Table of Contents

1.	Yellow Nail Syndrome	1
2.	Diffuse Subepidermal Calcinosis Cutis & Osteoma Cutis of the Scalp	3
3.	Interstitial Granulomatous Dermatitis with Plaques	5
4.	Congenital Multiple Glomuvenous Malformations (Glomangiomas)	7
5.	Primary Cutaneous Anaplastic Large Cell Lymphoma in the Setting of Lymphomatoid Papulosis	9
	UNKNOWN CASE	
6.	Unspecified Granulomatous Dermatitis	12
7.	Case in Search of a Diagnosis	15
8.	Granulomatous Arteritis	18
9.	Treatment of Primary Cutaneous B-Cell Lymphoma, Follicular Center Cell Type with Rituximab, a Monoclonal Anti-CD20 Antibody	21
10.	Subcutaneous Fat Necrosis of the Newborn	24
11.	Granulomatous Dermatitis & Lymphangitis Consistent with Melkersson-Rosenthal Syndrome	26
12.	Acquired Acrodermatitis Enteropathica in a Patient with Type 1 Diabetes Mellitus & End-Stage Renal Disease	28
13.	Cutaneous Rosai-Dorfman Disease	30
14.	Lupus Vulgaris	32
15.	“Blue” Neurofibromas	34
16.	Treatment of Lichen Striatus with Topical Tacrolimus 0.1% Ointment	36
17.	Progressive Acral Fibromatosis of the Hands	38
18.	Darier Disease (Keratosis Follicularis)	41
19.	Cutaneous Polyarteritis Nodosa Associated with Antiphospholipid Antibody Syndrome	43
20.	Lupus Pernio (Sarcoidosis)	45
21.	Verruciform Xanthoma of the Vulva	48
22.	Metastatic Carcinoid	51

PRESENTERS

Leslie Bernstein, MD and Sarah Stein, MD

HISTORY OF PRESENT ILLNESS

The patient is a 9-year-old girl with a past medical history significant for recurrent pneumonias who presented with a 1-year history of discoloration of the fingernails and toenails. Many of her nails shed, but regrew 3-months prior to presentation. The patient's nails grew slowly and were hard, making them difficult to trim.

PAST MEDICAL HISTORY

Chronic systemic-steroid-induced bilateral cataracts
Recurrent pneumonias
Recurrent bronchiectasis
Recurrent sinusitis

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

8% ciclopirox nail lacquer
Intermittent antibiotics for pulmonary flares
Albuterol inhaler/nebulizer

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Attends school

PHYSICAL EXAMINATION

All 10 toenails had a yellow-green "marble-like" discoloration and were thickened. The nail texture was "spongy," but with a smooth surface. Thumbnails had horizontal ridging. The proximal nail-folds of the fingernails and toenails were absent. Periungual regions were normal. Skin, hair and teeth appeared normal. No peripheral edema was appreciated.

LABORATORY DATA

Nail fungal culture was negative

Chest X-ray and CT scans showed bilateral pleural effusions, bronchiectasis and pansinusitis

Thoracentesis revealed an exudative effusion

Pulmonary function tests suggested mild obstructive lung disease

Complete blood count, basic metabolic panel, hepatic function panel, and sweat chloride test were all within normal limits

DERMATOPATHOLOGY

None

DIAGNOSIS

Yellow nail syndrome

TREATMENT & COURSE

Ciclopirox nail lacquer was discontinued and application of Carmol 40 solution was begun to soften the nails. The patient's family reported nail peeling within 6-weeks and some thinning of the nails, but no dramatic change. Vitamin E caplet application to the nails was initiated as well. The patient's respiratory condition is being followed by pulmonology.

REFERENCES

DeCoste SD, Imber MJ, Baden HP. Yellow nail syndrome. J Am Acad Dermatol 1990;22:608-11.

Pavlidakey GP, Hashimoto K, Blum D. Yellow nail syndrome. J Am Acad Dermatol 1984;11:509-12.

Venencie PY, Dicken CH. Yellow nail syndrome: report of five cases. J Am Acad Dermatol 1984;10:187-92.

PRESENTERS

Amy Farmer, MD, Maria Medenica, MD and Allan Lorincz, MD

HISTORY OF PRESENT ILLNESS

The patient is an 89-year-old African American woman with no significant past medical history who presented with an approximately 50-year history of white papules and nodules on her scalp that recently began to itch.

PAST MEDICAL HISTORY

Hypertension
Gastroesophageal reflux disease
Asthma

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

Metoprolol
Nifedipine
Lasix
Potassium Chloride
Prevacid
Combivent inhaler

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Non-contributory

PHYSICAL EXAMINATION

Multiple white to yellow, non-tender papules and nodules were present diffusely on the scalp, with overlying alopecia.

LABORATORY DATA

Parathyroid hormone, calcium, magnesium, phosphorous, creatinine, and liver function tests were all within normal limits.

Anterior-posterior and lateral skull radiographs demonstrated calcification of the subcutaneous tissues of the scalp. Minimal hyperostosis frontalis interna was also noted.

DERMATOPATHOLOGY

Biopsy of a scalp lesion showed parakeratosis and an epidermis that was focally acanthotic as well as focally thin. Just beneath the epidermis were large, blue-purple, amorphous deposits as well as bone formation. Von Kossa stain confirmed these blue-purple deposits to be calcium.

DIAGNOSIS

Diffuse subepidermal calcinosis cutis and osteoma cutis of the scalp

TREATMENT & COURSE

The patient has received topical agents to alleviate pruritus.

REFERENCES

Shmunes E, Wood MG. Subepidermal calcified nodules. Arch Dermatol. 1972;105:593-597.

Woods B, Kellaway TD. Cutaneous calculi: subepidermal calcified nodules. Br J Derm 1963;75:1-11.

Touart DM, Sau P. Cutaneous deposition diseases. Part II. J Am Acad Dermatol 1998;39:527-544.

PRESENTERS

Amy Farmer, MD and Maria Medenica, MD

HISTORY OF PRESENT ILLNESS

The patient is a 76-year-old white woman with osteoarthritis and carpal tunnel syndrome who presented with a 4-month history of an asymptomatic, indurated, erythematous plaque on her low back. Additional lesions developed over the subsequent 2-months on the bilateral lower extremities.

PAST MEDICAL HISTORY

Osteoarthritis
Basal cell carcinoma of the nose
Carpal tunnel syndrome
Diverticulosis

PAST SURGICAL HISTORY

Left shoulder arthroplasty
Right hip arthroplasty

MEDICATIONS

Neurontin
Metamucil
Fosamax
Aspirin
L-Lysine

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Non-contributory

PHYSICAL EXAMINATION

A 20 by 10 cm indurated erythematous plaque was present on the skin overlying the sacrum. The plaque had a firm, palpable rim with central clearing. Additional lesions of similar morphology were located symmetrically on the lower extremities.

LABORATORY DATA

ANA 1:320, diffuse (<1:160)

ESR 23 (0-20)

Lyme IgM & IgG antibodies, anti-dsDNA antibody, SSA & SSB antibodies, TSH, rheumatoid factor, complete blood count with differential were all within nomal limits

DERMATOPATHOLOGY

Skin biopsies from the sacrum and left popliteal fossa showed an unremarkable epidermis. In the entire dermis and focally in the subcutaneous tissue, there was an infiltrate which consisted of a small number of lymphocytes and a considerable number of foamy histiocytes, epithelioid cells, multinucleated giant cells as well as eosinophils, particularly in the lower dermis. This infiltrate was seen perivascularly and interstitially between collagen fibers.

DIAGNOSIS

Interstitial granulomatous dermatitits with plaques

TREATMENT & COURSE

The lesions are resolving without therapy.

REFERENCES

Long D, Thiboutot DM, Majeski JT, Vasily DB, Helm KF. Interstitial granulomatous dermatitis with arthritis. J Am Acad Dermatol 1996;34:957-961.

Tomasini C, Pippione M. Interstitial granulomatous dermatitis with plaques. J Am Acad Dermatol 2002;46:892-9.

Chu P, Connolly MK, LeBoit PE. The histopathologic spectrum of palisaded neutrophilic and granulomatous dermatitis in patients with collagen vascular disease. Arch Dermatol 1994;130: 1278-1283.

Aloi F, Tomasisni C, Pippione M. Interstitial granuolomatous dermatatitis with plaques. Am J Dermatopathol 1999;21:320-3.

PRESENTERS

Iris Kedar, MD, Maria Medenica, MD, and Sarah Stein, MD

HISTORY OF PRESENT ILLNESS

The patient is a 13-year-old young man who presented for evaluation of lesions on his face and trunk. The lesions on his face and right chest had been present since birth. The lesion on the right chest had increased in size slowly over time, and more recently new spots were noted around the lesion. The lesions were not painful, and had not bled. There was no history of GI bleeding or hematuria.

PAST MEDICAL HISTORY

Mild scoliosis

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

None

ALLERGIES

None known drug allergies

FAMILY HISTORY

The patient's mother did not have similar lesions, but the patient's father was not available for examination.

SOCIAL HISTORY

Attends school

PHYSICAL EXAMINATION

On the right chest, there was a 1 by 0.5 cm vascular reddish-blue macule surrounded by several pinpoint macules. On the right cheek, there were two 1 to 2 mm vascular purplish macules. Two similar reddish-purple macules were noted on the back. No pain was elicited on palpation. No pulsation was appreciated.

LABORATORY DATA

None

DERMATOPATHOLOGY

Biopsy of a lesion on the right chest showed irregular ectatic vascular spaces in the lower dermis surrounded by a single layer of endothelial cells and a few layers of glomus cells. The lesion was not encapsulated.

