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From the Dermatologikum Hamburg: Quiz

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The patient

A 48-year-old woman presented herself with an erythematous macule on the extensor surface of the right thigh. The lesion had been present for years, but recently the patient noticed some increase in size and changes in the coloration. This had prompted her to see a dermatologist (Figure 1). A biopsy was taken and photomicrographs are presented in Figures 2A-I. What is your diagnosis?

Answer and explanation

Hobnail lymphatic malformation (so-called hobnail hemangioma or targetoid hemosiderotic hemangioma)

Dilated and bizarre-shaped vessels are seen in the upper reticular dermis and extend into the papillary dermis. At the periphery of the lesion vessels are smaller in diameter and assume slit-like configurations between collagen bundles reminiscent of the macular stage of Kaposi's disease. At

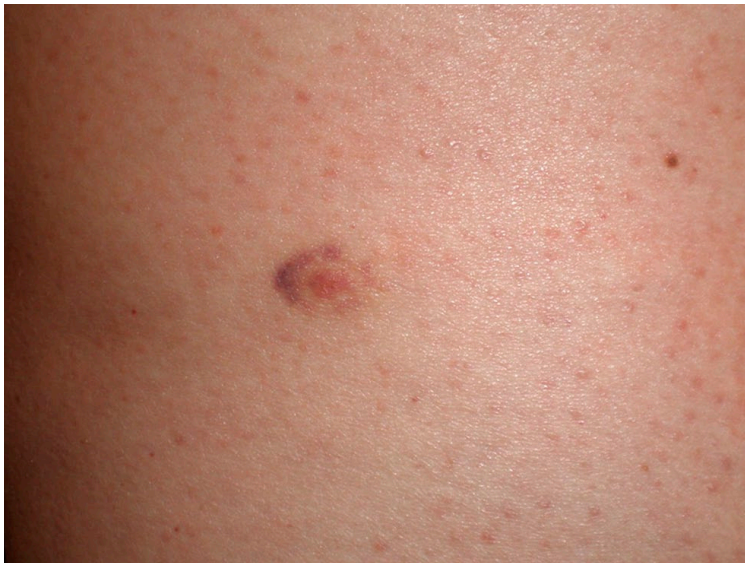

Figure 1. Clinical appearance. [Copyright: ©2011 Böer-Auer.]

