

# Posters Presented at the 18th Joint Meeting of the International Society of Dermatopathology, March 18–19, 2015, Holiday Inn Golden Gateway, San Francisco, California, USA

## Rhabdomyomatous Mesenchymal Hamartoma: A Case Report

Matthew B. Strausburg, MD<sup>1</sup>, Stefanie A. Ali, MD,<sup>2</sup> Youssef H. Tahiri, MD<sup>3</sup>, and Matthew Kuhar, MD<sup>1,3</sup>

*Indiana University School of Medicine, Departments of <sup>1</sup>Dermatology, <sup>2</sup>Pathology, and <sup>3</sup>Plastic Surgery.*

Rhabdomyomatous mesenchymal hamartoma (RMH) is a rare, benign hamartoma of the dermis and subcutaneous tissue first described in 1986. Since that time, 39 cases have been reported in the literature. Classically, these tumors are congenital, midline, and solitary. RMH has been associated with congenital anomalies such as cleft lip, bilateral sclerocorneas and retinal dysplasia. Additionally, RMH can be a syndromic feature of amniotic band syndrome, Goldenhar syndrome, and Delleman's syndrome. We report a case of a healthy 2-year-old boy who presented with a firm, flesh-colored, beak-like nodule located on the midline chin. The nodule was asymptomatic and present since birth. Histologically, it demonstrated a dermal proliferation of haphazardly-arranged striated muscle bundles, and other admixed mesenchymal elements including adipose, nerve, collagen, and blood vessels. It has been suggested that these hamartomas result from aberrant embryologic development or migration of mesodermal tissue. Our goal is to raise awareness of the existence of this rare benign entity and that it is a potential syndromic marker.

## Neural, Arterial, and Soft Tissue Calcification in an Unusual Case of Lupus Panniculitis

Stefanie A. Ali, MD<sup>1</sup>, and Matthew Kuhar, MD<sup>1,2</sup>

*Indiana University School of Medicine, Departments of <sup>1</sup>Pathology and <sup>2</sup>Dermatology.*

Dystrophic calcification is the most common type of calcinosis cutis and is associated with a variety of connective tissue disorders but only rarely cutaneous lupus erythematosus. Calcinosis cutis generally involves the skin, subcutaneous tissue, vasculature, muscles, and tendons, but there are no reports of peripheral nerve calcification. We report a case of a patient with known systemic lupus erythematosus who developed an unusual panniculitis with subcutaneous fat necrosis and calcification of the subcutaneous adipose, blood vessel walls, and peripheral nerve segments. Stains for infectious organisms were negative, and the patient improved with immunosuppressive therapy. We suggest this is an unusual presentation of lupus panniculitis with previously undescribed peripheral nerve calcification. Our case also supports a previous suggestion in the literature that when necrosis and calcification are seen in lupus panniculitis, it likely indicates co-existent systemic lupus. While there are reports of basal ganglia calcification in neonatal lupus, this is the first report to our knowledge of peripheral nerve calcification associated with lupus.

## Epithelial Sheath Neuroma: A Mimicker of Malignant Perineural Invasion

Stefanie A. Ali, MD<sup>1</sup>, and Simon Warren, MD<sup>1,2</sup>

*Indiana University School of Medicine, Departments of <sup>1</sup>Pathology and <sup>2</sup>Dermatology.*

Perineural invasion can be seen in common cutaneous carcinomas and is usually associated with malignancy, aggressive behavior, and poor prognosis. However, perineural invasion is not exclusive to malignant conditions. An epithelial sheath neuroma is a benign process of unknown etiology that can mimic malignant perineural invasion. Epithelial sheath neuromas are primarily seen in the elderly, typically presenting as solitary, pruritic, flesh-colored papules on the upper back. Characteristic histologic features include multiple enlarged peripheral nerve fibers ensheathed by mature squamous epithelium. The process is limited to the superficial dermis and is surrounded by a loose myxoid stroma, lymphocytic infiltrate, and sometimes prominent infundibular cysts. The neural elements stain positively with S100, and the epithelial components stain with cytokeratins. Given these features, the primary clinical implication is to avoid mistaking this benign entity for an aggressive malignancy, sparing the patient unnecessary treatment and concern.

## Fibroelastolytic Papulosis: A Case Report

Florencia Anatelli, MD, and Daniel L. Delatorre, MD

*Mid-Atlantic Pathology Services/Aurora Diagnostics, Sterling, Virginia.*

Fibroelastolytic papulosis (FEP) is a rare acquired elastic tissue disorder with only few reported cases in the literature. FEP encompasses a spectrum of clinical and histopathological findings previously described under 2 disorders: Pseudoxanthoma elasticum-like papillary dermal elastolysis and white fibrous papulosis of the neck. It affects elderly women and it is thought to result from intrinsic aging. Clinically, FEP presents with multiple asymptomatic flesh-colored papules with cobblestone appearance on the axillae, neck and trunk. Histologically, it is characterized by absence or decreased elastic fibers in the papillary dermis. It may be accompanied by thickening of the mid dermal collagen fibers. While the clinical findings are striking and resemble PXE, the histologic findings may be subtle, requiring in some instances multiple biopsies to make the diagnosis. Although FEP presents a benign course it may become cosmetically disturbing for the patient. No successful medical therapies have been reported as of today. We report a case of Fibroelastolytic papulosis in a 75-year-old white woman with progressive disease. We discuss the differential diagnosis with other elastic tissue disorders that may present with similar clinical and/or histopathologic findings.

## Degos' Disease Mimicking Lichen Sclerosus: Hyalinized Vessels Provide a Clue to the Diagnosis

Sri Krishna Chaitanya Arudra, Karen Wu, Stephen Moyer, and Jacqueline M. Junkins-Hopkins

*University of Toledo Medical Center, Toledo, OH; Pathology Specialists of New England, Elliot Hospital, Manchester, NH; Elliot Dermatology at River's Edge, Manchester, NH; and Ackerman Academy of Dermatopathology, NY, NY.*

**Background:** Degos' disease or malignant atrophic papulosis (MAP) of skin may present as a life-threatening cutaneo-intestinal syndrome, due to ischemic consequences of vascular insult. Biopsies of classic "porcelain-white" atrophic papules classically are reported to show wedge-shaped necrosis. Evolutionary stages may simulate dermatoses such as tumid lupus erythematosus, dermatomyositis, or lichen sclerosus et atrophicus (LSA). A case of MAP simulating LSA is presented and the histologic spectrum is reviewed.

**Case:** A 49 year old male with headaches, abdominal pain, bloody stools and Barrett's esophagitis presented with a "healing inflammatory lesion" on left flank. Shave biopsy revealed homogenized dermal sclerosis similar to LS. There was perivascular hyalinization. A diagnosis of MAP was made after correlating the histology with a clinical image.

**Conclusion:** Cutaneous lesions of MAP may rarely mimic LSA. Identifying hyalinized vessels and awareness of this histological presentation will assist with diagnosing this potentially life-threatening condition.

### Symmetrical Drug-Related Intertriginous and Flexural Exanthema Secondary to Tramadol Masquerading as Cellulitis

Naiara S. Barbosa, MD, and Alina G. Bridges, DO  
*Mayo Clinic, Rochester, MN.*

Symmetrical drug-related intertriginous and flexural exanthema (SDRIFE) is a subset of systemic contact dermatitis in which patients have classical baboon pattern exanthema secondary to a systemic medication to which no prior cutaneous or cross-sensitization has occurred. A 53-year-old female hospitalized for sudden onset of buttock redness was evaluated for possible cellulitis. She had a history of back pain for which tramadol had been initiated 2 days prior to admission. Examination was remarkable for non-tender symmetrical gluteal erythema with minor induration. Biopsy showed papillary dermal edema and mixed dermal inflammation composed of lymphocytes, histiocytes, eosinophils, and neutrophils. Blood and tissue cultures were negative. Given her clinical presentation, skin histology, and temporal correlation with first tramadol exposure, a diagnosis of SDRIFE was made. Tramadol was discontinued and wet dressings applied with rapid improvement. Although non-specific, histology can be extremely helpful in guiding diagnosis. SDRIFE is most commonly caused by aminopenicillins,  $\beta$ -lactam antibiotics, and chemotherapeutic agents. Cases induced by Tramadol have not been reported. Awareness and recognition of SDRIFE can expedite identification of offending drugs and clinical management.

### Vismodegib in Periorbital Basal Cell Carcinoma: A Case Report

Nicholas Bercovici, DO<sup>1</sup>, Bernardo Mucha, MD<sup>1</sup>, Robert Piorowski, MD<sup>1</sup>, Michael Murphy, MD<sup>2</sup>, and Zende Elaba, MD<sup>1</sup>

<sup>1</sup>Hartford Hospital, Hartford, CT; <sup>2</sup>University of Connecticut, Farmington, CT. Basal cell carcinoma (BCC) remains the most common form of skin cancer. Although readily treated by surgical removal, these tumors occasionally progress to an advanced state that is not amenable to surgery or radiation therapy or, rarely, spread to distant sites. Molecular studies have shown that 90% of BCCs show dysregulation of the hedgehog signaling pathway, resulting in uncontrolled proliferation of basal cells. Vismodegib is a small-molecule inhibitor of smoothed homolog (SMO), a transduction protein involved in the hedgehog pathway. Vismodegib is FDA-approved for BCC which has metastasized, relapsed post-surgery, or cannot be treated with surgery or radiation. We present a case of a 55-year-old male who presented with a large, non-resectable infiltrative BCC involving the right temple-orbital area. Vismodegib therapy resulted in remarkable tumor reduction in the temporal skin. However, periocular infiltration necessitated orbital exenteration. Although involvement of the eye limited treatment effect, this case illustrates Vismodegib as a promising option for patients with surgically non-resectable BCC.

### New-Onset Braf-Positive Cutaneous Reticulohistiocytoma-Like Histiocytosis in a Patient With Langerhans Cell Histiocytosis

Chinmoy Bhate, MD, William T. Johnson, DO, Arthur C. Huen, MD, PhD, Lisa M. Grandinetti, MD, Ronald Jaffe, MD, and Jonhan Ho, MD  
*Department of Dermatology, University of Pittsburgh Medical Center, Pittsburgh, Pennsylvania.*

Non-Langerhans cell histiocytoses have been previously described to occur concomitantly in patients with known Langerhans cell histiocytosis. We describe a 40 year old female patient with cutaneous Langerhans cell histiocytosis involving the scalp, inframammary folds, and inguinal creases who developed new-onset papules and nodules on the upper arm. Histologic examination of these cutaneous nodules demonstrated a dermal histiocytic proliferation of large cells with ground glass eosinophilic cytoplasm, scattered foamy macrophages and multinucleated giant cells, which stained positively for CD68, Factor XIIIa, and S100 but negative for CD1a and Langerin. Lesional cells were reactive against a BRAF immunohistochemical stain for the V600E mutation, which was confirmed by subsequent molecular and genomic testing of the tissue specimen. Identification of this mutation raised the possibility of Erdheim-Chester disease, a diffuse, multiorgan non-Langerhans cell histiocytosis which has recently been linked BRAF V600E mutations and which may have an association with Langerhans cell histiocytoses.

