

An Asymptomatic Slate-brown Papule on the Left Calf in a 56-year-old-man

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CASE REPORT

A 56-year-old man had noticed an asymptomatic slate-brown papule on the left calf for 4 years. The lesion underwent a gradual growth with recent rapid enlargement; no previous trauma history was noted. He was otherwise in good health. The lesion was surgically excised under local anesthesia.

An excisional biopsy from the left calf was stained with hematoxylin-eosin (Fig. 1, 2). Immunohistochemically, the tumor cells stained negatively with desmin and factor VIII RA. The iron stain shows hemosiderin dispersed within the tumor.

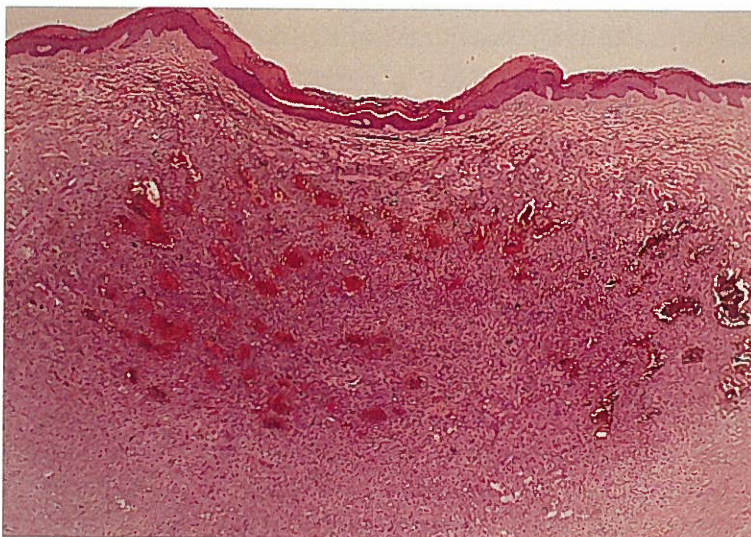


Fig. 1
(H & E stain, x40,)

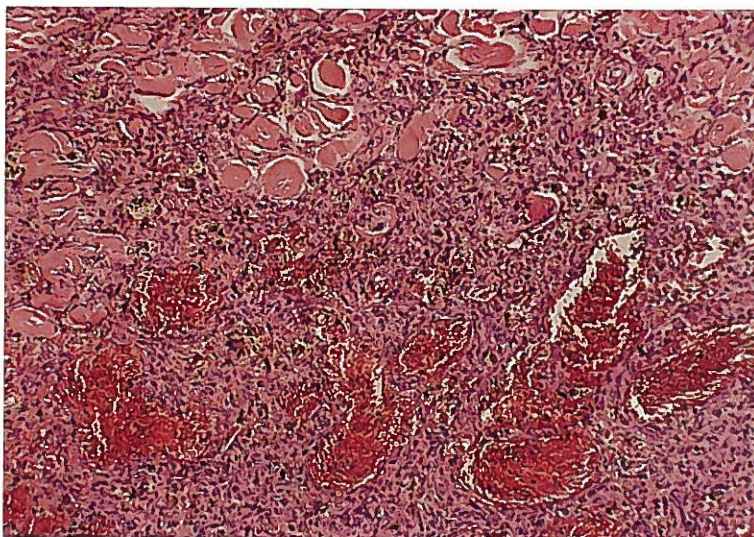


Fig. 2
(H & E stain, x400)

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DIAGNOSIS:*Aneurysmal Fibrous Histiocytoma (AFH)*

DISCUSSION

In 1966, Hairston and Reed¹ first reported 3 cases of "aneurysmal sclerosing hemangioma" of the skin. They observed pseudovascular spaces in area of classic sclerosing hemangioma. In 1979, Enzinger² described 41 cases under the named "angiomatoid malignant fibrous histiocytoma", characterized by a significant trauma history and high risk of recurrence and metastasis. In 1981, Santa Cruz and Kyriakos³ first reported 17 cases of "aneurysmal (angiomatoid) fibrous histiocytoma (AFH)" and considered it as a benign counterpart of Angiomatoid malignant fibrous histiocytoma.

AFH represents only 1.7% of all fibrous histiocytomas.⁴ Clinically, they are similar to ordinary fibrous histiocytoma, as a solitary nodule at many anatomical sites over a wide age range. However, they are most common in the lower limb/limb girdle(50%) of middle-age adults with slight predilection for females. Due to rapid growth, spontaneous bleeding, pigmentation and a larger average size, clinical diagnosis is frequently mistaken for a cyst, melanocytic tumor or hemangioma. Most cases lack definite trauma history. It is often asymptomatic, but sudden increase in size, change in color, spontaneous hemorrhage, pain or tenderness is also complained.⁵

Histopathologically, the tumors are relatively well demarcated. Epidermal changes, ranging from mild to prominent acanthosis, are present in 88% of cases.⁴ The formation of large blood-filled pseudovascular spaces and storiform proliferation of tumor cells in capillary-rich stroma are rather characteristic. Three types of tumor cells are noted---histiocyte-like cells, fibroblast-like cells, and intermediate cells that display combined features of the former two. The majority of the tumor cells belong to the intermediate cells. The variably sized, blood-filled spaces are not lined by endothelial cells but by tumor cells. The slow continuous extravasation of blood from capillaries in highly

cellular areas results from a poorly developed reticulin network.³ Intra-and extracellular deposition of hemosiderin is prominent. All tumors are highly polymorphic; cytological atypia was generally absent or mild. Mitotic activity ranges from one to ten per ten high power fields; however, atypical mitotic figures are never seen.⁴ Immunohistochemically, the tumor cells are positive for vimentin, factor XIIIa, Mac 387 and Co-Mo-2 and give negative results for factor VIII-RA, desmin, actin and S-100, supporting the fibrohistiocytic origin.^{4,5}

AFH should be completely excised due to its high local recurrence rate (19%), which is much higher than that of ordinary fibrous histiocytoma (<2%). Local extension to adjacent lymph node in a recurrent tumor was reported; nevertheless, true metastasis was not discovered.⁴

We present this rather uncommon case to remind that AFH should be considered as one of the differential diagnoses of cutaneous vascular tumors to prevent misdiagnosis as aggressive conditions.

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