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ORAL ABSTRACT PRESENTATIONS, WEDNESDAY 3-18-15, 8:00–10:00 AM

Evaluation of Symptoms in 27 Patients With Diagnosis of Acrodermatitis Enteropathica in Iran

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Acrodermatitis Enteropathica (AE) is an inherited autosomal recessive disorder caused by a mutation in the hZip4 zinc transporter gene, which reduces zinc absorption and all body zinc levels. The objective was to determine the symptoms and blood zinc level ranges of patients and determine their relationship. The study is a retrospective cross-sectional study that surveyed 27 infant patients with severe zinc deficiency that was suggestive of AE, who were referred to Razi Hospital's Dermatology Center in Iran during 1999 to 2004. The average age and disorder onset time were 8.9 months and 37 days after birth, respectively. The average serum zinc level was 45.3 µg/dL. The statistically significant symptoms of AE can be listed as perioral, perinasal, perigenital, and perirectal lesions, acral lesions and lesions on fingers, hands, elbow and feet, diarrhea and stomach ache, diffused alopecia, and neurological symptoms like fatigue, moodiness, irritability, photophobia, and anorexia. Physicians should be aware of this largely misdiagnosed disorder and its specific symptoms to increase recognition and early treatment of it in infancy when it is often fatal.

An Atypical Outbreak of HFMD in Adults

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Most cases of Hand, Foot, and Mouth Disease (HFMD) are seen in young children <5 years old presenting with fever and a vesiculobullous eruption of the hands, feet, and mouth. Here we report an atypical outbreak of 12 HFMD cases in May–December 2014 in an adult cohort in a large urban setting. Retrospective data analysis over the last 10 years (2005–2014) revealed an unusually high number of cases in the previous 8 months, comprising of 100% adult patients (age 27–76). Clinically as well as histologically, these cases resembled erythema multiforme. EM shows interface changes and individually necrotic apoptosis of keratinocytes. The HFMD cases showed more prominent reticular degeneration, more ballooning of basal keratinocytes, and less interface changes. Additionally, papillary dermal edema and a mixed perivascular inflammatory infiltrate with neutrophils were also seen in the HFMD cases. Clinically atypical variants were seen with both widespread (“eczema coxsackiemi”) and limited involvement in this cohort. Enteroviral tropism for anterior hand/feet keratinocytes suggests that these areas facilitate transmission through direct contact. The atypical anatomic presentation (33% of the cases) including on the knee, wrist, and forearm is not inconsistent with this theory.

A Clinically Validated Gene Expression Score Impacts Diagnosis and Management Recommendations of Melanocytic Lesions

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Background: Recently, a 23-gene expression signature was clinically validated as an adjunctive tool to differentiate benign nevi from malignant melanomas. The goal of this study was to quantify the impact of that test on diagnosis and management recommendations made by dermatopathologists.

Methods: Difficult to diagnose melanocytic lesions encountered during routine practice were submitted for gene expression testing and received a melanoma diagnostic score (MDS). Submitting dermatopathologists completed a survey on the pre-test diagnosis, level of diagnostic confidence, further evaluation to be performed, and management recommendations. The survey was repeated after receiving the MDS. Changes between the pre- and post-test surveys were analyzed.

Results: Thousand six hundred and ninety-five eligible cases were submitted by 79 dermatopathologists. When the MDS was available in diagnostically challenging cases, indeterminate diagnoses were reduced by 42.7%. Additionally, treatment recommendations were revised in 29.1% of all cases.

Conclusions: The MDS impacts diagnosis and management recommendations by dermatopathologists confronted with diagnostically challenging melanocytic lesions.

Beta-Papillomavirus Infection of Paradoxical Cutaneous Squamous Cell Carcinoma During Braf-Inhibition Therapy

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BRAF-inhibition (BRAFi) melanoma treatment carries a risk of secondary cutaneous squamous cell carcinoma (cSCC) and other cancers. Cutaneous beta-HPV may act with host and environmental factors in BRAFi-cSCC.

Methods: Primary BRAFi-cSCC tissue DNA, isolated from patients on BRAFi (vemurafenib or dabrafenib), was tested for cutaneous oncogenic viruses and somatic mutations. Clinical parameters were statistically analyzed with histopathology.

Results: Twenty-nine patients contributed 69 BRAFi-cSCCs, of which 22% had wart-like features (BRAFi-cSCC-WF). During vemurafenib therapy, BRAFi-cSCC-WF arose 11.6 weeks more rapidly than conventional-cSCC controlling for gender and UV-exposure (p -value = 0.03). HPV-17, HPV-38 and HPV-111 composed 49% of infections. Three novel beta-HPV genotypes were discovered. *RAS* mutations occurred in 63% of evaluated BRAFi-cSCC. *PIK3CA*, *EGFR* and unique *CKIT* and *ALK* mutations were also detected.

Conclusions: BRAFi-cSCCs demonstrate rapid onset, frequent wart-like histomorphology, beta-HPV infection, UV-damage and *RAS* mutation. These findings enhance our understanding of keratinocyte oncogenesis and broaden the knowledge base of multifactorial mediators of cancer.

Cellular Blue Nevomelanocytic Lesions: Analysis of Clinical, Histological, and Outcomes in 37 Cases

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Cellular blue nevomelanocytic lesions (CBNL) frequently pose diagnostic problems to pathologists and their biological potential may be difficult to establish. In this study, we have analyzed the clinical, histological, and outcomes data of 37 CBNL, as well as the molecular characteristics of 4 lesions. Our cohort of cases comprised of 8 cellular blue nevi (CBN), 17 atypical cellular blue nevi (ACBN), and 12 blue nevus-like melanoma (BNLM) with a mean follow-up of 5 years. Seven CBN cases with follow-up had a benign clinical course (average follow-up 4.7 years). Among 6 patients with ACBN with sentinel lymph node (SLN) biopsy results, 3 were positive, and a single additional case had 1 positive non-sentinel lymph node. All 14 cases of ACBN with follow-up were alive and without recurrence with mean follow-up of 5 years. Of the 8 melanoma cases with follow-up, 2 patients with sentinel and non-sentinel lymph node involvement succumbed to their disease (average follow-up period of 4.8 years). Array comparative genomic hybridization (aCGH) was performed on 2 ACBN and 1 BNLM: One of the 2 ACBN showed chromosomal aberrations, and the 1 BNLM showed multiple chromosomal gains and losses. Multiplex polymerase chain reaction (PCR) was performed on 1 ACBN, and no mutations were found. From these results, we conclude that ACBN occupy an intermediate position within the spectrum of CBN and BNLM.

