



## **Primary Malignant Fibrous Histiocytoma of Proximal Humerus with Pathological Fracture- Case Report**

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### **Authors' contributions**

*This work was carried out in collaboration among all authors. Author MDS Conception and critical revision of the article. Author AAN Data collection and Drafting the article. Author SSR Data analysis. Author SMD Final approval of the version to be published. All authors read and approved the final manuscript.*

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**Case Report**

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### **ABSTRACT**

Malignant fibrous histiocytoma (MFH) is a tumor of late adulthood that occurs in men more commonly than women. Proximal humerus involvement in PMFH is comparatively rare. A 45 year old female presented with complain of pain and mild swelling over the left shoulder with restricted range of movement since 15 days. Radiography of the affected part was suggestive of an osteolytic eccentric lesion with a pathological fracture. A trucut needle biopsy showed minimal tumour tissue with abundant necrosis and showed giant cells with pleomorphic mononuclear cells. So intralesional curettage followed by osteosynthesis with PHILLOS plating and bone cementing was done. Intra operative specimen sent for histopathological examination showed presence of fibroblast cells arranged in storiform pattern s/o MFH along with occasional multinucleated giant cells. Postoperatively patient was started with adjuvant chemotherapy. Patient had good results functionally and clinically with no signs of recurrence till follow up of 1.5 years.

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## 1. INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a tumor of late adulthood that occurs in men more commonly than women.<sup>1</sup> But its osseous counterpart is an unusual entity, accounting for less than 2% of all primary malignant tumors [1]. Primary osseous MFH (PMFH) is a centrally located lesion found in the diaphyseal or metaphyseal region of the bone that causes aggressive bone destruction and a soft tissue mass. [2] PMFH is found in the extremities 70–75% of the time with about 50% of the cases in the lower extremities.<sup>3</sup> The common sites in order are the distal femur, proximal tibia and proximal femur. MFH was reported as somatic soft tissue sarcoma in many studies but bony involvement is relatively rare [3].

Proximal humerus involvement in PMFH is comparatively rare. Pathological fracture is noted in 20% of all patients with MFH of bone at the first presentation [4]. Here, we report a rare case of PMFH of the proximal humerus and its management, presenting with a pathological fracture along with osteolytic lesion.

## 2. CASE PRESENTATION

A 45 year old female presented with complain of pain and mild swelling over the left shoulder since 15 days. Patient gives history of a trivial trauma 10 days after the onset of the above complaints. A thorough clinical examination revealed slight swelling and tenderness of the proximal aspect of the left upper arm, restricted and painful movements of left shoulder, without vascular and neurological involvement. All preoperative routine blood investigations were within normal limits. Radiography of the affected

part was suggestive of an osteolytic eccentric lesion with a pathological fracture [Fig. 1]. A trucut needle biopsy was done, which showed minimal tumour tissue with abundant necrosis and degenerative changes. Sections studies showed giant cells and pleomorphic mononuclear cells. Patient was diagnosed provisionally with a possibility of giant cell tumour. Based on the trucut needle biopsy, patient underwent surgical intervention in form of intralesional curettage followed by osteosynthesis with PHILLOS plating and bone cementing [Fig. 2]. Intra operative specimen was sent for histopathological examination, which showed presence of fibroblast cells arranged in storiform pattern along with occasional multinucleated giant cells [Fig. 3]. These cells showed mild to moderate pleomorphism with 3-4/10 HPF of mitoses. Immunohistochemistry study showed positive for CD68. Biopsy report confirmed the diagnosis of MFH with low malignant potential. Post-operatively, patient was immobilize for 2 weeks in shoulder arm pouch with immobilizer after which gradual physiotherapy for up to 6 months was given to achieve good functional range of movement of shoulder. This intra operative biopsy report was different from the initial biopsy taken pre operatively. Thereafter patient was referred to a higher centre for adjuvant chemotherapy and was followed up regularly every 3 months to examine for any signs of recurrence or metastasis. We managed patient with adjuvant postop chemotherapy and followed this patient till 1.5 years, showing no clinical or radiological signs of any recurrence. Patient showed good radiological and functional results with no sign of recurrence or metastases at final follow-up of 1.5 years [Figs. 4 and 5].

