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Angiomatoid Fibrous Histiocytoma: Case Report

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ABSTRACT

Background: Angiomatoid Fibrous Histiocytoma (AFH) is a rare soft tissue tumor that occurs most often in children, teenagers and young adults. In clinical and radiographic terms, the lesion is easily mistaken for hematoma, soft tissue hemangioma or malignant fibrous histiocytoma. While the lesion is rare, because of the potential for local recurrence and metastasis, this lesion must be accounted for in the differential diagnosis of a soft tissue mass.

Objective: A case report of angiomatoid fibrous histiocytoma; the clinical, radiological and pathological outcomes.

Case Report: Our case report of a 30 years old female who presented with left knee soft tissue mass anterior to the patella, which was discomforting, painful mass. X-ray and MRI of the left knee showed a soft tissue mass in the adipose tissue not attached to the underlying musculature. Wide margin resection was done,

Result: Histopathology report findings were consistent with angiomatoid fibrous histiocytoma. CT staging done postoperatively with no sign of metastatic lesion. Follow up MRI left knee done showed no sign of recurrence.

Conclusion: Angiomatoid fibrous histiocytoma is a rare soft tissue tumor with intermediate malignant potential. Although non-specific, a mass with MRI characteristics, including cystic areas, a fibrous pseudocapsule and internal blood spots on the extremity of a child, adolescent or young adult, AFH must be considered as part of the differential diagnosis. A wide surgical excision with clear margins and post-excision follow-up is required.

Keywords: Angiomatoid Fibrous Histiocytoma, CT, MRI, X-ray.

INTRODUCTION

Angiomatoid fibrous histiocytoma (AFH) is a rare type of soft tissue tumor. It commonly affects extremities, such as arms, legs, hands or feet. AFH is a soft tissue neoplasm of low malignant potential, typically occurring in the superficial soft tissues of the extremities in children and young adults. Occurrence outside somatic soft tissues is most uncommon [(1)].

AFH is a tumor that most frequently affects children and young individuals between the ages of 5 and 25 years (median 13 years). This tumor is rare, representing approximately 0.3% of all soft tissue tumors. Although incidence can be underestimated because of overlapping histopathological results [(2)]. Presentation usually involves a slow-growing, painless mass in the deep dermis and subcutaneous tissue [(3)]. It most frequently occurs at sites of normal lymphoid tissue, such as the antecubital fossa, axilla, inguinal and supraclavicular regions [(4)]. The majority of cases occur at the extremities, although there are reported cases in the head and neck region (10%) and trunk [(1,5)]. A small proportion of patients exhibit systemic symptoms, including pyrexia, anemia and malaise, suggesting production of tumor cytokines [(6)]. Symptoms of pain and tenderness are rare [(7)].

CASE REPORT

A 30-year-old woman showed up with a painful mass on her left knee for 18 months. Pain increases with an activity such as walking and an activity involving knee flexion. Wearing trousers or kneeling pressure is painful. The patient does not recall a history of a left knee injury or traumatic events. The pain used to be on/off, but now it is continuous. Pain is discomforting and interfering with her daily activity. No constitutional symptoms were presented.

Physical exam of her left knee demonstrated anterior to the patella, superficial mass under the skin, rounded, measuring 2.1x1.8 cm without erythema, not tender, palpable mass, soft, mobile, not attached to the skin or the underlying fascia. Radiographs of the left knee (Figures 1, 2, 3) were unremarkable. Subsequent magnetic resonance imaging (MRI) of the left knee (Figures 4, 5, 6) showed a well-circumscribed solitary left knee prepatellar soft tissue mass measuring: 3.5 x 3.4 x 1 cm, in the subcutaneous adipose tissue superficial to the patella and quadriceps tendon without involvement of the underlying musculature.

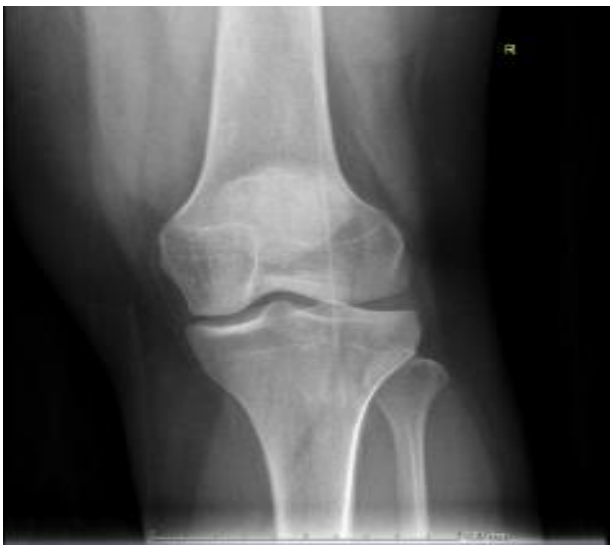

Figure (1): Anteroposterior view left knee x-ray done on initial presentation, lesion can't be appreciated in all views.

