8-1-2021

Rare lymphatic malformation of external ear canal with history of sclerotherapy

Shannon Rodgers
Henry Ford Health, SRodger5@hfhs.org

Jason Pimentel
Henry Ford Health, JPIMENT1@hfhs.org

Recommended Citation

Rodgers S, and Pimentel J. Rare lymphatic malformation of external ear canal with history of sclerotherapy. American Journal of Dermatopathology 2021; 43(SUPPL 8):S12.

cystic structures. Histopathologic analysis demonstrated a proliferation of histiocytic cells with mitotic figures, zonal necrosis, eosinophils, and focal osteoclast-like giant cells. Immunohistochemistry staining was positive for Vimentin, S100, CD1a, CD68 and CD 207. Laboratory testing (LDH, ESR, TSH/T4, and urinalysis) was essentially normal. Complete blood count with differential was normal except for mild thrombocytosis. At three-week follow-up, the patient noted sudden painful lymphadenopathy. Imaging studies revealed tumor extension from the scalp into the inner and outer tables of the calvarium with extradural invasion. Prominent lymphadenopathy was seen within the supraclavicular and cervical chains. Palatine tonsillar enlargement was also noted. Skeletal survey and chest X-ray did not show bony involvement. Based on the clinical, histological and imaging findings, the patient was diagnosed with multifocal Langerhans Cell Histiocytosis - eosinophilic granuloma with intracranial extension and massive lymphadenopathy. After evaluation with oncology, the patient and family opted for clinical monitoring since the scalp lesion and lymphadenopathy were now asymptomatic and minimized in size.

Rare Lymphatic Malformation of External Ear Canal With History of Sclerotherapy

Shannon Rodgers and Jason Pimentel
Department of Pathology, Henry Ford Health System, Detroit, MI.

An 8-year-old boy with history of conductive hearing loss, presented with a 3.1 x 3.1 x 2 cm left postauricular mass diagnosed as lymphatic malformation on imaging. Treatment with bleomycin sclerotherapy led to successful improvement of the malformation. Seven months post-treatment, he presented to his audiologist with several months of intermittent bleeding and pain from the ear. Upon examination by otolaryngology, a 0.5 cm papillary lesion was discovered in the external ear canal which had not been identified previously. A biopsy demonstrated a papillary proliferation of dilated thin walled lymphatic channels expanding multiple dermal papillae, consistent with lymphatic malformation. We propose that this lesion could represent extension of the original lesion or a new lesion secondary to regional lymphatic damage from prior sclerotherapy. Lymphatic malformation is rare within the external ear canal. Moreover, this case developed after treatment by regional sclerotherapy. To our knowledge, this sequela has not been previously documented in the literature but may be underreported. Therefore, routine regional follow up may be warranted in any periauricular sclerotherapy for lymphatic malformation.

Extracutaneous Pyoderma Gangrenosum in an African American Woman

Gabriella Santa Lucia, Alexia Demaio, and Samantha Karlin
Department of Dermatology and Dermatologic Surgery, Medical University of South Carolina, Charleston, SC.

Rare extracutaneous manifestations of pyoderma gangrenosum (PG) are potentially fatal and difficult to diagnose. Successful diagnosis is based on integrating a combination of radiologic, histologic, and clinical findings. A 52-year-old female with a history of cutaneous PG and unidentified lung nodules was admitted to the hospital for severe tibial ulcerations. Repeat chest CT revealed new necrotic nodules and multiseptated consolations. Plan was to initiate Humira for cutaneous PG lesions, however concern for pulmonary infection warranted lung biopsy prior to starting immunosuppression. Histopathology of the left lower lobe biopsy showed dense neutrophilic inflammation with abscess formation and without granulomas. Atypical infection such as nocardia was suggested, ruling out vasculitis and rheumatoid nodules. The patient continued to deteriorate after linezolid and imipenem administration. Immunosuppressive therapy was initiated after lung histopathology matched that of cutaneous PG lesions, suggesting the nodules were an extracutaneous PG manifestation. Both cutaneous and pulmonary lesions improved on immunosuppression. Extracutaneous PG is a diagnosis of exclusion as it mimics other infectious pathologies. Initial reads of radiologic and histologic studies may be misinterpreted as infectious etiologies without careful clinical correlation. Delay in correct diagnosis can lead to significantly worse patient outcomes and extraneous medical workup.

