

Poster 655

Focal dermal elastosis: a case report of a rare pseudoxanthoma elasticum-like entity

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We report the case of a 54-year-old Caucasian female with a five-year history asymptomatic papules. There was no family history of similar lesions. Physical examination revealed greater than 100, 2-4 mm, firm, yellow, dermal papules located on the neck, antecubital and popliteal fossa, flexor surface of both forearms, and inner thighs. Skin biopsy showed a focal increase in the concentration of elastic fibers. There was no fragmentation, calcification, or phagocytosis of elastic fibers. We rendered a diagnosis of late-onset dermal elastosis. Late-onset dermal elastosis is a rare entity with only a few cases reported in the literature. Histopathologically, biopsies of late-onset dermal elastosis show a focal increase in normal-appearing elastic fibers. Tissue lacks evidence of calcification and elastolytic change distinguishing this entity from pseudoxanthoma elasticum and elastolytic entities. We believe that awareness of this entity is important as it can clinically simulate other processes associated with systemic manifestations.

Poster 656

RESIDENT

Sea urchin spine deposition: histopathology and review of the literature

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Background Sea urchins belong to one of the five classes of echinoderms. Their skeleton is composed of ten fused plates that encircle the body, with long, movable spines that reach up to 30 cm long and 1 cm thick. These spines can induce clinically significant foreign body reactions upon contact with the skin. Previous studies published in the biology and materials science literature have confirmed that the spines are large, porous, biogenic single crystals of magnesium-rich calcite with three-dimensional meshwork architecture. To our knowledge, this represents the first pathologic documentation of the structure and morphology of sea urchin spines as they appear on hematoxylin and eosin (H&E) staining. Observations A healthy 57-year-old man presented after surfing in the Caribbean with several skin-colored, mildly tender papules on the left palm, as well as an erythematous papule overlying a foreign body on the right second toe. A biopsy from the left wrist demonstrated granulomatous inflammation with foci of central fibrinous debris. A biopsy from the papule on the right toe showed stratum corneum with scale-crust and fragments of a polarizable mineralized foreign material arranged in a lattice-like configuration. The gross specimen consisted of a firm brown spine with

ridged edges and a somewhat blunt tip. Conclusions The historical, clinical, gross, and histologic findings in our patient confirm the diagnosis of a granulomatous foreign body reaction to sea urchin spines. To our knowledge, these findings represent the first dermatopathologic description of sea urchin spine as seen on H&E staining to be presented in the dermatopathology literature.

Poster 657

RESIDENT

Granulomatous and fibrotic reaction to expanded polytetrafluoroethylene used as injectable filler

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A 53 year old female presents with intermittent lip swelling over the past 25 years which had worsened over the previous three to four weeks. Her past medical history is significant for multiple injections of expanded polytetrafluoroethylene (ePTFE) suspended in saline for lip augmentation 25 years ago. Physical examination revealed symmetrical swelling of the upper and lower lips with vague palpable nodularity. Biopsy showed superficial and deep dermal granulomatous inflammation containing polarizable material and significant hyalinized fibrosis. The findings were consistent with foreign body granuloma secondary to ePTFE. The patient showed significant improvement with oral prednisone, however the granulomas did not completely resolve and even worsened when a prednisone taper was attempted. Expanded polytetrafluoroethylene is a hydrophobic material composed of carbon and fluorine molecules, which has widely been used for graft material in endovascular and urogynecological surgeries and as implant material in reconstructive surgeries. Its popularity is largely due to the low risk of infection and biological inertness with no reports of bioincompatibility since its introduction in 1971. The process of breaking down ePTFE into particles and suspending it in saline to use as an injectable filler for lip augmentation is not currently approved and its use in this form has not previously been documented in the literature. This case is unique in that it demonstrates a use of ePTFE not formally reported and to our knowledge is the first report of bioincompatibility in the form of foreign body granuloma and dense fibrosis to ePTFE.

Poster 658

FELLOW

A visible reaction to an invisible tattoo

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Invisible (or blacklight) tattoos are fast becoming the trend in the world of tattoo art, and with their rise comes

the onset of associated complications. Though there have been many reports of cutaneous reactions to traditional tattoo pigments, literature regarding reactions to invisible tattoos is scarce. We report the case of a 28-year-old man who presented with an inflammatory eruption of 2 months' duration confined to the area of a recently placed invisible tattoo; the eruption was diagnosed as granulomatous dermatitis to a foreign material. Under fluorescent light, a refractile foreign material was identified in the biopsy specimen, which we believe to be melamine, one of the invisible tattoo's five ingredients. Previous cases of cutaneous reactions to invisible tattoos were attributed to polymethylmethacrylate, not a component of the tattoo in this case. To our knowledge, this is the first case implicating melamine as the cause of a granulomatous tattoo reaction. Given the rising popularity of invisible tattoos, we present this case to raise awareness of the risks associated with this alternative tattoo trend.

Poster 659

FELLOW

Pleomorphic congenital ichthyosis with superimposed fungal colonization

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We present a rare case of a self-limiting pleomorphic ichthyosis in a neonate with superimposed fungal colonization. The patient, a 32-week gestational age female neonate, presented with a diffuse epidermal scaling at birth. The patient was born via spontaneous vaginal delivery, complicated by preterm premature rupture of membranes. Upon delivery, the patient was noted to have a diffuse, fine epidermal scaling with superficial fissures involving the trunk, extremities, palms and soles. Microscopic examination revealed a mild compact hyperkeratosis with follicular plugging, retention of the granular layer, and absence of inflammation. Hyphae, but not yeast, were present in the stratum corneum, in follicular ostia and also identified on a skin scraping specimen. The patient's symptoms improved significantly within one week. Only minimal residual scaling of the palms remained. The term, congenital ichthyosis, is applied to a heterogenous group of genetic disorders of abnormal keratinization present at or within a few weeks of birth. The classic phenotypes are "Harlequin foetus", lamellar, and congenital ichthyosiform erythroderma/ erythrodermic ichthyosis. The term pleomorphic ichthyosis has been coined to classify diseases that fall in the spectrum of congenital ichthyosis but are milder, even self-improving, and overlap with the three classical types. A unique feature of this entity is the rapid clearing of the lesions over a span of weeks to months, with only localized residual scaling. Cutaneous fungal colonization may be present in neonates and seems inversely proportional to gestational age. Detection of superficial fungal colonization could be over interpreted and mask a congenital ichthyosis.

Poster 660

RESIDENT

Multiple familial trichoepitheliomas: a rare genodermatosis

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A nine-year old girl presented with approximately 20 tan-white, shiny papules on her nose, cheeks, and forehead. Some lesions demonstrated central umbilication. A clinical diagnosis of molluscum contagiosum was made, and the patient was treated with topical imiquimod, cimetidine, and liquid nitrogen ablation. The patient continued to develop additional papules. Multiple lesions were biopsied, and histopathologic exam of each lesion revealed a thinly stranded basaloid proliferation with peripheral palisading, horn cysts, and a prominent connective tissue sheath with occasional papillary mesenchymal bodies. These features were diagnostic of trichoepithelioma. Further questioning revealed that the patient's father had developed in childhood multiple facial lesions which reportedly resolved before adulthood. A diagnosis of multiple familial trichoepitheliomas (MFT) was made. MFT exists on a spectrum of rare adnexal tumor syndromes associated with mutations of the tumor suppressor gene CYLD. This case demonstrates that the syndrome may mimic molluscum contagiosum clinically and also illustrates the importance of biopsy for definitive tumor diagnosis and elicitation of family history for clues to this hereditary disease.

Poster 661

FELLOW

Unilateral, linear, zosteriform Darier disease in a blaschkoid distribution masquerading as varicella zoster

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Darier disease (DD), an autosomal dominant genodermatosis caused by a mutation of the ATP2A2 gene at 12q23-24.1, typically manifests with keratotic crusted red-brown papules in a bilateral seborrheic distribution. The lesions of the much rarer unilateral, linear variant are confined to a limited area and often lack typical features such as nail changes and a family history. A blaschkoid distribution of lesions, unilaterally, on a background of normal skin, reflects heterozygosity for a postzygotic somatic mutation during early embryogenesis. A consequence of possible gonadal mosaicism is transmission of the diffuse, non-segmental form of DD to the next generation. A 53-year-old Caucasian male hospital inpatient was referred to Dermatology for suspected varicella zoster. He reported a left-sided skin eruption, first noted 32 years ago, during military service, which waxed and waned, without resolution. No family members had similar conditions. The eruption was pruritic, but not

painful. Examination revealed red-brown, mildly scaly, blaschkoid plaques studded with red-brown, keratotic micro-papules and monomorphic vesicles over the left mid abdomen, left upper shoulder and left lower back. The distribution was blaschkoid rather than dermatomal. A punch biopsy revealed suprabasilar acantholysis with dyskeratosis, consistent with linear DD. In situ hybridization for HSV 1 and 2 and VZV was negative. Treatment with 0.1% tretinoin cream daily to the area was started. This case illustrates how a rare entity may inadvertently masquerade as a more common disease process, if unrecognized. In this instance, the correct diagnosis avoided inappropriate antivirals and isolation while providing the opportunity for meaningful therapy and genetic counseling.