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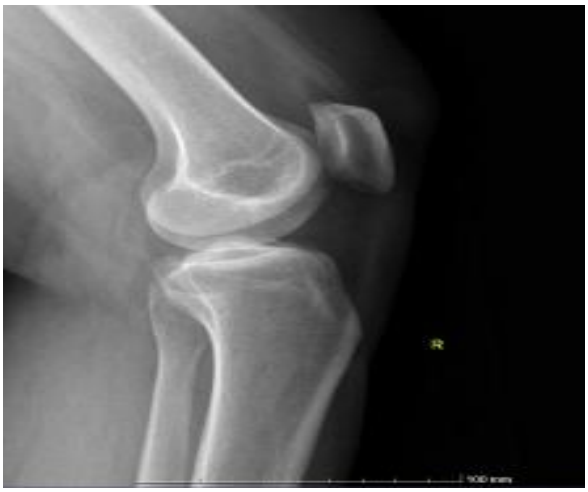


Figure (2): Lateral view left knee.

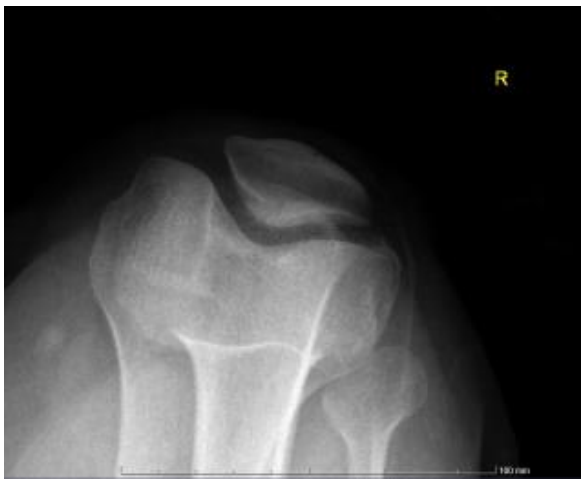


Figure (3): Skyline view left knee.

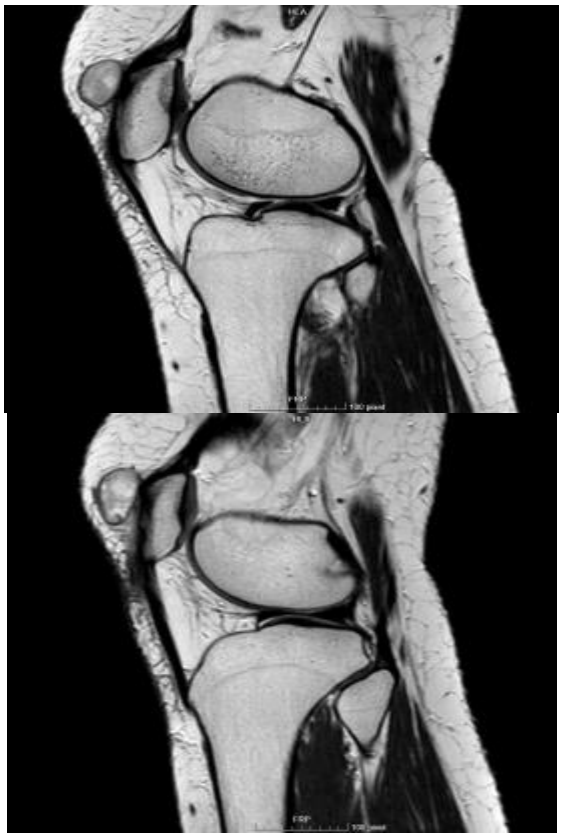


Figure (4): T2 weighted image fast spin echo sagittal view showing well-circumscribed, ovoid, mixed cystic and solid mass is present in the subcutaneous fat of the anterior left knee without evidence of muscle infiltration. The lesion is heterogeneously hyperintense. The periphery of the mass demonstrates the typical low signal on unenhanced images representing a fibrous pseudocapsule