The Novel Use of Pre-Operative Epidermal Coloring of Very Small Dermatological Specimens—Protocol For Reduction of Lost Specimens

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Small tissue biopsies are often difficult to visualize and can be easily lost or mishandled as they travel from the patient to the dermatopathology lab. We hypothesized that full epidermal surface coloration of small skin lesions with a skin marker (genetic violet ink) prior to performing a shave biopsy would make gross specimens easier to identify without impacting microscopic appearance. Live evaluation of 4 inked and 4 non-inked gross (2–3 mm) specimens in covered and uncovered formalin containing jars by fifty consecutive healthcare personnel showed that inked specimens were significantly ($p < 0.001$) easier to visualize than non-inked specimens. Additionally, a blinded dermatopathologist evaluated 25 inked and 25 non-inked specimens microscopically and determined that use of this inking process did not interfere with histopathologic assessment or impede diagnosis. This pilot study describes an easily implementable quality improvement measure that may decrease the rate of loss and mishandling of specimens.

Comparative Analysis of Cytokeratin 15, TDAG51, Cytokeratin 20 and Androgen Receptor in Sclerosing Basaloid Neoplasms and Variants of Basal Cell Carcinoma

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Background: Desmoplastic trichoepithelioma (DTE), morpheaform basal cell carcinoma (BCC) and microcystic adnexal carcinoma (MAC) are sclerosing basaloid neoplasms with overlapping histopathologic features. We compared cytokeratin 15, (CK15), T cell death—associated gene 51 (TDAG51), cytokeratin 20 (CK20) and androgen receptor (AR) in differentiating these tumors and also assessed their expression in BCC subtypes.

Methods: Fifteen cases of DTE, 15 cases of infundibulocystic BCC, 18 cases of micronodular BCC, 18 cases of morpheaform BCC and 6 cases of MAC were assessed for CK15, TDAG51, CK20 and AR expression.

Results: Qualitative positivity rates of CK15 ($p = 0.636$) and TDAG51 ($p = 0.768$) were comparable among the sclerosing basaloid neoplasms, while AR ($p < 0.0001$) and CK20 ($p = 0.015$) were significantly different. The CK20+AR- immunophenotype was 100% sensitive and specific in diagnosing DTE. A CK20-AR+ immunophenotype was 95.24% specific and 83.33% sensitive for BCC, while the CK20-AR- immunophenotype was 83.33% sensitive and 90.91% specific for MAC. CK15, CK20, and AR were positive in 87%, 53%, and 67% of infundibulocystic BCC cases, respectively.

Conclusions: The combination of CK20 and AR exhibited the greatest utility in differentiating sclerosing basaloid neoplasms. AR showed limited ability to discriminate infundibulocystic BCC from trichoepithelioma, but CK15, TDAG51 and CK20 do not reliably differentiate between them.

Analysis of Histologic Features of Acral-Lentiginous Melanoma (ALM) in 627 Patients

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Objective: We are presenting a series of patients with ALM, analyzing potential adverse histologic features.

Method: Retrospective review of pathology/medical records for 627 ALM patients (1999–2014).

Results: The mean Breslow thickness was 2.3 mm (0.21–21 mm), 78.9% presented as Clark levels IV or V. Ulceration identified in 42.6% and regression in 15.1% of cases. Perineural invasion (PNI) was evident in 22.1% cases, lymphovascular invasion (LVI) in 16.5%, and microscopic satellitosis in 5.7%. LVI was significantly associated with local recurrence ($p = 0.0129$), diagnosed in 11.7% of patients. Positive sentinel lymph nodes (SLN) were identified in 50.6% cases. Univariate predictors of positive SLN included thickness, ulceration, LVI and satellitosis ($p < 0.0001$). The follow-up was 43.7 months (0.3–187.5). Patients with positive SLN were significantly more likely to develop distant metastases (46% vs 23%) ($p < 0.0001$) and to die from their disease (29% vs 11%) ($p < 0.0001$).

Conclusion: This is one of the largest cohorts of patients with ALM studied to date. The frequency of LVI and PNI invasion are the highest reported and may account for a significantly high rate of positive SLN and satellitosis/in transit metastases.

Ungual Bacteriosis: Massive Bacterial Colonization Per Se Could Be Pathogenetic in the Nail

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In the 70s, myriad of bacteria in Pap smears was known as vaginal bacteriosis, not considered to be a disease. In 1984, the condition was upgraded to “bacterial vaginosis,” then considered a disorder. Nowadays, the CDC considers that bacterial vaginosis is the most common vaginal infection in women aged 15–44. For observers of nail clippings and avulsions, it is obvious that the not infrequent massive population of bacteria among the nails keratins may qualify as unguinal bacteriosis in patients without evidence of onychomycosis. These massively overgrown Gram-positive cocci resemble those found in much lesser amounts in the usual nail. When pathogenic, they tend to induce plate keratolysis or form thick biofilms in the plate

surfaces or fill massively the unguis spaces. This report proposes that the time is prime to consider the possibility that such overwhelming population of bacteria can itself be deleterious to the nail and could induce dystrophy, onycholysis, onychoschizia and, even, surface nail caries. Although it could be argued that the bacteria are secondary inhabitants lodged in abnormal nail spaces induced by nonbacterial etiologic factors, the author will present occurrences that suggest that the bacteria are, on their own, the culprits of the nail abnormalities and not simply commensals. In addition, some fungal infections due to keratolytic fungi, probably molds, are also attended by a very dense mixture of bacteria that obscure the hyphae. This association could be symbiotic but a copathogenic role of the bacteria along the fungi in the destruction of the nail plate could not be entirely excluded. If this hypothesis were plausible, the medical nail caretaker could have additional resources to fight the many nail disorders that are pseudomycotic. Nail hygiene, besides ablation and light soaping of the feet, could evolve into bacteriostatic or bactericidal measures that could improve dystrophic nails that were primarily harboring bacteriosis.

Epithelioid Cell Histiocytoma of the Skin With Clonal *ALK* Gene Rearrangement

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Objective: Epithelioid cell histiocytoma (ECH) is a benign cutaneous tumor of unknown etiology. We aimed to identify molecular aberrations in ECHs demonstrating anaplastic lymphoma kinase (*ALK*) protein expression.