DIAGNOSIS

Congenital multiple glomuvenous malformations (glomangiomas)

TREATMENT & COURSE

The patient and family were reassured that these lesions are benign. He has had no treatment, but is interested in having these lesions removed.

REFERENCES

Blume-Peytavi U, Adler YD, Geilen CC, et. al. Multiple familial cutaneous glomangioma: a pedigree of 4 generations and critical analysis of histologic and genetic differences of glomus tumors. J Am Acad Dermatol 2000;42:633-9.

Brouillard P, Vikkula M. Vascular malformations: localized defects in vascular morphogenesis. Clin Genet 2003;63:340-51.

Brouillard P, Boon LM, Mulliken JB, et. al. Mutations in a novel factor, glomulin, are responsible for glomuvenous malformations ("glomangiomas"). Am J Hum Genet 2002;70:866-74.

Calduch L, Monteagudo C, Martinez-Ruiz E, et. al. Familial generalized multiple glomangiomyoma: report of a new family, with immunohistochemical and ultrastructural studies and review of the literature. Pediatr Dermatol 2002;19:402-8.

Carvalho VO, Taniguchi K, Giraldi S, et. al. Congenital plaquelike glomus tumor in a child. Pediatr Dermatol 2001;18:223-6.

Iqbal A, Cormack GC, Scerri G. Hereditary multiple glomangiomas. Br J Plast Surg 1998;51:32-7.

PRESENTERS

Amy Priess, MD, Eugene Mandrea, MD, Maria Medenica, MD, and Sarah Stein, MD

HISTORY OF PRESENT ILLNESS

The patient is a 17-year-old Caucasian man with lymphomatoid papulosis and atypical nevi who presented with a 1-month history of a large ulcerated lesion on the right wrist, and similar lesions on the left lower extremity. The patient was not aware of a precipitating event, but did acknowledge a recent trauma to the lesion of the right wrist. The patient has had multiple skin papules, some of which have been ulcerated in the past, which have all healed spontaneously. He has had numerous skin biopsies, as recent as 9-months prior to this presentation. All biopsies were consistent with lymphomatoid papulosis. The review of systems was unremarkable. The patient denied any fatigue, fevers or weight loss, and stated he was otherwise feeling well.

PAST MEDICAL HISTORY

Lymphomatoid papulosis
Atypical nevi
Mononucleosis

PAST SURGICAL HISTORY

Excision of atypical nevi

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

No family history of skin cancers

SOCIAL HISTORY

Active high school student

PHYSICAL EXAMINATION

This was a well-appearing, well-developed young man in no apparent distress. Examination of the skin revealed a 4 by 4 cm fungating and ulcerated lesion with minimal exudate on the right wrist. On the left lower extremity, there was a 3 by 4 cm crusted nodule on the medial knee and a 4 by 4 cm ulcerated lesion with a small amount of exudate on the lateral calf. There was a 3 by 3 cm violaceous plaque on the right anterior thigh. There were violaceous, crusted plaques elsewhere on the legs. A firm, mobile, nontender lymph node was palpable on the right upper inner arm. There was no other significant lymphadenopathy. No hepatosplenomegaly was noted.

LABORATORY DATA

EBV EA antibody titer 40 (< 10)

EBV capsid IgG 40 (< 10)

Complete blood count with differential, complete metabolic panel with LDH, HTLV I/II, HIV, quantitative immunoglobulin A, G, and M levels, lymphocyte subsets, ESR, EBV nuclear antigen antibody and EBV capsid IgM were all within normal limits

Infused CT scan of the chest, abdomen and pelvis demonstrated moderate splenomegaly and multiple retroperitoneal and a few mesenteric nodes, all measuring less than 1 cm. The lungs and pelvis were unremarkable. A repeat CT after treatment demonstrated resolution of previously seen splenomegaly and adenopathy.

DERMATOPATHOLOGY

Biopsy of the right arm showed an absent epidermis. In the dermis, there was a dense infiltrate of numerous lymphoid cells, many of which were large and had irregular, large, hyperchromatic nuclei. Scattered small lymphocytes were seen as well. Immunohistochemical studies were performed. T cell markers, including CD2, CD3, CD4 and CD5, were variably positive. CD20, a marker for B cells, showed occasional positivity. The largest number of cells was positive for CD30, a marker for activated B and T cells. The cells were anaplastic lymphoma kinase fusion protein (ALK) negative.

DIAGNOSIS

Primary cutaneous anaplastic large cell lymphoma in the setting of lymphomatoid papulosis

TREATMENT & COURSE

The patient was referred to radiation oncology and underwent a 21-day course of radiation therapy to the lesions on his right wrist, left knee and calf. The patient tolerated the treatment extremely well, with no appreciable adverse events. At the completion of treatment, the right wrist lesion measured 3 by 3.5 cm (compared to 4 by 4 cm at the start of treatment), the left medial knee lesion measured 1 by 2 cm (compared to 3 by 4 cm at the start of treatment) and was nearly crusted over, and the left lateral calf lesion measured 2.5 by 3 cm (compared to 3 by 4 cm at the start of treatment). At 2-months follow-up, these lesions have continued to improve. Lesions on the right wrist and left knee are completely healed leaving a pink scar. The left calf lesion is a pink scar with an eschar. Treatment with methotrexate is being considered.

REFERENCES

Aoki M, Niimi Y, Takezaki S, Azuma A, Seike M, Kawana S. CD30+ lymphoproliferative disorder: primary cutaneous large cell lymphoma followed by lymphomatoid papulosis. Br J Dermatol 2001;145:123-126.

Bekkenk MW, Geelen FAMJ, van Voorst Vader PC, Heule F, Geerts ML, van Vloten WA, Meijer CJLM, Willemze R. Primary and secondary cutaneous CD30+ lymphoproliferative disorders: a report from the Dutch Cutaneous Lymphoma Group on the long-term follow-up data of 219 patients for diagnosis and treatment. Blood 2000;95:3653-3661.

Demeierre MF, Goldberg LJ, Kadin ME, Koh HK. Is it lymphoma or lymphomatoid papulosis? J Am Acad Dermatol 1997;36:765-772.

Drews R, Samel A, Kadin ME. Lymphomatoid papulosis and anaplastic large cell lymphomas of the skin. Sem Cut Med Surg 2000;19:109-117.

LeBoit PE. Lymphomatoid papulosis and cutaneous CD30+ lymphoma. Am J Dermatopathol 1996;18:221-235.

Rifkin S, Valderrama E, Lipton JM, Karayalcin G. Lymphomatoid papulosis and Ki-1+ anaplastic large cell lymphoma occurring concurrently in a pediatric patient. J Pediatr Hematol/Oncol 2001;23:321-323.

Troussard X, Galateau F, Moreau A, Reman O, Leporrier M, Leroy D. Spectrum of CD30 lymphoproliferative diseases from lymphomatoid papulosis to anaplastic large cell lymphoma. Int J Dermatol 1996;35:842-848.

Willemze R, Beljaards RC. Spectrum of primary cutaneous CD30 (Ki-1)-positive lymphoproliferative disorders, a proposal for classification and guidelines for management and treatment. J Am Acad Dermatol 1993;28:973-980.

Willemze R, Kerl H, Sterry W, Berti E, Cerroni L, Chimenti S, Diaz-Perez JL, Geerts ML, Goos M, Knobler R, Ralfkiaer E, Santucci M, Smith N, Wechsler J, van Vloten WA, Meijer CJLM. EORTC classification for primary cutaneous lymphomas: a proposal from the Cutaneous Lymphoma Study Group of the European Organization for Research and Treatment of Cancer. Blood 1997;90:354-371.

Zackheim HS, Vonderheid EC, Ramsay DL, LeBoit PE, Rothfleisch J, Kashani-Sabet M. Relative frequency of various forms of primary cutaneous lymphomas. J Am Acad Dermatol 2000;43:793-796.

PRESENTERS

Michael Jude Welsch, MD, Anthony Mancini, MD, Christopher Shea, MD, Maria Medenica, MD, and Sarah Stein, MD

HISTORY OF PRESENT ILLNESS

The patient is a 6-year-old Hispanic girl who presented with a 2-year history of asymptomatic lesions on the face. The lesion started as a small red area on the left cheek and had progressed slowly over the course of 2 years. The patient had a short course of oral steroids previously that reportedly led to some improvement of the lesions. Complete review of systems was negative for additional complaints. The patient was previously evaluated at Children's Memorial Hospital (CMH) in 2002.

PAST MEDICAL HISTORY

Non-contributory

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Developmentally normal 6-year-old. Her parents are immigrants from Mexico, but the patient was born in Chicago with no travel history outside of the US.

PHYSICAL EXAMINATION

There were red, indurated, well demarcated plaques on bilateral cheeks and periorally.

LABORATORY DATA

Abnormalities were noted in SGOT 48 (8-37), SGPT 78 (8-35), ACE 58.2 (8-52), p-ANCA 21.7 (<12.1), ANA 1:160, speckled (1:160), and IgG 1492 (640-1420)

Complete blood count, complete metabolic panel, CRP, ESR, IgA & IgM, anti-dsDNA, ASCA were all within normal limits

Tissue cultures for bacteria, mycobacteria, and fungi were negative

The chest x-ray was normal

Ophthalmology evaluation showed no ocular pathology

GI series with small bowel follow through revealed no evidence of inflammatory bowel disease. Elevated p-ANCA may be seen in ulcerative colitis, but the patient is asymptomatic. Colonoscopy is being considered.

Liver biopsy in 2002 at CMH demonstrated micro and macro vesicular steatosis with focal mild lobular inflammation and hepatocellular dropout that may be compatible with nonalcoholic steatohepatitis. No diagnostic features of autoimmune hepatitis were identified.

