Painful, Nonhealing, Violaceus Plaque on the Right Breast

Patrick McMullan, PhD; Lorin A. Bibb, MD; Katalin Ferenczi, MD

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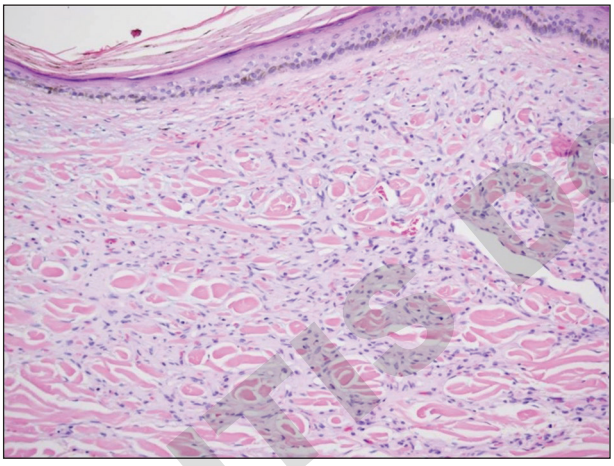

H&E, original magnification ×100.

A 42-year-old woman with a medical history of hypertension and smoking tobacco (5 pack years) presented with a painful, nonhealing, violaceous, reticulated plaque with ulceration on the right breast of 3 months' duration. Histopathology revealed diffuse, interstitial, bland-appearing spindle cells throughout the papillary and reticular dermis that were distributed between the collagen bundles. Dermal interstitial spindle cells were positive for CD31, CD34, and erythroblast transformation specific–related gene immunostains. Factor XIIIa and human herpesvirus 8 immunostaining was negative.

THE BEST **DIAGNOSIS IS:**

a. angiosarcoma
b. dermatofibroma
c. dermatofibrosarcoma protuberans
d. diffuse dermal angiomatosis
e. Kaposi sarcoma

PLEASE TURN TO PAGE 26 FOR THE DIAGNOSIS

From the Department of Dermatology, University of Connecticut Health Center, Farmington.
The authors report no conflict of interest.
Correspondence: Katalin Ferenczi, MD, University of Connecticut Health Center, Department of Dermatology, 21 South Rd, Farmington, CT 06032 (ferenczi@uchc.edu).
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THE **DIAGNOSIS:** Diffuse Dermal Angiomatosis

Diffuse dermal angiomatosis (DDA) is an acquired reactive vascular proliferation in the spectrum of cutaneous reactive angioendotheliomatoses. Clinically, DDA presents as violaceous reticulated plaques, often with secondary ulceration and sometimes necrosis.[1-3] Diffuse dermal angiomatosis more commonly presents in patients with a history of severe peripheral vascular disease, coagulopathies, or infection, and it frequently arises on the extremities. Diffuse dermal angiomatosis also has been shown to develop on the breasts, particularly in patients with pendulous breast tissue. Vascular proliferation in DDA is hypothesized to be from ischemia and hypoxia, leading to angiogenesis.[1-3] Diffuse dermal angiomatosis is characterized histologically by the presence of a diffuse proliferation of spindled endothelial cells distributed between the collagen bundles throughout the dermis (quiz image and Figure 1). Spindle-shaped endothelial cells exhibit a vacuolated cytoplasm. On immunohistochemistry, these dermal spindle cells classically stain positive for CD31, CD34, and erythroblast transformation specific–related gene (Erg) and stain negative for both human herpesvirus 8 (HHV-8) and factor XIIIa.

