Aneurysmal Fibrous Histiocytoma

Bahram Azadeh,
Agustin Chong Lopez,
Shaikha Al-Thani,
Department of Pathology and Laboratory Medicine
Hamad Medical Corporation, Doha, State of Qatar

Abstract

A 45-year-old Indian man had a painful, slow growing swelling on the anterior aspect of the right thigh, clinically assumed to be a sebaceous cyst, associated with varicose veins and venous oedema of the right leg. The excised mass (25x23x15 mm) consisted of a large blood-filled space (20mm) devoid of endothelial cells surrounded by a wall (2 to 6 mm) with features of a fibrous histiocytoma. Tumor cells were positive for vimentin, macrophages were positive for MAC375 and focally for S100. Scattered T-lymphocytes present were CD3 positive, CD20 negative. The cyst lining was negative for factor VIIIRAg and CD34, Staining for actin, desmin, cytokeratin, Ki67 and PCNA was negative. Aneurysmal fibrous histiocytoma is a rare variant of cutaneous fibrous histiocytoma characterized by large blood-filled spaces devoid of endothelial cells. Because of the large size and rapid growth secondary to hemorrhage it may be clinically confused with cysts, benign and malignant tumors.

Introduction

Aneurysmal fibrous histiocytoma is a rare variant of cutaneous fibrous histiocytoma, which contains large blood-filled spaces. Clinically, they are large, blue, black, or dark red, cystic tumors which may be associated with symptoms of pain and rapid growth. Clinico-pathological features of aneurysmal fibrous histiocytomas are so different from the common cutaneous fibrous histiocytomas that malignancy may be considered in some cases.

In the original description of 17 cases reported by Santa Cruz & Kyriakos,1981; similar cases that they quote from earlier literature, and in more recent series, aneurysmal fibrous histiocytoma has been clinically confused with haemangioma, neurofibroma, nonspecific cysts, gumma, malignant melanomas, and sarcomas. 1-4

The clinical diagnoses considered in the present case report was neurofibroma or sebaceous cyst.

Case Report

A 45-year-old Indian man presented on 18.7.98 with a painful, slow growing swelling on the anterior aspect of the right thigh, of one-year duration, associated with varicose veins (since1991) and venous oedema of the right leg. He had a past history of a hair-line fracture of the right calcaneum after falling (29.9.97), a left varicocele found on investigation of secondary infertility (19.1.95), and a herniated L5-S1 paravertebral disc confirmed by CT Scan. The mass, which was assumed to be a sebaceous cyst or neurofibroma, was excised. On cut-section, the specimen (25x23x15 mm) consisted of a large blood-filled space 20 mm in diameter with a wall 2 to 6 mm in thickness (Fig 1), histologically located in deep dermis and subcutis (Fig. 2-4). There were no endothelial cells in the cyst lining (Fig. 2, 3 & 8). Spindle cells in a storiform pattern with scattered lymphocytic infiltrates were evident in the solid part of the tumor (Fig 3-5). Abundant foamy and siderotic macrophages (Fig. 6) in the cyst lining and in the wall, were associated with hemosiderin depositions (Fig.7), new and old collagen fiers (Fig. 8). There were no lymphoid follicles, cytological atypia or mitotic figures. Immunohistochemistry (ABC method) revealed vimentin staining in the tumor cells. Macrophages were positive for MAC375 and focally for S100 protein. Negative staining for factor VIIIRAg and CD34 confirmed the absence of endothelial cells in the cyst lining. Most of the scattered lymphocytes present were T-cells (CD3 positive, CD20 negative). Staining for actin, desmin, cytokeratin, Ki67 and PCNA was negative.

