Painful Violaceous Nodule With Peripheral Hyperpigmentation

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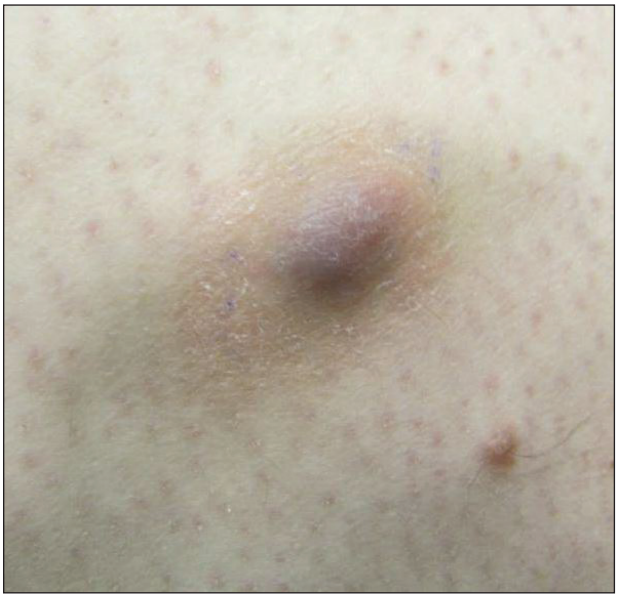

A 30-year-old man presented for evaluation of a painful lesion on the left thigh of 3 to 4 years' duration. Pain was exacerbated on physical exertion and was relieved by application of ice packs and use of over-the-counter analgesics. The patient denied any bleeding from the lesion. No other medical comorbidities were present. Physical examination demonstrated a pink, scaly, 3×2-cm patch with peripheral hyperpigmentation overlying a central, moderately firm, violaceous, 10×15-mm nodule on the left anteromedial thigh. The lesion was excised and sent to pathology.

WHAT'S THE **DIAGNOSIS?**

a. aneurysmal dermatofibroma
b. Kaposi sarcoma
c. malignant melanoma
d. pilomatricoma
e. pyogenic granuloma

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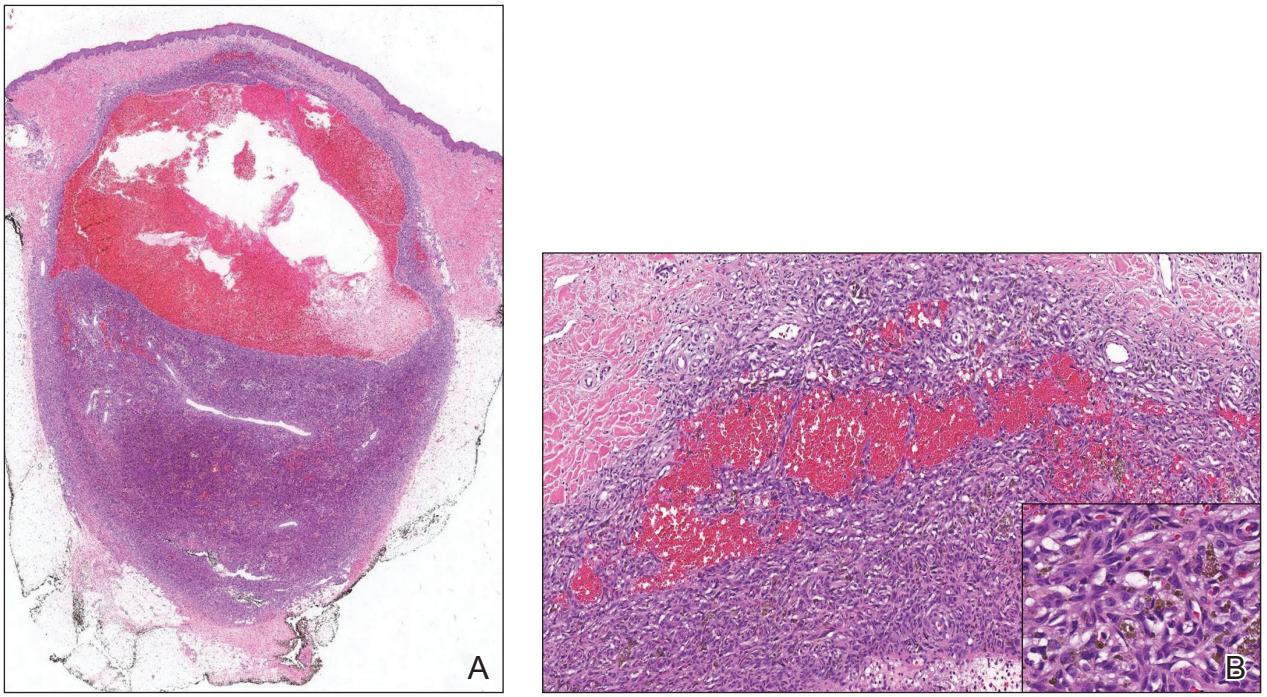
THE **DIAGNOSIS:** Aneurysmal Dermatofibroma

