Angiomatoid Fibrous Histiocytoma in an Elderly Male - An Unusual Presentation of a Rare Case

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Abstract:

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue neoplasm of low malignant potential which is often misdiagnosed clinically. It typically occurs in the superficial soft tissues of the extremities in children and young adults. It is characterised by recurrences and rarely metastases. Surgery remains the mainstay of the management. Here, we present a rare case report of AFH in a 65-year-old male who was diagnosed on fine needle aspiration as spindle cell sarcoma. The patient underwent wide local excision. The patient is under follow up. There is no evidence of metastases and patient is disease free till 3 years after excision.

Keywords: Angiomatoid fibrous histiocytoma, soft tissue tumour, low malignant potential, recurrence

Introduction:

Angiomatoid malignant fibrous histiocytoma (AFH) was first described by Enzinger in 1979.[1] It is a rare soft tissue tumor characterised by low malignant potential, and it rarely metastasizes. It accounts for 0.3% of all soft tissue sarcomas and is usually found in the deep dermis and subcutaneous tissues of extremities of children and young adults.[2] A few cases of AFH has been described in adults in literature.[3,4] AFH has diverse clinicopathological features and is also associated with certain genetic attributes.[2] Because of its varied clinical presentation, it is difficult to establish preoperative diagnosis of AFH. Here, we report a case of AFH in an elderly male patient.

Case Report:

A 65-year-old man presented in a surgery outpatient department at a rural tertiary care hospital in central India with complaints of swelling over the right leg for 2 months. It was insidious in onset and was gradually progressive in nature. The patient also gave the history of trauma to the right leg at the same site 3 months back. There was no history of weight loss or decreased appetite. Physical examination was unremarkable. Local examination revealed a swelling measuring 7x4 cm present over the anterolateral aspect of the right upper leg without tenderness.

Magnetic resonance imaging right leg revealed a well-defined lesion of size 14.5x5.8x5.4 cm in an anterior intramuscular compartment in the proximal part of right leg involving the tibialis anterior and right extensor digitorum longus muscles with multiple dilated and tortuous blood vessels in the subcutaneous plane. The lesion was abutting right proximal tibia with thinning of its cortex, abutting and compressing anterior crural fascia, tibialis posterior muscle and its tendon, tibialis anterior vessels and interosseous membrane [Figure 1]. A suspicion of soft tissue sarcoma was raised and histopathological correlation was advised.

Fine needle aspiration cytology (FNAC) was done from the site. It revealed low cellular smears, chiefly consisting of spindled cells arranged mostly singly with high N:C ratio, clumped chromatin and prominent nucleoli [Figure 2]. The possibility of spindle cell sarcoma was raised.