Discussion

Aneurysmal fibrous histiocytoma differs from the common fibrous histiocytoma by a larger size, pigmentation and rapid growth due to intralesional hemorrhage. In some cases of cutaneous fibrous histiocytomas, slow extravasation of blood from capillaries is believed to cause slit-like tissue cracks which continue to enlarge under pressure to form finally the cavernous or angiomatoid areas devoid of endothelial linings characteristic of aneurysmal fibrous

Correspondence:
Bahram Azadeh MD FRCPath,
Chairman
Pathology and Laboratory Medicine,
Hamad Medical Corporation, PO Box 3050,
Doha, State of Qatar.
Tel: +974-392591
Fax: +974-392183

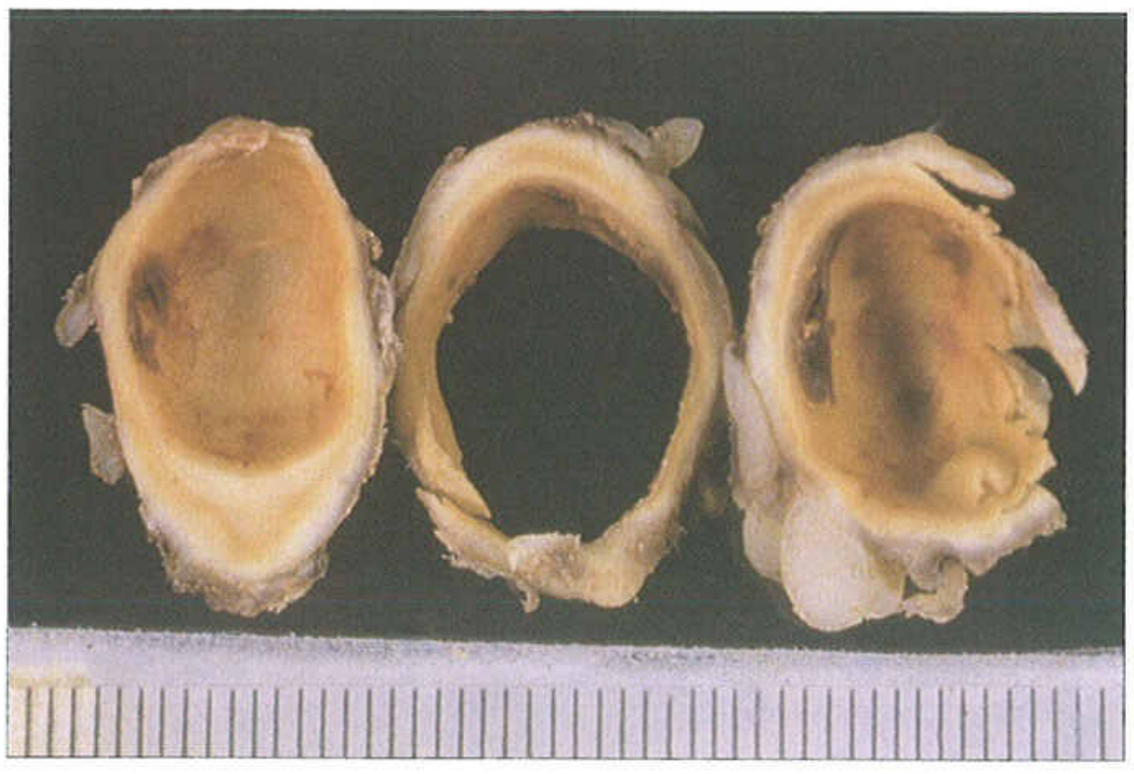

Fig.1. Aneurysmal Fibrous Histiocytoma. Blood clots removed from the cystic space to reveal a yellowish-tan, smooth lining.