### PRE OP X RAY



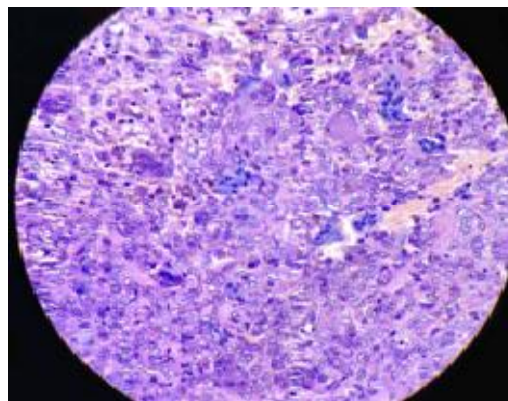
**Fig. 1. Preoperative radiograph of left proximal humerus AP and lateral views showing an osteolytic eccentric lesion with a pathological fracture**

**POST OP X RAY**



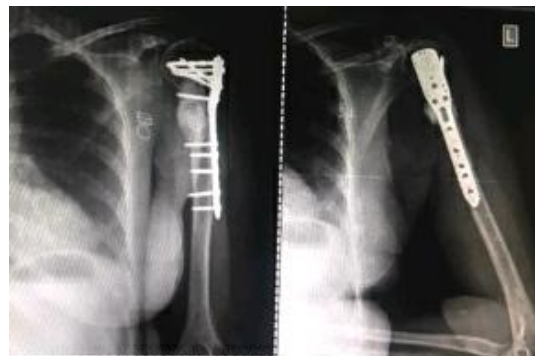
**Fig. 2.** Postoperative radiograph of left proximal humerus AP and lateral views showing intracortical curettage followed by osteosynthesis with PHILLOS plating and bone cementing

**HISTOPATHOLOGY REPORT**



**Fig. 3.** Intra operative specimen showing presence of fibroblast cells arranged in storiform pattern along with occasional multinucleated giant cells

**LAST FOLLOW UP X RAY**



**Fig. 4.** Last follow up at 1.5 years postoperative radiograph of left proximal humerus AP and lateral views showing united pathological fracture of left proximal humerus

### FOLLOW UP MOVEMENTS



**Fig. 5. Final follow up at 1.5 years postoperative showing full range of motion of left shoulder joint**

### 3. DISCUSSION

MFH usually presents with a soft tissue mass with or without cortical erosion [5]. Radiologically, MFH is an aggressive, permeative lesion, which often lacks distinctive features found in other high-grade primary bone malignancies [6]. There is not normally a periosteal reaction. MRI is very helpful for MFH in local staging as well as surgical planning because it assesses the degree of intramedullary extension (and dimensions) and invasion of the adjacent physeal plates, joints, muscle compartments and neurovascular bundles [7].

<sup>18</sup>F-FDG PET/CT has high sensitivity, specificity and accuracy for MFH based on the semiquantitative measurements of the glucose consumption and also it felt to be an extremely useful tool for the detection of bone metastases and recurrence [8]. But in this case as patient due to financial issue didn't undergo for such investigations.

MFH classification showed a broad range of histological appearances and consisted of 5 subtypes: storiform, pleomorphic, myxoid, giant cell and inflammatory [9]. The mainstay treatment for MFH-B is preoperative chemotherapy followed by wide resection with adjuvant chemotherapy [10]. Chemotherapy reduces the tumor bulk and increases the chances of a limb sparing procedure. Prognosis depends on local recurrence and metastases. The prognosis of MFH becomes worse as a lesion is larger and deeper in the soft tissue. MFH metastasizes to the lungs, lymph nodes, liver and bone [11]. In several studies postoperative radiation therapy has

been recommended for patients in postoperative histological evaluation [12]. In current case, interestingly due to contradictory biopsy report, we initially treated patient as giant cell tumor and later we treated patient postoperatively with adjuvant chemotherapeutic agents (Methotrexate + Adriamycin) for MFH and using this protocol, remissions are achieved significantly with better functional improvement. Combination of Intralesional curettage and adjuvant chemotherapy appears to be the most promising and easily managed at rural level where there is unavailability of radiation and related high cost therapies. The prognosis of MFH of bone is poor with having reported 5-year survival rate of 38.3% [13]. The surgical management in relatively younger age group were associated with a better prognosis as compared to older age group having comorbidities [14]. Bone tumours have variable biological behaviour with prognosis depending on local or distant recurrence and metastases [15]. The other review of the literature reveals different rates of metastases and local recurrences between 30-70% [16]. Several studies reported that due to adjuvant chemotherapy the chances of local or metastatic diseases shows gradual reduction. As certain studies shown that intralesional surgery have high chances of local recurrence and metastatic possibilities but we still didn't report any recurrence or metastases in this case [17]. Some studies also reported that adjuvant radiation therapy is not as effective as chemotherapy [18]. In the present report, the lower rate of metastasis was due to the effectiveness of chemotherapy [19]. As the patient was last followed up till 1.5 years, we require a minimum 5 years of follow up to rule out recurrence so as to assess survival rate.

#### 4. CONCLUSION

MFH treated with a surgery and adjuvant chemotherapy without radiotherapy can also give satisfactory outcome. We also conclude that current approach for low malignant MFH showed good results functionally as well as clinically with no signs of recurrence. Therefore, consideration can be made to administer chemotherapy in addition to intralesional curettage in order to improve survival rates in low PMFH without radiation.

#### CONSENT

It is not applicable.

#### ETHICAL APPROVAL

It is not applicable.

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#### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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