Biopsy of the lesion revealed a circumscribed dermal nodule comprised of storiform arrangements of enlarged, plump, fibrohistiocytic cells punctuated by variably sized clefts and large cystic spaces filled with blood that lacked an endothelial lining. No bizarre nuclear pleomorphism, atypical mitoses, or tumor necrosis were identified. The overlying epidermis exhibited mild acanthosis with broadening of the rete ridges. Proliferative spindled cells entrapped dermal collagen bundles at the periphery. Hemosiderin-laden macrophages were present throughout the proliferation and in the adjacent dermis (Figure). These findings supported the diagnosis of aneurysmal dermatofibroma (ADF).

Aneurysmal dermatofibroma, also known as aneurysmal fibrous histiocytoma, is a rare variant of dermatofibroma that was described by Santa Cruz et al[1] in 1981 and represents 2% to 6% of dermatofibromas.[1,2] Aneurysmal dermatofibromas often lack the characteristic clinical and dermoscopic findings of conventional dermatofibromas, creating a diagnostic challenge for the clinician.[3] Incomplete excision of this benign tumor was associated with a local recurrence rate of 19% (5/26) in one study,[4] in contrast with the exceedingly low rate of local recurrence (<2%) attributed to conventional dermatofibromas.[2,4]

Clinically, ADFs commonly appear as blue-brown nodules on the arms and legs, often with a history of rapid and sometimes painful growth.[1] Clinically, an ADF can have vascular, cystic, or melanocytic features that, in the context of lacking typical clinical findings of a dermatofibroma, can complicate clinical diagnosis; for example, ADFs can demonstrate several melanomalike features including atypical vessels, chrysalis structures, blue-white structures, a pinkish-white veil, irregular brown globulelike structures, an atypical pigment network, color variegation, a multicomponent pattern, and ulceration.[3] Alternatively, ADFs can present with a vascular tumor–like pattern consisting of white areas and globular blue-red areas or a polymorphous vascular pattern with a peripheral collarette.

Our case illustrates the classic histologic appearance of an ADF. Large cavities and slitlike spaces filled with blood distinguish this entity from conventional dermatofibroma and other dermatofibroma variants; for example, cellular dermatofibroma is a benign variant of



Low-power image showing the classic appearance of aneurysmal dermatofibroma with a characteristic cystic space filled with blood (A) (H&E, original magnification ×10). Storiform arrangement of fibrohistiocytic cells surrounding blood-filled clefts that lack an endothelial lining (B)(H&E, original magnification ×100). Note the dermal collagen trapping at the periphery of the nodule. Higher magnification highlights the plump fibrohistiocytic cells and hemosiderin-laden macrophages (inset; H&E, original magnification ×400).

dermatofibroma that exhibits crowded fascicular architecture without an increase in vascular spaces. Aneurysmal dermatofibromas also should be distinguished from angiomatoid fibrous histiocytoma, which has intermediate malignant potential despite a similar-sounding name and a similar nodular appearance with large blood-filled spaces; however, many cases are located predominantly in the subcutis with epithelioid morphology, desmin immunohistochemical reactivity, and prominent tumor-associated lymphoid proliferation that can be mistaken for a lymph node.[5] Furthermore, in contrast with vascular tumors, the blood-filled spaces of ADFs do not have an endothelial lining.

In summary, ADF is a rare dermatofibroma variant that has a variety of clinical presentations, often masquerading as a cyst, vascular tumor, or melanocytic neoplasm. The classic histopathologic features confirm the diagnosis. Although ADFs can be painful and have a tendency to recur, these lesions have a benign clinical course.

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