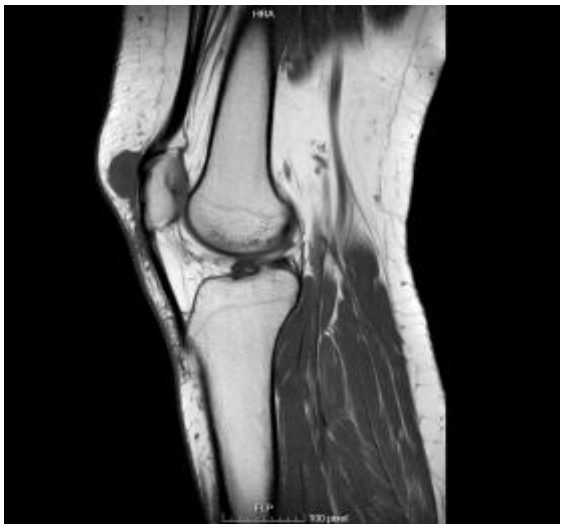
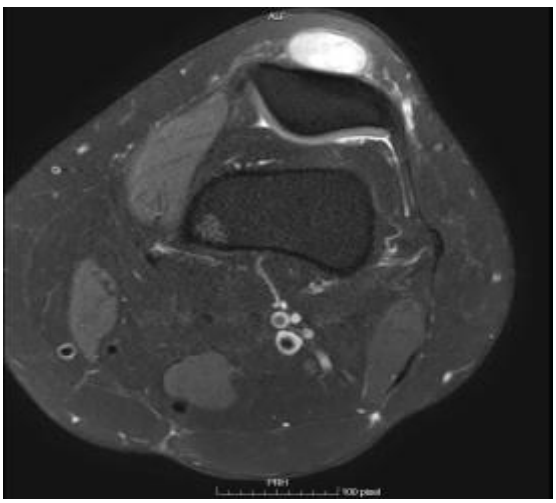
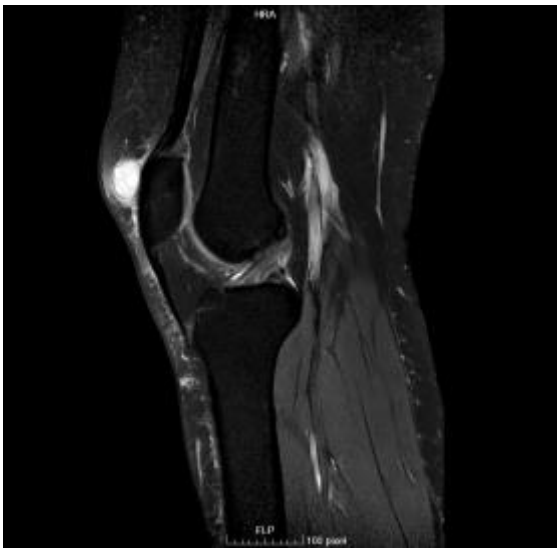


Figure (5): T1 weighted image fast spin echo sagittal view. The lesion is homogeneously hypointense.





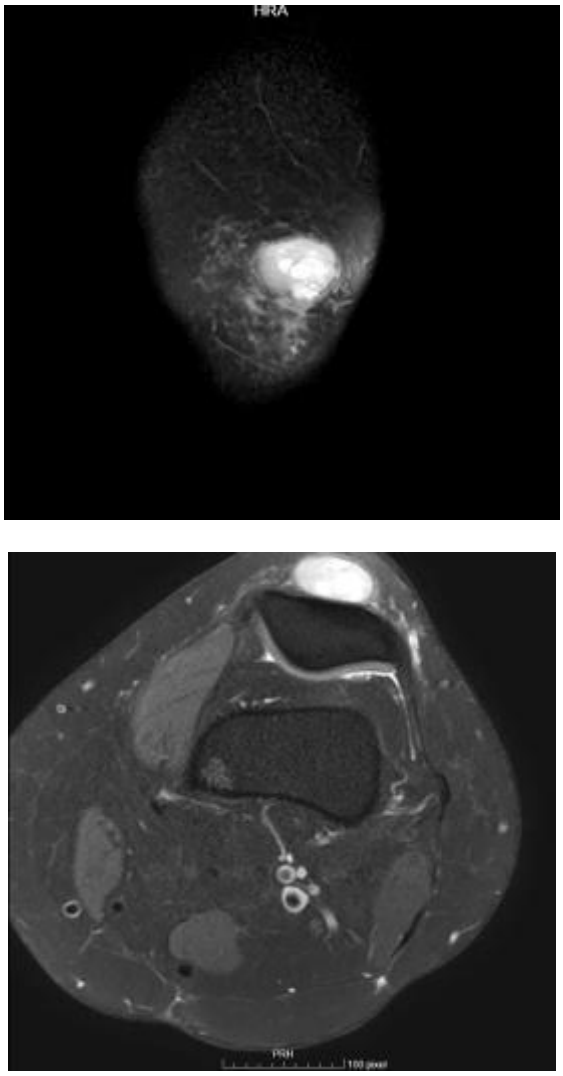


Figure (6): PDW SPAIR (Proton Density Weighted Spectral Attenuated Inversion Recovery) MRI images sagittal, coronal and transverse, the lesion is heterogeneously hyperintense.