### Angiolymphoid Hyperplasia With Eosinophils Mimicking Primary Cutaneous Follicle Center Lymphoma

Katherine Brick, MD, David Arps, MD, Douglas Fullen, MD, and May Chan, MD

*University of Michigan Department of Pathology.*

A 44-year old woman presented to a surgeon for excision of recurrent lesions on the forehead and cheek. Upon histopathologic review, both specimens demonstrate a dense multinodular lymphoid infiltrate sparing the epidermis. The infiltrate consists of multiple lymphoid follicles with polarized germinal centers and surrounding mantle zones extending into the superficial subcutis. A proliferation of small vessels with plump to epithelioid endothelial cells is present in the upper central portion of the lesion. The lymphoid infiltrate is composed of both CD3-positive T cells and CD20-positive B cells. The CD4:CD8 ratio is 4:1. Germinal centers are positive for Bcl-6 and CD10 but negative for Bcl-2. Ki-67 highlights approximately 80% of the germinal center cells. B-cell immunoglobulin gene rearrangement is negative, arguing against a primary cutaneous follicle center lymphoma (PCFCL). This is the first reported case of angiolymphoid hyperplasia with eosinophilia (ALHE) mimicking PCFCL on the face. A prior report described similar findings on the extremities. Recognition of an unusually robust lymphoid component in ALHE will avoid misdiagnosis of PCFCL and inappropriate treatment.

### Cytokeratin-Negative Proximal-Type Epithelioid Sarcoma Versus Rhabdoid Tumor

Larissa A. Chismar, MD, and Mark Jacobson, MD

*Albert Einstein College of Medicine Department of Pathology, Bronx, NY; and Albert Einstein College of Medicine Department of Pathology, Bronx, NY.*

We present the case of a 13-year-old girl who presented with a changing nodule on the vertex of the scalp. A biopsy showed a proliferation of epithelioid cells with atypical vesicular nuclei in a myxoid stroma. A subset of the cells had prominent round cytoplasmic rhabdoid inclusions. The epithelioid cells were positive for vimentin and focally positive for EMA and CD34. They were negative for cytokeratins (AE1/3, CAM5.2, CK903), S100, CD31, desmin, SMA, and INI-1. FISH studies confirmed the presence of a homozygous deletion of *SMARCB1*. This genetic abnormality may be seen in epithelioid sarcoma and rhabdoid tumors. Distinction between these entities can be extremely difficult. In this case, a diagnosis of epithelioid sarcoma

was favored because of CD34 positivity. The patient was treated with wide local excision and continues to be monitored for recurrence of disease.

### Inherited Epidermolysis Bullosa—A Rare Case Presentation

Dr. P.L. Dhand<sup>1</sup>, Dr. Fuzail Ahmad<sup>2</sup>

Professor and Head, Department of Pathology<sup>1</sup>, Postgraduate student<sup>2</sup>, R.D. Gardi Medical College, Ujjain, Madhya Pradesh, India.

**Introduction:** Epidermolysis bullosa (EB) is a heterogenous group of rare disorder characterised by blistering of skin and mucosa on exposure to mechanical stress. On the level of dermoepidermal separation at the basement membrane, 3 major groups have been defined—simplex, junctional and dystrophic. The mode of inheritance is mostly autosomal dominant in EB simplex and autosomal recessive in junctional EB.

**Objective:** This poster highlights key findings both clinical and diagnostic that have been published in the field of inherited EB within past few years.

**Method:** It includes history, clinical examination, histopathology and immunofluorescence.

**Clinical Features:** A 15 year old patient presented with recurrent, multiple fluid filled lesion, multiple patches over body with hair loss, bleeding gums and swollen gums.

**Microscopy:** H/E stained section shows subepidermal cleavage inspissated with RBCs without acantholysis.

**Result and Conclusion:** The case was diagnosed as inherited EB. The proposed poster should be of value to clinicians and researchers emphasizing both clinical and molecular features of inherited EB and has sufficient flexibility incorporated in its structures to permit further modifications in the future.

### Blastic Plasmacytoid Dendritic Cell Neoplasm, a Rare and Aggressive Malignancy

Nicole Dominiak, MD, Courtney McFaddin, MD, John Maize, Sr, MD, John Metcalf, MD, and Jonathan Ralston, MD

Department of Pathology, Medical University of South Carolina, Charleston, SC.

Blastic plasmacytoid dendritic cell neoplasms (BPDCN) are rare, aggressive hematological malignancies with an extensive predilection for cutaneous involvement. These lesions are known to have aggressive clinical courses, with extremely poor prognoses. We present a case of a blastic plasmacytoid dendritic cell neoplasm from the face of a 74 year old male with a recent history of weight loss and a rapidly growing nodule on his left cheek. The biopsy exhibits a dense dermal infiltrate of atypical mononuclear inflammatory cells, which spare the epidermis. These cells are of medium size with irregular nuclear borders and prominent macronucleoli. Immunohistochemical staining was most consistent with a diagnosis of BPDCN, with strong positivity for CD45, CD56, CD4, BCL-2, and CD123. Uniquely, our lesion also stained positively for CD79a and CD10, usually only positive in a minority of lesions. As these lesions are so rare and since early aggressive treatment is warranted, even in cutaneous limited disease, a delay in diagnosis can be incredibly detrimental to the patient.

### Cutaneous Rosai-Dorfman Disease With Extensive Dermal Fibrosis and Increased Number of IGG4+ Plasma Cells

Olena Dorokhova, and Mark Jacobson

Montefiore Medical Center, Bronx, NY.

Cutaneous Rosai-Dorfman disease (CRDD) is a proliferative disorder of S100+ histiocytes that shares histopathologic features of abundant plasma cells and stromal fibrosis with IgG4-related sclerosing disease (IRSD). We present a case of a 68-year-old female with multiple nodules on her abdomen. Histologically, the lesion was characterized by marked dermal fibrosis as well as scattered large polygonal histiocytes with abundant pale cytoplasm and vesicular nuclei, that were positive for S100 and CD68 and negative for

CD1a. These cells showed emperipolesis, and were intermingled with a florid, mixed inflammatory cell infiltrate containing numerous plasma cells. The infiltrate was primarily located in the subcutaneous tissue. The maximal number of IgG4+ plasma cells was up to 15 per HPF. We present this case to highlight the extensive dermal fibrosis with plasma cells that is a rare finding in CRDD, and reinforce the value of deep skin biopsy in this clinical setting. Although the numbers of IgG4+ plasma cells in our case were lower than those reported in IRSD, their presence and dermal fibrosis suggests that CRDD may be related to IRSD.

### Invasive Gata3-Positive Mucinous Sweat Gland Adenocarcinoma Arising From the Eyelid

Maria D. Estopinal, MD<sup>1</sup>, John Lazarchick, MD<sup>1</sup>, Lee Fucich, MD, and Erin Carlquist, MD<sup>1</sup>

<sup>1</sup>University of South Alabama, Mobile, AL.

**Background:** Mucinous sweat gland adenocarcinoma is a rare primary adnexal neoplasm of the skin. The tumor is most commonly located on the head with predilection for the eyelid. Microscopically there are lobules of neoplastic cells floating in pools of mucin which are immunoreactive for cytokeratins, EMA, CEA, estrogen and progesterone receptors. GATA3 antibody has recently been used for the evaluation of cutaneous carcinoma including adnexal tumors. Extracutaneous primary mucinous adenocarcinoma must be ruled out due to histopathologic similarities.

**Objective:** To report a primary invasive GATA3-positive mucinous sweat gland adenocarcinoma of the eyelid.

**Methods:** An anterior orbital mass involving the right lower eyelid was biopsied from a 74-year-old female. Hematoxylin-eosin and immunoperoxidase stains encompassing CK7, CK20, CDX2, S100 and GATA3 were performed.

**Results:** The histopathologic findings supported the diagnosis of mucinous adenocarcinoma arising from eccrine glands of the eyelid. The neoplastic cells were strongly positive for CK7 and GATA3 antibodies and negative for CK20, CDX2 and S100.

**Conclusions:** To our knowledge this is the first case of a GATA3-positive mucinous sweat gland carcinoma of the eyelid.

### Metaplastic Carcinoma of the Skin With Elements of Squamous Cell Carcinoma, Undifferentiated Sarcoma, and Osteosarcoma

Katherine Fening, MD and Kristopher McKay, MD

Section of Dermatopathology, University of Alabama at Birmingham.

An 80 year-old female presented with a 5-centimeter subcutaneous nodule on the back. A partial sampling examined at a referring facility reportedly demonstrated malignant fibrous histiocytoma. However, on complete evaluation of the re-excision specimen, elements of squamous cell carcinoma and undifferentiated pleomorphic sarcoma were noted. There were also foci of malignant osteoid. A diagnosis of metaplastic carcinoma of the skin (carcinosarcoma) was made. This entity is rare and usually occurs on the head of elderly patients, although other sites of involvement have been reported. There is intimate association of distinct epithelial and mesenchymal components, both of which are typically frankly malignant. The epithelial component may consist of basal cell carcinoma, squamous cell carcinoma, or an adnexal neoplasm. The mesenchymal component typically consists of undifferentiated sarcoma, chondrosarcoma, fibrosarcoma or osteogenic sarcoma. This case highlights the inherent pitfalls of partial sampling and contributes to the relatively limited number of reported cases.

### Oliver Adams Syndrome. Case Report of Oliver Adams Syndrome in Dominican Republic (DR)

Gerardo Flores Solaegui<sup>1</sup>, Loryart Mars Grullón<sup>1</sup>, Raisa Acosta<sup>2</sup>, and Silvia Marte<sup>3</sup>

<sup>1</sup>Dermatology, <sup>2</sup>Dermatology/Dermatopathology; Instituto Dermatológico Dominicano y Cirugía de piel Dr. Huberto Bogaert Díaz, Santo Domingo,

*República Dominicana; <sup>3</sup>Pediatric Dermatology, Robert Reid Hospital, Santo Domingo, República Dominicana.*

Adams Oliver Syndrome (OSA) was first described in 1945 by Adams and Oliver and characterized by the presence of aplasia cutis defects at the vertex and transverse extremities, with variation of expression. Today its prevalence is unknown. Vascular abnormalities such as congenital telangiectasia cutis marmorata, pulmonary hypertension, portal hypertension, and retinal hypervascularization. Other characteristics may occur such as psychomotor retardation and congenital heart defects. The management of this entity requires a multidisciplinary approach to improve the quality of life, treat complications, and prevent damage. Patient is currently closely monitored by several specialists and presenting improvement of injury.

### Traumatic Neuromas of the Penis

Desiree A. Godar<sup>1</sup>, Valerie Laniosz<sup>1</sup>, Alina G. Bridges<sup>1</sup>, and Andre M. Oliveira<sup>2</sup>

<sup>1</sup>Mayo Clinic Dept of Dermatology; and <sup>2</sup>Mayo Clinic Dept of Pathology, Rochester, MN.