A Novel Histologic Finding in a Rare Disease: Scarring Alopecia in Hereditary Mucoepithelial Dysplasia

Kerrie Satcher, MD, Jennifer Schoch, MD, and Addie Walker, MD
University of Florida, Department of Dermatology, Gainesville, FL.

Hereditary mucoepithelial dysplasia (HMD) is a rare, autosomal dominant disorder with characteristic cutaneous findings, non-scarring alopecia, ophthalmologic, and pulmonary disease. The culprit gene is unknown; diagnosis relies on clinical findings and family history. We describe a 13-year-old boy with an extensive family history of HMD, who presented with red scrotal and intertriginous plaques, diffuse xerosis, and alopecia with scale, consistent with a diagnosis of HMD. Scalp biopsy showed psoriasiform acanthosis with columns of parakeratosis overlying invaginations of superficial epidermis with dyskeratosis- features previously described in cutaneous HMD. Our patient's biopsy also demonstrated findings of scarring alopecia including dermal fibrosis, diminished sebaceous glands, follicular rupture, concentric perifollicular fibroplasia and mixed inflammation—features not previously described. Our case adds to the limited HMD literature by describing histologic features of scarring alopecia in a condition previously viewed as non-scarring. Recognition of these histologic findings may be a key step in diagnosing HMD, a rare disorder with important implications for visual and respiratory health.

Keloidal Plaques in A Patient With Diffuse Cutaneous Systemic Sclerosis

Jane Scribner, MD, Laura Winterfield, MD, MPH, and Jessica Forcucci, MD
Medical University of South Carolina, Charleston, SC.

A woman in her 50s with a history of diffuse cutaneous systemic sclerosis, previously managed by rheumatology with mycophenolate and IVIG, presented with a new onset of painful plaques on her chest, breasts and axillae. They progressed over 12 months and were not responsive to topical clobetasol. Biopsy demonstrated a proliferation of fibroblasts with hyalinized thick keloidal collagen bundles in a haphazard distribution extending from the mid-reticular dermis to the dermal-subcutaneous junction with sparing of the papillary dermis. A significant inflammatory infiltrate was not identified. Keloidal morphea, also known as nodular scleroderma, is considered one of rarest clinical forms of scleroderma. Keloidal morphea commonly presents as asymptomatic plaques favoring the chest, back, and proximal extremities, and arise in sclerodermatous areas. The histopathologic variants include sclerodermatous, keloidal, and mixed patterns. The etiology of keloidal morphea remains unclear, but may involve a genetic predisposition and keloidal response to inflammation of the skin in patients with scleroderma. The authors have no relevant relationships to disclose.

Unusual Case of Primary Cutaneous Adnexal Carcinoma With Negative P63 and P40

Shiyanbola O.O.,* and Rosa G.†
*Departments of *Pathology, University of Wisconsin, Madison, and †Associated Pathologists/St Mary's Hospital, Madison, WI.*

We present a case of primary cutaneous adnexal carcinoma in the left groin of a 75-year-old woman with no history of malignancy, demonstrating a cribriform/tubular morphology, with ER, PR and GATA-3 expression but negative for p40 and p63. An initial biopsy was diagnosed as adenocarcinoma, favor metastatic breast carcinoma. Repeat p40 and p63 stains on the excision were negative. Background mammary-like glands were not identified. CT of the chest, abdomen and pelvis, mammogram and MRI of the breast were negative. A PET scan was negative, with the exception of the