Methods: Two lesions found in a male and female patient (aged 68 and 33 years, respectively), each presenting as a solitary small asymptomatic skin nodule involving the extremities, were evaluated by light microscopy, immunohistochemistry, fluorescent in situ hybridization (FISH), and next generation sequencing.

Results: Both tumors were superficial dermal proliferations of large epithelioid factor XIIIa-positive cells set in a fibrovascular stroma and flanked by an epidermal collarette. Immunoreactivity for *ALK* protein was observed in the majority of the cells. FISH testing demonstrated a clonal *ALK* gene rearrangement. By sequencing, the fusion partners were identified as *vinculin* and *sequestosome 1* genes.

Conclusions: We postulate to consider ECH as a distinct entity of neoplastic etiopathogenesis conceivably driven by the *ALK* gene rearrangement. This study further expands the spectrum of tumors associated with this molecular event.

Uncommon Clinical Presentations of Leprosy: Apropos of 3 Cases

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Abstract: Leprosy is a chronic infectious disease of skin and nerves caused by *Mycobacterium leprae*. In the era of leprosy elimination we now encounter cases with uncommon presentations. Here we present 3 such cases.

Case 1: A 25 years old man presented with erythema, edema and tenderness localized to right hand with overlying pustules. He also had 2 subcutaneous abscesses over right arm and chest wall. Fine needle aspiration of the abscess over arm showed multiple acid fast bacilli.

Case 2: A 52 years old man presented with polyarthralgia, fever and testicular pain. On investigations was found to be seropositive for hepatitis C virus. Thus a provisional diagnosis of poly arteritis nodosa was considered. Two weeks later he presented with multiple tender, erythematous nodules over arms and legs. Skin biopsy from these nodules showed foamy macrophage granulomas with multiple acid fast bacilli.

Case 3: A 54 year old man presented with erythematous scaly papules and plaques over face, arms and legs following insecticide spray. Skin biopsy done with possibility of contact dermatitis showed foamy macrophages with acid fast bacilli in fite stain.

Prurigo Pigmentosa: Clinicopathological Analysis of 32 Cases With Emphasis on its Etiology

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Background: Prurigo pigmentosa is unique among inflammatory diseases of the skin and the singularity of it is manifest both clinically and histopathologically.

Objective: To evaluate the clinical and histopathological features of prurigo pigmentosa and propose its etiological factors.

Methods: All the medical records, photographs and histopathological slides of 32 patients diagnosed with prurigo pigmentosa in Taiwan.

Results: The patient's age ranged from 11 to 79 years (F:M = 2.2:1). Lesions were primarily reddish-brown located on back area occurring during summer season. Minocycline alone or in combination with steroid was effective among antibiotic treatment. The major microscopic features found were association of bacterial colonies revealing either folliculitis or perifolliculitis (21/32). Others were perivascular and dermal lymphocytic infiltration (32/32), spongiosis (21/32), necrotic keratinocytes (17/32), basal cell vacuolization (12/32), dermal melanophages (20/32) and eosinophils (14/32).

Conclusion: Our data suggest that bacterial involvement may play an important role in the pathogenesis of prurigo pigmentosa. However, clinicopathologic correlation and thorough long-term follow-up are necessary to establish a diagnosis of prurigo pigmentosa. Thus, detail investigations for its etiology and pathogenesis have yet to be determined in future.

Braf Expression in Thin and Thick Melanomas: An Immunohistochemical Study

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Braf expression in melanoma is used to determine the utility of kinase inhibitor therapy. Despite routine testing by PCR, little is known of the expression in early stage melanomas and this mutations may be progressive with extent of disease. To test this hypothesis, anti-VE1 Braf antibodies were used in 10 thin melanomas (range 0.28–0.6 mm) compared to 12 thick melanomas (range 1.35–5.0 mm). Five intradermal nevi with 10 metastatic melanomas and 5 Spitz nevi were used as positive and negative controls respectively. Selected melanomas also had PCR performed. All melanomas were obtained from relatively sun protected sites. The staining was graded from 1 to 3 (I:0–33% of melanocytes stained; II:34–66%; III:67–100%). All cases studied showed grade III staining. In only one of the thin melanomas was there staining (in the epidermis and dermis). In 4 of the thick melanomas, there was staining (3 in the epidermis and dermis, 1 ulcerated in the dermis only). Although the epidermal staining was unusual, this preliminary study suggests a possible change in phenotype as melanomas age and progress.

Galectin-3 Expression in Primary Cutaneous CD30-Positive Lymphoproliferative Disorders and Transformed Mycosis Fungoides

☆WINNER BEST ORAL ABSTRACT AT MEETING☆

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We investigated 11 primary cutaneous anaplastic large cell lymphoma (PCALCL), 47 lymphomatoid papulosis (LYP) and 14 cases of transformed mycosis fungoides with CD30 expression (MF-T) for Galectin-3 expression. A Galectin-3 score was applied using a photo-based morphometric evaluation program. Double staining for CD30 and Galectin-3 was performed. The Galectin-3 expression in CD30 (+) tumour cells was significant lower in MF-T compared to CD30-positive lymphoproliferative disorders (CD30 LPD) ($p < 0.001$), but we found no difference between PCALCL and LYP. Galectin-3 was in PCALCL more often localized in the cytoplasm in contrast to LYP, in which an equal distribution in the cytoplasm and the nucleus was more common. The lower Galectin-3 expression in MF-T compared to CD30 LPD may represent an additional criterion to differentiate both entities. The different sublocalization of Galectin-3 signal might reflect a different biological function.

Lymphoplasmacytic Plaque—A Series of 6 Patients. Diagnostic Approach and Review of the Literature

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We investigated 6 own lymphoplasmacytic plaques (LPP) and 9 cases reported in the literature for their clinical and histologic characteristics and their diagnostic work-up. The objective was to define clinical and histologic criteria of LPP and to develop a diagnostic flow-chart. We could differentiate 3 main histological patterns (superficial band-like only, (deep) dermal only, and mixed). Acanthosis and interface dermatitis are key feature in cases with a superficial band-like or mixed infiltrate. Granulomas and giant cells could be only found in 34% of the cases. The number of plasma cells was variable accounting for 5 to 40% of the infiltrate. The number of blood vessels was increased in the majority of the cases. "Free-floating" collagen bundles surrounded by histiocytes (pseudorosettes) were identified as a new histological feature. An infectious agent was excluded in all cases.