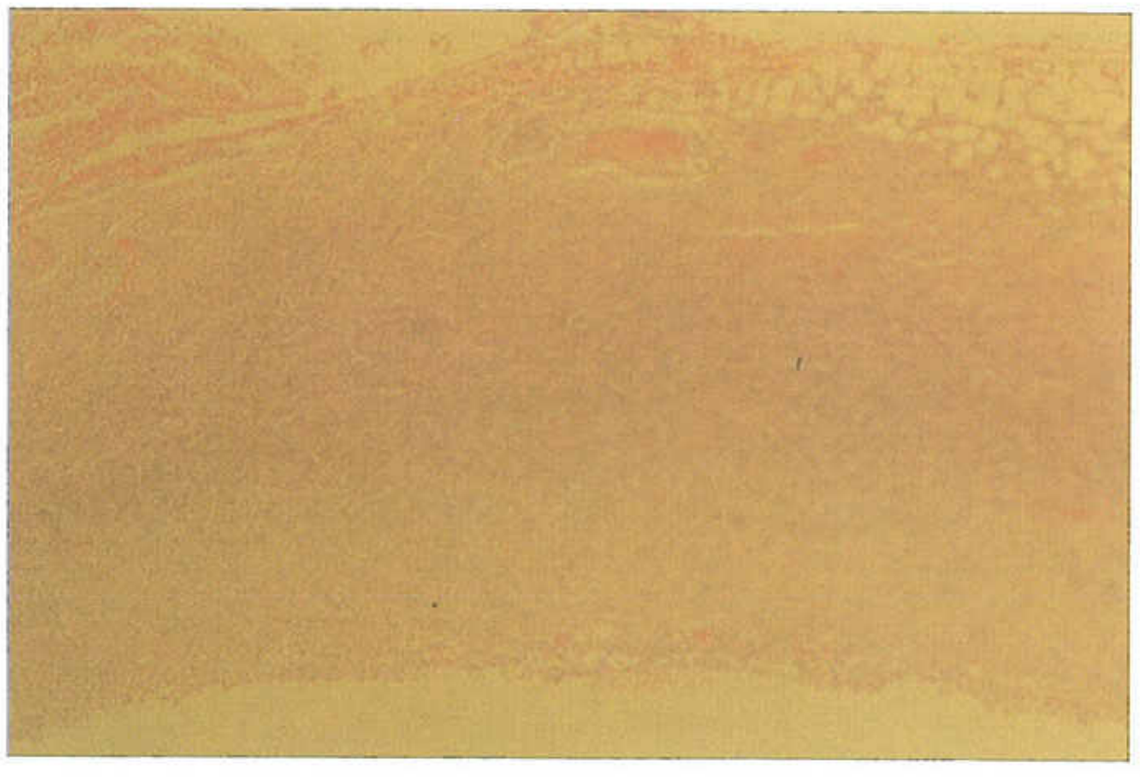

Fig. 2. The cyst wall, consisting of compact connective tissue fibers and cellular elements, has extended to subcutaneous adipose tissue (H&E).

