"All Popliteal cysts are not Baker's cyst: A rare case of Angiomatoid Fibrous Histiocytoma"

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February 16, 2023

Abstract

An angiomatoid fibrous histiocytoma is a rare soft tissue neoplasm of intermediate biologic potential, is often misdiagnosed because of its clinical and radiological similarity to other conditions. Surgery is the mainstay of management and can effectively control local recurrence and metastasis if properly evaluated preoperatively.

TITLE OF THE CASE REPORT: **"All Popliteal cysts are not Baker's cyst: A rare case of Angiomatoid Fibrous Histiocytoma"**

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ABSTRACT:

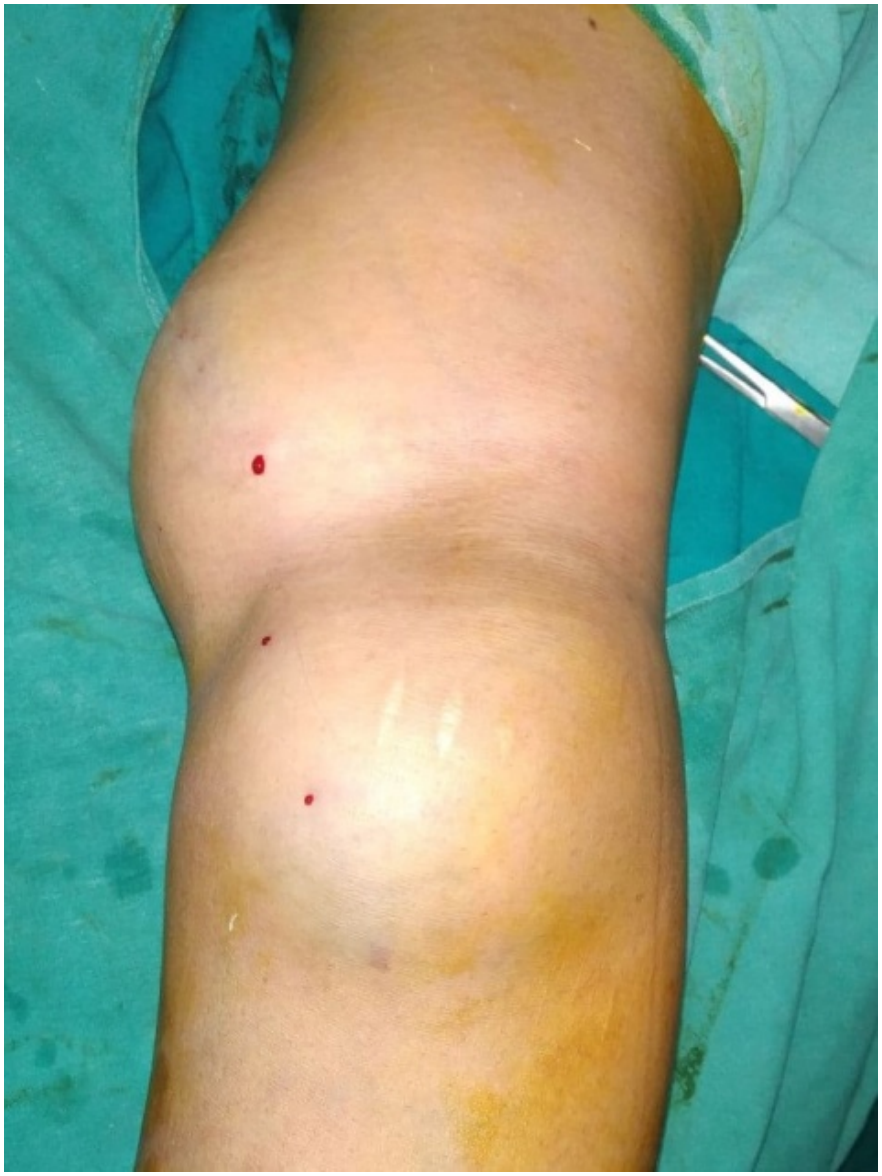
Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue neoplasm of intermediate biologic potential that occurs mostly in extremities and is often misdiagnosed initially, because of its clinical and radiological similarity to other diseases.[1-4] .Clinically it is very difficult to differentiate it from other tumors like angiosarcoma or a simple organised hematoma [2] .Similarly, Histopathology is not always revealing. Surgery is the mainstay of management and can effectively control local recurrence and metastasis if planned beforehand. Herein ,we describe a case report of a 55 year-old female who presented with a swelling in the popliteal region which on clinical and radiological evaluation revealed a popliteal cyst and was managed by an unplanned excisional biopsy. We present this case to emphasize to clinicians that the chances of improper management of such tumors with unplanned excision are high , if such differentials are not kept in mind preoperatively.