DERMATOPATHOLOGY

Biopsy of a lesion on the left cheek showed an epidermis with slight spongiosis. The dermis had a superficial and deep perivascular and interstitial infiltrate of lymphocytes, numerous plasma cells, occasional eosinophils, histiocytes, and multinucleated giant cells. Well-formed granulomas were identified at the base of the specimen. Observation with polarized light was negative for birefringent material. The GMS stain was negative for bacteria and the Fite stain was negative for mycobacteria.

Biopsy of the oral mucosa revealed an extensive lymphohistiocytic infiltrate surrounding adnexal structures and salivary gland tissue. There were a few collections of histiocytes forming vague-defined granulomas surrounded by lymphocytes and plasma cells. The dense infiltrate also involved the interface between the mucosa and submucosa and was seen surrounding nerve bundles. Special stains, including PAS, GMS, gram and for acid fast bacteria, were negative.

DIAGNOSIS

Unspecified Granulomatous Dermatitis

TREATMENT & COURSE

A trial of twice daily application with a high potency topical steroid was initiated. The patient has been referred to rheumatology for further evaluation. Treatments prior to our evaluation included oral prednisone for 3 months. Oral erythromycin and sulfasalazine were ineffective. Topical therapies included aclometasone, tacrolimus, and adapalene. The patient reportedly cleared while on systemic corticosteroids but recurred off treatment and had minimal response to the alternate treatments.

REFERENCES

English III, JC, Patel PJ, Greer KE. Sarcoidosis. J Am Acad Dermatol 2001;44:725-43.

Kahn K, Schwarzenberg SJ, Sharp H, Greenwood D. Role of serology and routine laboratory tests in childhood inflammatory bowel disease. Inflamm Bowel Dis 2002;8:325-9.

van der Waal RI, van der Meij EH, van de Scheur MR, Starink TM, van der Waal I. Cheilitis Granulomatosa: and overview of 13 patients with long-term follow up-results of management. Int J Dermatol 2002;41:225-9.

PRESENTERS

Lisa Carroll, MD, Cheuk Yung, MD, and Maria Medenica, MD

HISTORY OF PRESENT ILLNESS

The patient is a 39-year-old Caucasian man who presented with a 6-month history of an asymptomatic eruption on the face and neck. The eruption began as a solitary lesion on the face and progressed to involve the face and neck. As multiple new lesions developed and the older lesions failed to regress, the patient presented to our clinic for further evaluation and treatment. He denied central nervous system, ocular, respiratory, gastrointestinal, and musculoskeletal symptoms.

PAST MEDICAL HISTORY

Non-contributory

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Non-contributory

PHYSICAL EXAMINATION

There were multiple, pink-tan, indurated papules and plaques ranging from 0.4 to 1.2 cm on the forehead, temples, lateral cheeks, and antero-lateral neck. Lesions appeared yellow on diascopy. The surrounding skin was normal in appearance without sclerodermoid changes.

LABORATORY DATA

Complete blood count, comprehensive metabolic panel, ESR, urine and serum protein immunoelectrophoresis, and hepatitis serologies were all within normal limits

Fasting lipid profile showed normal triglycerides 109 (30-149) and HDL 48 (40-80) with minor abnormalities in total cholesterol 205 (120-199) and LDL 135 (60-129)

Chest X-ray and CT scan of the chest, abdomen and pelvis were within normal limits

DERMATOPATHOLOGY

Biopsy of lesions from the right face, left temple, and forehead showed an unremarkable epidermis. In the dermis, there was a dense infiltrate of spindle-shaped cells and epithelioid cells with round to oval nuclei and abundant pale cytoplasm. No nuclear or cytologic atypia or mitotic figures were identified. These cells were arranged in a whorled pattern, at times resembling nests, particularly in the upper dermis. Occasional lymphocytes as well as a moderate number of plasma cells were also noted. Colloidal iron staining was positive for mucin. Immunohistochemical studies were positive for S100 and factor XIII (limited to the dendritic cells surrounding the cells arranged in nests), neuron-specific enolase (NSE) (in a few of the cells arranged in nests), and lysozyme in an occasional cell. Immunohistochemical staining with CD68, desmin, muscle-specific actin (MSA), smooth-muscle actin (SMA), and epithelial membrane antigen (EMA) were negative. Electron microscopy demonstrated collections of histiocyte-like cells, some of which contained numerous lipid droplets. Some of the lipid droplets were electron lucent, indicating a high content of saturated fatty acids. Some of the lipid droplets show mild electron density with a lucent center or a lucent halo. Comma-shaped structures and Birbeck granules were not observed.

An outside hospital biopsy from the face showed a superficial and deep dermal interstitial proliferation of fibroblast-like spindle cells with bland cytologic features, factor XIII positivity, and CD68 and S100 negativity. There were lymphocytic aggregates containing numerous plasma cells. Alcian blue staining confirmed the presence of dermal mucin. Colloidal iron staining performed in our laboratory on the same tissue showed focal deposits of mucin.

DIAGNOSIS

Case in search of a diagnosis

TREATMENT & COURSE

The patient continues to develop additional lesions on the face and neck. None of the lesions have spontaneously resolved. Treatment has been deferred pending a diagnosis.

REFERENCES

Breier F, Zelger B, Reiter H, Gschnait F, Zelger BWH. Papular xanthoma: a clinicopathological study of 10 cases. J Cutan Pathol 2002;29:200-206.

Caputo R, Veraldi S, Grimalt R, Gianotti R, Tosti A, Varotti C, de Kaminsky AR. The various clinical patterns of xanthoma disseminatum. Dermatology 1995;190:19-24.

Ferrando J, Bombi JA. Ultrastructural aspects of normolipidemic xanthomatosis. Arch Dermatol Res 1979;266:143-159.

Gianotti F, Caputo R. Histiocytic syndromes: a review. J Am Acad Dermatol 1985;13:383-404.

Loggini B, Pingitore R, Avvenente A, Giuliano G, Barachini P. Lichen myxedematosus with systemic involvement: clinical and autopsy findings. J Am Acad Dermatol 2001;45:606-608.

Pedicelli C, Angelo C, Palermi G, Paradisi M. Cellular variant of neurothekeoma. Euro J Dermatol 2002;12:209-211.

Pomann J, Rudner E. Scleromyxedema revisited. Int J Dermatol 2003;42:31-35.

Poswig A, Hinrichs R, Megahed N, Hunzelmann N, Kirchberg K, Brenneisen, Krieg T, Scharffetter-Kochanek K. Discrete papular mucinosis—a rare subtype of lichen myxoedematosus. Clin Exp Dermatol 2000;25:289-292.

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

Sanchez RL, Raimer SS, Peltier F, Swedo J. Papular xanthoma: a clinical, histologic, and ultrastructural study. Arch Dermatol 1985;121;626-631.

Wen CY, Ito M, Chen L-D, Nakayama T, Nakashima M, Tsuneda K, Sekine I. Cellular neurothekeoma: A case report with an immunohistochemical study. Acta Med Nagasaki 2002;47:71-73.

Zelger BG, Steiner H, Kutzner H, Maier H, Zelger B. Cellular 'neurothekeoma:' an epithelioid variant of dermatofibroma? Histopathology 1998;32:414-422.

PRESENTERS

Lisa Carroll, MD and Christopher Shea, MD

HISTORY OF PRESENT ILLNESS

The patient is a 78-year-old Caucasian woman who presented with a 6-week history of progressive, tender, cord-like nodules on her arms and legs. Review of systems was significant for decreased appetite with a 30-pound weight loss over the past 10 months, atypical chest pain, intermittent abdominal pain, intermittent hematuria, swelling and erythema of the feet, bilateral hip arthralgias, bilateral thigh myalgias, left foot drop, and severe headaches preceded by transient episodes of aphasia. She denied any pulmonary symptoms.

PAST MEDICAL HISTORY

Idiopathic hypertrophic subaortic stenosis by echocardiograms
Osteoporosis
Hyperparathyroidism
Polyarticular arthritis

ALLERGIES

No known drug allergies

MEDICATIONS

Furosemide
Colchicine
Verapamil
Clopidogrel
Sertraline
Fexofenadine
Centrum silver

FAMILY HISTORY

Non-contributory

PHYSICAL EXAMINATION

Multiple, firm, erythematous, cord-like, linear plaques measuring between 2 and 6 cm in length were present on both forearms and both lower extremities. The lower legs and feet were slightly edematous. The plantar surface of the toes bilaterally appeared violaceous. Neither livedo reticularis nor cutaneous ulceration was noted. 2+ dorsalis pedis, radialis, and carotid pulses were palpable bilaterally. The temporal arteries were nontender. A functional systolic murmur (II/VI) was detected upon cardiac auscultation.

LABORATORY DATA

Complete blood count and complete metabolic panel were within normal limits except for mild abnormalities in hematocrit 35% (36-47%), MCV 79 (81-99), and alkaline phosphatase 139 (30-120)

Serum immunoelectropheresis was suspicious for monoclonal IgM and kappa bands

Urine immunoelectropheresis was suspicious for low concentration monoclonal kappa chain

Urinalysis, TSH, T4, ESR, CRP, c-ANCA, p-ANCA, RPR, and hepatitis panel were all within normal limits

Echocardiogram was unremarkable

Chest x-ray showed no nodules, hemorrhage, or acute cardiopulmonary abnormalities

Fresh tissue culture was negative for bacteria, fungus, and mycobacteria

HISTOPATHOLOGY

Biopsy from the right arm showed a nodular collection of epithelioid and multinucleated cells forming a palisade around a central area of necrosis within the deep dermis and subcutaneous tissue. Elastic tissue stain demonstrated an internal and external elastic lamina associated with the large dermal and subcutaneous nodule. The external lamina exhibited areas of disruption associated with the inflammatory infiltrates. The PAS and GMS stains were negative for fungi. The Fite stain was negative for mycobacteria. Observation with polarized light was negative for birefrigent foreign material. The spindle cells located at the periphery of the nodule were strongly positive for anti-smooth muscle actin (SMA) and muscle specific actin (MSA) and negative for desmin and S100.

