Asymptomatic ecchymotic plaque on the left leg

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CLINICAL FINDINGS

A 38-year-old male presented with occasionally itchy, irregularly shaped, ecchymotic plaque with a necrotic center on the left shin. The lesion started around 10 years back and progressively enlarged to reach 4×2 cm in size. (Fig. 1) It had not responded to topical medications. Cryotherapy was tried but with minimal and transient improvement. According to the patient, it started as a small papule and gradually increased in size and started to form a hypertrophic crust. The patient did not complain of any systemic illness and there was no family history of similar lesions.

On clinical examination, there was a single well-defined ecchymotic plaque with a violaceous border and a necrotic center. (Fig. 1) There was no tenderness or pain on palpation. Rest of the dermatological and systemic examination of the patient was normal. Nails and mucous membranes were not affected and showed no significant abnormalities. Routine laboratory investigations including CBC, blood sugar, hepatic and renal profile revealed no abnormal findings. X-ray of the lower limb showed no underlying bony involvement.

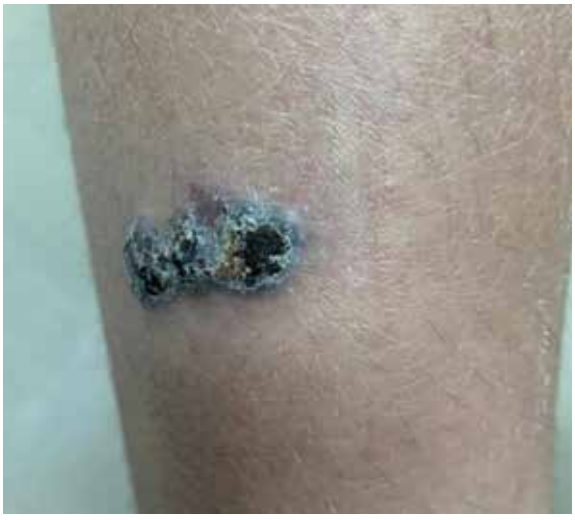

Fig. 1 Irregularly shaped, ecchymotic plaque with a necrotic center on the left shin

What is your clinical differential diagnosis?

- Solitary angiokeratoma
- Kaposi's sarcoma
- Angiosarcoma
- Melanoma

Excisional biopsy was performed for complete removal of the lesion and histological analysis revealed a dermal vascular proliferation, specifically prominent, dilated, thin-walled vessels in the mid dermis. Prominent endothelial cells created a "hobnail" appearance. Erythrocyte extravasation and hemosiderin deposition were present. (Fig. 2,3,4,5)

DIAGNOSIS

Targetoid Hemosiderotic Hemangioma

DISCUSSION

Targetoid Hemosiderotic Hemangioma (THH) was first described in 1988.[1] It is also known as Hobnail Hemangioma, a vascular, benign solitary

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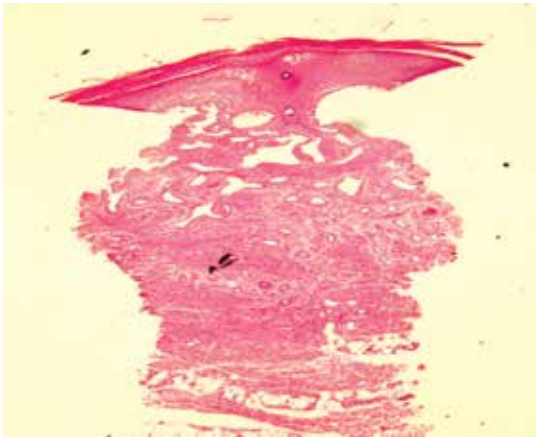