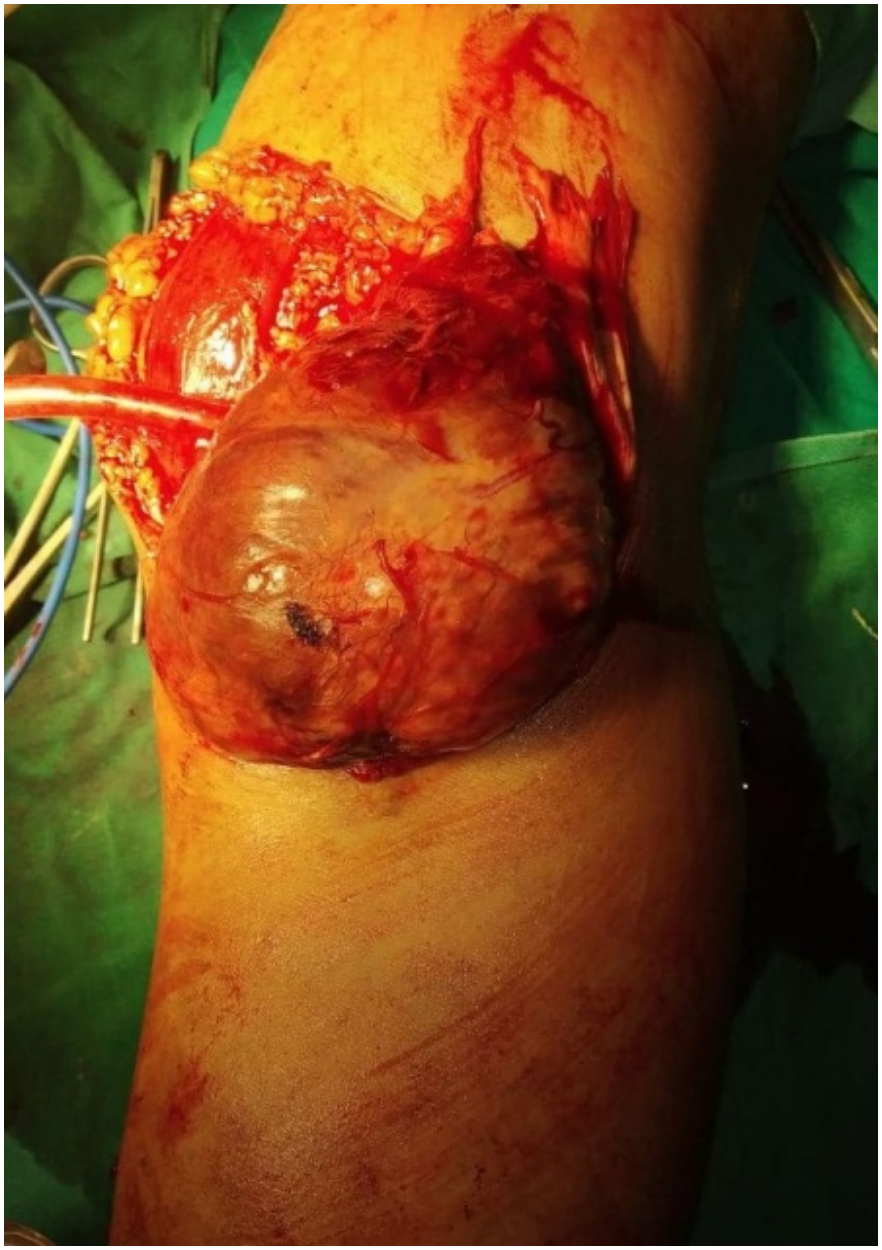
*INTRODUCTION:*Angiomatoid fibrous Histiocytoma(AFH) is a rare soft tissue tumor with intermediate biologic potential that is commonly found in the extremities of children , adolescents and young adults in deep dermis and subcutaneous plane[1,2,4,5] .It was initially described as angiomatoid malignant fibrous histiocytoma by Enzinger[1] ,but it is no longer regarded as malignant because of its benign appearance and favourable prognosis[2,5] .Because of its indolent clinical presentation and radiological features , it is often misdiagnosed for a benign condition such as a hematoma, hemangioma or simple cysts leading to