Biopsy from the left leg showed a slightly thin epidermis, a deep dermal lymphohistiocytic infiltrate, and a subcutaneous artery with prominent thrombosis within its lumen. A focus of necrosis within the lumen was surrounded by numerous neutrophils, lymphocytes and focal collections of multinucleated giant cells. Acute and chronic inflammatory cells extensively infiltrated the vascular wall. Multinucleated giant cells were also present within the arterial wall. The giant cells, both within the vessel and scattered in the tissue, were positive for CD68. The PAS and GMS stains were negative for fungi. The gram stain was negative for bacteria. The elastic tissue stain demonstrated an internal and external elastic lamina.

DIAGNOSIS

Granulomatous arteritis

TREATMENT AND COURSE

Treatment with prednisone 40 mg once daily for 3-weeks resulted in flattening of most lesions, with residual post-inflammatory hyperpigmentation. The patient was slowly tapered off prednisone, with no evidence of recurrence after 1 month without prednisone.

REFERENCES

Churg A, Churg J, eds. Systemic Vasculitides. New York: Igaku-Shoin Medical Publishers;1991.

Bauza A, Espana A, Idoate M. Cutaneous polyarteritis nodosa. Br J Dermatol 2002;146:694-699.

Crowson AN, Mihm, Jr MC, Magro CM. Cutaneous vasculitis: a review. J Cutan Pathol 2003; 30(3):161-173.

Fiorentino DF. Cutaneous vasculitis. J Am Acad Dermatol 2003;48:311-340.

Frances C, Boisnic S, Bletry O, Dallor A, Thomas D, Kieffer E, Godeau P. Cutaneous manifestations of Takayasu arteritis. Dermatologica 1990;181:266-272.

Perniciaro CV, Winkelmann RK, Hunder GG. Cutaneous manifestations of Takayasu's arteritis. J Am Acad Dermatol 1987;17:998-1005.

Skaria AM, Ruffieux P, Piletta P, Chavaz P, Saurat JH, Borradori L. Takayasu arteritis and cutaneous necrotizing vasculitis. Dermatology 2000;200:139-143.

PRESENTERS

Mario Lacouture, MD, Joseph Baron, MD, Maria Medenica, MD, and Anne Laumann, MBChB, MRCP

HISTORY OF PRESENT ILLNESS

The patient is a 51-year-old African American man who presented for lesions that appeared 4-months after the completion of a course of radiation therapy, with a complete response, for biopsy-proven, follicle center cell type, primary cutaneous B-cell lymphoma. Lesions were similar to initial lesions, which consisted of tender, 0.5 to 2.0 cm, red-brown papules and nodules.

PAST MEDICAL HISTORY

Colon cancer

PAST SURGICAL HISTORY

Hemicolectomy

MEDICATIONS

Procardia
Clonidine
Vancenase
Dolobid

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Alcohol abuse

PHYSICAL EXAMINATION

Approximately 6 well-defined 0.5 to 3.0 cm erythematous nodules were present on his left face.

LABORATORY DATA

Complete blood count and comprehensive metabolic panel were within normal limits except for minor abnormalities in MCV 76.4 (81-99), SGOT 38 (8-37), and SGPT 49 (8-35)
Total body CT showed no evidence of lymph node enlargement
The patient declined a bone marrow biopsy

DERMATOPATHOLOGY

Biopsy of the lesion on the left face showed an unremarkable epidermis. The dermal findings included a dense superficial and deep interstitial infiltrate of atypical lymphocytes with small, round to oval, hyperchromatic nuclei and scant cytoplasm. Scattered, ill-defined nodular areas were also present within the infiltrate. These areas contained lymphocytes with larger irregular nuclei, more dispersed chromatin, occasional prominent nucleoli, and more abundant cytoplasm, suggestive of follicle formation. An immunostain for the B-cell marker CD20 stained the atypical lymphocytes, including those within the nodular areas. These B-cells did not coexpress CD5. Immunostains for the T-cell markers CD3 and CD5 labeled small reactive lymphocytes scattered throughout the infiltrate.

DIAGNOSIS

Primary cutaneous B-cell lymphoma, follicle center cell type

TREATMENT & COURSE

Treatment with Rituximab, a monoclonal anti-CD20 antibody, was started, at weekly intravenous infusions of 375 mg/m^2, for 4 weeks, following premedication with oral acetaminophen 650 mg, intravenous diphenhydramine 50 mg, and intravenous hydrocortisone 100 mg. The patient tolerated the infusions well, resulting in a complete response.

REFERENCES

Fung MA, Murphy MJ, Hoss DM, Grant-Kels JM. Practical evaluation and management of cutaneous lymphoma. J Am Acad Dermatol 2002;46:325-357

Heinzerling LM, Urbanek M, Funk JO et al. Reduction of tumor burden and stabilization of disease by systemic therapy with Anti-CD20 antibody (Rituximab) in patients with primary cutaneous B-cell lymphoma. Cancer 2000;89:1835-1844.

Kerl RWH, Sterry W, Berti E et al. EORTC classification for primary cutaneous lymphomas: A proposal from the cutaneous lymphoma study group of the European Organization for the Research and Treatment of Cancer. Blood 1997;90:354-371.

Maloney DG, Smith B, Appelbaum FR. The anti-tumor effect of monoclonal anti-CD20 antibody therapy includes direct anti-proliferative activity and induction of apoptosis in CD20 positive non-Hodgkin's lymphoma cell lines [abstract]. Blood 1996;88(Suppl 1):637a.

Massengale WT, McBurney E, Gurtler J. CD20-negative relapse of cutaneous B-cell lymphoma after anti-CD20 monoclonal antibody therapy. J Am Acad Dermatol 2002;46:441-443.

Sabroe RA, Child FJ, Woolford AJ, Spittle MF, Russell-Jones R. Rituximab in cutaneous B-cell lymphoma: a report of two cases. Br J Dermatol 2000;143:157-161.

Tedder TF, Engel P. CD20: a regulator of cell cycle progression of B-lymphocytes. Immunol Today 1994;91:1644-1652.

Willemze R, Kerl H, Sterry W, Berti E et al. EORTC classification for primary cutaneous lymphomas: a proposal from the Cutaneous Lymphoma Study Group of the European Organization for Research and Treatment of Cancer. Blood 1997;90:354-371.

PRESENTERS

Lucile White, MD, Maria Medenica, MD, and Sarah Stein, MD

HISTORY OF PRESENT ILLNESS

The patient is a 14-day-old African-American infant who presented to clinic for evaluation of a growth on her back. The lesion was noted at birth to be a pink patch, but over the past 2 weeks it had progessively enlarged and become more violaceous. The child was eating and sleeping normally. Prior to her appointment, she had received a 5-day course of antibiotics for the lesion on her back. The infant was born via a full-term, vaginal delivery and found to have a tight nuchal cord. Her APGAR was 6, but after oxygen administration increased to 8 and 9. The mother had received standard prenatal care, but experienced prolonged rupture of membranes. The infant's septic work-up at birth was negative.

PAST MEDICAL HISTORY

Non-contributory

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

The infant lives with her mother and grandmother.

PHYSICAL EXAMINATION

On physical exam, this was a well-appearing, active infant. Left of the midline in the thoracic area, she had a 2.0 by 1.5 cm smooth, violaceous, soft, fluctuant nodule. The vertebrae were palpable to the right of the lesion. During the punch biopsy, the lesion expressed a clear fluid mixed with a thick, soft, light-yellow substance.

DERMATOPATHOLOGY

Biopsy of the lesion showed an unremarkable epidermis. The mid-dermis contained a dense perivascular infiltrate and interstitially increased number of fibroblasts with occasional giant cells and necrosis of lipocytes. The subcutaneous tissue showed needle-shaped clefts in the adipocytes and foamy histiocytes.

DIAGNOSIS

Subcutaneous fat necrosis of the newborn

TREATMENT & COURSE

Upon follow-up, the lesion had flattened and was fading. Her calcium level at day of life 28 was normal.

REFERENCES

Balfour E, Antaya R, Rossitza L. Subcutaneous fat necrosis of the newborn presenting as a large plaque with lobulated cystic areas. Cutis 2002;70:169-173.

Burden A, Krafchik B. Subcutaneous fat necrosis of the newborn: a review of 11 cases. Pediatr Dermatol 1999;16:384-387.

Requena L, Yus E. Panniculitis. Part II. Mostly lobular panniculitis. J Am Acad Dermatol 2001;45:325-361.

Taeusch HW, Ballard RA, Fletcher J, eds. Avery's Diseases of the Newborn. 7th Ed. Philadelphia: WB Saunders;1998:1198.

Tran JT, Sheth AP. Complications of subcutaneous fat necrosis of the newborn: a case report and review of the literature. Pediatr Dermatol 2003;20:257-261.

PRESENTERS

Lucile White, MD, Christopher Shea, MD, and Anne Laumann, MBChB, MRCP

HISTORY OF PRESENT ILLNESS

The patient is a 32-year-old Hispanic woman with no significant past medical history who presented with a 5-year history of bilateral eyelid swelling, more prominent on the right than the left, with mild facial edema. She subsequently has experienced persistent facial edema and worsening right eyelid swelling. Her periorbital edema was exacerbated in the morning and with increased temperature. Ten years ago she noted nodular areas on her right cheek which resolved with intralesional steroid injections. She denied neurologic, pulmonary, and bowel symptoms. She denied any oral changes.