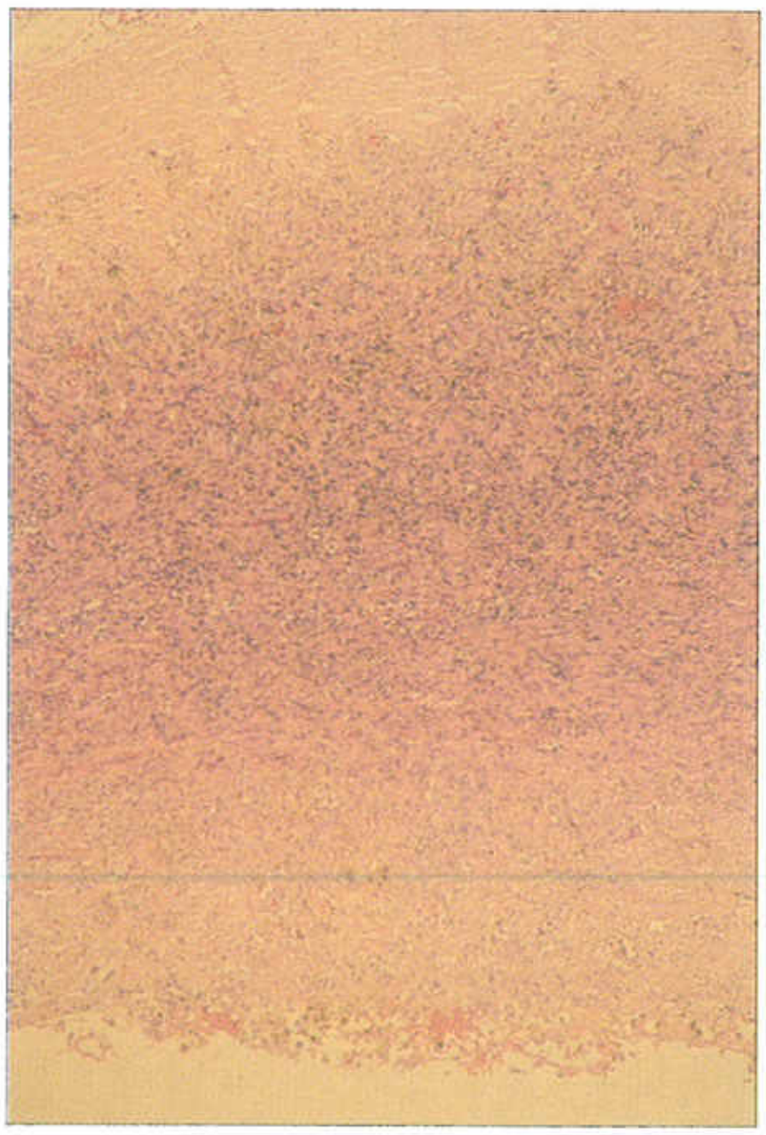

Fig. 3. There is no endothelial cell in the lining of the cystic space (H&E).