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Reassigned to Poster Session 1

Poster 663

RESIDENT

Acantholytic dyskeratosis: a possible mosaicism in segmental Darier's disease

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A 33-year-old female presented for a routine skin evaluation. Incidental linear, brown, scaly, papules were identified on her right abdomen, present for approximately 13 years. No other skin lesions were detected, and her nails and oral cavity appeared normal. She had no past medical history, family history, or prior treatment for this condition. Clinically, the differential diagnosis included segmental Darier's disease and an epidermal nevus. A biopsy revealed acanthosis, hyperkeratosis, mild papillomatosis and acantholytic dyskeratosis with acantholysis resulting in suprabasilar clefting with corps ronds and rare grains. The histologic differential diagnosis included Darier's disease, Grover's disease, Hailey-Hailey disease, acantholytic acanthoma and an acantholytic dyskeratotic epidermal nevus. A diagnosis required clinical-pathologic correlation. Grover's disease is symmetrically distributed on the chest and back, most prevalent in middle-age men and often exacerbated by heat and sweating, in a remitting-recurring pattern. Hailey-Hailey disease presents as macerated scaly plaques in intertriginous areas. An acantholytic acanthoma is a solitary papule. An acantholytic dyskeratotic epidermal nevus is congenital or present since early childhood. It would be unusual to present after puberty although this is a typical manifestation for segmental Darier disease, the likely diagnosis. The segmental nature of this patient's lesions may explain the absence of nail changes or family history. Genetic testing for ATP2A2 Darier gene mutations has low sensitivity in skin specimens. Gene mutation analysis of offspring lymphocytes may assess the risk of living children. Genetic counseling should be offered to patients with suspected mosaicism of the ATP2A2 for possible generalized Darier phenotype in offspring.

Poster 664

Frozen section pathology diagnosis for non-melanoma skin cancers-correlation with permanent section pathology diagnosis

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Frozen section (FS) pathology is routinely used for margin assessment of basal cell carcinoma (BCC), squamous cell carcinoma (SCC) and melanocytic proliferations including melanoma (lentigo maligna). FS can also be used for the primary diagnosis of several skin lesions. The aim of this study was to determine the accuracy of frozen section pathology diagnosis of non-melanoma skin cancers including actinic keratoses, squamous cell carcinoma and basal cell carcinoma, by assessing the degree of diagnostic correlation between frozen section and permanent section pathology for NMSCs. We performed a retrospective chart review of 300 cases in which frozen section diagnoses were compared with permanent section diagnoses for non-melanoma skin cancers (NMSCs). Our results indicated that frozen section and permanent section pathology were in agreement 83.3% of the time (95% CI, 79.1 - 87.6%), with the highest concordance rates for BCC. Unbalanced sampling for tumor in tissue submitted for FS may account for most of the discordance between frozen and permanent sections. In our practice, frozen sections can be processed and slides generated for review within 2 hours, in comparison to permanent section processing which takes on average, 1 - 2 days to generate a final report. The rapidity of FS tissue processing and pathology reporting makes this technique useful in dermatologic practice for immediate diagnosis and management of NMSCs. This is especially beneficial for patients as the use of frozen section pathology for diagnosis allows for expedited treatment of skin lesions, and reduces the anxiety associated with waiting for permanent section diagnoses.

Poster 665

Evolution of dermatopathology illustrated through successive editions of a classic text

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The 50th Anniversary of the American Society of Dermatopathology (ASDP) offers an opportunity to assess the growth and development of dermatopathology in the United States. Lever's Histopathology of the Skin has been released in ten editions from 1949 until present day. The initial edition is a small and compact publication whereas the most recent edition is replete with color images, on line resources, and discussions of many disease entities not

recognized in the initial edition. This Poster presentation compares the content, presentation, and topics in successive editions of this classic text in an effort to portray changes in the practice of dermatopathology. The introduction of color images, immunohistochemistry, and other notable developments will be presented in a timeline format. The viewer will have the opportunity to appreciate the remarkable growth and dynamic development of dermatopathology over the last half century as we celebrate the Golden Anniversary of the ASDP.

Poster 666

Beauty is just skin deep: second annual dermatopathology and cutaneous immunopathology art competition

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Images in dermatopathology not only are important in diagnosing skin disease for patients but also have inherent aesthetic value. We hosted the Second Annual Dermatopathology and Cutaneous Immunopathology Art Competition to promote appreciation for the beauty of microscopic images of the skin. Entries were judged based on their aesthetic appeal, originality, and potential educational value. Eleven photomicrographs were entered into the competition. The winning entry, entitled, "In Honor of All Those Diagnoses That Seem So Many Light Years Away", depicted changes of epidermodysplasia verruciformis that were manipulated by photo-editing software to resemble a comet. "The Interface of Love" depicted heart-shaped changes at the dermal-epidermal junction, with clip-art hearts superimposed. "Deep Impact", a high-powered view of an asteroid body, won third place. The microscope can reveal beauty in a wide range of skin specimens when used by an operator with an artistic eye.

Poster 667

RESIDENT

Pre-bisection of a single skin biopsy for immunofluorescence does not increase the risk of a technically inadequate study: a review of 3450 specimens

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Dermatologists sometimes bisect skin biopsies for dual evaluation, placing one half of the specimen in formalin for routine hematoxylin-and-eosin analysis and the other half for direct immunofluorescence. We evaluated the frequency of this practice and whether these samples were associated with a higher rate of studies that were technically inadequate for interpretation, e.g., complete or near complete disruption of the basement membrane zone, or missing epidermis or mucosal epithelium. Of 3450 cases consecutively submitted for direct immunofluorescence over a 6 year period, 434 were

pre-bisected specimens (12.6%). Of the bisected cases, 23 (5.3%) were technically inadequate. In non-bisected (wholly submitted) specimens, 153 of 3016 (5.1%) were technically inadequate. The frequency of technically inadequate specimens between bisected and non-bisected specimens was not significantly different ($p = 0.84$). In conclusion, bisection of skin biopsies by the submitting provider does not substantially increase the risk of a technically inadequate direct immunofluorescence study result.

Poster 668

RESIDENT

Cutaneous granular cell reaction: a potential mimic of granular cell tumor in superficial biopsies

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Exuberant collections of macrophages with granular eosinophilic cytoplasm may accumulate at sites of previous surgery or trauma and have been termed granular cell reactions (GCR). We report a case of a 56 year-old female who developed a firm nodule based in a scar resulting from excision of a mesenchymal neoplasm 30 years earlier. Biopsy demonstrated a nodular infiltrate of cells arrayed in sheets and cords dividing collagen bundles and surrounding skin appendages within the superficial dermis. Constituent cells were medium-sized and had abundant coarse granular, eosinophilic cytoplasm with distinct cell borders, round to ovoid nuclei and small nucleoli. There was no evidence of cytologic atypia or mitotic activity. A granular cell tumor (GCT) was suspected. Immunohistochemistry demonstrated the granular cells to be positive for CD68, and negative for S100, CD34, cytokeratin cocktail, desmin, and SMA. Closer inspection of H&E sections revealed hemosiderin and hematoidin pigment deep to the collection of granular cells. Based on this collection of findings, a diagnosis of granular cell reaction was rendered. Our case illustrates that cutaneous granular cell reactions may mimic granular cell tumors. The distinction between GCR and GCT is important as the latter represents a neoplasm, which may rarely locally recur when atypical or metastasize when frankly malignant. In contradistinction GCR is a benign reactive process. Clinical correlation, appreciation of hemoglobin breakdown products and immunohistochemistry for S-100 and CD68 are the key to making the distinction between these lesions, particularly when faced with limited clinical information and a small or superficial biopsy.

Poster 669

RESIDENT

Localized lymphedema can explain the phenomenon of labial hypertrophy: a case series

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Background: Labial hypertrophy (LH), or redundant vulva, is thought to represent an anatomic variant of aesthetic and/or functional importance. Most existing discussions of LH evaluate surgical methods and outcomes rather than the pathology of the labial tissue itself, which is often reported as normal. Objective: To describe the histopathology of LH. Materials and methods: Consecutive cases of labioplasties performed for LH were retrieved from a 10-year period. Clinical and pathologic features were recorded. Results: Thirty-three labioplasty specimens from 30 women were identified. The patients had a mean age of 34, with a range from 14-62 years. LH was asymmetric in 50%. Associations included pregnancy, vulvar tear during vaginal delivery, and obesity. Three patients experienced recurrence of LH requiring a second labioplasty. Histopathologically, all LH specimens showed signs of lymphedema: dermal edema, fibroplasia, dilated lymphatic vessels, and uniformly distributed spindle and dendritic cells. Sebaceous hyperplasia and lichenification were common. Demodex infestation was evident in 1 patient. Immunohistochemical staining with D2-40 confirmed the presence of numerous dilated lymphatics, and staining for estrogen and progesterone receptors demonstrated their expression on stromal cells. Conclusions: Rather than an anatomic variant, labial hypertrophy appears to be a phenomenon caused by localized lymphedema. Therefore, in the evaluation of LH, identification of causative and exacerbating factors of lymphedema, such as trauma/scarring, inflammation, and obesity, should be factored into its management.