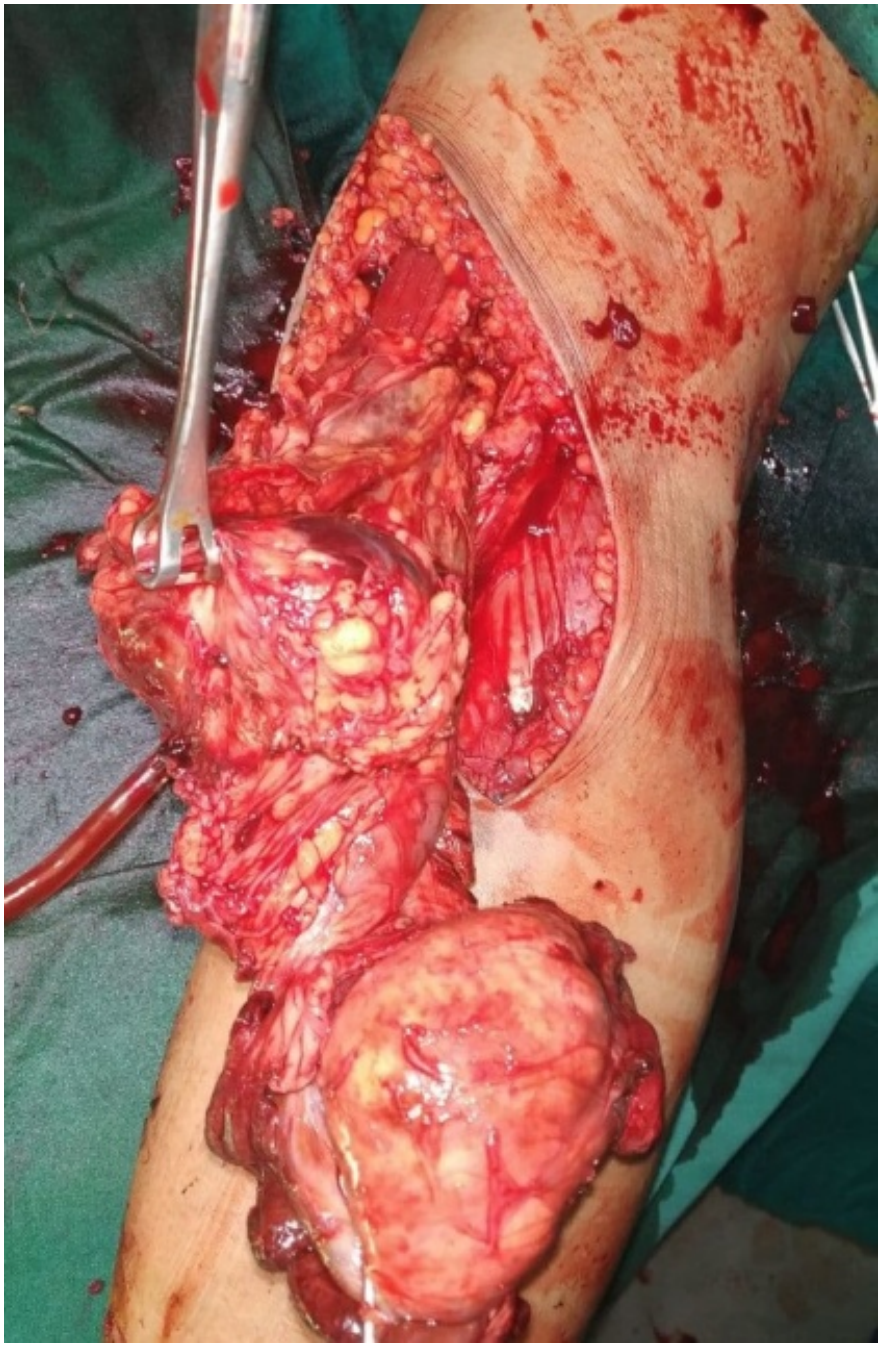
inappropriate treatments such as unplanned excision[5]. Although the prognosis of AFH is not poor , it recurs in up to 15 % of cases and metastasizes in fewer than 1 % of cases[2] .So, keeping AFH as a differential in cases with soft tissue masses can guide us to do an extended resection of the tumor with wide margins and minimise the chances of recurrence.

