to cephalosporins, tetracyclines and aminoglycosides, and sensitivity to fluoroquinolones including levofloaxacin, trimethoprim/sulfamethoxazole, imipenem and amoxicillin/clavulanate potassium.

HISTOPATHOLOGY

DP002172-03 (right calf) – Deep suppurative and granulomatous folliculitis. Mycobacterial, fungal and bacterial stains were negative.

DP002762-03 (left lower leg) – Acute and chronic dermal inflammation including neutrophils, lymphocytes and histiocytes. Mycobacterial, fungal, and bacterial stains were negative.

DIAGNOSIS

Atypical mycobacterial infection of the bilateral lower extremities

TREATMENT AND COURSE

The patient was started on oral levofloxacin with gradual improvement of her lesions. After sensitivities returned, amoxicillin/clavulanate potassium was added to her regimen. *Mycobacterium fortuitum* is classified as a fast-growing bacillus, and infection can be seen in immunocompetent patients often with associated with water exposure. During her pedicure, the patient's feet were soaked in a whirlpool footbath which may have been colonized with the offending organism.

REFERENCES

Winthrop KL, Abrams M, Yakrus M, et al. An outbreak of Mycobacterial furunculosis associated with footbaths at a nail salon. *NEJM* 2002; 346:1366-1371.

Gebo KA, Srinivasan A, Perl TM, et al. Pseudo-outbreak of *Mycobacterium fortuitum* on a human immunodefieciency virus ward: transient respiratory tract colonization from a contaminated ice machine. *Clin Infect Dis* 2002; 35(1):32-38.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 18

Presented by Joaquin Brieva, M.D. and Naomi Donnelley, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 70-year-old man presented with a eighteen-month history of an erythematous, pruritic eruption on his upper chest, arms and shoulders. He was initially seen and biopsied by an outside physician and diagnosed with granuloma annulare. Topical corticosteroids and tacrolimus had provided little to no relief. He denied constitutional systems and specifically denied having fevers, chills, arthralgias or any personal or family history of sun sensitivity.

PAST MEDICAL HISTORY

Hypercholesterolemia
Spinal Stenosis

MEDICATIONS

Aspirin
Atenolol
Calcium
Simvastatin

ALLERGIES

Penicillin

FAMILY HISTORY

Coronary artery disease
No known connective tissue disease

SOCIAL HISTORY

Retired attorney. Recently stopped smoking. Occasional alcohol use.

PHYSICAL EXAMINATION

He has multiple annular and serpiginous, indurated erythematous plaques on the chest and sun-exposed areas of the arms. His face, mucous membranes, palms and soles are uninvolved. He has diffuse underlying dermatoheliosis.

LABORATORY RESULTS

The following were negative or within normal limits:
Complete blood count, comprehensive chemistry panel, anti-nuclear antibody and specific antinuclear antibodies including: Anti-Scl-70, Anti- Jo-1, Anti-Smith, Anti-RNP, Anti-SSA (Ro) and Anti-SSB (La).

The following were abnormal:
Anti-histone antibody – 1.2 (normal is < 1.0).

HISTOPATHOLOGY

DP000512-03 (right arm) – Consistent with annular elastolytic granuloma

DIAGNOSIS

Annular elastolytic granuloma

TREATMENT AND COURSE

The patient improved slightly with sun avoidance. He continued to use his topicals intermittently and was started on pentoxifylline. He continues to have persistent plaques.

REFERENCES

Hanke C.W. et al. Annular elastolytic giant cell granuloma. A clinicopathologic study of five cases and a review of similar entities. *J Am Acad Dermatol* 1979 Nov;1(5):413-21.

Meadows K.P. et al. Erythematous annular plaques in a necklace distribution. Annular elastolytic giant cell granuloma. *Arch Dermatol* 2001 Dec;137(12):1647-52.