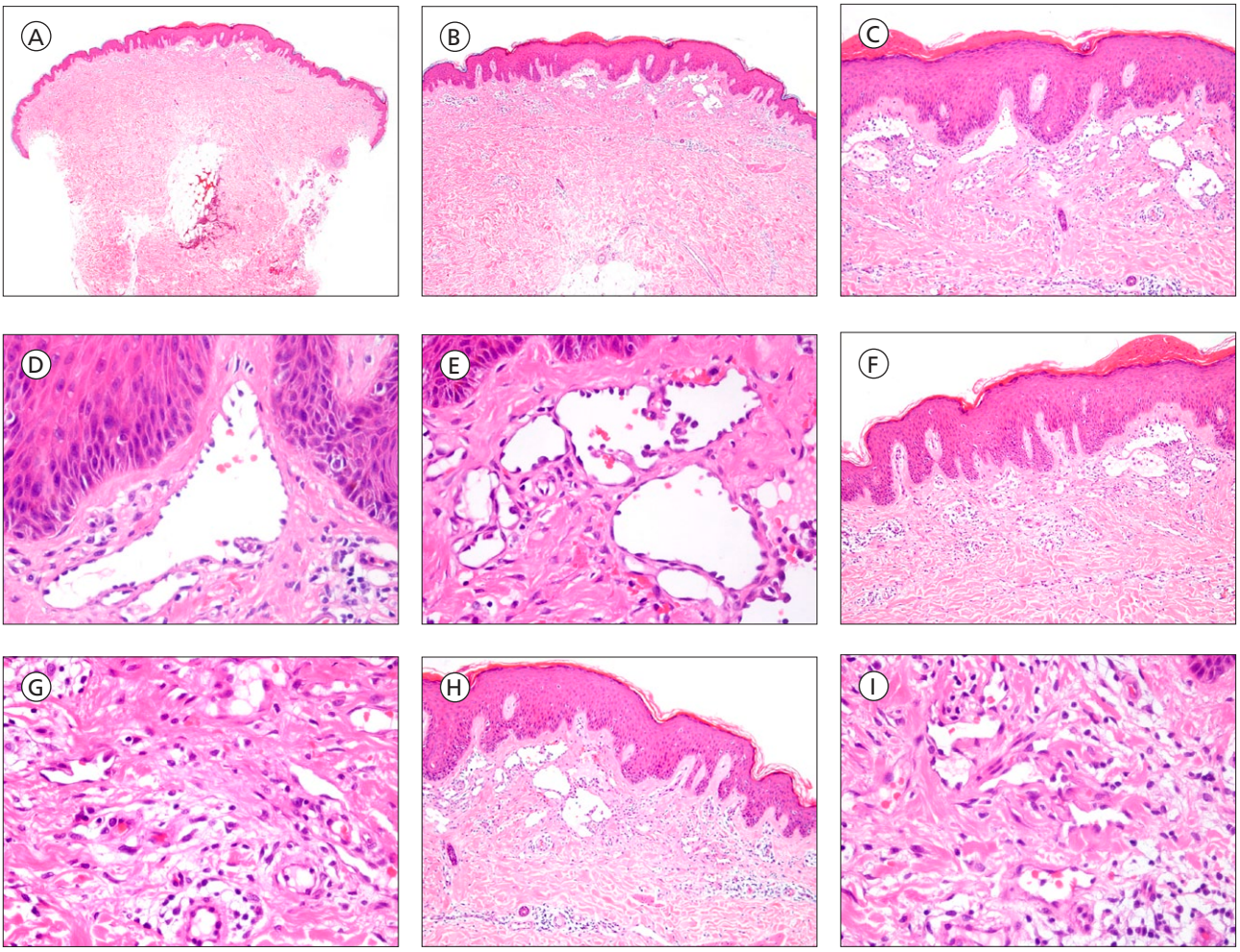

Figure 2A-I (left and above). Histopathology. [Copyright: ©2011 Böer-Auer.]

higher magnification, the vessels are seen to be lined by a single layer of endothelial cells with plump nuclei. Nuclei have a typical hobnail appearance in some vessels, whereas others show what has been described as a matchstick morphology. In some foci, papillary projections protrude into the lumen of a vessel.

The lesion is covered by a broad zone of parakeratosis. Numerous extravasated erythrocytes are present between collagen bundles. A sparse infiltrate of lymphocytes, histiocytes, and few plasma cells is also present. Iron stain highlights numerous hemosiderophages (Figure 3).

In clinicopathological correlation, the findings seen in this patient are stereotypical of a fully developed lesion of so-called hobnail lymphatic malformation (HLM) (formerly called hobnail hemangioma or targetoid hemosiderotic hemangioma). The targetoid appearance seen clinically is explained by the hemorrhage and the hemosiderin deposits that form a halo around the angioma.

HLM arises commonly on the extremities and trunk and lesions are usually less than 1 cm in diameter. The clinical presentation is not always as characteristic as seen here. Studies on larger series of patients have shown that often lesions present as angiomatous macule or papule without any halo [1-3]. This variation has been interpreted as different stages in the process of evolution and devolution of the lesion, as result of hormonal influences, or as a result of previous trauma to a preexisting angioma. In the case shown here, the broad zone of parakeratosis indicates that the lesion has, in fact, been irritated from the outside.

The clinical variation of the lesion is reflected in the number of different diseases that have been considered as clinical differential diagnoses of patients reported in the literature [1-3]. It includes such conditions as disparate as melanocytic lesions, infantile hemangioma, tufted angioma, Kaposi sarcoma, insect bites, erythema multiforme, and dermatofibroma.

Not only clinically but also histopathologically HLM shows some variation. Mentzel et al studied 62 cases and described a biphasic growth pattern of dilated vessels lined by hobnail endothelial cells in superficial parts of the lesion and collagen dissecting narrow vessels in deeper parts of the lesion. Some lesions resembled cavernous lymphangioma or lymphangioma circumscriptum, others had features reminiscent of retiform hemangioendothelioma, progressive lymphangioma and so-called Dabska's tumor [3]. Late lesions have been described as showing collapsed vascular lumina, fibrosis, and abundant hemosiderin [3].

