

Poster SESSION 2

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NEOPLASIA, CARCINOGENESIS, TUMOR BIOLOGY

Poster 500

FELLOW

Porocarcinoma arising in an apocrine poroma with follicular differentiation

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Poroma has been traditionally classified as an eccrine neoplasm, but occasional cases displaying sebaceous and/or follicular differentiation have resulted in the concept of "apocrine poroma." Although follicular differentiation is seen in apocrine poromas, carcinomas arising in this setting are rare. We describe a case of porocarcinoma arising in an apocrine poroma with follicular differentiation. A 60-year-old African-American woman presented with a 20-year history of a forehead mass that had enlarged and become painful in the past month. Clinical examination showed a 1 cm hyperpigmented, exophytic, tender, bleeding nodule. Histologically, the tumor showed predominantly classic poroma features characterized by uniform, basaloid epithelial cells with occasional ductules. Focal areas of the tumor displayed nests of basaloid epithelial cells with peripheral palisading, adjacent follicular stroma, and rare papillary mesenchymal bodies. Sebaceous differentiation was absent. A small focus of poroma cells with severe cytologic atypia, consistent with porocarcinoma, was identified at a peripheral edge of the tumor. The tumor was excised using Moh's micrographic surgery and showed residual poroma but no residual porocarcinoma. This case represents an uncommon variant of poroma and illustrates the potential for malignant transformation of longstanding apocrine poromas with follicular differentiation.

Poster 501

RESIDENT

A white spotted zebra: an unusual case of multiple tumors of follicular infundibulum

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The differential diagnosis of multiple facial bumps is wide and ranges from common lesions like acne to less common genodermatoses. We present an unusual cause of multiple painless nonpruritic hypopigmented macules on the cheeks of a 55-year-old man, present for two years. The clinical differential diagnosis included leprosy and vitiligo. The lesions were not anesthetic to touch and ranged in size from 2mm-3cm. There was no history of underlying malignancy or other cutaneous findings. Histopathological examination of two separate facial lesions showed plate-like, anastomosing cords of cytologically bland squamoid and basaloid epithelium extending from the undersurface of the epidermis. An elastin stain showed

a brush-like network of elastic fibers at the border of the tumor. These findings are consistent with tumor of the follicular infundibulum (TFI). The multiple form of this adnexal tumor has been described in only 6 % of the cases. The pathogenesis of TFI is still controversial, but the absence of the staining for epithelial antigen in this case supports that TFI is a distinct entity and not a variant of basal cell carcinoma. Some TFI have recently been proposed to represent an epidermal reaction pattern due to association with other underlying lesions in 25% of cases, and occasional absence of increased elastic fibers. However, this case of multiple lesions is not associated with underlying pathology, and shows distinct elastin staining, favoring primary neoplasia. Although rare, TFI should be considered in the differential of multiple facial bumps, with a unique and characteristic presentation as hypopigmented macules.

Poster 502

RESIDENT

A case of cytokeratin-20 negative Merkel cell carcinoma in a patient treated with infliximab

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Merkel cell carcinoma (MCC) is a rare, aggressive, neuroendocrine tumor that presents as a painless, rapidly-growing tumor in the skin of white elderly patients in the setting of ultraviolet exposure and immunosuppression. Treatment with tumor necrosis factor (TNF)-a blockers such as infliximab may be a cause of this immunosuppression. The pathologic diagnosis of MCC is based mainly on a characteristic immunohistochemical staining pattern with cytokeratin 20 (CK20). A small minority of MCCs, however, are CK20 negative, which sometimes poses a diagnostic dilemma. We present the case of a patient with rheumatoid arthritis treated with infliximab and low-dose prednisone who developed a painless, rapidly-growing lesion found to be a CK20-negative MCC. To our knowledge, this is the first report of a CK20-negative MCC diagnosed in the setting of systemic immunosuppression with a TNF-a blocker. The significance of the association between the development of a CK20-negative MCC and treatment with infliximab has yet to be determined. This case highlights the challenges in diagnosis of MCC with immunohistochemical staining as well as the risk of MCC in patients with long-standing immunosuppression, which may be due to treatment with TNF-a blockers.

Poster 503

RESIDENT

Reticulated acanthoma with sebaceous differentiation: another sebaceous neoplasm associated with Muir-Torre syndrome?

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Cutaneous lesions in Muir-Torre syndrome (MTS) are characterized by various types of sebaceous neoplasms. In 1993, Steffen and Ackerman coined the term “reticulated acanthoma with sebaceous differentiation (RASD)” to describe a histopathologically distinct sebaceous neoplasm that was first designated “superficial epithelioma with sebaceous differentiation”. To date, there has only been speculation about the relationship between RASD and MTS. We report the case of a 53 year-old gentleman who presented with a 1.0 cm yellowish plaque on his lower back that was clinically thought to be a sebaceous neoplasm. The patient also had a history of multiple sebaceous adenomas typical of MTS. His family history was remarkable for multiple colon cancers. Microscopically, the lesion showed a broad superficial epithelial neoplasm characterized by a proliferation of spinous keratinocytes and scattered clusters of mature sebocytes at the base of the rete ridges. The stratum corneum was parakeratotic and inflamed. Pseudo-horn cyst and cytologic atypia were absent. Within the papillary dermis, there were dilated capillaries admixed with mild chronic inflammation. The diagnosis of reticulated acanthoma with sebaceous differentiation was rendered. Immunohistochemically, the tumor cells in the patient’s RASD and previously identified sebaceous adenomas showed a loss of MSH6 protein expression, whereas there was no deficiency for MLH1, MSH6, and PMS2. We present this case to highlight the potential pathogenic link of RASD and MTS. This benign neoplasm which can be mistaken for various other cutaneous lesions with sebaceous differentiation deserves wider recognition for its occurrence as part of MTS.

Poster 504

RESIDENT

Primary cutaneous carcinosarcoma with unique immunohistochemistry and cytogenetic findings

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Primary cutaneous carcinosarcoma (PCC) is an exceedingly rare malignancy, infrequently reported in the literature. We present a 51-year-old male with a 3 month history of a rapidly growing hemorrhagic polypoid mass on the skin of the lower right back. Surgical excision revealed an exophytic mass measuring up to 4.0cm. Histopathologic evaluation revealed a biphasic tumor consisting of nests of baso-squamous cells with marked pleomorphism, vesicular

nucleoli and moderate mitotic activity. The stromal component was sharply demarcated from the surrounding fascicles of spindle cells and demonstrated cytologic atypia with pronounced pleomorphism and prominent nucleoli. Immunohistochemical analysis revealed the epithelial component to be strongly positive for CK5/6, CK903, and p63, as well as moderately positive for pan-cytokeratin. The spindle cell (sarcomatoid) component was positive for vimentin, and smooth muscle actin (SMA). Cytogenetic analysis performed by FISH (fluorescence in situ hybridization) revealed a loss of the long arm of chromosome 6, a loss of the short arm of chromosome 9 and monosomy of chromosomes 9 and 11. Both epithelial and mesenchymal cells demonstrated these abnormalities in similar percentages. Cytogenetic analysis of primary cutaneous carcinosarcomas has not previously been reported. We report novel cytogenetic findings which are analogous to those found in carcinosarcomas in both the lung and uterus. This provides additional support to the divergence hypothesis which posits that such tumors arise from a single totipotential stem cell that differentiates into separate epithelial and mesenchymal components.

Poster 505

RESIDENT

Atypical generalized eruptive histiocytosis clonally related to chronic myelomonocytic leukemia with loss of Y chromosome

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Generalized eruptive histiocytosis (GEH), described in 1963 by Winklemann and Muller, is a reactive and self-healing non-Langerhans histiocytosis. Rare cases of atypical GEH have been reported in patients with hematopoietic malignancy, but the biological relationship between the two disorders is not known. We report an 84-year-old man with chronic myelomonocytic leukemia who presented with coalescing erythematous papules and plaques on the Posterior neck, ear, and lower lip, followed by development of blast crisis. Skin biopsy revealed a thick band-like dermal infiltrate of cells that exhibited morphologic features of macrophages or histiocytes and prominent elastolytic phagocytosis. These cells demonstrated a mature immunophenotype, expressing CD14 and CD68, with partial expression of CD13 but not CD1a, CD43, CD56, CD123, Langerin, or S-100 protein. Karyotype and fluorescence in situ hybridization analyses showed loss of the Y chromosome in bone marrow and skin specimens, providing evidence of a clonal relationship between the cutaneous eruption and the underlying chronic myelomonocytic leukemia. The presence of the same clone in skin and bone marrow specimens from our patient supports the possibility that atypical GEH is a marker for underlying hematopoietic malignancy. Discovery of additional cases may shed further light on the pathogenesis of this rare entity.

Poster 506

RESIDENT

Squamous cell carcinoma developing in a pediatric patient with incontinentia pigmenti

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A 16 year-old caucasian girl with a clinical history of incontinentia pigmenti (IP) presented with an enlarging, tender 1.6 by 1.4 centimeter verrucous nodule arising within a previously-stable hyperpigmented streak on her left calf. On a deep shave biopsy the nodule showed compact hyperkeratosis with an irregularly acanthotic and crowded epidermis broadly infiltrating the dermis. Individual and aggregated dyskeratotic cells, squamous eddies and atypical keratinocytes with prominent nucleoli and mitotic figures were appreciated. Within the dermis was a moderate lymphocytic infiltrate without eosinophils. These features were consistent with a well-differentiated squamous cell carcinoma (SCC) and the nodule was excised using Mohs surgery. IP is an uncommon X-linked dominant disorder with mutations in the nuclear factor kappa beta (NFkB) pathway. Affected patients progress through three or four clinical phases - vesicular, verrucous, hyper- and sometimes hypo-pigmented streaks, normally stabilizing by age two. Rarely-reported late manifestations include pseudogliomas, retinoblastomas, pheochromocytomas and painful subungual fibromas. This is the fourth reported case of SCC or keratoacanthoma in a patient with IP. Two prior reports involved atypical presentations of IP with persistent verrucous nodules preceding development of acanthomas. The third case lacked a detailed histopathologic description. Our case thus provides the first clear example of invasive SCC within a stable hyperpigmented patch of IP. The mechanism of carcinogenesis in this case is unclear, particularly since IP usually induces a pro-apoptotic state in affected cells. This suggests a more complex and persistent role for NFkB and its upstream regulators in IP and in tumorigenesis.

Poster 507

RESIDENT

Low-fat (fat-free) pleomorphic lipoma: a diagnostically challenging variant

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Pleomorphic lipoma primarily occurs on the Posterior neck or shoulder of adult men. This tumor is often located in the deep dermis or subcutaneous tissue and is challenging for dermatopathologists to make the correct diagnosis, especially when little or no fat is present. In light of a relatively recent case series by Sachdeva et al, the authors report a case of low-fat (fat-free) pleomorphic lipoma mimicking ancient myxoid schwannoma. An 84-year-old female presented for a slowly enlarging subcutaneous nodule involving the hand. A clinical diagnosis of cyst was rendered and the lesion was excised. Microscopically,

the lesion was composed of bland spindle cells arranging in short parallel bundles admixed with scattered hyperchromatic multinucleated giant cells (florete cells). These cells were embedded in a myxoid stroma containing ropey collagen. In some sections, the tumor was completely devoid of mature adipocytes. The differential diagnosis considered by the referring pathologist included myxoma, ancient schwannoma, atypical lipomatous tumor, and myxofibrosarcoma. By immunohistochemistry, the majority of cells strongly expressed CD34. The keratin, desmin and actin immunostains were negative. Notably, scattered S-100 protein positive cells were admixed within the lesion. On the basis of these clinical and pathological characteristics, we diagnosed the tumor as a low-fat pleomorphic lipoma. Local recurrence of this tumor has not been observed for greater than one year. We hope that awareness and recognition of this distinct entity will lead to more accurate diagnosis and avoidance of unnecessary work-ups.

Poster 508

RESIDENT

Aggressive digital papillary adenocarcinoma

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A 58 year-old man presented to dermatology clinic complaining of a recurrent nodule on the dorsal right middle finger. The patient had an excisional biopsy 6 years prior, with subsequent near-complete resolution of the nodule after the biopsy. He then developed recurrence of the nodule with progressive growth and pain prompting presentation for re-evaluation. Examination revealed a 2 by 1 centimeter firm nodule within the prior biopsy site on the dorsum of the right middle finger near the distal interphalangeal joint. Records from the original procedure were reviewed, revealing previous pathologic diagnosis of hidradenoma papilliferum. Given the incongruence of the diagnosis of a hidradenoma papilliferum with an acral site, the original slides were reviewed and a revised diagnosis of aggressive digital papillary adenocarcinoma was made. The patient subsequently required amputation of the digit and workup to evaluate for metastasis. Aggressive digital papillary adenocarcinoma is a rare tumor thought to be of eccrine differentiation, which most commonly occurs on the digits of older men. It has high potential for local recurrence and can metastasize. These tumors are frequently misdiagnosed at the time of presentation and can be mistaken clinically for benign entities such as cysts and infections. Inexperienced pathologists may falsely lead the clinician to a benign diagnosis and delay in definitive treatment, as occurred in our case. We present the clinical and histological presentation of aggressive digital papillary adenocarcinoma and emphasize the importance of the clinical pathological correlation in order to distinguish it from other benign entities.

Poster 509

RESIDENT

Mast cell sarcoma: report of a rare case and review of the literature

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We report a case of mast cell sarcoma arising in the lower lip of an otherwise healthy 20-year-old male with distant history of cutaneous mastocytosis. Biopsy of the lesion revealed an infiltrating mass of rounded tumor cells with pale cytoplasm, nuclear pleomorphism, atypia, and prominent mitotic figures. The tumor cells were positive for CD4, CD30, CD2, CD43, CD68, MITF, EMA, CD117, and tryptase. The lesion was excised with negative margins and the patient was started on imatinib. Mast cell sarcoma is a very rare tumor classified within the spectrum of mastocytoses by WHO, however with an aggressive behavior. We have reviewed all seven cases reported in the literature. These cases have been located in the larynx, ascending colon, brain, tibia, uterus, small intestine, and skin. The age at presentation ranged from 4 to 74 years. Six cases reported no history of mastocytosis. All cases reported no history of other malignancies. Three cases reported evolution to mast cell leukemia. The survival has ranged from 14 months to four years. C-kit mutation (D816Y) and translocation FIP1L1-PDGFR were analyzed in two cases and were found negative. One case had c-kit mutation (N822K) that is associated with a good response to imatinib. The patient with the longest survival was treated with combination chemotherapy and imatinib. In conclusion, this is an example of the exceptionally rare case of mast cell sarcoma. Although there is a shortage of information on this tumor, our patient's currently successful treatment with imatinib offers beneficial information with possible future implications.

Poster 510

RESIDENT

Cutaneous squamous cell carcinoma with sarcomatous and osteoclast-like giant cell features: a case report

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Osteoclast-like giant cell (OLGC) proliferations have been reported in a variety of neoplasms involving the uterus, kidney, liver, lung, salivary gland, breast, and pancreas. Proliferations of OLGCs have been reported as a rare phenomenon in 7 cases of cutaneous squamous cell carcinoma (SCC) but their significance remains unclear. We report the case of a 92 y/o male with a 1.8 cm crusted nodule of the back. Histologically, the lesion showed a diffuse dermal proliferation of atypical polygonal to spindle-shaped epithelioid cells extending

into the subcutaneous fat. Numerous mitotic figures, some atypical, were noted. Admixed within the malignant cells were abundant multinucleated giant cells with a benign morphology resembling osteoclasts. The atypical epithelioid cells showed immunoreactivity for p63, CK-34BE12, CK-MNF116, vimentin and were negative for S-100, and HMB-45. The OLGC component reacted only with CD68 and vimentin. A diagnosis of invasive, poorly differentiated SCC with sarcomatoid features and OLGC proliferation was made. The main differential diagnosis considered in this case is osteoclast-like giant cell tumor of the skin which is histologically similar to those seen in the bone or soft tissue. However, such tumors would lack malignant features and reactivity for cytokeratins. Most reports describe OLGC proliferations in SCC to be a reactive phenomenon while others propose it to be coexistent with the sarcomatoid component of SCC. In this particular case, the lack of cytological atypia in the giant cells, absence of reactivity to epithelial markers, and immunoreactivity for histiocytic marker CD68 favor a reactive process.

Poster 511

RESIDENT

Metastatic high grade urothelial carcinoma to the scalp: a case report

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Urothelial carcinoma ranks as the fourth and tenth most common cancer in men and women, respectively. The main sites of metastatic spread of bladder carcinoma are liver, lung, and bone. Cutaneous metastasis from primary urothelial carcinoma is rare and reported in only 0.84% of patients. We report a case of a 72 year old male who underwent a radical cystoprostatectomy and pelvic lymphadenectomy for a pT4aN2 urothelial carcinoma. Five months post-surgery, he presented with a 2-cm pink nodule on the right temple and a 6-cm depressed sclerotic plaque on the left frontal scalp. Punch biopsies revealed diffuse dermal infiltrates of nested to linearly arranged atypical cells, similar in histomorphology to the primary bladder carcinoma. A panel of immunoperoxidase stains (positive for 34BE12, CK7, CK20, p63, Uroplakin III) was consistent with metastatic urothelial carcinoma and the patient was referred for chemotherapy. Very few cases of cutaneous metastases of bladder carcinoma are reported in the literature. The anogenital and abdominal areas are the most common sites, probably resulting from direct seeding of cancer cells. Scalp metastasis is very rare and has been described for large cell neuroendocrine carcinoma of the bladder but not for transitional cell carcinoma. The clinical appearance of cutaneous metastases of urothelial carcinoma is not distinctive and may resemble various common dermatologic disorders. Lesions have been reported to be nodular, inflammatory,

sclerodermoid, and even vascular in presentation. It is important to recognize cutaneous metastatic lesions as they often represent an early sign of generalized recurrent disease.

Poster 512

RESIDENT

Neurofibroma with floret-like multinucleated giant cells in a patient with no known history of neurofibromatosis

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Neurofibroma is a benign nerve sheath tumor with many variants. One rare type contains floret-like multinucleated giant cells (FMGCs) but otherwise has the usual neurofibroma histological features. Less than 50 of these are reported in the literature. Since many were removed from patients with neurofibromatosis, an association has been suggested. Two studies have addressed this idea, but much about the topic remains unknown. The current patient is a 74-year-old African American male with no known history of neurofibromatosis who presents with a longstanding skin-colored to depigmented and pedunculated left temple papule. Histology shows neurofibroma with numerous FMGCs. Immunohistochemical stains show spindle cells positive for S-100 and vimentin but negative for CD34 and CD68. The FMGCs are positive for vimentin and CD34 but negative for S-100 and CD68. This histology and staining pattern are consistent with past reports of neurofibroma with FMGCs. Although this patient has no history of neurofibromatosis, this tumor type has been described in similar patients but to a lesser degree than in those with neurofibromatosis. The differential includes two fibroblastic tumors: giant cell fibroblastoma (GCF) and pleomorphic fibroma (PF). PF is a benign fibroblastic tumor with pleomorphic, hyperchromatic cells or giant multinucleated cells. The S-100 negative spindle cells differentiate it from neurofibroma with FMGCs. GCF contains FMGCs, but unlike neurofibroma with FMGCs, it has pseudovascular spaces and onionskin pattern perivascular lymphocytes. This is important because GCF has intermediate malignancy. Neurofibromas are benign tumors treated with excision, and the FMGC variety is assumed to follow the same course.

Poster 513

RESIDENT

Eccrine spiradenoma, cylindroma and chondroid syringoma coexistent in the same lesion

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A 57 year old man presented with a 20 year history of a dermal nodule that had enlarged to 3 cm over 2 months on

his left upper back clinically thought to be an epidermoid cyst. An excisional biopsy revealed a proliferation of nodules of basaloid cells with areas of ductal differentiation and areas of hyaline pink deposits. In one area there were two cell types: cells with larger pale nuclei, and cells with smaller hyperchromatic nuclei. There were other areas composed of smaller islands of cells comprised of the same two types and hyaline deposits enveloped by dense eosinophilic cuticles and areas of ductal differentiation. In addition there were areas of myxoid and hyaline stroma with thin cords and islands of monomorphic basaloid cells with areas of ductal differentiation. This adnexal neoplasm had areas of spiradenoma, cylindroma, and chondroid syringoma (mixed tumor). One previous case of concomitant spiradenoma and chondroid syringoma has been described in the literature and one case of concomitant spiradenoma and malignant mixed tumor. In two cases chondrosarcomatous differentiation has been reported. Spiradenomas and cylindromas are not uncommonly present within the same lesion and may share a mutation in the CYLD gene, but the combination of either with a chondroid syringoma is rare. The etiopathogenesis of all three tumors is hotly debated with arguments for eccrine versus apocrine nosology. This case is interesting because of the three different adnexal neoplasms represented, suggesting a possible common histogenesis.

Poster 514

RESIDENT

A digital papillary adenocarcinoma presenting as a bony metastasis

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Digital papillary adenocarcinoma is an uncommon tumor presenting as a painless nodule on distal extremities; most commonly on fingers or toes. Histopathology shows a poorly circumscribed, dermal and subcutis-based lesion with tubuloalveolar and ductal structures. Within cystic lumina, protruding papillary projection can be seen. However, a cribriform pattern without papillary projections may be observed. Mitotic activity is variable. Additional findings may include: Poor glandular formation, necrosis, bone and soft tissue invasion, and nuclear pleomorphism. Immunohistochemical studies are positive for S100, CEA, and cytokeratins. Biologic behavior cannot be reliably predicted based on histology or clinical characteristics. Here, we present a 34-year-old male whom presented with a left calcaneus bone mass. X-ray revealed a lytic lesion which was subsequently biopsied to reveal a neoplastic process with both glandular and squamous pearl-type formation. The background stroma had focal hyalinization and small round cells. CEA staining showed patchy positivity, favoring a metastatic eccrine carcinoma. On follow-up, a nondescript soft tissue tumor was identified on the left medial great toe, and a punch biopsy was submitted for histopathologic

evaluation. Microscopic sections revealed a lesion with features identical to the bone lesion. A diagnosis of Digital Papillary Adenocarcinoma was rendered. The tumor was composed of ducts lined by cuboidal cells with a papillary architecture present in some areas. Cystic spaces were interposed between and around the more solid areas. The mitotic rate was three per square millimeter. Large areas of necrosis and bony destruction were seen histologically in the subsequent BKA specimen.

Poster 515

RESIDENT

Panfollicular carcinoma

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Panfolliculoma is a term used to describe benign follicular neoplasms with differentiation towards both the upper and lower segments of the hair follicle. We report here two cases of carcinoma with panfollicular differentiation. First patient was a 48-year old male who presented for evaluation of a 1 cm by 0.4 cm pink papule of the nose. A shave biopsy revealed intradermal basaloid islands with cytologic atypia and nuclear pleomorphism, as well as cysts with matrical differentiation and inner root and outer root sheath differentiation. The lesion infiltrated into the deep dermis. The second patient was a 77-year old male who had a shave biopsy of a 0.4 cm pink papule on the left helical rim to rule out a basal cell carcinoma. Similarly to the first patient, H&E revealed basaloid islands, cords and strands extending deep into the dermis, exhibiting both inner and outer root sheath differentiation and high mitotic index. Complete re-excision through Mohs was achieved in both cases. To our knowledge, these are the first cases of this unique carcinoma reported to date.

Poster 516

RESIDENT

Penile fibroepithelioma of Pinkus and basal cell carcinoma: two case reports and literature review

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Basal cell carcinoma (BCC) is the most common cause of skin cancer and is usually seen on sun-exposed sites. BCC are subtyped depending on the growth pattern including nodular, superficial, infiltrative, micronodular, and depending on the author can include the fibroepithelioma of Pinkus (FEP). Only one case of penile FEP has been described in the literature. We present the clinicopathologic features of two cases of single, penile lesions clinically thought to be Human Papilloma Virus-associated, non-melanocytic skin cancer; however, histologically, the tumors had palisading basal cells, clefting, and mucin pools, all classic features of

basal cell carcinoma. Penile BCC are rare and usually the nodular subtype. The first case is a 76 year old with a shave biopsy showing a well-circumscribed, reticuloform lesion originating from the basal keratinocytes. On high power, thin vascular septae with peripheral palisading cells, clefting and mitoses were seen consistent with a FEP. The second case is a shave biopsy from a 69 year old with an erythematous papule on the left penile shaft showing similar histologic features with the classic nodular pattern. Penile BCC's are extremely rare, estimated at less than 0.25% of all cases of basal cell carcinoma. FEP was first described as a premalignant fibroepithelial tumor of the skin by Herman Pinkus in 1953; however, recent evidence supports the lesion to be malignant with loss of TP53 and dysregulation of the PATCH gene. Although rare, FEP/BCC do occur on the penis; therefore, early diagnosis is imperative for the possibility of conservative treatment options.

Poster 517

RESIDENT

A novel case of combined basal cell carcinoma, clear cell syringoma and common blue nevus

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Basal cell carcinomas, clear cell syringomas and blue nevi are all skin tumors that usually occur as solitary lesions. We encountered a novel case of a single cutaneous lesion with these three different tumors occurring together. An 81-year-old Asian woman presented with a 4 month history of a pruritic lesion on the left cheek. She denied any personal or family history of skin cancer. Physical examination revealed a 10 x 8 mm light blue/brown dusky thin plaque without surface changes on the left cheek. The patient had a past medical history significant for diabetes mellitus. A shave biopsy was performed and the histopathological analysis revealed a combined tumor consisting of basal cell carcinoma, clear cell syringoma and common blue nevus. The three distinct intermingled neoplasms consisted, respectively, of small basaloid islands with scant peritumoral mucinous stroma, epithelial strands and duct-like structures lined by a two cell layer of cuboidal epithelial cells with clear cytoplasm, and a proliferation of slender pigmented dendritic melanocytes (confirmed by positive Mart-1 immunohistochemical stain) admixed with melanophages. While combinations of two of these tumors have been previously reported, to our knowledge, this is the first time that all three have been found to coexist in the same location.