Ozkaya-Bayazit E. et al. Annular elastolytic giant cell granuloma: sparing of a burn scar and successful treatment with chloroquine. *Br J Dermatol* 1999 Mar; 140(3):525-30.

CHICAGO DERMATOLOGICAL SOCIETY CASE # 19

Presented by Joan Guitart, M.D. and Lisa Rhodes, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 39 year-old woman presented with a one-week history of a slightly painful eruption in her bilateral axillae. She denies pruritus. She has no history of similar eruptions, and denies using any new products to the area. She avoided using deoderant for one week with no improvement.

PAST MEDICAL HISTORY

Schizoaffective disorder

MEDICATIONS

Clozapin
Paroxetine
Stool softener

ALLERGIES

None

FAMILY HISTORY

Noncontributory

SOCIAL HISTORY

Noncontributory

PHYSICAL EXAMINATION

Bilateral axilla with confluent hyperkeratotic reddish-brown papules coalescing into verrucous plaques

HISTOPATHOLOGY

DP005777-03 (right axilla) – Epidermis shows verrucous projections with prominent parakeratosis and granular cytoplasmic changes. Human papilloma virus (HPV) immunohistochemistry was negative.

DIAGNOSIS

Axillary granular parakeratosis

TREATMENT AND COURSE

We recommended calcipotriene topical cream twice a day. Unfortunately, the patient was lost to follow-up.

REFERENCES

Contreras ME, et al. Axillary intertriginous granular parakeratosis responsive to topical calcipotriene and ammonium lactate. *Int J Dermatol* 2003; 42(5): 382-3.

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 21

Presented by Anthony Mancini, M.D., Fernanda Greco, M.D., and Peter Bachmann, M.D.
Division of Dermatology, Children's Memorial Hospital, an affiliate of the Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS
This 10 year-old Caucasian boy presented with a 12-month history of papules on his left arm and left chest. The lesions were asymptomatic and gradually progressed. The patient was otherwise healthy and had no history of recent infections or any constitutional symptoms.

PAST MEDICAL HISTORY
Verrucae vulgaris
Seborrheic dermatitis

MEDICATIONS
None

ALLERGIES
No known drug allergies

FAMILY HISTORY
Systemic lupus erythematosus in paternal grandmother

SOCIAL HISTORY
Noncontributory

PHYSICAL EXAMINATION
There are flesh-colored to slightly yellow, firm, non-tender, non-inflamed papules coalescing into linear plaques over the left anterior chest and left medial arm, extending down toward the left forearm.

LABORATORY RESULTS
The following were negative or within normal limits:
Complete blood count with differential, chemistry panel, thyroid function studies, urinalysis, quantitative immunoglobulin study, serum protein electrophoresis, antinuclear antibody (1:40, speckled), anti-double-stranded DNA, anti-extractable nuclear Ag, anti-Scl 70, anti-Jo1, anti-Smith, anti-SS-A (Ro), anti-SS-B (La) and anti-U1RNP.

HISTOPATHOLOGY
311-Q28-0512-0 (left upper chest) – Dermal mucinosis with mild superficial and deep lymphocytic infiltrate

DIAGNOSIS
Linear mucinosis

TREATMENT AND COURSE
No satisfactory therapeutic options exist for this idiopathic form of mucinosis. This patient's condition has been stable so far with neither spontaneous resolution nor progression.

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 20

Presented by Amy Paller, MD and Jill Weinstein, MD
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 13 year-old girl presented for evaluation of areas of scarring and redness on her face, bilateral extremities and trunk which began during her first year of life and had progressed. At birth, the patient was noted to have an occipital sore as well as areas of scarring and telangectasias on her face. The occipital sore healed with scarring and telangectasias and was subsequently surgically reduced. The facial scarring persisted and became more telangectatic with time. At six months of age, the patient was noted to have similar lesions on the left side of the body which progressed to linear streaks involving a significant portion of her bilateral upper and lower extremities and trunk. The lesions are slightly uncomfortable but not tender or pruritic.