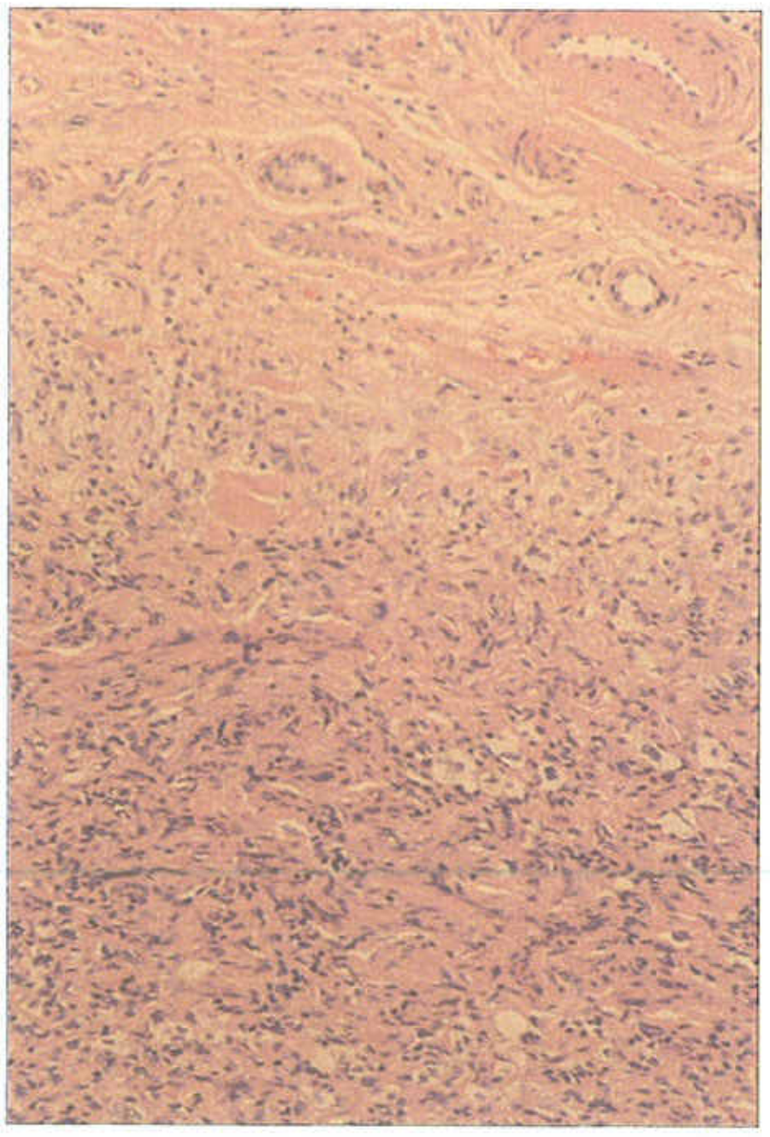

Fig. 4. Histiocytes and spindle cells in a somewhat storiform pattern have infiltrated the dermis to the level of sweat glands (H&E).

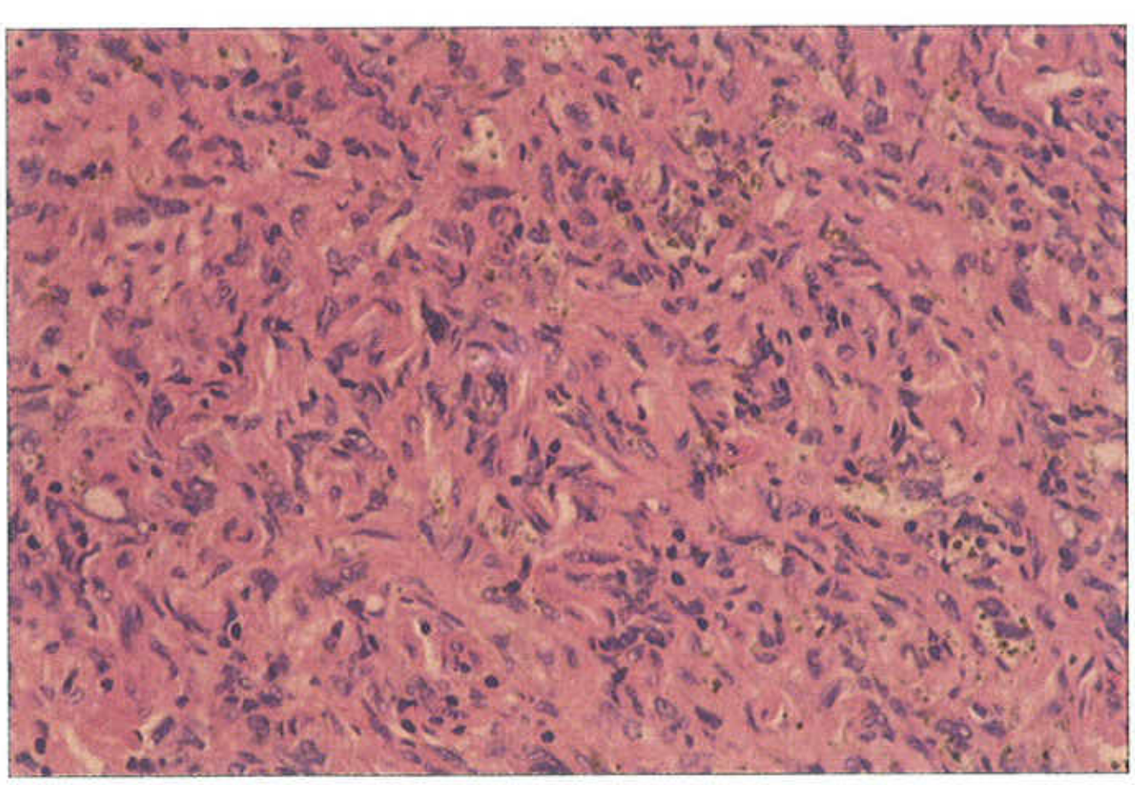

Fig. 5. Higher power of spindle cells in a storiform pattern (H&E).