The patient underwent surgical excisional biopsy.

On pathological examination, the lesion demonstrated characteristic features suggestive of angiomatoid fibrous histiocytoma. Specimen showed a neoplasm composed of pseudo-encapsulated lesion with peripheral lymphoid aggregates. There were nodules, sheets, short fascicles of monomorphic bland spindle to ovoid eosinophilic cells in a focal myxoid background. The neoplasm contained hemorrhagic cyst-like spaces. The neoplastic cells had focal severe pleomorphism. The surgical resection margins were negative.

On immunohistochemistry, the tumor cells showed CD99 focally +ve, CD 68 focally +ve, Desmin rare positivity, smooth muscle actin focally +ve, S100 -ve, CD34 -ve.

Staging CT scan of the chest, abdomen and pelvis 5 weeks after excision was not showing any signs of metastasis. Follow-up MRI imaging 6 months after resection showed no evidence of recurrence.

Ethical considerations:

An approval of the study was obtained from Soliman Fakeeh Hospital, Jeddah (Saudi Arabia) Academic and Ethical Committee. The patient and her relative were informed that the case would be published as case report and this was accepted. This work has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

DISCUSSION

This lesion initially were described as "angiomatoid malignant fibrous histiocytoma" by **Enzinger** in 1979 [(1)], the histological aspect was controversial. At present, the exact line of differentiation remains unknown. But it's not called "malignant" anymore. Based on its benign microscopic appearance and favorable prognosis [(8)]. Furthermore, the 2002 World Health Organization (WHO) classification removed it from the malignant subtype of fibrous histiocytoma of sarcoma (now a synonym for undifferentiated pleomorphic sarcoma). And categorized as tumors of uncertain differentiation as angiomatoid fibrous histiocytoma [(4)].

It is difficult to make a preoperative diagnosis of AFH without distinct clinical or imaging findings that can lead to diagnosis. However, malignant soft tissue tumors were stratified by age and location in order to narrow the differential diagnosis. For instance, based on a study of 39,179 soft tissue lesions over a 10-year period, 80% of the rare malignant soft tissue masses in the patients aged 6-15 in the hand and wrist, upper extremity, axilla and shoulder, lower extremity, hip, groin and buttocks, or trunk are most likely or second most likely to be AFH [(9)].

In most cases. Histological findings are more precise and diagnostic than radiological findings. Radiographs and CT scans may show heterogeneous mass. And possibly hint at cystic and enhancing components. but inferior to MRI soft tissue details and findings. MRI imaging has historically been used for staging and follow up.

Although a latest report drawing attention to the potential utility of positron emission tomography in staging [(10)]. The diagnosis of AFH relies on histopathology and immunohistology findings. At a macroscopic level, the AFH is usually firm and circumscribed. The characteristic microscopic appearance includes distributions of ovoid to spindle cells with bland, vesicular nuclei, lymphoplasmacytic infiltrate with intervening blood-filled cystic spaces, and a fibrous pseudocapsule [(11)]. Immunohistochemistry provides variable positivity for desmin, CD68 and CD-99 [(12)]. Finally, cytogenetic analysis is added to the diagnostic of AFH, with the fusion gene EWSR1-CREB1 present in most AFH [(13)].

Since the tumor was first described, many studies have been conducted to determine the malignant

potential of AFH with variable conclusions. Cumulative results from a meta-analysis of several studies show that most patients (73.2%) are disease-free after local excision, and a minority (23.2%) developed recurrence and 8.7% developed metastasis within 24 months of surgery [(14)]. The overall mortality rate is 4.3%, which may be underestimated due to short monitoring periods [(15)]. While, the tumor is often described in the literature as a low malignant potential lesion, it is officially classified as intermediate malignant potential [(13,16)]. The clinical behavior of the tumor cannot be determined on the basis of clinical or histological parameters [(3,14,17)]. However, the development of local recurrence and metastases demonstrated a correlation with invasion in the deep fascia or muscle as surgically evaluated [(14)]. The treatment consists of surgical resection.

We report a case of AFH in the left knee of an adult female. The case is consistent with the clinical, radiographic and histopathological characteristics of AFH.

The patient was free of recurrence following surgical excision approximately 25 months following surgery. Due to the intermediate malignancy potential of this lesion, the patient will need continuous clinical and radiographic monitoring.

CONCLUSION

Angiomatoid fibrous histiocytoma is a rare soft tissue tumor with intermediate malignant potential. Although non-specific, a mass with MRI characteristics, including cystic areas, a fibrous pseudocapsule and internal blood spots on the extremity of a child, adolescent or young adult, AFH must be considered as part of the differential diagnosis. A wide surgical excision with clear margins and post-excision follow-up is required.

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