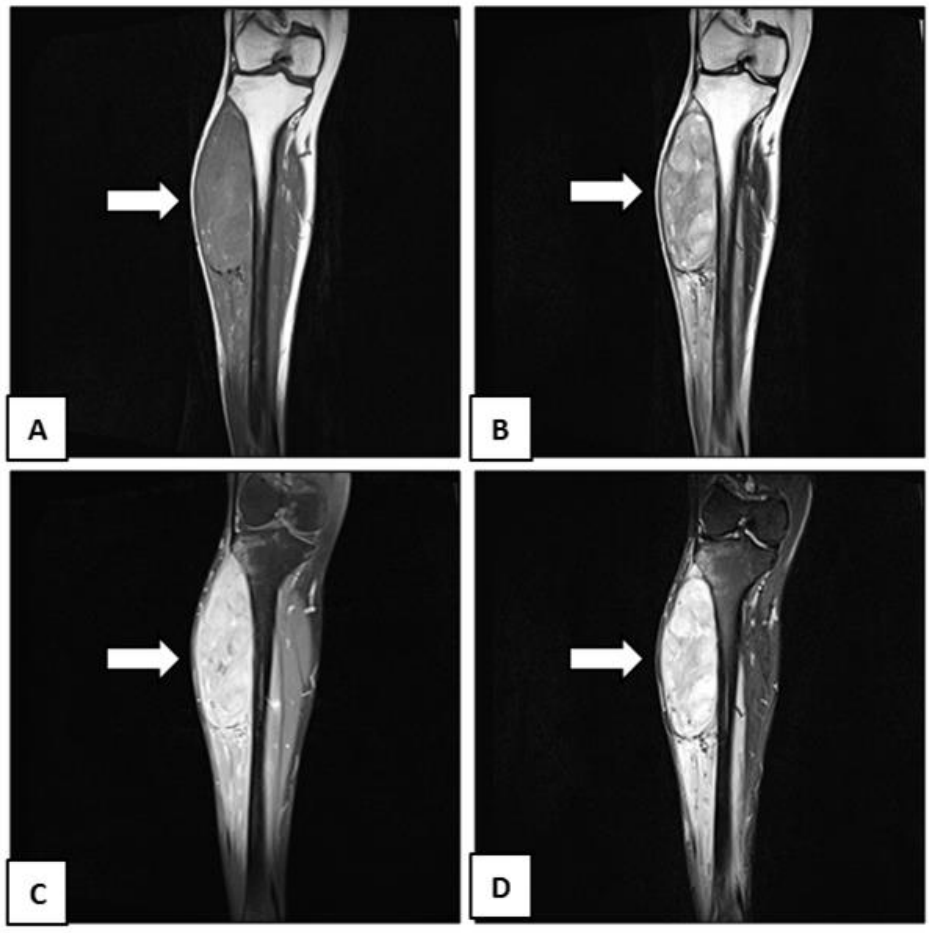

Figure 1: A well-defined spindle shaped mass in anterior intermuscular compartment of proximal right leg involving tibialis anterior & right extensor digitorum longus muscles. The lesion shows intermediate signal intensity with few hyperintense foci on T1W sequence (Fig A); shows heterogeneous hyperintensity on T2W (Fig B) & STIR (Fig C) sequences and shows heterogenous post contrast enhancement (Fig D).

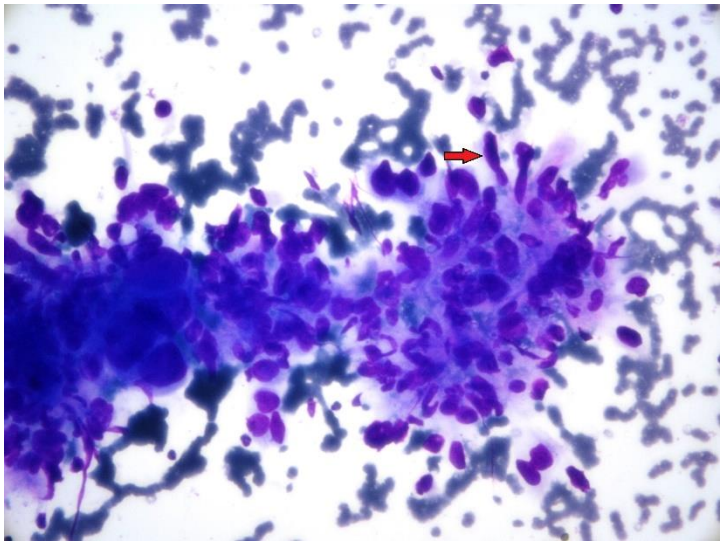

Figure 2: FNAC showing spindled cells (red arrow) with high N:C ratio, clumped chromatin and prominent nucleoli (Giemsa, magnification = 100x).