Poster 670

RESIDENT

Malignant mesothelioma diagnosed by cutaneous biopsy

Laurel Stevens, MD

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Malignant mesothelioma is an uncommon malignancy that originates from the pleura, peritoneum, pericardium or tunica vaginalis of the testis. Cutaneous involvement and primary diagnosis by cutaneous biopsy are exceedingly rare. We report a case of a 50-year-old female who presented with an indurated plaque on her right flank for six months. The patient had a recent history of unexplained, recurrent pleural effusions. Pleural fluid cytologic studies, scalene fat pad biopsy, pleural biopsy, and mediastinal lymph node biopsy were all negative. The patient denied exposure to asbestos. Skin biopsy of the indurated plaque on the flank revealed infiltration of the subcutis by epithelioid cells in sheets and cords with

tubule formation. The tumor cells expressed cytokeratin (AE1/AE3), calretinin, and WT-1 but were negative for S-100 protein, thyroid transcription factor-1, CD31, CD33, CD34, CD45 and myeloperoxidase by immunohistochemistry. A diagnosis of epithelioid-type malignant mesothelioma was made based on the histologic and immunohistochemical findings. Mesothelioma rarely affects the skin. Although there are reports of distant cutaneous metastasis, cutaneous involvement is most often due to direct extension or seeding of tumor cells during diagnostic or therapeutic procedures. Most reports of mesothelioma affecting the skin involve patients previously diagnosed with mesothelioma or with history of significant exposure to asbestos. This case illustrates a rare primary diagnosis of malignant mesothelioma by skin biopsy. Despite the rarity of this diagnosis on cutaneous biopsy, it is important to keep mesothelioma in the differential diagnosis when evaluating cytokeratin-positive epithelioid tumors of the skin.

Poster 671

RESIDENT

Tinea versicolor, vitiligo, or both? an exploration of the mechanisms of hypopigmentation in Malassezia infection

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An 85 year-old man with cutaneous T cell lymphoma presented with hypopigmented and dyspigmented patches over the chest and upper back. Biopsies showed a patchy reduction of melanin at the dermoepidermal junction by Fontana staining and diminished melanocytes by MITF staining. Due to focally preserved melanocytes, this was initially diagnosed as an early, incomplete, or recovering focus of vitiligo. However, subsequent biopsies demonstrated a similar decrement in the number of melanocytes and melanin production with concurrent tinea versicolor (TV) infection. Recent culture studies of human melanocytes have demonstrated that Malassezia alkaloids are capable of inducing apoptosis in a dose-dependent manner. This mechanism of depigmentation is different than the traditionally understood inhibition of tyrosinase enzymes by Malassezia metabolites. This unusual case prompts the following question: What do we really know about the mechanisms of hypopigmentation in tinea versicolor (Malassezia) infection?

Poster 672

RESIDENT

Neonate with epidermolytic ichthyosis displaying classic histopathologic findings of epidermolytic hyperkeratosis

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Epidermolytic hyperkeratosis (EHK) is characterized histopathologically by hyperkeratosis, vacuolization of the spinous layer and large keratohyaline granules in the keratinocyte cytoplasm. Ultrastructurally, cells show clumped keratin tonofilaments, which is caused by mutations in KRT1 or KRT10. Varying degrees of dyskeratosis may be present. The clinical entity epidermolytic ichthyosis (previously known as bullous congenital ichthyosiform erythroderma or bCIE) displays the finding of EHK histologically and is one of the few neonatal ichthyoses which can be diagnosed via histopathological findings. We present a full term neonate born with a white semi adherent membranous covering which was eroding away, leaving a bright red base. Within hours, the material became hyperkeratotic and thick. He had no prenatal complications. Upon initial examination, the initial clinical differential did include a collodion membrane (for which a large differential exists including lamellar ichthyosis and non-bullous CIE). However, histopathology revealed classic findings of EHK allowing for a rapid diagnosis and subsequent further workup and counseling of family members. Our patient was monitored in the neonatal intensive care unit for one week, where he did well with supportive care. We present this case for its classic presentation of a rare genodermatosis, and to further the clinical pathological correlation with neonatal erythrodermas and ichthyoses. There are many neonatal presentations which are non specific and cannot be truly diagnosed at birth, but the diagnosis of epidermolytic ichthyosis can be confidently made with the histopathologic findings of EHK.

Poster 673

RESIDENT

Pacinian corpuscle hyperplasia

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A 34-year-old man presented with a 3-year history of a tender blue macule on the left thumb, which he sustained 3 years earlier when cut while using a paint sprayer. Physical examination showed a nearly triangular blue-gray macule about 1cm long, overlying tender indurated dermal tissue. A 2mm punch biopsy specimen was obtained and on gross examination, it showed a solid dark blue dermis with several 1- 1.8mm white, glistening, rice-grain-shaped globules on the distal aspect of the tissue core.

H&E stained tissue specimens showed a dermis full of pigment-laden macrophages and multinucleated giant cells with haphazard arrangement of nuclei. Subjacent to the dermis were hypertrophic Pacinian corpuscles with an abnormally elevated number of concentric lamellae. The dermal changes represent a foreign body reaction to the paint. The enlarged Pacinian corpuscles are the likely source of pain and represent a rare condition called Pacinian corpuscle hyperplasia or, sometimes, Pacinian neuroma. This case lends support to the widely held theory that Pacinian hyperplasia is due to either gross trauma or repeated micro-trauma to the hand. Although rare (with approximately 78 reported cases through 2011), hyperplastic Pacinian corpuscles should be remembered when considering the differential diagnoses of painful subcutaneous lesions of the digits, especially in manual laborers, who may have suffered prior trauma to the area.

Poster 674

RESIDENT

Erdheim-Chester disease presenting as multiple reticulohistiocytic granulomas

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Erdheim Chester disease (ECD) is a rare multi-system non-Langerhans cell histiocytosis characterized by infiltration of involved organs by foamy histiocytes and Touton giant cells. The most common site of involvement is the long bones of axial skeleton followed by nervous system. Cutaneous involvement occurs rarely as the presenting feature but more commonly as part of a multi-organ involvement and typically includes xanthelasmas and occasionally red-brown papules and nodules composed of dermal infiltrate of foamy histiocytes and Touton giant cells resembling xanthogranulomas. We present an unusual case of ECD where the skin biopsies showed reticulohistiocytic granulomas. The patient is a 41-year-old man with past medical history of pituitary dysfunction who presented to the neurology clinic for severe vertigo and difficulty walking. Extensive work-up including MRI of brain and CSF was largely unrevealing. He subsequently developed multiple hyperpigmented papules on the trunk and arms which were biopsied. The biopsies revealed a well-defined dermal collection of histiocytes and multinucleated giant cells with abundant eosinophilic, finely granular cytoplasm with a "ground-glass" appearance typical of reticulohistiocytic granuloma. The possibility of multicentric reticulohistiocytosis was suggested. Additional work-up included bone scans and x-rays of the lower extremities that showed osteosclerotic lesions. Based on the clinico-radiologic features and skin biopsy findings of non-Langerhans histiocytosis, a diagnosis of ECD was established. We draw attention to the unusual morphologic finding of reticulohistiocytic granulomas rather than the typical xanthogranulomas with foamy macrophages reported in ECD, and suggest that ECD should be considered in the differential diagnosis of multiple reticulohistiocytic granulomas.

Poster 675

RESIDENT

Online interactive dermatopathology - is there a need for dermatopathology teaching resources on the internet?

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In July 2010, we launched an interactive website called DermOID - Online Interactive Dermatopathology. Our website is free and offers regular multiple-choice quizzes with images of histopathology and in-depth explanations. Upon registration, users answer a survey regarding level of interest, daily workload, amount of dermatopathology training, purpose for using DermOID, availability of online resources, and preferred learning modalities. Analysis of 258 responses showed most registrants were resident physicians (42%), dermatopathologists (16%), pathologists (14%), dermatologists (13%), and others. As expected, dermatopathologists had the highest daily workload of skin specimens, dermatopathologists and dermatologists reported more residency training in dermatopathology, and fellows and residents showed the most interest in using DermOID for board exam preparation. The majority of respondents utilized the internet either always (20%) or often (55%), and 94% of users preferred to utilize some combination of internet and printed resources for learning. Despite an interest, significant proportions of users were either not aware of internet dermatopathology teaching resources (20%) or thought there were few such websites (47%). Also, 38% of respondents thought there were few interactive dermatopathology teaching websites, and 39% were unaware of the number of such websites. Following two years, over 280 individual dermatopathology cases and over 250 registered users, we believe that DermOID fills an important niche in online dermatopathology education. Our work is on-going and we hope to further tailor our site to the individual needs of users. Furthermore, the technology used in DermOID suggests possibilities for future use in telepathology and continuing medical education (CME).

