Ill-Defined Macule on the Abdomen

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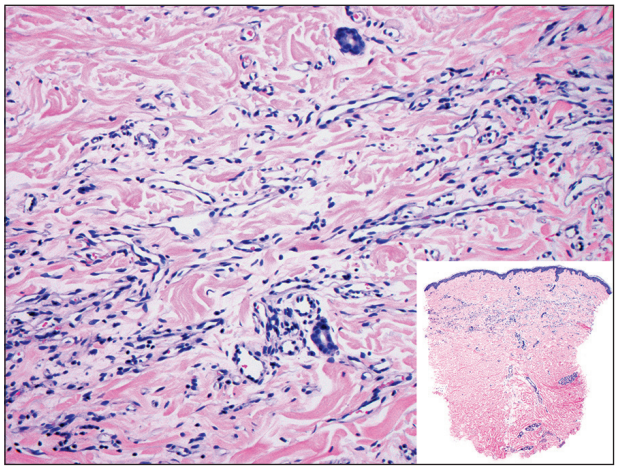

H&E, original magnification ×200 (inset, original magnification ×40).

A 38-year-old woman presented with an asymptomatic lesion on the abdomen. On physical examination, there was a 5×2-mm, solitary, ill-defined pink macule on the right side of the abdomen. The patient denied recent change in size or color of the lesion, prior trauma, or a personal or family history of similar lesions. Due to the uncertain diagnostic appearance, a punch biopsy was performed.

THE BEST **DIAGNOSIS IS:**

a. cutaneous angiosarcoma
b. Kaposi sarcoma
c. microvenular hemangioma
d. targetoid hemosiderotic hemangioma
e. tufted angioma

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THE **DIAGNOSIS:** Microvenular Hemangioma

Microvenular hemangioma is an acquired benign vascular neoplasm that was described by Hunt et al[1] in 1991, though Bantel et al[2] reported a similar entity termed *micropapillary angioma* in 1989. Microvenular hemangioma typically presents as a solitary, slowly enlarging, red to violaceous, asymptomatic papule, plaque, or nodule measuring 5 to 20 mm in diameter. It usually is located on the trunk, arms, or legs of young adults without any gender predilection. Microvenular hemangioma is rare.[3] The etiology has not been elucidated, though a relationship with hormonal factors such as pregnancy or hormonal contraceptives has been described.[2]

Histopathologically, microvenular hemangioma has a characteristic morphology. It is comprised of a well-circumscribed collection of thin-walled blood vessels with narrow lumens (quiz image).[4] The blood vessels tend to infiltrate the superficial and deep dermis and are surrounded by a collagenous or desmoplastic stroma. The endothelial cells are normal in size without atypia, mitotic figures, or pleomorphism. A mild lymphoplasmacytic inflammatory infiltrate sometimes is present. Microvenular hemangioma expresses many vascular markers confirming its endothelial origin, including CD34, CD31, WT1, factor VIII–related antigen, and von Willebrand factor.[3] Moreover, WT1 staining suggests the lesion is a vascular proliferative growth, as it usually is negative in vascular malformations due to errors of endothelial development.[5] In addition, it lacks expression of podoplanin (D2-40), which also supports a vascular as opposed to a lymphatic origin.[4]

Cutaneous angiosarcoma is a rare and highly aggressive malignant neoplasm of the vascular endothelium with a predilection for the skin and superficial soft tissue. Clinical presentation is variable, as it can arise sporadically, commonly on the scalp and face of elderly patients, in areas of chronic radiation therapy, or in association with chronic lymphedema (Stewart-Treves syndrome).[6] Sporadic neoplasms appear clinically as purpuric macules, plaques, or nodules and are more common in elderly men than women. They are aggressive tumors that tend to recur and metastasize despite aggressive therapy and therefore carry a poor prognosis.[7] Histopathologically, well-differentiated tumors are characterized by irregular dissecting vessels lined with crowded inconspicuous endothelial cells (Figure 1). Cutaneous angiosarcoma is poorly circumscribed with marked cytologic atypia, and the vessels can take on a sinusoidal growth pattern.[8]

Kaposi sarcoma (KS) is a virally induced lymphoangioproliferative disease, with human herpesvirus 8 as the implicated agent. There are 4 principal clinical variants of KS: epidemic or AIDS-associated KS, endemic or African KS, KS due to iatrogenic immunosuppression, and Mediterranean or classic KS.[9] Cutaneous lesions vary from pink patches to dark purple plaques or nodules that commonly occur on the lower legs[10]; however, the clinical appearance of KS varies depending on the clinical variant and stage. Histopathologically, early lesions of KS exhibit a superficial dermal proliferation of small angulated and jagged vessels that tend to separate into collagen bundles and are surrounded by a lymphoplasmacytic perivascular infiltrate. These native vascular structures often are surrounded by more ectatic neoplastic channels with plump endothelial cells, known as the promontory sign (Figure 2).[11] With more advanced lesions, the proliferation of slitlike vessels becomes more cellular and extends deeper into the dermis and subcutis. Although the

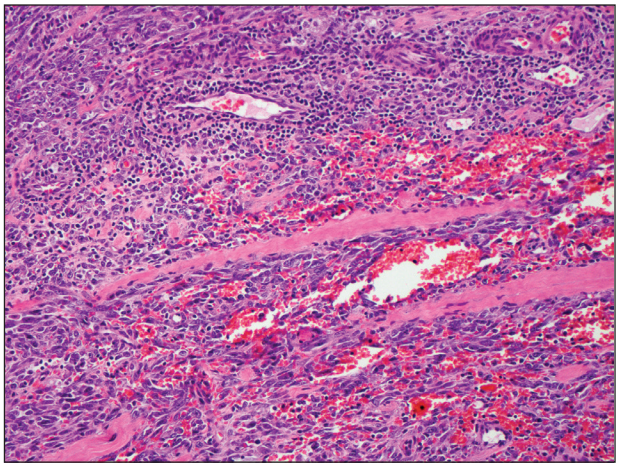

FIGURE 1. Cutaneous angiosarcoma. Dermal proliferation of irregular dissecting vessels lined with crowded inconspicuous endothelial cells (H&E, original magnification ×200).

