Targetoid hemosiderotic hemangioma

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A 30-year-old male presented with an asymptomatic erythematous patch over his left thigh of five years duration with an evolving central blackish growth over the same patch since the last six months. There was no history of any trauma preceding the lesion. Physical examination showed a raised blackish compressible papule over the anterior aspect of the left thigh, 0.5 cm in diameter, surrounded by a peripheral purpuric ring with a few pale areas [Figure 1]. There was no other cutaneous or systemic abnormality. Excision biopsy showed vascular proliferation with dilated and bizarre-shaped vessels in the papillary dermis and extending into the upper reticular dermis. Numerous extravasated erythrocytes were present between collagen bundles. A sparse lymphohistiocytic infiltrate was noted at the base of lesion. At higher magnification, the vessels were found to be lined by a single layer of endothelial cells with no papillary projections protruding into the lumen. Deposition of hemosiderin, though noted throughout, was more pronounced in areas of extravasation of erythrocytes [Figure 2 and 3].

On clinicopathological correlation, a diagnosis of targetoid hemosiderotic hemangioma (THH) was made. The clinical targetoid appearance is explained by hemorrhage from the vascular proliferation (causing a purpuric ring, with hemosiderin deposits contributing to the central black color).

THH arises commonly over the extremities and trunk. The classic clinical presentation of THH is that of a single, annular, targetoid-appearing lesion made up of a violaceous papule surrounded by an ecchymotic ring. These lesions are generally less than 1 cm in diameter. The clinical presentation may not always be as characteristic as seen in present case. Lesions may present as angiomatous macule or papule without any halo. This variation has been interpreted as different stages in the process of evolution and devolution of the lesion, as result of hormonal influences, or trauma to a pre-existing angioma. Pale areas within purpuric ring represent spontaneous devolution.[1]

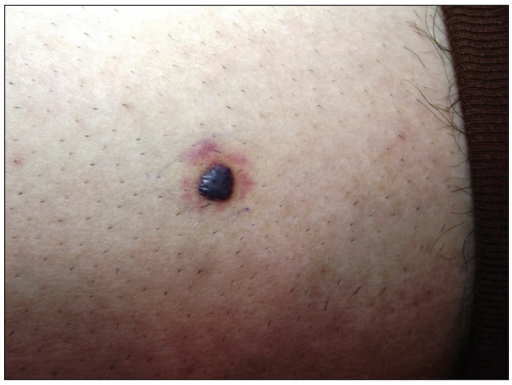

Figure 1: Hyperpigmented papule with ecchymotic ring having some pale areas also on anterior thigh

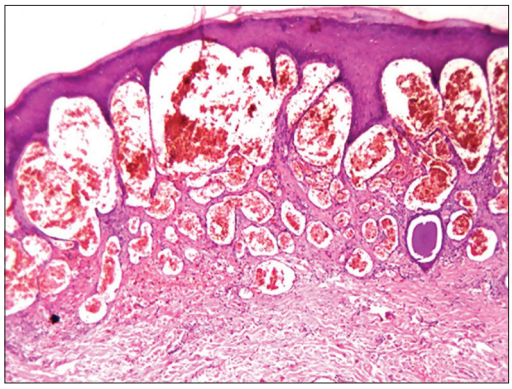

Figure 2: Well-circumscribed vascular proliferation in papillary dermis and extending into upper reticular dermis (H and E, ×40)

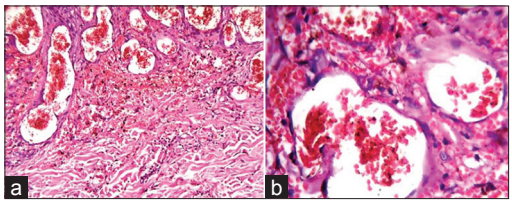

Figure 3: (a) High power view showing bizarre-shaped vascular proliferations with erythrocyte extravasation and hemosiderin deposition. (H and E, ×100) (b) Vascular channels lined by single layer of endothelial cells, extravasated erythrocytes and diffuse hemosiderin deposition. (H and E, ×400)

Clinical differential diagnoses include melanocytic lesions, infantile hemangioma, tufted angioma, Kaposi sarcoma, insect bite, erythema multiforme, and dermatofibroma, depending on the stage of evolution. The histological differential diagnoses include early Kaposi's sarcoma, epithelioid hemangioma, progressive lymphangioma, and well-differentiated angiosarcoma.[1,2]

THH shows both clinical and histopathological variations. In a study of 62 cases by Mentzel *et al*., a biphasic growth pattern of dilated vessels lined by hobnail endothelial cells in superficial parts of the lesion and narrow vessels in deeper parts of the lesion dissecting the collagen were described. Some lesions resembled lymphangioma circumscriptum, retiform hemangioendothelioma, progressive lymphangioma, and Dabska's tumor, while some resembled cavernous lymphangioma (as in the present case).

Late lesions have been described as showing collapsed vascular lumina, fibrosis, and abundant hemosiderin. Findings in the present case were consistent with late lesion.[2,3]

The lymphatic endothelial cell marker D2-40 (podoplanin) has been found strongly positive in most of the case series. Thus THH has recently been shown to be a lymphatic malformation.[1,4]

Simple excision is curative and recurrence is not usually seen.[2,3]

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