Poster 518

An aggressive clinical presentation of a relatively uncommon benign adnexal neoplasm - eccrine syringofibroadenoma

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A 57 year-old African American male presented to our clinic with a four year history of a slow growing, asymptomatic lesion on his left foot. His medical history was remarkable for diabetes, peripheral neuropathy and renal insufficiency and surgical history for amputation of his first three toes of the left foot secondary to osteomyelitis in 2008. Physical examination revealed an impressive diffuse cerebriform tumor involving 75% of the dorsal aspect of the left foot with focal ulceration, hyperkeratosis and yellow crusting. Microscopic examination of an initial shave biopsy demonstrated anastomosing strands of uniform epithelial cells extending from the base of the epidermis into the mid-dermis with fibrosis, containing a moderately dense, superficial, perivascular lymphoplasmacytic infiltrate. A wedge biopsy three months later to rule out malignancy showed histopathologic findings similar to those observed in the initial biopsy. Eccrine syringofibroadenoma (ESFA) is a rare benign tumor of the acrosyringium, with approximately 70 reported cases to date. It usually presents as a solitary plaque located on an extremity and may occur spontaneously, or can be associated with inherited and acquired conditions. Of the five different variants described, reactive ESFA can be associated with lichen planus, elephantiasis, HPV type 10, bullous pemphigoid, and diabetic neuropathy (which is most likely the case in our patient). Malignant transformation into squamous cell carcinoma or clear cell acanthoma have been reported. While there are only a few case reports of ESFA in patients with diabetes and peripheral neuropathy, this case represents a florid clinical presentation of ESFA.

Poster 519

RESIDENT

Solid carcinoma: a rare skin adnexal neoplasm revisited

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BACKGROUND: Primary apocrine or eccrine carcinomas are rare and their nosology is still evolving. First described in 1997, solid carcinoma is now considered a discrete entity and apocrine origin has been postulated. We report an additional case of solid carcinoma, the second including its immunohistochemical profile. CASE REPORT: A 63 year old male presented with a firm 3 cm diameter

Posterior scalp nodule. A punch biopsy revealed a skin adnexal neoplasm of indeterminate malignant potential, not specifically classified. The subsequently excised tumor extended widely to the surgical margin, exhibited perineural invasion and was interpreted as a microcystic adnexal carcinoma. Re-excision yielded clear surgical margins. RESULTS: Histologically, innumerable solid aggregations of neoplastic epithelial cells filled the dermis and extended into the subcutis. The aggregations varied in size and ranged from round or ovoid nests to elongated columns or cords. Deeper aggregations were generally smaller. The stroma was fibrotic and variably cellular. The neoplastic cells were round and uniform in size, with monomorphous nuclei and pale or clear cytoplasm. Tubular structures and mitoses were rare to absent. Perineural invasion was present. The neoplastic cells expressed cytokeratin and p63 but not GCDFP-15 or S100. CONCLUSION: Solid carcinoma is a rare but distinctive skin neoplasm. Apocrine origin is postulated but still debated. Biologically, this is an indolent but relentless locally destructive neoplasm that must be removed completely. Mohs micrographic surgery therefore appears to be the treatment of choice.

Poster 520

RESIDENT

Squamous cell carcinoma with nodal metastases arising in a proliferating trichilemmal cyst

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A 64-year-old male presented with a large scalp lesion. Imaging studies revealed a 2.5 x 1.4 cm mass within the subcutaneous soft tissue over the left occipital bone, and a wide and deep resection was performed. On histologic examination the tumor consisted of dermal nodules of squamous epithelium, many of which formed cysts with trichilemmal keratinization associated with necrosis and parakeratotic cells. In multiple foci, there were abrupt transitions from cyst walls comprised of keratinocytes with mild cytologic atypia to those with marked cytologic atypia and numerous mitotic figures. No connection to the epidermis was seen. A diagnosis of moderately to poorly differentiated carcinoma in association with a proliferating trichilemmal (pillar) cyst was made. Following additional imaging studies, the patient underwent a further neck and scalp resection with bilateral lymph node neck dissection. Histologic sections revealed a poorly differentiated carcinoma, associated with necrosis, involving subcutaneous adipose tissue and underlying skeletal muscle. Multiple lymph nodes were positive for metastatic carcinoma. Proliferating trichilemmal (pillar) cyst (PTC) is a tumor with differentiation towards the outer root sheath of the hair follicle. Recurrence following complete surgical excision and malignant transformation are rare with metastatic spread described in single case reports. A unique feature of this case was the very abrupt transition from mildly atypical squamous epithelium to squamous epithelium exhibiting marked nuclear atypia and hyperchromasia, atypical mitoses, and geographic

necrosis. This case illustrates the importance of careful sampling and thorough evaluation of proliferating pilar cysts for the possibility of a malignant transformation.

Poster 521

RESIDENT

Tricholemmal carcinoma in situ involving a tricholemmal cyst

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An 89-year old male presented with a clinically cystic 4 mm papule on the left temple. The clinical impression was a benign cyst. Pathologic examination revealed a small dermal-based cystic lesion with markedly atypical-appearing clear to squamoid cells lining the cyst wall, consistent with carcinoma in situ involving the cyst. The cells showed abundant glycogen-containing cytoplasm (confirmed by PAS stains with and without diastase), consistent with tricholemmal differentiation, and areas of tricholemmal/pilar type keratinization (without a granular layer) were also present, consistent with tricholemmal carcinoma in situ. Ki-67 and p53 immunohistochemical stains were strongly positive (with greater than 20% of nuclei staining on Ki-67, and > 80% on p53) in the cyst-lining cells, further supporting the interpretation of carcinoma in situ. Multiple deeper level sections were examined, but did not show any evidence of an associated invasive carcinoma. Tricholemmal (pilar) cysts are common benign adnexal tumors, and atypia/dysplasia or carcinoma in situ arising within them is exceedingly rare. Previously, only one case of a tricholemmal cyst with carcinoma in-situ has been reported. That case was associated with an atypical fibroxanthoma. We report the second case of (tricholemmal) carcinoma in-situ involving a tricholemmal cyst, which was not associated with another tumor or evidence of invasive carcinoma.

Poster 522

RESIDENT

Recurrent atypical fibroxanthoma with satellite metastasis

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Atypical fibroxanthoma (AFX) is a tumor of uncertain etiology that develops on sun-exposed regions of elderly males. It is widely considered to act indolent clinically, despite its highly atypical cytologic features. Reports of metastatic AFX are rare in literature, and recurrence is uncommon. We report a case of recurrent AFX exhibiting satellite metastases. A 76-year-old male presented with recent growths of multiple red, nodular, and well-circumscribed lesions each measuring 1.2 cm in a radiating pattern on left scalp vertex. The site was treated twenty

months prior for AFX with Moh's micrographic surgery. Pertinent history includes occupational radiation exposure, and directed radiation treatments of the right face and ear. MRI and PET imaging revealed no distant metastases or calvarial involvement. Wide local excision of the area showed multiple, expansive, well-defined nodules involving reticular dermis, subcutis, and galea. Pleomorphic epithelioid and spindled cells with marked nuclear atypia and hyperchromasia were seen with conspicuous mitotic figures. Small and distinct satellite nodules around the periphery of the main tumors were identified. No evidence of necrosis, vascular or perineural invasion was seen. Immunohistochemistry was positive for vimentin, and negative for pancytokeratin, cytokeratin 5/6, S100, and CD34. Previous biopsies showed similar histologic and immunophenotypic features. Diagnosis of AFX exhibiting satellite metastasis was favored over a high-grade sarcoma after consideration of both clinical and pathologic history. We report a previously undocumented behavior of a recurrent AFX with satellitosis, which falls somewhere in the middle of an ever widening spectrum of documented AFX behaviors ranging from indolent to malignant.

Poster 523

RESIDENT

Sebaceous carcinoma of the vagina/vulva

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We present a 70-year-old female with a vaginal/vulvar lesion. A 0.5 x 0.4 x 0.2 cm unoriented soft tissue specimen was submitted for pathologic diagnosis. The biopsy specimen shows a rare neoplasm that displays both features of a squamous cell carcinoma or basal cell carcinoma, and distinct sebaceous gland differentiation. This neoplasm resembles the sebaceous carcinomas seen on the face of eyelid. Extraocular sebaceous carcinomas are uncommon neoplasms. It is known that sebaceous glands are present on the vulva; however cases of sebaceous carcinoma in the vulva are exceedingly rare. Due to the small number of reported cases of sebaceous carcinoma of the vulva, definitive information regarding prognosis and optimal treatment has not been established. The few cases that have been reported support the general opinion that this neoplasm is aggressive and metastatic disease carries a poor prognosis.

Poster 524

RESIDENT

EGFR and MYC gene copy number aberrations are less common in keratoacanthoma than squamous cell carcinoma

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Background: Epidermal growth factor receptor (EGFR) and MYC genomic aberrations have been described in cutaneous squamous cell carcinoma (SCC) but have

not been widely investigated in keratoacanthoma (KA). Methods: Two tissue microarrays were created containing 2-mm cores of 50 KAs, 20 SCCs, and 10 verrucae vulgares. EGFR and MYC gene copy number aberrations and amplification were evaluated by fluorescence in situ hybridization. EGFR and MYC protein expression was evaluated by immunohistochemistry. Results: EGFR gene copy number aberrations were seen in 39% of KAs, 50% of atypical KAs, and 74% of SCCs. Increased MYC copy number was observed in 30% of KAs, 67% of atypical KAs, and 89% of SCCs. MYC gene amplification was more common in SCC than KA, while EGFR gene amplification was rare and not significant. MYC protein overexpression was more common in SCC than KA, but there was no difference in EGFR protein overexpression in SCC and KA. Conclusions: Genetic differences exist between KA and SCC. Greater differences were observed in MYC assays than EGFR assays, suggesting MYC may better differentiate KA and SCC. Cases of prototypic KA are best reported as KA rather than as a variant form of SCC.

Poster 525 **RESIDENT**

Alopecia neoplastica in a 36 year-old man: syringoid eccrine carcinoma extensively infiltrating the scalp

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Syringoid eccrine carcinoma is a rare tumor, usually of the scalp, presenting as a slow growing plaque. Local recurrence after excision is quite frequent. Freeman and Winkelman originally described this entity as a basal cell tumor with eccrine differentiation. However, given its cytologic appearance and the fact that the tumor cells contain eccrine enzymes, the lesion was later given its current designation. Here, we present a 36-year-old male with an eight month history of a scalp plaque associated with no hair growth. A punch biopsy was performed to reveal a deeply infiltrative eccrine neoplasm, showing little maturation and less lumen formation than a syringoma. The histologic findings were consistent with a syringoid eccrine carcinoma. Excision of the lesion showed perineural invasion and positive deep resection margins. Two subsequent re-excisions of the lesion were performed, both revealing positive margins and extensive perineural invasion. Histopathology of this uncommon tumor resembles a syringoma, composed of numerous tubular structures lined by atypical basaloid cells of one or more layers. In addition, this lesion may display ductal, cystic, and comma-like epithelial components. Yet syringoid eccrine carcinomas differ from syringomas by the presence of an infiltrative growth pattern and anaplasia. Additionally, the lesion differs from microcystic adnexal carcinoma (MAC) in that MACs display squamoid features and a desmoplastic stroma, whereas syringoid eccrine carcinomas have a basaloid cell pattern and lack of stromal response. The tumor occurs within the dermis and often

extends into the deep subcutis. Perineural invasion is not uncommon.

Poster 526 **RESIDENT**

Kaposi sarcoma resembling dermatofibroma - a report of two cases

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Kaposi sarcoma (KS) is a vascular neoplasm associated with HHV-8. We present two cases of KS with unusual histopathological features. The first patient is a 27-year-old man with intermittently treated HIV who was admitted to the inpatient service with shortness of breath and several months history of scattered purple patches and plaques with facial and lower extremity edema. CD4 count was 113 and viral load 724,541. The second patient is a 47 year old man with untreated HIV who presented with a diffuse pruritic eruption of one year duration; according to the patient, viral load was undetectable and CD4 count was not known. On exam, the patient was found to have scattered violaceous flat topped papules and plaques on the upper extremities; on the lower extremities, there were erythematous scaly hyperpigmented plaques. Punch biopsies from the right arm of the first patient and the left wrist of the second patient were performed. Both biopsies show a perivascular, periadnexal and interstitial proliferation of spindle-shaped cells throughout the dermis. Interstitial cells were scattered between collagen bundles. Few slit-like spaces with extravasated erythrocytes and adjacent siderophages, highlighted with the Perls' stain, were appreciated. With immunostains for CD31 and D2-40, the spindle-shaped cells were positive and demonstrated many vascular channels with compressed lumina. Some of the spindle cell nuclei were positive with HHV-8. These two cases are unusual because on routine histology the vascular channels were inconspicuous and the spindle cells mimicked fibroblasts. Immunostains were helpful in finalizing the diagnosis.

Poster 527 **RESIDENT**

Ki-67 and p16 staining characteristics of trichilemmal cysts and proliferating trichilemmal tumors

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INTRODUCTION: Trichilemmal cysts (TC) are benign cystic tumors which recapitulate the isthmus-catagen portion of hair follicles. Proliferating trichilemmal tumors (PTT) can arise de novo or in association with a TC and can be locally aggressive. This study evaluates the expression of

p16 and the ki-67 proliferation rate to determine if there are differentiating characteristics. METHODS: 24 TCs and 13 PTTs were stained for p16 and ki-67. Quick score evaluation of p16 staining (negative = 0, weak = 2-6, moderate = 8-10, strong = 12,15,18) and ki-67 proliferating percentages were tallied. RESULTS: 19 of 24 (79%) cases of TC demonstrated negative (2) or weak staining (17) for p16. 5 of 24 (21%) demonstrated moderate staining. There were no strong staining cases. 7 of 13 (54%) of PTT demonstrated strong p16 staining. The remaining 6 cases demonstrated moderate (3) or weak (3) staining. At least one focus of strong full thickness staining was seen in 12 of 13 cases of PTT. Total ki-67 proliferation rates were not statistically different, 7.4% in TC and 8.1% in PTT. However, proliferation rates above the basal layer demonstrated a significant difference statistically ($p < 0.001$) with 0.5% proliferation rate in TC and 2.4% in PTT. CONCLUSION: 1. Strong full thickness p16 staining was seen only in PTT. 2. Exclusion of basal layer ki-67 staining, demonstrated significantly different proliferation rates (0.5% versus 2.4%) between TC and PTT 3. Full thickness p16 expression and increased suprabasal Ki67 proliferation rate may assist in differentiating borderline cases of PTT arising in TC.

Poster 528

RESIDENT

Cutaneous lymphadenoma: a case report

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Originally referred to as benign lymphoepithelial tumor of the skin, cutaneous lymphadenoma is an adnexal neoplasm with distinct morphologic and immunohistochemical profiles. It is a slow-growing tumor predominantly found on the head and neck of young to middle-aged adults. Although it has been well documented, this report includes a photograph of the clinical presentation. We present a 49-year-old man with a long-standing (at least 16 years), stable, firm, flesh-colored papule located behind the earlobe that showed focal telangiectasia. The clinical differential included intradermal nevus, dermatofibroma, and basal cell carcinoma. Histologically, the neoplasm is well circumscribed and unencapsulated with a collection of basaloid nests set in a dense collagenous stroma. The basaloid nests are further characterized by a significant lymphocytic and histiocytic infiltrate. The histogenesis still remains uncertain. Excision is curative.

Poster 529

RESIDENT

Multiple collagenomas as a sign of multiple endocrine neoplasia type 1 (MEN1)

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Multiple endocrine neoplasia type I (MEN1) is an autosomal dominant hereditary cancer syndrome characterized by parathyroid adenomas, pancreatic neuroendocrine tumors, and pituitary adenomas. Benign cutaneous hamartomas, most commonly angiofibromas, collagenomas, and lipomas, are also associated with MEN1. Herein, we describe a 36 year old male with a history of recurrent kidney stones related to hypercalcemia who presented with a 2.3 cm enhancing pancreatic tail lesion and multiple "skin tags" of the anterior and Posterior trunk, neck, and face. Upon further questioning, the patient revealed a long-standing history of hypertension and recurrent hyperparathyroidism, as well as an extensive family history of hyperparathyroidism and a pituitary tumor in his mother. Laboratory studies revealed an elevated calcium level of 11.4 mg/dl (normal range = 9-11 mg/dl), parathyroid hormone level of 128.8 pg/ml (normal range = 10-65), and prolactin level of 185.5 ng/ml (normal range = 2.6 - 13.1 ng/ml). MRI of the pituitary revealed a 1.3 cm heterogeneously enhancing mass, which coupled with the patient's elevated prolactin levels, was consistent with a prolactinoma. An endoscopic ultrasound with fine needle aspiration of the pancreatic mass revealed atypical cells positive for synaptophysin, chromogranin A, and CD56, suspicious for a pancreatic endocrine tumor. Biopsy of the "skin tags" revealed dense collagen bundles consistent with collagenoma. This constellation of findings supported the diagnosis of MEN1 in this patient. We review the cutaneous findings of MEN1 and highlight the importance of recognizing these lesions as they may suggest a diagnosis of MEN1 prior to the onset of endocrine symptoms.

Poster 530

RESIDENT

Large glomus tumor with oncocytic and clear cell change mimicking an adnexal neoplasm

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A 30-year-old male presented with a painful, 4 cm left plantar foot mass. Excisional biopsy revealed a multinodular dermal and subcutaneous neoplasm composed of uniform, basaloid cells at the periphery of the nodules transitioning to oncocytic, epithelioid cells with focal clear cell change in the central portions of the nodules. Although the tumor showed remarkable similarity to an eccrine poroma or dermal duct tumor, no definitive duct formation was identified. The tumor cells displayed focal but strong expression of smooth muscle actin but were negative for cytokeratin (AE1/AE3) and S-100

protein by immunohistochemistry. The immunophenotype confirmed the diagnosis of glomus tumor with oncocyctic change. Re-excision showed no residual neoplasm. Glomus tumors are neoplasms that resemble the normal glomus apparatus. They represent less than 2% of soft tissue tumors and typically present as a solitary mass in the extremities, usually in the subungual region of the fingers. Oncocyctic change in glomus tumors is a recognized but uncommon finding that often results in diagnostic confusion with adnexal neoplasms. Clear cell change is an exceedingly rare finding in glomus tumor that has not been reported in the literature to our knowledge. The presence of both of these features in our case caused diagnostic confusion with an adnexal neoplasm. However, the absence of duct formation prompted immunohistochemistry to differentiate between glomus tumor and an adnexal neoplasm. This case demonstrates that glomus tumors can show both oncocyctic and clear cell change and bear striking resemblance to adnexal tumors.

Poster 531

RESIDENT

PAX2 expression in hidradenomas with clear cell features: a potential morphologic and immunohistochemical mimic of renal cell clear cell carcinoma

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Background: When presented with an epithelial malignancy in the dermis, the differential always includes metastatic carcinoma. Renal Cell Carcinoma (RCC) is well known to have metastatic potential to a wide variety of organs. Though relatively uncommon, RCC has been shown to metastasize to the skin, and the head and neck region is the most common site. These metastatic foci recapitulate the morphology seen in primary kidney tumor, with nests of cells with clear cytoplasm imbedded in a delicate vascular network. Since the incidence is low, metastatic RCC is typically not on the top of the differential when presented with a dermal based epithelial lesion composed of clear cells. Nodular hidradenomas (eccrine acrospiroma) can have similar morphology, especially the clear cell variant. Purpose: Arriving at the correct diagnosis is crucial to proper patient management and this may require additional studies such as immunohistochemical stains. PAX2 is a well known marker for RCC that has previously been undescribed in nodular hidradenomas. We study the expression of PAX2 in hidradenomas. Material and Methods: We studied a series of 12 cases of nodular hidradenomas with clear cell features and renal cell carcinoma as control and immunostain them with PAX2 and other markers for adnexal epithelium. Results: All hidradenomas stain uniformly for PAX2. Conclusions: PAX2 cannot be used to reliably distinguish hidradenoma, a benign adnexal tumor, from metastatic RCC. We present a panel of markers that can be used to confidently make the distinction between hidradenoma with clear cells and metastatic RCC.

Poster 532

RESIDENT

Primary cutaneous apocrine adenocarcinoma

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A 50 year old male presented with a three year history of multiple tender "boils" in the left axilla that progressed to increase in size, number, pain and drainage associated with a 20 lb weight loss. He failed to respond to several I&D and antibiotic courses. Skin biopsy confirmed apocrine adenocarcinoma. He had normal mammogram, GI endoscopy and PSA level. PET/CT showed several enhancing 2 cm hypermetabolic cutaneous and subcutaneous lesions in the left axilla. He received neo-adjuvant radiation followed by wide local excision with left axillary lymph node dissection. Six out of 13 nodes were positive. Six months later, he re-presented with a new lesion on the left anterior shoulder and was found to have disease recurrence. The repeat PET/CT showed 20 new right base pulmonary nodules. Cutaneous apocrine adenocarcinoma is a rare malignant sweat gland tumor. 186 cases were identified in 17 SEER database from 1973-2006. The annual incidence is 0.0049-0.0173/100,000. Commonly involved sites include axillae (53%), head and neck, nipple and anus. This is a slow-growing cancer but often has metastases by the time of diagnosis because they were frequently mistaken for abscesses or keloids. Most of these malignant tumors are believed to develop de novo from normal apocrine glands. Evidence of decapitation secretion seen on H&E is a useful clue to distinguish apocrine origin. These tumors are positive for cytokeratin7, gross cystic disease fluid protein-15 and androgen receptors. Primary cutaneous apocrine adenocarcinoma has a high recurrence incidence, like our case, due to the delayed diagnosis.

Poster 533

FELLOW

Angiomatous Kaposi's sarcoma: a variant that mimics hemangiomas

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We describe 14 cases of angiomatous Kaposi's sarcoma (KS), a distinct histological variant of KS first mentioned by Ackerman and Gottlieb in 1988. Intriguingly, so far this variant of Kaposi's sarcoma has not attracted much attention and has not been studied in detail. Its hallmarks are dilated vessels with round lumens, stuffed with erythrocytes, and in many cases, fewer spindle cells than in conventional KS. The lesions can easily be mistaken for hemangiomas, especially if the history does not alert the pathologist to risk factors for KS. Immunohistochemistry showed prominent D2-40 staining of the neoplastic vasculature, but not the pre-existing vessels, suggesting lymphatic differentiation despite the erythrocyte filled, round lumens. This observation provoked consideration as

to how connections between lymphatics and blood vessels play a role in the pathogenesis and progression of Kaposi's sarcoma. To test whether D2-40 staining of round vessels with erythrocytes was distinctive, we stained sinusoidal hemangiomas and cellular angiolipomas, both of which have these structures, and contrast the findings in KS with these.

Poster 534

Inguinal endometriosis-associated clear cell adenocarcinoma

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Endometriosis is a gynecological medical condition in which endometrial tissues of uterine corpus appear outside the uterine cavity, most commonly in the ovaries. Inguinal endometriosis is rare, occupying less than 1% of all cases of endometriosis. Secondary tumors arising from endometriosis have been reported in various organs, although most occur in the ovaries. We present a case of clear cell adenocarcinoma arising from inguinal endometriosis. Clear cell adenocarcinoma is known as an ovarian malignant neoplasm, most of which cases are associated with ovarian endometriosis. The patient is a 64-year-old Japanese female with a 3-year history of right inguinal mass. She had a left mastectomy for breast cancer 28 years ago, a total hysterectomy for adenomyosis 14 years ago, and a colectomy for colon cancer 6 months ago. Her inguinal mass had gradually grown to over 10 cm. Systemic examination including CT and PET showed no metastasis. Since skin biopsy did not lead to a definite histological diagnosis, tumorectomy was performed. The tumor size of the specimen was 13 x 10 x 8 cm. The tumor had two skin ulcers and was attached to the right inguinal canal. Histology showed a well-circumscribed multinodular neoplasm in both dermis and subcutis. The neoplastic cells are flat or cuboid with pale cytoplasm, and atypical nuclei formed numerous dilated lumens, in a so-called tubulocystic pattern. Endometriosis coexisted in the stroma of the neoplasm and in the tissue of the inguinal canal.

Poster 535

RESIDENT

Reactive vs neoplastic atypia in epidermal lesions

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Introduction: Distinguishing reactive atypia from neoplastic atypia is sometimes challenging. We sought to compare the incidence and severity of cytologic atypia in malignant epidermal lesions, benign epidermal proliferations with inflammation, and inflammatory skin disorders. Materials and Methods: 101 biopsies were retrieved and the epithelial cells were evaluated for diffuse solid nuclear hyperchromasia, crowding, irregular nuclei, high nuclear/

cytoplasmic (N/C) ratio, enlarged central nucleolus, single vs multiple nucleoli, irregularity of nucleoli, hyperchromatic parakeratosis, mitotic figures, necrotic nuclei and staining quality of the cytoplasm. Cytological features were graded on a semiquantitative scale from 0 to 4. Results: Nuclear hyperchromasia, crowding, irregular nuclei, high N/C ratio, a large single prominent nucleolus, coarse parakeratosis, atypical mitotic figures, eosinophilic color of cytoplasm and glassy eosinophilic cytoplasm were statistically more frequent in malignant neoplasms. Atypical mitotic figures and glassy eosinophilic cytoplasm were uncommon findings, but were only noted in malignant neoplasms. Among the more frequently seen features, irregular nuclei, crowding, high nuclear-cytoplasmic ratio, enlarged prominent nucleolus were the most predictive for malignancy. Conclusion: We conclude that in the setting of an atypical cutaneous squamous proliferation, irregular nuclei, crowding and atypical mitotic figures along with diffuse nuclear hyperchromasia, high N/C ratio, enlarged single prominent nucleolus, coarse parakeratosis, color of cytoplasm and glassy eosinophilic cytoplasm are reliable cytological indicators of malignancy. In contrast, the mitotic rate, presence of multiple nucleoli, irregular or crumbly nucleoli, clumping of chromatin, and presence of central small nucleoli are not helpful in this distinction.

Poster 536

RESIDENT

Indeterminate cell histiocytosis: two cases of a rare dendritic cell tumor

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Indeterminate cell histiocytosis is a rare cutaneous neoplasm composed of a proliferation of dendritic cells that share immunophenotypic features of Langerhans cells (S-100+ and CD1a+) but lack Birbeck granules. They may also stain with some macrophage markers, such as CD68. We report two cases of this uncommon disease, demonstrating its wide range of biologic behavior. One case is a 37-year-old female presenting with a 3 cm pretibial nodule and inguinal lymphadenopathy. Histologic sections showed a dense dermal proliferation of enlarged epithelioid cells with eosinophilic cytoplasm and irregular nuclei. There were numerous mitoses and foci of necrosis. By immunohistochemical analysis, the infiltrate was positive for S100 and CD1a, dimly positive for CD68, and negative for other markers including CD3, CD20, and Mart-1. A Langerin stain was negative, suggesting lack of Birbeck granules. A second case is of a 36-year-old male presenting with a small brown papule on the upper thigh. This lesion was confined to the superficial dermis, with no appreciable mitotic activity or necrosis. The immunoprofile was the same as the first case. There was no reported recurrence or evidence for systemic disease. Indeterminate cell histiocytosis has been described in both pediatric and adult populations, and can occur as a solitary lesion or as multifocal disseminated lesions. Extracutaneous manifestations have also been reported. While the clinical

course is usually benign, it is important to follow these patients more closely as indeterminate cell histiocytosis may have systemic involvement and has been associated with other hematopoietic neoplasms including acute leukemias.