Reappraisal of the Histopathological Features of Bullous Pemphigoid

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Bullous pemphigoid (BP) is the most common autoimmune bullous disease with a diagnosis made based on the clinical manifestation, histopathological findings, results of direct immunofluorescence, and serological test results. Because subepidermal bulla is the hallmark of BP in a biopsy specimen, other histopathological features are often ignored. We retrospectively studied histopathological features of 110 BP cases and found several remarkable features other than subepidermal bulla. Of the 110 BP cases, subepidermal bulla was seen in 98 cases, of which 65 also showed linear necrosis of basal cells, 25 showed intraepidermal bulla, and 26 showed adnexal involvement. Adnexal involvement was confirmed by the presence of floating adnexal epithelium within the bulla in 24 cases. Acantholysis was seen in 20 cases harboring subepidermal bulla, whereas vacuolar alteration was seen in 34 cases with subepidermal bulla and 8 cases without. Although eosinophils were the predominant inflammatory cells within the bulla and also in the dermis and epidermis in most cases, neutrophils predominated in the epidermis in 15 cases and within the bulla in 16 cases.

Palisaded Neutrophilic and Granulomatous Dermatitis/ Interstitial Granulomatous Dermatitis. Report of 5 Cases

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Palisaded neutrophilic and granulomatous dermatitis (PNGD) and interstitial granulomatous dermatitis (IGD) are rare dermatoses, commonly believed to be the same entity or different sides of the same pathology. Both share a wide clinical spectrum, but have some different histopathological features and are associated with rheumatologic and other systemic diseases. We describe 5 cases, 3 of them with features of PNGD and 2 of IGD. Two PNGD cases were associated with uveitis; 1 case of IGD had autoimmune thyroiditis and a direct relationship with a reactive depressive state. Two cases were not associated with any other condition. Patients with PNGD showed indurated and raised erythematous papules and plaques, and patients with IGD displayed erythematous flat plaques. Histopathologically, PNGD cases evidenced a diffuse neutrophilic infiltrate with leukocytoclasia and histiocytes with degenerated collagen, and IGD cases showed an almost exclusive diffuse lympho-histiocytic infiltrate with some collagen degeneration. We demonstrate cases of PNGD and IGD with distinct clinical-pathological entities.

Ischemic Fasciitis: Lessons From 16 Cases

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Ischemic fasciitis (IF), is a pseudosarcoma resulting from repeated pressure and consequent intermittent ischemia of soft tissue. Although common in bedridden patients, activity-related intermittent pressure rather than immobilization was found to be the cause in 7 of 12 cases. The elbow and forearm were most commonly affected (4 cases each). In 75% of cases, a history sufficient to explain lesion development was obtained, usually after biopsy. Preoperative diagnoses included: mass, cyst, and nodule. In no instance was IF predicted clinically or radiologically.

Illustrative Case: An active 61-year-old woman presented to dermatology with a painless 1.0 x 1.5 cm swelling on the extensor surface of her proximal forearm temporally related to pressure from a new Pilates-reformer regime. MRI revealed thickening within the dermal soft tissues of the proximal forearm replacing the normal subcutaneous fat signal. Enhancement in MRI with contrast was read as an inflammatory process. Histologically, fibrous involution of fat with ischemic cell dropout was bordered by atypical fibroblasts and reactive vascularity. This zonation, especially over a bony prominence, should lead to history and radiologic consultation that avoids further surgery.

Cutaneous Metastases, 7 Years Report

Monica Ruiz-Ballon, Mauricio Postigo-MacDowall, Rosario Paz-Castro, and Claudia Mares-Cuadros

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Objective: To review and describe all cases of cutaneous carcinoma metastases during a 7 year period, in our service.

Methods: We retrospectively review all cases with histopathologic diagnosis of metastatic carcinoma from 2007 to 2014. Clinical and pathologic data were collected.

Results: Overall, 17 cases were identified in the database from 8854 primary malignancies (incidence of 0.19%) and 1.62% from all skin cancers. Average age of patients was 58.8 years old. 70.6% of cases were females and 29.4% were males. Clinical morphology: 59% was a nodule, 17% a macule, 12% a papule, 6% an erythematous plaque and 6% a depressed lesion. Primary tumor: 29.4% were from breast, 17.6% from lung; skin, kidney, ovary, cervix and endometrium accounted for 5.88% each and 23.5% were of no known primary.

Conclusions: Our series demonstrate breast as the most common primary for cutaneous metastases, followed by lung; the most common clinical appearance was a nodule. These findings correlates with previously reported cases.

Macular Arteritis Associated With Concurrent HIV and Hepatitis B Infections: Evidence for a Disease Spectrum Association With Cutaneous Polyarteritis Nodosa

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We report the first case of macular arteritis (MA) with concurrent HIV and HBV infections. Of particular interest in MA is the striking discordance between the clinical presentation and histopathological findings, a fact that dermatologists and dermatopathologists should be aware. The case showed typical findings of MA with a predominantly lymphocytic infiltrate and intraluminal thrombosis. Both HIV and HBV have been reported as viral inducers of cutaneous polyarteritis nodosa (CPAN). Their association with MA in this case supports the hypothesis that MA and CPAN represent a single disease spectrum of vasculitides, with MA representing the chronic, lymphocytic and indolent stage, and CPAN, the neutrophilic, acute stage with a risk for systemic progression. Lymphocytic thrombophlic arteritis (LTA), a third, uncommon disease is in between MA and CPAN on a spectrum. Features of this case and other published cases provide strong evidence that there is a single, mild-to-severe, chronic-to-acute disease spectrum of MA-LTA-CPAN.

“DERMATOPATHOLOGY TRAINEE WORLD CUP,” WEDNESDAY 3-18-15, 10:30 AM–12:30 PM

Trapp (T-Cell-Rich Angiomatoid Polypoid Pseudolymphoma) With Atypical Features

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TRAPP is a unique type of T-cell pseudolymphoma. It is commonly encountered in the head and neck. Clinically, it looks-like pyogenic granuloma. Histologically, it is composed of T-cells, with variable number of plasma cells, histiocytes and may be few eosinophils. Prominent vessels lined by plump endothelial cells. T-cells are admixture of CD4 and CD8 cells. Most studies suggested a reactive process since most cases were polyclonal. We present an atypical case of TRAPP in 62-year-old female. She presented with a small reddish papule on the nose for several months. Shave biopsy showed localized dense CD3-positive T-cell infiltrate with equal number of CD4 and CD8 cells. CD20 was negative. Atypical T-cells with scattered mitosis were prominent. CD7 was completely negative. CD30 showed few scattered positive cells. T-cell gene rearrangement showed clonality. There were prominent blood vessels with hobnail-like endothelial cells. Patient underwent blood work up including CBC with differential, blood smear, and flow cytometry. They were within normal range. The patient reported complete healing of the lesion after the shave biopsy and regular follow up confirmed it.