Cutaneous fibrous histiocytoma, more commonly referred to as dermatofibroma, is a common benign lesion that presents clinically as a solitary firm nodule most commonly on the extremities in areas of repetitive trauma or pressure. It classically exhibits dimpling of the overlaying skin with lateral pressure on the lesion, known as the dimple sign.[4] Histologically, dermatofibromas share similar features to DDA and demonstrate the presence of bland-appearing spindle cells within the dermis between the collagen bundles, resulting in collagen trapping. However, a distinguishing histologic feature of a dermatofibroma in comparison to DDA is the presence of epidermal hyperplasia overlying the dermatofibroma, leading to tabled rete ridges (Figure 2). Spindle cells in dermatofibromas are fibroblasts and have a distinct immunophenotype that includes factor XIIIa positivity and negative staining for CD31, CD34, and Erg.[4,5]

Dermatofibrosarcoma protuberans (DFSP) is a rare malignant soft-tissue sarcoma that clinically presents as a firm, flesh-colored, dermal plaque on the trunk, proximal extremities, head, or neck.[5] Histologically, DFSP can be distinguished from DDA by the high density of spindle cells that are arranged in a storiform pattern, extending

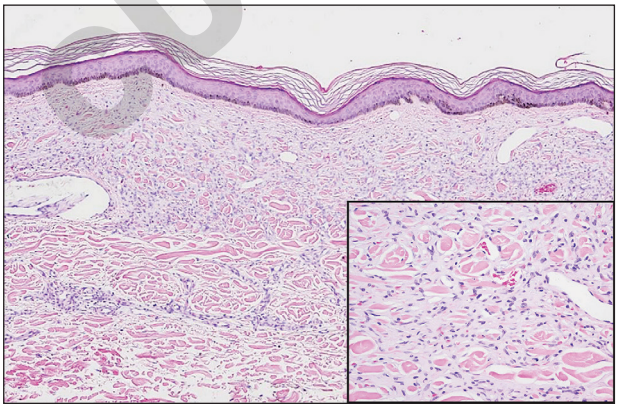

FIGURE 1. Diffuse dermal angiomatosis. A broad bandlike proliferation of spindle cells in the papillary and upper reticular dermis with vacuolated cytoplasm and enhanced collagen deposition (H&E, original magnification ×100 [inset: H&E, original magnification ×400]).

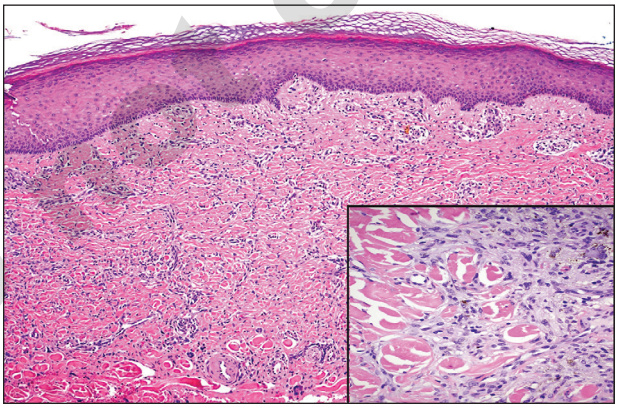

FIGURE 2. Dermatofibroma. Epidermal hyperplasia with tabled rete ridges overlying a bland-appearing spindle cell proliferation within the papillary and reticular dermis and collagen trapping (H&E, original magnification ×100 [inset: H&E, original magnification ×400]).

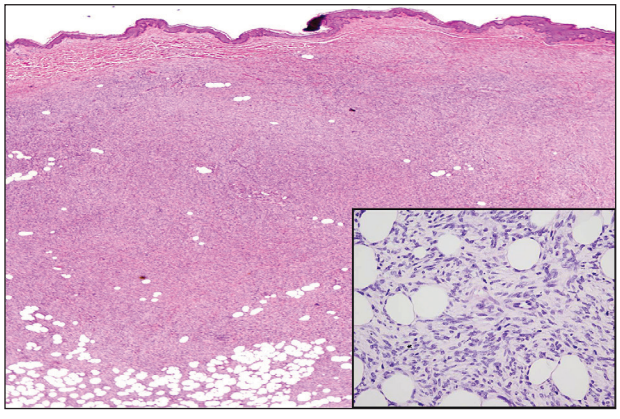

FIGURE 3. Dermatofibrosarcoma protuberans. A dense and highly cellular dermis with spindle cells arranged in a storiform pattern that extend and infiltrate the subcutaneous fat in a honeycomblike pattern (H&E, original magnification ×100 [inset: H&E, original magnification ×400]).