Poster 676

RESIDENT

Minocycline induced hyperpigmentation coexisting with pigmented purpuric dermatitis

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Minocycline is a semi-synthetic tetracycline antibiotic that is often used to treat acne vulgaris and rosacea. Skin pigmentation may occur as an adverse effect in 2.4 to 14.8% of patients treated with minocycline. Minocycline induced hyperpigmentation of the skin has 3 unique

manifestations. Type I presents as blue-gray pigmentation in scars. In Type II, blue-gray macules and patches appear on previously normal skin and there is a predilection for the anterior legs. Type III consists of brown discoloration of sun-exposed sites. A 75-year-old man presented with multiple asymptomatic non-palpable brown-black macules ranging from 2 to 4 mm in diameter distributed on bilateral anterior shins. He had a remote history of malignant melanoma occurring on his face and was being treated with chronic minocycline therapy for rosacea. A biopsy specimen demonstrated diffuse brown-black pigment deposition in macrophages in the superficial dermis that stained positive for iron with Prussian Blue stain and positive for a melanin-like substance with Fontana-Masson stain. The biopsy specimen also revealed a mild interface dermatitis with patchy superficial perivascular lymphoid infiltrates and extravasation of erythrocytes and hemosiderin deposition that was consistent with pigmented purpuric dermatitis. The etiology of pigmented purpuric dermatitis is unknown but it may be related to venous insufficiency, drug or contact hypersensitivity, and it may be the initial presentation of a T cell lymphoproliferative disorder.

Poster 677

FELLOW

Pigmented Apocrine Hamartoma: A Rare Lesion of the Vulva

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We describe two patients, ages 10 and 38, who presented with dark pigmented papular lesions of long duration located on the vaginal labia. Histologically, both lesions showed squamous mucosa with dermal cystically dilated apocrine glands tightly associated with and focally connected to the epithelium. The glands contained proteinaceous luminal secretions with melanin pigment. These glands were lined by a double-layered epithelium with luminal melanin laden cuboidal cells and an outer myoepithelial layer. The cuboidal cells had a moderate amount of eosinophilic cytoplasm. Immunohistochemical stains for Melan-A, HMB-45 and S-100, with appropriate controls, highlighted an increased number of dendritic melanocytes colonizing the lining of these dilated glands, correlating with the pigmented character of the lesion. The histological features and the immunoprofile are consistent with pigmented apocrine hamartoma of the vulva. Although this lesion most commonly occurs in the female genital area, it has been described at other anatomic sites including the face, trunk and axilla. All affected patients reported in the literature were between the ages of 7 to 19 years of age. Thus, our 38-year-old patient is the oldest reported patient with this lesion. The limited literature on this lesion suggests a benign clinical course with no local recurrences or progression to malignancy after a short-term clinical follow up. It is important to recognize

this entity and include it in the differential diagnosis of pigmented lesions of the vulva.

Poster 678

RESIDENT

Verruciform xanthoma arising in the anogenital area: a potential possible diagnostic pitfall

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Verruciform xanthoma is a papillomatous mucosal or cutaneous lesion that presents a potential diagnostic pitfall when arising in the anogenital region. A 65-year-old male presented with a pedunculated, hypopigmented nodule on his scrotum. The clinical differential diagnosis included condyloma acuminatum, squamous cell carcinoma, and verrucous carcinoma. Histopathologic examination showed epidermal papillomatosis with hyperkeratosis, focal parakeratosis and absent to minimal squamous atypia. The low-magnification differential diagnosis included condyloma acuminatum and verrucous carcinoma. On high-magnification examination, aggregates of histiocytes with vacuolated cytoplasm (xanthoma cells) were identified in the papillary and superficial reticular dermis. Given the epidermal features and presence of xanthoma cells in the superficial dermis, the lesion was diagnosed as a verruciform xanthoma. Verruciform xanthoma is thought to represent a reactive process secondary to epithelial damage. Although the majority of verruciform xanthomas arise on oral mucosa, rare cutaneous cases present in the anogenital region. When occurring in the anogenital region, there is potential for diagnostic error as they can clinically and histologically mimic condyloma acuminatum, verrucous carcinoma, and squamous cell carcinoma. Accurate diagnosis of verruciform xanthoma is dependent on careful evaluation of papillomatous and verrucous cutaneous lesions for characteristic xanthoma cells in the superficial dermis.

Poster 679

FELLOW

A congenital cervical midline cleft

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Congenital cervical midline cleft is a rare ventral midline anomaly with fewer than 100 cases reported in the English literature. It most commonly occurs on the neck and is usually noted at or soon after birth as a skin tag adjacent to a sinus tract. Most authors believe that the anomaly occurs secondary to a failure in fusion of the 1st and 2nd branchial arches or failure of midline mesodermal penetration. We present a 9 month-old boy who underwent excision of his congenital midline defect. Histologic examination of the surgical excision specimen revealed dermal skeletal muscle bundles, a sinus tract lined by stratified squamous epithelium with underlying fibrosis, and rudimentary salivary glands consistent with a congenital cervical midline cleft.

Poster 680

RESIDENT

Amyloid nodule: a rare mimic of giant cell fibroma

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A 63 year-old Caucasian female complained of a mobile mass in left cheek and lip present for approximately six months. The lesion was freely movable, nontender, pink, and measured 1.5 cm in diameter. When excised, the lesion easily shelled out. The clinical differential diagnosis was a fibroma, lipomatous tumor, or dermoid cyst. A benign salivary gland neoplasm was also considered. Grossly, the nodule was solid and gray, without necrosis or hemorrhage. Microscopically, the nodule consisted of a core of dense, eosinophilic, acellular connective tissue with a peripheral zone of spindle cells, giant cells and histiocytes. CD68 stain highlighted the giant cells and histiocytes. The diagnosis of giant cell fibroma was strongly considered. However, special stains for amyloid including crystal violet, congo red and thioflavin T were all strongly positive for homogenous deposition of amyloid, primarily within the central portion of the lesion with less staining at the periphery. Review of the medical literature revealed that amyloid nodules of the oral cavity are quite rare, and the condition has only been cited once in the dermatopathology literature. A few case reports noted an exuberant giant cell component, and so amyloid nodules have been misdiagnosed as giant cell tumors. This case represents a close histological mimic of giant cell fibroma. Establishing the correct diagnosis of amyloid nodule is important for alerting the clinician to a possible underlying plasma cell dyscrasia or other lymphoproliferative disorder.

Poster 681

RESIDENT

Epithelioid sarcoma: a report of two cases

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Epithelioid sarcomas are rare, soft tissue sarcomas with a propensity for local recurrence and regional metastasis. We report two cases to increase awareness of this entity in the dermatology community. Case 1 is a 26 year-old man with a 9 year history of an 8 cm ulcerated mass on his right calf. Case 2 is a 68 year-old man with a 6 month history of an enlarging 5 cm painful mass on his right distal thigh. Biopsy revealed tumors comprised of bland, large, epithelioid cells, occasional mitotic figures and focal necrosis. Immunocytochemical stains were positive for CD34, vimentin, pancytokeratin, EMA and negative for p63, CK7, CK20 and S-100. This constellation of findings is consistent with epithelioid sarcoma (ES). ES presents as a slow-growing tumor on the distal extremities of young men and is often associated with tendons, fascia, periosteum and nerves. Because of its rarity, bland histology, and mimicry of granulomatous lesions and squamous cell carcinoma, the diagnosis is easily missed.

Unlike other sarcomas, nodal metastases occur in 22-48% of ES patients and can arise 20 years after diagnosis. Survival at 5 and 10 years is 68% and 61%, respectively. Independent predictors of survival are: stage, operability, age younger than 16 years, and negative lymph node involvement. The mainstay of treatment is wide local excision with sentinel lymph node dissection. Adjuvant radiation may prevent local recurrence. Thus, ES is an aggressive soft tissue sarcoma that should be considered in the differential diagnosis of epithelioid infiltrates in the skin.

Poster 682

Two rare cases of lipomatous neurofibroma

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We report two cases of lipomatous neurofibroma, a recently described, rare variant of neurofibroma. The first patient was a 64 year-old female who presented with a lesion on the mid back that clinically was consistent with a neurofibroma. The second patient was a 22 year-old male who presented with a painless right index finger mass of 10 years duration. The mass was excised and consisted of a 2.0 cm white to pink rubbery firm well-circumscribed nodule. A shave biopsy from the first case revealed a well-circumscribed dermal spindle cell proliferation within a collagenous stroma with focally interspersed adipose tissue. The excisional biopsy from the second case demonstrated a soft tissue well-circumscribed spindle cell proliferation within a finely fibrillary collagenous and myxoid stroma with focally interspersed adipose tissue. The spindle cells in both cases displayed elongated to wavy nuclei with evenly dispersed chromatin. Mitoses were inconspicuous. Scattered mast cells were present in the background. Immunohistochemical staining was performed in case two and demonstrated that the spindle cells were positive for S100, CD34, and FXIIIa and negative for SMA, desmin, and CD57. The presence of adipose tissue intermingled with neurofibroma is a rare occurrence with the etiology ascribed to tumor senescence, metaplasia, or chronic injury. Our two cases are also notable for their location outside of the head and neck where they are most commonly seen.