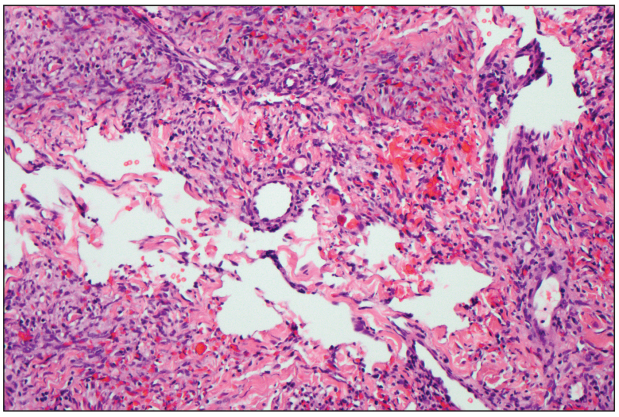

FIGURE 2. Kaposi sarcoma. Angulated and jagged vessels surrounded by a lymphoplasmacytic perivascular infiltrate and ectatic neoplastic channels, known as the promontory sign (H&E, original magnification ×200).

histopathologic features vary with the stage of the lesion, they do not notably vary between clinical subtypes.

Targetoid hemosiderotic hemangioma, also known as hobnail hemangioma, is a small, benign, vascular tumor that usually affects the trunk, arms, and legs in young to middle-aged adults without a gender predilection. Clinically, it appears as a small, solitary, red to purple papule or macule that typically is surrounded by a pale thin area and a peripheral ecchymotic ring, creating a targetoid appearance, thus the term *targetoid hemosiderotic hemangioma*.[12] Histopathologically, there is a prominent dermal vascular proliferation. In the papillary dermis, there are dilated superficial vessels lined with a single layer of endothelial cells characterized by a plump, hobnail-like appearance that protrude into the lumen (Figure 3). In the deeper dermis, the vascular spaces are angulated and slitlike and appear to dissect through collagen bundles. Hemosiderin, thrombi, extravasated erythrocytes, and a lymphocytic infiltrate also are often seen.[13]

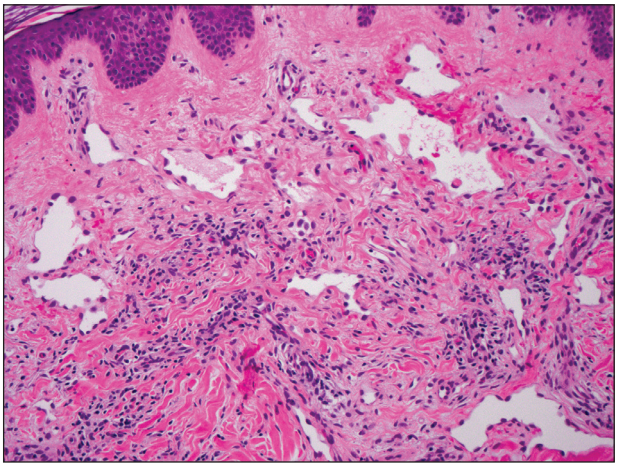

FIGURE 3. Targetoid hemosiderotic hemangioma. Dilated superficial vessels lined by plump, hobnail-like endothelial cells that protrude into the lumen (H&E, original magnification ×200).

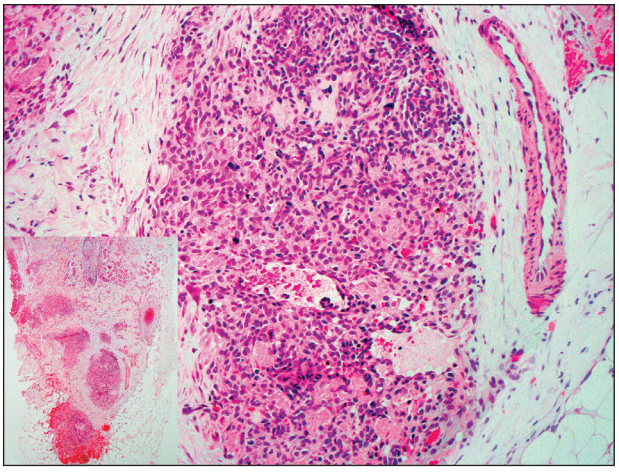

FIGURE 4. Tufted angioma. Discrete lobules or tufts of tightly packed capillaries in a cannonball-like appearance throughout the dermis (H&E, original magnification ×200 [inset, original magnification ×40]).

Tufted angioma is a rare benign vascular lesion that usually presents as an acquired lesion in children and young adults, though it may be congenital. It is commonly localized to the skin and subcutaneous tissues. Clinically, the lesions appear as red to purple patches and plaques that typically are located on the neck or trunk. More than 50% of cases present during the first year of life and slowly spread to involve large areas before stabilizing in size.[14] Partial spontaneous regression may occur, but complete regression is rare.[15] Lesions usually are asymptomatic but may be painful during periods of platelet trapping (Kasabach-Merritt phenomenon), which may develop in congenital cases. Tufted angioma is named for its characteristic histopathologic appearance, which consists of multiple discrete lobules or tufts of tightly packed capillaries in a cannonball-like appearance throughout the dermis and subcutis (Figure 4).[14,15]

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