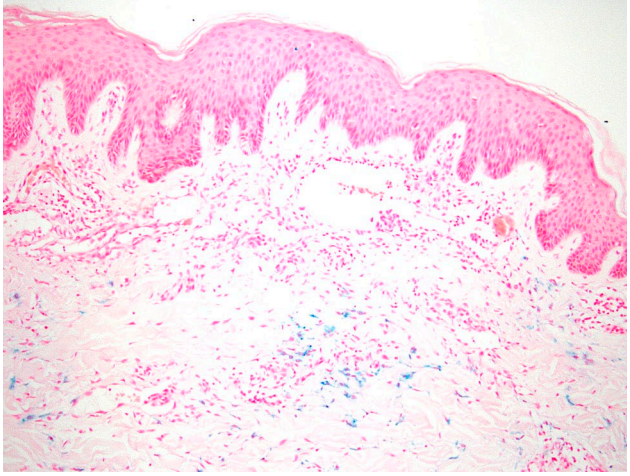

Figure 3. Iron staining. [Copyright: ©2011 Böer-Auer.]

It is important to note that hobnail endothelial cells are not unique to hobnail angioma but may also be seen in retiform hemangioendothelioma, progressive lymphangioma, and papillary intralymphatic angioendothelioma. But the most important differential diagnosis of HLM is the macular stage of Kaposi's disease. The promontorium sign, slit-like proliferations of capillaries, hemorrhage, hemosiderophages, and infiltrates containing plasma cells can be seen in both conditions [2-5]. However, intraluminal papillary projections and hobnail appearance of endothelial cells is usually not seen in Kaposi's disease and vessel proliferations of a macular stage of Kaposi's disease almost always spare the papillary dermis, whereas hobnail angioma typically involves the papillary dermis. Moreover, lesions of Kaposi's disease are usually positive for human herpesvirus type 8.

The condition presented here was first described in 1988 by Santa Cruz and Aronberg. They recognized it as a distinctive entity and characterized it clinically and histopathologically [1]. They were impressed by the clinical appearance, which was targetoid in all of their cases and termed it "targetoid hemosiderotic hemangioma." The term "hobnail hemangioma" was introduced in 1999 by Guillou et al, who attempted to emphasize the histopathological hallmark of the condition, namely, endothelial cells shaped like hobnails and matchsticks [2].

HLM is conventionally classified as a benign vascular tumor, but it has been a matter of controversy whether it can be differentiated from blood vessels or lymphatics and whether it is a true neoplasm or a malformation. Franke et al, in 2004, showed that endothelia of hobnail hemangiomas exhibit an antigenic profile similar to normal lymphatics, the majority of neoplastic vascular channels expressing CD31 but not CD34 and lacking pericytes [4]. Moreover, the lymphatic endothelial cell marker D2-40 (podoplanin) was strongly positive in all lesions studied by them. Therefore, they suggested that what had been called hobnail hemangioma really was a lymphangioma and that naming it "hemosiderotic angioma" or "hemosiderotic cutaneous lymphangioma" would be more appropriate [4].

Very recently, in January 2012, immunostaining with the endothelial marker Wilms tumor 1 (WT1) has been investigated in larger series of the tumor and has illuminated the nature of it. WT1 is considered to be expressed by vascular neoplasms but not by malformations. Trindade and coworkers studied 52 cases of HLM and none of them expressed WT1. In 10 of their cases, the authors also performed other stainings and found lesions to be positive with D2-40 and negative with Ki-67 and human herpesvirus 8 latent nuclear antigen. The authors concluded that these results supported a lymphatic line of differentiation, but because of WT1 negativity it should be classified as a lymphatic malformation. They suggested the term "superficial hemosiderotic lymphatic malformation" for the condition [5].

At the same time, Al Dhaybi et al investigated 12 pediatric cases of HLM with almost the same set of immunohistochemical markers. They found D2-40 immunostaining to be positive in all cases, while WT1 was largely negative. The authors also concluded that hobnail hemangioma should be classified as lymphatic vascular malformation and proposed the name "targetoid hemosiderotic lymphatic malformation" [6].

In short, what has been described as hobnail hemangioma (or targetoid hemosiderotic hemangioma) has recently been shown to be a lymphatic malformation based on immunohistochemical staining characteristics. That is why, here, we replaced the former term hobnail hemangioma with the term hobnail lymphatic malformation.

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