An unusual case of dorsal hand papules and nodules

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Funding sources: This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Conflicts of interest: None to declare.

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.

Clinical findings

A 56- year-old female 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 two-week wait pathway. Clinically, this was a hyperpigmented firm nodule that was suspected to be a dermatofibroma. However, over the ensuing two years, multiple small dermal hyperpigmented papules and nodules continued to develop over the dorsal aspects of the hands only (Figure 1a-c).

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 which was first described in 1985[2]. They are considered rare, with approximately 150 reported cases [1,2]. However, the condition may be significantly under recognised given the subtle histological features. It predominantly affects middle-aged to elderly females[1]. 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[1]. Whilst localised, multifocal, and generalised variants have been identified, the localised form is the most prevalent[2]. Often, erythematous to violaceous papules and plaques are described in this condition, ranging from 2-15mm in size[2]. This case uniquely highlights the appearance of multinucleate angiohistiocytoma in Fitzpatrick Type VI patients. 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[1].

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Figure legends

Figure 1 Multiple small hyperpigmented papules measuring between 2-8 mm at second presentation (b) Image detail of the lesions over the left hand shown in (a) and first lesions to develop (c) Details of the lesions on the right hand seen in (a)

Figure 2 Image on low power from the dorsal aspect of the left hand. The dermal collagen appears sclerotic or hyalinised, with an increase in small calibre vascular channels.

Figure 3 Image on high power. The red arrows indicate multinucleated cells with angulated cytoplasm.

CPD Questions

Learning objective: To understand the diagnostic features of multinucleate angiohistiocytoma

Question 1. A patient presented with 10 asymptomatic pigmented, smooth papules which 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 with rounded cytoplasm.
(d) Storiform spindled cells and the presence of multinucleated with angulated cytoplasm.
(e) Storiform spindled cells and the presence of multinucleated cells with rounded cytoplasm.

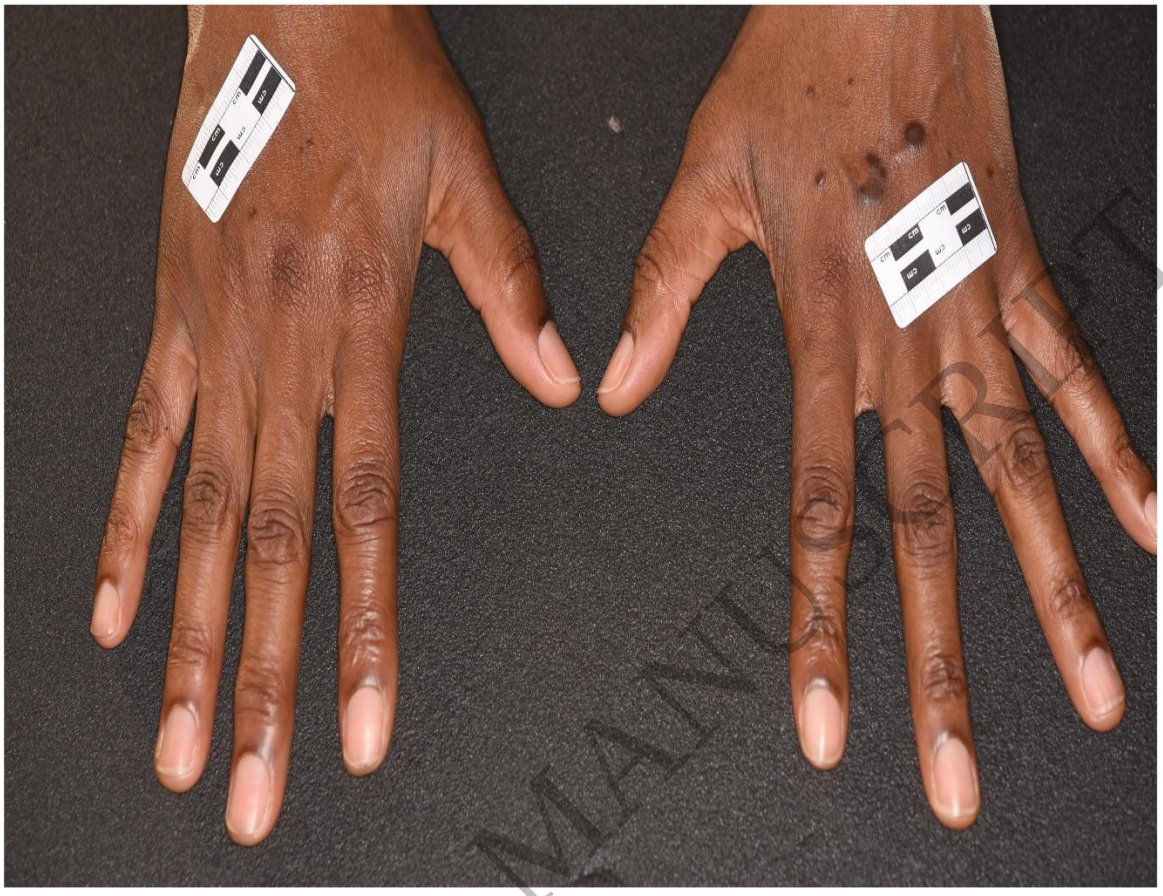

Figure 1A
312x160 mm (x DPI)