CASE PRESENTATION :

A 45 year old female presented to us with a history of right knee pain for 6 months and swelling in the posterior aspect of the knee for 1 year, which has been gradually increasing in size. The patient had difficulty squatting and sitting cross-legged . There were no constitutional symptoms. Past medical, surgical and family history were not significant. On examination, there were two soft cystic , fluctuant, non-tender swelling , each around 7*5 cm arising from the posteromedial aspect of the right knee lying longitudinally in craniocaudal fashion (Figure 1).There were no skin changes .USG findings revealed a fluid filled , well circumscribed, dumbbell shaped mass with 2 cysts, each of sizes around 6*5*3 cm communicating with each other arising from the posteromedial knee with a neck at its deepest extent, extending into the joint space between the semimembranosus and medial head of gastrocnemius suggestive of popliteal cyst. The patient underwent en bloc surgical excision(Figure 2,3,4) .On histopathological examination, the lesion demonstrated characteristic features of Angiomatoid Fibrous Histiocytoma including multinodular growth of myoid spindled or histiocytoid cells with a distinctive syncytial appearance, pseudo-angiomatous spaces filled with blood and surrounded by tumour cells , a thick fibrous pseudo capsule with peritumoral lymphoplasmacytic cuffing with occasional germinal center formation, areas of prominent hemosiderin deposition and foam cells within the tumour cells (Figure 5,6). Immunohistochemistry for supporting the diagnosis was planned but due to resource constraints, we could not manage the same.