During the year prior to presentation, the patient used betamethasone dipropionate lotion for the affected areas on the body and desonide lotion for the affected areas on the face. She reports that these medications might have decreased the redness. The patient is otherwise healthy. She wears glasses for myopia but has no other ocular or visual problems. She denies bone pain or history of bony abnormalities. She has a history of a thermal burn to her inner thighs which healed with significant hypopigmentation and scarring.

PAST MEDICAL HISTORY

Induced vaginal delivery at 38.5 weeks gestation, no complications
Surgical reduction procedure for occipital telangectatic scar
Thermal burn to bilateral inner legs

MEDICATIONS

Betamethasone dipropionate lotion
Desonide lotion

ALLERGIES

No known drug allergies

FAMILY HISTORY

No history of any focal skin abnormalities

SOCIAL HISTORY

High school student, lives with mother and father.

PHYSICAL EXAM

Well-appearing girl with focal hypoplastic areas on the face and atrophic, telangectatic streaks in the distribution of Blaschko's lines on her trunk as well as upper and lower extremities. Focal areas of hypopigmented macules are visible within the atrophic streaks, especially on the posterior lower extremities. There is also a scarred, telangectatic plaque on the vertex of her scalp. Eye exam is grossly normal. There is no involvement of the hands and feet, and no dental or bony abnormalities.

LABORATORY RESULTS
The following were negative or within normal limits:
Radiographic studies of left and right humeri and femora.

HISTOPATHOLOGY
DP004283-03 (left thigh) – Suggestive of vascular malformation

DIAGNOSIS
Goltz Syndrome (focal dermal hypoplasia)

TREATMENT AND COURSE
Pimecrolimus cream applied to the affected areas has resulted in moderate improvement, especially of the lesions on the lower extremities.

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CHICAGO DERMATOLOGICAL SOCIETY CASE # 22

Presented by Joaquin Brieva, M.D. and Stephanie Mehlis, M.D.
Department of Dermatology, Feinberg School of Medicine, Northwestern University

HISTORY OF PRESENT ILLNESS

This 41 year-old man presents with a history of blister formation since childhood. The blisters affect his hands, feet, knees and other sites of trauma. He also had photosensitivity as a child. His symptoms improved as he aged. His skin developed a mottled appearance during adolescence. He also complained of scaling and erythema of his palms and soles and a tightness of his hands.

One year prior to his presentation, the patient had a squamous cell carcinoma removed from his left hand. Despite months of radiation, the squamous cell carcinoma metastasized to his axillary lymph nodes. At an outside hospital, he underwent axillary lymph node dissection. His postoperative course was complicated by abscess formation at the site of dissection and subsequent bacteremia. He was transferred to Northwestern Memorial Hospital for further evaluation.

PAST MEDICAL HISTORY

Hypertension
Esophageal and urethral strictures status post repair

MEDICATIONS

Absorbase
Dalteparin
Morphine sulfate
Ondansetron
Paravastatin
Vancomycin
Zolpidem

ALLERGIES

No known drug allergies

FAMILY HISTORY

No family history of any blistering disease. Parents are nonconsanguineous.

SOCIAL HISTORY

Single, no children.

PHYSICAL EXAM

The patient's skin has generalized poikiloderma with mottled hypopigmented patches, reticulated erythema and telangiectasias. His palms and soles show diffuse erythema and hyperkeratosis with sclerodactyly of his hands. The dorsal aspects of his hands and feet have "cigarette paper-like" thinning with dry scale.

DIAGNOSIS

Kindler syndrome with metastatic squamous cell carcinoma

TREATMENT AND COURSE

The patient started combination chemotherapy and radiation therapy for metastatic squamous cell carcinoma. He continued to have difficulty healing after the lymph node dissection and passed away from metastatic squamous cell carcinoma several months after his admission.

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