Poster 537

RESIDENT

Cutaneous spindle cell angiosarcoma following irradiation mimicking Kaposi sarcoma

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A 70 year-old, immunocompetent woman presented with a violaceous, 2 cm skin nodule with surrounding ecchymoses in the left thigh. Imaging showed subtle, diffuse skin thickening with infiltration of the subcutis. Ten years prior to present admission the patient was treated for a pleomorphic sarcoma of the left thigh with wide local excision, radiation and chemotherapy. A biopsy of the nodule was interpreted at an outside institution as Kaposi sarcoma. Upon our review of the case, the histologic features were remarkably similar to Kaposi sarcoma with a dermal-based proliferation of uniform spindle cells arranged in slit-like vascular channels. Anastomosing vascular channels or marked nuclear atypia were not seen. However, given the history of irradiation, we considered the possibility of a spindle cell angiosarcoma. The tumor cells expressed CD31 but were negative for Human Herpesvirus 8 (HHV-8) by immunohistochemistry confirming the diagnosis of angiosarcoma. The tumor was re-excised with negative margins, and the patient is alive with no evidence of disease at 20 months after diagnosis. Angiosarcoma almost always displays marked nuclear atypia and anastomosing vascular channels. However, it may rarely be composed predominantly of spindle cells and mimic other spindle cell neoplasms. The distinction between Kaposi sarcoma and angiosarcoma is critical in regard to prognosis and therapy. This case illustrates an important potential pitfall in the diagnosis of spindle cell malignant vascular tumors and emphasizes the necessity of careful review of the clinical history and appropriate utilization of ancillary tests in arriving at the correct diagnosis.

Poster 538

RESIDENT

Syringocystadenoma papilliferum mimicking an anal condyloma

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Syringocystadenoma papilliferum is a benign sweat gland neoplasm that typically presents as a hairless nodule on the head and neck. Approximately a third of cases arise in association with nevus sebaceus. Aging sporadic lesions not associated with nevus sebaceus may exhibit verrucous

changes in the overlying epidermis; however, there are few reports of syringocystadenoma papilliferum arising in association with condyloma acuminatum. Furthermore, the perianal region is an exceedingly unusual location for this rare tumor. We report the case of a 46 year old man with a history of type-2 diabetes who presented with a pedunculated, keratotic perianal lesion measuring 1.0 cm. Microscopic examination revealed classic features of syringocystadenoma papilliferum with verrucous transformation of the overlying epidermis. Superficial keratinocytes exhibited cytoplasmic vacuolization and nuclear hyperchromasia reminiscent of koilocytes. In situ hybridization assays for HPV 6/11 and wide-spectrum HPV were negative. Ki-67 immunostain highlighted only basilar keratinocytes and a p16 immunostain was negative. These results argued against an HPV related lesion. Our case presentation illustrates a unique combination of an unusual site with a potential diagnostic pitfall in distinguishing verrucous features of syringocystadenoma papilliferum from syringocystadenoma papilliferum arising in association with condyloma acuminatum. We show how key histological findings and supporting studies may aid in the appropriate diagnosis.

Poster 539

RESIDENT

Expression of c-Met and Her2/neu in Merkel cell carcinoma: potential markers for targeted therapy

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Targeted therapy plays an increasingly important role in the era of personalized medicine. Aberrant hepatocyte growth factor/mesenchymal-epithelial transition factor (HGF/c-Met) receptor tyrosine kinase pathway has been implicated in melanoma, breast, colon, and renal carcinoma. Human epidermal growth factor receptor 2 (Her2/neu) amplification has been well established in breast carcinoma and also shown in gastroenteropancreatic neuroendocrine tumors. Both c-Met and Her2/neu represent candidates for targeted therapy, and neither has been studied in Merkel cell carcinoma. We evaluated the role of these two signaling pathways in the Merkel cell tumorigenesis by characterizing the expression profiles of three immunohistochemical markers (c-Met, phospho-Met, and Her2/neu). Fifteen cases of Merkel cell carcinoma were stained with Her2/neu, c-Met, and p-Met (phospho-met, activated form of c-Met). In 8 cases, Her2/neu stains were performed on all tumor blocks to exclude tumor heterogeneity. The results for c-Met and p-Met were recorded in a semiquantitative fashion (0: negative, 1+: 1-25% of positive tumor cells, 2+: 26-50%, 3+: 51-100% and staining intensity as 1-3+). Her2/neu scoring was assessed per American Society of Clinical Oncology guidelines. None of the cases (0/15) showed Her2/neu amplification. The vast majority of cases showed expression with

c-Met (13/15, 87%), and 10 cases showed moderate to strong staining intensity in more than 25% of the lesional cells. Weak staining for p-Met was observed in 3/15 (20%) cases. Her2/neu pathway does not appear to be involved in Merkel cell tumorigenesis. In contrast, c-Met was moderately to strongly expressed in 87% of cases, suggesting involvement of the HGF/c-Met pathway.

Poster 540

Benign clear cell nodular hidradenoma involving the lymphatic system: do “benign” metastases exist?

Catherine Stefanato, MD FRCPath

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Background - The term ‘benign metastasis’ has been sometimes applied to distant locations of neoplasms with clear-cut histopathologic features of benignity and with a completely favorable clinical outcome. **Materials and methods** - We report eight cases of morphologically benign clear-cell nodular hidroadenoma (CCNH) involving the lymphatic system. **Results** - In five cases the cutaneous tumor showed small foci of intravascular deposits of neoplastic cells; in two cases the cutaneous tumor presented with a synchronous tumor in a regional node; in one case a nodal location CCNH was found with no evidence of any primary in the skin. All the cases were treated with conservative surgery and none of them showed disease progression during a follow-up period ranging from 2 to 11 years. **Conclusions** - As an alternative to the hypothesis of a low-grade malignancy, the theory of ‘benign metastasis’ of CCNH can be considered when lymphatic spread is found in the absence of other histopathologic features of malignancy. Since lymphatic spread does not automatically imply malignancy, a provisional diagnosis of ‘atypical’ CCNH can be proposed for such cases.

Poster 541

Benign epithelioid peripheral nerve sheath tumor of the skin

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Benign epithelioid peripheral nerve sheath tumor (BEPNST) is a term proposed to describe those neoplasms that lack otherwise usual conventional histological and immunohistochemical features of either a benign neurofibroma or a schwannoma. They are rare in the skin and can be diagnostically challenging. Malignant variants

of these tumors have been more commonly reported than their benign counterpart. We report a case of BEPNST in a 78 year old male who presented with a six to eight month history of an asymptomatic, slow growing, 2.2-cm lesion on his left neck. The entire lesion was excised. Hematoxylin and Eosin stained sections showed a well circumscribed tumor in the dermis. The tumor was composed of trabeculae, loosely arranged nests and tight, cohesive nests of epithelioid tumor cells in a myxohyaline stroma. The epithelioid tumor cells comprised 75% of the lesion, demonstrated oval to round nuclei, conspicuous nucleoli and moderate to abundant pink cytoplasm with a distinct cytoplasmic borders. Occasional enlarged, hyperchromatic nuclei, bizarre giant cells and rare mitotic figures were noted. The spindle cell component comprised about 25% of the tumor and is composed of bland, fine, spindle cells arranged in short, interlacing fascicles. They had elongated nuclei and moderate pink cytoplasm. The tumor cells were uniformly and strongly positive for S100 protein alone and were negative for most of the epithelial and other neural markers. The tumor did not demonstrate histological features specific for either a neurofibroma or a schwannoma and a diagnosis of BEPNST was made.

Poster 542

RESIDENT

Myxoinflammatory fibroblastic sarcoma: a case report

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A 51-year old man presented with a solitary firm nodule on the proximal lateral side of his penile shaft. The lesion had been slowly growing for the past 6 months. It became painful over a period of 3 weeks. Complete excision of the mass revealed a 1.7x 1.2x 1 cm firm, pink-white nodule. Histologic examination showed a mixture of myxoid, hyaline, and inflammatory cells. The inflammatory area consisted of lymphocytes, histiocytes, scattered neutrophils and eosinophils. Within the hyalinized and myxoid zones, there were aggregates of bizarre-appearing tumor cells, often with giant macronucleoli, reminiscent of cytomegalovirus-infected cells or Reed-Sternberg cells. Rare pseudolipoblast-like fibroblastic cells were present within the myxoid areas. Sparse mitotic figures were present. No foci of necrosis or hemorrhage were seen. The neoplastic cells expressed vimentin strongly and were negative for CD45, EMA and CD30. The morphology and staining pattern in this case were consistent with myxoinflammatory fibroblastic sarcoma. Myxoinflammatory fibroblastic sarcoma (MIFS) is a rare, soft tissue of low-grade malignancy that most often occurs in acral location, although rare cases have been reported in more proximal locations, including the trunk. MIFS has significant potential for aggressive local recurrence, and limited potential for lymph node and distant metastases. MIFS require wide excision, with consideration of adjuvant radiotherapy in selected cases. To our knowledge, there is no previous report of this lesion on penile shaft. Familiarity

with this feature is essential to accurate diagnosis and effective patient management.

Poster 543

RESIDENT

Primary cutaneous mucinous adenocarcinoma

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Primary cutaneous mucinous adenocarcinoma is a rare malignant adnexal neoplasm (approximately 200 cases have been reported) with evidence favoring an origin in the deep coil of the eccrine sweat duct. Patients tend to be men, older than 60, and present with a firm skin-colored papule or nodule predominantly on the face or scalp, particularly in the peri-orbital region. Histologic features include clusters of relatively bland epithelial cells floating in pools of mucin separated by fibrous bands of stroma, involving the dermis and occasionally the subcutis. The lesions have a penchant to recur with positive and even narrow margins; however, only a small proportion locally metastasize. We describe the case of a 57 year old man complaining of a 3-4 month history of a 5 mm peri-orbital firm nodule. Histologic examination showed the characteristic findings diagnostic of primary cutaneous mucinous adenocarcinoma. We also discuss the importance of clinico-pathologic correlation in distinguishing between primary and metastatic mucinous carcinoma, which, on histologic and immunohistochemical grounds, is often not a trivial task.

Poster 544

Clear cell acanthosis associated with dermal duct tumor

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Clear cell acanthosis was first described by Ackerman et al.(1985). This histologic pattern is characterized by the focal distribution of pale cells in an acanthotic epidermis. It may occur as the main histologic alteration not only in clear cell acanthoma(CCA), but also in some other underlying conditions, such as seborrheic keratosis, melanocytic nevus or vulgaris verruca. Only a few cases were searched. We report a very rare case of dermal duct tumor with overlying clear cell acanthosis in a 18-year-old Korean girl. The patient presented a pink ovoid papule on her right chin that was unresponsive for intermittent topical steroids for several years. Histopathologic examination showed psoriasiform epidermal hyperplasia and multiple, focal irregularly distributed nests of keratinocytes with pale-staining cytoplasm. There were some exocytosis of neutrophils in the epidermis, and the dermis showed papillary dermal edema, enlarged vessels and mixed inflammatory cell infiltration that are resembling

with CCA. However, unlike CCA, the lateral margins of the pale cells were poorly demarcated and alternation of pale cells and normal keratinocytes were seen in other rete ridges. In addition, several nests of small uniform cuboidal cells showing maturation towards ductal lamina were present at the level of eccrine gland. These epidermal changes were clear cell acanthosis associated with dermal duct tumor.

Poster 545

A case of cellular angiofibroma (angiomyofibroblastoma-like tumor) in the male inguinal lesion

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Cellular angiofibroma (CA) is a benign mesenchymal tumor, occurring mainly in the superficial soft tissue of the genital region. It is uncommon tumor with approximately 20 reported cases in Japan. Bland spindle cell component and abundant small- and medium-sized vessels with mural hyalinization are characteristic histopathological findings. We describe a cellular angiofibroma in a 61 year-old Japanese male. He noticed a subcutaneous nodule on the suprapubic lesion in October 2010. The nodule gradually enlarged and he visited our hospital on August, 2011. Magnetic resonance imaging and ultrasonography depicted a 50mm well-circumscribed solid subcutaneous tumor. Microscopically, the lesion was well-circumscribed with fibrous capsule. It was composed of small spindle cells arranged in short fascicles and mixed up with numerous small- and medium-sized hyalinized vessels. There were alternating areas of compact short spindle cells and less cellular, loosely textured areas. The cells had monomorphic bland nuclei and scant mitotic figures. Immunohistochemically, these spindle cells were strongly positive for CD34 and negative for S-100 protein, desmin, and alpha-smooth muscle actin. The lesion was completely excised and no recurrence was observed 8 months after the excision. We considered this is a relatively typical case of a cellular angiofibroma, although the diagnosis of solitary fibrous tumor and cellular spindle cell lipoma are also considered. Cellular angiofibroma is uncommon but important mesenchymal neoplasm for dermatopathologist which is recognized as a distinct benign neoplasm with morphological and immunohistochemical overlap with angiomyofibroblastoma, cellular spindle cell lipoma and mammary-type myofibroblastoma.

Poster 546

Linear eruptive collagenomas on chest and neck

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Eruptive collagenomas are rare type of acquired connective tissue nevi. Multiple asymptomatic, firm, flesh-colored or pink nodules or plaques are clinically presented. Usually, symmetrical distribution is observed over the trunk and upper extremities. The histologic nature of the lesions has not been fully delineated. However, by definition, collagen fibers are predominantly increased in dermis. Herein we report a case of linear eruptive collagenomas, a rare clinical finding for connective tissue nevi. Less than ten cases of eruptive collagenoma showing linear or zosteriform distribution are found through literature searching. A sixteen-year-old male patient presented multiple papules and nodules on the chest and neck for several years. They were asymptomatic and unnoticeable at usual posture but revealed multiple erythematous papules and nodules after full extension. Interestingly, their distribution was linear from central portion of ant. chest to left side of post. neck. Five months ago, these lesions were treated at other hospital under the diagnosis of herpes zoster without improvement. His family history of similar cutaneous lesions and past history were unremarkable. His lab tests to find associated systemic diseases were all negative or within normal limits. Histological findings of biopsy specimen from a nodule on his chest revealed thickened collagen fibers in the dermis. Masson-Trichrome staining confirmed proliferation of collagen fibers. Elastic staining showed relatively decreased elastic fibers in the dermis. Alcian blue staining was positive in entire dermis.

Poster 547

RESIDENT

Decreased expression of desmoglein-3, E-cadherin and syndecan-1 in cutaneous acantholytic compared with invasive well-differentiated SCC

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Background: Acantholytic squamous cell carcinoma (ASCC) is an uncommon subtype of cutaneous squamous cell carcinoma. Evidence of reduced E-cadherin and syndecan-1 expression in ASCC suggests that abnormal expression of cell-cell adhesion molecules may play a role in the acantholytic morphology. Expression of desmoglein-3 (DSG-3) has not been characterized in ASCC. Objective: By immunohistochemistry, we sought to study expression of DSG-3, E-cadherin and syndecan-1 in ASCC compared with invasive, well-differentiated SCC (SCC). Methods: Twenty-two cases each of ASCC and SCC were matched for patient age within one decade, gender, and tumor site. Immunohistochemical staining patterns

of antibodies to DSG-3, E-cadherin and syndecan-1 were graded on a semi-quantitative scale for staining intensity (SI) and the degree of circumferential staining (CS) about the cell membrane. Three fields (high-, average-, and low-staining) were evaluated on each specimen. Results were assessed by conditional logistic regression and χ^2 analysis. Results: For ASCC compared to SCC, DSG-3 SI was reduced for average- (P=.0004) and low- (P=.003) fields; CS was reduced in average- (P=.0002) and low- (P=.003) fields. E-cadherin SI in ASCC was reduced for high- (P<.0001), average- (P=.02) and low- (P=.003) fields; CS was reduced in high- (P=.0009), average- (P=.03) and low- (P=.0007) fields. Syndecan-1 SI was reduced in high- (P=.004) and low- (P=.01) fields with reduction in CS only in low- (P=.02) fields. Conclusion: We found that expression of DSG-3, E-cadherin and syndecan-1 is reduced in most specimens of ASCC compared with SCC. These findings may explain the differences in biologic behavior between ASCC and SCC.

Poster 548

RESIDENT

Verruciform xanthoma on the forearm in the absence of chronic skin disease or syndrome

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An 82-year-old man presented with a 8mm pink, crusted, crateriform plaque with a yellowish center, and a hyperpigmented rim, located on the left forearm. It had grown over the past 6 months. Clinically, it resembled a keratoacanthoma and was excised. Microscopic analysis revealed hyperkeratosis, parakeratosis, papillomatosis and moderate acanthosis with no atypia of keratinocyte nuclei. Discrete collections of foamy histiocytes were present throughout the dermis with slight fibroplasia. This was diagnosed as verruciform xanthoma (VX), a rare benign lesion classically presenting on the oral mucosa or on anogenital skin. The etiology of VX is not yet completely understood, and is currently a topic of debate (see J Cutan Pathol 39:391 as well as Arch Dermatol. 148:260 and Arch Dermatol. 148:262). Only 40 reports of extra-oral and extra-genital VX exist, with the vast majority of these occurring on the background of a chronic skin disease or syndrome, including: a) conditions of chronic inflammation or trauma, such as lichen sclerosis, recessive dystrophic epidermolysis bullosa, and pemphigus vulgaris, b) chronic lymphedema, c) chronic graft versus host disease, or d) congenital epidermal nevi, especially those associated with the Congenital Hemidysplasia with Ichthyosiform Nevus and Limb Defects (CHILD) syndrome. Our patient had no history of chronic dystrophic skin disease or syndrome. This case highlights an extremely rare, if not unique, presentation of a solitary VX on the upper-extremity of an otherwise healthy individual.

Poster 549

RESIDENT

Rare presentation of oral verrucous carcinoma on the lip

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had perioral lesions of 10 year duration. The patient had been previously diagnosed with herpes simplex and treated with acyclovir. His past medical history was significant for AIDS, (CD4 count of 5); he was not on HAART secondary to non-compliance. The patient reported slight difficulty eating and speaking, but denied lip pain. Physical examination revealed small 3-6mm exophytic papules coalescing into a verrucous plaque that obliterated the entire mucosal and cutaneous lip, extending past the vermilion border. A wedge biopsy demonstrated compact parakeratotic cornified material, with papillomatosis and downward proliferation of keratinocytes showing dyskeratosis and glassy atypia. The tumor showed lobular architecture with a bulldozing pattern of invasion. Based on the clinical presentation and pathological findings the patient was diagnosed with oral verrucous carcinoma. Verrucous carcinoma is a rare variant of squamous cell carcinoma (SCC) that is usually found on the lower lip of elderly patients. While the most prevalent tumor of the oral cavity is invasive SCC, verrucous carcinoma on the lip represents only 2.8% of all oral neoplasms. Verrucous carcinoma has a significantly better prognosis than conventional SCC of mouth with >95% survival at 5 years. Dermatologists frequently associate SCC on the lower lip with Caucasian males and significant sun exposure. We present a rare case of verrucous carcinoma of the lower lip in an African American individual with acquired immunodeficiency syndrome.

Poster 550

RESIDENT

Primary cutaneous adenoid cystic carcinoma of the scalp with p16 expression

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Adenoid Cystic Carcinoma (ACC) is a rare carcinoma that typically arises in salivary glands but can also occur in other sites including skin. Primary salivary ACC is a locally aggressive tumor with late recurrence and late metastasis. Primary cutaneous ACC (PCACC) are found predominately on the scalp. PCACC are more indolent than salivary ACC, and, despite a high incidence of local recurrence, metastases are exceedingly rare. Fewer than sixty cases of PCACC have been reported in the English literature. A 62-year-old Caucasian male presented with a 6 mm mobile, blue-tinted nodule on the left mid-scalp unchanged for several years. Excisional biopsy showed a nodular, epithelial neoplasm within the dermis

and superficial subcutis. The central portion showed a solid pattern containing ductules and canalicular spaces. Peripherally, the tumor exhibited a cribriform morphology with well-circumscribed islands of basaloid epithelium exhibiting sharply-demarcated luminal spaces. Immunohistochemistry of the lesion demonstrated focal positivity for p16. A subsequent wide local excision showed no residual carcinoma. The patient remains disease free four months after diagnosis. Focal p16 expression has been reported in salivary gland ACC but has only been reported rarely in PCACC. The majority of reported salivary ACC with p16 protein expression were not positive for high-risk human papillomavirus by in-situ hybridization. PCACC is a rare malignancy that should be considered in the differential diagnosis of adnexal neoplasms, and, when occurring on the head and neck, must be distinguished from cutaneous involvement by salivary ACC.

Poster 551

Interdigitating dendritic cell sarcoma: a rare cutaneous case

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Interdigitating dendritic cell sarcoma (IDCS) is an exceedingly rare sarcoma with the largest series to date consisting of 4 cases. Many lesions display an aggressive clinical course. IDCS arises from S100 positive non-lymphoid accessory cells which are primarily located in the T-cell areas of lymphoid tissues. As such, lymph node involvement is the most common clinical presentation. Rare cases of both primary and metastatic skin lesions have been reported. We present a case of cutaneous IDCS arising in the right thigh of a 51 year-old male. The tumor was composed of a proliferation of atypical epithelioid cells with large convoluted nuclei, vesicular chromatin, and abundant amphophilic cytoplasm. On immunohistochemistry, the cells were positive for S100, fascin, and EMA. The cells were negative for a variety of markers including: pan cytokeratins, Mart-1, SOX-10, CD1a, CD 68, and CD 30. Given the S100 positivity, the differential diagnosis includes melanoma as well as cutaneous histiocytoses, such as Langerhans cell histiocytosis. Although rare, given the aggressive nature of this lesion, and possible associations with underlying disease, IDCS should enter the differential diagnoses of S100 positive cutaneous neoplasms.

Poster 552

RESIDENT

Sarcomatoid basal cell carcinoma: histologic, immunohistochemical, and molecular analyses of 2 cases

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Sarcomatoid carcinoma or carcinosarcomas of the skin are rare. Basal cell carcinoma (BCC) with osteosarcomatous differentiation is the second most common cutaneous sarcomatoid carcinoma, following squamous cell carcinoma with heterologous mesenchymal differentiation. There are only eleven cases of BCC with an osteosarcomatous component reported in the literature, with limited documented molecular analyses. We report the clinical, histologic, and molecular features of two additional cases. The cases occurred in elderly men, who are free of recurrent disease at 14 and 6 months, respectively, following excision. Both cases had a neoplastic basaloid epithelial component which stained positively for BerEP4 and high molecular weight cytokeratin (HMW cytokeratin), consistent with BCC. Admixed with the BCC was an osteosarcomatous component, comprised of spindle cells with osteoclast-like giant cells in the first case, and spindle cells with bone and cartilage in the second case. The sarcomatous component stained negative for both BerEP4 and HMW cytokeratin. Molecular analyses for common mutations in 17 cancer genes by a SNaPshot genotyping assay revealed only the presence of a mutation in TP53 at position 818G>C (R273P) in one of our cases; this case also had p53 overexpression in the epithelial and sarcomatous components by immunohistochemistry. Inactivating mutations in TP53, a tumor suppressor gene, and the corresponding p53 overexpression are found in 40 to 90% of BCC and are reported to be associated with epithelial to mesenchymal transition (EMT). Our findings suggest that factors other than TP53 may also be involved with EMT in BCC, especially in those lacking TP53 mutations.

Poster 553

Presentation of extra-mammary Paget's disease on the Posterior neck with immunohistochemical evaluation

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Extra-mammary Paget's disease outside of the anogenital region or axillae is rare. We present an unusual case of extra-mammary Paget's disease arising on the left Posterior neck of a 63 year old Caucasian male which was clinically thought to represent basal cell carcinoma or Bowen's disease. Shave biopsy and subsequent re-

excision showed an intra-epidermal Pagetoid carcinoma without evidence of underlying adenocarcinoma. Immunohistochemical studies including CK 7, Cam 5.2, CK 5/6, CK 19, CK 20, S-100, Mart-1, synaptophysin, and CEA were performed in the evaluation of the lesion. The results confirmed the diagnosis of extra-mammary Paget's disease and helped exclude Pagetoid squamous cell carcinoma in situ, malignant melanoma in situ, and the remote possibility of in situ Merkel cell carcinoma. The utility of the individual antibodies employed is discussed with respect to histologic differential diagnosis and results compared to those reported in the literature. A particularly interesting finding was the strong expression by the tumor cells for CK 19, a reported marker for follicular stem cells. However, the more specific marker CK 15 was negative, suggesting a more differentiated adnexal cell of origin. The current case appears unique for its presentation in sun-damaged skin outside of mammary, anogenital, or axillary regions and re-emphasizes that extra-mammary Paget's disease must be entertained for any Pagetoid neoplasm of the skin.

Poster 554

RESIDENT

Sinonasal non-keratinizing squamous cell carcinoma with nasal skin extension as the initial presentation

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Sinonasal non-keratinizing squamous cell carcinoma, previously designated as transitional cell carcinoma or cylindrical cell carcinoma, is an uncommon malignant neoplasm with distinct histopathological features, considered to be a low-grade malignancy that usually occurs in elderly patients. Extensive local invasion is uncommon. Here we report a case of 90 year old woman whose original presentation was as erythematous nasal skin nodules, biopsy of which showed a dermal tumor with features of sinonasal non-keratinizing squamous cell carcinoma. No epidermal dysplasia was present. A subsequent CT scan confirmed the presence of an endophytic tumor on the nasal sidewall. The initial presentation of sinonasal non-keratinizing squamous cell carcinoma as a skin lesion is previously unreported to our knowledge. Correct diagnosis requires accurate evaluation of the histopathology, as well as a comprehensive knowledge of pathology specific to this anatomic location.