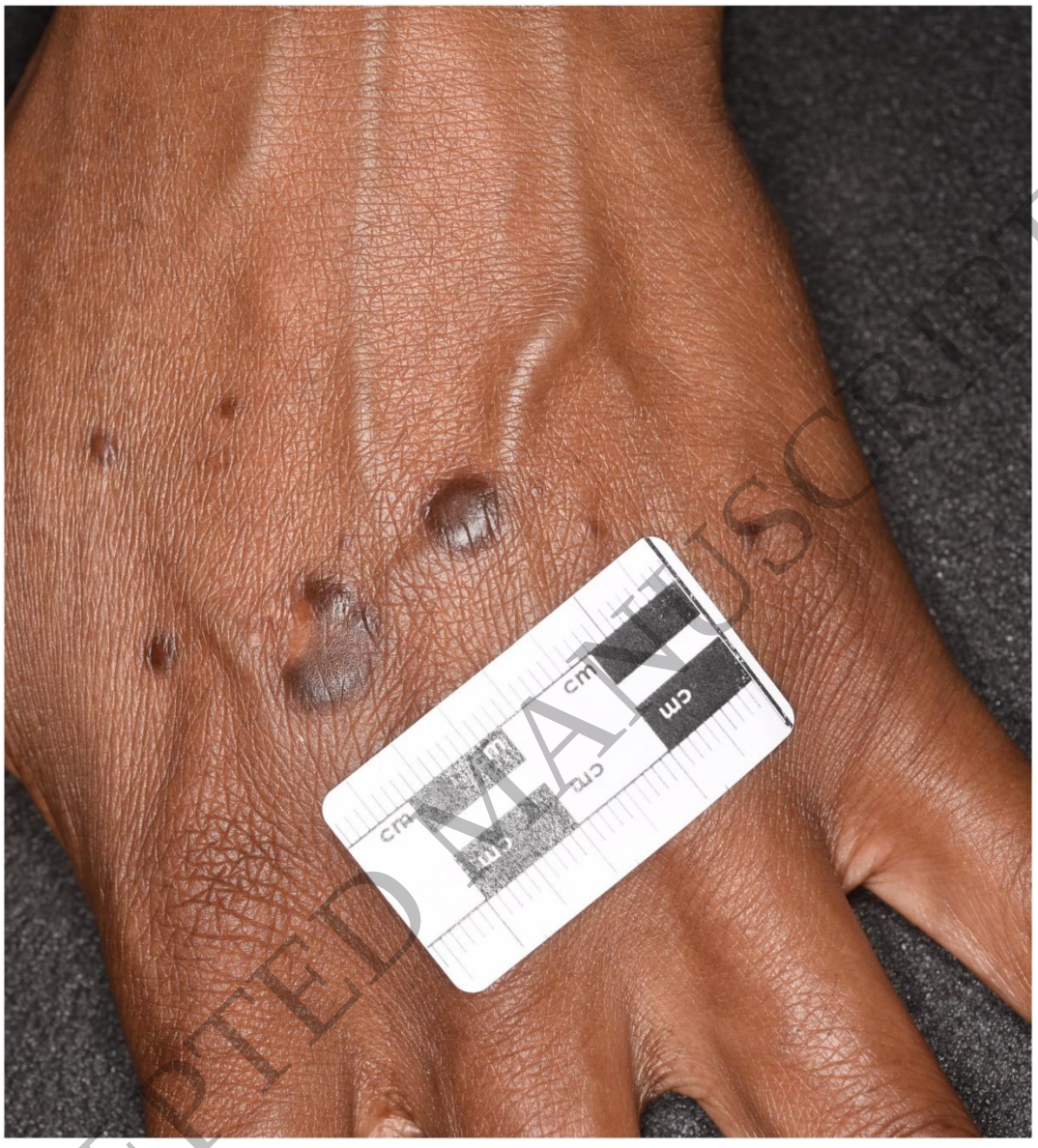

Figure 1B
141x155 mm (x DPI)