PAST MEDICAL HISTORY

Non-contributory

PAST SURGICAL HISTORY

Right blepharoplasty

MEDICATIONS

Zoloft
Venoblend vitamin (Ester C, grape seed oil, and butcher's broom)

ALLERGIES

Sulfa drugs

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

She works as an office coordinator and denies alcohol or tobacco use

PHYSICAL EXAMINATION

Nonpitting edema was present in the upper eyelids, right greater than left. The entire face was mildly edematous.

LABORATORY DATA

Complete blood count with differential, iron, transferrin, comprehensive metabolic panel, ANA, SSA & SSB antibodies, anti-ENA-RNP & -Smith antibodies, C1 esterase inhibitor, and ACE were all within normal limits

RAST allergy testing was positive for cat dander and grass

A head MR demonstrated marked swelling of the right upper eyelid. Similar but less severe changes were noted on the left side.

DERMATOPATHOLOGY

Biopsy of the right eyelid showed numerous well-formed granulomas composed of epithelioid and multinucleated histiocytes within the dermis and soft tissue. Many of these had a modest infiltrate of lymphocytes at their periphery. Caseation necrosis was not identified. Observation with polarized light was negative for foreign material. In many areas, collections of foamy, epithelioid histiocytes were also present within the lymphatic spaces. GMS and Fite stains were negative.

DIAGNOSIS

Granulomatous dermatitis and lymphangitis consistent with Melkersson-Rosenthal syndrome

TREATMENT & COURSE

Prior to presenting to the University of Chicago, she received metronidazole cream and laser treatment for rosacea. As the edema worsened, she was treated with intralesional steroids, systemic prednisone, tetracycline, and a 3-week course of Accutane. Her course of Accutane was prematurely discontinued due to hypertriglyceridemia. In November 2001, she had a right blepharoplasty. The eyelid edema recurred 8 to 12 months after the surgery. We placed her on oral niacinamide 1.5 g twice daily, minocycline 50 mg once daily, hydroxychloroquine 200 mg twice daily, and topical tacrolimus twice daily. The patient noted no improvement after several months and discontinued the medications. She desires another blepharoplasty, but the surgeon has requested that medical management be exhausted prior to repeating surgery. We are considering infliximab or etanercept therapy.

REFERENCES

Ang KL, Jones NS. Melkersson-Rosenthal syndrome. J Laryngol Otol 2002;116:386-388.

Heinz C, Weinman I, et al. Bilateral conjuntival lesion in Melkersson-Rosenthal syndrome. Br J Ophthal 2001;85:1267-1268.

Mignogna MD, Fedele S, Lo Russo L, Lo Muzio L. The multiform and variable patterns of onset of orofacaial granulomatosis. J Oral Pathol Med 2003;32:200-5.

Rogers R. Granulomatous cheilitis, Melkersson-Rosenthal syndrome, and orofacial granulomatosis. Arch Dermatol 2000;136:1557-1558.

Shapiro M, Peters S, Spinelli HM. Melkersson-Rosenthal Syndrome in the periocular area: a review of the literature and case report. Ann Plast Surg 2003;50:644-648.

Van der Waal RI, Schulten EA, et al. Cheilitis granulomatosa: overview of 13 patients with long-term follow-up results of management. Int J Dermatol 2002;41:225-229.

PRESENTERS

Linda Wang, MD, Maria Medenica, MD and Shail Busbey, MD

HISTORY OF PRESENT ILLNESS

The patient is a 41-year-old white woman with a history of alcohol abuse and type 1 diabetes mellitus complicated by end-stage renal disease who presented with a 5-month history of a pruritic eruption that began on the upper extremities and progressed to involve the trunk and lower extremities. The patient also experienced hair loss and diarrhea. Topical corticosteroids and antifungals as well as oral antibiotics did not improve her condition.

PAST MEDICAL HISTORY

Alchohol abuse
Chronic pancreatitis
Alchohol withdrawal seizures
Hypertension
Type 1 diabetes mellitus
Retinopathy
Neuropathy
Left foot ulcer and osteomyelitis
End-stage renal disease on hemodialysis
Anemia
Hypothyroidism
Depression

PAST SURGICAL HISTORY

Left arm arteriovenous fistula placement
Peritoneal dialysis catheter placement
Left foot debridement
Left foot trans-metatarsal amputation

MEDICATIONS

Thiamine
Levoxyl
Panokase
Multivitamin
B complex vitamin
Aciphex
Humulin NPH
Epogen
Acetaminophen with codeine
Augmentin

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Non-contributory

PHYSICAL EXAMINATION

There was decreased density of thin brittle scalp hair as well as scaly erythematous patches around the mouth, arms, distal extremities, perineum and buttocks.

LABORATORY DATA

Serum zinc 0.35 mcg/mL (0.66 to 1.10 mcg/mL)

Abdominal ultrasound showed borderline hepatomegaly, chronic pancreatitis, borderline echogenic kidneys consistent with renal disease

DERMATOPATHOLOGY

Biopsy from the left inner thigh revealed confluent parakeratosis with absence of the granular layer and a moderate number of vacuolated cells in the upper layer of a minimally acanthotic epidermis. The papillary dermis showed a few ectatic capillaries, and a minimal lymphohistiocytic infiltrate.

DIAGNOSIS

Acquired acrodermatitis enteropathica

TREATMENT & COURSE

With oral zinc sulfate supplementation, her diarrhea resolved, her hair regrew, and her skin eruption cleared. As diabetes and end stage renal disease are both risk factors for developing acquired acrodermatitis enteropathica, the patient continues on zinc sulfate supplementation.

REFERENCES

Bouda M. Zinc deficiency as a cause of dermatosis in a patient with type I diabetes and renal insufficiency. Vnitr Lek 1994;40:445-7.

Wahid MA, Fathi SAH, Aboul-Khair MR. Zinc in human health & disease. Ric Clin Lab 1988;18:9-16.

PRESENTERS

Linda Wang, MD, Eugene Mandrea, MD, Maria Medenica, MD and Shail Busbey, MD

HISTORY OF PRESENT ILLNESS

The patient is a 33-year-old woman with no significant past medical history who presented to our clinic during the 3rd month of pregnancy with a 6-month history of an asymptomatic plaque on the right chest wall that began as a red papule resembling a "mosquito bite." She denied a history of insect bite, trauma, and a personal or family history of cutaneous lymphoma. An outside hospital biopsy showed a dermal infiltrate composed of small lymphocytes, histiocytes, and scattered plasma cells. Immunohistochemical studies revealed the cells to be predominantly B cells, but T cells were present. The biopsy was interpreted as reactive lymphoid hyperplasia with the recommendation to perform fresh tissue studies if the lesions persisted or recurred. Due to the persistence of the lesion, the patient presented for further evaluation and treatment.

PAST MEDICAL HISTORY

Non-contributory

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

Prenatal vitamins

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Non-contributory

PHYSICAL EXAMINATION

On the right chest wall, there was a 5 by 4 cm, erythematous to violaceous, indurated plaque. No pain was elicited by palpation.

LABORATORY DATA

None

DERMATOPATHOLOGY

Biopsy of the lesion on the right chest wall showed an unremarkable epidermis and papillary dermis. The remaining dermis showed a dense infiltrate composed predominantly of small lymphocytes, a large number of which were CD3-positive and many of which were CD20-positive by immunohistochemical staining. Also noted were scattered plasma cells, which did not exhibit monoclonality as assessed by kappa or lambda light chains. There was a small groups of large histiocytes with round nuclei, pale vacuolated cytoplasm, and indistinct cell membrane, almost all of which were S100-positive and a modest number of which were CD68-positive by immunohistochemical staining. Emperipolesis of lymphocytes and plasma cells was observed. Gene cell rearrangement by polymerase chain reaction (PCR) revealed a polyclonal B- and T-cell lymphoid proliferation without evidence of a clonal population.

DIAGNOSIS

Cutaneous Rosai-Dorfman disease

TREATMENT & COURSE

The lesion has become less indurated, but remains of stable size at 5 by 4 cm.

REFERENCES

Kroumpouzos G, Demierre MF. Cutanous Rosai-Dorfman disease: histopathological presentation as inflammatory pseudotumor. A literature review. Acta Derm Venereol 2002;82:292-296.

Chu P, Leboit PE. Histologic features of cutaneous sinus histiocytosis (Rosai-Dorfman disease) clinically limited to the skin. J Cutan Pathol 1992;19:201-206.

Kang JM, Yang WI, Kim SM, Lee MG. Sinus histiocytosis (Rosai-Dorfman disease) clinically limited to the skin. Acta Derm Venereol 1999;79:363-365.

Child FJ, Fuller LC, Salisbury J, Higgins EM. Cutaneous Rosai-Dorfman disease. Clin Exp Dermatol 1998;23:40-42.

Saenz-Santamaria MC, Reed JA, Ochs RL, McNutt S. Asymptomatic nodules on the chest. Arch Dermatol 1997;133;231-236.

Annessi G, Giannetti A. Purely cutaneous Rosai-Dorfman disease. Br J Dermatol 1996;134;749-753.

Pérez A, Rodríguez M, Febrer I, Aliaga A. Sinus histiocytosis confined to the skin: case report and review of the literature. Am J Dermatopathol 1995;17:384-388.

