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CED
Clinical and Experimental Dermatology
Clinicopathological Case

An unusual case of dorsal hand papules and nodules

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Clinical findings

A 56-year-old woman presented with a 4-year history of evolving lesions over the dorsal hands (Figure 1a–c). She had a background history of hypertension, for which she was prescribed amlodipine and losartan. She initially presented in 2020 with a single slow-growing asymptomatic nodule over the dorsum of the left hand, prompting urgent referral under the 2-week-wait pathway. Clinically, this was a hyperpigmented firm nodule that was suspected to be a dermatofibroma. However, over the ensuing 2 years, multiple small dermal hyperpigmented papules and nodules continued to develop over the dorsal aspects of the hands only (Figure 1a–c).

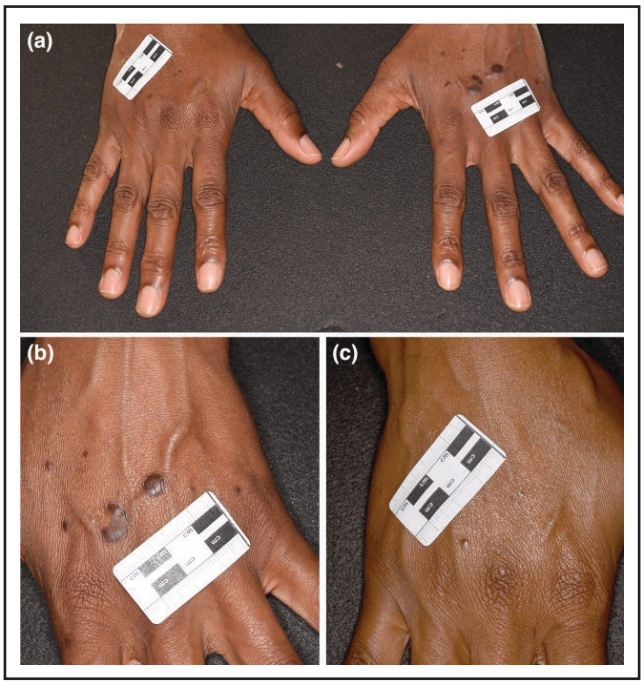

Figure 1 These images were taken during the patient's second presentation in June 2022, 2 years after her initial presentation with a single lesion. (a) This image shows multiple small, hyperpigmented papules, ranging in size between 2 and 8 mm, all located on the dorsal surface of the patient's hands. (b) Image detail of the lesions on the left hand. (c) Image detail of the lesions on the right hand.

Histopathological findings

Two diagnostic skin biopsies were performed (Figures 2 and 3). These revealed a dermal proliferation of small calibre vascular channels (Figure 2) and a notable presence of multinucleated cells with angulated cytoplasm (Figure 3).

What is your diagnosis?

Multinucleate angiohistiocytoma.

Discussion

Multinucleate angiohistiocytoma is an idiopathic, benign vascular proliferation that was first described in 1985.[1] They are considered rare, with approximately 150 reported cases.[1,2] However, the condition may be significantly underrecognized given the subtle histological features. It predominantly affects women between the ages of 45 and 65 years.[2] The pathogenesis of the condition is unknown. Given the chronic inflammatory changes seen on histology, this may be a reactive phenomenon.[3]

The dermal clustered papules or nodules are asymptomatic. They may become confluent and are commonly located over the dorsum of the hands, face and, less frequently, on the lower legs.[2] Although localized, multifocal and generalized variants have been identified, the localized form is the most prevalent.[1] Often, erythematous-to-violaceous

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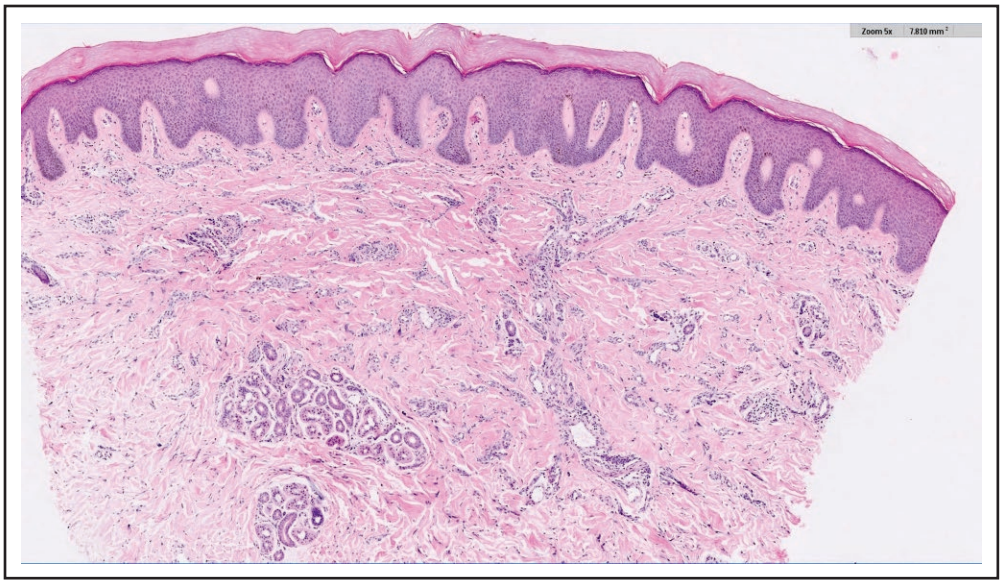

Figure 2 Image on low power from the dorsal aspect of the left hand (haematoxylin and eosin, ×5). The dermal collagen appears sclerotic or hyalinized, with an increase in small calibre vascular channels.

papules and plaques are described in this condition, ranging from 2 mm to 15 mm in size.[1] This patient's case highlights the appearance of multinucleate angiohistiocytoma in someone with Fitzpatrick type VI skin. Differential diagnoses include dermatofibroma, Kaposi sarcoma and lichen planus.