Poster 555

RESIDENT

Trichoblastic carcinoma on the scalp of a 60 year-old African American female

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Trichoblastic carcinoma is a rare malignant adnexal neoplasm which normally arises from the scalp on fair-skinned individuals due to UV-B exposure. This tumor is capable of developing dermal fat invasion and malignant behavior. This case report examines a 60 year-old African American female with a slow-growing pigmented lesion on the scalp. A biopsy taken at initial presentation showed a transected pigmented nodular basal cell carcinoma. Due to its large size and atypical presentation, the lesion was widely excised. The resection specimen showed a neoplasm composed of large nodules of basaloid cells filling the dermis, extending to all margins and into the subcutaneous tissue. Some portions of the tumor show peripheral palisading and tumor-stroma clefting, characteristic of basal cell carcinoma and other portions show nodules resembling follicular germ cells with close association with minimal amounts of mesenchyma, resembling trichoblastoma. However, other portions of the tumor consist of large basaloid cells with large nuclei, open chromatin, nuclear crowding, nuclear pleomorphism and mitoses. These nodules had central, comedo-type necrosis and lack tumor-stroma clefting. Many of the nodules are also heavily pigmented. Although the lesion had features of a pigmented trichoblastoma, and focally of basal cell carcinoma, due to the size, malignant cytology and mesenchymal association with the large nests, we present this case as trichoblastic carcinoma, adding to the literature another case of this rare tumor.

Poster 556

RESIDENT

Rapidly evolving Kaposi-form hemangioendothelioma complicated by Kasabach-Merritt syndrome, with dramatic response to sirolimus therapy

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Kaposiform hemangioendothelioma (KHE) is a very rare and aggressive tumor of infancy that is oftentimes associated with a consumptive phenomenon called Kasabach-Merritt Syndrome. We present the case of a 4-week-old child who developed the sudden appearance of bruising on her back. Serum studies revealed a progressive thrombocytopenia with subsequent consumptive coagulopathy. Within hours, the purpuric lesion demonstrated rapid growth with progressive infiltration of the underlying tissues. A deep biopsy was performed, and histologic evaluation showed a deep dermal and subcutaneous vascular tumor

composed of numerous spindle-shaped endothelial cells lining small slit-like or crescentic blood vessels. A CD31 immunohistochemical stain was strongly and diffusely positive in the tumor, confirming its vascular nature. Staining for HHV-8 was negative, excluding Kaposi sarcoma, which shares similar histologic features with KHE. Given the depth of the tumor, its large size, and sudden appearance in infancy, these findings were most consistent with KHE complicated by Kasabach-Merritt syndrome. The patient was initially treated with propranolol and systemic corticosteroids followed by vincristine, but experienced little improvement in platelet counts. A recently published report of success with sirolimus in a small cohort of patients prompted trial of this medication. Within weeks, she had dramatic shrinkage of the tumor, as well as normalization of platelet counts, and she continues to improve on sirolimus; therefore, further excision of this tumor was not pursued.

Poster 557

RESIDENT

Basal cell carcinoma on genital skin

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Basal cell carcinoma (BCC) is the most common skin cancer overall but is extremely uncommon in sun-protected areas such as genital skin, occurring in 0.05% of cases. When occurring on genital skin, it is associated with a more aggressive course with higher recurrence rates believed to be due from neglect, patient hesitation, or unawareness on the part of both the patients and physicians. BCC is mainly caused by long-term UV radiation that causes DNA damage in various genes. PTCH and p53 gene mutations have been found in both sporadic and BCC associated with Gorlin's Syndrome. The etiology of cancer in sun-protected areas has not been determined. We examined a total of 10 cases of BCC diagnosed over 10 years, including clinical impression, anatomical site, patient age, and BCC subtype and immunohistochemical staining with p53 to further elucidate etiology of this neoplasm. Approximately 80% were from the vulva of female patients average aged 70.4 years. As with traditional BCC, the nodular variant was most common (40%). A case of fibroepithelioma of Pinkus, which develops more frequently in the genital and groin was observed as well.

Poster 558

RESIDENT

Type II cutaneous meningioma: a potential diagnostic pitfall in p63 positive cutaneous neoplasms

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Background: Cutaneous meningiomas are divided into three types. Type I lesions present at birth, predominantly in the subcutaneous tissue of the scalp, forehead and vertebrae and are derived from ectopic arachnoid cells that develop during embryogenesis. Type II lesions are found in adults around sensory organs of the head, cranial and spinal nerves, and are derived from arachnoid cells found surrounding nerves after they penetrate the dura. Type III lesions are due to direct extension or metastasis from dural based neoplasms. Dural meningiomas (especially grades II and III) have been shown to express p63. The aim of our study is to examine the expression of p63 in type II cutaneous meningiomas. Methods: Biopsies taken from two patients were evaluated for histopathologic features and expression of p63, EMA, cytokeratin, S100, CD31. Results: Case 1 (female, 74, forehead, lesion), case 2 (female, 89, scalp, lipoma vs cyst). In both cases the lesional cells demonstrated spindled to epithelioid cells with elongated cytoplasmic processes arranged in a whorling pattern. Pseudovascular spaces were noted in case 1. There was no significant cytologic atypia or necrosis, but case 2 had up to 5 MFs/10 HPFs. Immunohistochemical staining showed diffuse expression of EMA and p63. The tumors were negative for cytokeratin, S100, and CD31. Conclusions: 1. Type II cutaneous meningiomas express p63. 2. p63 is not helpful in the differentiation of type II and type III cutaneous meningiomas. 3. Type II cutaneous meningiomas should be considered in the differential diagnosis of cutaneous neoplasms which express p63.

Poster 559

RESIDENT

An unusual locally invasive combined squamous and melanocytic neoplasm: a case report with complex differential diagnosis

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Dermal based combined squamous and melanocytic neoplasms are emerging clinicopathologic entities that appear on sun-exposed areas of elderly patients. The histopathologic differential includes two provisional diagnostic entities: (i) dermal squamomelanocytic tumor

and (ii) melanocytic matricoma, and (iii) markedly rare histologic variant of pilomatrical carcinoma, so called pilomatrical carcinoma with intralesional melanocytes. Due to their rarity, the biological behavior of these rare cutaneous neoplasms remains uncertain. Herein, we present a novel case of dermal based squamous and melanocytic neoplasm. A 72-year-old man presented with a pigmented papule on nasal ala clinically concerning for basal cell carcinoma. Histopathologic evaluation demonstrated atypical melanocytic cells architecturally intimately intermixed with single units and clusters of atypical squamous cells. Two notable features of this case are focal matrical differentiation, characterized by shadow cells with accompanying granulomatous reaction, and locally invasive growth, characterized by multifocal perineural invasion. We hypothesize that since both dermal squamomelanocytic tumors and melanocytic matricomas may be locally aggressive, the two putative diagnostic entities may in fact be better classified as incipient pilomatrical carcinomas.

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RESIDENT

Proximal-type epithelioid sarcoma in a 54-year-old woman

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Epithelioid sarcoma is a rare tumor that characteristically presents as a subcutaneous or deep dermal mass on the distal extremities of young adults. A more aggressive "proximal" or large-cell type has been described and occurs in middle-aged and older adults most commonly on proximal body sites. This variant is reported to recur more frequently and metastasize earlier than the classic type of epithelioid sarcoma. We present the case of a 54-year-old woman with an asymptomatic lesion on the right pubic region. Her dermatologist excised the lesion with a clinical diagnosis of lipoma. On pathologic examination there is a poorly circumscribed, 1.8 cm nodular neoplasm in the subcutis composed of confluent sheets of atypical epithelioid cells with enlarged hyperchromatic vesicular nuclei, coarse chromatin, and rhabdoid cytologic features. The neoplasm is embedded in a fibrous and focally myxoid stroma. Immunohistochemical staining is positive for broad-spectrum cytokeratin (AE1/AE3), vimentin, low molecular weight keratin (CAM 5.2), and epithelial membrane antigen. Focal loss of INI-1 expression in the large epithelioid cells was observed. The neoplasm was negative for S-100, carcinoembryonic antigen, estrogen receptor, CD34 and TTF-1. The patient has no evidence of a prior malignancy. A diagnosis of proximal-type epithelioid sarcoma was made.

Poster 561

RESIDENT

Integrated analysis of homozygous deletions in merkel cell carcinomas

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We have performed a quantitative research data analysis of copy number deletions and transcriptome in merkel cell carcinomas. We found that the average number of all major copy number deletions to be 18 per tumor. We have integrated these data with previous gene expression analysis of the Reference Sequence genes and have identified genes commonly affected by copy number deletions. Pathways represented for genetic inactivation included those involved in intracellular signaling and DNA topological change. Three hundred and twenty three genes were identified in commonly deleted genomic loci that also demonstrated reduced expression in primary merkel cell carcinomas. Our analysis provides an integrated revisit of publically available archived research data that may prove useful for future cancer therapy.

Poster 562

A case of mantle adenoma

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Mantle adenoma is a rare, benign skin tumor with a predilection for the face which Ackerman AB described in the book. It is characterized histopathologically as cords and columns of interconnecting epithelium that emanate from an infundibulum. Case report: A 79-year-old man had a 30 mm tumor on the left occipital region of his head which had grown progressively larger for 7 years. The histopathological findings showed cords or columns of undifferentiated epithelial cells with sebaceous differentiation interconnected in a reticulate manner with lumen formation. There was deposition of mucin and an increase of collagen fiber in the stroma. Immunohistochemical study revealed that the tumor cells were partially positive for epithelial membrane antigen and weakly positive for GCDFP-15 (Gross cystic disease fluid protein-15) and negative for AE1/AE3, CEA, CAM5.2, BerEP4, alpha-smooth-muscle actin and adipophilin. We diagnosed this tumor as mantle adenoma. Although our patient showed negative immunohistochemical staining for adipophilin, we diagnosed this tumor as mantle adenoma based on the histopathological findings of HE stain.

Poster 563

RESIDENT

Rare case of retroperitoneal sarcoma associated with Muir-Torre syndrome

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Muir-Torre syndrome (MTS) is defined by the combination of at least one cutaneous sebaceous neoplasm and a primary synchronous or metachronous internal malignancy (typically gastrointestinal). Now known to be a subtype of hereditary nonpolyposis cancer syndrome (HNPCC), MTS is a genetically characterized by autosomal-dominance due to germline inactivating mutations in genes encoding mismatch repair proteins; MSH2, or less commonly MLH1, representing the typical underlying molecular mechanism. We report the very rare and unusual case of a 58 year-old man diagnosed with MTS established from a previous cutaneous sebaceous neoplasm, who subsequently presented with a large 14.3 cm left adrenal mass. Sections revealed a malignant neoplasm composed of spindled cells with high mitotic activity (>5 per high power field). Immunohistochemical studies revealed that the tumor was weakly reactive to SMA while being negative for desmin, CD117, CD31, CD34, S100 protein, and pan-cytokeratin. Further immunohistochemical analyses revealed intact nuclear expression of MLH1, but complete loss of nuclear MSH2 expression in neoplastic cells, establishing a clear relationship to his MTS. We document that on very rare occasion, sarcomas can be part of the MTS internal tumor spectrum.

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Reassigned to Poster Session 1

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RESIDENT

Cystic panfolliculoma: a rare follicular neoplasm

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Panfolliculoma is a rare, follicular neoplasm which differentiates towards both the upper and lower portions of the hair follicle. Fewer than 20 panfolliculomas have been described. We present the second case in the literature of panfolliculoma with cystic change, to our knowledge. A 23-year-old female presented with a 1 cm nodule on her left Posterior thigh. The clinical differential diagnosis included an epidermal inclusion cyst and nevus lipomatosus superficialis. Histopathologically, the neoplasm consists of a well demarcated, symmetrical follicular tumor present within a fibrotic stroma. The fibrosis is concentric around the hair follicles. The follicles

are dilated with cystic change. The hair bulbs in the lesion are wide and present in the superficial dermis. There are prominent trichohyaline granules. Differentiation towards the isthmus and infundibulum are also present. There is a surrounding suppurative and granulomatous infiltrate. A Ber-EP4 immunohistochemical stain is weakly positive in the germinative cells, but not the follicular papillae. The entire epithelium, including the follicular epithelium, stains diffusely and strongly with CK903. CK5/6 highlights the outer root sheath strongly in the suprabulbar zone, isthmus and inferior portion of the infundibulum. Within the superficial portion of the infundibulum, there is localization of CK5/6 to the outer and basal layers of the outer root sheath. This CK5/6 staining pattern is different from that of the other reported case of cystic panfolliculoma. In summary, this cystic panfolliculoma is another interesting example of a rare entity with a slightly different immunoprofile than the other previously described case.

Poster 566

Basal cell carcinoma with pilomatricoma-like changes

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Basal cell carcinoma (BCC) may exhibit variable morphology and types of differentiation. A rare variant features pilomatricoma-like changes and has only been described in nine previous instances, usually occurring as ulcerative papules or nodules on the face or upper extremities. It must be distinguished from benign and malignant pilar tumors. We report an 82 year old man with history of multiple non-melanoma skin cancers who presented with a papule on his right helix of uncertain duration. Shave biopsy of this lesion demonstrated irregular aggregations of atypical basaloid cells emanating from the undersurface of the epidermis. The atypical basaloid aggregates demonstrated peripheral palisading of nuclei, nuclear hyperchromasia, and focal individual cell necrosis. At the advancing edge of the neoplasm, there were smaller angulated nests that infiltrate the dermis. The neoplastic aggregates exhibited stromal clefting. Some of the dermal basaloid aggregates demonstrated features reminiscent of hair matrix, and cornified cells with karyolytic nuclei (“shadow cells”) were also seen. These findings were compatible with BCC with pilomatricoma-like changes. It is important to recognize this rare type of BCC, which may potentially be mistaken for pilomatricoma or pilomatrical carcinoma. Treatment with excision has been described without recurrence or metastasis.

Poster 567

RESIDENT

A case of primary cutaneous mucinous adenocarcinoma of the eyelid

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We report a case of an 82 year-old woman who presented with a left lower eyelid lesion for 6 months. Skin excisional biopsy showed a well-demarcated dermal tumor composed of cohesive cells in solid and cribriform growth patterns set in a lake of mucin. Focal duct formation was noted. The tumor cells had abundant eosinophilic cytoplasm and mildly pleomorphic nuclei. Frequent mitoses were present. By immunohistochemistry, the tumor cells were positive for estrogen receptor, progesterone receptor, patchy positive for mammoglobin, and rare cells were positive for p63. Myoepithelial cells at the periphery of tumor islands were highlighted by focal positivity for myosin and type IV collagen. The tumor cells were negative for PAX-5, D2-40, CEA and CD117. Whole-body computed tomography scan showed the absence of primary mucinous adenocarcinoma elsewhere which excluded the possibility of metastatic origin. Follow-up at 3 months after excision showed no recurrence or metastasis of the tumor. Combined clinical, morphological and immunohistochemical profiles supported the diagnosis of primary cutaneous mucinous adenocarcinoma involving the eyelid. Primary mucinous adenocarcinoma of the skin is rare and the data in the published literature are limited to case reports and small case series. These tumors are presumed to be a subtype of eccrine sweat gland carcinoma. The tumors involving the eyelid may extend into the orbit and metastasize to regional lymph nodes, if incompletely excised. Metastasis from primary mucinous adenocarcinoma of the breast and gastrointestinal tract, though a rare possibility, needs to be excluded with careful full clinical investigation.

Poster 568

RESIDENT

A case of congenital metastatic rhabdoid tumor presenting as an exophytic periorbital tumor and multiple subcutaneous nodules

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Rhabdoid tumor is a rare, aggressive malignancy first reported in 1978. Extrarenal rhabdoid tumors have been described in the central nervous system, skin, soft tissues, liver and gastrointestinal tract amongst others. We report a case of a newborn boy born with a large exophytic periorbital tumor as well as several subcutaneous nodules scattered on the head, neck, trunk and extremities.

Radiologic studies revealed metastases in the cerebellum and liver. An excisional biopsy revealed solid sheets of epithelioid and focally spindle-shaped neoplastic cells with a moderate amount of clear to eosinophilic cytoplasm, and enlarged, irregular, vesicular nuclei with conspicuous nucleoli. Vimentin was diffusely positive and INI-1 was not expressed in tumor cells. Interestingly, a small portion of tumor cells expressed myogenin which is not commonly expressed by malignant rhabdoid tumors. This case reviews the presentation of the rare and highly aggressive rhabdoid tumor and demonstrates that rhabdoid tumors many express myogenin in a small percentage of tumor cells. The significance of myogenin positivity in rhabdoid tumor remains to be determined.

Poster 569

RESIDENT

Carcinoma cuniculatum: a potential diagnostic pitfall on punch biopsy of the penis

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An 84 year old man presented with a plaque on the glans penis for eight months. Examination revealed a scaly, indurated plaque, measuring 1.2 x 0.5 cm, without inguinal lymphadenopathy. A superficial punch biopsy was performed, which showed an atypical squamous proliferation with mild cytologic atypia but a focal infiltrative pattern. Due to concern for an underlying malignant lesion, a repeat biopsy was performed, which revealed multiple anastomosing keratinaceous cysts. A diagnosis of squamous cell carcinoma, polycystic type (carcinoma cuniculatum) was rendered and the patient underwent a partial penectomy. Macroscopic examination showed multiple interconnecting cysts with keratinaceous contents, involving 80% of the penile circumference. Microscopic examination revealed a well-differentiated SCC with a branching and burrowing pattern, diagnostic of carcinoma cuniculatum. Carcinoma cuniculatum type of penile SCC has been described recently as a well-differentiated tumor characterized by a labyrinthine network of sinuses resembling rabbit burrows, from which the term cuniculatum is derived. It predominantly affects older males, and has a good prognosis. The diagnostic challenges in our case were the punch biopsy specimens, which showed mild epidermal cytologic atypia as well as keratin filled cystic structures without significant atypia. A diagnosis of the carcinoma cuniculatum variant of SCC versus an epidermal inclusion cyst, in a similar scenario could be quite challenging. We present this case for its rarity and to highlight the diagnostic difficulty that a dermatopathologist could face due to a punch biopsy of this lesion.

Poster 570

Squamous cell carcinoma arising within an inverted follicular keratosis

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Inverted follicular keratoses (IFK) are benign epithelial neoplasms, typically firm nodules, developing from the follicular infundibulum. An IFK may be mistaken histologically for a squamous cell carcinoma. Malignant degeneration of an IFK into a true squamous cell carcinoma is rare. We describe a 65-year-old woman with a pedunculated papule of the left upper eyelid. Histologic examination revealed hyperkeratosis overlying an acanthotic proliferation of benign appearing keratinocytes and a dilated infundibulum and squamous eddies. Deep to this was a proliferation of atypical, somewhat spindle, keratinocytes demonstrating pleomorphism and nuclear atypia, indicative of a squamous cell carcinoma arising in association with an inverted follicular keratosis.

Poster 571

RESIDENT

Cutaneous leiomyosarcoma and proliferating trichilemmal tumor: two cases illustrating the importance of clinico-pathologic correlation

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Clinico-pathologic correlation is of the utmost importance in the evaluation of inflammatory cutaneous diseases and its importance in the diagnosis of cutaneous neoplasms should not be overlooked. We present two such cases: the first, a 67 year old man with a subcutaneous nodule overlying the proximal interphalangeal joint initially diagnosed as squamous cell carcinoma, despite the absence of epidermal aberration on clinical exam. Consideration for amputation and the poor clinico-pathologic correlation prompted a second review of the histology revealing a well-circumscribed neoplasm with rapid keratinization towards the center as well as a lack of both cytologic atypia and mitotic activity, consistent with a proliferating trichilemmal tumor. The second case is a 59 year old man with a skin-colored pedunculated papule with the clinical appearance of an acrochordon; however, on account of its recent history of rapid growth, a biopsy was performed. Subsequent histologic examination revealed a superficial spindle cell neoplasm with nuclear pleomorphism, mitotic activity, and an infiltrative border. Immunohistochemical staining showed diffuse smooth muscle actin reactivity, consistent with a superficial cutaneous leiomyosarcoma. These cases further strengthen the importance of communication between the dermatologist and pathologist and subsequent clinico-pathologic correlation in reaching the correct diagnosis.

Poster 572

RESIDENT

Basal cell carcinoma arising in a connective tissue hamartoma

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Basal cell carcinoma is known to occur in epidermal nevi. The development of a basal cell carcinoma in a dermal connective tissue hamartoma has not been reported; however, nonmalignant basaloid epidermal hyperplasia has been seen in this setting. We describe 62-year-old man with a gluteal cleft grape-like fleshy mass, which, at the time of excision, had developed serous drainage. It had developed rapid growth over the span of several months and had been considered a condyloma clinically. Histopathology revealed multifocal papillary dermal nodules of basaloid cells with epidermal connection and surrounding stromal retraction. An excess of irregularly arranged fibroblasts and disorganized smooth muscle was seen amid tumor nodules. We considered the aforementioned basaloid epidermal hyperplasia as well as nonmalignant basaloid follicular hamartoma; however, in light of its rapid growth clinically and histologic pattern, we felt a true basal cell carcinoma had developed. A subsequent excision revealed no carcinoma but an abundance of connective tissue elements indicative of connective tissue hamartoma, confirming the impression on the initial sections that a basal cell carcinoma had occurred in the setting of a connective tissue nevus in a non-sun exposed site.

Poster 573

RESIDENT

Skin hyperpigmentation and keratoses at sites of friction in a patient with promyelocytic leukemia

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We report a case of 34-year-old man who presented with pruritic hyperpigmented and keratotic papules and plaques on his neck, groin and palms. Patient had a history of relapsed promyelocytic leukemia treated with arsenic trioxide. Physical examination revealed hyperkeratotic flat-topped grey papules and plaques within the field of hyperpigmentation over his neck and lower abdomen as well as skin-colored keratotic punctate papules on his palms. A shave biopsy from the abdomen demonstrated acanthotic epidermis, mild hypergranulosis and vacuolated keratinocytes in the epidermis. In the dermis, there was a sparse superficial perivascular lymphocytic infiltrate associated with melanophages. Solar elastosis was not present. These histologic findings were consistent with clinical impression of arsenical keratosis. Exposure to arsenic may occur as a result of occupational exposure, ingestion of contaminated water, or from medicinal use. Skin is a common manifestation of arsenic toxicity. The

earliest findings with arsenic toxicity are pigmentary changes, more commonly described at sites of friction and referred to as "raindrops on a dusty road." More specifically, the raindrops consist of less pigmented lesions on a background of diffuse hyperpigmentation. The high affinity of arsenic for sulfhydryl groups makes keratinocytes a sensitive target for arsenic-induced toxicity and explains development of arsenical keratoses, Bowen's disease, SCC and BCC. Arsenical keratoses are pre-cancerous lesions with predilection for sites of friction. They tend to occur after 2 years of arsenic exposure, and after a latency of 10 years can transform into SCC. Careful attention to distribution, patient history, and histopathology enables accurate diagnosis.

Poster 574

RESIDENT

Pigmented matrical carcinoma: a case report with comparison with pilomatricoma

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Pigmented matrical Carcinoma (PMC), an extremely rare malignant follicular neoplasm, which is characterized by tumor recognized as matrical carcinoma (MC) with intralesional melanocytosis. We report a case of 5.2x3.5x3.0 cm, tan-red, ulcerated polypoid dermal nodule occurring in the shoulder of an 86-year-old male. Histopathologically, the tumor demonstrated multilobular proliferation of pleomorphic matrical cells arranged in solid sheets and unusual pattern not described previously- thin inter-anastomosing cords resembling cribriform pattern - with multiple small globules of corneocytes reminiscent of 'shadow cells'. There were numerous atypical mitotic figures, 8-12 per high-power-field, and foci of tumor necrosis. Also noted was a focal area of pigmented dendritic melanocytes admixed with tumor cells. Tumor cells showed strong nuclear immunohistochemical (IHC) reactivity with β -catenin, p63 and MIB-1. Aggregates and scattered dendritic cells within the tumor were Melan-A and S-100 positive. Trabeculae-cribriform growth areas demonstrated CD56 membranous reactivity. These histological and IHC findings confirm PMC. Four cases of pilomatricoma (PMX) were selected for the presence of pigments and melanocytic participation using Fontana Masson stain, S-100 and Melan-A. These PMX's demonstrated β -catenin nuclear reactivity, membranous E-cadherin with scattered S-100 and Melan-A reactive dendritic cells. Another case of melanocytic matricoma (MM), revealed closely packed melanocytes at the periphery of aggregated matrical cells with matrix cells demonstrating β -catenin nuclear reactivity. The presence of melanocytes in association with matrical cells suggests neoplastic proliferation of cells possibly with biphasic differentiation. Lastly, the expression of β -catenin by PMC, MM and PMX confirms the role of Wnt/ β -catenin mutation pathway in their tumorigenesis.

Poster 575

RESIDENT

Carcinoma erysipeloides: an uncommon manifestation of metastatic mucoepidermoid carcinoma

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Carcinoma erysipeloides (CE) is an uncommon but distinct form of cutaneous metastasis characterized by lymphangitic spread of tumor resulting in lymphatic obstruction and erythema of the involved skin clinically resembling erysipelas. It is most commonly seen with breast cancer but has been reported with other primary tumors. Only a few cases of cutaneous metastases from mucoepidermoid carcinoma of salivary glands have been published and to our knowledge CE pattern has not been previously reported. We present a rare case of cutaneous metastasis from mucoepidermoid carcinoma presenting as CE. The patient is a 64-year-old woman with history of mucoepidermoid carcinoma of the right hard palate and right cervical lymph node metastasis who underwent radical neck dissection and radiation therapy. Two years after the neck dissection, she presented with right neck erythema and tenderness which spanned an area of 25 x 10 cm and extended from the sternal notch to the base of the skull, surrounding the well healed surgical incision scar. She denied fevers, pain or new masses and did not display signs of cellulitis or infection. Skin biopsies from the erythematous region showed dermal lymphatic permeation by clusters of carcinoma cells consistent with mucoepidermoid carcinoma. CE typically mimics conditions such as erysipelas, cellulitis and radiation dermatitis. CE should be kept in the differential diagnosis of erythematous skin lesions even in the context of tumors where cutaneous metastases are uncommon such as mucoepidermoid carcinoma. Skin biopsy should be performed for the histopathologic confirmation of the diagnosis in these unusual clinical presentations.