The patient underwent wide local surgical excision. A linear incision was taken over the lateral aspect of right leg and deepened. Approximately 15x6x6 cm tumor mass was seen probably originating from the tibialis anterior muscle and involving the extensor digitorum muscle. It was extended till tibia without any bone involvement. A sharp and blunt dissection with a 2 cm margin wide local excision of the lesion was done. The remaining muscles where repaired. A drain was kept in situ and closure was done in layers with compressive dressing. Grossly, the tumor was 17x6.5x3 cm. The cut surface showed gray white to gray red mass and was firm in consistency. H&E stained sections showed highly cellular tumor cells mixed with focal areas of hemorrhagic cyst like spaces [Figure 3]. There was also presence of abundant chronic inflammatory cells [Figure 4]. The individual tumor cells showed pleomorphism. The mitotic figures were 10 to 12 mitoses per 10 high power fields. There was no evidence of vascular as well as neural invasion [Figure 5]. Immunohistochemistry (IHC) was done using a formalin-fixed and paraffin embedded tissue blocks. The tumor cells were found to be positive for Vimentin, (EP21, Rabbit monoclonal Primary antibody, Cell Marque) [Figure 6], while it were negative for Desmin (EP15, 1:100, Rabbit monoclonal

primary antibody, Cell Marque) [Figure 7] and S100 (Anti-human S-100 protein, 15E2E2, purified bovine S-100 protein, Biogenex) [Figure 8]. A diagnosis of AFH was made based on histopathology and immunohistochemistry findings. Intra operative and post-operative recovery of the patient was uneventful. The surgeon decided to not to give chemotherapy for this patient considering the intraoperative findings and histopathological findings. The patient is under follow up. There is no evidence of metastasis and patient is disease free till 3 years after excision.

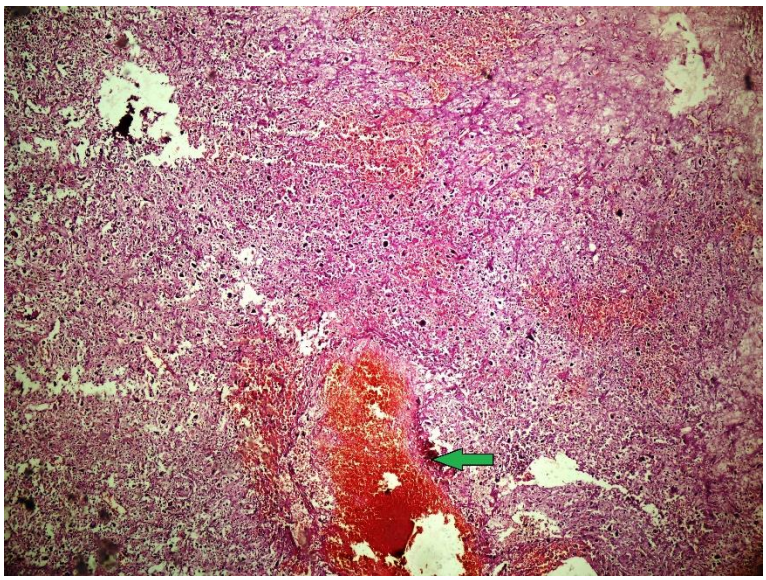

Figure 3: Section showing highly cellular tumour cells mixed with focal areas of hemorrhagic cyst like spaces (green arrow) (H&E, magnification = 40x).

