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Pili multigemini of the eye: unusual and rare presentation

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Cutaneous malignant melanoma is rare in children and adolescents and accounts for only 1.3% of cases in the United States. Merely 0.3-0.4% occur during the first decade of life. Congenital and infantile melanoma most often occurs in the head and neck. Superficial spreading and nodular melanoma are the most common histologic subtypes. Acral melanomas have been described, but are exceedingly rare. We report a case of acral melanoma in a 15-month-old child. Physical examination showed a large black papule on the plantar foot with an adjacent satellite lesion. Initial biopsy was concerning and complete excision revealed an atypical compound melanocytic proliferation with pagetoid migration and sheets of atypical melanocytes forming nodules within the dermis and extension into the subcutaneous fat. The melanocytes showed diffuse positivity for HMB-45 and significant loss of p16 expression. This diagnosis was rendered with hesitance and difficulty, as melanoma is rare in this age group and location. Therefore, a high level of suspicion and awareness of the pathologic criteria used to differentiate benign and malignant melanocytic lesions is necessary to avoid delayed diagnosis and treatment.

Avoid Making A Diagnosis “Off the Cuff”

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A 17-year-old male presented with a 2 cm forehead mass present for 2 years. Clinically, the mass was favored to be a lipoma. On excision, the lesion consisted of well circumscribed histiocytoid cells with mild pleomorphism and a prominent lymphoplasmacytic cuff.

The community pathologist performed microorganism special stains, which were negative. At this point the lesion was assumed to be a hematopoietic neoplasm, prompting a hematopathology and hematology-oncology consultation. Tumor cells were positive for CD163 and Desmin and negative for Lu-5 and SMA. An EWSR1-CREB1 fusion was identified and a diagnosis of angiomatoid fibrous histiocytoma (AFH) was rendered. AFH usually arises in the extremities of young adults and teenagers. In this case, the characteristic dense lymphoplasmacytic cuff surrounding the tumor was initially confused for a hematopoietic neoplasm or infectious process. Community pathologists should be aware of this entity and the fact that it can present on the scalp and forehead to properly guide immunohistochemical staining and correct clinical follow up. These lesions can recur and rarely metastasize, so appropriate follow up is necessary.

Pili Multigemini of the Eye: Unusual and Rare Presentation

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Pili Multigemini (PM) is an uncommon, pilar dysplasia characterized by clusters of hair shafts emerging from a single follicle. We present a rare and unique case of PM involving the eye. A 43 year old Caucasian male, presented with a prolonged history of a recurrent lesion at the right upper eyelid, present for two years. Clinical examination revealed a subcutaneous cyst with a prominent follicular pore with a differential diagnosis of epidermoid cyst, nevus and pili bifurcati. Excisional biopsy was performed and microscopy showed a benign malformed hair follicle containing multiple different hair shafts enclosed in a common outer root sheath; consistent with the diagnosis of Pili Multigemini. Pili Multigemini was first described by Flemming in 1883 and is frequently found in the beard of adults and scalp of children. To our knowledge, this is the first case of Pili Multigemini involving the eye. It is noteworthy to consider during diagnostic workup; as well as to exclude other follicular and inflammatory abnormalities involving the eye.

A Case Report of Eccrine Syringofibroadenoma

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Eccrine syringofibroadenoma is a rare benign adnexal tumor with eccrine sweat duct proliferation. Less than 80 cases have been reported, either associated with reactive, neoplastic or syndromic settings. Due to its rarity and variable clinical presentation, it is easily misdiagnosed without pathology exam. Here we report a case of eccrine syringofibroadenoma. A 48-year-old African-American male with personal history of diabetes, hypertension and diabetic glomerulosclerosis and remote history of local treated cutaneous wart at right heel presented to dermatology clinic for the recurrent slow growing, enlarged wart. The cutaneous excisional biopsy shows peculiar histological features including thin anastomosing strands arising from the epidermis, bland cytomorphology, fibrovascular stroma, and lace-like architecture. Although architectural features mimic a relative common cutaneous lesion, fibroepithelioma of Pinkus, a variant of basal cell carcinoma, focal eccrine ductal differentiation and eccrine immune profile supports the diagnosis of syringofibroadenoma.