Multinucleate Cell Angiohistiocytoma.

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Multinucleate Cell Angiohistiocytoma

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Case Presentation

Patient: 74 year-old Caucasian female.

History of Present Illness: The patient presents with a lesion on the right thigh for eight years duration. She describes the lesion as bothersome and spreading. Denies itching, burning or pain. The lesion was previously biopsied and diagnosed as Lyme disease and bacterial infection. Patient tried topical corticosteroids without improvement.

Medical History: Hypertension, seasonal allergies, colon cancer, ovarian cancer

Family History: Hypertension, cardiac disease, breast cancer

Social History: Patient is single, retired, denies alcohol, denies tobacco use

Medications: Amlodipine, vitamin D3, vitamin B

Previous Treatment: Topical corticosteroids

Physical Examination: Well-circumscribed red to tan papules and plaques with central atrophy. Lesions are strikingly linear on the left medial thigh and one lesion on the right.

Biopsy: *Advanced Dermatology Associates, LTD* (AD16-12471, 10/21/2016) Left medial thigh, mid-medial and inferior: "Proliferation of poorly grouped capillaries and small venules in the upper to mid dermis with unique multinucleate giant cells against a fibrotic stroma. A predominantly lymphoid perivascular infiltrate is also noted with few plasma cells, neutrophils, and eosinophils. Special stains (AFB-Fite, Gram, GMS) are negative for infectious organisms. Vascular endothelial cells are positive for antibodies to vimentin and CD34. Mononuclear cells express vimentin, Factor XIIIa, and CD68. Multinucleate cells are strongly positive for vimentin, but variable for the expression of CD68."

Reason for Presentation: Interest

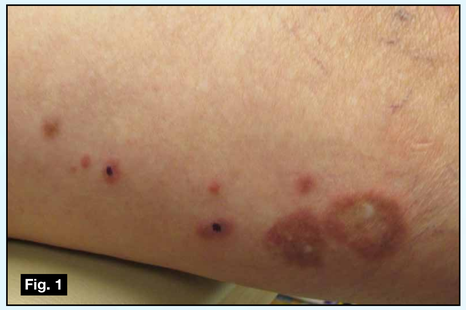

Figure 1: Multiple, red-brown plaques with central atrophy in a linear distribution located on the right lateral thigh. (Purple dots indicate biopsy locations).

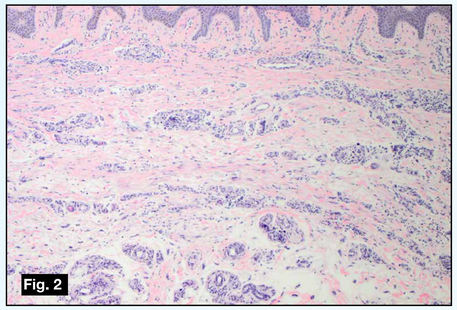

Figure 2: (H&E, 10x) Proliferation of poorly grouped capillaries and small venules within a collagenous fibrotic stroma in the superficial and mid dermis.

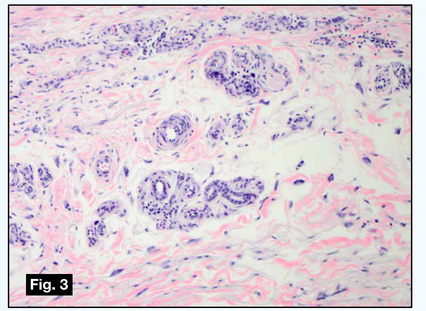

Figure 3: (H&E, 20x) Groups of capillaries and venules with scattered multinucleate giant cells in the superficial and mid dermis.

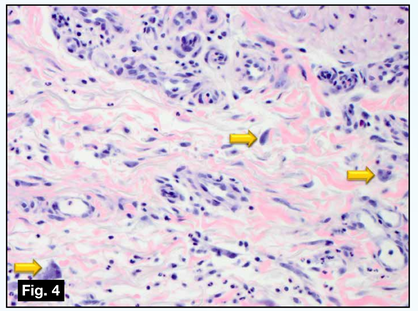

Figure 4: (H&E, 40x) High power view demonstrating the unique multi-nucleate cells (yellow arrows) scattered through the proliferation of vessels.

Discussion

Multinucleate cell angiohistiocytoma (MCAH) is an uncommon vascular fibrohistiocytic proliferation that often presents clinically as grouped red-purple papules and nodules. Lesions commonly arise on the dorsal hands and lower extremities. Localized, multifocal, and generalized variants have been reported. The localized variant is the best described and most common.

Multiple hypotheses regarding etiology and pathogenesis have been proposed. The favored concept is that these lesions arise as a reactive rather than a neoplastic process. A few reports of spontaneous remission support the reactive hypothesis. Another theory proposes that MCAH arises secondary to female hormone influence due to the identification of estrogen receptor alpha expression on the interstitial and multinucleate cells of the lesions. This theory also serves to explain why MCAH occurs more frequently in women with a ratio of 3:1 female to male. However, identification of estrogen receptor positivity has not been consistent among reported cases.

Diagnosis of multinucleate cell angiohistiocytoma requires a clinicopathological correlation. Clinically, the differential diagnosis of the violaceous papules and nodules includes granulomatous entities including sarcoidosis and granuloma annulare or vascular proliferations such as Kaposi's sarcoma and acroangiodermatitis. Histopathological analysis of MCAH does not show granulomatous inflammation expected in the first two entities nor large vessels with a dense capillary proliferation and hemosiderin deposition seen in acroangiodermatitis. Instead, MCAH pathology reveals an upper-to-mid-dermal proliferation of narrow vessels within thickened collagen bundles, as well as, large angulated multinucleate giant cells with palisading nuclei and eosinophilic cytoplasm. Expression of CD68 can be variably positive or sometimes negative. Immunohistochemical staining can aid in diagnosis showing vimentin positivity and S100 negativity of the infiltrates with variable positivity of Factor XIIIa. Lesional analysis of Kaposi's sarcoma should not demonstrate multinucleate giant cells and immunohistochemistry would reveal HHV-8 positivity.

The course of multinucleate angiohistiocytoma is considered progressive but benign. Treatment is not required, but may be desired by patients secondary to cosmetic concern. Lesions may be treated topically or intralesionally with corticosteroids prior to initiation of more aggressive treatment. Reports of treatment by way of surgical excision, cryotherapy, pulsed dye laser, argon laser, intense pulsed light, and carbon dioxide laser have all shown efficacy anecdotally. The rationale behind the use of IPL and PDL is based on their oxy-hemoglobin targets and the vascular nature of these lesions.

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