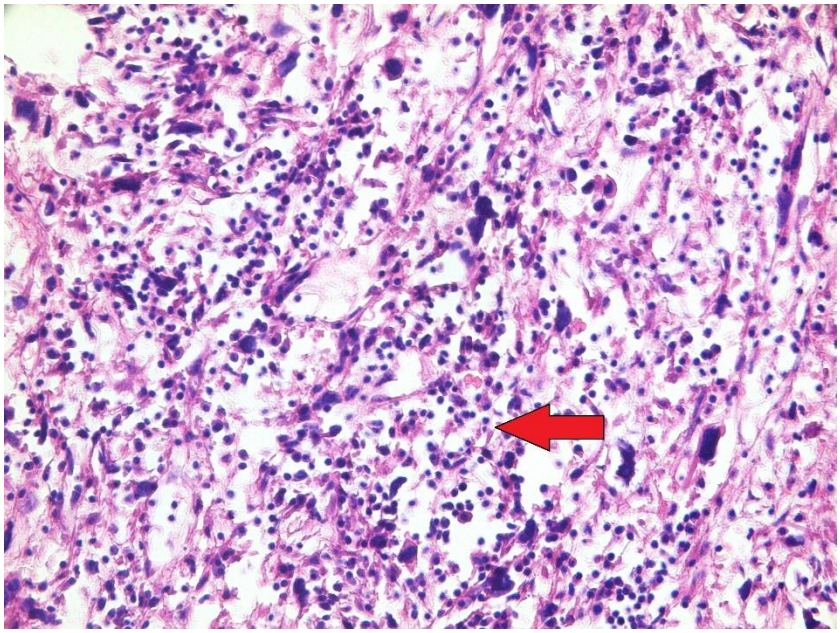

Figure 4: Section showing presence of abundant chronic inflammatory cells (red arrow) (H&E, magnification = 100x).

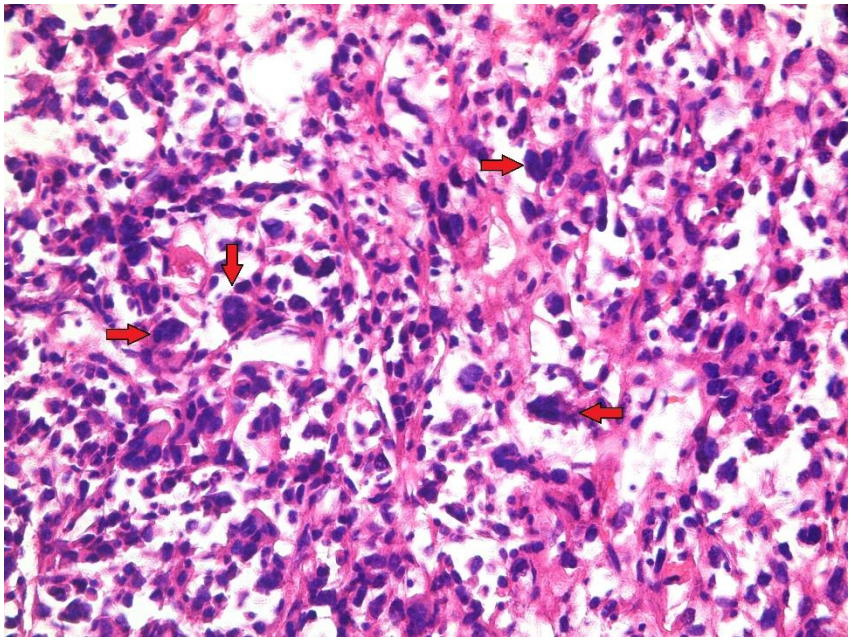

Figure 5: Section showing tumour cells with pleomorphism and mitoses (red arrow) (H&E, magnification = 100x).