PRESENTERS

Mario E Lacouture, MD, Christopher R Shea, MD, and Allan Lorincz, MD

HISTORY OF PRESENT ILLNESS

The patient is 37-year-old man who presented with a slowly enlarging, erythematous plaque on his left cheek that had been present since childhood. The lesion had been diagnosed elsewhere as sarcoidosis, based on the histopathologic finding of granulomas. Treatment with intralesional steroids and pulsed-dye laser had been unsuccessful. There were no systemic symptoms or history of infection, BCG immunization, trauma, or arthropod bites.

PAST MEDICAL HISTORY

Non-contributory

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

The patient lived in southern Soviet Union until his late adolescence, when he moved to Israel. He presently lives with his wife and daughter in the United States.

PHYSICAL EXAMINATION

On physical examination, a 4 by 3 cm elevated, erythemato-violaceous plaque with well-defined, irregular borders was present anterior to the left tragus. The surface was smooth, with telangiectasias and a central 2 by 1 cm pink scar, where laser treatments had been performed. An apple-jelly appearance on diascopy was evident. Abdominal examination was normal and no lymphadenopathy was present.

LABORATORY DATA

Mantoux test with 1 tuberculin unit (TU) of purified protein derivative (PPD) was positive, with erythema and induration of 14 mm noted after 48 hours

Tissue culture yielded *Mycobacterium tuberculosis* (MTB), which was sensitive to isoniazid, streptomycin, rifampin, ethambutol, and pyrazinamide

Polymerase chain reaction (PCR) assay targeting the 16s ribosome RNA of MTB complex did not reveal the presence of MTB DNA in a lesional biopsy sample

DERMATOPATHOLOGY

Biopsy from the left cheek showed slight epidermal spongiosis and a superficial and deep nodular and interstitial infiltrate of histiocytes including numerous multinucleated giant cells forming granulomas, with a peripheral infiltrate of lymphocytes and plasma cells. Observation with polarized light revealed only a few collections of birefringent foreign material within the cytoplasm of occasional giant cells. The PAS stain was negative for fungi. There was focal necrosis noted at the center of the granulomas. The Fite stain was negative for mycobacteria.

DIAGNOSIS

Lupus vulgaris

TREATMENT & COURSE

The patient has received isoniazid 300 mg, and pyridoxine 25 mg once daily. The lesion has decreased in size since the patient initiated therapy.

REFERENCES

Barbagallo J, Tager P, Ingleton R, Hirsch R, Weinberg JM. Cutaneous tuberculosis. Diagnosis and management. Am J Clin Dermatol 2002;3:319-328.

Gooptu C, Marks N, Thomas J, James MP. Squamous cell carcinoma associated with lupus vulgaris. Clin Exp Dermatol 1998;23:99-102

Kivanc-Altunay I, Baysal Z, Ekmekci TR, Koslu A. Incidence of cutaneous tuberculosis in patients with organ tuberculosis. Int J Dermatol 2003;42:197-200.

Kumar B, Muraldihar S. Cutaneous tuberculosis: a twenty-year prospective study. Int J Tuberc Lung Dis 1999;3:494-500.

Motta A, Feliciani C, Toto P, De Benedetto A, Morrelli F, Tulli A. Lupus vulgaris developing at the site of misdiagnosed scrofuloderma. J Eur Acad Dermatol Veneorol 2003;17:313-315.

Woo PN, Batta K, Tan CY, Colloby P. Lupus vulgaris diagnosed after 87 years, presenting as ulcerated ‘birthmark.’ Br J Dermatol 2002;146:524-540.

PRESENTERS

Leslie Bernstein, MD, Christopher Shea, MD, and Sarah Stein, MD

HISTORY OF PRESENT ILLNESS

The patient is a 16-year-old man with a past medical history significant for Neurofibromatosis type 1 who presented with a 2-month history of tender, blue nodules on the trunk and extremities.

PAST MEDICAL HISTORY

Neurofibromatosis type 1
Pseudoarthrosis and radial fracture
Paraspinal plexiform neurofibromas with spinal cord compression

PAST SURGICAL HISTORY

External fixation device placement for radial fracture

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Attends high school

PHYSICAL EXAMINATION

Approximately 15 bluish, noncompressible, dermal nodules were present on the anterior and posterior trunk and proximal extremities. Also noted were diffuse café-au-lait macules, axillary freckles, and soft papules typical of cutaneous neurofibromas.

LABORATORY DATA

Complete blood count and sedimentation rate were within normal limits

Brain MRI showed multiple neurofibromata in the cervical canal with foramina widening, multiple intradural neurofibromata along the canal, and cord compression

DERMATOPATHOLOGY

Biopsy from the abdomen demonstrated an unremarkable epidermis. In the mid dermis and portions of the subcutaneous tissue, there was a proliferation of delicate spindle cells within a myxoid stroma. A plexiform configuration was noted. Areas of typical neural differentiation was identified toward the center of several of these aggregates. Mast cells were noted.

DIAGNOSIS

"Blue" neurofibromas

TREATMENT & COURSE

Clarinex 5 mg once daily was recommended, but the patient declined this treatment. The patient is being followed to monitor for progression of lesions.

REFERENCES

Eichenfield LF, Levy ML, Paller AS, Riccardi VM. Guidelines of care for neurofibromatosis type 1. J Am Acad Dermatol 1997;37:625-30.

Kluwe L, Tatagiba M, Funsterer C, Mautner V-F. NF1 mutations and clinical spectrum in patients with spinal neurofibromas. J Med Genet 2003;40:368-71.

Babovic S, Bite U, Karnes P, Babovic-Vuksanovic D. Liposuction: a less invasive surgical method of debulking plexiform neurofibromas. Derm Surg 2003;29:785-7.

PRESENTERS

Leslie Bernstein, MD, Christopher Shea, MD, and Shail Busbey, MD

HISTORY OF PRESENT ILLNESS

The patient is a 20-year-old man with no significant past medical history who presented with an 8-year history of an asymptomatic, semicircular, right-sided truncal rash following trauma to this site. The rash did not resolve despite treatment with hydrocortisone cream, a topical antifungal agent and oral diflucan.

PAST MEDICAL HISTORY

Eczema

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Non-contributory

PHYSICAL EXAMINATION

On the right lateral trunk in a semi-annular distribution, there were erythematous, flat-topped, coalescing papules covered by fine scale in the vicinity of a few small scars

LABORATORY DATA

None

DERMATOPATHOLOGY

Biopsy of the lesion revealed focal parakeratosis and an extensive interface dermatitis with vacuolization of the basal layer and colloid bodies. A lymphocytic infiltrate was noted at the basal layer, the superficial and deep vascular plexus and around a deep eccrine gland. No significant cytologic atypia was evident.

DIAGNOSIS

Lichen striatus

TREATMENT & COURSE

The patient was treated with tacrolimus 0.1% ointment twice daily. The lesions resolved after several weeks of treatment.

REFERENCES

Hauber K. Lichen striatus: clinical features and follow-up in 12 patients. Eur J Dermatol 2000;10:536-9.

Hofer T. Lichen striatus in adults or 'adult blaschkitis?' There is no need for a new naming. Dermatology 2003;207:89-92.

Nutter AF, Champion RH. Lichen striatus occurring as an annular eruption: an acquired 'locus minoris resistentiae.' Brit J Dermatol 1979;101:351-2.

Goyal S, Cohen B. Pathological case of the month. Arch Pediatr Adol Med 2001;155:197-8.

PRESENTERS

Amy Farmer, MD, Maria Medenica, MD, and Shail Busbey, MD

HISTORY OF PRESENT ILLNESS

The patient is a 61-year-old Hispanic man with a 90-pack-year smoking history and coronary artery disease who presented to Rheumatology and subsequently Dermatology clinics with a 1-year history of progressive swelling, pain, sensitivity to cold, and decreased range of motion of both hands. Review of systems was significant for early satiety, progressive dysphagia, and a firmness left of midline on the neck. The patient denied a history of vibratory trauma, foreign travel, and a personal or family history of connective tissue disease.

PAST MEDICAL HISTORY

Hypertension
Coronary artery disease
Gastroesophageal reflux disease
Hyperlipidemia

PAST SURGICAL HISTORY

Appendectomy
Hernia repair

MEDICATIONS

Prednisone
Flexeril
Tramadol
Nifedipine
Plavix
Metoprolol
Ecotrin
Aciphex
Zocor
Glucosamine
Timolol

ALLERGIES

Penicillin

FAMILY HISTORY

Diabetes
Cerebral vascular accident

SOCIAL HISTORY

Born in Puerto Rico, lived last 38-years in the United States with no foreign travel in past 30-years
Worked as an automobile mechanic, but not with vibrating tools
90-pack-year smoking history
Occasional alcohol use

PHYSICAL EXAMINATION

Both hands and wrists were violaceous, severely indurated, and tender to palpation. Firm nodules were present on the palmar surfaces. Active and passive range of motion was absent in all digits. There was a firm mass left of midline on the neck.

LABORATORY DATA

ANA 1:320, speckled (1:160)
ESR 26 (0-20)
Complete blood count with differential, comprehensive metabolic panel, HIV serologies, hepatitis serologies, rheumatoid factor, anti-ds DNA antibody, SSA & SSB antibodies, anti-Scl-70 antibody, anti-ENA-RNA & -Smith antibodies, and complement levels were all within normal limits
MRI of the hands demonstrated soft tissue swelling of the digits, and slightly narrow carpal tunnels
Upper endoscopy revealed gastroesophageal reflux disease
Contrast CT of the neck is pending

DERMATOPATHOLOGY

Biopsy of the left palm showed an unremarkable epidermis. In the entire dermis as well as subcutaneous tissue, there were parallel fascicles composed of both slender and occasional plump fibroblasts between dense collagen bundles as well as thick eosinophilic keloid bundles. Immunohistochemistry study with markers for muscle was performed. Desmin was negative. Muscle specific actin (MSA) and smooth muscle actin (SMA) highlighted vessels that contained muscle in their walls. In addition, the MSA stain demonstrated a modest number of positive fibroblasts within this growth.