True and False Cytokeratin Immunoreactivity in Sentinel Lymph Nodes (SLNs) Resected From Merkel Cell Carcinoma (MCC)

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Background: Detection of MCC in SLNs often requires cytokeratin immunohistochemistry (IHC). We evaluated a range of cytokeratins to optimize detection of MCC SLN metastases.

Table 1. Immunoreactivity comparison (%)

	CK20	OSCAR	CAM5.2	AE1/AE3
Sensitivity	82	100	100	89
Golgi pattern	83	74	67	76
Cytoplasmic Pattern	0	4	0	0
Mixed Pattern	17	22	33	24
Focal	9	0	4	12

Design: Twenty-eight MCC metastatic SLNs and 15 MCC negative SLNs form the study set. IHC slides for CK20, CAM 5.2, AE1/AE3, OSCAR were scored by 2 pathologists for reactivity distribution and pattern. For comparison 6 melanoma SLNs were stained with CAM5.2 and OSCAR.

Results: 1. All cytokeratins had mixed patterns with Golgi predominating (75%) (Table 1). 2. OSCAR and Cam5.2 were easily detected, but exhibited anomalous reactivity in negative MCC SLNs (6/15) and melanoma SLNs (4/6). 3. OSCAR and Cam5.2 detected MCC in 2 SLNs negative by CK20 and AE1/AE3.

Conclusions: Cytokeratins OSCAR, Cam5.2, and AE1/AE3 are more sensitive for detecting MCC in SLNs than CK2. Anomalous expression of OSCAR and Cam5.2 requires interpreting with caution.

Pineal Gland Mass and A Skin Nodule

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A 47 year old female presented to the ER reporting a 1 week history of dizziness, headache, nausea, vomiting, and memory loss. A head CT discovered a hyperattenuating, well-circumscribed 1.7 cm pineal region mass. Initial differential included a primary pineal parenchymal neoplasm, a tentorial meningioma, or a germ cell tumor but the tumor was not found by Neurosurgery to involve the pineal gland and exhibited poorly differentiated cells. An extensive small round blue cell differential existed but was narrowed to lymphoma, Ewing's sarcoma, small cell carcinoma, and melanoma. Immunohistochemistry was positive for vimentin, CD99, and focal p53, but negative for PLAP, EMA, chromogranin/synaptophysin, GFAP, CAM 5.2, IDH-1, and NFP. Follow-up IHC was negative for Lu-5, but positive for NSE, MART-1, MiTF, HMB-45, and S-100. Primary CNS melanoma was considered but a full body skin exam revealed a large 3 × 4 cm exophytic violaceous firm fixed lesion on her right lower knee. Similar staining was noted but the lesion could not be confirmed as the primary lesion. This presentation will discuss primary CNS melanomas, secondary metastatic disease to the brain, as well as Melanoma of Unknown Primary.

NCOA2 Immunohistochemistry in Cutaneous Indeterminate Cell Histiocytosis Versus Langerhans Cell Histiocytosis

✧THIRD PLACE WINNER, DERMATOPATHOLOGY TRAINEE WORLD CUP COMPETITION✧

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Diagnosis of Langerhans cell histiocytosis (LCH) relies on langerin immunohistochemistry (IHC) or electron microscopy to assess for Birbeck granules, which are specific to LCH. When these studies are negative, the diagnosis of indeterminate cell histiocytosis (ICH) is possible. In light of the absence of diagnostic tools with positive predictive value for ICH, however, this entity remains controversial. Alteration of *NCOA2*, which encodes

a transcription factor involved in fat homeostasis, has been noted in at least 1 case of ICH. We evaluated NCOA2 IHC in 15 cases of cutaneous histiocytosis. All ICH (3) and juvenile xanthogranuloma (JXG, 3) cases demonstrated nuclear positivity for NCOA2, while all other histiocytoses (5 LCH, 2 Rosai-Dorfman, 2 histiocytosis not otherwise specified) were negative. The extent of giant cells and localized presentations distinguished cases of JXG from ICH. Cases of LCH and ICH showed essentially identical histology. In this collection of cases, NCOA2 was a useful tool for separating ICH and LCH.

Elizabethkingia Meningosepticum: Case Series Reveals an Under-Recognized Cause of Cutaneous Infection

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Objective: Discuss an under-recognized and potentially fatal cutaneous pathogen.

Background: *Elizabethkingia meningosepticum* is a ubiquitous soil and water-dwelling Gram-negative bacillus. It is a known cause of neonatal meningitis and nosocomial mini-epidemics. Infections have a high mortality rate due to polymicrobial resistance.

Methods: An updated review of our institution's electronic medical record for the years 1997-2014 revealed 25 cases of *E. meningosepticum*.

Results: Twenty-four percent of cases were cutaneous infections. In addition, 56% of all infected patients and 76% of infected adults had a cutaneous trauma at time of infection. The mean age of patients infected with *E. meningosepticum* was 30.4 years including 17 adults and 8 children. The bacterium was also isolated from non-cutaneous sites including blood, CSF, sputum, bronchoalveolar lavage and a catheter tip. Review of the current literature reveals that adult infection is considered rare, and reports of the organism causing cutaneous infections are quite limited.

Conclusion: Our findings suggest that *E. meningosepticum* is a more significant pathogen in cutaneous infections in the adult population than what is supported in the current literature.

Histologic Variation in Cellular Neurothekeoma: A Morphologic Spectrum of an "Uncommon" Cutaneous Tumor

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Cellular neurothekeoma (NTK) is a relatively uncommon, dermally-based tumor that usually occurs in the head/neck region of children and young adults. There is a female predilection and lesions typically present as one-to-multiple sub-centimeter papules. NTK follows a benign clinical course and, when fully excised, rarely recurs. Cellular NTK exhibits a lobular proliferation of spindled-to-plump "epithelioid" cells confined to the dermis upon a variably hyalinized stroma. Their immunohistochemical profile is fairly distinct and often essential in rendering a correct diagnosis. NKI/C3 is always positive and the majority of cases express PGP9.5, NSE, and at least partial MITF. Additional useful markers include S100A6, SMA, factor XIIIa, PG-M1, and CD68. S100 is always negative. Cellular NTK may display a number of histologic appearances that may prove diagnostically challenging. Furthermore, the differential diagnosis includes tumors of melanocytic, fibrohistiocytic, and neural origin. Knowledge of the morphologic spectrum of NTK is necessary to avoid potential pitfalls. We present several examples of cellular NTK from our institutional experience including those showing granular cell, chondroid, and fibrohistiocytic features.