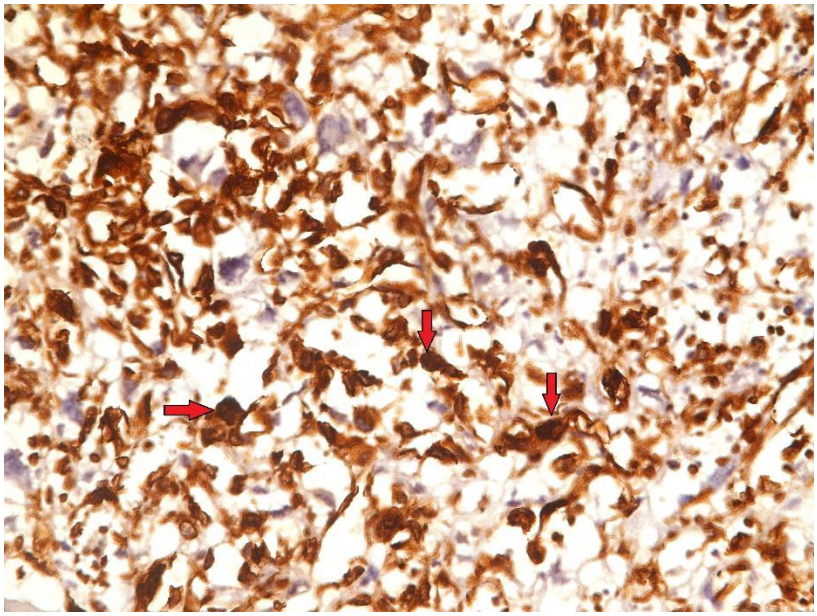

Figure 6: Positivity for Vimentin (magnification = 100x).

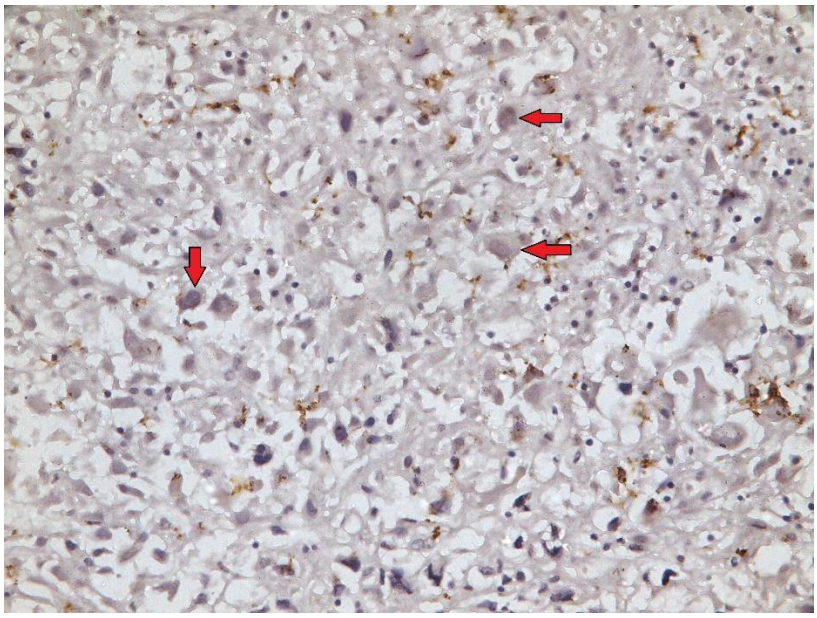

Figure 7: Negativity for Desmin (magnification = 100x).

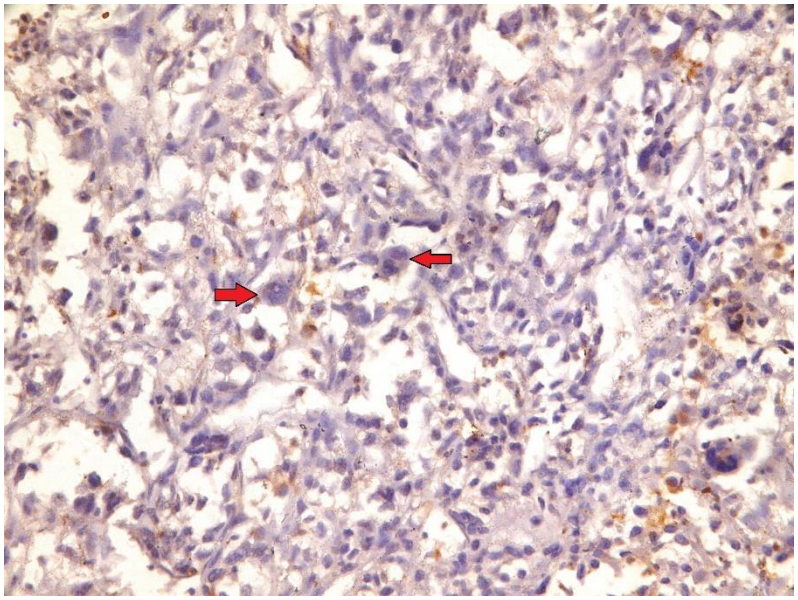

Figure 8: Negativity for S-100 (magnification = 100x).