Fig. 2 Dilated vascular channels in mid dermis

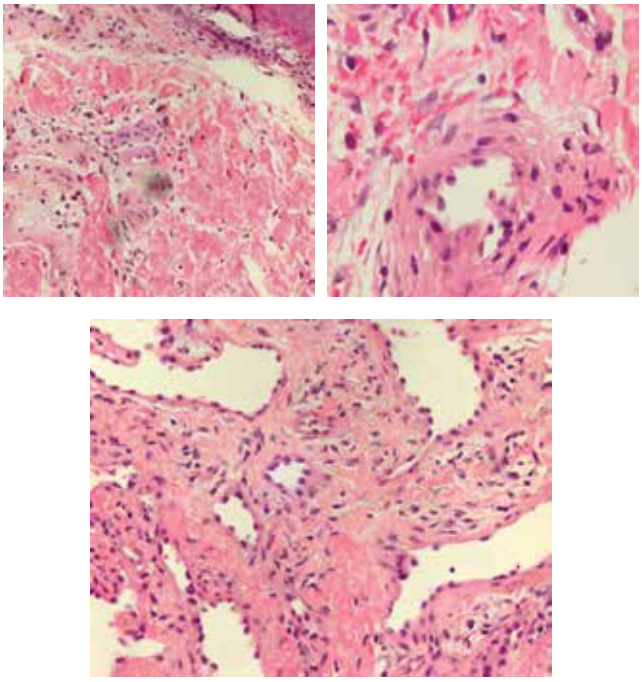

Fig. 3,4,5 Vascular channels lined with hobnail endothelial cells with extravasated RBC's and siderophages

lesion of unknown origin, probably lymphatic, which affects young or middle-aged people.[2] The most common clinical feature is a solitary violaceous papule or plaque surrounded by a pale, thin area and a peripheral ecchymotic ring, simulating a target, preferably located on the trunk or extremities.[3,4]

The targetoid appearance is not universal; cases with non-targetoid appearance have been described.[5] In the case of a non-targetoid or atypical presentation, it can be mistaken for other cutaneous vascular (hemangioma, angiokeratoma, Kaposi's sarcoma), melanocytic (melanocytic nevus, dysplastic nevus, melanoma) and fibrohistiocytic (dermatofibroma, fibroma) tumors.[6]

The exact pathogenesis of THH is unknown, although it has been postulated that trauma is the only known predisposing factor, for example due to irritation from a belt or insect bites.[7] Trauma could lead to the development of micro-shunts, in which the pressure of the capillaries would cause the filling of the lymph spaces of the lesion with erythrocytes, and contribute to the formation of aneurysmal microstructures. The obstruction of some efferent lymphatic vessels would result in inflammation, fibrosis and interstitial hemosiderin deposits.[8,9] Hormones may also influence the clinical morphology, resulting in the cyclic changes of waxing and waning diameter and peripheral colour.[10]

Dermoscopy shows well-demarcated red or red-bluish lakes (depending on the extent of involvement in the dermis). Black macules may also be seen and represent hemorrhagic crusts.[2]

Histolopathological findings varies according to the developmental stage in which the lesion was biopsied. In the initial stages, there is a biphasic pattern: in the papillary dermis, there are dilated vessels lined by a single layer of prominent epithelioid-like endothelial cells with solid intraluminal projections and hobnail appearance; and in the deep dermis, the vascular spaces are angulated and slit-like, resembling lymphatic vessels, which are concentrated around sweat glands, often forming small hemangiomatous nodules, dissecting the collagen bundles.[11] Extensive extravasation of red blood cells, inflammatory lymphocytic aggregates and fibrin thrombi are also present.[12] In later stages, the vascular lumen appears collapsed and there is

extensive deposition of hemosiderin in the stroma, as well as fibrosis.[11,12]

Immunohistochemically, the dilated and slit-like vascular vessels are labeled by lymphatic endothelial markers D2-40 and VEGFR-3 and negative or only focally positive for vascular endothelial markers such as CD34 and CD31, as well as for alpha-smooth muscle actin, which marks surrounding pericytes.[11-13]

The histopathologic differential diagnosis includes entities such as patch-stage Kaposi sarcoma and angiosarcoma. The dilated channels with intraluminal papillary projections and hemosiderin deposition, seen in THH, are absent in Kaposi sarcoma. Unlike Kaposi sarcoma, THH does not contain plasma cells and does not stain positively with human herpesvirus 8.[14] Angiosarcoma may be differentiated by increased endothelial cell atypia and a highly infiltrative pattern. The clinical appearance and histopathology of THH distinguishes it from other endothelial tumors bearing the so-called hobnail cytomorphology, such as retiform hemangioendothelioma, progressive lymphangioma, and papillary intra-lymphatic angioendothelioma.[15]

There is no effective preventive treatment for THH. Simple excision is curative and allows a correct histological diagnosis. Because it is a benign condition, such lesions are only removed for diagnostic or cosmetic reasons, since literature reports clearly indicate that neither local invasion nor dissemination takes place. There are no reports of recurrence after excision of the lesion.[16]

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