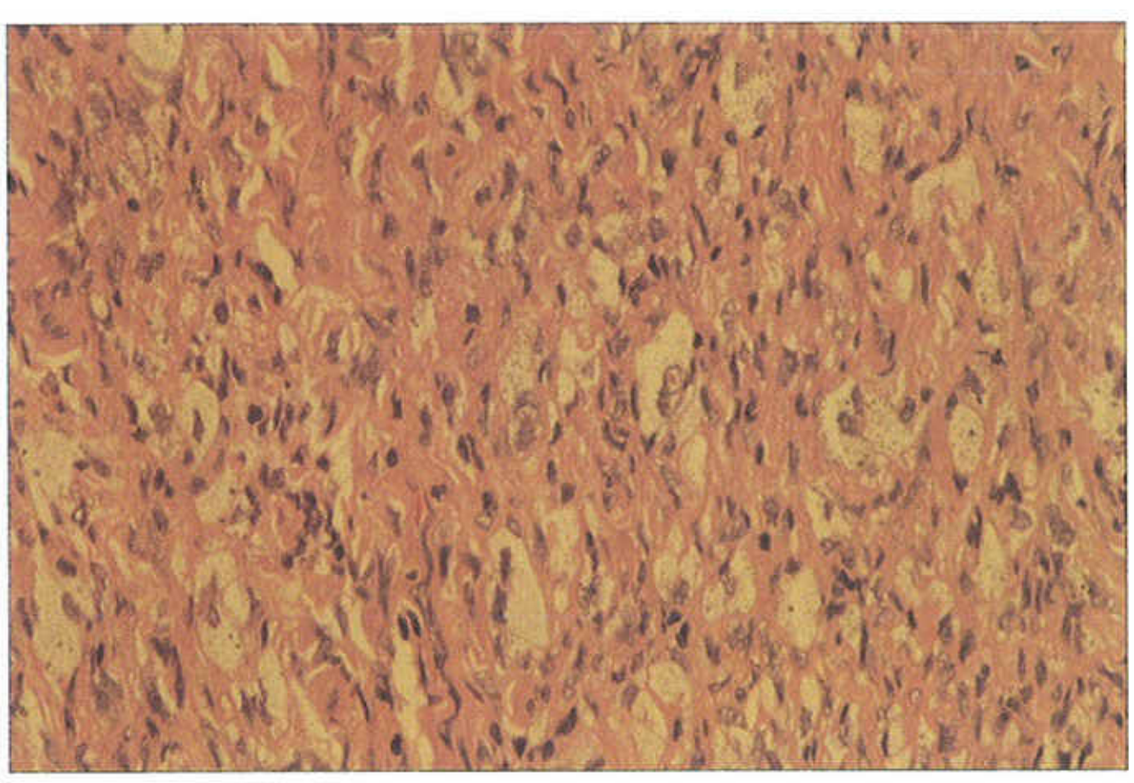

Fig. 6. The solid part of the tumor shows numerous foamy macrophages along with the usual features of a fibrous histiocytoma (H&E).