Histopathology typically shows a combination of increased dermal vascularity in the presence of angulated multinucleate cells, with the latter required to make the diagnosis. Multinucleate angiohistiocytoma is characterized by an increased number of factor XIIIa-positive fibrohistiocytic interstitial cells and multinucleated cells with angular contours located in the dermis.[4] Although not pathognomonic of multinucleate angiohistiocytoma, the presence of multinucleated giant cells is the most specific histopathological finding (3–10 hyperchromatic nuclei and basophilic cytoplasm).[4] The multinucleated cells are stained by vimentin and, alternatively, by CD68.[4] Mononuclear dendritic cells are positive for vimentin, factor XIIIa, MAC387 and lysozyme. Endothelial cells, in turn, are positive for vimentin, CD31, CD34 and factor VIII.[4]

Treatment options are limited, and trials of topical/intralesional corticosteroids, surgical excision and laser therapy have been reported with limited success.[2]

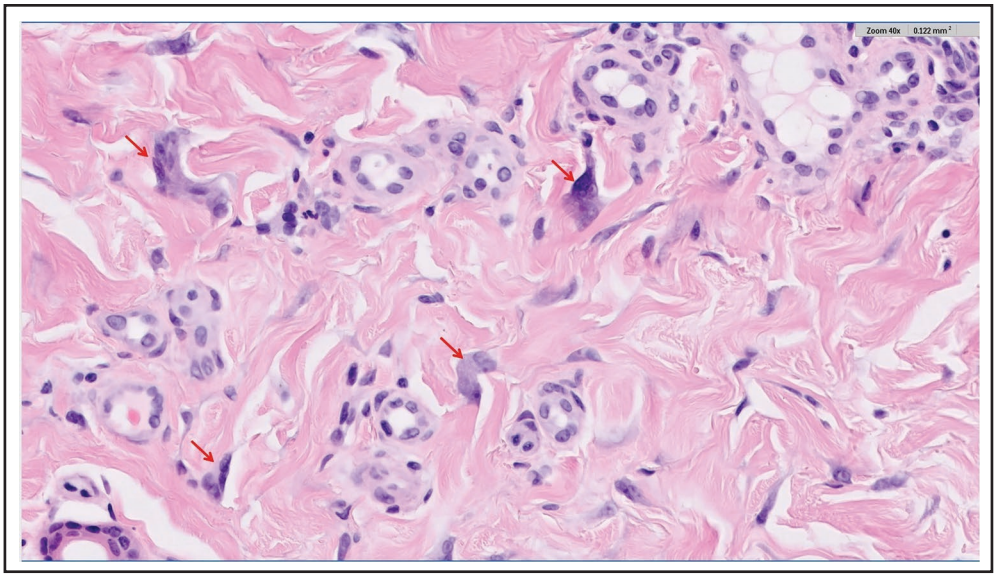

Figure 3 Image on high power (haematoxylin and eosin, ×20). The red arrows indicate multinucleated cells with angulated cytoplasm.

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Conflicts of interest

The authors declare no conflicts of interest.

Data availability

The data underlying this article are available in the article and in its online supplementary material.

Ethics statement

Not applicable.

Patient consent

Written patient consent for publication was obtained.

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CPD questions

Learning objective

To understand the diagnostic features of multinucleate angiohistiocytoma.

Question 1

A patient presents with 10 asymptomatic pigmented smooth papules that have been gradually increasing in size over the past 3 years. What distribution would be most typical of multinucleate angiohistiocytomas?

(a) Dorsal and palmar aspects of the hands.
(b) Dorsal aspects of the hands and anterior thigh region.
(c) Dorsal aspects of the hands and face.
(d) Dorsal aspects of the hands and feet.
(e) Dorsal aspects of the hands and shoulders.

Question 2

Which best describes the histopathology that would point to a multinucleate angiohistiocytoma over a dermatofibroma?

(a) Increased vascularity and the presence of multinucleated cells with angulated cytoplasm.
(b) Increased vascularity and the presence of multinucleated cells with rounded cytoplasm.
(c) Proliferation of fibrohistiocytic cells and the presence of multinucleated cells with rounded cytoplasm.
(d) Storiform spindled cells and the presence of multinucleated cells with angulated cytoplasm.
(e) Storiform spindled cells and the presence of multinucleated cells with rounded cytoplasm.

Instructions for answering questions

This learning activity is freely available online at https://oupce.rievent.com/a/JBGWKP

Users are encouraged to

- Read the article in print or online, paying particular attention to the learning points and any author conflict of interest disclosures.
- Reflect on the article.
- Register or login online at https://oupce.rievent.com/a/JBGWKP and answer the CPD questions.
- Complete the required evaluation component of the activity.

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