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A slowly enlarging purple nodule on the arm

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The patient

A 50-year-old Caucasian gentleman presented with a one-year history of a gradually enlarging pigmented lump on the right forearm. On examination there was a 1 cm blue-purple nodule, which was hard in consistency (Figure 1). A mobile dermal cystic component was palpated after lateral pressure.

Further dermoscopic examination revealed a homogeneous blue-purple pigmentation surrounded by a subtle brown hyperpigmentation and a central white area with fine linear whitish structures. No pigment network was noticed (Figure 2).

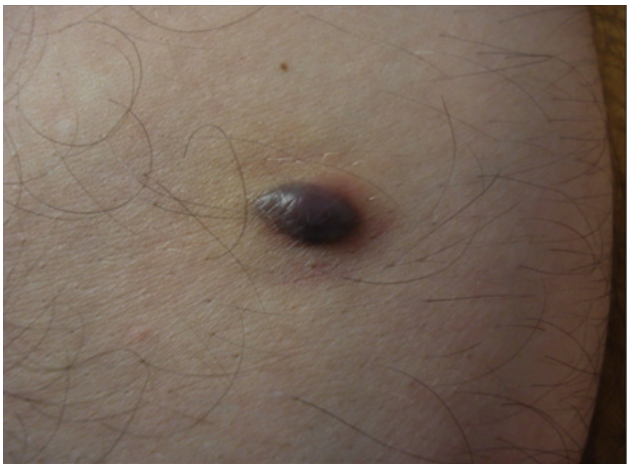

Figure 1. Well-defined, blue-purple nodule on right forearm. [Copyright: ©2016 Ramyead.]

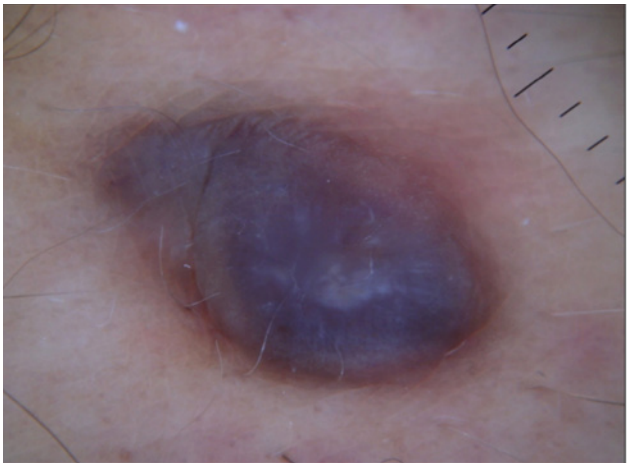

Figure 2. Homogeneous bluish purple pigmentation under dermoscopy surrounded by a subtle brown hyperpigmentation and a central whitish area with fine shiny linear whitish structures. [Copyright: ©2016 Ramyead.]

Histopathological examination was performed to exclude melanoma. It revealed an ill-defined nodular dermal proliferation of spindle-shaped fibroblasts and myofibroblasts arranged as short intersecting bundles. There were multiple dilated vascular channels and numerous hemosiderin-laden macrophages (Figure 3).