Poster 683

RESIDENT

Low-grade fibromyxoid sarcoma with nuclear pleomorphism arising in the subcutis of a child

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A 10-year-old male presented with a back mass clinically suspicious for a cyst. No past medical history was available. The mass was excised and displayed a well-circumscribed, subcutaneous neoplasm. The tumor was composed of bland spindle cells arranged in hyper- and hypocellular zones in a variably fibrotic and myxoid background with arcades of small curved blood vessels. Additionally, scattered cells displayed large, hyperchromatic, pleomorphic nuclei. The tumor cells were focally positive for EMA, but negative for S100 protein and desmin by immunohistochemistry. The tumor cells were positive for FUS gene rearrangement by fluorescence in situ hybridization (FISH) using breakapart probes confirming the diagnosis of low-grade fibromyxoid sarcoma (LGFMS). The patient was treated with subsequent wide local excision and is alive with no evidence of disease 7 months after diagnosis. LGFMS typically occurs in the deep soft tissues of adults where it is characterized by a low histologic grade but a significant risk of late recurrence and distant metastasis. LGFMS is rare both in children and in superficial locations, and nuclear pleomorphism is exceptionally rare in LGFMS. The young age of the patient, small tumor size, superficial location, and presence of scattered pleomorphic cells in this case resulted in significant diagnostic difficulty which was resolved by the use of FISH for the FUS gene rearrangement.

Poster 684

FELLOW

An interesting case of low grade pleomorphic malignant fibrous histiocytoma, pleomorphic undifferentiated sarcoma, on the nose of a 72 year-old man

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Pleomorphic malignant fibrous histiocytoma (MFH), also known as pleomorphic undifferentiated sarcoma (PUS), is a soft tissue sarcoma which is a diagnosis of exclusion as a sarcoma not otherwise classifiable. It is considered one of the most common sarcomas in late adulthood, occurring often in Caucasian males. It is considered a fibroblastic or undifferentiated neoplasm. Location of this tumor is usually in the extremities or retroperitoneum with metastases to the lungs, bone, and liver. Often, they are clinically described as enlarging, painless masses. Histopathology demonstrates high cellularity, marked nuclear pleomorphism, an increased mitotic rate, spindle cell morphology, and necrosis in high grade

lesions. Treatment consists of resection; chemotherapy use is controversial. We present a case of a 72-year old Caucasian male with an enlarging mass on his nose. An initial superficial biopsy was performed a month before the resection. This initial biopsy showed changes of a benign hemangioma. Over the following month, the lesion continued to grow and became deep red in color. An excision was performed. Initially, we thought it would be an angiosarcoma. Histologically, it is a cellular tumor with a storiform pattern, focal marked nuclear pleomorphism, scattered mitoses, and focal necrosis. After an exhaustive panel of immunohistochemical stains, only CD10 demonstrated consistent positivity. A diagnosis of atypical fibroxanthoma was considered; however, the degree of nuclear pleomorphism and focal necrosis ruled against it. This case is interesting because of its unusual location and clinical findings.

Poster 685

RESIDENT

Subcutaneous myxoid liposarcoma

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We report a case of myxoid liposarcoma presenting as a painful mass (3.8 x 2.2 x 2.2 cm) on the dorsal left foot of a 40-year-old man. The mass was first noted by the patient 6 years prior to presentation and had slowly enlarged over time. Imaging studies revealed a complex multilobulated lesion within the subcutis overlying the fourth and fifth distal metatarsals and metatarsophalangeal joints without involvement of underlying bony structures. Initial clinical impression was that the mass represented a complex ganglion cyst. Histopathologic review following lesional biopsy and subsequent forefoot amputation revealed a well-circumscribed but unencapsulated multilobular mass. The mass demonstrated a myxoid stroma, a delicate branching capillary vasculature, and was composed of bland hyperchromatic spindled to round cells and lipoblasts. There was enhanced cellularity at the periphery of the lobules but the round cell component comprised less than 5% of the tumor cells and no necrosis was identified. Fluorescence in situ hybridization (FISH) for DDIT3 (CHOP) gene rearrangement confirmed the diagnosis of myxoid liposarcoma. No evidence of a primary or synchronous lesion was identified. Careful histologic evaluation with recognition of true lipoblastic differentiation and confirmation by gene rearrangement studies are essential in the diagnosis of myxoid liposarcoma and in differentiating it from myxofibrosarcoma, a more common and aggressive subcutaneous sarcoma. Myxoid liposarcoma can rarely present as a subcutaneous mass and should be considered in the differential diagnosis of cutaneous myxoid tumors in adults.

Poster 686

FELLOW

Superficial paramucosal clear cell sarcoma of the soft parts resembling melanoma in a 13 year old boy

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Clear cell sarcoma (CCS) of tendons and aponeuroses/malignant melanoma (MM) of soft parts is an aggressive rare neoplasm that is characterized by a nested or fascicular pattern of spindle cells and a pathognomonic reciprocal translocation, t(12;22)(q13;q12), that results in fusion of the EWS and ATF1 genes. CCS most commonly presents as a deep soft tissue tumor that most frequently affects the extremities of young female adults, typically around the ankle and the foot. Numerous recent studies have recognized the importance of a cutaneous clear cell sarcoma variant (CCCS), which can mimic a broad spectrum of entities, including spindle cell melanoma, spindle cell squamous carcinoma, cutaneous leiomyosarcoma and atypical fibroxanthoma. We report a case of a 13-year-old boy with CCCS who presented with a few month history of an asymptomatic papule on the lower lip, suggestive of a mucocele. Biopsy of the lesion demonstrated a wedge shaped neoplasm arranged in nests and fascicles of epithelioid to oval shaped cells with pale cytoplasm, open chromatin and prominent nucleolus. The superficial component was closely opposed to the basal epidermis resembling the junctional nests of a melanocytic neoplasm. The process extended into and involved the striated muscle of the lip. The cells expressed S-100, CD99 and synaptophysin, with focal HMB-45 and MITF positivity by immunohistochemistry. Fluorescence in situ hybridization confirmed the presence of the, t(12;22)(ESWR1-ATF1) translocation. We present this case to demonstrate this novel presentation of clear cell sarcoma in a paramucosal location extending from an immediate submucosal location into the striated muscle of the lower lip.

Poster 687

RESIDENT

Spindle cell hemangioma of the scrotum: a case report

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Spindle cell hemangioma is a rare, benign vascular lesion first described by Weiss and Enzinger in 1986. Initially these lesions were classified as low grade angiosarcomas, however further study revealed that despite a high local recurrence rate, patients never developed metastatic disease. Both men and women are equally affected, and

they can occur in every age group. There are reports of spindle cell hemangioma in the oral cavity, penis, vulva, pancreas and bone, but the most common location is the upper and lower extremities. Histologically, spindle cell hemangiomas consist of cavernous vessels surrounded by cellular spindle cell areas with slit-like vascular spaces. The differential diagnosis includes malignant neoplasms such as Kaposi's sarcoma and angiosarcoma, but a low mitotic rate and lack of cytologic atypia or necrosis underlies its benign nature. We report a case of a 53-year-old man who presented with an asymptomatic firm 1.8 cm nodule on his right scrotum. He had no history of trauma or evidence of scrotal varicoceles, nor did he have a history of any sexually transmitted diseases. No other lesions were noted. Excision revealed an ectatic vascular and spindle cell tumor with areas of thrombosis. HHV-8 immunohistochemical stain was negative, and CD34 marked endothelial cells surrounding ectatic vascular spaces and medium-sized vessels. KI-67 marked less than 5% of the nuclei of endothelial cells. After thorough review, the lesion was diagnosed as a spindle cell hemangioma. To our knowledge this is the first reported case of a spindle cell hemangioma presenting on the scrotum.

Poster 688

RESIDENT

A rare case of a malignant squamomelanocytic tumor

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We report a case of malignant squamomelanocytic tumor, an extremely rare entity. A 73 year-old man presented with a lesion on the left neck. The clinical impression was basal cell carcinoma. A shave biopsy revealed a biphasic population of malignant cells in varying sized nests within the dermis. The squamous component consisted of sheets of malignant squamoid cells demonstrating focal keratinization and squamous "whorls". This population was closely intermingled with nests and sheets of malignant melanocytes. The epidermis displayed a markedly atypical melanocytic proliferation at the dermoepidermal junction with extensive infiltration of adnexal structures and scattered atypical pagetoid melanocytes. Immunohistochemical staining for pankeratin, cytokeratin 5/6, and p63 highlighted the squamous component. Immunohistochemical staining for S-100, Melan-A, HMB-45, MITF, and tyrosinase highlighted the melanoma component. The constellation of pathologic findings was consistent with a malignant squamomelanocytic tumor with both invasive melanoma and invasive squamous cell carcinoma components. The patient is free of disease after three months. The full spectrum of biologic behavior of squamomelanocytic tumors is not firmly established, but rare metastases have been reported. As more cases of squamomelanocytic tumor are reported and their outcomes documented, it may be possible to determine their biologic potential and establish standardized criteria for classification, prognostic, and therapeutic purposes.