Are Genital High Grade Squamous Intraepithelial Lesions (HSIL) Suspected Prior to Biopsy by Dermatologists in Men?

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Objective: Clinicians often submit genital lesions, with benign pre-biopsy diagnoses which are histologically diagnosed as HSIL. The objective is to determine how frequently clinicians consider HSIL in the pre-biopsy diagnosis.

Methods: A retrospective record review is performed of cases diagnosed as genital HSIL in men at a National Dermatopathology lab between 1/1/2008 and 1/1/2014. Since HSIL is relatively new terminology advised by the Lower Anogenital Squamous Terminology consensus conference (LAST), cases are identified with diagnoses of HSIL, Bowen's disease, Bowenoid papulosis and squamous cell carcinoma in situ. The records are examined to identify pre biopsy diagnoses, history of skin cancer and HPV infection.

Results: One hundred and fifty-six specimens from 148 patients are identified. In 53% of specimens there was no documented suspicion of malignancy (HSIL, SCC, basal cell carcinoma or melanoma). In 28% of specimens, the pre-biopsy diagnoses were limited to clinically pigmented lesions (seborrheic keratosis, lentigo, dysplastic nevus or melanoma). In 40% of specimens, a pigmented lesion was listed in the differential diagnosis.

Conclusions: Dermatologists often fail to consider the diagnosis of HSIL in male genital lesions. HSIL should be considered in the differential diagnosis of all genital lesions in men.

Boggy Scalp and Hair Loss: A Case of Lipedematous Alopecia

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A 51-year-old Caucasian female presented with a 1-year-history of scalp pruritus which was accompanied by thickness of the scalp and gradual hair loss. The patient had no significant past medical history and had not recent illnesses. Family history was unremarkable. Physical examination revealed patchy areas of alopecia most prominent along the vertex scalp. The scalp was mildly tender on palpation and had a boggy consistency. Histologic sections revealed hyperkeratosis and mild acanthosis. A scant mixed superficial perivascular, and perifollicular infiltrate was present. No significant dermal or perifollicular fibrosis was detected. There was prominent subcutaneous adipose tissue extending into the dermis and around hair. Based on the clinical and pathologic findings patient was diagnosed with lipedematous alopecia (LA). LA is a rare disease of unknown etiology characterized by thick, boggy scalp with varying degrees of hair loss. Although initially described to occur in Afro-American patients, several reports on Asian and Caucasian patients have lessened the role of racial factors in the pathogenesis of this disease. We report a case of LA in a Caucasian patient who was initially diagnosed with "scarring alopecia." The clinical and histologic features for this rare entity are described.

Diagnostic Pitfalls in Localized Massive Lymphedema

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Localized massive lymphedema is a rare pseudoneoplasm that occurs in morbidly obese patients and can mimic a well-differentiated liposarcoma (WDL). It develops due to obstruction of subcutaneous lymphatics due to large, dependent skin folds and occurs on the abdomen or medial lower extremities. We present the case of a 42-year-old morbidly obese man who presented with a rapidly enlarging tumor on the left upper thigh. Excision of a 25 pound mass was performed. Histopathology showed massive expansion of thickened, subcutaneous fibrous septae by edema fluid, foci of atypical fibroblasts, and reactive capillary proliferations on the periphery of fat lobules. Typical features of WDL including lymphoid aggregates and foci of atypical cells with multilobated, hyperchromatic nuclei were not seen. Fluorescent in situ hybridization testing for MDM2 (12q15) did not demonstrate amplification. Although the clinical and histopathologic findings in this case are classic, the diagnosis was not considered by the referring physicians, which in our experience is not uncommon and reflects a general

lack of awareness of this rare entity. Thus, we report this case to raise awareness and to help prevent misdiagnosis as WDL.

Atypical Apocrine Tumor of Anogenital Mammary-Like Glands Presenting in Cowden Syndrome: A Novel Association?

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Purpose: Anogenital mammary-like glands (AMG) are appendages of anogenital skin. Rarely, they transform into epithelial and stromal tumors that morphologically resemble breast tumors. Cowden Syndrome (CS) is an autosomal-dominant (PTEN gene) cancer-susceptibility syndrome that potentially leads to skin and breast cancers. A benign proliferative AMG lesion in the setting of CS is described here for the first time.

Methods: Twenty-seven-year-old female with CS (R130Q-PTEN mutation) presented with a tender, 1-cm polypoid perianal lesion. A full thickness excision was performed.

Results: Grossly, the cut surface showed a lobulated lesion with glistening tan-pink surface. Microscopy revealed a circumscribed, lobulated, proliferative lesion with papillary, micropapillary, and focal cribriform architecture that associated with a fibromyxoid stroma. This strikingly resembled the breast fibroepithelial lesions. Immunostaining was positive for ER, PR, GCDPF-15 and GATA3.

Conclusions: As CS carries a known risk for breast carcinoma, this case raises the question of increased risk for perianal malignancies arising in AMG in CS setting. Association of proliferative AMG pathogenesis and PTEN mutation needs further investigation.

Overlap Porphyria Cutanea Tarda and Lupus: A Report of 2 Cases

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We present 2 patients with co-existing porphyria cutanea tarda (PCT) and lupus. The first, a 63 year old woman on hemodialysis for polycystic kidney disease, initially presented with a blistering dorsal hand eruption. Skin biopsy showed thickened blood vessels Urineporphyrins were elevated, consistent with PCT. She was also noted to have a macular telangiectatic eruption on the chest, which triggered workup for connective tissue disease. Skin biopsy showed vacuolar interface dermatitis and blood tests found, elevated ANA, and elevated anti-Ro antibodies, consistent with an overlapping diagnosis of subacute cutaneous lupus. A 52 year old patient with known systemic lupus erythematosus developed photodistributed bulla on the upper extremities. Skin biopsy from the hand showed both subepidermal bulla and vacuolar interface dermatitis with dermal perivascular neutrophilic inflammation. Urine porphyrins and ferritin were elevated at the time of the eruption. Skin blistering resolved after therapeutic phlebotomy. Though rare, the overlap of PCT and lupus should be considered in patients with suggestive clinical features to ensure optimal treatment.