Traumatic penile neuromas are benign nerve sheath tumors resulting from regeneration of nerves that have been damaged by trauma. They present as firm skin colored papules that may be painful or asymptomatic. A 36-year-old gentleman presented with a 2-month history of asymptomatic penile lesions. No preceding trauma was identified. Prior treatment included oral metronidazole and fluconazole as well as topical steroids and Aldara with no improvement. Examination revealed multiple 1 mm skin-colored papules on the dorsal and ventral penis. A biopsy revealed an increase proliferation of dermal nerve bundles and Meissner corpuscle-like structures embedded in a fibrous stroma in the papillary dermis. S100 staining highlighted the nerve fibers and Meissner corpuscle-like structures. These findings were consistent with the diagnosis of traumatic penile neuromas. Traumatic penile neuromas are rare. Various forms of inciting trauma have been described in the literature including vigorous sexual activity, biopsy and circumcision. It is important to consider a biopsy in penile lesions, especially those that are unresponsive to treatment, as traumatic neuromas can be mistaken for condyloma acuminata clinically and treatment for condylomata may further promote development of new lesions. Adams Oliver Syndrome (OSA) was first described in 1945 by Adams and Oliver and characterized by the presence of aplasia cutis defects at the vertex and transverse extremities, with variation of expression. Today its prevalence is unknown. Vascular abnormalities such as congenital telangiectasia cutis marmorata, pulmonary hypertension, portal hypertension, and retinal hypervascularization. Other characteristics may occur such as psychomotor retardation and congenital heart defects. The management of this entity requires a multidisciplinary approach to improve the quality of life, treat complications, and prevent damage. Patient is currently closely monitored by several specialists and presenting improvement of injury.

### An Atypical Presentation of Galli Galli Disease

Tania M. Gonzalez Santiago, MD, Lawrence E. Gibson, MD, and Lisa Drage, MD

Mayo Clinic, Department of Dermatology, Rochester, Minnesota.

Galli-Galli disease (GGD) is a rare variant of the genodermatosis Dowling-Degos disease (DDD) with the histologic finding of acantholysis. The pathogenesis has long been unknown, but a recent report described a 2 base-pair deletion, resulting in a frameshift mutation in the KRT5 gene in patients with DDD. GGD is characterized by reticulated hyperpigmentation predominantly affecting the flexures along with pruritic, erythematous, scaly papules in the same distribution as well as on the trunk and proximal extremities. The differential diagnosis of GGD includes classic DDD, transient acantholytic dermatosis, Darier's disease, and epidermolysis bullosa with mottled pigmentation. Galli-Galli disease is best considered a subtype of Dowling-Degos disease with clinical and histologic features similar to transient

acantholytic dermatosis. We report 2 patients with GGD exhibiting erythematous scaly plaques and lentigo-like macules on the lower extremities, rather than the characteristic reticulate hyperpigmented macules of large body folds. In our cases, the history along with the histopathologic features supports a diagnosis of GGD. A high index of suspicion is needed to diagnose GGD that lacks the characteristic reticulate hyperpigmentation of large body folds.

### Eruptive Keratoacanthoma En Plaque

Jennette Gruchy<sup>1</sup>, David J. Leffell<sup>2</sup>, and Rossitza Lazova<sup>2,3</sup>

<sup>1</sup>Department of Pathology, Dalhousie University, Halifax, Nova Scotia, Canada; <sup>2</sup>Department of Dermatology, and <sup>3</sup>Department of Dermatology and <sup>3</sup>Pathology, Yale University School of Medicine, New Haven, CT.

Several variants of keratoacanthoma have been described since its first inception in 1889. Generally this entity has a benign clinical course and often demonstrates spontaneous regression. Rarely, histologic features include perineural and lymphovascular invasion, particularly in cases of the head and neck area. However, limited reports suggest these do not appear to have an adverse impact on prognosis. We present a 46-year old woman who initially developed a solitary 7-mm keratotic papule on the right nasolabial fold, diagnosed histologically as keratoacanthoma. Subsequently, in the same area, a 3-cm erythematous plaque studded with multiple milia-like 1-2 mm papules was formed. Biopsies of 2 such papules were diagnosed histologically as keratoacanthomas with perineural invasion. A re-excision of the entire plaque showed multiple keratoacanthomas with perineural and intravascular invasion. Review of the literature revealed a similar case reported by Washington and Mikhail as "eruptive keratoacanthoma en plaque." Their patient developed multiple keratoacanthomas on the right cheek as a scaly erythematous plaque studded with small cysts in the setting of immune suppression. Our patient, despite intravascular invasion extending focally to a surgical margin, has not developed any recurrences or sinister complications 8 years after the re-excision. This supports the notion that keratoacanthoma with perineural and lymphovascular invasion does not necessarily predict a more ominous course. In view of this, aggressive treatments, particularly on the face, should be given careful consideration.

### Primary Cutaneous CD4+ Pleomorphic Small/Medium-Sized T-Cell Lymphoma Arising Within an Elastolytic Giant Cell Granuloma

Lauren Guren, MD, and Anthony Fernandez, MD

Cleveland Clinic Department of Dermatology.

An 80-year-old man presented with a 1-year history of an enlarging nodule on his back. He had a biopsy 6 months prior, consistent with an elastolytic giant cell granuloma. The nodule increased in size. On his back was a red, firm, annular plaque and at the inferior portion was a 1.5 cm smooth, red nodule. A punch biopsy was taken revealing a normal epidermis while the superficial, mid and deep dermis is largely displaced by an atypical lymphoid infiltrate composed of small to medium-sized lymphocytes. Immunohistochemical stains showed that the atypical cells express CD3, CD4, CD5, CD43, betaF1 with a loss of CD7. Molecular studies for T-cell gene rearrangement were performed and demonstrate a clonal T-cell population. Gene rearrangement was also performed on the previous biopsy of the elastolytic granuloma which showed a non-clonal cell population. These findings are consistent with a primary cutaneous CD4+ pleomorphic small/medium-sized T-cell lymphoma. For treatment, the patient was referred to radiation-oncology where he received external beam radiation treatments, leading to resolution of the nodule and annular plaque. This case highlights a unique presentation of a rare T-cell lymphoma which based on our literature search, a similar presentation has been described in only 1 additional case.

### Reactive Angioendotheliomatosis Presenting as Cellulitis in a Patient With Metastatic Melanoma

Hatem Hassanein H, MS<sup>1</sup> and Jane Messina, MD<sup>2</sup>

<sup>1</sup>University of South Florida, Morsani College of Medicine, Tampa, FL; and  
<sup>2</sup>Senior Member, Moffitt Cancer Center Departments of Anatomic Pathology and Cutaneous Oncology

A 47-year-old-male diagnosed with Stage IIIC melanoma of unknown primary involving the left axilla presented with an enlarging 13 × 8 cm left axillary nodule and a 15 cm area of erythematous induration of the overlying skin unresponsive to topical, oral and IV antibiotics. Excisional biopsy of the 5 cm nodule revealed metastatic melanoma, positive for HMB-45, Melan-A, pan melanoma, MITF and S-100. In the overlying skin, a dermal proliferation of small capillary-like spaces filled the reticular dermis. The spaces were lined by cuboidal to focally hobnail endothelial cells, surrounded by pericytes and lymphocytes. CD31 highlighted these vascular spaces and actin was focally positive in the walls of larger vessels. A diagnosis of reactive angioendotheliomatosis (RAE) was made. RAE is a reactive vascular proliferation associated with underlying vascular obstruction from a variety of conditions, most commonly hematolymphoid neoplasms. RAE demonstrates various clinical presentations as well, including cellulitis. This is the first report of RAE presenting in the setting of metastatic melanoma.

### An Annular Presentation of Primary Cutaneous Follicle Center Lymphoma

Hinds BR, McCalmont TH, and Pincus LB

*Departments of Pathology and Dermatology, University of California, San Francisco.*

Unusual clinical morphologies have been reported in primary cutaneous follicle center lymphoma (PCFCL), namely miliary or acneiform papules and gyrate or figurate plaques with a proclivity for the head/neck and trunk, respectively. A 60-year-old woman presented with an eruption involving the right zygomatic cheek for 6 weeks. Physical examination revealed multiple 2–3 mm erythematous papules coalescing into an arcuate plaque with a palpable rope-like border. A punch biopsy revealed a dermal lymphocytic infiltrate comprised mostly of small to medium-sized epithelioid and spindled lymphocytes. Immunophenotyping showed that most lymphocytes expressed CD20. Additionally, a bcl-6 stain exhibited a diffuse pattern, while a CD21 stain did not reveal follicular dendritic cell networks. A bcl-2 stain only labeled T-cells. Based on this staining, a diagnosis of follicle center lymphoma was rendered. Clinical workup was negative for systemic disease. Herein, we present a new annular clinical guise of PCFCL, which simulates granuloma annulare. We alert dermatologists and dermatopathologists alike to another potential pitfall in the clinical diagnosis of PCFCL. Recognition will avoid untoward diagnostic and treatment delays.

### Intravascular Large B-Cell Lymphoma: A Rare Systemic Disease Diagnosed by Skin Biopsy

Omer Ibrahim, MD, Anthony Fernandez, MD, PhD, and Wilma Bergfeld, MD  
*Department of Dermatology, Cleveland Clinic Foundation, Cleveland, OH.*

#### Case Report:

**Purpose:** To highlight the gross and microscopic findings of a very rare and potentially devastating condition, and the utility of skin biopsy in establishing diagnosis of a disease characterized by extremely nonspecific symptoms and varying presentations. A 74-year-old Caucasian man with 6-month history of pancytopenia and panhypopituitarism was admitted to the hospital for evaluation of a 1-week history of a purpuric eruption, “bruises” on his chest, and concurrent hematuria. Examination revealed numerous violaceous papules coalescing into purple plaques over the anterior chest, bilateral anterior shoulders, and upper abdomen. All the papules and plaques were completely blanchable on diascopy. Pathologic examination of a representative lesion revealed dilated superficial and deep dermal blood vessels stuffed with large anaplastic lymphocytes with immunoreactivity for CD-20. Laboratory review revealed pancytopenia. Imaging of the brain revealed an engorged pituitary gland. Bone marrow biopsies were normal. The patient was diagnosed with Asian-variant intravascular large B-cell lymphoma (IVLBCL) with

cutaneous, urologic, and pituitary involvement and accompanying hemophagocytic syndrome. Treatment with systemic anti-CD20 chemotherapy was initiated; however the patient expired a few days later due to hypotension and cardiopulmonary collapse.