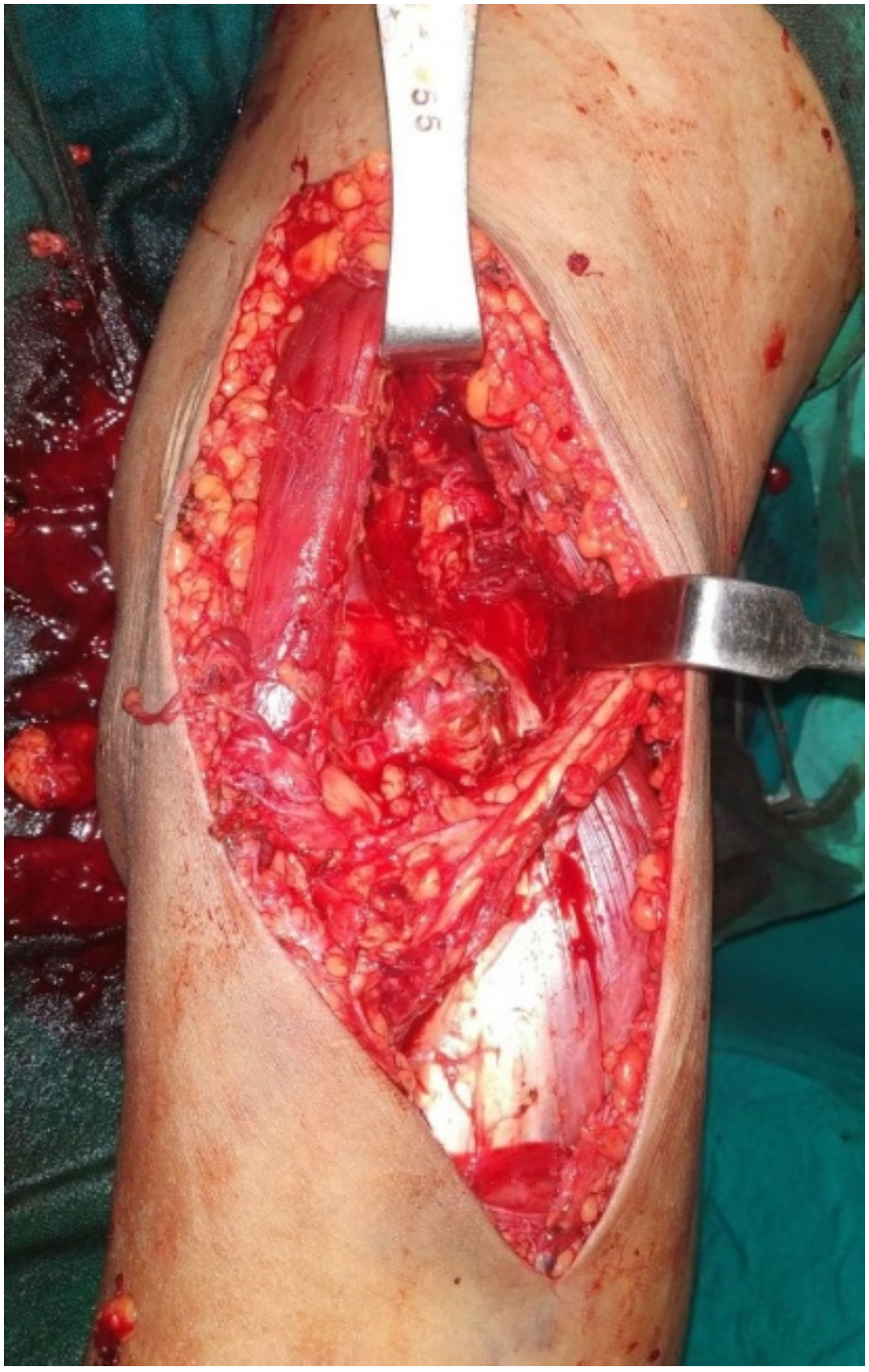
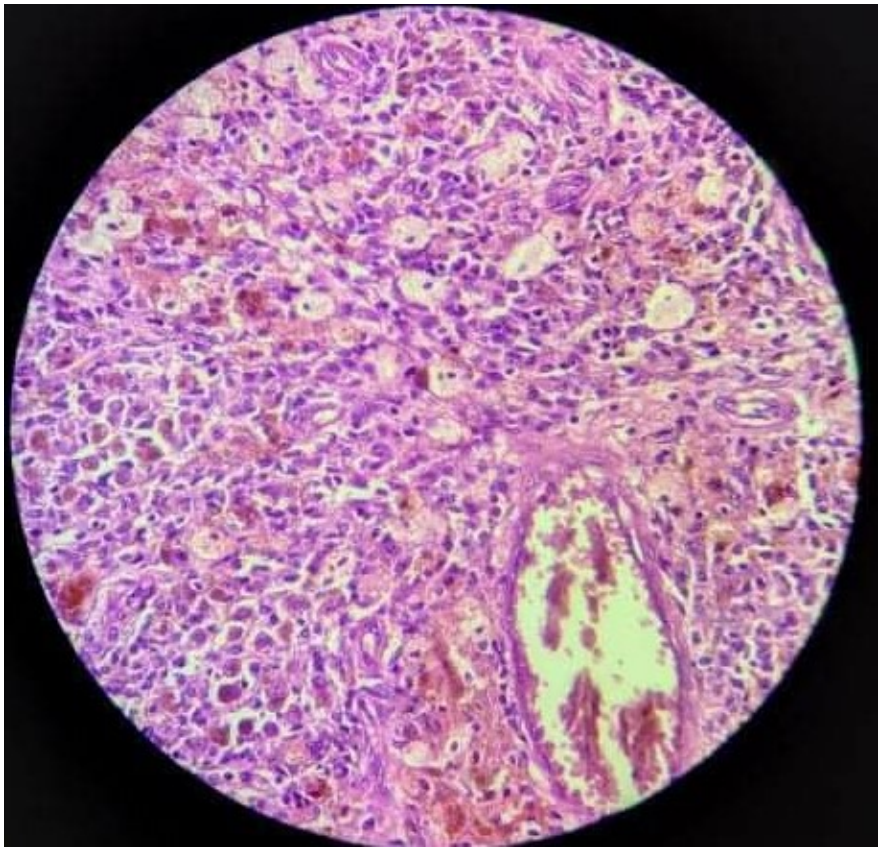
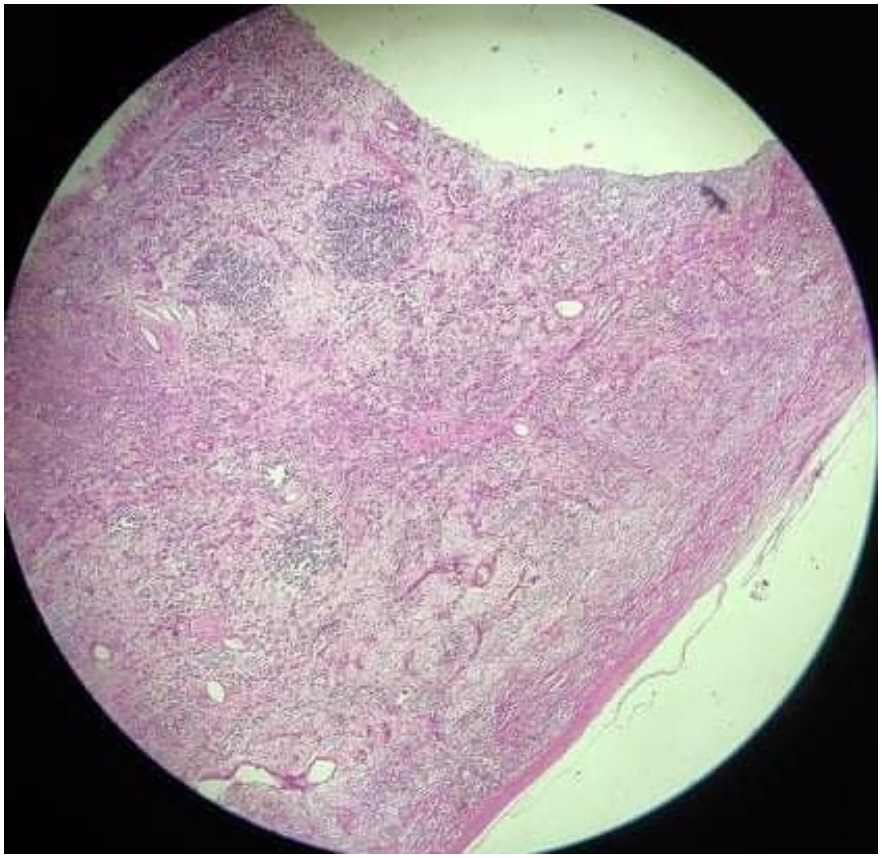