Histologic Features of Non-Melanoma Skin Lesions Treated With Imiquimod

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Object: Describe the histologic features of skin biopsies performed on lesions treated with topical imiquimod.

Methods: This is a case series in which a retrospective electronic record review was performed utilizing "imiquimod" as the search term. Our search identified 5 non-melanoma cases meeting criteria.

Results: Inflammatory responses included dense, often lichenoid type lymphoid infiltrates. One case demonstrated atypical lymphocytes. Epidermal

changes included dyskeratotic keratinocytes and vacuolar interface alteration. Residual lesions were not identified in any biopsies.

Conclusions: Topical imiquimod is a dermatologic agent indicated to treat genital warts, actinic keratoses and superficial basal cell carcinomas. Lesions treated with imiquimod can pose clinical diagnostic challenges as erythema and ulceration are not uncommon responses thereby making it difficult to ascertain whether a lesion has been adequately treated. Biopsies are therefore performed to assure adequate treatment. While although the histologic findings of lesions treated with imiquimod could lead to further diagnostic uncertainty, as both the epidermal and inflammatory response can mimic malignant entities, in our series, pathologists were able to discern whether or not residual lesions remained in all cases.

Disseminated Histoplasmosis in a Traveler: A Potential Diagnostic Pitfall

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A 33 year-old male veteran presented to a hospital with a 4-week history of diarrhea and fever after returning from Iraq. Clinical evaluation revealed a new diagnosis of HIV-AIDS with a CD4 count of 21. He was subsequently started on HAART. One week later, he developed an asymptomatic erythematous maculopapular eruption involving the face, trunk, and upper and lower extremities. The clinical differential diagnosis included drug eruption, hemophagocytic lymphocytosis syndrome, Kaposi's sarcoma, and viral exanthem. A punch biopsy from the right arm demonstrated abundant histiocytes in the superficial dermis containing intracellular PAS and GMS-positive microorganisms. A bone marrow biopsy revealed numerous macrophages with similar intracytoplasmic microorganisms. The histopathologic differential diagnosis included disseminated histoplasmosis and leishmaniasis. The organisms appeared to lack kinetoplasts under light microscopy. Immunohistochemical staining for *Histoplasma capsulatum* was positive and PCR testing for *Leishmania spp.*, was negative. Tissue fungal cultures grew *H. capsulatum*, and urine Histoplasma antigen was positive, further supporting the diagnosis. Our case highlights the value of synergistic histologic, microbiological, and molecular assessment in distinguishing between histoplasmosis and leishmaniasis. These entities may be difficult to differentiate in an immunocompromised patient with a significant travel history.

Malignant Melanoma Growth Requires CD98 Expression

☆FIRST PLACE WINNER, DERMATOPATHOLOGY TRAINEE WORLD CUP COMPETITION☆

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Background: Malignant melanomas are a heterogeneous group of malignancy commonly studied using assays to detect DNA sequence alterations.

Objective: We sought to identify alterations in protein levels that are hallmarks of malignant melanomas. We focused on identifying cell-membrane proteins that could represent new targets for molecular therapy. We chose to study CD98, an essential amino acid carrier composed on heavy chain (CD98hc, i.e. SLC3A2) and corresponding light chain (CD98lc, i.e. SLC7A5), and evaluate whether this marker is upregulated on the cell surface of melanomas.

Methods: We evaluated 5 cases of benign melanocytic nevi, 4 cases of atypical melanocytic nevi, and 50 cases of malignant melanoma for CD98 expression. Cases were scored for staining intensity ranging from 0 (negative staining) to 3+ (diffuse strong staining).

Results: CD98 was markedly upregulated on the cell surface of 70% (35/50) of malignant melanomas, often at high levels. CD98 was completely negative in all 5 benign melanocytic nevi and was detected at low levels in 3/4 atypical nevi. In addition, we demonstrated that CD98 was required for in vitro proliferation of several melanoma cell lines.

Conclusion: CD98 protein levels are upregulated in melanomas. CD98 could potentially be used as a diagnostic marker in ambiguous melanocytic lesion, in which the diagnosis of melanoma is entertained. Furthermore, melanomas expressing this marker could be subjected to targeted therapy with available anti-CD98 agents.

A Rare Case of Axillary Sweat Gland Carcinoma With Osteosarcomatous Transformation

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Primary cutaneous apocrine sweat gland carcinomas are rare tumors which most commonly involve the axillae. Some are indolent, while others may show aggressive behavior and metastatic spread, with the lymph nodes the most common site of metastasis. Complete surgical excision is the treatment of choice. Various histologic patterns have been reported, but mesenchymal transformation has not been previously described in these tumors. A 63-year-old man presented with painful, acute enlargement of a previously stable and asymptomatic pea-sized nodule in the left axilla with overlying skin changes. Histopathologic sections demonstrated a poorly differentiated primary sweat gland carcinoma with prominent areas of sarcomatoid transformation, composed primarily of osteosarcomatous heterologous differentiation. Nested and cystic areas contained epithelial cells with strong, diffuse CK7 immunoreactivity. Molecular analysis was consistent with an osteosarcoma. This case highlights a previously unreported histologic variant of axillary sweat gland carcinoma and emphasizes the importance of correlating clinical and histopathologic findings within the context of abnormal or unexpected molecular results.

Correlation of P21 Expression in Head and Neck Squamous Cell Carcinoma With Clinicopathologic and Prognostic Parameters

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Purpose: p21 is closely related predictive marker, having bad prognostic effect in Head and Neck squamous cell carcinoma (HNSCC). The purpose of my study is to determine the expression of p21 in HNSCC with various parameters.

Materials and Methods: One hundred and ten patients of HNSCC were included in the study. p21 expression was noted and correlated with various parameters.

Results: Out of 110 cases, p21 positive cases were 88 (80%) while p21 negative were 22 (20%). Nodal metastasis was seen in 51 cases (57.95%) with positive p21 expression as compared to 4 cases (18.8%) with negative p21 expression (p value = 0.04). Thirty-nine cases (44.31%) showed recurrence with p21 positive carcinomas while 3 cases (13.63%) showed recurrence with p21 negative carcinomas (p value = 0.08). Two year survival rate was 56.81% (n = 88) with p21 positive cases while p21 negative cases showed 90.90% (n = 22) 2 year survival rate (p value = 0.20).