Poster 689

FELLOW

Pigmented apocrine hamartoma of the vulva

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Pigmented vulvar lesions are present in approximately 10% of females. The differential diagnosis of a pigmented lesion in the vulva includes melanosis, verruca vulgaris, seborrheic keratosis, melanocytic nevi, melanoma, and pigmented vulvar intraepithelial neoplasia (squamous cell carcinoma-in situ). A 23-year-old woman was noted to have a small blue papule in the 3:00 area of the left labia minora. The patient was asymptomatic, and an excisional biopsy was performed to exclude the presence of a melanocytic lesion, specifically melanoma. The biopsy revealed a dermal lesion comprised of cystic spaces lined by a cuboidal apocrine epithelium, which was surrounded by an outer layer of myoepithelial cells. The apocrine epithelium contained an abundance of brown melanin pigment and melanocytes, which were highlighted by the Melan-A stain. Some of the melanocytes exhibited a dendritic morphology. The cystic structure was filled with debris and desquamated cells. Pigmented apocrine hamartomas occurring on the vulva are uncommon but they should be considered in the differential diagnosis of a pigmented lesion on the labia minora of a young woman.

Poster 690

FELLOW

Malignant eccrine spiradenoma

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There is paucity of reports about the extremely rare phenomenon of malignant transformation in eccrine spiradenoma (ES). We present three cases of enlarging tumors in female patients aged 39 to 89 years. Biopsies revealed both a benign appearing population composed of deeply basophilic epithelial lobules in the dermis consisting of "lymphocytoid" cells with small hyperchromatic nuclei and ductal-type cells with larger vesicular nuclei and a malignant proliferation of cells with nuclear pleomorphism and atypia. Three patterns were observed. One case showed gradual transition from benign ES to malignancy associated with focal clear cell change. Another case revealed a benign ES component associated with geographic areas of necrosis and increased mitotic rate. The third case showed a complete replacement of ES by sheets of atypical population of cells exhibiting marked mitotic activity and infiltrative behavior. Immunohistochemical analysis revealed high positivity for Ki-67. Considerable proliferative activity was documented by mitotic rate in the tumors reaching 15 per 10 high power fields (x40) counted. Two cases had membranous staining for Her2Neu while one case showed focal positivity for

ER; PR was negative. Positive p53 staining was also noted. Although wide local excision with tumor free margins and sentinel node testing were performed in all cases, the presence of additional stains may suggest the possibility of hormonal therapy and multiple kinase inhibition as adjunctive therapeutic options.

Poster 691

RESIDENT

The future of diagnostic dermatopathology: telepathology using the Apple iPad2®

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BACKGROUND: We examined the efficiency of using an Apple iPad2® remotely to diagnose dermatopathology cases via telepathology. **DESIGN:** 110 previously diagnosed dermatopathology specimens were retrieved by medical record and reviewed. Inflammatory lesions and excisions were excluded, leaving 89 cases. Five months later, the cases were transmitted for teleconsultation to the original diagnosing dermatopathologist. A high resolution video camera (Nikon DS-L2, Ver 4.4) mounted on a pathology microscope was used to transmit digital video of a slide to an Apple iPad2® at the pathologist's remote location via live streaming at an interval time of 500ms and a resolution of 1280/960. Concordance to the original diagnosis and the seconds elapsed to reaching the diagnosis was recorded. **RESULTS:** 25.8% of cases were melanocytic, while 74.2% were non-melanocytic. 92.1% of cases were diagnosed immediately. Of these cases, 98.8% of the telediagnoses were concordant with the original. 7.9% of cases were deferred for conventional diagnosis. Of these cases, 57.1% of cases had a differential diagnosis of squamous cell carcinoma (SCC) versus inflamed keratosis (ISK) and 42.9% of cases had a differential diagnosis of a subtle melanocytic lesion on sun-damaged skin. In 1.2% of cases, there was a clinically significant difference in diagnosis between the conventional diagnosis and the telepathological diagnosis. The average time to diagnosis was 39.6 seconds (range 10 to 218 seconds). **CONCLUSION:** Telepathology performed by dermatopathologists via an Apple iPad2® may serve as a reliable and rapid technique for the diagnosis of routine cases with some diagnostic caveats in mind.

Poster 692

RESIDENT

Graft versus host disease associated eosinophilic fasciitis (GVHD-EF)- a diagnostic challenge

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A 37 year-old man presented with insidious development of pruritic, indurated, slightly scaly plaques with decreased hair growth on the lateral thighs and skin tightening. History was significant for acute lymphoblastic leukemia with bone marrow transplantation three years prior. Biopsy revealed a layered pattern of collagen homogenization, with deep dermal and superficial pannicular sclerosis highlighted by trichome and PAS stains. A deep lymphoplasmacytic infiltrate embedded in a layered pattern within sclerotic collagen fibers was observed. Rare basal keratinocytic apoptosis most prominent at hair follicles was noted without frank interface dermatitis or epidermal atrophy. Deep sclerosis raised the differential of scleroderma and deep morphea, unlike morpheaform GVHD where papillary dermal sclerosis and epidermal atrophy would be typical. However, the presence of apoptosis in the basilar layer in combination with deep sclerosis overall favored a diagnosis of GVHD associated eosinophilic fasciitis (GVHD-EF). The classification of chronic cutaneous GVHD extends far beyond the classic division of lichenoid and sclerodermoid, and knowledge of scleroderma-like forms of chronic GVHD is continuing to expand. As in this case, peripheral eosinophilia is absent in 40% of GVHD-EF. Magnetic resonance imaging can aid in establishing the diagnosis of fascial involvement with GVHD when uncertainty persists. Clinical implications of misdiagnosis are significant, as treatments for sclerodermoid types of chronic cutaneous GVHD, for example with extra-corporeal photopheresis, vary significantly from more superficial forms and delays in appropriate treatment should be avoided. Increasing exposure to allogeneic stem cell transplantation warrants further delineation and understanding of the range of cutaneous GVHD presentations.

Poster 693

RESIDENT

Langerhans cell histiocytosis associated with lichen sclerosis of the vulva

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Langerhans cell histiocytosis (LCH) is characterized by a clonal proliferation of bone marrow-derived Langerhans cells. The condition spans a wide clinical spectrum, and can manifest as localized, multifocal, or disseminated disease. While cutaneous involvement is relatively common, LCH restricted to the vulvar area is a rare phenomenon and can occur in different clinical settings. Although this form of disease does not disseminate in these patients, it can be aggressive with local recurrences despite excision and

radiation therapy. We present a case of a 68-year-old female with a one-month history of pruritic lesions on her vulva. Physical examination showed whitish plaques with scattered nodular areas on the labia majora. A vulvar biopsy demonstrated a background of lichen sclerosus with foci of oval to polygonal cells with moderately abundant eosinophilic cytoplasm and folded nuclei showing frequent nuclear grooves. Immunohistochemical staining showed that the cells were positive for CD1a and S-100, confirming the diagnosis of LCH. On further workup, there was no evidence of disseminated disease involving other organs. While vulvar LCH is uncommonly seen, and with only one previous case report in the literature arising in the setting of lichen sclerosus, this case illustrates the importance of recognizing this condition and ensuring proper clinical follow-up to rule out a systemic involvement.

Poster 694

RESIDENT

The utility of deeper tissue sections in dermatopathology

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Guidelines are lacking for appropriate utilization of deeper sections in dermatopathology. Prior limited data suggest that deeper sections are more helpful in diagnosing neoplastic than inflammatory skin diseases. Better characterization of specific skin lesion types benefiting from deeper sections may provide useful information to improve diagnostic accuracy and reduce costs. We performed a prospective study using 150 routine biopsies for which deeper sections were pre-ordered before reviewing the initial hematoxylin-and-eosin section. Diagnoses before and after deeper sections were recorded. Utility, the primary endpoint, was defined as the percent of cases in which the deeper sections altered the final diagnosis. In our series, most cases were neoplastic(97%; melanocytic and keratinocytic neoplasms were equally represented) vs. inflammatory(3%); submitted as shave(93%) vs. punch(7%) biopsies. Data analyses revealed that deeper sections altered diagnosis in 9% of cases. Clinical significance, a secondary endpoint, was defined as whether the diagnostic information provided by deeper sections altered clinical management. A clinically-significant change occurred in 7% of all cases, and more often for keratinocytic(64%) than melanocytic(36%) neoplasms. There was one diagnosis of invasive melanoma detected on deeper sections, which was not detected initially. Thus, 68 melanocytic lesion cases required deeper sections in order to detect one melanoma. In our laboratory, processing cost of 3 deeper sections is \$1.56. Assuming pathologist time spent evaluating a set of deeper sections is equivalent to the time spent evaluating a new case, and that the approximate reimbursement per case is \$119.16, then the cost of detecting one melanoma is estimated at \$8,208.96.