Poster 576

RESIDENT

A case of high grade sarcoma in a patient with a history of recurrent meningioma and stereotactic radiosurgery

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We present a 69 year-old male who underwent biopsy of a right frontal parasagittal meningioma in April 1991 after new onset seizures. He declined surgery until November 1997. Recurrence of the meningioma was treated with stereotactic radiosurgery in 2005. He recently presented with falls, mental status changes, and scalp ulcers. An MRI of the brain showed a mass measuring up to 5.1 cm, involving the bilateral frontal lobes subjacent to the prior

craniotomy site. Surgery revealed scalp tissue which was fibrous and adherent to the skull. Tumor invaded the superior sagittal sinus and left frontal lobe, and was completely resected. Microscopic examination of the intradural, extraaxial component of the tumor revealed a meningioma. There was neoplasm located in the scalp, calvarial bone and intradural extra-axial compartment with features that differed from the meningioma and consisted of a hypercellular proliferation with high mitotic activity, bizarre pleomorphic nuclei, and areas of necrosis in a desmoplastic background, consistent with high grade sarcoma. In light of the location and the patient's history of radiation; the findings are suggestive of a post-radiation sarcoma. Immunohistochemistry of the meningioma showed was positive for EMA, and negative for PR. Ki-67 for the meningioma was 7%, and for the sarcomatous component it was 35%. Immunohistochemistry of the high grade sarcoma was positive for Vimentin, and CD68, and negative for smooth muscle actin (SMA). The unique histologic findings along patient's history of radiotherapy present as an exceedingly rare combination of concurrent meningioma and malignant sarcoma.

Poster 578

Nestin expression in a case of carcinosarcoma with basal cell carcinoma and osteosarcoma elements

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Carcinosarcoma is a rare malignant neoplasm with epithelial and mesenchymal components, with the most common being squamous or basal cell carcinoma (BCC) with osteosarcomatous differentiation. BCC carcinosarcomas appear primarily on sun-exposed sites, typically as nodules. Although most cases have not recurred, there has been a case of metastasis, suggesting greater malignant potential in BCC carcinosarcoma compared to BCC. CASE REPORTA 69-year-old asymptomatic man presented with a small stable mass on the medial aspect of his right calf for several years. He was treated with Keflex for presumed granuloma, but it continued to grow, necessitating an excisional biopsy. Biopsy demonstrated a basaloid epithelial tumor with peripheral palisading and rare cleft retraction consistent with BCC, which segued into a malignant mesenchymal component forming osteoid. Mitotic figures and cytologic atypia were conspicuous in the mesenchymal portion. Immunohistochemistry demonstrated loss of cytokeratin and p63 staining within the sarcomatous component. Nestin was expressed in sarcomatous areas, but not in the carcinomatous component. MRI showed isolated involvement of the skin. There was no evidence of residual tumor on wide re-excision, and clinical follow-up for ten months showed no evidence of recurrence. CONCLUSIONBCC carcinosarcoma is a rare cutaneous neoplasm with poorly understood malignant potential. We report for the first time the expression of the

sarcoma marker nestin in the sarcomatous component of carcinosarcoma. Carcinosarcoma should be considered in any cutaneous neoplasm showing a malignant mesenchymal component. Further study is needed to better define the molecular and clinical features of these tumors.

Poster 579

RESIDENT

Histological tracking of vismodegib treatment for basal cell carcinoma: pitfalls and pearls

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Introduction: Vismodegib, a targeted inhibitor of Smoothed, the key component of the hedgehog signaling pathway (HhSP), is the first FDA-approved HhSP targeting agent for treatment of advanced BCC. Histological assessment of treatment response to vismodegib has not been precisely defined. Case: A 61-year-old man was referred for a large, ulcerated tumor of the chest. The mass measured 25 x 24 x 4 cm (fig 1). Histology revealed nodular and focally keratotic basal cell carcinoma (fig 2). Given the inoperable status of the tumor, the patient was started on vismodegib therapy. After one month of treatment, dramatic tumor shrinkage was achieved resulting in 50% reduction in clinical dimensions (fig 3). However, biopsy of residual tumor done at 4 weeks showed poorly differentiated squamoid features not evident on multiple samples of the initial tumor (fig 4). Continued treatment, guided by clinical benefit, resulted in marked further tumor regression. Biopsy sampling at 12 weeks revealed complete histological clearance of BCC and showed only granulomatous inflammation. Comment: The histological appearance of BCC from patients undergoing vismodegib therapy has not been clearly defined. Treatment resistance has been shown in some patients on vismodegib during investigative trials. However, it is unclear whether the microscopic appearance of vismodegib-resistant BCC differs from that of the original tumor. This case illustrates that squamoid differentiation may occur in BCC during treatment with vismodegib. This finding does not seem to indicate tumor resistance to vismodegib by selection of more aggressive tumor elements, since the BCC continued to rapidly shrink with ongoing treatment.

Poster 580

Primary cutaneous ganglioneuroma: a report of a case

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Ganglioneuroma is a rare benign skin tumor arising from the neural crest. There are two classifications

of ganglioneuroma: cutaneous ganglioneuroma and ganglion cell choristoma. Few cases of primary cutaneous ganglioneuroma have been reported. We report a case in a 61 year old male presenting with a soft nodule in the middle of the back. Careful questioning yielded no other relevant medical history and no history of tumors as a child. Histologically, there were two populations in the dermis, a spindle cell proliferation with wavy nuclei, and a second population of large polygonal cells with vesicular nuclei. The first population was S100 positive and the second was S100 negative and focally positive for NSE, synaptophysin, and PGP 9.5 diffusely positive. The implication is that this is a Schwann cell and ganglion cell proliferation representing a cutaneous ganglioneuroma or ganglion cell choristoma. Although, this is likely an isolated lesion, exclusion of a prior history of a neuroblastoma or other neural tumors is essential.

Poster 581

RESIDENT

Extramammary Paget's disease associated with a superficial basal cell carcinoma, Pinkus type

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We highlight the case of a 77-year-old woman who presented with a vulvar lesion that was clinically suspicious for extramammary Paget's disease. A punch biopsy revealed elongated, interconnected slender nests of basaloid cells, with a surrounding fibrovascular matrix, in patterns of so-called fibroepithelioma of Pinkus, a recognized variant of superficial basal cell carcinoma. At higher magnification, however, numerous individual atypical cells with abundant intracellular mucin were distributed throughout the lesion in pagetoid patterns. These cells were found to be positive for cytokeratin 20 and negative for cytokeratin 7. The histologic diagnosis of "superficial basal cell carcinoma with concomitant extramammary Paget's disease" was rendered. An association between Paget's disease and epidermal hyperplasia has been described, although documented cases exhibiting the striking degree of fibroepitheliomatous hyperplasia demonstrated by this case are limited and this phenomenon is unrecognized by many pathologists. It has been proposed that the fibroepithelioma of Pinkus-like changes might merely be the result of an inductive process initiated by malignant pagetoid cells. It is essential to rule out internal malignancy, especially since cytokeratin 20 staining in the genital area has been associated with synchronous internal malignancy, especially colorectal carcinoma. Additionally, the observation of fibroepitheliomatous hyperplasia in the genital region should trigger a search for Paget's cells, which can easily be missed when they are few in number. It is important for pathologists to be aware of this under-recognized combination of pathologic processes.

Poster 582

RESIDENT

Ocular oncocyoma: report of two cases

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Oncocytomas are benign neoplasms, which can occur at a variety of sites but only rarely in the eye. Ocular adnexal oncocyomas usually involve the caruncle. They occur most commonly in elderly women. We report two cases of ocular oncocyoma. First case: a 63 year-old woman with a one-year history of a slow growing red small mass located in the caruncle of the lower eyelid that caused occasional pruritis. Second case: an 84 year-old man with a five-year history of a bright red 5-6 mm papule on the lower-mid eyelid. Histologically, both lesions were well circumscribed and comprised of polyhedral cells exhibiting abundant, finely granular acidophilic cytoplasm and round to oval paracentral nuclei usually containing a single prominent nucleolus. Cells were arranged in confluent sheets and forming glandular spaces with secretory material. The second case showed both tubular and papillary configurations. Sparse intermingled goblet cells were also apparent. Mitotic activity was absent in both cases. Nuclear atypia and infiltrative growth were absent. Electron microscopy revealed cytoplasm packed with mitochondria of varying sizes and shapes. Ocular adnexal oncocyomas most often involve the caruncle and are most commonly seen in elderly women. However, rarely it arises from the eyelid. Oncocyoma in men is rarely reported. Surgical excision usually is curative however very rare malignant transformation has been reported. Although oncocyoma of the lacrimal gland have been rarely reported in the dermatopathology literature, it is important to be recognized by the dermatopathologists because of its ability for malignant transformation.

Poster 583

RESIDENT

A case of primary cutaneous leiomyosarcoma of the scrotum

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Primary cutaneous leiomyosarcomas are uncommon dermal or subcutis tumors. The most common sites include the extremities, scalp, and trunk. However, this tumor may infrequently arise in the genitalia, nipple, and upper lip. This lesion most often affects males in the sixth decade and may present as a painful/tender mass. Dermal-based cutaneous leiomyosarcomas arise from the arrector pilorum, or in cases of scrotal lesions, dartos muscle. Here we present a 58-year-old male whom presented to the clinic with a left scrotal mass which was subsequently excised. Histology of the lesion showed a well circumscribed, malignant neoplasm with markedly

pleomorphic nuclei, hyperchromasia, and numerous mitoses. Atypical mitoses were also observed. Positive staining for SMA, MSA, and Desmin aided in classifying this neoplasm as an interesting case of primary cutaneous leiomyosarcoma of the scrotum. Histopathology of leiomyosarcomas shows interlacing fascicles of spindled cells with cigar-shaped nuclei. Nuclear pleomorphism can be variable. Mitotic activity is at least one per 10 high-power fields in cellular areas. Due to the resemblance of this tumor to other desmoplastic tumors of the skin, immunohistochemistry is necessary for making the diagnosis. The presence of vimentin, smooth muscle actin, and h-caldesmon can be demonstrated. Desmin is likely to be present in most lesions. In a study by Kaddu, et al., the two most commonly encountered growth patterns were nodular and diffuse. The nodular tumors are usually very cellular, have abundant mitoses, and atypical nuclei. On the other hand, diffuse tumors have a lower cellularity, well differentiated cells, and subtle mitoses.

Poster 584

RESIDENT

Giant cell angiofibroma: tumor or localized periorbital lymphedema?

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A 56 year old female presented with a longstanding, slowly enlarging mass of the lower left eyelid. Excision demonstrated skin and soft tissue expanded by edema, spindled fibroblasts, and multinucleated giant cells. Lesions with apparently identical clinical and histopathologic features have been classified as neoplasms designated 'Giant cell angiofibroma' (GCA). Since similar histopathologic features may be encountered in localized lymphedema around the orbit and in many other anatomic sites, we suspect that at least some GCAs represent an unusual presentation of localized lymphedema, not neoplasms.

Poster 585

RESIDENT

Cutaneous metastases from Signet-ring cell carcinoma of the esophagus

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A 63-year-old male with a history of signet-ring cell carcinoma (SRCC) of the esophagus presented with a 2 month history of painful growths on his trunk. The esophageal cancer had been treated with neoadjuvant chemotherapy (cisplatin and 5-fluorouracil), radiation, and esophagectomy 10 months prior to presentation. Physical examination demonstrated an erythematous firm nodule on the left Posterior shoulder and within a surgical scar on the right trunk. Biopsies from both lesions confirmed a diagnosis of metastatic esophageal SRCC. Within the dermis, there were numerous interstitial

glandular structures with focal signet ring differentiation coursing amongst the collagen bundles. Many atypical cells and mitotic figures were also noted. Esophageal SRCC is a rare histological variant of esophageal adenocarcinoma. The reported incidence is 0.1-0.6%; it is highly aggressive and associated with a poor prognosis. Cutaneous metastasis of esophageal SRCC is very rare; most patients die within 3 months after being diagnosed with any cutaneous metastasis from the esophagus. Other tumors can demonstrate signet-ring cell differentiation including melanoma, primary cutaneous non melanoma skin cancers, lymphoma, and other adenocarcinomas. An immunohistochemical panel may aid in determining the etiology of SRCC. Cytokeratin expression of CK7+/CK20- is a reliable indicator of an esophageal origin. Chemotherapy, radiation, and surgery are frequently employed for treatment. However, an effective treatment regimen has yet to be determined due to the limited number of cases observed. Pre-operative chemoradiation followed by esophagectomy has been reported to improve survival compared to esophagectomy alone.

Poster 586

RESIDENT

Digital cutaneous lesions as initial presentation of visceral malignancy

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Select visceral malignancies rarely present as digital cutaneous metastases. We report two patients without a prior history of malignancy who initially present with rapidly progressive swelling of the finger tip. Skin biopsies subsequently led to the discovery of their underlying malignancies. The first patient is a 75 year old man who presented with a swollen, bleeding right fifth finger that got progressively worse over a course of three weeks. Two months prior to presentation he reported hurting himself on a lawn mower. Biopsy showed a CK20, CEA, and CDX-2 positive adenocarcinoma. Further workup demonstrated a 4 cm rectosigmoid adenocarcinoma. The patient was started on chemotherapy but has been lost to follow up. The second patient was a 64 year old man who presented with a three week history of pain and swelling of his left index finger which he believed to be related to a spider bite. Biopsy showed a metastatic hepatocellular carcinoma that stained positive for HepPar and polyCEA with a canalicular pattern. Imaging showed widely metastatic disease, and the patient deceased three months after the initial diagnosis. We describe the second case of an occult hepatocellular carcinoma and the third case of an occult colorectal carcinoma manifesting as digital cutaneous metastases as the initial symptom.

Poster 587

RESIDENT

A case of pseudovascular squamous cell carcinoma
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Pseudovascular or pseudoangiosarcomatous squamous cell carcinoma (SCC) is a rare, aggressive subtype of squamous cell carcinoma which may histologically mimic angiosarcoma in light of its growth pattern and significant acantholysis. We describe a 71-year-old man with a left upper eyelid exophytic papule which upon shave biopsy demonstrated focal proliferation of atypical, epithelioid keratinocytes along the dermoepidermal junction. There was also a diffuse and infiltrative dermal proliferation of atypical epithelioid and spindle cells exhibiting extensive acantholysis with pseudovascular channels and cystic spaces, some of which contained numerous erythrocytes. A panel of immunohistochemical stains was performed; both the intradermal and dermal neoplastic cells exhibited immunoreactivity against CK 5/6 and p63. The superficial, more well-differentiated portion of the neoplasm stained positively for pancytokeratin and pre-keratin while deeper, poorly-differentiated neoplastic cells had only focal immunoreactivity. The tumor cells failed to demonstrate immunoreactivity against CD31, CD34, S100, or Melan-A immunostains. The histologic immunohistochemical features of this specimen confirmed the presence of a pseudovascular SCC. The patient was subsequently treated with Mohs micrographic surgery.

Poster 588

RESIDENT

Aggressive digital papillary adenocarcinoma in an 11 year-old female

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Aggressive digital papillary adenocarcinoma (ADPA) is a rare malignant tumor of the cutaneous eccrine glands that typically occurs in elderly males and classically presents as a minimally painful mass on the volar surface of fingers and toes. Histologically, it is characterized as a solid/cystic lesion with tubuloglandular structures and luminal papillary projections that may exhibit nuclear pleomorphism, brisk mitotic rate, and necrosis. However, ADPA may also show benign features, including circumscription, well differentiated glandular structures, lack of significant cytologic atypia, and low mitotic index, but these features do not correspond to the lesion's biologic potential. It has been proposed that all ADPA have the potential for recurrence and metastasis and therefore should be treated with aggressive surgical therapy (amputation or radical excision). We present the case of an 11-year-old female with no significant past medical history who developed a 2 cm

soft tissue mass of the right middle finger adjacent to the proximal interphalangeal joint. Excisional biopsy showed a well-circumscribed eccrine tumor with papillary projections, minimal cytologic atypia, and scattered mitoses, consistent with aggressive digital papillary adenoma/adenocarcinoma. ADPA is a rare lesion and to our knowledge has not been previously reported in patients younger than 15 years. Awareness of this entity enables clinicians to correctly diagnose and aggressively treat this lesion that has high potential for recurrence and metastasis.

Poster 589

Sebaceous adenoma recurring as sebaceous carcinoma

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A 76 year old man presented with a keratotic papule on the right elbow. A shave biopsy demonstrated multifocal sebaceous lobules with several layers of undifferentiated basaloid cells, interpreted as sebaceous adenoma. The dermal margin was involved. Eight years later, the patient presented with an 8mm hard dermal nodule at the site of the previous biopsy. A deep shave biopsy showed a spherical dermal nodule of basaloid cells with sebaceous differentiation. The nuclear/cytoplasmic ratio was high, and numerous mitotic figures were easily identified, consistent with sebaceous carcinoma. Re-excision with clear margins was performed. Although lack of MLH-1 staining was seen in both specimens, the patient had no personal or family history of malignancy; therefore, further evaluation of Muir-Torre Syndrome was not pursued. Sebaceous neoplasms are uncommon tumors with differentiation towards the sebaceous lobule. In 1998, Nussen and Ackerman first introduced the idea that sebaceous "adenoma" is sebaceous carcinoma. Most textbooks still consider adenoma to be benign and clearly separate from carcinoma. To our knowledge, this is the first case of an adenoma progressing or recurring as carcinoma. Clear margins should be obtained, if possible, when treating so-called sebaceous "adenoma."

Poster 590

RESIDENT

Correlation study of the expression of enhancer of zeste homolog 2 and cyclin D1 in cutaneous squamous cell carcinoma

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Enhancer of Zeste Homolog 2 (EZH2) has been shown to be involved in the progression of multiple human cancers. Our previous study indicated that EZH2 is over-expressed in cutaneous squamous cell carcinoma (SCC). EZH2 is present downstream of the pRb-E2F pathway,

which is regulated by cyclin D1. A correlation between the expression of EZH2 and cyclin D1 was reported in the mucosal SCC of the head and neck region. To extend our previous study and explore if such a correlation also exists in cutaneous SCC, we performed immunohistochemistry by the anti- EZH2 and cyclin D1 antibodies using tissue microarrays (59 cases) plus 11 separate cases of cutaneous SCC. EZH2 is detected in 53 out of the 70 cases of cutaneous SCC. Cyclin D1 is detected in 24 out of the 70 cases. Among the 53 cases with positive EZH2 immunostaining, 23 cases are also cyclin D1 positive. Among the 17 cases with negative EZH2 immunostaining, 16 cases are cyclin D1 negative. Pearson correlation analysis shows that the expression of EZH2 is correlated with that of cyclin D1 (two-tailed $p < 0.05$). These results implicate that cyclin D1 and EZH2 may share the same tumorigenic pathway in a subset of cutaneous SCC.

Poster 591

RESIDENT

Coalescing multifocal (plaque-type) syringoma of the face mimicking a microcystic adnexal carcinoma

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A 65 year-old woman presented with a 0.5 cm ill-defined, firm plaque on the glabella. A 3 mm punch biopsy was performed and interpreted at an outside institution as a "basaloid neoplasm with sweat duct differentiation", favoring syringoma. Because of the unusual degree of dermal involvement, the pathologist raised the possibility of a microcystic adnexal carcinoma (MAC). A subsequent excisional biopsy demonstrated an infiltrating growth pattern of syringomatous elements which prominently involved the deep reticular dermis and subcutis. The case was submitted for expert consultation and a diagnosis of MAC was rendered. The patient was referred for Mohs micrographic surgery. During the operation, the surgeon had difficulty establishing negative margins, and after thirteen stages, decided to arrest the procedure and submit the specimens for pathologic review. The permanent sections showed similar histological findings to the prior biopsies, however, it became apparent that the lesion was discontinuous and multifocal. Moreover, the lesion was largely confined to the dermis and no perineural invasion was identified. Based on these findings, a diagnosis of coalescing multifocal (plaque-type) syringoma was favored, and confirmed by a second expert consultant. This case illustrates the importance of recognizing the unusual histologic features of this rare variant of syringoma. While the extent to dermal involvement and infiltrative appearance may raise the possibility of MAC, there is no perineural invasion and the lesion is multifocal. Paradoxically, this latter feature makes misdiagnosis as a malignancy particularly treacherous, because of the possibility for extensive and potentially disfiguring surgery on cosmetically sensitive regions.

Poster 592

RESIDENT

High grade pleomorphic sarcoma (malignant fibrous histiocytoma) with HMB-45 immunohistochemical expression

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We present an extremely unusual case of malignant fibrous histiocytoma (MFH) that demonstrated focal HMB-45 immunohistochemical positivity. An 83-year-old woman presented with a right knee mass that was increasing in size. An MRI showed a 7.6 x 5.2 x 6.4 cm septated and circumscribed mass in the medial soft tissues adjacent to the proximal tibia. The mass appeared to involve the subcutaneous fatty tissues, but was external to the muscular fascia with no involvement of bone. Microscopic examination of the excised mass revealed a heterogeneous lesion with high-grade areas composed of markedly pleomorphic to spindled cells arranged in a storiform pattern with hemorrhage and necrosis, intermixed with myxoid areas with lower grade nuclear features. Rare multinucleated tumor cells with vacuolated cytoplasm demonstrated HMB-45 positivity. However, both S-100 and Melan A were negative, making a diagnosis of melanoma unlikely. We previously reported an unusual case of atypical fibroxanthoma with similar tumor cells exhibiting focal HMB45 reactivity (J Cutan Pathol 2004; 31:284-286) – this patient is alive and well 8 years later. This pattern of tumor cell HMB45 immunopositivity is important to be aware of to avoid a misdiagnosis of melanoma.

Poster 593

RESIDENT

Detection of polyomavirus by PCR and IHC in Merkel cell carcinomas of unknown primary origin

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Merkel cell carcinoma (MCC) is one of the most aggressive primary cutaneous malignancies with a presumptive line of differentiation similar to the epidermal Merkel cells. Recently, a novel member of the polyoma virus family, named Merkel cell polyomavirus (MCPyV) was identified to be integrated in the MCC tumor cells genome. Most MCCs develop within the skin of the head and neck or on the extremities and commonly metastasize to regional lymph nodes. However, over the last 20 years, there have been reports of MCC involving lymph nodes without a primary skin lesion identified at the time of diagnosis. Whether nodal MCCs of unknown primary origin (MCCUP) occur as a result of a metastasis with regression of the primary lesion or represent bona-fide primary MCCs occurring in lymph nodes remains a controversial and unresolved issue.

In a retrospective search, we identified 3 cases of MCCUP involving inguinal, axillary, and neck lymph nodes. Our aim was to evaluate the status of MCPyV in our MCCUP cases by polymerase chain reaction (PCR) using 4 different primer pairs amplifying sequences within the T antigen and VP1 gene of MCV and by immunohistochemistry (IHC). We found that all our 3 MCCUP cases were positive by PCR and 2 of 3 cases (66%) were positive by IHC for MCPyV. Our results seem to indicate that MCCUP and cutaneous MCC cases have similar MCPyV detection rates.

Poster 594

RESIDENT

CD56 positive Merkel cell carcinoma with crush artifact

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The patient was an 85 year-old female who presented with a history of a rapidly growing lesion on her left cheek. Physical examination revealed a 0.9 cm pink asymptomatic nodule on the left cheek. A punch biopsy showed dermal aggregates of atypical round blue cells with lacey chromatin embedded in a dense diffuse lymphoid infiltrate with extensive crush artifact. Differential diagnosis included Merkel cell carcinoma and NK-cell lymphoma. Immunohistochemistry revealed that the lobular aggregates of tumor stained diffusely positive for CD56 and with CK20 in a paranuclear dot pattern. Chromogranin and TTF-1 staining were negative, while synaptophysin was positive. CD3 stained the dense surrounding T cell lymphoid infiltrate while CD20 stained few admixed B cells. All the findings supported a diagnosis of Merkel cell carcinoma over an NK-cell lymphoma. As previously reported in the literature, CD56 positivity in Merkel cell carcinoma with crush artifact may present a potential for diagnostic confusion with NK-cell lymphoma.

Poster 595

Hereditary leiomyomatosis and renal cell carcinoma (HLRCC) syndrome: a case report

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Hereditary leiomyomatosis and renal cell carcinoma (HLRCC) is an autosomal dominant familial syndrome characterized by the development of cutaneous and uterine leiomyomas as well as renal tumors. The mutation of this condition has been identified in the fumarate hydratase gene. The histology of the renal tumors has just been recently characterized. They tend to have a poor prognosis and demonstrate frequent spread to regional lymph nodes. A 37 year old male presented with abdominal pain and hematuria. CT scan demonstrated

a right renal mass. The patient underwent a right renal nephrectomy which demonstrated a high grade renal cell carcinoma with metastasis to inter-aortal caval lymph nodes. Morphologically, the tumor was composed of areas of collecting duct morphology and foci more typical of type 2 papillary renal cell carcinoma. The composite tumor cells demonstrated prominent eosinophilic cytoplasm and large nuclei with prominent nucleoli. Immunostains demonstrated that the tumor cells were negative for cytokeratin 7 and CD10 and positive for AMACR and pancytokeratin. Further investigation into the patient's history revealed that there was a prior diagnosis of multiple cutaneous leiomyomas and one leiomyosarcoma. Indeed, in one report the association of multiple leiomyomas and renal cell carcinoma (RCC) was strongly highlighted. The morphology and immunostaining pattern of the RCC together with the cutaneous leiomyomas/ leiomyosarcoma is compatible with HLRCC. The patient is currently enrolled in an NIH-sponsored HLRCC study along with family members. This case highlights the need for proper diagnosis of this syndrome by the pathologist facilitating early detection of these aggressive tumors.

Poster 596

RESIDENT

Workup of suspected shark bite: when to consult dermatology?

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A 54 year old man stumbled up to a lifeguard tower in Pacific Beach, San Diego, California, complaining of dizziness after swimming in the ocean. He was found by lifeguards to have a large wound on his left shoulder. Suspecting a shark attack, they rushed him to the Emergency Department, where he revealed that the lesion had been slowly growing for the past 25 years and expanding more rapidly in the past 6 months. He also reported weakness of the ipsilateral arm and general fatigue. Physical exam revealed an 18 x 19.5cm actively bleeding ulcerated plaque with an erythematous, rolled border on the left shoulder. Smaller satellite lesions were noted. Skin biopsy demonstrated nests of atypical basophilic cells infiltrating the dermis with retraction artifact, consistent with basal cell carcinoma. Positron emission tomography (PET) showed extensive uptake in the left shoulder with multiple hypermetabolic foci in bilateral lungs suspicious for metastatic disease. Fine needle aspirate of the lung revealed tight clusters of small cells with high nuclear-cytoplasmic ratio, hyperchromatic chromatin, irregular nuclear membranes, and metachromatic basement membrane material, against a background of pulmonary alveolar tissue, consistent with metastatic basal cell carcinoma. Although metastatic basal cell carcinoma (MBCC) is well documented in the literature, it is an exceedingly rare complication of a very common malignancy. Previously associated with high morbidity and mortality, this prognosis may be changing with the advent of hedgehog signaling pathway inhibitors. We will review

important diagnostic considerations and highlight new treatment modalities for MBCC.