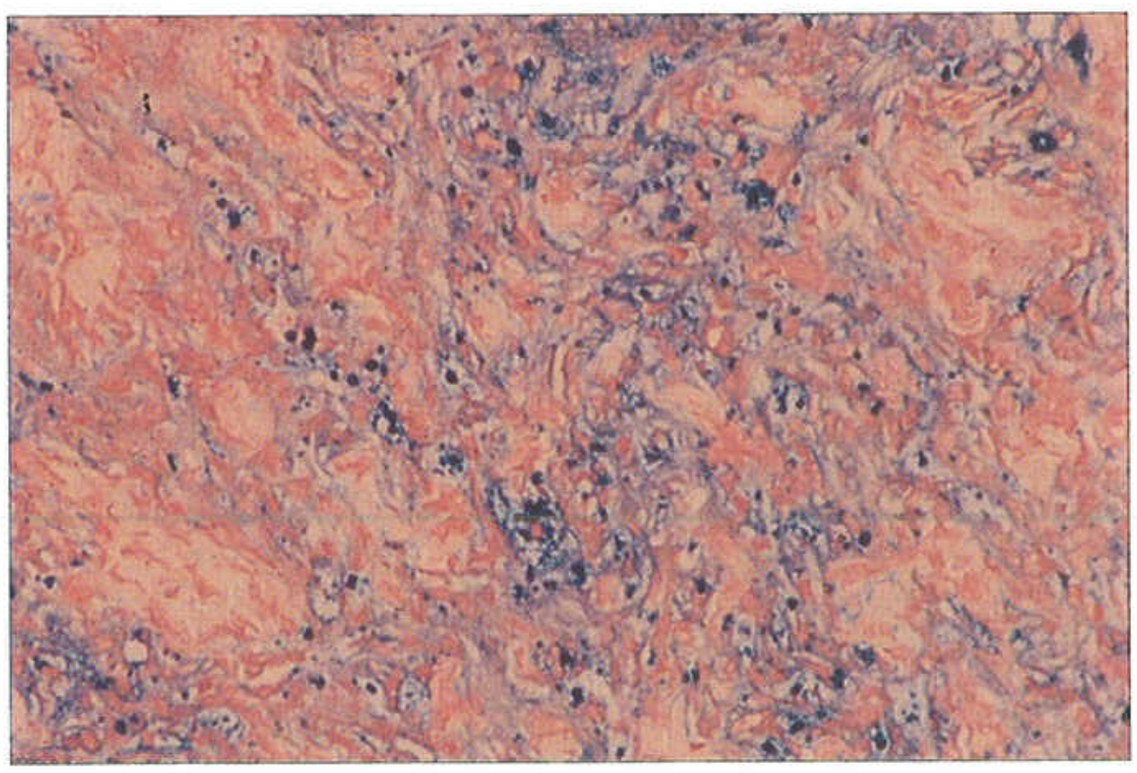

Fig. 7. Bluish deposits of hemosiderin pigments in the wall (Perlsí).

