

Asymptomatic solitary nodule on the left arm

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A solitary erythematous nodule on the arm

A 31-year old Kuwaiti female patient presented with a solitary skin lesion on the left arm since 1 year. The lesion started as a small red lesion that slowly progressed into a large nodule with erythematous halo. The lesion was painless and no other symptom was reported. There was no previous history of similar lesion and there was no history of any systemic disease.

On examination, there was a solitary well-defined exophytic red nodule, measuring 1.1x0.9 cm. The surface of the nodule was intact and showed slight glistening. The lesion was non-tender and it was soft in consistency (Fig. 1). On the periphery, there was a halo of normal skin surrounded by a rim of purpuric lesion (Fig. 2). No abnormal findings were observed in the axillary lymph nodes and no other skin lesions were seen on general examination.



Fig. 1 A female patient with a red nodule with a purpuric rim on the left arm.



Fig. 2 The lesion shows a targetoid like appearance.

Routine laboratory investigations including complete blood count, kidney function tests, liver function tests, and thyroid function tests were normal. Radiological examination (X-ray) showed no attachment to underlying structures.

What is the clinical diagnosis?

1. Aneurismal dermatofibroma
2. Targetoid hemosiderotic hemangioma
3. Tufted angioma
4. Capillary hemangioma

Total surgical excision was performed for the nodular part of the lesion and the biopsy specimen was subjected for histological examination. The skin sections showed pandermal proliferation of blood vessels extending from the papillary dermis into the deep portion with more accentuation in the upper dermis (Fig. 3). The blood vessels were different sizes and all were lined by a single layer of plumed endothelial cells with a characteristic

arrangement in a linear row (Fig. 4). The stroma showed marked extravasations of erythrocytes with scattered hemosiderin deposits (Fig. 5).

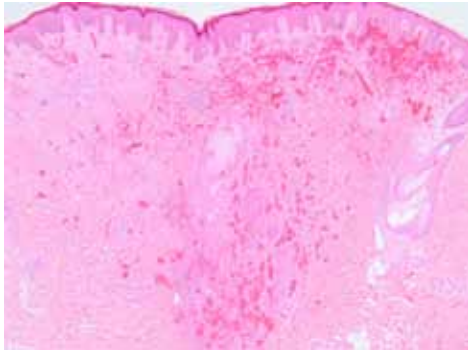


Fig. 3 Pandermal proliferation of blood vessels extending from the papillary dermis into the deep portion

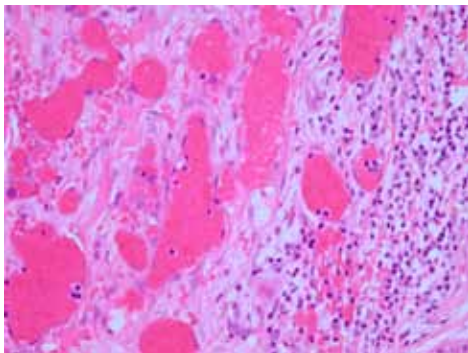


Fig. 4 Different sizes blood vessels that are lined by a single layer of plump endothelial cells.

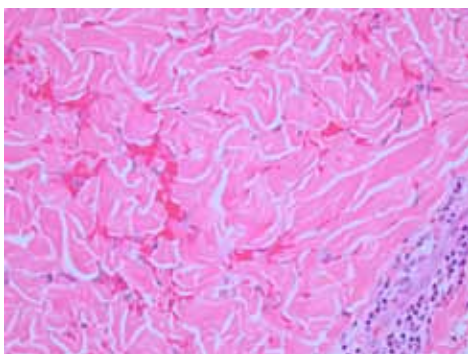


Fig. 5 The stroma showed marked extravasations of erythrocytes with scattered hemosiderin deposits.

The diagnosis is

Targetoid hemosiderotic hemangioma (THH)
(hobnail hemangioma)

DISCUSSION

Targetoid hemosiderotic hemangioma (Hobnail hemangioma) is a rare benign vascular neoplasm, which was first described by Santa Cruz and Aronburg in 1988.¹ Clinically a typical clinical appearance of THH appears as an asymptomatic small (2-3mm), solitary, red, purple, and/or brown papule that is typically encircled by a pale area and peripheral ecchymotic ring giving a targetoid appearance.² However, these features are only present in a small percentage of cases and, most often, the clinical appearance is that of a red-blue or brown papule. The characteristic targetoid appearance is due to peripheral hemorrhage and subsequent deposition of hemosiderin.³

Published reports of hobnail hemangioma reveal an equal gender incidence, with an age range of presentation of 5-67 years. It is identified more frequently in younger persons.⁴ There are 10 case reports in the Korean literature. In those cases, skin lesions developed at 3-32 years. The lesion of THH is often solitary, but multiple lesions can occur. The lesions are mainly distributed on the trunk and extremities.⁵

The main histopathological feature of THH is a dermal vascular proliferation with biphasic growth patterns. In the papillary dermis, the ectatic vascular spaces are often lined with a single layer of prominent plump endothelial cells protruding into the lumen of the vessels (hobnail endothelial cells).⁶ There are occasional intraluminal papillary projections. The hobnail endothelial cell has scanty cytoplasm and rounded nuclei that protrude into the lumina.⁷

The vascular channels in the deeper dermis become much less conspicuous and eventually disappear completely. The whole architecture is wedge-shaped, with a prominent superficial component.

In the later stages, extensive stromal hemosiderin deposits are commonly seen.⁸ The histopathologic differential diagnosis of THH include patch stage Kaposi's sarcoma, solitary angiokeratoma, progressive lymphangioma, eosinophilic hemangioma, and retiform hemangioendothelioma (RHE).⁹ The etiology of THH is unknown, one well-accepted mechanism is that of THH developing from trauma to a pre-existing hemangioma. Some patients with THH have been reported in whom the lesions changed periodically during menstruation under the influences of sex steroid hormones, but immunohistochemical staining failed to demonstrate either estrogen or progesterone receptors in these cases.¹⁰

It was also postulated that THH is better considered as a lymphatic malformation but not a lymphatic neoplasm on the basis of the WT1 negativity.¹¹ Santonja and Torrelo suggested a vascular origin for hobnail hemangioma since it had positive reaction with Factor VIII-related antigen, CD31 and CD34.¹²

Immunohistochemistry usually shows a strong immunopositivity for CD31 and factor VIII. CD34 is moderately immunopositive and VEGF is usually negative. CD31 is the best marker for benign and malignant vascular tumors.¹³ Previous studies have documented CD31 positivity in majority of the cases and CD34 positivity in minority of the cases. Furthermore, a limited number of cases may stained positively for VEGF and D2-40, which are the markers of vascular tumors with presumed lymphatic differentiation.¹⁴ Results of such studies suggested a lymphatic line of differentiation of neoplastic cells in hobnail hemangioma. However, immunoexpression of VEGF and D2-40 in angiomatous entities has alternatively been interpreted by some as a possible origin from

stem cells that would be able to differentiate either into lymphatic cells or blood vessels.¹⁵

Treatment options for THH are limited and non-specific. Excision of small lesions is the most accepted therapeutic option. Therapeutic sclerosis of the ectatic spaces by the injection of sclerosing solutions is theoretically possible but would probably not affect the lesion beyond the small area treated.¹⁶

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