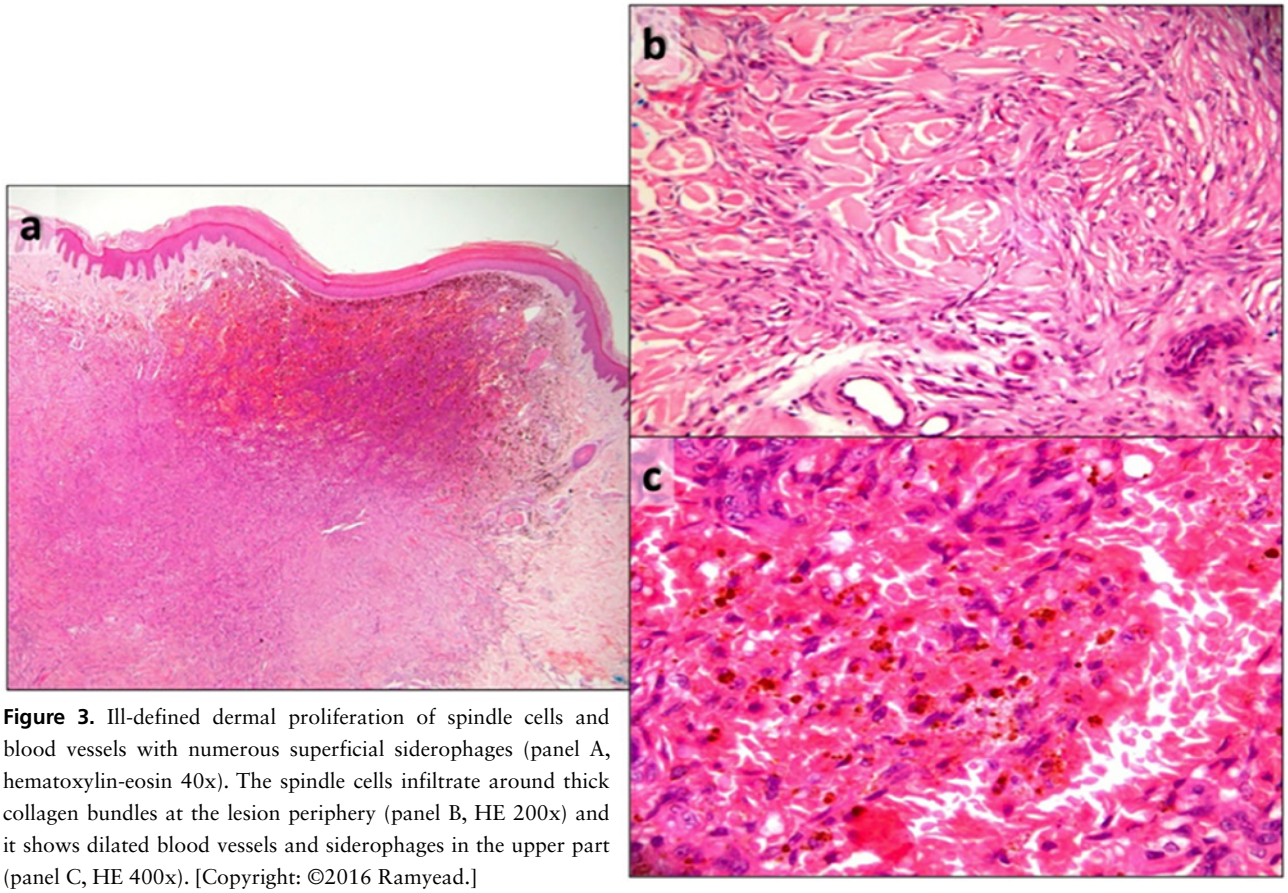

Figure 3. Ill-defined dermal proliferation of spindle cells and blood vessels with numerous superficial siderophages (panel A, hematoxylin-eosin 40x). The spindle cells infiltrate around thick collagen bundles at the lesion periphery (panel B, HE 200x) and it shows dilated blood vessels and siderophages in the upper part (panel C, HE 400x). [Copyright: ©2016 Ramyead.]

What is your diagnosis?

Answer and explanation

Aneurysmal dermatofibroma

Dermatofibromas are benign fibrous histiocytomas, representing the second most common cutaneous fibrohistiocytic lesion after acrochordons (skin tags). They present as firm, dome-shaped papules, which are commonly hyperpigmented and are primarily seen in adults on the lower extremities [1]. The dimpling of the skin with lateral compression or "Fitzpatrick's sign" is considered by many to be pathognomonic for dermatofibromas. Despite the description of this sign in all major textbooks, not all dermatofibromas dimple (as in our case), and all that dimple are not dermatofibromas. The exact etiology of dermatofibromas is unknown, however, they have been associated with trauma and insect bites [2]. They are usually solitary, but cases of multiple synchronous lesions (eruptive dermatofibromas) have been reported, especially associated with autoimmune disorders, in particular, systemic lupus erythematosus, atopic eczema and human immunodeficiency virus [3-5].

Histologically, dermatofibromas are characterized by a nodular dermal proliferation of spindled fibroblasts and myofibroblasts, arranged in intersecting fascicles with entrapment of thick collagen bundles in the superficial and mid dermis. There is hyperplasia of the epidermis with flat confluent rete ridges. The proliferating cells are positive for vimentin, factor XIIIa and CD68 and negative for CD34.

At least 20 different histological variants of dermatofibromas have been described with the aneurysmal subtype being unusual (2% of all cases) [6-8]. Aneurysmal dermatofibroma, also known as aneurysmal fibrous histiocytoma (AFH) was initially described in 1981 [9]. It presents as single dark papules or nodules, most commonly located on the lower limbs, which occasionally grow quickly and may bleed. They are clinically alarming and are often confused with melanocytic or vascular neoplasms [10]. Dermoscopy shows homogeneous pigmentation with different hues from blue-purple to red-brown colors. The presence of fine whitish linear structures corresponding to fibrosis is commonly seen, but these findings are non-specific and an excision biopsy is mandatory to exclude melanoma [10,11]. A rainbow pattern initially described in Kaposi sarcomas [12] has also been reported in aneurysmal dermatofibromas [13]. It can only be observed under polarized dermoscopy and is a result of the passage of polarized light through increased dermal fibrosis. Histologically, AFH consists of blood-filled spaces lacking endothelium within the fibrohistiocytic tumor. Perl's stain is positive, identifying stromal hemosiderin deposition, while the spindle cells are negative for endothelial (CD34) and melanocytic (S100) markers [6]. The pathogenesis is unknown, however, the blood is thought to represent extravasation of erythrocytes from capillaries. Important clinical differential diagnoses include malignant melanoma, hemangioma and Kaposi's sarcoma.

In summary, in the presence of a gradually enlarging, blue-purple, hard nodule with homogeneity on dermoscopy, there should be a high index of suspicion for an aneurysmal dermatofibroma. The dermoscopic pattern which may include fine white lines may be identical to some nodular melanomas, however the slow growth and long term evolution indicates its benign nature. These tumors require histological confirmation.

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