DIAGNOSIS

Progressive acral fibromatosis of the hands

TREATMENT & COURSE

The patient's pain was not controlled with Flexeril, Vioxx, or Ultram, and his range of motion did not improve with prednisone 20 mg once daily, colchicine 0.6 mg once daily, minocycline 100 mg once daily or methotrexate 7.5 mg once weekly with folic acid 1 mg once daily. For the last 4-months, the patient has received treatment with prednisone 10 mg daily and colchicine 0.6 mg twice daily without improvement of his progressive dysphagia or range of motion.

REFERENCES

Connelly TJ. Development of Peyronie's and Dupuytren's diseases in an individual after single episodes of trauma: a case report and review of the literature. J Am Acad Dermatol 1999;41:106-108.

Sullivan TP, King LE, Boyd AS. Colchicine in dermatology. J Am Acad Dermatol 1998;39:993-999.

PRESENTERS

Iris Kedar, MD and Christopher Shea, MD

HISTORY OF PRESENT ILLNESS

The patient is a 19-year-old woman who presented with a 5-year history of pruritic "dark spots" on her legs. One year after their onset, she noted the spots on her abdomen. Several months prior to presentation, the lesions had spread to her face and arms. She used hydrocortisone cream for several months, which helped with the itching, but the rash persisted.

PAST MEDICAL HISTORY

Non-contributory

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

None

ALLERGIES

No known drug allergies

FAMILY HISTORY

There was no history of relatives with a similar skin condition. Her father's medical history is unknown.

SOCIAL HISTORY

The patient has one child

PHYSICAL EXAMINATION

Hyperpigmented, keratotic, 2 to 5 mm papules were present on the face, chest, abdomen, arms, and legs. There was relative sparing of the extensor surface of the arms and back. Palms and soles exhibited pits and punctate keratoses. Nails exhibited subungal hyperkeratoses, V-shaped notching, and longitudinal red and white longitudinal lines.

LABORATORY DATA

None

DERMATOPATHOLOGY

Biopsy of a lesion from the abdomen showed orthokeratosis and focal parakeratosis. Extensive acantholysis and dyskeratosis were noted, as well as corps ronds. The dermis had a superficial perivascular infiltrate of lymphocytes and melanophages.

DIAGNOSIS

Darier Disease (Keratosis Follicularis)

TREATMENT & COURSE

The patient was started on tazarotene 0.1% gel applied nightly initially to her face and abdomen. She noted a decrease in the number of lesions, and the remaining lesions were flatter. She is now using tazarotene 0.1% gel on all affected areas. Before starting this medication a β-hCG was checked and was negative. A complete blood count, comprehensive metabolic panel, and lipid panel were also checked because of the concern for systemic absorption of tazarotene with extensive body surface area use, and in anticipation of possibly starting isotretinoin in the future.

REFERENCES

Berg D, Bassett AS. Darier's disease: current understanding of pathogenesis and future role of genetic studies. Int J Dermatol 1993;32:397-400.

Burge SM, Wilkinson JD. Darier-White disease. A review of the clinical features in 163 patients. J Am Acad Dermatol 1992;27:40-50.

Burge S. Management of Darier's disease. Clin Exp Dermatol 1999;24:53-6.

Burkhart CG, Burkhart CN. Tazarotene gel for Darier's disease. J Am Acad Dermatol 1998;38:1001-2.

Cooper SM, Burge SM. Darier's disease: epidemiology, pathophysiology, and management. Am J Clin Dermatol 2003;4:97-105.

Sheridan AT, Hollowood K, Sakuntabhai A, Dean D, Hovnanian A, Burge S. Expression of sarco/endo-plasmic reticulum Ca2+-ATPase type 2 isoforms (SERCA2) in normal human skin and mucosa, and Darier's disease skin. Br J Dermatol 2002;147:670-4.

PRESENTERS

Iris Kedar, MD, Nadera Sweiss, MD, Maria Medenica, MD, and Allan Lorincz, MD

HISTORY OF PRESENT ILLNESS

The patient is a 63-year-old woman who presented with an 8-month history of painful nodules on both legs. Several weeks prior to presentation, she noted swelling and erythema of her legs. Review of systems was remarkable for intermittent pain of her left index finger. She denied fevers or other systemic symptoms.

PAST MEDICAL HISTORY

Leprosy diagnosed at age 32, treated for 10 years with dapsone, rifampin, and thalidomide
Hypercholesterolemia
Hypertension

PAST SURGICAL HISTORY

Tubal ligation

MEDICATIONS

Atorvastatin
Metoprolol

ALLERGIES

Penicillin

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Non-contributory

PHYSICAL EXAMINATION

Tender, erythematous nodules were present on both legs. Livedo reticularis was present on the arms and legs.

LABORATORY DATA

ANA 1:1280, speckled and nucleolar (1:160)
Lupus anticoagulant dilute tissue thromboplastin inhibition 1.2 (<1.1) & dilute Russell's viper venom time 41.1 (31-39) (confirmatory test positive)

Beta 2 glycoprotein IgM antibody 112.3 (<20) & IgA antibody 20.5 (<20)
Anticardiolipin IgM antibody 22 (0-15.6)
CRP 1.0 (<0.6)
Complete blood count, comprehensive metabolic panel, ESR, hepatitis B and C serologies, rheumatoid factor, p-ANCA, SSA & SSB antibodies, anti-ENA-RNP & -Smith antibodies were all within normal limits

DERMATOPATHOLOGY

Incisional biopsy on the right lower leg showed an artery in the lower dermis with a thick and edematous wall. There was focal invasion of neutrophils and modest deposition of fibrin in the vessel wall. A mixed cell infiltrate of neutrophils, lymphocytes, and histiocytes surrounded the vessel. GMS and Fite stains were negative for fungal elements and acid fast organisms respectively.

DIAGNOSIS

Cutaneous polyarteritis nodosa associated with antiphospholipid antibody syndrome

TREATMENT & COURSE

The patient was treated with prednisone 20 mg, clopidogrel 75 mg, and aspirin 81 mg once daily. She responded with resolution of her nodules and livedo reticularis. The prednisone has since been tapered. She continues on clopidogrel and aspirin.

REFERENCES

Bauza A, Espana A, Idoate M. Cutaneous polyarteritis nodosa. Br J Dermatol 2002;146:694-9.

Borrie P. Cutaneous polyarteritis nodosa. Br J Dermatol 1972;87:87-95.

Chen K. Cutaneous polyarteritis nodosa: a clinical and histopathological study of 20 cases. J Dermatol 1989;16:429-42.

Daoud MS, Hutton KP, Gibson LE. Cutaneous periarteritis nodosa: a clinicopathological study of 79 cases. Br J Dermatol 1997;136:706-13.

Diaz-Perez JL, Winkelmann RK. Cutaneous periarteritis nodosa. Arch Dermatol 1974;110:407-14.

Minkowitz G, Smoller BR, McNutt S. Benign cutaneous polyarteritis nodosa. Relationship to systemic polyarteritis nodosa and to hepatitis B infection. Arch Dermatol 1991;127:1520-3.

Pak H, Montemarano AD, Berger T. Purpuric nodules and macules on the extremities of a young woman. Arch Dermatol 1998;134:231-2, 234-5.

PRESENTERS

Amy Priess, MD, Maria Medenica, MD, and Christopher Shea, MD

HISTORY OF PRESENT ILLNESS

The patient is a 44-year-old African-American woman with a past medical history significant for asthma, hepatitis C, and sarcoidosis, which initially appeared as painful, pruritic papules on the left buttock, and progressed to involve the face and lungs. She presented to our clinic with a 10-year history of erythematous papules, plaques and atrophic scarring of the central face, including the nose, upper lip, and cheeks. She had received no treatment at the time of her diagnosis.

PAST MEDICAL HISTORY

Sarcoidosis
Asthma
Hepatitis C
Angina

PAST SURGICAL HISTORY

Repair of left forearm fracture

MEDICATIONS

Acetaminophen with codeine
Aspirin
Prednisone
Gabapentin
Albuterol inhaler
Ranitidine
Nitroglycerin patch

ALLERGIES

No known drug allergies

FAMILY HISTORY

Cousin with sarcoidosis

SOCIAL HISTORY

Smokes 5 cigarettes per day

PHYSICAL EXAMINATION

Red, smooth papules, edematous plaques, and atrophic scarring were present on the nose, upper lip, and cheeks. A saddle nose was also noted. Two hypopigmented plaques were present on the right lower extremity and left buttock.

LABORATORY DATA

Hepatitis C virus antibody was repeatedly reactive

Complete blood count with differential, comprehensive metabolic panel, G6PD, RPR, HIV, hepatitis B surface antigen and antibody were all within normal limits

Chest x-ray showed bilateral hilar lymphadenopathy and interstitial changes

Chest CT showed hilar lymphadenopathy, and moderate to severe centrilobar emphysema

Pulmonary function tests demonstrated a mild obstructive lung disease and decreased diffusing capacity

CT of the maxillary and facial sinus showed inflammatory changes of the sinuses as well as a deformity of the left maxillary sinus

DERMATOPATHOLOGY

Biopsy from the face showed a minimally acanthotic epidermis. Throughout the entire dermis, there were large islands of epithelioid cells admixed with a few lymphocytes and surrounded by a thin layer of lymphocytes. GMS and Fite stains were negative for fungal elements and acid-fast organisms, respectively.