### Eosinophilic Pustular Folliculitis (Ofuji’s Disease) in an Elderly Filipino Woman

Mahwish Irfan, MD, Joan Tamburro, DO, and Steven D. Billings, MD  
*Cleveland Clinic, Cleveland, Ohio.*

A 69-year-old Filipino woman presented with a 5-year history of pruritic papules of the face and chest, previously diagnosed as chronic actinic dermatitis. She denied systemic symptoms or new medications. She noted mild improvement with prednisone, dapson, and methotrexate, but continued to have flares. Examination revealed an edematous facial appearance with multiple red-brown scaly papules, pustules, and plaques on the face and chest. Her scalp had depigmented macules with perifollicular sparing of pigment. Labs revealed eosinophilia. Punch biopsy showed mild spongiosis with intraepidermal eosinophilic pustules and a rich dermal folliculocentric eosinophilic infiltrate with normal mucin, consistent with eosinophilic pustular folliculitis. This rare skin disease is characterized by recurrent crops of pruritic papules and pustules on the face and trunk. Lesions usually resolve spontaneously with hyperpigmentation and scaling. Peripheral eosinophilia is common and patients have no systemic symptoms. EPF responds to indomethacin and symptomatic treatment of pruritus but many patients are misdiagnosed and treated with systemic immunosuppressants. Thus, early recognition and biopsy are essential and may spare affected patients side effects associated with unnecessary systemic medications.

### Granulomatosis With Polyangiitis Presenting With Cutaneous Ulcers

Mahwish Irfan, MD, Pooja Khera, MD, and Melissa Piliang, MD  
*Cleveland Clinic, Cleveland, Ohio.*

A 64-year-old man presented with a 6-month history of hemoptysis, cough, and painful cutaneous ulcers. ROS revealed fever, epistaxis, and an 80-pound weight loss. He had a 35-pack-year smoking history raising concern for lung malignancy. Examination revealed an ill-appearing man with deep, punched-out ulcers on the mid-back and neck. A chest-x-ray showed a cavitary mass. A punch biopsy of an ulcer showed leukocytoclastic vasculitis with granulomatous inflammation. Biopsy of the lung mass revealed acute and necrotizing granulomatous inflammation. Additional work-up revealed a positive C-ANCA antibody and chronic sinusitis on CT scan, establishing the diagnosis of granulomatosis with polyangiitis (GPA), formerly known as Wegener’s granulomatosis. GPA is a life-threatening small vessel vasculitis. The clinical triad includes systemic necrotizing vasculitis, necrotizing granulomatous inflammation of the respiratory tract, and necrotizing glomerulonephritis. Cutaneous signs such as ulcers, nodules, digital infarcts, and oral ulcers may be the initial presenting signs. Biopsy of these lesions can help clinch the diagnosis, especially in cases with deteriorating clinical course and extensive differential diagnosis. Our patient was treated with a systemic steroids and rituximab with rapid and marked improvement.

### Granulomatosis With Polyangiitis Mimicking Classic Inflammatory Bowel Disease-Associated Pyoderma Gangrenosum

Scott A. Kindle, MD, Michael J. Camilleri, MD, Lawrence E. Gibson, MD, and Mark D.P. Davis, MD

*Department of Dermatology, Mayo Clinic, Rochester, Minnesota.*

Granulomatosis with polyangiitis (GPA) affects multiple organs including the skin. A 26-year-old female with a history of inflammatory bowel disease

(IBD) treated with adalimumab presented with leg ulcerations of pyoderma gangrenosum. The ulcerations initially improved with infliximab. She presented 7 months later with worsening leg ulcers, a new ulcer in the right conchal bowl, and nasal septal perforation. Anti-neutrophil cytoplasmic antibody (ANCA) testing was positive. Biopsy of the nasal septum showed vasculitis and biopsy of the leg ulcer showed granulomatous inflammation, both consistent with a diagnosis of GPA. On review of her bowel biopsies, vasculitis was noted on 1 specimen, but her diagnosis remains Crohn's disease. Treatment with rituximab and prednisone was initiated. Within 2 months the ulcers were almost healed. This presentation illustrates that GPA can present with findings identical to IBD-associated pyoderma gangrenosum. Lack of response to infliximab plus the nasal septal and auricular lesions led to ANCA testing and the diagnosis. The development of GPA after adalimumab also raises the possibility of adalimumab-induced GPA.

### A Case of Iododerma Secondary to Amiodarone Therapy

Ivanka Kovalyshyn, DO, Shilpi Khetarpal, MD, and Anthony Fernandez, MD, PhD

*Departments of Dermatology and Pathology, Cleveland Clinic Foundation, Cleveland, OH.*

We report a case of iododerma secondary to amiodarone therapy in a 72-year-old patient with a history of atrial fibrillation, type 2 diabetes, chronic kidney disease, hypertension and congestive heart failure. Patient presented with a 2- month history of pruritic skin eruption on upper extremities. Physical examination revealed numerous red to violaceous scaly papules and pustules symmetrically distributed on both forearms. Laboratory tests revealed a serum creatinine of 3.79 mg/dL (normal range 0.70 to 1.40 mg/dL) and serum iodine level of 42,305 ng/mL (normal range 40 to 92 ng/mL). A punch biopsy of the forearm papule revealed pseudoepitheliomatous hyperplasia, spongiosis with vesiculation, intraepidermal neutrophilic microabscesses, focal subepidermal bulla formation and mixed dermal infiltrate. PAS and GMS stains were negative for infectious organisms. Given the histologic features and negative stains, as well as our patient's markedly elevated serum iodine level, the diagnosis of iododerma secondary to amiodarone therapy was made. Amiodarone was discontinued and within 2 weeks, patient noted significant improvement of his skin lesions. In conclusion, patients with renal insufficiency and exposure to amiodarone can rarely present with iododerma. It is important to recognize this rare presentation to ensure accurate diagnosis and correct management.

### Case Presentation: Progressive Symmetrical Erythrokeratoderma

Charlotte LaSenna, Mariya Miteva, and Paolo Romanelli

*Department of Dermatology and Cutaneous Surgery, University of Miami Miller School of Medicine, Miami, FL.*

**Case:** Seventeen-year-old female history of recurrent, multiple, erythematous, scaly lesions affecting arms and legs since birth. Physical exam showed widespread, symmetrically distributed, erythematous, annularly-circumscribed hyperkeratotic plaques over the extensor aspects of extremities and mild palmar keratoderma.

**Pathology:** H&E of lesional skin showed papillated epidermal acanthosis with hypergranulosis, parakeratosis, and compact orthokeratosis. The granular layer showed single vacuolated keratinocytes and intranuclear granules in the upper layer of cells.

**Diagnosis:** Progressive Symmetrical Erythrokeratoderma (PSEK).

**Course:** Oral isotretinoin and topical calcipotriene led to clearance of lesions, however recurrence occurred upon cessation of therapy.

**Discussion:** Erythrokeratodermias are a heterogenous group of inherited disorders. In PSEK, genetic analyses and in vivo studies on transgenic mice have demonstrated a frameshift mutation of the loricrin gene causing delayed termination of transcription. The histopathology of PSEK is nonspecific. Pathogenically, the N-terminal domains of profilaggrin are aggregated with

mutant loricrin in condensed nuclei, which are viewed in the cornified layer as parakeratosis.

### A Rare Case of Squamous Cell Carcinoma With Melanoma In Situ

Michael C. Royer, MD\*, Mark S. Lincoln, MD\*, Luke S. Chung, MD†, and George P. Lupton, MD†

*\*Department of Pathology and Laboratory Services, Walter Reed National Military Medical Center, Bethesda, MD; and †Department of Dermatopathology, Joint Pathology Center, Silver Spring, MD.*

Neither melanoma in-situ nor squamous cell carcinoma is particularly uncommon lesions, especially in the older population. However, the combination of the 2 is distinctly unusual. We herein describe such a case. The patient, a 72 year old male, presented with a hyperpigmented macule on the right preauricular area, which appeared to be consistent with lentigo maligna melanoma. Histologic examination showed a superficial squamous cell carcinoma with marked cytologic atypia and atypical mitoses. The lesion contained a confluent proliferation of atypical pigmented cells along the dermal-epidermal junction. Similar cells were noted in the non-neoplastic epidermis, with extension into adnexae. The cells were highlighted with melanocytic immunohistochemical stains, confirming the diagnosis of melanoma in-situ. AE1/AE3 highlighted the squamous proliferation. The histologic features are those of a superficial squamous cell carcinoma with melanoma in situ and represent a rare presentation of coexistent tumors. Similar collision tumors have previously been described in the literature.

### Isolated Cutaneous Rosai Dorfman of the Scalp

Nicholas Logemann<sup>1</sup>, Damon McClain<sup>2</sup>, and Michael Royer<sup>1</sup>

*<sup>1</sup>Walter Reed National Military Medical Center, Bethesda, MD; and <sup>2</sup>Naval Hospital Camp Lejeune, Camp Lejeune, NC.*

A 22 year old male presented with a several month history of a non-healing erythematous papule on the scalp irritated by close haircuts. A biopsy was performed which revealed a dermal histiocytic proliferation of large cells with abundant pale cytoplasm of varying morphologies, including xanthomatous, reticulohistiocytic and multinucleate types. Within the cytoplasm of scattered histiocytes were intact lymphocytes, or emperipolesis. Immunohistochemistry was positive for S100 and negative for CD1a. A systemic workup was then performed which failed to reveal any evidence of systemic disease. The patient remains in good health. Cutaneous Rosai Dorfman is a rare histiocytic proliferation occurring more commonly in young adults. The prognosis is favorable especially if complete excision can be performed. The clinical presentation can be varied, though some sources site a predilection for the face. Our patient adds to the few cases in the literature of this rare entity. It also highlights the protean manifestations of cutaneous Rosai-Dorfman as it clinically mimicked a benign cyst or keloid with no suspicion for a lymphoproliferative disorder.

### Atypical Presentation of Sezary Syndrome With CD4+/CD7+/CD26-T Cells and Marked Epidermotropism

Gabriela A. Maloney, and Marylee Branieceki, MD

*Midwestern University, Downers Grove, IL; Advocate Lutheran General Hospital, Park Ridge IL.*

Sezary Syndrome (SS) is a rare leukemic variant of cutaneous T-cell lymphomas characterized by cerebriform cells (Sezary cells), erythroderma, pruritus and circulating neoplastic T-Cells. We present a case of a 46 year old female with tenderness, pruritus, hyperpigmented lesions and lymphadenopathy. She later developed severely pruritic cutaneous lesions in both breasts that started as bruises and progressed to erythroderma covering 70% of her body. Skin biopsies of the breast were performed and revealed atypical small to medium sized epidermotropic CD4+/CD7+ T-cell lymphocytes with

Pautrier microabscesses. Peripheral blood flow cytometry revealed atypical circulating Sezary type T-cell lymphocytes but the Sezary cell count was not  $>1000/\mu\text{l}$ . A clonal T-cell rearrangement was detected and the CD4:CD8 ratio was approximately 15:1. The neoplastic T-cells did not express CD56 or CD26. Axillary nodal tissue also showed lymphoid neoplastic involvement with a similar immunoprofile. This case of Sezary Syndrome is atypical due to the retention of T cell CD7 expression, significant atypical lymphocytic epidermotropism that histologically mimicked mycosis fungoides and circulating Sezary cells (CD4+/CD7+/CD26-) of less than  $1000/\mu\text{l}$ . It is important to recognize this unique presentation and be able to differentiate it from MF as this can significantly alter the prognosis and treatment while having an impact on patient's life.