Poster 597

RESIDENT

Chemotherapy-related leukemia cutis: a case report and review of hematopoietic malignancies related to breast cancer treatment

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A 36-year-old woman with a history of breast cancer, status post mastectomy, treated with doxorubicin and cyclophosphamide in 2011, currently on tamoxifen, presented with multiple oval, pink macules and flat papules on the trunk. There was minimal surface change besides faint scale; KOH preparation demonstrated rare evidence of hyphae. A skin biopsy revealed a diffuse infiltrate of atypical mononuclear cells with scattered mitoses intercalating between collagen bundles throughout the dermis. Immunohistochemistry confirmed a diagnosis of myelogenous leukemia cutis. Subsequent bone marrow biopsy revealed treatment-related acute myeloid leukemia with chromosome translocation 9:11 and MLL gene arrangement. Given the high prevalence of breast cancer in the general population and that chemotherapy is used to treat all stages of breast cancer, hematopoietic malignancies related to these treatments are increasingly relevant to the diagnosis of skin disease in breast cancer patients. We review hematopoietic malignancies associated with current breast cancer targeting therapy, highlighting their cutaneous manifestations.

Poster 598

RESIDENT

Pagetoid intraepidermal urothelial carcinoma of the glans penis: a case report with helpful immunohistochemical features

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Pagetoid intraepidermal carcinoma (Paget's disease) can present along nearly any skin surface and is associated with a variety of neighboring or distant malignancies. We present an 80 year old man referred to dermatology for evaluation and treatment of an asymptomatic, thin red plaque abutting the urethral meatus. His medical history is significant for distant prostatic adenocarcinoma treated with radical prostatectomy and distant ureteral-based urothelial carcinoma. A punch biopsy specimen revealed single cytologically pleomorphic, hyperchromatic, and atypical cells scattered throughout the epidermis. The atypical cells stained positively for CK7 and CK20 and negatively for S100, MART-1, PSA, and CEA, consistent

with urothelial carcinoma. Subsequent cystoscopic tissue biopsy demonstrated urothelial carcinoma in situ. The differential diagnosis of pagetoid intraepidermal carcinoma of the penile glans includes urothelial carcinoma, gastrointestinal carcinoma, malignant melanoma, renal carcinoma, and squamous cell carcinoma. Histopathologic evaluation with judicious immunohistochemical profiling is critical to focus the differential diagnosis and guide subsequent workup.

Poster 600

RESIDENT

Evaluation of mismatch repair protein MSH2 in cutaneous squamous cell carcinoma (SCC) and basal cell carcinoma (BCC)

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Defects in the DNA mismatch repair (MMR) machinery leads to microsatellite instability (MSI) and play a key role in the pathogenesis of Muir-Torre syndrome, an inherited cancer syndrome characterized by sebaceous neoplasms in association with visceral malignancy. Despite extensive studies on the loss of MMR proteins in sebaceous neoplasms, limited studies are available to address their roles in more common cutaneous epithelial tumors such as basal cell carcinoma (BCC) and squamous cell carcinoma (SCC). Our group previously showed that MSH2 is the mostly commonly lost MMR protein in sebaceous tumors. In this study, a total of 26 patients with BCC and/or SCC were separated into three cohorts: those with (1) synchronous/metasynchronous sebaceous adenomas that demonstrated loss of MSH2 protein (n=5), (2) sebaceous adenomas that demonstrated intact MSH2 expression (n=7), and (3) no synchronous/metasynchronous sebaceous tumors (n=14). Some patients harbored more than one BCCs or SCCs. A total of 20 BCC/SCC were identified in cohort 1, 16 in cohort 2, and 14 in cohort 3. The expression of MSH2 protein was evaluated in these lesions. We demonstrated that 10% of BCC/SCC (2/20) showed loss of MSH2 protein in cohort 1, 6% (n=1/16) in cohort 2, and 0% (n=0/14) in cohort 3. There is no significant increase in MSH2 loss in the cohort with synchronous/metasynchronous sebaceous adenoma that demonstrated MSH2 defects. In addition, only a minority of BCC and SCC demonstrated MMR protein abnormality, and MSI does not appear to play a role in the pathogenesis of these epithelial tumors.

Poster 601

Ductal adenocarcinoma of the breast metastatic to scalp mimicking a pilar cyst: a case report and a review of cutaneous scalp metastases

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Metastatic carcinomas to the scalp are classically described as painless rapidly growing nodules, patches, or plaques. In rare cases, however, they present as tender subcutaneous nodules. We present a case of ductal adenocarcinoma of the breast metastatic to the scalp mimicking a pilar cyst. A 49-year-old female presented with a 6cm by 4cm left breast ulcer with foul smelling yellow-green exudate and erythematous ulcerated nodules on the left axilla. Core biopsies of the breast and axillary nodules confirmed the diagnosis of ductal adenocarcinoma that is ER+PR-Her2-. Further examination of the patient revealed a 4cm by 3cm by 2cm subcutaneous tender non-mobile nodule on the right occipital scalp without epidermal change or alopecia. Computed tomography of the brain revealed a right parietal bone lytic lesion. Ultrasound-guided biopsy of the nodule revealed uniformly malignant ductal epithelial cells arranged as nests and cords, with pleomorphic nuclei, coarse chromatin, inconspicuous nucleoli, and pink cytoplasm. Immunohistochemical evaluation revealed concordance with breast primary. This case highlights the variety of clinical morphologies with which cutaneous metastases can present.

Poster 602

RESIDENT

Primary cutaneous adenoid cystic carcinoma: a case report

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A 65-year-old woman with no significant past medical history presented with a subcutaneous nodule on her left upper Posterior thigh. The lesion had been slowly growing for the past 5 years and it was getting tender over a period of 4 weeks. On exam, a 2 cm subcutaneous, firm nodule with reddish blue discoloration of overlying skin was seen. Complete excision was performed. Histologic sections revealed an intradermal neoplasm without an overlying epidermal connection. Tumor lobules were composed of basaloid cells with minimal cytoplasm, arranged in cords and strands in a multifocal growth pattern. There was mild nuclear pleomorphism. Many lobules demonstrated a cribriform pattern. The nests did not demonstrate peripheral palisading of tumor cells. Foci of perineural invasion were appreciated. The lesion showed infiltrative

border. The morphology in this case was consistent with primary cutaneous adenoid cystic carcinoma (PCACC). Wide local re-excision was performed. twenty-five months after the re-excision, the patient remains asymptomatic without any evidence of recurrence or metastasis. Adenoid cystic carcinoma (ACC) is a malignant neoplasm of the major salivary glands. ACC also rarely occurs in the minor salivary glands, esophageal gland, bronchial glands, mammary glands, and skin. Cutaneous ACC is very rare; about 60 cases have been published. It is believed to arise from eccrine and apocrine glands. The microscopic differential diagnosis of PCACC includes metastatic cutaneous adenoid cystic carcinoma, cylindroma, adenoid basal cell carcinoma, mucinous apocrine carcinoma, and apocrine mixed tumor of the skin. We discuss the histopathologic findings, clinical course and relevant differential diagnoses.

Poster 603

FELLOW

Primary carcinosarcoma of the skin: a series of 10 cases

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Carcinosarcomas, biphasic tumors composed of co-existing malignant epithelial and mesenchymal components, are extremely rare in the skin. Primary cutaneous carcinosarcomas (PCCs) often have a spindled morphology with a subtle epithelial component. However, identification of both the elements is critical since these tumors are frequently associated with a poor prognosis. To better define the histologic characteristics, we identified 10 cases of PCCs over a period of 17.5 years (1995 to 2012). The patients' age ranged from 57 to 97 years (mean 77.4). There was a slight male predominance (6:4) and the male patients were on an average 6 years younger than the women at presentation (81 vs. 75). Six of the 10 lesions were located on sun-exposed regions. Fifty percent of tumors were ulcerated and there was prominent inflammatory infiltrates in 2 cases. Squamous cell carcinoma (SCC) was the most common epithelial component (5 cases), all of which were moderately to poorly-differentiated with spindled areas or individually scattered cells. Basal cell carcinoma (BCC, 3 cases) was the next frequent pattern followed by a malignant adnexal neoplasm and a mixture of neuroendocrine carcinoma, SCC and BCC. Undifferentiated pleomorphic sarcomas made up the mesenchymal components in all tumors, 2 of which also contained multinucleated giant cells. The presence of epithelial and mesenchymal elements was confirmed by comprehensive immunophenotyping. In summary, we describe a series of 10 cases of primary cutaneous carcinosarcoma and discuss the clinical presentation, histologic features, immunophenotypical patterns and differential diagnosis of these rare tumors.

Poster 604

FELLOW

Follicular induction overlying a dermatofibrosarcoma protuberans

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The term "induction" has been used to describe epidermal changes above dermatofibroma. These changes include simple acanthosis, often with basaloid/follicular or sebaceous differentiation. Dermatofibrosarcoma protuberans (DFSP) is a fibrohistiocytic tumor of low malignant potential, while giant cell fibroblastoma (GCF) is considered as a juvenile variant of DFSP. DFSP is generally characterized by more uniform spindle cells, which are based in the dermis and subcutaneous fat and exhibit a more prominent storiform pattern than the one seen in dermatofibroma. In contrast to that characteristic hyperplastic overlying a dermatofibroma, the epidermis overlying DFSP usually attenuated. Superficial shave biopsies of DFSP may lack characteristic features of DFSP. We present a 14 year old female with a DFSP whose overlying epidermis exhibited follicular induction, a finding which has not been previously associated with DFSP. Her excisional biopsy revealed a DFSP with a characteristic storiform pattern of CD34(+) spindle cells infiltrating the dermis and subcutaneous fat. Focally noted were multinucleated giant cells, a feature which is more reminiscent of giant cell fibroblastoma. Overlying the tumor, multiple small epithelial islands of basaloid cells arising from the base of the epidermis showed palisading and associated spindled cells resembling hair bulbs. The COL1A1-PDGFB fusion was detected via FISH. The patient underwent three wide local excisions and 5 stages of Mohs surgery to clear the lesion. In summary, we report a case of DFSP, which showed a follicular induction. This epidermal change has not been reported in DFSP, and may present a diagnostic pitfall in superficially sampled lesions.

Poster 605

FELLOW

CD10, procollagen-1 and p53 unhelpful in differentiating cutaneous spindle cell neoplasms

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Cutaneous spindle cell neoplasms are a common and difficult diagnosis in the field of dermatopathology. Several immunohistochemical stains have been purported to be helpful in the distinction of these tumors. P53 is a marker that has not been well studied in deciphering spindle cell cutaneous neoplasms and CD10 and procollagen-1 (PC-1) staining patterns have not been well studied in leiomyomas and leiomyosarcomas (LMS). We immunohistochemically

stained 15 cases of leiomyoma, 14 cases of cutaneous LMS, 1 case of subcutaneous LMS and 15 cases of AFX with CD10, PC-1, and p53 and graded positivity on a scale of 1 to 3. Cases were considered positive for CD10 if they showed membranocyttoplasmic positivity, PC-1 if it showed cytoplasmic staining, and p53 if it showed nuclear positivity. Out of the leiomyoma cases only 1/15 cases showed weak positivity for CD10, no cases showed PC-1 positivity and only 2 cases showed weak to moderate p53 positivity. Twelve out of 14 cases of cutaneous LMS demonstrated weak to strong CD10 positivity, 10/14 cases showed weak to strong staining for PC-1 and 7/14 cases demonstrated weak to strong positivity for p53. All (15/15) cases of AFX demonstrated weak to strong positivity with both CD10 and PC-1 and 14/15 cases of AFX were weak to strongly positive for p53. We conclude that PC-1 and CD10 are sensitive but not specific for the diagnosis of AFX and do not appear to adequately differentiate from cutaneous LMS (p=0.2461 and 0.1067 respectively for pair-wise comparisons).

Poster 606 **FELLOW**

Squamous cell carcinoma (SCC) with pilomatrical differentiation

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While a single case of primary SCC of the lung with pilomatricoma-like features has been previously noted in the literature, to date, SCC with pilomatrical differentiation has not been reported in the skin. We present a case of 72-year-old male who had a 1.0 cm non-healing erythematous lesion on the right elbow for 6 months. Histopathologic examination of a biopsy revealed variably-sized dermal nodules, with mild to moderately atypical squamous epithelium, with keratin pearls and focal epidermal connection. Woven in, were islands with basophilic cells at periphery and central eosinophilic shadow cells. Taken together, these features were interpreted as being most consistent with SCC with pilomatrical differentiation. Given that pilomatricomas usually contain mutations of CTNNB1, a gene encoding the 92Kda protein β -catenin, immunostaining with β -catenin was performed. β -catenin gene mutations of hair matrix cells stabilize β -catenin proteins, which accumulate in the cytoplasm and translocate to the nucleus. These nuclear proteins then activate gene transcription resulting in abnormal matrical cell proliferation and pilomatricoma formation. As expected, in basophilic cells, nuclear staining was the predominant pattern while in non-basaloid squamous epithelial areas, membranous staining was evident. The patchy nuclear β -catenin staining in foci of atypical squamous epithelium within the epidermis favored the histopathologic diagnosis of SCC with pilomatrical differentiation, as opposed to a collision lesion of SCC and pilomatricoma. While scattered previous reports document the potential for misinterpretation as carcinoma of FNAs

from pilomatricoma, to the best of our knowledge, this is the first report of cutaneous SCC with pilomatrical differentiation.

Poster 607 **FELLOW**

Extraocular sebaceous carcinoma in-situ arising in association with squamous intraepidermal neoplasia: case report and review

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Extraocular sebaceous carcinoma is a rare neoplasm. The diagnosis can be difficult to make and requires a high degree of suspicion. Tumors do not necessarily arise in pre-existing sebaceous glands, are usually invasive at the time of diagnosis, and, like their ocular counterparts, can exhibit pagetoid intraepidermal spread. Cases of purely in-situ extraocular sebaceous carcinoma are extremely rare and somewhat controversial. Review of the literature reveals three cases, one each on the arm, the temple, and the neck. The extraocular sebaceous carcinoma in-situ on the neck arose in association with an actinic keratosis. We report two cases of extraocular sebaceous carcinoma in situ arising in association with squamous intraepidermal neoplasia, one in an actinic keratosis, and one in a squamous cell carcinoma in-situ. Both neoplasms exhibited keratinocytic atypia of the epidermis and a proliferation of well differentiated but atypical sebaceous epithelium. Whether or not these tumors have dual sebaceous and squamous differentiation, pure sebaceous differentiation, or alternatively metaplastic sebaceous differentiation, is unresolved.

Poster 608 **FELLOW**

Epidermal panfolliculoma: advanced follicular differentiation confined to the epidermis

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A 55 year old male patient presented with an asymptomatic skin lesion on his right leg of two months duration. The patient had a previous history of recurrent genital warts. Physical examination revealed a skin colored verrucous nodule 8X8mm in size on the anterior aspect of the right leg. Additionally, the patient had multiple genital condyloma which had been previously biopsied and the diagnoses confirmed. The clinical impression of the lesion was seborrheic keratosis and was shaved for biopsy. Histology demonstrated a slightly elevated plaque like lesion with basket weave stratum corneum, coarse hypergranular layer, focal acanthosis and papillomatous projections of the epidermis. Within the epidermis there were foci of keratinocytes

in a cystic configuration maturing to a basket woven corneum representing infundibular differentiation. The lesion exhibited an intraepidermal aggregate of heavily pigmented cells with refractile keratinization and ghost cells resembling matrical cells. In addition there were vertically oriented structures closely mimicking an early follicle composed of an inner layer of cells with eosinophilic cytoplasmic granules (trichohyaline granules) as seen in inner sheath differentiation. Rimming the cells with eosinophilic granules were keratinocytes with pale cytoplasm resembling outer root sheath cells. In the dermis there were scattered aggregations of fibroblasts and papillary mesenchymal bodies, as seen in follicular germ/hair papillae. The extent of follicular differentiation in epidermal lesions is usually restricted, mimicking one or two component of the pilar unit. In contrast this rare case demonstrates that advanced follicular differentiation can present as an epidermal lesion similar to the dermal papilloma.

Poster 609

FELLOW

A case of multiple cutaneous leiomyomas

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We present a case of multiple cutaneous leiomyomas. Clinical, histologic and genetic features of multiple cutaneous piloleiomyomas are reviewed, with emphasis on cutaneous leiomyomas as a potential dermatologic indicator of an underlying cancer-associated genodermatosis, hereditary leiomyomatosis and renal cell cancer syndrome (HLRCC). A 47 year-old woman presented with multiple, mildly tender papules, which were increasing in number. She developed the first lesions on the right upper arm approximately six years prior to presentation. Clinically, her skin exam was remarkable for numerous individual and grouped, 0.2 - 1.5 cm pink to red-brown smooth, rubbery papules and nodules, distributed on the right arm, central chest, right back and left lower leg. Her medical history was significant for a hysterectomy for symptomatic uterine leiomyomas. Her family history was remarkable for similar lesions in her mother and her daughter, which have not been formally diagnosed. A biopsy was obtained from a characteristic 0.4 cm papule within a larger cluster of papules scattered over a 10 cm area on the right back. Histologically, the initial biopsy showed a smooth muscle proliferation with occasional hyperchromatic nuclei and rare mitotic figures. Because of mitotic activity, there was concern for leiomyosarcoma, and additional sampling was performed. Punch biopsies of three different lesions showed similar features, although less mitotic activity, and in the context of the original biopsy, all lesions were diagnosed as benign leiomyomas. The patient was referred to genetics for fumarate hydratase testing. Considerations for genetic testing and clinical implications of multiple cutaneous leiomyomas are discussed.

Poster 610

FELLOW

Loss of mismatch repair protein expression in cutaneous squamous cell carcinomas of Muir-Torre patients

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In Muir-Torre syndrome a heritable defect in mismatch repair genes drives tumorigenesis, leading to sebaceous neoplasms and other visceral malignancies. It has previously been well-characterized that inherited and acquired defects in mismatch repair proteins can be reliably detected in sebaceous neoplasms with commonly available immunohistochemical stains. In order to examine whether defective mismatch repair drives tumorigenesis in non-sebaceous cutaneous malignant neoplasms in patients with Muir-Torre, we describe the staining patterns of cutaneous invasive squamous cell carcinomas, in situ squamous cell carcinomas, and basal cell carcinomas of six patients with sebaceous neoplasms that previously demonstrated defects of immunohistochemical expression of mismatch repair proteins. Three of these patients have gene-confirmed Muir-Torre syndrome and the gene status is unknown in the other three patients. Five in situ squamous cell carcinomas, three basal cell carcinomas, and four invasive squamous cell carcinomas were stained with antibodies to MLH1 (NovoC, clone E505, 1:200, Bond autostainer), PMS2 (BD, clone A16-4, 1:200, Dako autostainer), MSH2 (Calbiochem, clone FE11, 1:400, Dako autostainer), and MSH6 (BD, clone 44, 1:400, Dako autostainer). The only lesions demonstrating absence of mismatch repair protein expression were invasive squamous cell carcinomas in two patients with gene-confirmed Muir-Torre syndrome. All basal cell carcinomas and in situ squamous cell carcinomas demonstrated intact staining for all patients. Mismatch protein expression loss in the invasive squamous cell carcinomas correlated with the mutated gene for those patients.

Poster 611

FELLOW

Lessons from a challenging case of basal cell carcinomas mimicking trichoepitheliomas

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The application of immunohistochemical stainings has been advocated in the differential diagnosis of basal cell carcinoma vs. trichoepithelioma. We recently encountered a case of a 37 year old female with pearly facial papules. Among three biopsies done, one showed histologic features typical of a basal cell carcinoma, one was similar to a trichoepithelioma; the third one

has features of both. Interestingly, stains for Bcl2, Ki67, CD34, CD10 and CK20 could not conclusively distinguish basal cell carcinoma from trichoepithelioma in the latter two cases. The more the lesion morphologically resembled a trichoepithelioma, the more staining characteristics of trichoepithelioma it had. Because the two lesions possessed both morphological features of trichoepitheliomas and basal cell carcinomas, a working diagnosis of basal cell carcinoma, infundibulocystic type, was favored. Additional clinical information of this patient, as well as family history suggested that this patient had Nevoid Basal Cell Carcinoma syndrome (Gorlin syndrome), further corroborating the diagnosis of basal cell carcinomas. This case illustrates that Gorlin syndrome may imitate multiple 'trichoepithelioma-like' lesions present on tissue sections. It also highlights the fact that the immunohistochemical staining patterns being used to distinguish trichoepitheliomas from basal cell carcinomas were mostly derived from studies using more conventional types of basal cell carcinoma, thus may not be informative enough for the correct diagnosis of infundibulocystic type of basal cell carcinoma.

Poster 612

FELLOW

Axillary lesions in a patient with nevoid basal cell carcinoma syndrome: a diagnostic dilemma

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Nevoid Basal Cell Carcinoma syndrome (NBCCS) is a hereditary condition associated with multiple developmental abnormalities and a predisposition to basal cell carcinomas. We present a case of a 13 year old female with a history of NBCCS who presented to dermatology for a skin exam and was found to have multiple flesh colored to brown, pearly papules on the nose, peri-nasal and infra-orbital regions as well as multiple flesh-colored soft pedunculated papules in the bilateral axilla. Two of the axillary lesions were biopsied. The biopsies demonstrated an irregularly shaped proliferation of squamoid cells with small keratin cysts and branches of basaloid cells with peripheral palisading arising from the epidermis, extending into the superficial dermis. The surrounding stroma was fibrous to myxoid. CD34 labeled stromal cells but was negative in the epithelial component. BCL2 and BER-EP4 predominantly labeled the peripheral epithelial cells. CD10 labeled stromal cells with focal staining at the periphery of the epithelial cells. The morphologic features and immunohistochemical staining were felt to be most consistent with trichoepithelioma. This case represents a unique presentation of non-basal cell carcinoma skin lesions in a patient with NBCCS, and highlights challenges that may arise in diagnosing follicular neoplasms in such patients as well as issues that may arise in the process of cutaneous surveillance.

Poster 613

FELLOW

Cutaneous Ewing's sarcoma mimicking malignant melanoma: a potential diagnostic pitfall

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We report an unusual case of cutaneous Ewing's sarcoma involving the abdominal skin and subcutaneous tissue in a 19 year old female with no prior medical history. The case was originally misdiagnosed as a small cell melanoma based on diffuse and strong S100 immunoreactivity. Additional immunostains showed that neoplastic cells expressed CD99, CD56, and variable nuclear FLI-1. Neoplastic cells were negative for melanocytic markers including Melan-A, tyrosinase, MITF, and HMB-45, as well as cytokeratins, CD45, desmin, and c-terminus WT1 arguing against melanoma, carcinoma, lymphoma, rhabdomyosarcoma, and desmoplastic small round cell tumor. FISH showed EWS gene translocation. RT-PCR confirmed the presence of EWS-FLI1 fusion transcript, further supporting the diagnosis of Ewing sarcoma. We report the clinical, histologic, immunophenotypic, and molecular features of this unusual case and discuss the differential diagnosis of cutaneous small round blue cell tumors.

Poster 614

FELLOW

The role of immunohistochemistry in predicting Muir Torre syndrome in patients presenting with cutaneous sebaceous lesions

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Background: Sebaceous skin tumors can be sporadic or syndromic, associated with the Muir Torre syndrome (MTS). Previous studies have shown that immunohistochemistry (IHC) for mismatch repair proteins (MMR) can detect sebaceous lesions of MTS. We present immunohistochemical and genotyping results from a large case series. Materials, methods: The pathology archives were searched for all cutaneous sebaceous lesions diagnosed between 2003-2012, along with the patient clinical information. Immunohistochemical studies for the 4 MMR proteins and molecular data (germline mutations in mismatch repair genes) were retrieved. Results: We identified 137 sebaceous lesions, from 55 patients. Immunohistochemical studies and molecular testing were available for 33 patients. From this group, 22 patients had loss of one or more MMR proteins by IHC (67%) and 11 (33%) had intact MMR expression. 12/22 (55%) patients with MMR loss also had associated internal malignancies; 5/12 patients also harbored germline mutations. 10 patients had MMR loss (IHC), but no known visceral malignancy.

Interestingly, in 4/10 patients, germline mutation was identified. 11 patients had MMR intact; 5/11 had internal malignancies, 1/11 harbored germline mutation. Out of the 55 patients, 32 have confirmed MTS (by combined clinical and molecular criteria). Of note, in 7/32 (22%) patients, the sebaceous lesions diagnosis preceded the internal malignancies. Conclusion: We correlate clinical germline genotyping for MSI genes with IHC for MMR proteins. Our data indicates that loss of MMR by IHC in cutaneous sebaceous neoplasia is predictive of association with MTS. In a substantial subset of patients, characterization of the cutaneous sebaceous neoplasia leads to a clinical suspicion for MTS prior to the diagnosis of internal malignancy.

Poster 615

FELLOW

A case of syringoid eccrine carcinoma treated with Mohs surgical excision

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Syringoid eccrine carcinoma is an extremely rare tumor thought to arise from the eccrine apparatus, although its origin is controversial. It usually presents as a slow-growing plaque on the scalp, and less frequently, the extremities or trunk. We present a case of syringoid eccrine carcinoma in a 48 year old man, treated with Mohs excision. During a follow up visit for an excision of basal cell carcinoma, the patient noted a pruritic area on his scalp that had been present for one year. Examination showed a 5 x 3 cm depressed telangiectatic patch on the scalp vertex, with a clinical differential diagnosis including squamous cell carcinoma in situ and discoid lupus erythematosus. Two biopsies were performed, attempting to sample lesional and nonlesional areas. Both of the biopsies consisted of an infiltrative ductal epithelial proliferation with features of syringoid eccrine carcinoma. Mohs surgery was performed, spanning nine hours and requiring seven stages. The operative wound measured 12.8 x 7.5 cm and extended to the periosteum. Half of the wound was closed using a Posterior scalp rotation flap, with delayed skin grafting used for the remainder. The patient had an uncomplicated recovery with no evidence of recurrence three years later. Syringoid eccrine carcinoma has a propensity for local recurrence, so complete excision is imperative but can be difficult due to the tumor's infiltrative nature. It is this deeply infiltrative pattern that helps differentiate it from syringoma, while its basaloid cells distinguish it from the more squamoid appearance of microcystic adnexal carcinoma.

