Unusual targetoid nodule on the back

A 28-year-old woman presented with an asymptomatic recurrent nodule on her back for 6 months. The lesion was increasing in size and the patient noted variations in the nodule's color from pink to light brown. On examination, a non-tender, red to violaceous nodule with a thin pale brown margin and a peripheral ecchymotic ring measuring 1.6 cm × 1 cm was seen on the right posterior shoulder [Figure 1]. The remainder of the dermatological examination was unremarkable.

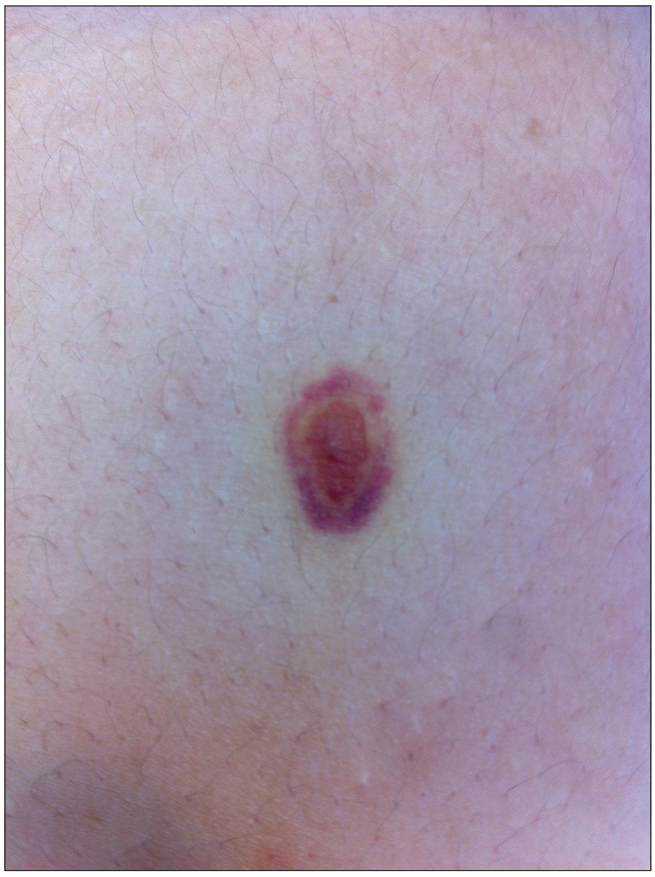

Figure 1: An erythemato-violaceous, non-tender, central nodule surrounded by a thin pale brown area and a peripheral ecchymotic ring measuring 1.6 cm × 1 cm on the right posterior shoulder

An excisional biopsy showed dilated vascular channels in the papillary dermis with a lining of prominent endothelial normotypic cells. Extravasated erythrocytes and perivascular superficial mononuclear inflammatory infiltrate were also seen [Figure 2]. Immunohistochemistry revealed intense positive reaction for the lymphatic endothelial cell marker podoplanin (D2–40) [Figure 3]. Perl's stain was also positive [Figure 4]. The study was negative for Wilms' tumor 1 gene.

What is Your Diagnosis?

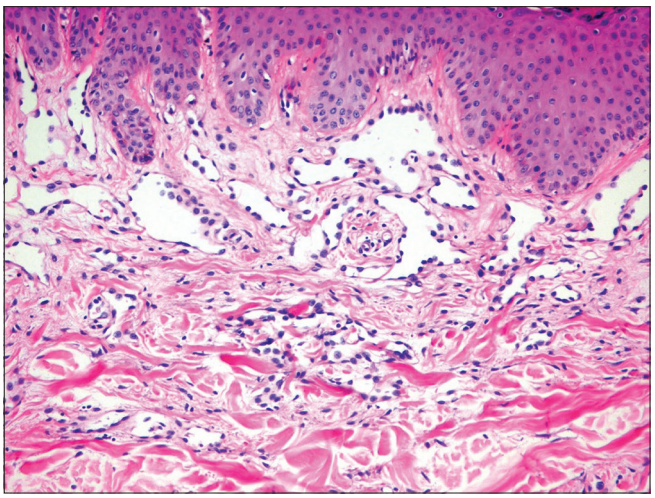

Figure 2: Dilated vascular channels in papillary dermis with a lining of prominent endothelial normotypic cells. Extravasated erythrocytes and perivascular superficial mononuclear inflammatory infiltrate and hemosiderophages were also found (H and E, ×100)

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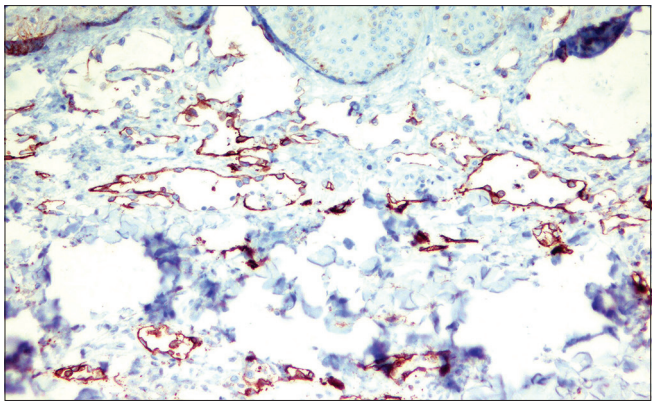