Figure 1:Posteromedial aspect of the knee with a swelling ,bilobed shape and with ill defined borders Figure 2:Intraoperative image showing a cystic mass in the popliteal fossa region Figure 3: white-tan, lobulated, haemorrhagic firm mass with thick capsule measuring 6.5*4.3*3.2 cm Figure 4: Figure showing the relations of the tumour after the excision. a)Medial head of Gastrocnemius b)Floor with exposed knee joint posteriorly c)Neurovascular bundle of popliteal fossa retracted medially d)Lateral head of Gastrocnemius.





Resected pathological specimen demonstrated: Figure 5: Pseudovessels: Around the blood vessels are tumor cells with admixed foamy cells and hemosiderin laden macrophages. (H and E, 100 X)

Figure 6: Section reveals Capsule: Within the capsule are various calibred pseudocystic like spaces and proliferating myoid to histiocyte like tumor cells. (H and E, 40 X)

DISCUSSION:

AFH was initially described by Enzinger[1] in 1979 as malignant angiomatoid fibrous histiocytoma. They constitute around 0.3% of all soft tissue tumors.[2-5]. However, the data may be understated because of the overlapping histopathological findings.[5] But, The World Health Organization (WHO) in 2013 classification placed it in the category of "intermediate tumors of uncertain differentiation" because of its better prognosis[2,4-13] .Although AFH can occur at any age but most patients present in the first three decades of life .In our case, the patient was in her forties. Although patients with AFH typically present with a painless subcutaneous soft tissue lump[5,10] , Costa and Weiss[3] reported that 18% of tumors invaded deep structures such as skeletal muscle. In our case, it was a painless swelling in the popliteal fossa region. Intraoperatively it was intraarticular in location and stump was found to be attached to the underlying bone. It most commonly arises in sites of normal lymphoid tissue such as the antecubital fossa, axilla, inguinal and supraclavicular regions [5,8,13].The majority of cases occur in the extremities, although cases have been reported in the head and neck region (10%) and trunk[1,5,6,13]. The cumulative findings of a meta-analysis of multiple studies demonstrate that the majority of patients (73.2%) are disease free after local excision and a minority (23.2%) develop recurrent disease and 8.7% metastasise within 24 months of surgery[3]. Metastasis occurs predominantly to regional lymph nodes but exceptionally to the lungs, liver, or brain.[13]