Poster 616

FELLOW

Eruptive angiokeratoma-like angiomas in the setting of chronic cutaneous graft versus host disease after allogenic bone marrow transplantation

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A 22-year-old man with acute lymphoblastic leukemia underwent allogenic bone marrow transplant (BMT) in June 2010 followed by immunosuppression with tacrolimus, mycophenolate and prednisone, as well as antimicrobial prophylaxis with dapson, acyclovir and voriconazole. 17 months after transplantation he developed chronic cutaneous graft-versus host disease (cGVHD). As of February 2012 apart from the development of actinic keratoses and squamous cell carcinomas (SCCs) that led to the discontinuation of voriconazole, multiple violaceous macules and nodules arose in the areas clinically affected by cGVHD. Microscopic findings of 4 biopsy specimens were similar and showed proliferations of dilated thin-walled vessels containing erythrocytes immediately beneath the epidermis that was hyperplastic, with compact hyperkeratosis and ulceration. Adjacent to such proliferations, sections showed chronic interface dermatitis compatible with c-GVHD. Vascular tumors in the setting of BMT or cGVHD, distinct from Kaposi sarcoma or bacillary angiomatosis, have rarely been reported except for 4 previous cases of eruptive cutaneous angiomas on a background of cGVHD. All cases appeared 1 to 3 years after BMT and were histologically classified as pyogenic granulomas or cavernous hemangiomas. The pathogenesis of this phenomenon remains obscure. Previous hypotheses have included a hyperplastic reaction to cutaneous injury; impaired vascular circulation with increased capillary pressure and, in turn, vascular ectasia; and a dysregulation of angiogenic factors, possibly related to the production of inflammatory cytokines by keratinocytes affected by cGVHD. We report the fifth case of eruptive angiomas in the context of cGVHD to help further define this entity and create awareness of this rare association.

Poster 617

FELLOW

Keratoacanthomas: an immunohistochemical analysis to define its relationship to squamous cell carcinoma using tissue microarray

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There has been debate for decades over whether keratoacanthomas are a specific subset of squamous cell carcinomas (SCCA) or if they are an entity sui generis. Since at least a subset of them involutes, they might represent a neoplasm with different biologic behavior as compared to traditional SCCA. Given the long established controversy and the newer techniques of evaluating substantial quantities of tissue samples via tissue microarray, immunohistochemical evaluation of samples via this method to define potential differences was performed. We built a tissue microarray with 38 keratoacanthomas, 27 keratoacanthomas with features of regression, 18 "atypical squamous neoplasms" (lesions with architectural features of keratoacanthomas but without cytologic atypia) and 20 well differentiated SCCA. A panel of immunohistochemical markers including Bcl-2, Bcl-6, p53, CD 117, CD34, p63, Cyclin D1, Ki-67, and E-cadherin was performed on each microarray. Subtle differences were observed, most notably with E-cadherin where a higher percentage of SCCA stained stronger than in the other three entities, particularly keratoacanthomas without regression (33% of SCCA stained with 3+ intensity as compared to 0% 3+ staining intensity in keratoacanthomas without regression). We concluded that there are slight differences in the immunohistochemical profile of keratoacanthomas without regression and well differentiated SCCA. Many immunohistochemical markers stained in a similar pattern, supporting the view that keratoacanthomas are a specific clinical subset of SCCA. However, the difference observed with E-cadherin provides evidence to support, at least, different protein expression in these two neoplasms, perhaps fueling the ongoing debate over the histogenesis of these lesions.

Poster 618

FELLOW

Correlation of pHH3 to Ki-67 protein expressions in leiomyosarcomas

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Phosphohistone H3 (pHH3) immunohistochemistry serves as a relatively specific mitotic marker. In contrast, the Ki-67 (MIB1) antibody labels all proliferating cells including those in interphase and during mitosis. However, is the

number of proliferating cells labeled by Ki-67 proportional to the number of cells in mitosis labeled by pHH3? Using ImageJ, a public domain, Java-based image-processing program, we analyzed Ki-67 and pHH3 labeling indices in five cases of cutaneous leiomyosarcoma. In each case, 10 fields of tumor were counted. Each field contained 454-612 cells. By use of Ki67 and pHH3, an average of 67.9 cells (13.6 %) were proliferating (range 62.5-74.6) and 3 cells (0.63%) were in mitosis (range 1.2-4.1), respectively. The pHH3:Ki-67 ratio in five tumors are 0.019, 0.022, 0.030, 0.055, and 0.073 with an average of 0.039 (3.9% of all proliferating cells were undergoing mitosis). The ratio of the latter two cases displayed increased tumor-infiltrating lymphocytes. Human cells in culture divide every 24 to 28 hours, spending ~1 hour in M-phase (~3-4%). The pHH3:Ki-67 ratio in our cases implies that the tumor cells spend ~4% of their time, or one hour per 24 hours, in M-phase (range 0.46-1.75 hours per 24 hours), similar to the time observed in cultured human cells, assuming that the individual mitotic activity is a randomized phenomenon. It appears that we see a good correlation of Ki-67 and pHH3 labeling in cutaneous leiomyosarcomas, with pHH3 staining being cleaner and easier to interpret. Herein, we provide data from an expanded study that includes melanoma.

Poster 619

FELLOW

Hair follicle nevus

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Hair follicle nevus is a rare hamartomatous follicular lesion that presents as a 3 to 10 mm, flesh-colored to erythematous, domed or pedunculated papule on the face. To our knowledge, only thirty cases have been reported in the literature. It is usually solitary; however four cases of multiple lesions have been reported. The classification, nomenclature, and even diagnostic criteria of the lesion have been subjects of debate and confusion since the term was first coined in 1928. Currently, hair follicle nevus is considered to be a follicular hamartoma consisting of a collection of closely spaced, variously oriented vellus follicles at different stages of maturation located high in the dermis. Small sebaceous glands, smooth muscle fibers within the stroma, and mononuclear inflammatory infiltrate may be present. The hair follicle nevus must be differentiated from trichofolliculoma and accessory tragus, among others. Careful clinical and histopathologic examination, including serial sections, is necessary. Treatment is surgical excision. We present a case of a hair follicle nevus arising in the left nasolabial fold of a 33-year-old black woman, present for over four months. A 3 mm punch biopsy completely excised the lesion. Microscopically, a well-circumscribed proliferation of closely-spaced, fully-formed hair follicles surrounded by prominent connective tissue sheaths was confined to the upper aspect of the dermis. Based on the findings, a diagnosis of hair follicle nevus was rendered.

Poster 620

FELLOW

Syringocystadenocarcinoma papilliferum: a report of 2 cases

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Syringocystadenocarcinoma papilliferum (SCACP) is a rare cutaneous adnexal tumor. We present two cases. Case 1 presents as an eroded nodule on the arm of a 75 yo woman. Histologically case 1 shows areas with typical syringocystadenoma papilliferum (SCAP) with papillomatous invaginations lined by apocrine epithelium with abundant plasma cell infiltrate. Arising from the SCAP is an invasive squamous cell carcinoma as well as an invasive adenocarcinoma. Case 2 presents as a scalp mass in a 68 yo man, present since birth, but enlarging and bleeding for the past year. Case 2 shows architecture reminiscent of SCAP with papillary structures with apocrine epithelial lining and abundant plasma cell infiltrate. Arising from the SCAP is a poorly differentiated invasive adenocarcinoma. SCACP is a rare tumor that has been shown to arise most commonly on the head and neck and in the perianal area. It is believed to arise from long-standing SCAP. Histologic features suggestive of typical SCAP are characteristically seen. Malignant transformation in the form of both in situ and invasive squamous cell carcinoma and adenocarcinoma have been reported. Notable histologic features that have been reported include central dirty necrosis in adenocarcinoma and pagetoid spread of tumor cells into the overlying epidermis. SCACP is a rare entity that should be considered in the differential diagnosis of a malignant adnexal carcinoma along with metastatic adenocarcinoma, squamous cell carcinoma and apocrine carcinoma. Although clinical experience is limited, SCACP has been treated with complete excision and thus far distant metastasis has not been reported.

Poster 621

FELLOW

A giant sarcomatoid basal cell carcinoma with heterologous osteosarcomatous elements on the arm of a centenarian

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A centenarian woman was brought to the dermatologist for evaluation of a nodule on the extensor aspect of her left arm. The woman was unable to provide information as to how long the tumor had been present. Examination revealed a 9.4 x 5 cm firm, exophytic tumor with superficial ulceration. The mass was excised and sent for histopathologic evaluation. Histologic sections demonstrated a multinodular basaloid neoplasm with characteristic features of a basal cell carcinoma (i.e. peripheral palisading, clefting with stromal mucin,

apoptotic cells). Deep to and merging with the typical basal cell carcinoma components were extensive sheets of pleomorphic spindle cells displaying “lace-like” osteoid deposition intimately associated with tumor cells, features diagnostic of osteosarcoma. These findings were interpreted as sarcomatoid basal cell carcinoma with heterologous osteosarcomatous elements. Sarcomatoid basal cell carcinoma with osteosarcomatous differentiation is a rare entity with few case reports and one small case series in the literature. To our knowledge, this is the largest example of this rare tumor type reported to date, and it expands the current literature on this unusual entity.

Poster 622

FELLOW

Anaplastic Kaposi’s sarcoma: a rare variant mimicking angiosarcoma

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A 65-year-old Nigerian male with a history of Kaposi’s sarcoma (KS) presented with recurrent disease of the left ankle. Previous treatment with excision, radiation, and intralesional bleomycin had led to temporary partial responses. Examination of the left ankle revealed a fungating mass, which was excised. Histology showed a nodular and focally necrotic tumor composed of large cells exhibiting nuclear pleomorphism, prominent nucleoli, and atypical mitotic figures. Scattered eosinophilic hyaline globules were noted. There was diffuse positive staining of tumor cells with CD31 and D2-40 and focal nuclear staining with HHV-8. Previous biopsies were reviewed and showed typical KS histology of trapped erythrocytes between spindled cells, which stained diffusely with HHV-8. Interestingly, progressive histologic dedifferentiation was appreciated on serial examination of previous biopsies with increasing cellular pleomorphism. “Anaplastic” or “pleomorphic” KS, a rare aggressive variant, has been described in classic, HIV-associated, and African endemic forms. It is characterized by significant cellular pleomorphism that often obscures the vasoformative nature of the lesion. Areas of typical KS histology and positive HHV-8 staining allow for distinction from angiosarcoma (AS). However, whether anaplastic KS represents high-grade transformation of KS or the development of true AS remains controversial. Further, while case reports have suggested that chronic edema, radiation, or immunosuppression may trigger anaplastic transformation of KS, this relationship has not been confirmed. As treatment and prognosis can differ between anaplastic KS and AS, correct diagnosis is critical. In our case, positive HHV-8 staining and previous biopsies consistent with typical KS support a diagnosis of anaplastic KS.

Poster 623

FELLOW

The mark of malignant invasion: laminin-332 alpha3 G45 domain in SCC carcinogenesis

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Because the dominant paradigm that cancer is a genetic disease—driven by unregulated oncogenes and tumor suppressor genes—has held sway, the stroma’s critical role in carcinogenesis is often overlooked. Going against the grain, we focus on the basement membrane protein laminin-332 and identify its retained alpha3 G45 domain in promoting squamous cell carcinogenesis, which we demonstrated in previous basic science experiments. The current study, the first that we are aware of, translates laminin-332 alpha3 G45’s biological findings into a human clinical context for diagnosing SCCs. We show that, in normal human skin, laminin-332 is minimally present in the basement membrane with total loss of the proteolyzed G45 fragment. By contrast, in our SCC case series, we reveal the abundance of cytoplasmic and extracellular staining of laminin-332 and G45 retention in invasive SCCs and, conversely, their absence in actinic keratoses and paucity in SCC in-situ. The results weaken the conventional notion that the basement membrane succeeds to protect the host from SCC invasion and offers insight into how carcinomas co-opt the host stromal defense. Further, we confirm that laminin-332 and G45 staining are positive in a small case series of patients with recessive dystrophic epidermolysis bullosa who develop SCCs. Lastly, we apply the diagnostic utility of these two stains in challenging cases in which inflamed seborrheic keratoses and lichen planus can mimic SCCs. Our findings argue that, in thinking outside of the box, the key to unlocking difficult SCC cases may very well lie outside of the cell.

Poster 624

FELLOW

An unusual immunophenotype of a cutaneous CD4+ T-cell lymphoma preceded by pyoderma gangrenosum-like lesions and cutaneous tuberculosis infection

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We report a case of a 68 year old gentleman who had ulceration in the groin area and lower extremities. Based on the clinical presentations and biopsy pathology, his lesions were treated with steroids, Kenalog injections and Cellcept for pyoderma gangrenosum and only achieved limited improvement. Later, tissue culture revealed mycobacteria tuberculosis and he received four-drug antituberculosis regimen. Despite therapies, his ulcerative lesions further progressed and a year later the biopsy showed a dense, diffuse dermal infiltrate of

large lymphocytes with vesicular to hyperchromatic nuclei and occasional prominent nucleoli that are positive for CD4 with aberrant loss of CD5 and CD7 and variable expression of CD25 and CD56 but with only rare TIA-1 expression. The T cell receptor gene rearrangement study is positive for a clonal T cell population. The differential diagnosis could include mycosis fungoides, adult T cell leukemia/lymphoma, T cell prolymphocytic leukemia and cutaneous small-medium pleomorphic T-cell lymphoma but the immunophenotype and the clinical presentations did not support the classic definition of any of the above categories. Since the tuberculosis infections are known to be associated with CD4 T cell functions and pyoderma gangrenosum might relate to abnormal T helper cell to T suppressor cell ratio, we report this unique case to document any potential associations among the manifestations in this unique type of lymphoma.

Poster 625

FELLOW

Two cases of squamous cell carcinoma arising within a sternal scar years after coronary artery bypass surgery

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Squamous cell carcinoma (SCC) arising in areas of chronic inflammation and vascular compromise is well-described. While cardiac procedures are common, reports of SCC arising in scars associated with coronary artery bypass graft surgery (CABG) are few. We describe two cases of elderly men with invasive SCC developing in their sternal CABG scars. One developed a 4 cm mass 17 years after CABG. Histopathology showed a moderately to poorly differentiated invasive SCC in the dermis with no epidermal connection, 3.75 mm deep, with peri- and intraneural invasion. No lympho-vascular invasion (LVI) was seen. Six months after surgical removal by Mohs procedures (X2) and adjuvant radiation, a new dermal lesion of poorly differentiated invasive SCC developed to the left of the scar. The other man developed a 1.5 cm cystic and solid lesion in his scar 3 years after CABG. Pathology revealed a moderately differentiated invasive SCC with perineural invasion (PNI) but no LVI. The tumor focally connected to the epidermis through a sinus with associated chronic inflammation. A year later the patient developed a 5.5 cm metastasis in the left axilla of moderately to poorly differentiated squamous cell carcinoma, likely representing a replaced lymph node with extracapsular extension. PNI and LVI were present in adjacent soft tissue. These two cases illustrate aggressive SCCs arising within CABG scars. The cases are histologically less differentiated than typical Marjolin’s ulcers; however both exhibit aggressive clinical behavior. Awareness of this possibility should lower the threshold to biopsy changing sternal scars, especially years after surgery.

Poster 626

FELLOW

Histopathologic features of keratoacanthoma that aid in differentiation from other common histologic mimics

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Keratoacanthoma (KA) is a benign neoplasm usually presenting as a hyperkeratotic nodule on sun-exposed skin. While often the diagnosis of KA is not difficult, it does share some histopathologic similarities to other skin conditions. These entities include prurigo nodularis (PN), hypertrophic lichen planus (HLP), verruca vulgaris (VV), and squamous cell carcinoma (SCC). Since a correct diagnosis of KA is important in the management of patients, we decided to study the histopathological differences between KAs and other common histologic mimics. We collected slides over the past 6 months of cases given any of the above diagnoses, and then singled out cases that demonstrated prominent hyperplasia with dyskeratosis and/or crateriform architecture, in order to determine which histological criteria may aid in differentiating between KAs and the other previously mentioned entities. We found that KA, when compared to the others, had endoexophytic growth, whereas VV were primarily exophytic, and PN and HLP were primarily endophytic. SCC was noted to be either the crateriform type or the KA type. The former lacked microabscesses, displayed severe atypia, and had a more predominantly endophytic growth, while the latter had microabscesses and an endoexophytic architecture but was distinguishable from KA by prominent atypia admixed into the typical dyskeratotic KA lobules. In addition, wedge-shaped hypergranulosis over the dyskeratosis and hemorrhage into the cornified layer suggested VV, dilated tortuous blood vessels along the dyskeratotic rete ridges was associated with PN, and presence of numerous degenerated keratinocytes and peripheral lichenoid infiltrate with saw-toothed rete ridges distinguished HLP.

Poster 627

FELLOW

An unusual clinical presentation of a rare histiocytic proliferative disorder

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Proliferations of indeterminate cell histiocytes (indeterminate cell histiocytoses, IDC) are rare and poorly defined in the literature. Reported clinical spectrum of presentations of IDC include solitary nodules and disseminated lesions consisting of papules or plaques on

the face, neck, trunk or extremities. IDC has been reported in both children and adults. We present a case of IDC with unusual clinical features of multiple skin colored papules associated with a cafe'-au- lait spot on the chest of an 8 year old boy. The papules initially appeared at the periphery of the spot and subsequently developed within the lesion. Several punch biopsies showed similar histologic features of a folliculocentric and dermal proliferation of histiocytoid cells with reniform nuclei and abundant cytoplasm with a mixed inflammatory background of lymphocytes and eosinophils. A subset of the dermal cells appeared more spindled/dendritic in shape. The cellular infiltrate diffusely expressed S-100 and CD1a with focal expression of CD68. The cells were negative for Langerin consistent with a diagnosis of IDC. The follow-up is still too short to assess a biological potential of this unusual presentation and to determine whether the indeterminate cell proliferation is reactive or represents a neoplastic process.

Poster 628

FELLOW

Source of gastrointestinal bleeding diagnosed with skin biopsies: a clinical, pathologic, and endoscopic correlation of blue rubber bleb nevus syndrom

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Blue rubber bleb nevus syndrome is characterized by multiple compressible vascular malformations of the skin. Involvement of the gastrointestinal tract is also known to occur. We present a case of a 29 year-old male who had a long history of intermittent, severe, hematochezia approximately twice per month. Multiple blue-gray, tender, lesions were also noted on exam, several of which were found on the sole of the left foot. Three of these lesions were biopsied and the histologic findings included multiple dilated thin-walled vascular channels occupying the entire thickness of the dermis, with extension into the subcutis. The vessels were well-formed, and no infiltrative cellular process was identified. The endothelium lining the vascular spaces was bland. Focal areas of intravascular thrombus with organization were present. Immunohistochemical stains were prepared on all three biopsies; the endothelium was highlighted with a CD31 stain. A stain for podoplanin (D2-40) was negative, thus excluding lymphangiomas. The patient was referred to a gastroenterologist who performed an endoscopic examination of the patient's gastrointestinal tract and found a dome-shaped, red-purple lesion in the gastric antrum adjacent to the pylorus, and another similar appearing lesion in the descending colon. Neither lesion showed signs of infection or active bleeding, so no treatment was performed. Close clinical follow-up and management of bleeding is the planned course of management.

Poster 629

FELLOW

Dermal and intraepidermal Merkel cell carcinoma presenting with squamous cell carcinoma in situ

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Merkel cell carcinoma (MCC), usually presents as a dermal tumor and only occasionally, it is confined to the epidermis. A group of MCC are known to arise in collision with other tumors, usually with squamous cell carcinoma (SCC), referred to as combined-MCC, and its development have been suggested to be associated with chronic ultraviolet damage and also from abnormal differentiation of a common stem cell. A very few cases of MCCIS combined with SCC have been reported in the literature. We present three collision cases of MCC with SCC, in one of the cases the MCC component was MCCIS. Case 1 and 2 presented in a 81 year-old male in the left temple and 77 year-old male in the anterior neck, respectively, microscopically, both tumors showed similar appearance characterized by malignant basophilic cells in irregular aggregates in the dermis consistent with MCC, the cells were positive for chromogranin and CK20, additionally, the epidermis showed acanthosis with lack of nuclear maturation consistent with SCCIS. Case 3 presented in a 77 year-old female in the nose, microscopically, sections revealed SCCIS and in between the lesion, there were areas showing irregularly shaped aggregates of basophilic hyperchromatic cells, these cells were positive for chromogranin and CK20. These cases are reported to make clinicians aware of this uncommon presentation of MCC.

Poster 630

FELLOW

A case of cutaneous inflammatory myofibroblastic tumor

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Inflammatory myofibroblastic tumors, sometimes called plasma cell granulomas, are rare benign spindle cell proliferations that are more commonly found in the lungs, bladder and gastrointestinal system than on the skin. We report this cutaneous case found on the arm of a 19 year old male. The nodular lesion clinically resembled a lipoma. On histologic examination, the specimen shows a predominantly storiform arrangement of elongated and bland spindle cells without significant atypia or pleomorphism in a collagenous stroma. There is a mixed inflammatory infiltrate consisting of plasma cells (some with Russell bodies), lymphocytes, histiocytes, and few eosinophils present densely in random foci but also interspersed throughout. Few distinct lymphoid follicles are present. The non-transected edges of the lesion show a fibrous pseudocapsule, suggesting good circumscription.

Immunostaining of the spindle cells is positive for smooth muscle actin, weakly positive for CD68, and focally positive for procollagen I and CD34. These findings highlight the myofibroblastic nature of the tumor and show some concordance with previously reported cases.

Poster 631

FELLOW

The mitotic marker phosphohistone H3 (PHH3) in the assessment of pilar leiomyomas and cutaneous leiomyosarcomas

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The accurate determination of the mitotic count is a critical component in the classification of tumors which exhibit cellular atypia such as leiomyomas and leiomyosarcomas. But the process is often plagued by difficulties. Phosphohistone H3 (PHH3) immunostain which is a relatively specific marker of mitosis has been shown to be an easier method to determine the mitotic index (MI) in a variety of tumors. In our study we evaluated the utility of the anti PHH3 antibody in quantifying mitotic figures in 20 pilar leiomyomas (LM) and 20 cutaneous leiomyosarcomas (LMS) comparing PHH3 and hematoxyline and eosin (H&E) stained sections. Mitotic figures were more readily identified on PHH3 sections. Minimal difference in MI between PHH3 and H&E sections not exceeding 4/10 high power fields (HPF) in the LMS group and 0-1/10 HPF in the LM group were observed. The differences were not statistically significant in both groups (p=0.7). The higher MI obtained on PHH3 section of both the LM and LMS group is most likely due to the ability of PHH3 to detect early mitosis characterized by the phosphorylation of histone H3 which begins in prophase but difficult to identify on H&E sections. Out of the 4 leiomyosarcoma lesions which showed a higher MI on H&E, three cases (75%) had a considerable number of degenerating/pyknotic nuclei mimicking mitotic figures on H&E. We conclude that the PHH3 is an easier and rapid immunostain to quantify mitotic figures in smooth muscle neoplasm of the skin in comparison to the conventional H&E.

Poster 632

FELLOW

Primary cutaneous myxoid spindle cell squamous cell carcinoma: a rare variant with CD10 expression

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Primary cutaneous myxoid spindle cell squamous cell carcinoma (MSC SCC) is a rare variant of SCC with only a handful of reports in the literature. In 2010, Yang, et al. published criteria for the diagnosis of MSC SCC including: a predominant population of poorly differentiated spindle cells, prominent myxoid stroma involving >50% of the

lesion, and neoplastic spindle cells positive for at least one cytokeratin. We report two cases of MSC SCC that fulfill the above criteria. Both cases occurred in elderly men who presented with crusted papules on the ear; histologically, the lesions consisted of pleomorphic spindle cell tumors in a prominent myxoid stroma. The spindle cells in both cases expressed pankeratin and p63; additionally, a subset of the neoplastic cells expressed CD10. Both tumors negatively expressed CD31, S100, Cam 5.2, smooth muscle actin, and desmin. To the best of our knowledge, CD10 expression in MSC SCC has not been previously reported. CD10 is useful in the diagnosis of atypical fibroxanthoma (AFX), where its expression is positive in the majority of AFX. It is not specific for AFX, however, and may be expressed in a number of other cutaneous tumors, including a minority of SCC. Our current findings raise several important points. Firstly, MSC SCC is a rare tumor that must be differentiated from other malignancies, such as myxofibrosarcoma and myxoid AFX. Secondly, CD10 expression in MSC SCC perhaps lends further evidence that spindle cell SCC and AFX are closely related tumors, if not variants of the same entity.

Poster 633

FELLOW

Cutaneous myoepithelioma with cartilaginous metaplasia: a new (but not surprising) histopathologic feature

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Cutaneous myoepitheliomas are rare, benign skin tumors composed of myoepithelial cells without ductal differentiation. Cartilaginous metaplasia within cutaneous myoepitheliomas has not been described, although myoepithelial cells are known to differentiate along both epithelial and mesenchymal lineages. Cutaneous mixed tumors/chondroid syringomas are also on the spectrum of myoepithelial neoplasia and display distinctive chondromyxoid stroma; however, these lesions have prominent ductal differentiation, in contradistinction to myoepitheliomas. We present a 37-year old man with a flesh-colored nodule on the left flank. Lesional histopathology showed a dome-shaped tumor with a broad central zone of cartilaginous differentiation. Outside the cartilaginous zone, epithelioid cells with eosinophilic cytoplasm formed a vague syncytium without a ductal component. Epithelial membrane antigen and S100 protein strongly stained the nuclei of the chondrocyte-like cells, as well as the surrounding epithelioid tumor cells, supporting a diagnosis of cutaneous myoepithelioma. Focal staining with cytokeratin antibodies, Cam 5.2 and AE1-3 was observed, while cytokeratin 116 staining was negative. Melan-A and calponin staining were both negative. Our case demonstrates that cartilaginous differentiation in cutaneous myoepitheliomas may be prominent and striking—a finding which is not surprising due to the ability

of myoepithelial cells to differentiate along mesenchymal lines. Cartilaginous metaplasia could pose a potential pitfall for misclassification of a myoepithelioma as a benign cartilaginous tumor. To the best of our knowledge, this is the first reported case of a cutaneous myoepithelioma with cartilaginous metaplasia.

Poster 634

FELLOW

Fibrokeratoma: non-digital type

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We present six cases of fibrokeratoma arising on non-digital sites. Digital fibrokeratoma is a tumor growth that presents as a firm flesh-colored projection, most commonly on the fingers and toes, with occasional nail involvement. The lesion occurs mainly in adults and arises more frequently amongst males. There are, however, rare reports in which fibrokeratomas have developed on other sites including the upper lip, palm, sole, elbow, wrist, calf and prepatellar regions. We present five cases of non-digital fibrokeratomas arising on the elbows, trunk, knee, and heel. Fibrokeratomas could be misdiagnosed as other generic lesions such as cutaneous horn, verruca vulgaris, Koenen tumor, acrochordon and supernumerary digit. The etiology of this entity remains unknown; however, chronic and repetitive minor trauma may be implicated.