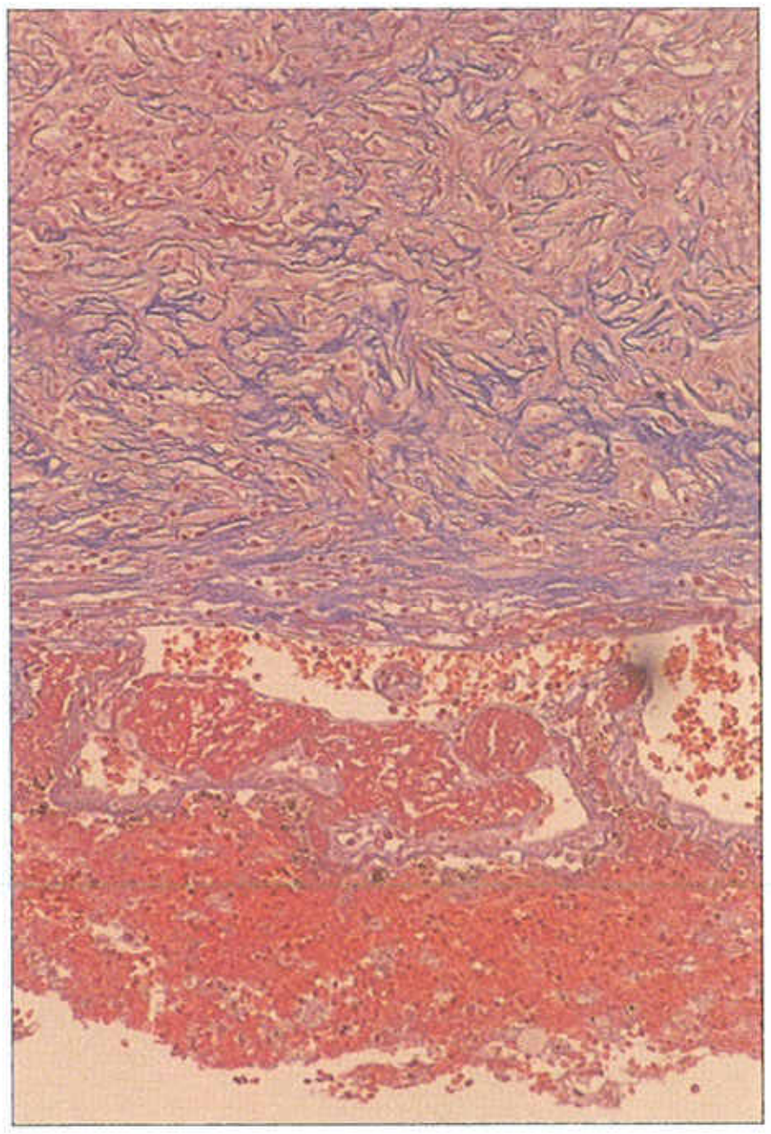

Fig. 8. Old and new collagen fibers are present in the solid part of the tumor. The lining of the aneurysmal component is devoid of endothelial cells and is covered by blood clot (Trichrome).

histiocytomas [1]. The solid portions of the tumor have the usual features of a fibrous histiocytoma. Histologically, aneurysmal fibrous histiocytomas may be misdiagnosed as vascular tumors including hemangioma, spindle cell hemangioendothelioma, cutaneous angiosarcoma and Kaposi sarcoma [1-4]. The most important tumor to be distinguished from aneurysmal fibrous histiocytoma is angiomatoid malignant fibrous histiocytoma, a rare soft tissue sarcoma, usually subcutaneous in the extremities of young individuals with median age of 13 years [5]. Local recurrence is common in angiomatoid malignant fibrous histiocytoma, but only about 20% of cases have been said to metastasize and this is not always associated with a fatal outcome [6]. Aneurysmal fibrous histiocytoma occurs in middle-age adults and lacks the cellular pleomorphism and the prominent lymphoplasmacytic infiltrates of angiomatoid malignant fibrous histiocytoma. A local recurrence rate of 19% reported in one series of aneurysmal fibrous histiocytoma is possibly due to incomplete excision of the often larger aneurysmal lesions [3]. A documented history of trauma, as in our patient, has been reported in a 12-year-old girl with aneurysmal fibrous histiocytoma involving subcutaneous tissue of the scalp [7].

References

1. Santa Cruz DJ, Kyriakos M. Aneurysmal (ìAngiomatoidî) Fibrous Histiocytoma of the Skin. Cancer 1981; 47: 2053-2061.

2. Sood U, Mehregan AH. Aneurysmal (angiomatoid) fibrous histiocytoma. J Cutan Pathol 1985; 12: 157-162.

3. Calonje E, Fletcher CDM. Aneurysmal benign fibrous histiocytoma: clinicopathological analysis of 40 cases of a tumor frequently misdiagnosed as a vascular neoplasm. Histopathology 1995; 26: 323-331.

4. Zelger BW, Zelger BG, Steiner H, Ofner D. Aneurysmal and haemangiopericytoma-like f ibrous histiocytoma. J Clin Pathol 1996; 49: 313-318.

5. Enzinger FM. Angiomatoid malignant fibrous histiocytoma. A distinct fibrohistiocytic tumor of children and young adults simulating a vascular neoplasm. Cancer 1979; 44:2147-2157.

6. Fletcher CDM. Angiomatoid ìmalignant fibrous histiocytomaî: An immunohistochemical study indicative of myoid differentiation. Hum Pathol 1991; 22: 563-568.

7. Kim CJ, Cho JH, Chi JG. Aneurysmal (ìangiomatoidî) fibrous histiocytoma in a child. J Korean Med Sci 1989; 4: 159-162.