Discussion:

AFH is classified as an intermediate tumor in the WHO classification.[2] It was initially considered as a variant of malignant fibrous histiocytoma, but a study of follow up of 108 cases showed a favorable prognosis, which ultimately led to its recognition as a distinct entity.[5] AFH are no longer regarded as "malignant" in the present era as precise line of differentiation is still unknown. Thus, the entity has now been removed as a subtype of malignant sarcoma and is placed under the category of "intermediate tumors of uncertain potential" according to 2013 World Health Organization (WHO) classification.[2] The exact line of differentiation for AFH still remains unknown.[2] Literature shows reports of AFH in the age group of children and young adults and very rarely in adults.[2,3,4,5] The median age for presentation is 21 years with a usual slight male preponderance. Ajlan AM et al[3] reported two cases of AFH in 28 and 85 years old patients while, Saito K et al[4] found ages ranging from 8 to 50 years old in a study on seven cases of AFH. Extremities are the commonest location of occurrence for AFH. Other extra somatic soft tissue locations includes mediastinum, lung, retroperitoneum, ovary, lung, bone and brain.[7]

Grossly, the tumor measures around 2-4 cm in size, however few studies also showed large sized AFH.[8] The mass is usually firm in consistency with hemorrhagic cystic spaces.[2] Microscopically, AFH shows the presence of a fibrous pseudocapsule, round to spindled fibrohistiocytic cell proliferation, a plasmalymphocytic infiltrate and a pseudoangiomatous pattern.[2,8] The presence of hemorrhagic cystic spaces cannot be explained in these tumors as clinically patients don't have either thrombocytopenia or any defect in coagulation system.[7] Morphologically, there can be presence of solid or cellular variants of AFH occasionally. The various differential diagnoses for AFH include myxoid tumors like extra skeletal myxoid chondrosarcoma, low grade fibromyxoid sarcoma and myxoid liposarcoma. These can be differentiated by absence of myxoid material, rhadbdomyoblasts, myxoid spindle cells on morphology. There are no specific immunohistochemical markers for the exact diagnosis of AFH till date. Around 50% of these tumors express desmin.[9] Epithelial membrane antigen is usually positive. It can be positive for few other myxoid markers like calponin, smooth muscle actin or rarely h-caldesmon but negative for skeletal markers like myogenin.[9] Our case was positive for vimentin and negative for desmin and S100.

Molecular studies, like fluorescent in situ hybridization (FISH) helps to detect the rearrangement of EXSR1-ATF-1 or FUS-ATF1 fusion transcripts.[7] Tanas et al[10] reported that 76% of cases of AFH in their study were diagnosed by FISH to harbor EWSR1 rearrangement. It helps to confirm the diagnosis of AFH in case of where typical morphological and immunohistochemical features are absent. These cases are usually treated with surgical excision with a wide local excision.[6,11] However; radiation therapy is utilized in cases where marginal excision is planned depending on surgeon's point of view. The literature shows different recurrence and metastasis rate in these cases. Enzinger FM[1] found local recurrence and metastasis in 63% and 21% cases, while Pettinato G et al[12]found

recurrence in 25% and metastasis in 5% cases. In our cases, there is no evidence of metastasis or recurrence till 3 years after excision.

Majority cases of AFH present in 2nd to 3rd decade. A very few cases has been reported in elderly age group as that of our case as per our knowledge.

Conclusion:

AFH is an uncommon soft tissue tumor, which can be found even in an elderly. Lack of specific clinical manifestations, radiological and histopathological features make the diagnosis is more difficult. Treatment includes appropriate surgical excision in order to obtain negative surgical margins, and it is essential to keep the patient under follow up to look for any clinical evidence of tumor recurrence.

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