DIAGNOSIS

Lupus Pernio (Sarcoidosis)

TREATMENT & COURSE

Treatment was initiated with clobetasol 0.05% ointment applied twice daily. She was referred to Pulmonary clinic for evaluation and treatment of her lung disease. The patient was started on prednisone 40 mg once daily with clinical improvement noted at 4-weeks follow-up. After a normal baseline eye examination, hydroxychloroquine 200 mg twice daily was begun. Prednisone was tapered and infliximab started. The patient's first infusion of infliximab was complicated only by arthralgias. At follow-up 6-weeks post-infusion, there was no clinical change, and thus, the patient declined further treatment with infliximab. Allopurinol up to 300 mg once daily was then initiated. Treatment with methotrexate was deferred because of the patient's positive hepatitis C status.

REFERENCES

Albertini JG, Tyler W, Miller OF III. Ulcerative sarcoidosis: case report and review of the literature. Arch Dermatol 1997;133:215-219

English, JC III, Patel PJ, Greer KE. Sarcoidosis. J Am Acad Dermatol 2001;44:725-43

Mallbris L, Ljungberg A, Hedblad MA, Larsson P, Stahle-Backdahl M. Progressive cutaneous sarcoidosis responding to anti-tumor necrosis factor-α therapy. J Am Acad Dermatol 2003;48:290-293

Moller DR. Treatment of sarcoidosis – from a basic science point of view. J Int Med 2003;253:31-40

Schwartz RA, Robertson DB, Tierney LM Jr. McNutt NS. Generalized ulcerative sarcoidosis. Arch Dermatol 1982;118:931-933

Thomas KW, Hunninghake GW. Sarcoidosis. JAMA 2003;289:3300-3303

PRESENTERS

Amy Priess, MD, Christopher Shea, MD, and Shail Busbey, MD

HISTORY OF PRESENT ILLNESS

The patient is a 33-year-old Caucasian woman with Turner's syndrome, hypertension, hypothyroidism, hypercholesterolemia, and abnormal pap smears who presented with a 1-year history of verrucous lesions of the vulva which were occasionally pruritic. The lesions had an insidious onset and were not associated with bleeding, ulceration, or crusting. There was no history of local trauma. Neither treatment with high-potency topical steroids nor imiquimod altered the lesions.

PAST MEDICAL HISTORY

Turner's syndrome
Hypertension
Hypothyroidism
Hypercholesterolemia
Abnormal PAP smears
Diverticulitis

PAST SURGICAL HISTORY

Colon resection
Tonsillectomy

MEDICATIONS

Levothyroxine
Atorvastatin
Triamterene

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

The patient is sexually active and uses condoms.

PHYSICAL EXAMINATION

Verruciform, hyperkeratotic, well-circumscribed plaques are present on the inner labia majora.

LABORATORY DATA

None

DERMATOPATHOLOGY

Biopsy of a vulvar lesion showed parakeratosis and neutrophils within the stratum corneum as well as marked epidermal hyperplasia without significant atypia. The papillary dermis contained collections of foam cells and also acute and chronic inflammatory cells. The PAS stain was negative for fungi.

DIAGNOSIS

Verruciform xanthoma

TREATMENT & COURSE

The patient was given reassurance and offered excision as a treatment option, which she declined.

REFERENCES

Barr RJ, Plank CJ. Verruciform xanthoma of the skin. J Cutan Pathol 1980;7:422-8.

Cuozzo DW, Vachher P, Sau P, Frishberg DP, James WD. Verruciform xanthoma: a benign penile growth. J Urol 1995;153:1625-1627.

Duray PH, Johnston YE. Verruciform xanthoma of the nose in an elderly male. Am J Dermatopathol 1986;8:237-240.

Griffel B, Cordoba M. Verruciform xanthoma in the anal region. Am J Proctol Gastroenterol Colon Rectal Surg 1980;31:24-25.

Huguet P, Toran N, Tarragona J. Cutaneous verruciform xanthoma arising on a congenital lymphoedematous leg. Histopathol 1995;26:277-279.

Jensen JL, Liao SY, Jeffes EWB. Verruciform xanthoma of the ear with coexisting epidermal dysplasia. Am J Dermatopathol 1992;14:426-430.

Kimura S. Verruciform xanthoma of the scrotum. Arch Dermatol 1984;120:1378-1379.

Leong FJWM, Meredith DJ. Verruciform xanthoma of the vulva. A case report. Pathol Res Pract 1998;194:661-665.

Mountcastle EA, Lupton GP. Verruciform xanthomas of the digits. J Am Acad Dermatol 1989;20:313-317.

Orchard GE, Jones EW, Jones RR. Verruciform xanthoma: an immunohistochemical study. Br J Biomed Sci 1994;51:28-34.

Ronan SG, Bolano J, Manaligod JR. Verruciform xanthoma of the penis. Light and electron-microscopic study. Urol 1984;23:600-603.

Shafer WG. Verruciform xanthoma. Oral Surg Oral Med Oral Pathol 1971;31:784-789.

PRESENTERS

Michael Jude Welsch, MD, Shail Busbey, MD, and Christopher Shea, MD

HISTORY OF PRESENT ILLNESS

The patient is a 51-year-old African-American man who presented for evaluation of non-painful, subcutaneous nodules on his torso, arms, and face that had appeared insidiously over the past month and a half. The patient was being worked-up for diarrhea when lesions in the liver suggestive of metastases were noted on computed tomographic scan. The patient was scheduled for liver biopsy prior to presentation. The patient had experienced weight loss of 12 pounds, cramping abdominal pain, and frequent loose, non-bloody stools. Flushing, respiratory, and cardiovascular symptoms were absent.

PAST MEDICAL HISTORY

Hypertension

PAST SURGICAL HISTORY

Non-contributory

MEDICATIONS

Nifedipine

ALLERGIES

No known drug allergies

FAMILY HISTORY

Non-contributory

SOCIAL HISTORY

Smoker
History of alcohol abuse, none in past two years

PHYSICAL EXAMINATION

There were scattered, well-defined, hard, mobile, 3 to 8 mm subcutaneous nodules on the shoulders, torso, and face

LABORATORY DATA

Complete blood count was within normal limits except for hemoglobin 8.6 (13.5-17.5) and hematocrit 27% (41-53%)

Basic metabolic panel, liver function tests, and prostate specific antigen were all within normal limits

Infused CT scan showed diffuse hypodensities in the liver with extensive retroperitoneal lymphadenopathy encasing the aorta and inferior vena cava. A single, hyperdense nodule in the left lower lung field was also identified.

Colonoscopy revealed one tubular adenoma

DERMATOPATHOLOGY

Biopsy from the back was performed. Monomorphous small round cells with eosinophilic cytoplasm in small aggregates were present within a fibrous stroma in the subcutis. Numerous mitotic figures, some atypical mitotic figures, and numerous apoptotic cells were noted. Positive immunohistochemical staining was seen for chromogranin and synaptophysin. AE1/AE3 anticytokeratin mixture strongly labeled the tumor in the dermis and subcutis. Anticytokeratin 20 labeled the tumor weakly positive at the edges. The tumor was negative for cytokeratin 7.

DIAGNOSIS

Metastatic carcinoid

TREATMENT & COURSE

The patient's diarrhea was treated with loperamide and supportive care. The patient expired 6 months later as a consequence of septic shock.

REFERENCES

Archer CB, Rauch HJ, Allen MH, MacDonald DM. Ultrastructural features of metastatic cutaneous carcinoid. J Cutan Pathol 1984;11:485-490.

Bart RS, Kamino H, Waisman J, Lindner A, Colen S. Carcinoid tumor of skin: Report of a possible primary case. J Am Acad Dermatol 1990;22:366-370.

Courville P, Joly P, Thomine E, Ziade J, Soubrane JC, Kuhn JM, Lauret P. Primary cutaneous carcinoid tumor. Histopathol 2000;36:566-567.

Durward G, Blackford S, Roberts D, Jones MK. Cutaneous scleroderma in association with carcinoid syndrome. Postgrad Med J 1995;71:299-300.

Eriksson B, Janson ET, Bax NDS, et al. The use of new somatostatin analogues, lanreotide and octastatin, in neuroendocrine gastro-intestinal tumors. Digestion 1996;57:77-80.

Hajdu SI, Winawer SJ, Myers WPL. Carcinoid tumors: A study of 204 cases. Am J Clin Pathol 1974;61:521-528.

Lebtahi R, Cadiot G, Sarda L, et al. Clinical impact of somatostatin receptor scintigraphy in the management of patients with neuroendocrine gastroenteropancreatic tumors. J Nucl Med 1997;38:853-858.

Makridis C, Ekbom A, Bring J, et al. Survival and daily physical activity in patient treated for advanced midgut carcinoid tumors. Surgery 1997;122:1075.

McCracken GA, Washington CV, Templeton SF. Metastatic cutaneous carcinoid. J Am Acad Dermatol 1996;35:997-998.

Öberg, K. Carcinoid tumors: molecular genetics, tumor biology, and update of diagnosis and treatment. Curr Opin Oncol 2002;14:38-45.

Orlefors H, Sundin A, Ahlström H, et al. Positron emission tomography with 5-hydroxy-tryptophan in neuroendocrine tumors. J Clin Oncol 1998;16:2534-2541.

Ritzel U, Leonhardt U, Stockmann F, Ramadori G. Treatment of metastasized midgut carcinoids with dacarbazine. Am J Gastroenterol 1995;90:627-631.

Williams ED, Sadler M. Clarification of carcinoid tumors. Lancet 1963;1:238-239.