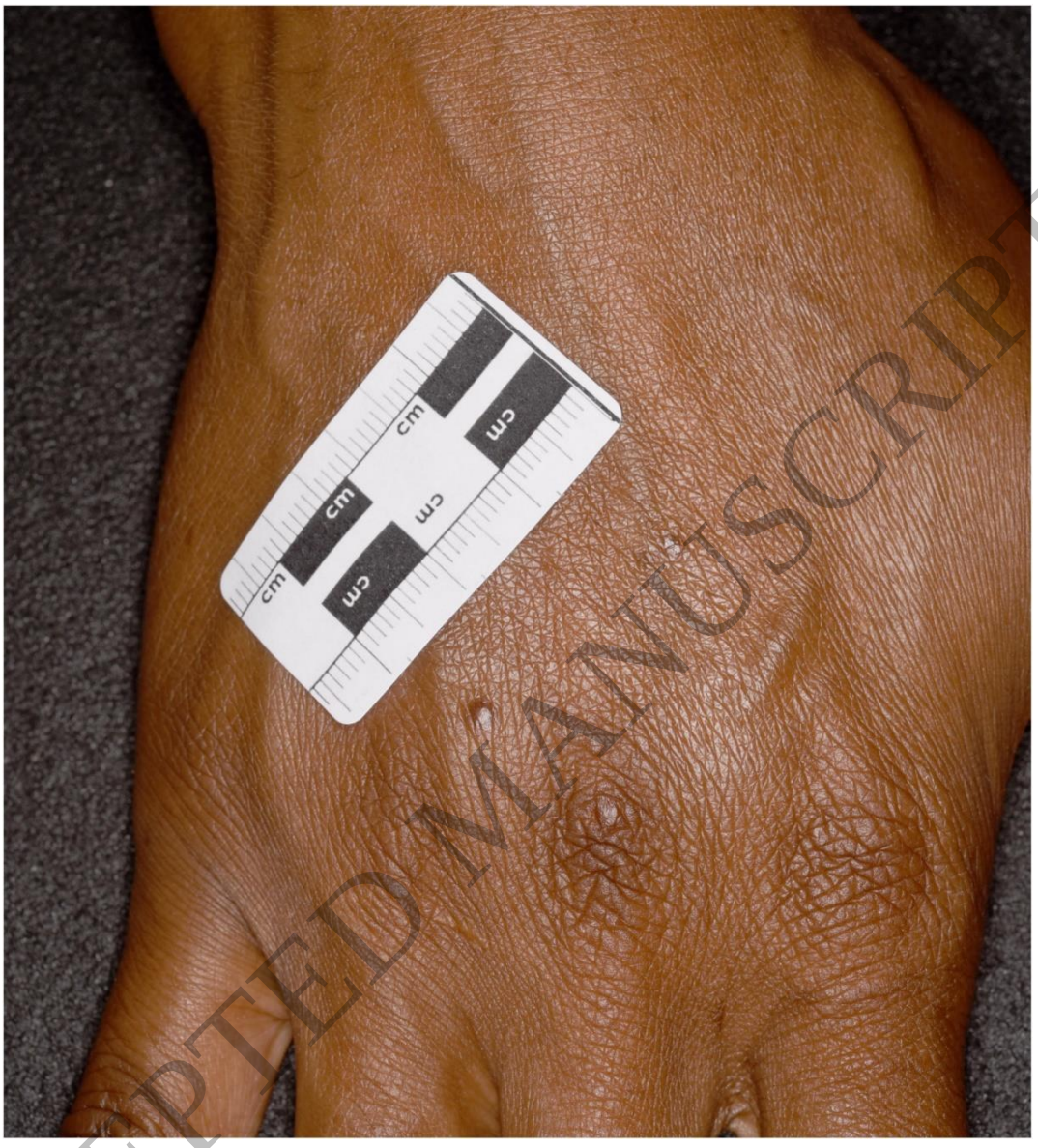

Figure 1C
141x155 mm (x DPI)