Poster 635

FELLOW

Morphometric analysis of keratoacanthomas

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Keratoacanthoma (KA) is characterized by a rapid growth of a well-differentiated squamous cell carcinoma with prominent cystic components, spontaneous regression, and excellent prognosis. One hundred and twenty four cases of KAs with complete tumor contours on sections, selected from consecutive 471 KAs, are analyzed. Parameters include areas of tumor proper (neoplastic epithelium), keratin-filled space, tumor mass (tumor proper plus keratin-filled space) and the whole tumor contour (tumor mass plus underlying stroma, regression fibrosis and inflammatory infiltrate) are measured by utilizing trichrome staining and Adobe Photoshop program. Our analysis reveals cyst/cavity component of KAs accounts for over 70% of the tumor areas in one third of the cases and over 80% of the tumor areas in 10% of cases. Overall, the cyst/cavity accounts 60% of the tumor areas with a range of 25-91%. Tumor proper to whole tumor ratio is 32% with a range of 7.28 - 59.02%. That is in majority of the KAs the neoplastic epithelial component accounts for only a minority of the total tumor area/volume. In over 20% of the KAs, the ratio is less than 20%. The tumor proper volume to space volume ratio likely will be close to the

area ratios calculated. The presence of significant cystic growth and large cyst/cavity spaces in KAs may explain the rapid growth of the tumors. Extensive inflammation and regression could also contribute to this rapid growth. Correlation of duration of the tumors with microscopic features including presence of tube-like projections, neutrophil pustules, flat cystic configuration, cyst/cavity to tumor ratio.

OTHER

Poster 636

RESIDENT

Spiradenocylindroma arising in epidermoid cyst on the penis

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We report an unusual case of an extremely rare spiradenocylindroma associated with a follicular-cystic lesion arising on the penile shaft. The patient presented with 3 month history of a bleeding lesion on his penis, which was initially concerning for squamous cell carcinoma. Upon excisional biopsy, histologic sections showed a large, centrally dilated follicular-cystic structure with a granular layer, findings suggestive of an epidermal inclusion cyst (epidermoid cyst). Attached to and surrounding this structure were cords and nest of small basophilic cells in a dense, sclerotic stroma, some of which forming a “jigsaw puzzle”-like pattern typical of cylindroma. Other sections were notable for larger lobules with infiltrating lymphocytes, findings more typical of a spiradenoma. These two tumors are not uncommonly found in association with each other, as they are closely-related adnexal ductal neoplasms; however, the association with, or emergence from, an epidermal cyst has not been previously reported, and the location on the penis is clearly unusual. The association of spiradenocylindroma with an epidermal inclusion cyst also is consistent with folliculosebaceous-apocrine unit derivation and apocrine differentiation in these tumors.

Poster 637

FELLOW

Trichoadenoma arising in an unusual location

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Trichoadenoma is a rare, benign tumor of follicular origin that was first described by Nikolowski in 1958. It usually presents as a small, less than 1.5 cm, solitary papule that occurs most commonly on the face and buttocks. Here, we report a 38-year-old African American male who presented with a 4 x 3 cm nodule in the groin that had been present for over 20 years. The initial differential diagnosis based on cutaneous examination included calcified epidermal inclusion cyst; however, on histologic examination the lesion proved to be trichoadenoma. Histologic examination

revealed a well-circumscribed tumor characterized by epithelioid cells forming follicular structures with dilated cystic cavities, resembling the cross-section of the infundibular portion of a hair follicle. No hair shafts were present, and focal areas of foreign body reaction were seen. This patient represents an atypical clinical presentation of trichoadenoma occurring as a large cyst-like subcutaneous nodule in the groin. The presentation of trichoadenoma may be atypical. In such cases, the correct diagnosis may only be reached through histologic examination.

Poster 638

Cutaneous ciliated cyst of the scalp: a case report supporting ciliated metaplasia of eccrine glands

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Cutaneous ciliated cysts (CCC) are rare benign cysts known to occur in the lower extremities of females of reproductive age. Reports of CCC demonstrate that the cyst lining is closely related histologically and immunohistochemically to fallopian tube epithelium. These cysts become clinically symptomatic during high estrogen states such as puberty and pregnancy. The theory of Mullerian heterotopia has been proposed to explain the pathogenesis of this benign entity. However, the concept of abnormal Mullerian rest migration during embryogenesis fail to adequately explain some cases of CCC. We present a case of a scalp CCC that alternatively supports the theory of ciliated metaplasia of eccrine glands. A 53-year-old African American female presented with a life-long history of a firm grey mildly painful nodule on the vertex scalp. Clinical differential diagnoses included nevus sebaceous, blue nevus, and adnexal neoplasm. A biopsy of the lesion revealed a collapsed unilocular cyst in the reticular dermis lined by columnar epithelium, with areas of pseudostratification and apocrine differentiation. Portions of the epithelium consisted of tall ciliated columnar cells with terminal bar. Immunohistochemical evaluation of the cyst lining revealed positive staining for CAM 5.2, epithelial membrane antigen (EMA), carcinoembryonic antigen (CEA), and p63. The cyst lining showed weak positivity for progesterone receptor (PR) and negative immunoreactivity for estrogen receptor (ER). The staining pattern of this ciliated cyst strongly supports eccrine gland differentiation, as opposed to Mullerian origin. There is growing evidence that ciliated cutaneous eccrine cysts are indeed distinct from cutaneous Mullerian cysts.

Poster 639

RESIDENT

Familial basaloid follicular hamartoma: a case report

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We report a case of a 60 year old male patient who had multiple painless skin bumps on his face and upper trunk since his childhood that increased in size over the past few years. On physical examination, multiple papules on his face and arms were present with accentuation around his nose and nasolabial folds. He has been closely followed with multiple biopsies of head and upper trunk lesions which showed small, well-demarcated epithelial proliferation composed of bland squamoid and basaloid cells in anastomosing cords and were diagnosed as basaloid follicular hamartomas (BFH). Basaloid follicular hamartoma is an uncommon benign cutaneous malformation of hair follicles. Because of its clinical and histologic similarities to basal cell carcinoma (BCC) and rare reports of BCC development in BFH, correct identification is important to avoid unnecessary procedures and improve patient monitoring.

typically solitary but usually lack intracranial extension or underlying bony defect. Their immunohistochemical profile with positive EMA and vimentin as well as negative CD31 and CD34 helps distinguish them from vascular malformations such as lymphangioma. These tumors behave in a benign fashion, and surgical excision is curative. Nevus sebaceus is a common congenital hamartomatous lesion composed of folliculosebaceous, apocrine and connective tissue elements. Our case is the first to date to report a meningothelial hamartoma in association with a nevus sebaceus.

Poster 640

RESIDENT

Meningothelial hamartoma associated with a nevus sebaceus

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A 20 month-old girl presented with a 1.5x1.8 cm vascular-appearing cystic lesion underlying a 6x8 cm yellow verrucous plaque on the vertex of the scalp. A staged excision was planned after an ultrasound failed to reveal any underlying structural abnormality of the skull. Histopathology from the initial excision revealed an acanthotic epidermis with budding immature folliculosebaceous follicles and a multiloculated cystic structure extending from the mid dermis into the subcutis. The cystic area was lined by flattened, endothelial-like cells and was surrounded by spindled to dendritic cells infiltrating the nearby collagen. Both the lining and interstitial cells were positive for epithelial membrane antigen (EMA), vimentin, and Factor XIIIa but negative for CD31 and CD34. Some of the dendritic cells stained positively for S100. The diagnosis of nevus sebaceus with associated meningothelial hamartoma was made. Further excisions are planned in the future. Meningothelial hamartoma is a rare lesion characterized by ectopic meningothelial elements in the skin or subcutaneous tissue that was distinguished from meningiomas in 1990 due to the presence of connective tissue elements in addition to the meningothelial cells. These lesions are almost always located on the scalp of young children and are

Poster 641

Reassigned to Poster Session 1

Poster 642

RESIDENT

A rare and lethal disease: junctional epidermolysis bullosa - Herlitz type

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Junctional epidermolysis bullosa (JEB) is an autosomal recessive mechanobullous disease that clinically presents as tense blisters on the extremities and trunk, with variable involvement of the mucosa and nails. Epidermolysis bullosa is extremely rare with the overall U.S. incidence being approximately 19 per one million live births, and JEB only represents a small portion of these. We present the case of a 14-day-old female, normal at birth, who developed extensive tense bullae. Histologic examination of one biopsy specimen displayed a subepidermal blister and the epidermis showed signs of early necrosis. There was superficial papillary dermal necrosis with a lymphocytic infiltrate and scattered eosinophils. Given the clinical context these findings were suggestive of a healing blister from EBS. A second specimen revealed a subepidermal cleft with minimal dermal inflammatory infiltrate. PAS highlighted the basement membrane attached to the dermis beneath the blister. Electron microscopy showed a disruption in the basement membrane with separation through the lamina lucida and a marked reduction or absence of hemi-desmosomes along the basal pole of basal keratinocytes. Based on these findings a diagnosis of Herlitz variant of JEB was given. JEB can be divided into 3 major subtypes: Herlitz-JEB, non-Herlitz-JEB, and JEB with pyloric atresia. The Herlitz type is characterized by the complete absence of laminin-332, which leads to its characteristic EM findings. Herlitz-JEB carries a poor prognosis with death frequently occurring within the first 2 years of life. This case is presented for clinical interest given its rare nature and unique ultrastructural findings.

Poster 643

RESIDENT

Fibroelastolytic papulosis: report of a rare and under-diagnosed entity

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We report a 76-year-old woman with a mildly pruritic rash on the right Posterior neck for 2 years. She denied ocular complaints and history of vascular or ocular disorders. Physical examination demonstrated multiple small, subcutaneous, flesh-colored to yellow papules on the right Posterior neck that coalesced into a 6 cm plaque. Differential diagnosis included pseudoxanthoma elasticum (PXE), fibroelastolytic papulosis (FEP), secondary anetoderma, and focal dermal elastosis. Histologically, extensive elastolysis of the reticular dermis was seen. No calcium deposits, dermal mucin, or features of PXE identified. Given the clinical presentation of numerous yellow papules on the neck and histological findings of papillary dermal elastolysis, a diagnosis of FEP was made. FEP is the projected diagnostic term for cases formerly identified as pseudoxanthoma elasticum-like papillary dermal elastolysis (PXE-PDE) and white fibrous papulosis of the neck (WFPN). FEP is rare, with 7 cases reported since the term was proposed. FEP affects men and women on the neck, supraclavicular region, scalp, axillae, lower abdomen/inguinal region, and/or antecubital fossa. Clinically, isolated, asymptomatic to mildly pruritic small white-yellow papules develop, potentially coalescing into cobblestone-patterned plaques resembling PXE. Histology shows significantly decreased or completely absent papillary dermal elastic fibers, sometimes with thick collagen fibers. The pathogenesis of FEP is unknown; however, it may represent a pattern of intrinsic aging. No successful treatments have been reported, but antioxidants may have potential. We report this rare case to increase awareness of FEP, as the true incidence is likely unknown due to the asymptomatic and benign nature of the disease.

Poster 644

RESIDENT

Amyloidosis cutis dyschromica: a rare variant of primary cutaneous amyloidosis

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Amyloidosis Cutis Dyschromica (ACD) is a rare variant of primary cutaneous amyloidosis. First reported in 1970, ACD has been described as a familial dermatosis presenting in childhood or adolescence as asymptomatic reticulated hyper- and hypopigmented macules involving the body and face. We present a 30 year-old female with diffuse hyperpigmented, firm skin on the face and body beginning in childhood. She later developed hypopigmented macules

diffusely and pruritic bullae on her legs in her early twenties. The patient's brother had an identical presentation on physical examination. Hematoxylin and eosin staining on a punch biopsy of the thigh revealed pale pink, globular homogenous deposits in the papillary dermis, and immunohistochemical staining with CKAE1/AE3 confirmed the presence of keratin derived amyloid. These findings were consistent with the histologic diagnosis of lichen amyloidosis. We present this unique constellation of cutaneous findings in the setting of keratin derived amyloid deposition. This case highlights a less well-known cause of familial cutaneous dyschromia.

Poster 645

FELLOW

Cutaneous findings of experimental allostim treatment for glioblastoma

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We report the skin findings from intradermal injections of an experimental therapy for glioblastoma performed in Thailand. The patient was a 42-year-old female who presented in 2009 with seizures. After CT and MRI imaging, a left parietal lobe biopsy revealed glioblastoma, WHO grade IV. The patient received several neurosurgical resections, radiation, and chemotherapy over multiple years. Her latest therapy involved an experimental trial run thru the Thai National Cancer Center. The protocol involved intradermal and intravenous delivery of AlloStim, an intentionally MHC-mismatched CD4+ T cell product. The T cells are activated with anti-CD3 and anti-CD28 antibodies attached to paramagnetic epoxy beads (Dynabeads). The treatment left multiple dark pigmented macules on the bilateral forearms at the sites of injection ranging in size from 3 to 5 mm. The patient's tumor proved to be resistant to all therapy its spread became far more extensive. The patient ultimately succumbed to pneumonia after progression of the tumor and marked clinical deterioration. At autopsy, no adverse side effects were attributed to the experimental therapy. Excisional skin biopsies of the dark pigmented macules on the forearms showed innumerable small round refractile brown micro-beads within the superficial dermis. There was a surrounding fibroblastic proliferation but a lack of extensive lympho-histiocytic reaction, including a lack of multinucleated giant cells. As the availability and popularity of experimental cancer therapies increases throughout the world, this case documents the possible skin findings associated with these treatments.

Poster 646**RESIDENT****Papular mucinosis in the setting of congenital HIV****Mariko Yasuda, MD**Mariko Yasuda, MD¹; Maryanne Makredes, MD¹; April Deng, MD¹; Leah Belazarian, MD¹¹University of Massachusetts, Worcester, MA, USA

The primary cutaneous mucinoses are defined by abnormal amounts of mucin in the skin as the major histologic feature within clinically relevant lesions. Lichen myxedematosus (LM) is a rare primary cutaneous mucinosis in which mucin accumulates in the upper and mid reticular dermis, with variable fibroblast proliferation and diminished or absent fibrosis. We present an 18-year-old female with perinatally acquired HIV and a CD4 of 557 on Highly Active Antiretroviral Therapy (HAART), who presented with multiple asymptomatic, slightly yellowish papules on her elbows for 1 month. Punch biopsy of a lesion on the elbow revealed a dome-shaped papule with an increase in dermal mucin. Her presentation was most consistent with the rare discrete papular type of LM typically reported in HIV-infected males with CD4 counts <100. This case illustrates the importance of recognizing cutaneous mucinoses, like the discrete papular variant, which may coexist with HIV.

Poster 647**FELLOW****Thyroid transcription factor 1 and napsin A expression in cutaneous bronchogenic cysts****Agne Naujokas, DO**Agne Naujokas, DO¹; Anatoly Urisman, MD, PhD¹; Kirk Jones, MD¹; Philip LeBoit, MD¹¹University of California - San Francisco, San Francisco, CA, USA

Cutaneous bronchogenic cyst represents an uncommon developmental cyst commonly found near the suprasternal notch in children or adults. First described by Seybolt and Clagett in 1945, bronchogenic cysts are thought to originate from the tracheobronchial bud of the primitive foregut. Such cysts are lined by a pseudostratified columnar epithelium and may contain other components such as mucous glands, smooth muscle, and/or cartilage, thus resembling the elements seen in the bronchial wall. Other cysts such as branchial cleft cyst, thyroglossal duct cyst, cutaneous ciliated cyst, or mature teratoma can share histopathologic features with bronchogenic cyst and may cause a diagnostic difficulty. Due to its pulmonary origin, we assessed immunohistochemical staining for thyroid transcription factor 1 (TTF-1) and napsin A to seek a marker that would help differentiate bronchogenic cyst from its mimics. A computerized search for the 10-year period 2002-2012 revealed three cutaneous bronchogenic cysts, all of which showed positive nuclear staining for TTF1 in the ciliated cells lining the cyst. Napsin A, a recently described marker that has been noted to be superior to TTF-1 in distinguishing lung primary adenocarcinoma, was negative in all three cases. In summary, bronchogenic cysts are developmental cysts that can share some histopathologic

features with other cutaneous cysts. Immunohistochemical staining for TTF-1, but not napsin A, may be of value in facilitating a specific diagnosis.

Poster 648**RESIDENT****Colloid milium of the face and oral cavity: a rare presentation****Ivanka Kovalyshyn, DO**Ivanka Kovalyshyn, DO¹; Golar Honari, MD¹; Melissa Piliang, MD¹¹Cleveland Clinic Foundation, Cleveland, OH, USA

We report a case of an otherwise healthy 60-year-old Caucasian female who presented with asymptomatic yellow papules involving face and oral cavity that had been present for 15 years. Physical examination revealed multiple yellow translucent and dome-shaped papules localized to central face and mucosal aspect of lower lip, anterior gingiva, the lateral borders of the tongue and the ventral surface of the tongue. Hematological, serum biochemistry, and urinalysis results were all within normal range. Punch biopsy of the papule from the lower lip mucosa revealed nodules of eosinophilic fissured material in papillary dermis, with fibroblasts aligned along the edges of the fissures. These histologic features were consistent with colloid milium. Colloid milium (CM) is a rare deposition disorder of unknown cause that presents with translucent yellow papules. There has been only one reported case of CM involving oral cavity. Proposed mechanisms of colloid deposition include degeneration of collagen or elastin, or a fibroblast bi-product. Usually there is no associated systemic abnormality. Histologically, CM is composed of homogenous, fissured, eosinophilic nodules, which are PAS-positive and diastase resistant. Differentiation from amyloid may be challenging as colloid material can stain positively with crystal violet and Congo red and give fluorescence with thioflavin T. In contrast to amyloid, colloid does not stain with pagoda red or other cotton dyes. In equivocal cases, electron microscopy may be used as amyloid shows non-branching straight filaments, in contrast to branching wavy filaments in CM. We present a unique case of CM involving face and oral cavity.

Poster 649**RESIDENT****Calciophylaxis occurring in a patient with rheumatoid arthritis****Julie Tse, MD**Julie Tse, MD¹; Rex Smith, MD¹; Mai Hoang, MD¹¹Massachusetts General Hospital, Boston, MA, USA

Calciophylaxis is a microvascular occlusion syndrome caused by mural calcification of small- and medium-sized vessels with subsequent thrombosis leading to cutaneous ulceration, especially in areas of high adiposity. Calciophylaxis can rarely occur in the absence of end-stage renal failure or hyperparathyroidism. We report the case of a 67-year-old obese woman with quiescent rheumatoid arthritis who presented with a one-month

history of a painful ulcer on her buttock. An initial punch biopsy showed only denuded epithelium with minimal inflammation of the subcutis and focal fat necrosis; she was presumed to have toxic epidermal necrolysis secondary to lamotrigine use. However, her lesions progressed despite discontinuation of lamotrigine, and subsequent biopsies failed to pinpoint the diagnosis. Her renal function and parathyroid hormone levels were within normal limits. She developed violaceous plaques with overlying bullae on her inferior breasts, abdomen, buttocks, inner thighs, and proximal calves, which eventually ulcerated. Skin biopsies finally revealed calciphylaxis, confirmed by Von Kossa stain. Attempts to treat the patient with sodium thiosulfate, alendronate, and debridements were unsuccessful, and the patient succumbed to sepsis four months following her initial presentation. An autopsy revealed intramural vascular and extravascular calcium deposition in the skin, lungs, and kidneys, which was highlighted by Von Kossa stain. Calciphylaxis has been most closely associated with end-stage renal failure and hyperparathyroidism but should also be considered in patients lacking these risk factors but presenting with the characteristic distribution of lesions.

Poster 650

RESIDENT

Nodular reaction to porcine collagen dermal filler

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A 63 year-old woman with history of basal cell carcinoma presented with multiple painless skin-colored 4 to 7 mm nodules above the vermilion border of the upper lip. She reported injection of Evolence (ribose cross-linked porcine collagen) at the site one year earlier. The clinical impression was of a foreign-body granuloma. A punch biopsy showed unremarkable epidermis with solar elastosis in the papillary dermis. The deep reticular dermis contained well-demarcated multinodular amorphous eosinophilic deposits extending to the subcutis. These deposits were loosely circumscribed by thin fibrous septae. Areas within the nodules had a finely fibrillar appearance. Polarized light microscopy did not reveal refractile foreign bodies. No inflammatory infiltrate was present, but occasional fibroblasts and capillaries were seen at the periphery of the nodules. Given the clinical history, these findings are consistent with nodular reaction to porcine collagen dermal filler. Ribose cross-linked porcine collagen is reported to have increased biocompatibility and decreased induration relative to hyaluronic acid or bovine collagen products. Nonetheless, rare accounts exist of non-inflammatory nodule formation at sites of porcine collagen injection. As in this case, the upper lip seems predisposed to this nodular reaction. Given porcine collagen's microscopic similarity to native dermal collagen, the subtle histologic findings of porcine collagen injection may be overlooked without adequate clinicopathologic correlation.

Our case is the second histopathologic description of Evolence, and the first to our knowledge since Evolence was removed from the U.S. market in 2009. Awareness of these findings is necessary given the continued use of similar products internationally.

Poster 651

RESIDENT

An open source digital pathology system supporting multi-touch interaction for dermatopathology teaching

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Background: Whole slide imaging has great potential for teaching but the large image sizes are challenging with respect to data handling and remote access. We are developing an open source client-server digital pathology system allowing rapid viewing of whole slide images (WSIs) over standard networks on desktop computers and multi-touch mobile devices. Data access is managed using social network based authentication (single-site login). Ultimately the system will have a distributed data architecture that adapts dynamically as new data server resources are added. Methods: Dermatopathology WSIs are uploaded to a server and processed into a multi-resolution format for remote viewing. Images are organized into 28 curriculum-based sessions, accessed using "Facebook" authentication, and reviewed at weekly attending-led sessions with teleconferencing to off site residents. Sessions involve sequential viewing of WSIs on a web-based client, with projection of the computer desktop on a large screen for group viewing. Histologic features, differential diagnoses, and diagnoses are discussed. An answer key is posted retrospectively on "Facebook" for consolidation. Results: Approximately 500 of 5,000 dermatopathology WSIs are uploaded for remote viewing and teaching. Viewing of WSIs using a wired network connection is fast and responsive, similar to the interaction with a glass slide, and resident feedback has been positive. A strong preference for multi-touch devices (iPad) has been expressed. Conclusions: There has been enthusiastic support for teaching dermatopathology using the digital pathology system over the past year. Both verbal and blinded written resident evaluations received for each session indicate a subjective increase in diagnostic confidence.

Poster 652

Whole slide imaging method for collaborative dermatopathology research

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Currently available digital scanners allow for whole slide images of microscopic slides to be obtained for histopathologic study. Using such a scanner, we have compiled an image data base of 265 slides representing 211 patients with deep penetrating nevi. The slides were scanned at a magnification of 40x. The image files are JP2(JPEG 2000) format with an average file size of 1.12 GB ranging between 0.1 GB and 1.95 GB. The total storage for the cohort is 297 GB. The digital slide image base allows for collaborative research with dermatopathologists regardless of geographical location. The images may be viewed by a secure internet link or shared by virtual private network or portable hard drive using image viewing software. This method allows for a number of advantages over glass slide studies including: the ability to remove patient identifying information, tumor morphometric analysis, annotation of images, high quality photomicrography, simultaneous comparison of microscopic fields from different slides and archiving for future restudy or teaching. Companion data collection programs may be customized to allow study participants to submit data electronically. The digitization of microscopic slides and data collection streamline the investigative process and facilitate the analysis of data produced.

Poster 653

RESIDENT

Autopsy findings after imatinib treatment in a patient with nephrogenic systemic fibrosis

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The patient is a 67-year-old man with coronary artery disease and chronic kidney disease from diabetes. He was diagnosed with nephrogenic systemic fibrosis (NSF) based upon his clinical presentation of progressive hardening and tethering of skin with joint contractures in the setting of renal failure, and the diagnosis was confirmed by a skin biopsy. He had been exposed to gadolinium on four occasions. He underwent physical therapy, phototherapy, and subsequently received oral imatinib. His skin lesions improved, but never resolved completely. He continued to have difficulty with movement. An autopsy was performed to document the extent of his disease. Besides skin involvement and renal findings of nephrocalcinosis and advanced diabetic changes, there was patchy fibrosis in the heart involving both ventricles and the interventricular

septum. The liver showed chronic passive congestion with bridging fibrosis, at least partially accounted for by right-sided heart failure. The limited systemic fibrosis observed in this patient may reflect the treatment effects of imatinib. Case reports of patients improving on imatinib have been published. Our report is the first autopsy case examining a patient with NSF that demonstrates limited systemic fibrosis after treatment with imatinib.

Poster 654

FELLOW

Cutaneous meningothelial rests of the scalp and lower back: a case report with immunohistochemical studies and review of the literature

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Meningothelial cells are components of the meninges that envelope the brain. The presence of these cells outside of the central nervous system is a rare phenomenon. Cutaneous meningothelial rests are hamartomas that contain ectopic meningothelial cells located in the skin. These lesions are most commonly located on the scalp of children or young adults, often resembling a nevus. We report two cases of cutaneous meningothelial hamartomas, one located on the scalp of a 25 year old male and the other on the lower back of a 3-month-old male. Histologic presentation in both cases was consistent with a hamartomatous malformation composed of vascular, adipose and smooth muscle elements admixed with meningothelial nests. The meningothelial nests contained pale staining cells with a pink frothy cytoplasm and pinpoint nuclei reminiscent of neuropil. Signs of malignant transformation, such as nuclear atypia or increased mitotic activity, were not observed. Immunohistochemical staining with glial fibrillary acid protein (GFAP) highlighted the neural origin of the meningothelial nests. CD31 staining was present in the surrounding vascular proliferation, while epithelial membrane antigen (EMA) stained the glandular structures, confirming the hamartomatous nature of this lesion. The histologic and immunohistochemical findings are consistent with the diagnosis of a meningothelial hamartoma. The histologic identification of a cutaneous meningothelial rest is a dermatopathologic challenge that may require immunohistochemical confirmation. We also performed a review of the literature on the histologic and clinical findings of cutaneous meningothelial hamartomas.