Generalized Multinucleate Cell Angiohistiocytoma, An Exceedingly Rare Entity

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BACKGROUND

Multinucleate Cell Angiohistiocytoma (MCAH) is a rare fibrohistiocytic and vascular proliferation. To date, near to 140 cases of generalized and non-generalized MCAH have been described in the literature among which only less than 20 cases are generalized form.
The pathogenesis of MCAH is not well understood. Several mechanisms of pathogenesis, including hormonal, have been postulated. MCAH follows a benign clinical course, however, may persist for years. Spontaneous regression has been reported. Treatment may be sought for cosmetic reasons. Several successful treatment modalities have been described including topical or intralesional corticosteroids, surgical excision, and other non-surgical modalities such as laser therapy.

CASE PRESENTATION

A 67-year-old female presented with multiple asymptomatic red to violaceous papules on the left temple, neck, and upper chest. H&E sections from the lesions demonstrated a proliferation of small-sized vessels predominantly in the superficial dermis accompanied with sparse multinucleated fibrohistiocytic cells, lymphocytes and occasional neutrophils, and fibrosis. Colloidal iron stain failed to demonstrate mucin deposition and PAS stain was negative for fungal microorganisms.

DISCUSSION

MCAH is an uncommon vascular and fibrohistiocytic disorder with unknown pathogenesis. The generalized variant is exceptionally rare. Compared to the localized variant, generalized MCAH may affect both genders equally, may present with earlier onset of the disease, and mainly affects the trunk and extremities. The most characteristic histopathologic features are present in the superficial dermis and consist of proliferation of capillary-sized vessels, some with thickened walls and the presence of multinucleated cells. Dermal fibrosis is marked by compact haphazardly arranged collagen bundles. A sparse lymphocytic infiltrate, histiocytes, neutrophils, and plasma cells may also be seen. In our case, a wide differential diagnosis and clinical correlation was considered.

Given the presence of dermal fibrosis and increased number of fibroblasts in an interstitial pattern, scleromyxedema (papular mucinosis) was considered in the differential diagnosis, however, other features including presence of multinucleated cells argued against that diagnosis.

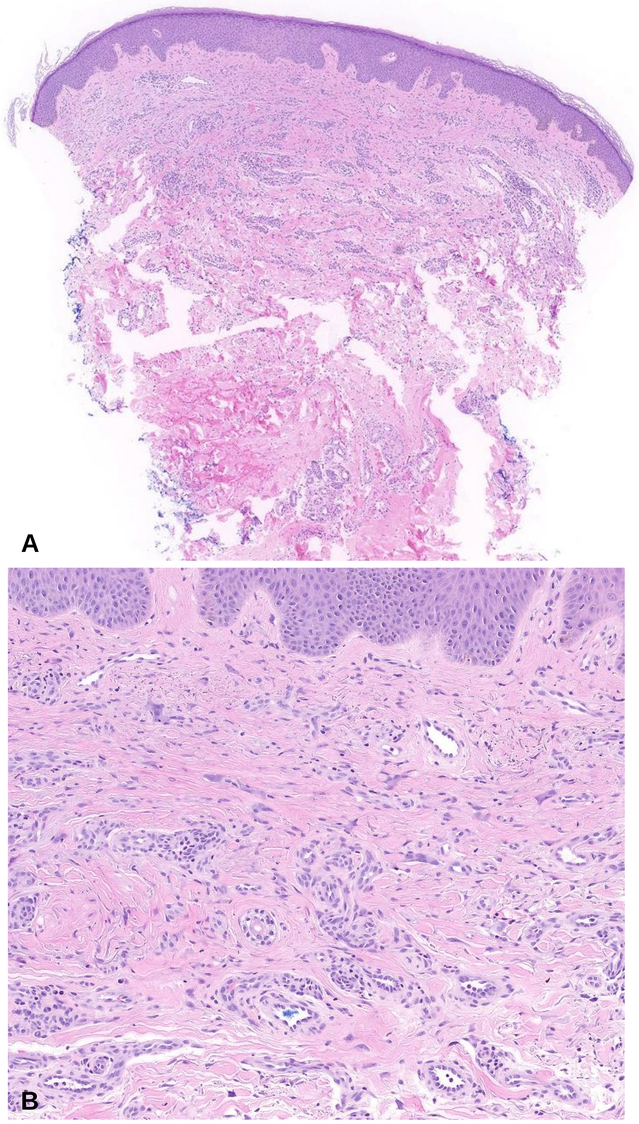

Figure 1: **A**. Significant increased number of vessels and dermal fibrosis are appreciated in this punch biopsy at scanning magnification (H&E stain, X 40). **B**. Dermal fibrosis, a sparse infiltrate of lymphocytes, stellate-shaped fibrocytes, some of which are multinucleated are evident in this higher power view and intermingled with many venules (H&E stain X400).

DISCUSSION (continue.)

No mucin deposition was highlighted with Colloidal Iron stain. Microvenular hemangioma is another consideration given the presence of many capillary-type vessels in the dermis, but additional element of fibrohistiocytic cells help to differentiate these two entities.
Kaposi sarcoma presents as painless purplish papules and may resemble MCAH clinically. However, the presence of multinucleated cells are a clue for MCAH and immunoproxidase stain for HHV8 is negative in all cases.

CONCLUSIONS

Generalized form of MCAH is an exceedingly rare entity and mainly affects face, upper trunk and extremities. The most characteristic histopathologic features include a proliferation of small-sized capillary-type vessels in the superficial and mid dermis intermingled with sparse multinucleated fibrohistiocytic cells and lymphocytes, and dermal fibrosis with thickened collagen bundles. Dermatologists and pathologists should be aware of the generalized form of this entity and consider it in the differential diagnosis of maculopapular rashes that mainly involve upper chest and extremities. The clinicopathologic correlation and recognition of this variant remains essential to differentiate it from other mimickers.

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