Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor with intermediate malignant potential, most commonly occurring in children, adolescents, and young adults. It accounts for approximately 0.3% of soft tissue neoplasms. The majority of cases occur in the extremities, are slow-growing, and are typically painless. We report a case of AFH on the scalp of a 40-year-old man, locally recurring within two years of the original operation. AFH is a rare condition with the potential of local recurrence and metastasis. It should be considered in the differential diagnosis of a soft tissue mass in a child or adolescent.

**INTRODUCTION**

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor with intermediate malignant potential, most commonly occurring in children, adolescents, and young adults. It accounts for approximately 0.3% of soft tissue neoplasms. The lesion is rare and has the potential for local recurrence and metastasis, so it is imperative to consider this lesion in the differential diagnosis of a soft tissue mass in a child or adolescent.\(^1\)

**CASE REPORT**

A 40-year-old man presented with the complaints of swelling on the right parieto-temporal region of the scalp since 2 years. The swelling was 2 cm × 2 cm, firm, non-tender and mobile. The patient was previously operated for a swelling at the same site 2 years back which had recurred. Excision biopsy revealed a skin covered grayish-white mass with a central hemorrhagic cystic cavity of 1.2 cm × 0.7 cm size [Figure 1]. Histopathology revealed a well-circumscribed (with pseudo-capsule) dermal tumor [Figure 2]. It had a central hemorrhagic cavity without endothelial lining, surrounded by tumor tissue composed of spindle cells arranged in short fascicles and storiform pattern, admixed with hemosiderin-laden macrophages [Figures 3-5]. At places, giant cells [Figure 6] and anastomosing vascular channels admixed with tumor and inflammatory cells were also seen.

**DISCUSSION**

AFH is a rare soft tissue neoplasm first described by Enzinger in 1979.\(^2\) It commonly arises over sites of normal lymphoid tissue such as the antecubital fossa, axilla, inguinal, and supraclavicular regions.\(^3\) Majority of the cases occur in the extremities although lesions occurring in the head and neck region (10%) and trunk have been reported.\(^2,4-6\)

Pathologically, AFH demonstrates four features: A fibrous pseudocapsule, a round or spindle cell fibrohistiocytic...
Swami, et al. Angiomatoid fibrous histiocytoma

Figure 1: Gross: 4 cm x 2 cm, grayish white skin covered tissue with area of hemorrhage

Figure 2: Microphotograph showing well-circumscribed tumor tissue in the dermis (H and E, x10)

Figure 3: Microphotograph showing hemorrhagic area surrounded by tumor tissue (H and E, x10)

Figure 4: Microphotograph showing spindle cells arranged in irregular and storiform fashion around hemorrhagic areas without endothelial lining (H and E, x40)

cell proliferation, a pseudoangiomatous pattern and a plasma-lymphocytic infiltrate. Immunohistochemistry show variable positivity for EMA, desmin, CD68 and CD99, whereas S100 protein and cytokeratin are negative.\(^7\)

WHO in 2002 removed AFH as a subtype of malignant sarcoma and placed it in the category of tumors of uncertain differentiation.\(^3\) The cumulative findings of a meta-analysis of multiple studies demonstrate that the majority of patients (73.2%) were disease free after local excision, while minority (23.2%) developed recurrent disease and 8.7% developed metastatic disease within 24 months post operation.\(^4\) Since AFHs are rare, it can lead to an erroneous diagnosis of a malignant disease.

Nodular Kaposi sarcoma is a possible mimic, with its circumscription, dermal location, bland spindle cell morphology and in particular, prominent vascular spaces. The pseudoangiomatoid spaces in AFH however, lack an endothelial lining, instead they are lined by flattened tumoral cells. This feature and the absence of CD34 and HHV8 should make distinction apparent.

Aneurysmal benign fibrous histiocytoma has a more polymorphous cellular pattern including giant cells and siderophages, retains the typical features of a dermatofibroma such as epidermal hyperplasia and peripheral collagen bundles and is desmin negative.

Metastatic tumor to the lymph node (including metastasizing cellular fibrous histiocytoma) may enter the differential diagnosis but closer inspection of AFH reveals that germinal centers are randomly arranged around the tumor, and true nodal architecture such as subcapsular and medullary sinuses is absent.
Palisaded myofibroblastoma shows delicate palisading spindle cells and thick bands or bundles of collagen fibers (“amianthoid fibers”) and is desmin negative.

Inflammatory pseudotumor may be considered in the differential diagnosis as it is occasionally seen to express desmin but lacks the pseudovascular spaces of AFH.

Spindle cell hemangioma typically occurs in the dermis and subcutis of the distal extremities. It contains cavernous vascular spaces, but in contrast to AFH, these are lined by an attenuated layer of endothelial cells. It is also poorly circumscribed and often contains epithelioid cells, many of which contain intracytoplasmic vacuoles.

The bland ovoid- to spindle cells of AFH can sometimes also be mistaken for granulomas although the more solid nature of the tumor and blood-filled spaces should make distinction possible. Desmin positivity can be seen in tenosynovial giant cell tumors, and AFH may contain giant cells, although this should not be a source of confusion, as the morphology of giant cell tumors, with their heterogeneous cellular composition including foamy macrophages, is characteristic.

In this tumor, it is important to perform the appropriate surgical excision to obtain negative surgical margins and to continue close post-operative surveillance with clinical examination to ensure that no signs or clinical evidence of tumor recurrence occurs.

**Financial support and sponsorship**
Nil.

**Conflicts of interest**
There are no conflicts of interest.

**REFERENCES**

1. Bauer A, Jackson B, Marner E, Gilbertson-Dahdal D. Angiomatoid fibrous histiocytoma: A case report and review of the literature. J Radiol Case Rep 2012;6:8-15.
2. Enzinger FM. Angiomatoid malignant fibrous histiocytoma: A distinct fibrohistiocytic tumor of children and young adults simulating a vascular neoplasm. Cancer 1979;44:2147-57.
3. Fletcher CD. The evolving classification of soft tissue tumors: An update based on the new WHO classification. Histopathology 2006;48:3-12.
4. Costa MJ, Weiss SW. Angiomatoid malignant fibrous histiocytoma. A follow-up study of 108 cases with evaluation of possible histologic predictors of outcome. Am J Surg Pathol 1990;14:1126-32.
5. Santa Cruz DJ, Kyriakos M. Aneurysmal (“angiomatoid”) fibrous histiocytoma of the skin. Cancer 1981;47:2053-61.
6. Regezi JA, Zarbo RJ, Tomich CE, Lloyd RV, Courtney RM, Crissman JD. Immunoprofile of benign and malignant fibrohistiocytic tumors. J Oral Pathol 1987;16:260-5.
7. Fanburg-Smith JC, Miettinen M. Angiomatoid “malignant” fibrous histiocytoma: A clinicopathologic study of 158 cases and further exploration of the myoid phenotype. Hum Pathol 1999;30:1336-43.