### Infantile Digital Fibroma: A Rare and Interesting Benign Soft Tissue Tumor

Etan Marks<sup>1</sup>, Evan Himchak<sup>1</sup>, Alexa Karkenny<sup>2</sup>, Jacob Schulz<sup>2</sup>, and Esperanza Villanueva-Siles<sup>1</sup>

Departments of Pathology<sup>1</sup> and Orthopedic Surgery<sup>2</sup> at Montefiore Medical Center, Bronx, NY.

An 8 month old male being followed at Montefiore Medical Center was brought in by his mother for a mass of the left second toenail bed that was noted to appear at about 2–3 months of age and had progressively enlarged. At the time of surgery the lesion was a 0.8 cm in greatest dimension non-mobile nodular growth, firm, with no signs of infection. The mass was overhanging the toenail, but the nail was uninvolved. When his mother tried to scrape the lesion, it bled easily and was not associated with any pain. The patient was taken to surgery and the lesion was removed. It revealed a dermal proliferation of hypo-cellular fascicles of fibroblasts and myofibroblasts with eosinophilic cytoplasmic inclusions (best seen on a trichrome stain). This led to the diagnosis of the very rare entity of Infantile Digital Fibroma. We present this case to highlight the importance of recognizing this entity, which has a propensity for recurrence, versus a malignant condition, such as infantile fibrosarcoma.

### Foreign Body Reaction to 3 Injectable Fillers

Jose E. Ollague, MD, and Mariya Miteva, MD

University of Miami, Department of Dermatology, Florida.

**Introduction:** A 62 year old white female patient presented with multiple indurated, small to medium subcutaneous nodules along the nasolabial fold, cheeks and chin, which became apparent in the last 3 months. No systemic symptoms associated. Significant personal history of multiple dermatological cosmetic procedures performed in the past. A 3 mm punch biopsy was performed, and the sample was stained with routine H&E.

**Results:** The histology revealed several fragments containing dermis, subcutaneous tissue and muscle. The entire dermis is replaced by dense florid granulomatous infiltrate of histiocytes and numerous giant cells surrounding rounded vacuoles of similar size (mimicking normal adipocytes). Interspersed sclerotic collagen fibers are appreciated. Focally, a similar granulomatous process encompasses an extracellular basophilic, amorphous material as well. At the subcutaneous level, multiple, vacuolar cystic spaces of different sizes (Swiss cheese pattern) are surrounded by foamy histiocytes.

**Conclusion:** These findings are consistent with a foreign body granulomatous reaction due to injectable polymethylmetacrylate, hyaluronic acid and silicone. To our knowledge, this is the first case of a foreign body reaction to 3 different injectable fillers.

### Cutaneous Presentation of IVLBCL: A Case Report

Rebecca Millius, MD, and Fangru Lian, MD

University of Arizona, Tucson, AZ.

Intravascular large B-cell lymphoma (IVLBCL) is an extranodal lymphoma defined by the growth of neoplastic lymphoid cells within the lumen of blood

vessels. Isolated cutaneous involvement is unusual and may represent a distinct subtype of the lymphoma. This disease carries an overall poor prognosis even with chemotherapy. The patient is a 37 year old woman who developed tender subcutaneous nodules in her bilateral lower extremities in 2007. Initial biopsies were not diagnostic. Her symptoms waned after 12–18 months. The nodules enlarged rapidly in 2013 and were associated with B symptoms. Excisional skin biopsy revealed cutaneous IVLBCL. She has had varying response to chemotherapy. Awareness of the cutaneous form of IVLBCL is important, as this entity can be limited to the skin vasculature. Biopsies should be full thickness from within the involved areas in order to maximize diagnostic material.

### References

Orwat DE, Batalis NI. Intravascular large B-cell lymphoma. *Arch Pathol Lab Med.* 2012;136:333–338.

Swerdlow SH. *WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues.* 4th ed. Geneva, Switzerland: WHO Press; 2008.

### An Unusual Candida Infection in an Immunocompromised Patient

Annie O. Morrison, MD, Ifeoma U. Nwadei, MD, Gina E. Johnson, MD, Matthew C. Fink, MD, Livia Van, MD, and Douglas C. Parker, MD, DDS  
Emory University, Atlanta Georgia.

A 63 year-old male presented with a new 2 cm, rubbery, brown, skin nodule on the posterolateral thigh with surrounding purpura. The patient had recently been diagnosed with acute myelogenous leukemia (AML). He had completed induction chemotherapy that included cytarabine, and was neutropenic and thrombocytopenic with persistent candidemia despite aggressive pharmacotherapy including micafungin. The differential diagnosis included septic emboli, leukemia cutis, vasculitis, trauma, drug eruption, and neutrophilic eccrine hidradenitis, which can be associated with cytarabine. Punch biopsy revealed a hyperkeratotic, acanthotic epidermis with mild, predominately lymphocytic perivascular inflammatory infiltrate. The subcutis showed numerous nodular aggregates of fungal yeasts. A fungal culture grew *Candida krusei*, confirming the diagnosis. *Candida krusei* is an opportunistic infection found in immunocompromised patients, particularly those with neutropenia. *C. krusei* is resistant to fluconazole, and has shown decreased susceptibility to other commonly used antifungals. Cutaneous manifestations can be the source of primary infection, but more commonly indicates an underlying systemic infection, as was the case in the above patient. Definitive diagnosis of cutaneous *C. krusei* requires microscopic tissue examination along with tissue culture with appropriate susceptibility testing.

### Bilateral Axillary Masses After Laser Photoepilation Therapy: A Unique Complication

Ifeoma U. Nwadei, MD<sup>1</sup>, Gina E. Johnson, MD<sup>1</sup>, and Krisztina Hanley, MD<sup>1</sup>  
<sup>1</sup>Emory University Department of Pathology, Atlanta, GA.

Techniques for unwanted hair removal have advanced over the past 8–10 years. The Alexandrite laser (755 nm) is considered by many cosmetic dermatology experts to be the gold standard in laser photoepilation. Complications from laser hair removal depend on skin type, body site and recent exposure to sun. They more frequently occur on sun-exposed skin and tend to be minor and temporary, including erythema and perifollicular edema. More severe effects include burns, blisters, hyper- or hypopigmentation or scarring. However, less is known of the histologic effects of laser therapy on skin and underlying soft tissue. A 39-year-old African-American female with polycystic ovarian syndrome and hirsutism presented with firm bilateral axillary masses after undergoing axillary laser photoepilation. An ultrasound evaluation demonstrated irregular solid hyperechoic masses with internal echoes in each axilla measuring 14 mm and 19 mm respectively. The breasts were symmetric, without skin or nipple abnormalities. Screening mammography showed no evidence of primary breast lesions. A core biopsy of the

right axilla revealed extensive subcutaneous fat necrosis with dermal fibrosis, and chronic inflammation. The patient was managed conservatively and continues to be asymptomatic 6 months later. This case describes a unique clinical and histologic complication of laser hair removal.

### Composite Lymphoma: Concurrent Cutaneous B-Cell Small Lymphocytic Lymphoma and Peripheral T-Cell Lymphoma in a Patient With Sezary Syndrome

WINNER OF THE BEST POSTER

Oluwakemi Onajin, MD, Tania Gonzalez Santiago, MD, and Alina G. Bridges, DO

Department of Dermatology, Mayo Clinic College of Medicine, Rochester, Minnesota.

Composite lymphoma (CL) is a rare entity defined by the co-occurrence of 2-or-more morphologically and immunophenotypically distinct lymphoma types. Our patient is a 91-year-old female who presented with erythroderma, lymphadenopathy, and a liver mass. Skin biopsy showed an atypical epidermotropic T-cell infiltrate with a positive clonal T-cell gene rearrangement consistent with cutaneous T-cell lymphoma. She was diagnosed with Sezary syndrome (SS). Liver biopsy demonstrated a CD5+ B-cell population with a dense T-cell infiltrate. She developed erythematous nodules on the occipital scalp and back. Skin biopsy revealed a diffuse dermal infiltrate composed of atypical T-cells and B-cells identical to that seen in the liver. Clonal T-cell and immunoglobulin receptor gene rearrangements were positive. Flow cytometry of the peripheral blood demonstrated CD5+, kappa light chain-restricted B-cells and CD4+ T-cells. She had concurrent cutaneous B-cell small lymphocytic lymphoma and peripheral T-cell lymphoma in addition to Sezary syndrome. To our knowledge, the simultaneous occurrence of these lymphomas in the skin has never been reported.

### Rickettsia Parkeri Rickettsiosis Presenting With a Papulovesicular Rash and Myopericarditis

Vishwas Parekh<sup>1</sup>, C. Blake Phillips<sup>2</sup>, Martin Rodriguez<sup>3</sup>, Gilbert J. Perry<sup>4</sup>, Kristopher McKay<sup>1,2</sup>, and Kathleen M. Beckum<sup>1,2</sup>

Departments of Pathology<sup>1</sup>, Dermatology<sup>2</sup>, Infectious Diseases<sup>3</sup>, and Cardiology<sup>4</sup>, University of Alabama at Birmingham, AL.

*Rickettsia parkeri* rickettsiosis (RPR), first reported in humans in 2004, is a rarely encountered spotted fever transmitted by the Gulf Coast tick (*Amblyomma maculatum*). RPR typically presents with fever, arthralgias, myalgias and cutaneous lesions of inoculation eschars and rash. Here we report a case with an unusual presentation of skin rash and myopericarditis. A 17-year-old Caucasian male presented with a 10-day history of fever, arthralgia, pleuritic-type chest pains and papulovesicular rash involving the left shin, foot and dorsal hand. A recent exposure to tick bites was noted. Laboratory results were consistent with myopericarditis. Skin biopsy demonstrated a superficial and deep dermal dense lymphocytic infiltrate with focal fibrinoid necrosis of the vessel walls, prominent interface changes with necrotic keratinocytes and focal epidermal necrosis. Rickettsiosis focused immunohistochemistry was reactive for spotted fever group *Rickettsia* species. Real-time PCR demonstrated *Rickettsia parkeri* in the skin lesion. The patient was afebrile within 3 days of doxycycline treatment and discharged on a 10-day course.

### Hyperkeratotic Variant of Porokeratosis in a Patient With HIV and Hepatitis C Virus Infections and a Therapy-Related Immunosuppressed State

Vishwas Parekh, MD<sup>1</sup>, Filamer D. Kabigting, MD<sup>2</sup>, and Jacqueline M. Junkins-Hopkins<sup>2</sup>

<sup>1</sup>Department of Pathology, University of Alabama at Birmingham, Birmingham, AL, and <sup>2</sup>Ackerman Academy of Dermatopathology, New York, NY.