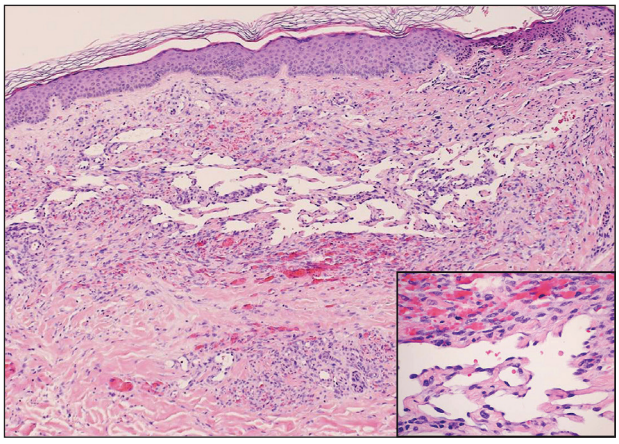

FIGURE 4. Kaposi sarcoma. A proliferation of spindle cells within the dermis, extravasated erythrocytes, and vessel formation around preexisting vessels (known as the promontory sign) (H&E, original magnification ×100 [inset: H&E, original magnification ×400]).

and infiltrating the underlying subcutaneous fat in a honeycomblike pattern (Figure 3). Spindle cells in DFSP typically show expression of CD34 but are negative for CD31, Erg, and factor XIIIa.[5]

Kaposi sarcoma (KS) is an endothelial cell–driven angioproliferative neoplasm that is associated with HHV-8 infection.[6] The clinical presentation of KS can range from isolated pink or purple papules and patches to more extensive ulcerated plaques or nodules. Histopathology exhibits proliferation of monomorphic spindled endothelial cells within the dermis staining positive for HHV-8, Erg, CD31, and CD34, in conjunction with extravasated erythrocytes arranged within slitlike vascular spaces (Figure 4). Additionally, KS classically exhibits aberrant endothelial cell proliferation and vessel formation around preexisting vessels, which is referred to as the promontory sign (Figure 4).

Angiosarcoma is a rare and highly aggressive vascular tumor arising from endothelial cells lining the blood vessels and lymphatics.[7,8] Clinically, angiosarcoma presents as ulcerated violaceous nodules or plaques on the head, neck, or trunk. Histologic evaluation of angiosarcoma reveals a complex and poorly demarcated vascular network dissecting between collagen bundles in the dermis (Figure 5). Multilayering of endothelial cells, papillary projections extending into the vessel lumina, and mitoses frequently are seen. On immunohistochemistry, endothelial cells demonstrate prominent cellular atypia and stain positive with CD31, CD34, and Erg.

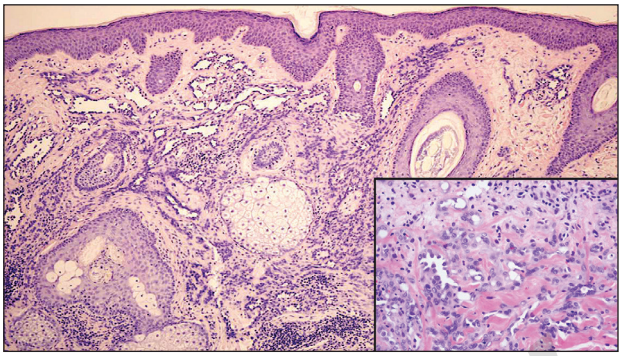

FIGURE 5. Angiosarcoma. Dissecting vascular spaces and papillary projections into the lumina with endothelial cells showing prominent cellular atypia (H&E, original magnification ×100 [inset: H&E, original magnification ×400]).

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