Making a pre-operative diagnosis of AFH is challenging with no distinct clinical or imaging findings to lead

to diagnosis. There is a vast variability of the differential diagnoses which includes: Aneurysmal bone cysts, schwannoma, synovial sarcomas, aneurysmal fibrous histiocytomas, follicular dendritic metastatic tumor of lymph nodes .[11,13] The presence of systemic symptoms, such as weight loss, malaise, fever, and anemia , which occur in some patients, can aid diagnosis[5,6,10,11] The cause of these constitutional symptoms are thought to be due to the tumoral cytokine production[5]. However, in the current case, the patient experienced no systemic symptoms.

The imaging modalities are not diagnostic but aid in the management. The Computed tomography may show a heterogeneous mass and possibly hint at cystic and enhancing components. MRI is superior to CT scan for the diagnosis and explaining the relationship with the surrounding structures[5] ·Although the MRI features of AFH described in the literature may aid diagnosis , none of them are considered to be significant[11] .MR has been traditionally used for postoperative follow-up. MRI was not done in our case due to non-availability in our institute and we proceeded with an excisional biopsy after the USG report.

Histopathological features of AFH have been well described[2,11].This includes the following features:(i) multinodular growth of myoid spindle or histiocytoid cells with a distinctive syncytial appearance, (ii) pseudoangiomatous spaces filled with blood and surrounded by tumor cells, (iii) a thick fibrous pseudocapsule with prominent hemosiderin deposition, and (iv) peritumoral lymphoplasmacytic cuffing with occasional germinal centre formation. These features were consistent to our case. AFH lacks a specific immunoprofile, so that immunohistochemistry is supportive rather than diagnostic. Immunohistochemistry variably demonstrates positivity for desmin, CD68 and CD99 [5,7,9-12] .Lastly, cytogenetic analysis has recently added to the diagnosis of AFH, with the EWSR1-CREB1 fusion gene present in the majority of AFH[5,8-12]

The prognosis of AFH is generally considered to be favorable[2,9-12] but because of its chances of local recurrence ,soft tissue sarcomas should be operated by a specialised surgeon and special consideration should be taken not to miss these tumors preoperatively . There should be wide surgical excision with clear margins. Extended resection of AFH is recommended because the tumor is considered intermediate and not purely benign as it requires continued clinic radiological surveillance [5] .

There were some limitations in the current study. We did not perform immunohistochemistry for desmin and did not examine fusion genes, which are often useful for differential diagnosis. However, a diagnosis of AFH could be made in specimen Biopsy. Given the intermediate malignant potential of this lesion, the patient will require continued clinical and radiographic surveillance.

CONCLUSION :

AFH is a rare disease that is often misdiagnosed initially. Patients can present with a clinical picture suggestive of other diseases, such as popliteal cyst as in our patient. In our series, AFH occurred in relatively older patient and was found in deep lesions. Although patients with AFH seem to have excellent prognoses, the chances of local recurrence and metastasis might be higher than initially expected. Therefore, Surgeons must be aware of AFH and include it in clinical, radiological, and histopathological differential diagnosis .

CONSENT: Written and informed consent was obtained from the patient for the publication of the case report and is available for the review by the editor of the journal.

AUTHOR CONTRIBUTION: Sriram Khati had evaluated, operated the case and has role in conceptualization, writing, editing, resource collection. Omkar Bist did the review and sources collection. Govinda Prasad Tiwari edited and reviewed the article.

FUNDING : None

DATA AVAILABILITY*:* Data described to support the findings in the study are openly accessible in the article.

COMPETING INTERESTS : Non conflict of interests.

ETHICAL STATEMENT : This is the retrospective case report and no sampling was used. The ethical approval can be waived.

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