Porokeratoses is a group of acquired and hereditary disorders of keratinization with a very characteristic clinical and histopathologic appearance. Typical lesions present as well-circumscribed slightly atrophic macules or patches with distinctive peripheral keratotic ridge, called cornoid lamella. Although a precise mechanistic explanation is lacking, ultraviolet radiation and immunosuppressed states are considered causally-associated with most cases of acquired porokeratosis. Hepatitis C virus (HCV) infection has been proposed as a link between the immunosuppressed states and development of acquired porokeratosis. Among the various recognized clinical entities that constitute this group, rare cases of hyperkeratotic variants have been described that may pose diagnostic challenge. Here we describe a remarkable case of hyperkeratotic variant of porokeratosis that occurred in a patient with known HIV and HCV infections and a coexisting therapy-related immunosuppressed state. In the light of previous literature, we postulate that the HCV infection may be a link between the immunosuppressed state and development of porokeratosis.

### CD56 and Granzyme B-Positive Mycosis Fungoides. Report of a Case

Mauricio Postigo<sup>1</sup>, Monica Ruiz<sup>1</sup>, Ferdinand deAmat<sup>2</sup>, and Jose Plaza<sup>3</sup>

<sup>1</sup>Anatomic Pathology and <sup>2</sup>Dermatology Services, Hospital Carlos Seguin, Arequipa, Peru; and <sup>3</sup>Department of Pathology, Division of Dermatopathology, Medical College of Wisconsin, Milwaukee.

Mycosis fungoides (MF) is the most common form of cutaneous lymphoma, follows an indolent course for many years and has a T-helper phenotype. We report the case of a 32 year old man with a 3 year history of erythematous and scaly papules and plaques on the face and extremities. Besides these, he only presented enlarged inguinal lymph nodes. On histopathology, in the skin, the infiltrate consisted of CD4, CD56 and granzyme B -positive, CD8-negative epidermotropic, perivascular and periadnexal T-cell infiltrate. The lymph node showed dermatopathic changes, without the phenotype described in the skin. There are MF CD8/CD56-positive and CD4/CD8-negative CD56+ described variants, and just only 1 report of a 85 year old CD4+, CD56+, and TIA-1+. Although this patient has an immunophenotype consistent with more aggressive forms of lymphomas, the clinical and histological features are characteristic of the indolent MF.

### Extranodal Rosai Dorfman Disease. Report of 8 Cases From Arequipa, Peru

Mauricio Postigo<sup>1</sup>, Monica Ruiz<sup>1</sup>, Ferdinand deAmat<sup>2</sup>, and Gilda Zea<sup>1</sup>

<sup>1</sup>Anatomic Pathology and <sup>2</sup>Dermatology Services, Hospital Carlos Seguin, Arequipa, Peru.

Rosai Dorfman disease is a non-clonal histiocytic proliferative disorder classically characterized by cervical lymphadenopathies with sinus expansion by large histiocytes with emperipolesis of lymphocytes and plasma cells. However, there are described extranodal cases, alone (23%) or in combination (43%) with nodal involvement. Here, we describe 8 cases of exclusive extranodal Rosai Dorfman disease, located in skin (5), soft tissue (2) and nasal mucosa (1). The median age was 43 years; 6 were female and 2 men. The location was in the trunk in 4 cases, one in the arm, the pinna and face. Five cases had jobs related to chemical exposure. Histologically, there was a vaguely nodular infiltrate composed of lymphocytes, plasma cells and large histiocytes, with ample cytoplasm, huge nuclei and prominent nucleoli; they evidenced emperipolesis of lymphocytes and plasma cells. These histiocytes were immunohistochemically reactive to S-100 and CD68. There was no evidence of clonal lymphocyte or plasma cell proliferation. Cases with large and multiple lesion persisted; larger tumors persisted, recurred and were locally infiltrative.

### Corrosion of Cutaneous Ferruginous Bodies Mimic Melanocytic Tumors and Other Lesions

B.D. Ragsdale, MD and Andrea Conway, BS  
Western Pathology, Inc. San Luis Obispo, CA.

Ferruginous foreign bodies that are embedded in the skin are excised when they become more symptomatic or mimic other lesions. This diagnosis should be thought of when distinctive corrosion products (CRPs), essentially rust, are seen in H & E sections. Twenty such specimens, mostly from men (17/20) and primarily from the extremities (14/20), were retrieved from file. CRPs are of 3 types in H&E slides: finely brown granules coarser than hemosiderin, blue globules and irregular glassy translucent yellow particles. Prussian blue iron stain renders only the first of these CRPs blue. None of this signature of iron corrosion is birefringent. Finding an empty space in dense fibrous tissue should prompt asking of the technical staff if a metallic foreign body was free in the container or removed during processing. Sometimes such impediments to cutting are removed from tissue or paraffin blocks and saved without mention in gross descriptions. Pre biopsy clinical radiographs and specimen x-ray of paraffin/blocks are alternate routes to confirmation that indeed a metallic foreign body was the cause. Scanning electron microscopy with energy dispersive x-ray spectroscopy (SEM/EDS) will confirm iron in the CRPs. Only in retrospect, will a minority of patients be able to voice a context for the original injury, 6 of the 20 in the present series, as varied as metal work, auto accidents and projectile trauma. Lack of patient recall does not negate the diagnosis when typical CRPs are evident.

### A Surfers' Knot, Untangled by Patient History

Bruce D. Ragsdale, MD<sup>1</sup>, Larisa M. Lehmer, MS<sup>2,3</sup>, Andrew J. Kaufman, MD<sup>3</sup>

<sup>1</sup>Western Pathology, Inc.; <sup>2</sup>UCISOM; <sup>3</sup>Center for Dermatology care, Thousand Oaks, CA.

One (10-cm) of 2 symmetrically rounded masses along the lower anterior rib cage margin was excised from a 44-year-old male and submitted with an outside pathologic report of low-grade liposarcoma recommending re-excision. No lipoblasts were in the adipocyte islands. The multiplicity and irregularity of islands of mature fat in copious dense fibrous tissue was unlike fibrolipoma that has trabeculations of non-dominant collagenous tissue. The patient, an avid surfer, indicated these bilateral lesions had been slowly enlarging over 19 years. His fellow short board riders have similar, smaller chest wall masses. Up to 80% of California surfers have surfer's "knots" or "nodules," movable subcutaneous non-tender masses most common anterior to proximal tibia and on feet. Intermittent pressure against a surfboard induces localized connective tissue overgrowth. Generally 1–6 cm in diameter, they create a "bio-pad" over bone. Aside from cosmesis, ulceration, often with low grade infection, can motivate surgery. Prevention is possible with protective padding. Clinical history is the route to avoid misdiagnosis and overtreatment as a lipomatous neoplasm.

### Cutaneous Rosai-Dorfman Disease at the Site of an Influenza Vaccination

Sophia Rangwala, MD, Wendi Wohltmann, MD, and James E. Fitzpatrick, MD  
Department of Dermatology, University Of Colorado Health Sciences Center, Aurora, Colorado.

Rosai-Dorfman disease (RDD) is a rare idiopathic histoproliferative disease characterized by massive painless cervical lymphadenopathy, fever, and weight loss. A dense proliferation of large histiocytes exhibiting emperipolesis and S100 immunoreactivity is seen histologically. One-third of cases involve an extranodal form of disease, with purely cutaneous disease having been reported in about 20 patients. Many cases of RDD have appeared to occur after a viral or bacterial illness, suggesting that the disease may be the result of an aberrant immunological host response to an infectious agent. We report the case of a 55-year-old female who developed pure cutaneous RDD after 1 month at the site of an inactivated influenza vaccination on the upper arm. This is the first reported association between the influenza virus and RDD. Additionally, this is the second reported association of RDD occurring at the site of a vaccination, lending further support to the

theory that RDD is the result of a dysfunctional immune response triggered by an infectious antigen.

### Malignant Melanoma and Cutaneous Mastocytosis- Is There a Pathogenetic Link?

Rachel Marchalik, BA<sup>1</sup>, and Jeave Reserva, MD<sup>2</sup>

<sup>1</sup>Georgetown University School of Medicine, Washington DC; <sup>2</sup>Medstar Washington Hospital Center, Washington DC.

Mastocytosis is a disorder characterized by the proliferation and accumulation of mast cells in tissues, most commonly the bone marrow and skin. Although melanocytes and mast cells arise from different cell lines, they both express transcription factors MITF and STAT3, and depend on KIT and its ligand, stem cell factor. While the majority of systemic mastocytosis exhibit KIT mutations, only 40% of melanomas demonstrate genetic aberrations in KIT and are usually different from the mutations seen in mastocytosis. However, to date, there have been 5 case reports published revealing a correlation between mastocytosis and melanoma. Three cases demonstrated systemic mastocytosis while the remaining 2 showed no ostensible systemic manifestation. We report a 56-year-old Caucasian woman with a past medical history of breast cancer and cutaneous mastocytosis (negative bone marrow biopsy) who presented with a pigmented macule on her upper back. Biopsy revealed a pT1b melanoma for which she underwent wide local excision. Five year follow up revealed no evidence of melanoma recurrence. We present this case to raise awareness of the possible relationship between cutaneous mastocytosis and melanoma.

**Acknowledgments:** None.

### Nuclear Dust Beyond Leukocytoclastic Vasculitis

Sonal Choudhary, MD, Mariya Miteva, MD, and Paolo Romanelli, MD  
Department of Dermatology University of Miami, Miami, Florida.

Nuclear dust (ND) is from breakdown of nuclei resulting in fallout. It presents as aggregates of small dark granules. It is most commonly identified with neutrophilic karrhyorhexis in leukocytoclastic vasculitis (LCV) although it is not restricted to neutrophils only by definition. It is less known that other vasculitic and non-vasculitic conditions may demonstrate ND. This knowledge helps to avoid over and misdiagnoses. We reviewed the literature to investigate all conditions other than LCV, which present with ND. A total of 112 articles were obtained by searching pubmed, MeSH, textbooks and monographs for "nuclear dust, karrhyorhexis, leukocytoclasia." We identified 32 entities. These include several categories, all of which demonstrate varying degrees of ND: 1) Vasculitides: medium size vessel, lymphocytic, eosinophilic and granulomatous vasculitis. 2) Antibody mediated disorders (Dermatitis herpetiformis, Bullous Lupus Erythematosus), 3) Neutrophilic dermatoses; 4) Inflammatory or infectious dermatoses (Granuloma annulare, Necrobiotic xanthogranuloma, Bacillary angiomatosis, certain Panniculitides) and 5) Miscellaneous.

**Conclusion:** The source of dust is not always neutrophilic and in a given case may be from involvement of multiple cell types (inflammatory, basal layer keratinocytes and endothelial cells).

### A Primary Cutaneous CD30-Positive T-Cell Lymphoproliferative Disorder Arising in a Patient With Multiple Myeloma and Cutaneous Amyloid

Ryan C. Romano, Daniel N. Cohen, Matthew T. Howard, and Carilyn N. Wieland

Mayo Clinic, Rochester, MN.