Poster 695

RESIDENT

Validation of anti-PHH3 immunostain in fibrohistiocytic tumors: a study of 96 cases

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Background: Accurate mitotic count is essential for classification of fibrohistiocytic lesions. Traditional counting of mitotic figures (MFs) using H&E sections is time consuming and prone to inter-and intraobserver variability. Our study aims to examine the performance of an antibody to phosphohistone H3 (PHH3) that labels MFs in all stages of mitosis in a diverse group of fibrohistiocytic tumors. Design: After obtaining IRB approval, 96 archival cases were selected. The series includes 40 dermatofibromas (DF), 28 atypical dermatofibromas (ADF) and 28 dermatofibrosarcoma protruberans (DFSP). All cases were stained with Hematoxylin-Eosin (H&E), PHH3 and MIB-1 stains. Mitotic count was performed by examining twenty 40X fields by two independent investigators. A consensus average Mitotic Index (MI) per 10 high power fields by H&E (HE-MI), PHH3 (PHH3-MI) and a MIB-1 labeling index (MIB-LI) were generated for each case. Student's T test and Spearman's test with a $p < 0.05$ were used. Results: PHH3 enabled rapid recognition of different mitotic phases. PHH3-MI was significantly higher than HE-MI [3.0 vs. 1.1/10HPF] ($p < 0.001$) in all cases. Similar results were observed in the subgroups of DF [1.2 vs. 0.2/10HPF], ADF [4.6 vs. 1.2/10HPF], and DFSP [4.1 vs. 2/10HPF] ($p < 0.001$) respectively. A strong correlation with PHH3-MI to MIB1-LI and PHH3-MI to HE-MI was demonstrated in ADF's [$r = 0.83$ and $r = 0.78$; $p < 0.001$] and DFSP's [$r = 0.78$ and $r = 0.61$; $p < 0.001$] respectively. Conclusions: PHH3-MI performs better than conventional HE-MI in fibrohistiocytic lesions particularly in the ADF and DFSP subgroups. Further studies are underway to assess PHH3-MI performance in benign and malignant melanocytic lesions.

Poster 696

Dermal lymphedema of the breast

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Patients with lymphedema of the breast present with discrete plaques of edema and erythema that clinically mimic cellulitis or inflammatory breast carcinoma. This condition was originally described in 1996 by Loprinzi et al in the post-surgical setting following treatment of breast carcinoma. More recently, in 2001, King's group described 2 patients also surgically treated for breast carcinoma who developed similar findings. We report 10 patients with erythematous plaques on the breast with a peau d'orange appearance. Nine were female and one was male. Eight

patients had a history of ipsilateral surgery for treatment of breast carcinoma. One patient had undergone sentinel lymph node biopsy from the axilla for melanoma, and the male patient was morbidly obese and spent the majority of his time lying down on the side of presentation. All biopsies showed ectasia of dermal vessels associated with edema and a sparse perivascular lymphocytic infiltrate. Immunohistochemical staining with D2-40 confirmed that the vessels were lymphatic. In those patients for whom follow-up was available, most showed improvement/resolution over a one to two year time period. Dermal lymphedema is an under-recognized condition that mimics inflammatory breast carcinoma and does not respond to treatment for cellulitis. It most commonly occurs in patients with prior surgery for breast carcinoma but may also be seen after axillary sentinel lymph node biopsy or from chronic dependency/stasis. It is important that dermatologists and dermatopathologists be aware of this condition to facilitate accurate diagnosis.

Poster 697

Cutaneous cholesterol embolization to the lower trunk: an under-recognized and pathologically elusive presentation mimicking heparin necrosis

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Cutaneous cholesterol embolization classically occurs to distal extremities in the setting of atherosclerotic disease, typically after instrumentation or initiation of anti-coagulation therapy. Dermatological manifestations include livedo reticularis, gangrene, nodules and purpura. Although livedo reticularis may extend from the legs to the trunk, gangrenous lesions predominantly involve the distal digits. We describe a 65 year old man with severe atherosclerotic disease who developed multiple eschars on the thighs, periumbilical area and sacrum after undergoing three cardiac catheterizations and stenting of the superficial femoral artery. The lesions were initially tender nodules presumably at sites of enoxaparin injections. Two biopsies from ulcerated skin did not reveal a definitive cause for the ischemia. A third, incisional biopsy of a new, non-ulcerated, subcutaneous nodule revealed multiple cholesterol emboli. This case illustrates the challenge in diagnosis of cholesterol emboli, both clinically and histologically. Cutaneous lesions are morphologically variable and often occur with systemic symptoms, mimicking numerous conditions. This patient's lesions were initially attributed to heparin necrosis due to localization on the abdomen. Periumbilical and lumbosacral involvement has not been emphasized in the literature and should be recognized as an uncommon but important cutaneous manifestation of cholesterol emboli. As the gold standard for diagnosis is histological confirmation, multiple biopsies may also be required. Dermatologists and dermatopathologists should have a low threshold of suspicion to facilitate timely diagnosis and treatment.

Poster 698

RESIDENT

New onset bullae in a case of long-standing mycosis fungoides

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Mycosis fungoides (MF) is known to have many different clinical and histological variants. As early as 1887, a rare variant associated with pemphigus-like bullae was described. This presentation has been termed mycosis fungoides bullosa when there is no clear etiology for the associated bullae. When evaluating new onset bullae in the setting of MF, common causes such as contact dermatitis, infections, and medication reactions must be considered. While within this differential diagnosis, autoimmune bullous disease has only rarely been reported to occur with MF. We describe a case of a 71-year-old male with long-standing MF who subsequently developed MF associated bullae secondary to bullous pemphigoid (BP) while under no topical, systemic, photo, or radiation treatment modalities. Two bullae associated with patches of MF were biopsied. The first specimen demonstrated evidence of BP with a subepidermal split containing numerous eosinophils with background histologic findings consistent with MF. Direct immunofluorescence confirmed these findings. A second biopsy site revealed histology consistent with MF. A subepidermal split without the characteristic findings of BP was noted as well. Immunohistochemistry was consistent with MF in both samples. This is the fourth reported case of MF associated BP, but the first confirmed case in which the bullae were located only at sites of MF and without confounding treatment that may have unmasked or caused the reaction. We hypothesize that the chronic inflammatory nature of MF could lead to basement membrane zone alteration exposing potential antigens ultimately resulting in a BP type reaction.

Poster 699

RESIDENT

Comparison of virtual microscopy and glass slide microscopy among dermatology residents during a simulated in-training examination

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Virtual microscopy is being increasingly explored as a tool for resident education, board certification, and dermatopathology practice. To examine the potential utility of virtual microscopy for dermatopathology education, whole scanned slides were incorporated into a mock in-training examination given to dermatology residents in our training program. 34 residents completed a 48-question dermatopathology exam with half of the questions using glass slides and half using whole scanned

slides. Efforts were made to allocate similar case difficulty between the two modalities. Approximately half of the trainees started with glass slides first and the other half with scanned slides. The number correct for glass slides was 13.8 versus 12.6 with scanned slides ($p=0.01$). First year residents performed equally for both methods ($p=1$), while second and third residents did significantly better with glass slides ($p=0.01$). Residents with no prior virtual slide experience performed better on glass slides ($p=0.01$). Those with greater than three times per month virtual slide experience performed equally (13.3 vs. 12.9, $p=0.27$). Our results show that practice with virtual imaging leads to nearly equal performance when compared to glass slides. 16 of 35 residents preferred glass slides, while 13 preferred virtual imaging and 6 would use either. Acceptance was more universal for virtual imaging in those who did not experience computer processing delays, as occurred at one testing site. The speed and clarity of virtual images have improved greatly over the years but remain the greatest impediments to implementing virtual slide review for testing purposes.

Poster 701

RESIDENT

Superficial acral fibromyxoma

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A 61-year-old African American man presented with an 8-month history of a solitary, asymptomatic, slowly growing mass on the right thumb. Physical examination revealed a 10 x 6 mm, firm, skin-colored, exophytic, nontender, subungual nodule with upward displacement of the nail plate. Full flexion and extension at the distal interphalangeal joint were noted. Histopathological examination of a shave biopsy specimen revealed compact hyperkeratosis overlying an acanthotic epidermis. The dermis had a circumscribed population of spindle cells without high-grade atypia, embedded in a myxocollagenous stroma. The lesion was strongly positive for CD34 and weakly positive for epithelial membrane antigen (EMA), but negative for S100 protein and smooth-muscle-specific actin. The diagnosis of superficial acral fibromyxoma (SAF) was made and the patient underwent complete excision. SAF is a rare, slow-growing, soft-tissue tumor, typically located in the periungual and subungual regions of the fingers and toes in middle-aged adults. While metastasis of SAF has not been reported, the recurrence rate is up to 42%; therefore, complete excision with long-term follow up is recommended. SAF usually presents as a dermal nodule composed of spindle-shaped cells, arranged in a storiform and fascicular pattern within a myxoid or collagenous stroma having increased numbers of blood vessels and mast cells. The immunophenotype (CD34+, CD99+, EMA+/-, SMA-, S100-, cytokeratin-) is characteristic. Expression of CD10 and nestin has been reported. The differential diagnosis includes myxoid neurofibroma, superficial angiomyxoma, myxoid dermatofibrosarcoma protuberans, low-grade

myxofibrosarcoma, sclerosing perineuroma, and acquired digital fibrokeratoma. Immunohistochemistry is a useful tool to distinguish among these soft-tissue tumors.