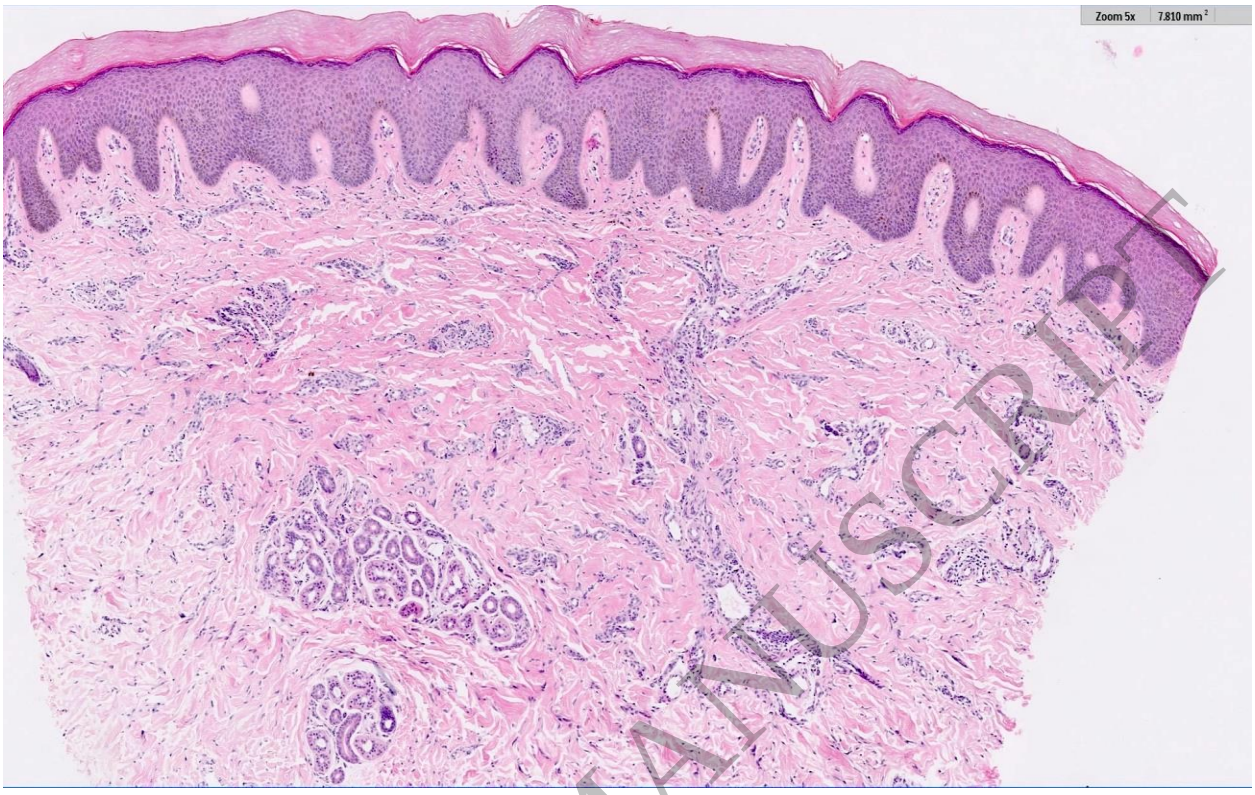

Figure 2
520x293 mm (x DPI)

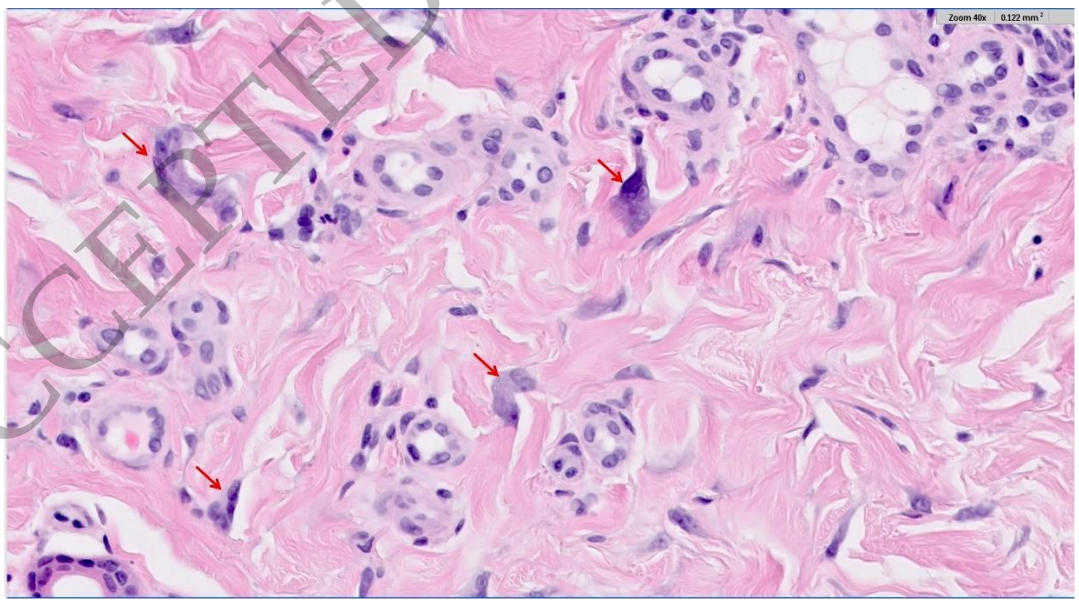

Figure 3
559x314 mm (x DPI)