Conclusion: Positive p21 expression in Head and neck squamous cell carcinomas is a strong bad prognostic factor and correlates with increased nodal metastasis, tumor recurrence and worse overall survival.

Going to Your Head: The Distribution of Cutaneous Metastases Is Predicted by Tissue Treg Density

☆SECOND PLACE WINNER, DERMATOPATHOLOGY TRAINEE WORLD CUP COMPETITION☆

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The distribution of cutaneous metastases is nonrandom, and favored sites, such as the scalp, are well-known. The determinants of this distribution, however, are poorly understood. Given that immunosurveillance influences tumor progression, we hypothesized that metastasis to a particular site might depend on the permissiveness of the immunologic microenvironment at that site, and, specifically, on the local density of regulatory T-cells (Tregs). We surveyed all cutaneous metastases evaluated at our institution from 1991 through 2014 (n = 1984) and calculated the frequency of metastasis to individual sites, controlling for regional surface area. We measured the density of tissue-resident CD3+FoxP3+ Tregs by flow cytometry using a separate cohort of mapped, normal skin samples (n = 140). We found that metastases were most common on the head and neck, especially on the postauricular scalp, with relatively fewer on the trunk, followed by the upper and lower extremities, respectively. The density of Tregs followed the same distribution, with highest levels on the head and neck and lowest on the lower extremities (p for trend <0.05). We propose that regional variations in Treg density may account for the distribution of cutaneous metastases.

Squamous Eccrine Metaplasia With Neurotropism: A Potential Overdiagnosis of Aggressive Squamous Cell Carcinoma

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We present a case of a 68-year-old male with biopsy-proven BCC of the left leg. A follow-up excision specimen showed nested basal cells attached to the epidermis with peripheral palisading and clefting, confirming the diagnosis. An irregular area of squamitized eccrine units and epidermal hyperplasia was associated with the biopsy site. This dermal squamous population surrounded a deep dermal nerve, raising the possibility of a second carcinoma. Nonetheless, the nuclei were cytologically bland and the nests had jagged contours characteristic of reactive biopsy site changes. The area of neurotropism was related to other squamitized eccrine ducts. This case differs from epithelial sheath neuroma because it involves an anatomically-normal deep dermal nerve and represents a focal finding associated with typical squamous eccrine metaplasia and reactive epidermal hyperplasia at a biopsy site.

Benign mimics of SCC exist and are potential diagnostic pitfalls. These entities include: pseudoepitheliomatous (pseudocarcinomatous) hyperplasia, inverted follicular keratosis, and, as reported herein, squamous eccrine metaplasia with neurotropism. Recognizing these patterns, applying appropriate diagnostic criteria, and maintaining close clinicopathologic correlation can prevent misdiagnosis.

Phimosis With Incidental Lichen Sclerosus Et Atrophicus: A Histopathologic Mimic of Mycosis Fungoides

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Background: Lichen sclerosus et atrophicus (LSA) in males characteristically involves the glans and foreskin, often resulting in acquired phimosis. LSA may not be clinically apparent, but has been identified histologically in over 60% of circumcision specimens from patients with phimosis. Histologic findings may be non-specific in early lesions, potentially simulating other inflammatory dermatoses. This inflammatory stage of LSA has been reported to simulate mycosis fungoides (MF). This histologic presentation is less well known in foreskin specimens. A case of a circumcision specimen showing histologic changes simulating MF due to subclinical LSA is presented, and the histologic features of phimosis are reviewed.

Case: A foreskin specimen was submitted for histologic review, with no initial clinical history. Histologically, there was psoriasiform epidermal hyperplasia without significant spongiosis and with epidermotropism of small, but irregular lymphocytes with perinuclear halos. Some of these were aligned along the basal layer. There was papillary dermal sclerosis. The patient had no known clinical rash.

Conclusion: Phimosis may have subclinical LSA and should be included in the list of MF simulators.

Majocchi's Granuloma May Be Missed Due to Negative Epidermal Fungal Stains

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Background/Objective: The diagnosis of Majocchi's granuloma (MG), typically confirmed by fungal stains, may be missed if fungi are absent in the stratum corneum. Our objective was to assess the frequency of fungal stain negativity in the stratum corneum of (MG) and histologic features.

Methods: Thirty-five cases of MG were obtained from the files of the Ackerman Academy of Dermatopathology from January 2012 to August 2014. Epidermal and dermal changes were analyzed on PAS and routine histology.

Results: 15/35 had identifiable hyphae in the epidermis. Epidermal changes included compact stratum corneum, parakeratosis and variable spongiosis. Dermal changes of vessel congestion, extravasation of erythrocytes, edema, and follicular spongiosis were common. Eosinophils, often admixed with neutrophils, were present in most of the sections. Some biopsies showed overlapping features with dermal hypersensitivity.

Conclusion: A diagnosis of MG may be missed due to the absence of fungus in the epidermis, especially if a hair is not present in the sections. Perivascular

mixed inflammation, compact hyperkeratosis, congestion, and purpura should prompt one to perform levels to assess for a follicle.

Mycosis Fungoides: When the Clinical Course Doesn't Fit

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A 68 year-old woman with a history of mycosis fungoides presented with progressive and painful ulcerations. Biopsies in 2011 showed an atypical dermal lymphoid infiltrate with epidermotropism and pilotropism that showed immunoreactivity to CD3, CD4, and CD7, and lacked expression for CD2, CD5, CD8, and TCR-Beta-F1. Although this pattern of immunostaining was atypical, she was diagnosed with folliculotropic mycosis fungoides. She trialed topical steroids, narrow band UVB, local radiation, methotrexate, chlorambucil, and bexarotene, yet continued to progress. Biopsy of a shoulder ulcer performed at our institution in 2014 showed a dermal infiltrate of atypical lymphoid cells with immunostaining demonstrating expression of CD3, without expression of CD4, CD5, CD7, CD8, or TCR-Beta-F1. TCR gamma-delta stain was positive. Systemic evaluation remains negative for lymphoma. This pattern supports a diagnosis of primary cutaneous gamma-delta T-cell lymphoma. In this case, lack of response to skin directed and systemic therapies prompted rebiopsy and the eventual diagnosis of primary cutaneous gamma-delta T-cell lymphoma. This demonstrates the need for careful immunophenotypic evaluation and the utility of the gamma-delta immunostain to identify this rare and aggressive cutaneous lymphoma.