Figure 3: Immunohistochemical study showed intense positive reaction for the lymphatic endothelial cell marker podoplanin (D2–40, ×100)

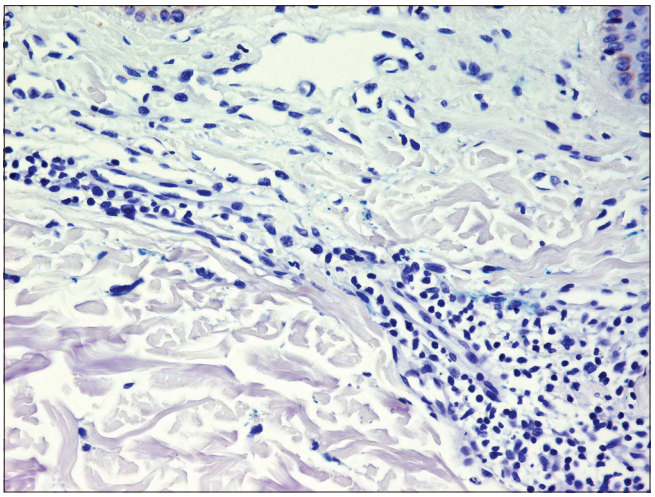

Figure 4: Isolated hemosiderophages around blood vessels at the periphery of the lesion (Perl's stain, ×400)

Answer

Targetoid hemosiderotic hemangioma.

Discussion

Targetoid hemosiderotic hemangioma, also known as superficial hemosiderotic lymphovascular malformation and previously known as hobnail hemangioma, is a rare lymphovascular lesion. Its variable and dynamic clinical manifestations present a diagnostic challenge to physicians.

Clinically, it is characterized by a "targetoid" appearance with a brown to violaceous papule or nodule surrounded by a thin, pale area and a peripheral ecchymotic ring that can expand or eventually disappear with the persistence of the central papule.[1] It presents in the third and fourth decade of life, mostly on the trunk and extremities, and has been present for weeks and up to 6 years before coming to medical attention.[2-4] It can be asymptomatic or tender. The clinical course varies among patients, some describe progressive growth of the lesion whereas others report no change. The peripheral ring can manifest intermittently or be persistent.

The exact etiology of targetoid hemosiderotic hemangioma remains unclear; physical trauma, inflammation and hormonal changes (because of the periodic changes related to menstrual cycle and pregnancy) have been proposed as causes.[2]

Histopathologic findings vary with the duration of the lesion. In early stages the dilated, irregular thin walled, ectatic vascular spaces in the papillary dermis are characteristic. Early lesions also show prominent endothelial cells, hemosiderin deposits, extravasated erythrocytes and a mild mononuclear inflammatory infiltrate without eosinophils, plasma cells or neutrophils. The findings in later stages are collapsed vascular lumina, fibrosis and abundant hemosiderin.[1]

To determine the origin of this lesion, several immunohistochemical markers have been analyzed. Positive reaction for CD31 proposes a vascular origin while D2–40 and vascular endothelial growth factor-3 support a lymphatic differentiation. The more generally accepted theory is that it is a biphasic lesion with lymphatic and vascular features.[3] On the other hand, the negative reaction for glucose transporter-1 and Wilms' tumor 1 suggests that it is a malformation rather than a neoplasm.[5]

Although entirely benign, its particular histology requires differentiation from vascular malignancies such as Kaposi's hemangioma, conventional angiosarcoma, Dabska tumor, and retiform hemangioendothelioma. This is why, even though the clinical manifestations can be strongly suggestive of superficial hemosiderotic lymphovascular malformation, histopathological study needs to be performed to rule out these neoplastic lesions.

Because of its benign nature, treatment is not always performed. Patients can be treated with local excision or excisional biopsy. Metastases or recurrence after excision have not been reported. In our patient the lesion was completely excised with no recurrences to date.

We agree that the term superficial hemosiderotic lymphovascular malformation is also a good term for this lesion that has several clinical presentations but characteristic histological findings; a biopsy therefore is the key to diagnosis.

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Conflicts of interest

There are no conflicts of interest.

Isidora García-Huidobro, Álvaro Abarzúa-Araya, Camila Downey, Sergio González[1]

Department of Dermatology, Facultad de Medicina, Pontificia Universidad Católica de Chile, [1]Department of Pathology, Facultad de Medicina, Pontificia Universidaad Católica de Chile, Santiago, Chile

Correspondence: Dr. Camila Downey, Department of Dermatology, Facultad de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile.
E-mail: camiladowney@gmail.com

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