CD30+ cutaneous lymphoproliferative disorders, a group of T-cell lymphomas including lymphomatoid papulosis (LyP) and cutaneous anaplastic large cell lymphoma, requires careful clinicopathologic correlation for diagnosis. Reports indicate an association with LyP and hematologic

malignancies. A 66-year-old woman with cutaneous amyloidosis secondary to multiple myeloma (MM) presented with erythematous and dark-brown papules involving the right arm, scalp and torso. Punch biopsies showed a non-epidermotropic dense dermal infiltrate of intermediate-sized plasmacytoid lymphocytes and multifocal amyloid deposition. The infiltrate was immunophenotypically a CD30-positive, ALK-negative T-cell lymphoproliferative disorder. She had no systemic involvement, and clinical correlation suggested a final diagnosis of LyP. Systemic amyloidosis, a complication of MM, has been reported in association with LyP but these reports have not documented cutaneous involvement. This case of a primary cutaneous CD30+ T-cell lymphoproliferative disorder consistent with LyP but with histologic features mimicking a B-cell proliferation arising in association with cutaneous amyloidosis highlights the importance of clinicopathologic correlation, a thorough immunohistochemical evaluation, and consideration for a second hematologic malignancy in association with LyP.

### Vulvar Erythrasma: Potential Cause for Persistent Symptomatology in Lichen Sclerosus

Michelle Schneider<sup>1</sup>, and M. Angelica Selim<sup>1</sup>

<sup>1</sup>Duke University Department of Pathology, Durham, North Carolina.

Erythrasma, a superficial infection caused by *Corynebacterium minutissimum*, is characterized by discoloration of intertriginous areas that are warm, moist and occluded. Appearing coral red on Wood lamp examination, it is asymptomatic or mildly pruritic. Once recognized, treatment is safe and effective. Erythrasma of the vulva is rare, with 1 case described in the English literature. We report 3 cases of erythrasma diagnosed on vulvar biopsy. 2/3 patients underwent biopsy for clinical suspicion of lichen sclerosus (LS), while the third had known LS and required evaluation for dysplasia. In all, pruritus and pain had recently worsened. Histologic examination revealed filamentous organisms in the stratum corneum consistent with *Corynebacterium*. One patient had minimal dermal fibrosis but no definitive changes of LS, the second was confirmed to have LS, and the third with known LS had no dysplasia identified. Secondary bacterial infections in LS are uncommon, with no reported cases of coexisting *Corynebacterium* infection. Erythrasma of the vulva is an important phenomenon to recognize as it may mimic LS, worsen the symptoms of established LS, or mimic development of dysplasia.

### Acquired Smooth Muscle Hamartoma of the Labia Majora: A Case Report

Kabeer Shah, and Andrew Folpe

Mayo Clinic, Rochester, MN.

Dermal smooth muscle hamartomas occur in 2 forms, congenital and acquired. Acquired smooth muscle hamartomas (ASMH) are much less common than are congenital lesions. Vulvar ASMH is especially uncommon, with only a single reported case. A 53-year-old woman presented with a 2 cm plaque of the right labia majora. The lesion had been present for 1 year, without change. An incomplete excision was performed. Sections showed involvement of the dermis and subcutis by a haphazard proliferation of small, hypocellular bundles of well-differentiated, smooth muscle actin and desmin-positive smooth muscle, lacking nuclear atypia or mitotic activity, diagnostic of ASMH. No additional surgery was performed, and the patient is currently asymptomatic. This appears to be only the second reported vulvar case of ASMH. Cases of ASMH have also been reported in the scrotum, although these may represent instead lymphedema-associated hyperplasias of Dartois smooth muscle. Lymphedema does not seem to be associated with genital ASMH in women. Vulvar ASMH should be distinguished from pilar leiomyosarcoma, and from myoid tumors of the genital region, including typical leiomyoma, cellular angiofibroma, and angiomyo-fibroblastoma.

### Necrolytic Migratory Erythema Resulting From Severe Nutritional Deficiency

Maria Sheron, MD, Anthony Fernandez, MD, PhD, and Melissa Piliang, MD  
Department of Dermatology, Cleveland Clinic, Cleveland, OH.

**Description:** A 33-year-old female with NASH cirrhosis and history of gastric bypass presented to the ICU for sepsis and a severe exfoliative rash. Dermatology was consulted for concern for Stevens-Johnson Syndrome/Toxic Epidermal Necrolysis. Review of systems was positive for erythema, pain, and edema of the lower extremities for 1 week. Examination revealed fissured, bright red orolabial mucosa, as well as dusky patches and desquamation on the posterior lower extremities and buttocks. Pathological examination revealed parakeratosis and necrosis overlying a subcornal split with keratinocyte pallor and vacuolization. The presence of a glucagonoma was ruled-out. Laboratory analysis demonstrated deficiencies of multiple amino acids, 25 hydroxyvitamin D, copper, biotin, manganese, and pre-albumin. The patient began a tailored nutritional program and rapid improvement of the skin was noted. It is important to recognize that Necrolytic Migratory Erythema may occur in the absence of a glucagonoma and have an atypical clinical presentation.

### Neuroblastoma-Like Schwannoma A Rare Variant With Unusual Morphology

Julie Snauwaert, Rajiv Patel, and Aleodor Andea

University of Michigan Health System, Department of Pathology, Ann Arbor, MI.

Neuroblastoma-like schwannoma (NLS) is a rare variant of schwannoma with only 15 cases reported to date. We document a new case of this unusual variant. A 38-year old male presented with a 1.3 cm, asymptomatic, slowly enlarging, mobile subcutaneous nodule that was first noticed about 1 year ago. Histology showed an encapsulated tumor in the deep dermis and subcutis. The most conspicuous feature was the presence of variably-sized rosette-like structures with irregular contours composed of an inner core of thin eosinophilic radiating collagenous fibers surrounded by small epithelioid cells with high N:C ratio, hyperchromatic nuclei, and occasional intranuclear pseudoinclusions. Despite the presence of some low-grade cytologic atypia, the lesion uniformly lacks overt pleomorphism, significant mitotic activity, necrosis, or cystic degeneration. Areas with Antoni B classic features were also noted. Similar to conventional schwannomas, the tumor cells were diffusely positive for S100. Differential diagnosis includes other tumors with pseudorosettes such as neuroblastoma, hyalinizing spindle cell tumor with giant rosettes and dendritic cell neurofibroma. Familiarity with this rare variant helps in establishing the correct diagnosis.

### Infliximab Treatment of Pyostomatitis Vegetans

Stanislav N. Tolkachjov, MD<sup>1</sup>, Jill K. Henley, DO<sup>2</sup>, Jason S. Reichenberg, MD<sup>3</sup>, and Alina G. Bridges, DO<sup>1</sup>

<sup>1</sup>Department of Dermatology, Mayo Clinic, Rochester, Minnesota; <sup>2</sup>Department of Internal Medicine, Loyola University, Maywood, Illinois; and <sup>3</sup>Department of Dermatology, UTSW-Austin, Austin, Texas.

Fifty-four year-old Caucasian male presents with 11-month history of painful, erythematous, crusted and eroded plaques with cobblestoning on the upper and lower mucosal lips. Crohn disease (CD), diagnosed more than 10 years ago, was treated with oral mesalamine and prednisone. Biopsies were consistent with pyostomatitis vegetans (PSV) showing pseudoepitheliomatous hyperplasia, spongiform pustule formation, and intraepithelial neutrophilic microabscesses. Symptoms did not improve after several months of prednisone at 60 mg, intramuscular triamcinolone, dapsone, mycophenolate mofetil, and topical corticosteroids and calcineurin inhibitors. Infliximab infusions of 5 mg/kg were initiated. Complete resolution was achieved within 4 months. The intervals between infusions were increased due to continued remission. PSV has

been associated with inflammatory bowel disease, irritable bowel syndrome, primary sclerosing cholangitis, and liver disease. PSV presents with cobblestoning of the oral mucosa with localized microabscesses, yellow pustules, and vegetations. Our case illustrates the typical clinical presentation, histopathologic findings, and successful treatment of an often refractory disease.

### Cystic Acantholytic Dyskeratosis of the Vulva

Kara Melissa T. Torres<sup>1</sup>, Nathan Cleaver<sup>1</sup>, and Jacqueline M. Junkins-Hopkins<sup>1</sup>  
<sup>1</sup>The Ackerman Academy of Dermatopathology, New York, NY.

**Background:** Acantholytic dyskeratosis of the vulva (ADV) is an uncommon condition of the vulvocrural skin that is histologically similar to Darier's and Hailey-Hailey disease, but lacks the family history of these disorders. Localization of the lesions to the perineal area further differentiates ADV from these disorders and from Grover's disease. Typically, there are isolated or grouped dome-shaped white or violaceous papules. Cystic lesions have not been described.

**Case:** A 53 year old female presented with an asymptomatic cyst on her left vulva of uncertain duration. Microscopic examination revealed cystic epithelium with areas of hypergranulosis, acantholysis, corps ronds and grain formation. There was no known history of generalized skin conditions with similar histology.

**Conclusion:** Acantholytic dyskeratosis in this location is uncommon. A review of the literature revealed a handful of cases, usually reported as papular acantholytic dyskeratosis. The cystic presentation noted in this patient has not been reported to our knowledge. The involvement of cystic infundibular epithelium is felt to be benign and similar to warty dyskeratomas seen on non-genital skin.

### Photo-Accentuated Dermatophytosis Mimicking a Photodistributed Lichenoid Drug Eruption

Katherine Tumminello, Kimberly Nicholson, Katherine Fening, and Kristopher McKay

University of Alabama at Birmingham, Section of Dermatopathology, Birmingham, AL; and Atlanta Dermatopathology, Atlanta, GA.

Dermatophytosis has been shown to mimic skin disorders such as photodermatitides. We present the case of a 48-year-old male with a history of type 2 diabetes and hypertension on numerous medications. He presented with a chief complaint of diffuse pruritus. The rash waxed and waned for years; his arms sometimes blistered during flares. There were large erythematous patches on the trunk and forearms with focal areas of papules coalescing into thin plaques. There was striking accentuation of the process in sun exposed sites, especially the chest and forearms. The clinical diagnosis was a photo accentuated lichenoid drug eruption. A biopsy revealed fungal hyphae in the stratum corneum. One theory suggests that photo-exacerbation may be due to increased capillary permeability in sun-damaged areas. This case demonstrates an atypical presentation of dermatophytosis which highlights the importance of remembering dermatophytosis in the differential diagnosis of dermatoses with unusual clinical features.

### Polypoid Melanoma, Nodular Type. A Case Report

Manuel Valdebran<sup>1</sup>, Karen N. Wu<sup>2</sup>, Andrew W. Wu<sup>3</sup>, and Jacqueline Junkins-Hopkins<sup>1</sup>

<sup>1</sup>Ackerman Academy of Dermatopathology, New York, NY; <sup>2</sup>Pathology Specialists of New England. Catholic Medical Center, Manchester, NH; and <sup>3</sup>Surgical Care Group Catholic Medical Center, Manchester, NH.

**Introduction:** Polypoid melanoma is a unique variant of melanoma with a characteristic exophytic growth pattern that shows no invasion of the reticular dermis, differentiating it from its nodular counterpart. Many times it can present as a benign non-pigmented lesion thereby delaying the diagnosis.