Poster 702

RESIDENT

Perineural squamous proliferation occurring without preceding biopsy: a mimic of squamous cell carcinoma

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Perineural invasion (PNI) by primary cutaneous squamous carcinoma (CSCC), i.e. invasion of the perineural space by malignant squamous epithelium, is associated with more aggressive behavior of the carcinoma. There are, however, histologic mimics of PNI that are associated with benign lesions or reactive conditions, including re-excision perineural invasion and reparative perineural proliferation. We report a case where a striking perineural squamous proliferation was found on the initial biopsy rather than in the (re)excision specimen. An 82 year old woman presented with a pruritic 2 cm erythematous plaque on her left mid upper back. An initial punch biopsy of the plaque showed a cytologically atypical but not overtly malignant squamous proliferation encasing small nerves in the superficial dermis. Epidermal and dermal changes suggested lichenification, but no CSCC was seen in multiple tissue levels examined. Small nerve trunks were unusually prominent in the upper dermis. There was no history of previous trauma or biopsy at this site, but as perineural tracking from an adjacent carcinoma could not be excluded, an excision was performed. The excision specimen showed proliferation of small nerves in the upper dermis, associated with adnexal structures, consistent with a neural hamartoma. No CSCC was found. This case is unusual in that there was no preceding biopsy, unlike previously reported cases of "re-excision" PNI. Perineural squamous proliferation in the first biopsy may represent a response to chronic irritation in the setting of a neural hamartoma. This case expands the differential diagnosis of benign reactive conditions mimicking PNI by SCC.

Poster 703

RESIDENT

Stromal changes in lentigo maligna, superficial basal cell carcinoma, and actinic keratosis

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Introduction. Many tumors are characterized by a distinctive stroma. Stromal changes in superficial basal cell carcinoma (BCC), lentigo maligna (LM), and actinic keratosis (AK) were studied. Materials and Methods.

We evaluated 101 specimen of AK, 100 specimens of BCC, and 100 specimens of LM for stromal changes. Results. Fibromyxoid stroma was present in 82% of BCC cases. Twenty five (25%) cases of BCC had red fibrous stroma and frequently demonstrated an aggressive pattern of tumor growth. Stromal changes in LM were noted in 58 (58%) of cases and were characterized by presence of fibrous stroma that displaced the solar elastosis downwards. Sixty five (64.4%) cases of AK showed absence of characteristic stromal changes. In the remaining AK cases, a red collagenous fibrous stroma resembling that of LM was present. Conclusion. In this study, fibromyxoid stroma was specific for BCC and if present at the margins suggests incomplete tumor excision. Red fibrous stroma can be seen in BCC, LM or AK. Its presence below BCC correlates with an aggressive growth pattern. Although the differences in stromas response to LM and AK are subtle, in AK the fibrotic stroma is less pronounced than in LM and is less likely to extend beyond the tumor. Familiarity with the stromal patterns may be helpful when examining histologic sections that only demonstrate tumor stroma.

Poster 704

FELLOW

Acquired unilateral dermal melanocytosis on the back of a 79-year old caucasian man

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A 79-year old man who was a longstanding patient in our institution's dermatology clinic presented for yearly follow-up. His past history was remarkable for Grover's disease on the chest and upper back which had subsequently cleared without treatment, as well as hypertension and hyperlipidemia. On skin examination, he was noted to have a new onset bluish 7 cm ill-defined smooth patch of the left side of the upper back. The patient was unaware of the lesion, and it was not present in the prior year's skin examination. There was no history of trauma or treatment to the area. Histopathologic examination of a 4mm punch biopsy of involved skin revealed an unremarkable epidermis, and scattered dendritic and spindled pigmented cells throughout the entire reticular dermis, focally concentrated around dermal vessels. These pigmented cells did not stain with Gomori's Iron, and did stain with Fontana Masson, confirming the presence of melanin pigment. Immunohistochemical stains for MITF and S100 highlighted scattered dendritic and spindled melanocytes throughout the reticular dermis of the involved skin. These findings confirmed the diagnosis of acquired dermal melanocytosis (ADM). ADM is a rare condition that most commonly presents on the face as acquired bilateral nevus of Ota-like macules (also called Hori's nevus), and is most prevalent in Asians. Extrafacial ADM is even rarer, and its presentation in the Caucasian population is limited to a few case reports in the literature. A review of the literature identified only two cases of ADM presenting on the back of Caucasian patients.

MIXED TOPICS

Poster 705

RESIDENT

Acute Generalized Exanthematous Pustulosis [AGEP] induced by clopidogrel

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A 54 year-old female with recent myocardial infarction was hospitalized for urgent cardiac catheterization. Clopidogrel was initiated after placement of a drug-eluting stent. Five days later, widespread edematous erythematous papules and small pustules appeared on intertriginous and flexural regions of her body. This eruption was associated with low-grade fever and neutrophilia. Her liver and renal function studies were normal. Skin biopsy revealed a subcorneal collection of neutrophils, and subtle vacuolopathy in the upper dermis. Clopidogrel was discontinued, and a new antiplatelet agent, ticagrelor, was initiated. The clinical and histopathologic findings established the diagnosis of acute generalized exanthematous pustulosis (AGEP). Four weeks later, the patient developed large areas of desquamation with complete resolution of her symptoms. We report the third patient case in the literature of clopidogrel-associated AGEPEP. The findings herein should alert dermatopathologists and dermatologists about this rare adverse drug reaction, especially in high risk cardiac patients where this medication is used to prevent complications after cardiac stenting.

Poster 706

RESIDENT

Atypical POEMS syndrome with calciphylaxis

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A 31-year-old woman with a two-year history of distal polyneuropathy and mat-like telangiectasias on the feet developed pericardial and pleural effusions, hypothyroidism, hypogonadism, diabetes mellitus, mild renal insufficiency, thrombocytosis, splenomegaly, papilledema, lacunar cerebral infarctions, and refractory ascites over a five month period, resulting in hospitalization. Dermatology was consulted regarding a two-week history of painful abdominal purpura. On examination, she was a young, cachectic woman with bulging flanks and bilateral lower extremity edema. Indurated, tender purpuric plaques and eschars were present on the abdomen, and blanching telangiectatic patches were noted on her dorsal feet. Scattered on her lips, trunk, and extremities were numerous hemangiomas. An extensive work-up, including serum and urine protein electrophoresis, bone scans, skeletal surveys, VEGF, and HHV-8 serology yielded normal results. A skin

biopsy from the abdomen revealed epidermal necrosis with subcutaneous vascular calcification consistent with calciphylaxis. Bone marrow biopsy demonstrated atypical megakaryocyte hyperplasia, with flow cytometry revealing a clonal lambda-restricted plasma cell population, consistent with a diagnosis of POEMS syndrome. After treatment with cyclophosphamide and dexamethasone, the neuropathy, volume overload, and vascular proliferations improved. POEMS syndrome is a paraneoplastic syndrome caused by plasma cell dyscrasias, occurring either as plasmacytomas or as diffuse bone marrow disease. The monoclonal population involved may be quite small, and in cases with a high degree of suspicion, bone marrow biopsy with flow cytometry should be performed. The association with calciphylaxis is rare, with only five previously reported cases; nonetheless, POEMS syndrome should be considered as a potential cause of non-uremic calciphylaxis.

Poster 707

RESIDENT

Evaluation of WT-1 expression in desmoplastic melanoma and mimicker lesions

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The desmoplastic variant of melanoma (DM) presents significant diagnostic challenges due to its resemblance to scars and other spindle cell neoplasms. It also has an immunohistochemical (IHC) profile in which only S100 is consistently positive. In this context, additional markers that are sensitive and specific for DM would be a helpful addition to the diagnostic armamentarium. A total number of 14 cases of DM and 10 dermal scars from excision specimens were evaluated with WT-1 IHC. Entities characterized by increased dermal spindle cells were also analyzed: clear cell AFX (1 case), MFH (1 case), spindle squamous cell carcinoma (SpSCC) (1 case), DFSP (3 cases). Staining intensity (0-3+) and the percentage of reactive cells were scored. Other markers evaluated included S100, MART-1, and HMB-45. All DM cases demonstrated WT-1 expression. On average, 90% of tumor cells in DM were positive for WT-1 with 3+ intensity (86%) and 2+ (14%). The scars showed a range of 1-40% of spindle cells staining with the following intensities: 3+ (60%), 2+ (10%), and 1+ (30%). There were no significant differences between DMs and scars with respect to WT-1 and S100 staining patterns. DFSP and MFH showed strong cytoplasmic reactivity in all lesional cells. The single spindle cell squamous cell carcinoma showed strong reactivity, though in a nuclear staining pattern. The clear cell AFX case was negative. In conclusion, WT-1 is a sensitive marker for DM and specificity appears to be comparable with S100 in differentiation from scars.

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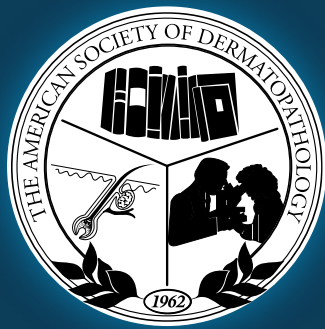
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