**Case Report:** A 64 year-old male presented with a 6-month history of a growing nodule on his left flank. On physical examination, a 3 cm, well circumscribed round erythematous nodule with a partially eroded surface was found. There was no evidence of lymphadenopathy. His past medical history was non-contributory. Histopathological examination of the lesion showed an atypical polypoid tumor composed predominantly of melan-A-positive melanocytes, both spindled and epithelioid in morphology, with a vertical growth phase and abundant mitoses. An HMB 45 immunostain demonstrated diffuse positivity with no evidence of a gradient throughout the lesion. A computerized tomography scan failed to reveal any evidence of metastatic disease.

**Conclusion:** The benign appearance of PM and its amelanotic nature may delay the diagnosis; at the time of presentation the lesions might have a significant vertical growth phase with an increasing risk of metastasis and poor prognosis.

### Cutaneous Extramedullary Hematopoiesis: A Rare Neoplastic MIMIC

John S. Van Arnam, M. Angelica Selim, and Anand S. Lagoo  
Duke University Pathology, Durham, North Carolina.

Myelofibrosis is a clonal neoplasm which results in replacement of marrow with connective tissue and leads to decreased and displaced hematopoiesis. Compensatory extramedullary hematopoiesis can occur in a wide variety of locations in the body-including rare instances in the skin. We present the case of a 65 year old woman with history of Jak2 positive polycythemia vera and secondary myelofibrosis with pancytopenia who presented with new painful dermal masses on her left shoulder. Biopsy demonstrated a dermal infiltrate of maturing myelocytic and histiocytic cells. The larger, often multinucleated cells demonstrated staining for CD41 and CD61, confirming their megakaryocytic lineage. CD34 highlighted only rare myelocytic cells and CD117 predominantly stained mast cells, suggesting against a diagnosis of leukemia cutis/granulocytic sarcoma. Although rare, correlation with clinical history and immunohistochemistry allows for identification of this rare phenomenon and distinction from myeloid leukemia.

### Report of a Dermal Adnexal Tumor Consistent With Trichogerminoma

John S. Van Arnam, M. Angelica Selim, and Thomas J Cummings  
Duke University Pathology, Durham, North Carolina.

Trichogerminoma is an exceptionally rare cutaneous adnexal tumor, representing a poor recapitulation of the multiple layers of the mature hair follicle along with primitive epithelial nests. We report a case of a 45 year old woman who presented with a subcutaneous nodule on her chest. Histologically, the tumor is characterized by a lobular basaloid proliferation with fibrotic stroma, well-demarcated from the surrounding tissue. There are scattered cell nests with dispersed chromatin imparting a pale appearance in contrast to the more hyperchromatic peripheral cells which demonstrate palisading. Immunohistochemistry for CK5/6, p63, and Bcl2 demonstrated strong ring-like staining in the periphery of the lobules with decreased or negative staining in the aforementioned nests, occasionally positive for CD10. The Ki67 index is moderate at 5%, with scattered staining throughout the lobules. Whether all trichogerminomas are a distinct entity or represent a trichoblastoma with more regions of more significant differentiation is unclear. Although most excisions usually are curative, there is a reported case of trichogerminoma evolving to undifferentiated carcinoma with metastasis which necessitates followup.

### An Umbilical Mucosal Polyp in an Otherwise Healthy 8-Month-Old Boy

Matthew Vasievich, MD, PhD, Ivanka Kovalyshyn, DO, Mahwish Irfan, MD, Anthony Fernandez, MD, PhD, and Allison Vidimos, MD  
Cleveland Clinic Foundation, Cleveland, Ohio.

**Objectives:** -Describe the clinical and histologic features of an umbilical mucosal polyp. -Discuss the differential diagnosis of an umbilical mass in

an infant. An otherwise healthy 8-month-old Caucasian male presented to the dermatology clinic for evaluation of a red, bleeding umbilical mass. The lesion had been present since loss of the umbilical stump at 1 week of life. It bled easily with Valsalva or trauma. On examination, the umbilicus demonstrated a bright red, friable, 5 mm papule. Differential diagnosis included umbilical mucosal polyp/omphalomesenteric remnant, pyogenic granuloma, umbilical hernia, and patent urachus. A shave biopsy revealed an ulcerated epidermis and granulation tissue with adjacent acanthosis and underlying normal appearing colonic mucosa with prominent lymphoid aggregates with well-developed germinal centers. These findings were consistent with a diagnosis of umbilical mucosal polyp. This case demonstrates the importance of considering a broad differential diagnosis when a midline lesion presents in an infant.

### Mutiple Simultaneous Dermatofibromas: Case Report and Review

David Wang, MD and Wendi Wohltmann, MD  
*Univeristy of Colorado School of Medicine.*

Dermatofibromas are rare, fibroblastic and myofibroblastic dermal neoplasms that present as discrete, indurated, plaque-like lesions, most commonly on the neck, trunk and proximal extremities. To our knowledge, only 1 case of multiple lesions has been reported; a pediatric patient. We report a unique case of dermatofibromas occurring simultaneously within 1 year as a 20 mm pigmented shoulder lesion, a 13 mm thigh induration, and a 15 mm pigmented calf lesion occurring in a 19 year old Hispanic woman. No significant medical or family history was reported. Her biopsies revealed characteristic histological findings- short fascicles of bland spindled cells oriented parallel to the epidermis and sparing adnexal structures. The lesional cells only labeled with Vimentin; Alpha-smooth muscle actin, Muscle actin, and Calponin were negative. Elastic fibers were preserved throughout the lesion. CD34 labeled a significant amount of background stroma. Apart from the multiplicity of this case, the immunohistochemical profile lends itself to an interesting discussion concerning the evolution of these myofibroblastic/fibroblastic tumoral cells and concerning a differential diagnosis that includes Atrophic DFSP, Fibroblastic Connective Tissue Nevus, and Plaque-like CD34 dermal fibroma.

### Follicular Herpesvirus Infection in a Tzanck Smear-Negative Patient

Katie Wang,<sup>1</sup> Meryl Rosen,<sup>2</sup> Alex Flamm, MD,<sup>3</sup> Raman Madan, MD,<sup>4</sup> Melissa Serravallo, MD,<sup>5</sup> and Ed Heilman, MD<sup>6</sup>  
<sup>1</sup>KCUMB, <sup>2</sup>SUNY Downstate School of Medicine,<sup>3-6</sup> SUNY Downstate Department of Dermatology.

We present the case of a 39-year-old man who came to the emergency department with a 3-day history of a pruritic, vesicular rash, accompanied by a low-grade fever. He was treated for syphilis 2 months prior, but denied HIV infection or other immunosuppressive conditions. The patient reported a history of chicken pox as a child. Physical examination was significant for a diffuse, vesicular, mildly erythematous rash on the head, trunk, extremities, left palm, and hard palate. Some excoriations were noted. We performed a Tzanck smear of the vesicles and obtained a 4 mm punch biopsy. Despite the presence of clear vesicles on erythematous bases, the Tzanck smear was negative. However, the histopathology stained in H&E revealed multinucleated giant cells located at the base of a hair follicle. In a Tzanck-negative, but clinically consistent case of herpesviridae infection, consider the possibility of follicular herpes and follow up with a skin biopsy.

### Interstitial Granulomatous Dermatitis in the Setting of Microscopic Polyangiitis

Daniel Winchester, MD, Alina Bridges, DO, and Julia Lehman, MD  
*Mayo Clinic, Rochester, MN.*

**Background:** Interstitial granulomatous dermatitis (IGD) is a rare cutaneous eruption typified by the development of erythematous papules or, rarely,

indurated cords. Microscopic features include interstitial granulomatous inflammatory infiltrate surrounding small foci of degenerated collagen. Systemic diseases such as rheumatoid arthritis and lupus erythematosus are found in 30% of cases. To date, no cases associated with microscopic polyangiitis (MPA) have been described.

**Case:** A 63 year old woman developed progressive dyspnea, fevers and an asymptomatic, faintly erythematous papular eruption on the bilateral lower legs. Labs showed a normocytic anemia, elevated ESR and CRP and perinuclear antineutrophil cytoplasmic antibodies (ANCA) with myeloperoxidase specificity. Skin biopsy showed interstitial histiocytes with partially degenerated collagen, consistent with IGD. Damage to the cutaneous vasculature was not evident.

**Discussion:** IGD is an immune complex deposition disease often associated with underlying systemic illnesses. MPA is a pauci-immune vasculitis presenting with fevers, weight loss, anemia, elevated ESR, and systemic involvement of the kidneys, lungs, and other organs. Often, the cutaneous manifestations of MPA include palpable purpura or erythematous macules and papules, often showing vasculitis on skin biopsy. This patient had the first reported case of IGD presenting in the setting of microscopic polyangiitis.

### High-Velocity Paint Gun Injuries: A Report of 2 Cases

Wendi Wohltmann, MD, Ryan Stevens, MD, and Joshua Wisell, MD  
*Dermatopathology Fellow; Dermatopathology Fellow; Department of Pathology, Assistant Professor; University of Colorado, Denver.*

Two patients presented with enlarging nodules at sites of high-velocity paint gun injury 7 and 15 years prior. One had a 3.5 cm flesh-colored tender dermal nodule on the medial left cheek. The second patient had a 5 cm tender exophytic nodule on the left ventral forearm. Punch biopsies showed a dense fibrohistiocytic infiltrate within the dermis arranged around polarizable crystalline fragments present within both the tissue and foreign body giant cells. There was copious black pigmentation composed of minute spheres (tattoo-like pigmentation, Perl's iron stain negative) present throughout both lesions. CD68 and procollagen-1 expressed strong positivity among the cells of interest. We believe the patient's nodules are resultant from the particles introduced into the skin traumatically from the high-pressure paint guns. The onset years after injury and the expanding nature were commonalities in each case. Clinical history is of paramount importance in these cases, as the histopathologic findings could be mistaken for foreign body reaction to tattoo or giant cell tumor of the tendon sheath in the proper anatomic location.

### Lymphangiectatic Variant of Eccrine Spiradenoma: A Rare Variant

Usama Yousif, MD, Amy Kerkvliet, MD, R.J. Summerer, DO, and Ali Jassim, MD, PhD  
*Sanford School of Medicine, University of South Dakota.*

Conventional eccrine spiradenoma is a benign, slow growing and painful tumor of the skin. While the tumor does not usually present a diagnostic dilemma, a rare variant with marked stromal lymphedema can be a challenge to interpret. We present a case of lymphangiectatic variant of eccrine spiradenoma in an 82-year-old white male who presented with a persistent left flank lesion for several months. The patient was initially asymptomatic and subsequently developed a suspected abscess that was excised to reveal a 6.5 cm subcutaneous mass. Microscopic examination reveals strands and cords of dark, epithelial, round to oval cells with inconspicuous nucleoli streaming between prominently dilated and congested vascular spaces. Within the cystic component there are small ductular structures. Additionally, prominent stromal lymphedema is present. To the best of our knowledge, there is only 1 reported case of this entity in the English literature. This case represents a diagnostic challenge and the purpose of reporting it is to alert surgical pathologists and dermatopathologists of the existence of this unusual variant of eccrine spiradenoma.