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Malignant Fibrous Histiocytoma of Shoulder Masquerading as Adhesive Capsulities

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Abstract: ***Malignant fibrous histiocytoma (MFH) is a rare malignant primary bone tumour that contains a mixture of fibrogenic cells that are histologically similar to histocytes. Histologically MFH appear as large multi-nucleated malignant cells, with abundant cytoplasmic nuclei. Incidence: MFH constitutes less than 2% of all primary malignant bone tumours. Common in males. A number of histological subtypes have been described including (1, 2). Storiform-pleomorphic: most common 50-60%, Myxoid: 25%, myxofibrosarcoma, Inflammatory: 5-10%, Giant cell: 5-10%, Angiomatoid, Location: distal femur (most common) >proximal tibia>proximal humerus (rare).***

Keywords: Malignant primary bone tumour, Malignant fibrous histiocytoma.

1. Introduction

Malignant fibrous histiocytoma (MFH)is a rare malignant primary bone tumour that contains a mixture of fibrogenic cells that are histologically similar to histocytes. Histologically MFH appear as large multi-nucleated malignant cells, with abundant cytoplasmic nuclei.

Incidence: MFH constitutes less than 2% of all primary malignant bone tumours. Common in males.

A number of histological subtypes have been described including (1,2)

- Storiform-pleomorphic: most common 50-60%
- Myxoid: 25%,myxofibrosarcoma
- Inflammatory: 5-10%
- Giant cell: 5-10%
- Angiomatoid

Location: distal femur (most common) >proximal tibia>proximal humerus (rare).

2. Case Report

A 42 year old man was referred to us with sudden onset of left shoulder pain. Initially he was diagnosed with a rotator cuff tendinopathy for which he received conservative treatment. After a short duration, he was involved in a road traffic accident, which aggravated the symptoms. Roentgenography of left shoulder was done, which showed a destructive lesion at metaphysis with cortical breach noted in the medial aspect of humeral surgical neck. Further anatomic imaging showed an aggressive tumor mass in the proximal diaphysis of humerus involving the humeral head and adjacent soft tissues.Lab parameters were normal and distant metastasis were ruled out. Biopsy was done and it showed a "High grade pleomorphic sarcoma".Patient received a course of chemotherapy, following which he underwent an en block resection of the tumor. Intra op specimen was sent for histopathological examination and it confirmed the diagnosis of "MALIGNANT FIBROUS HISTIOCYTOMA".

3. Conclusion

Treatment for MFH is similar to that of Osteosarcoma which involves chemotheraphy followed by surgical removal of the tumour. Prognosis is determined by the grade of tumour and the stage in which the patient presents to us. In general, poorer prognosis is associated with stage III or stage IV disease (5) A tumour located superficially in the subcutaneous tissues of the distal extremity, and measuring less than 5 cm, has a 5-year survival of 80%, whereas a proximal large (>5 cm) and deep tumour has a 5-year survival of 55% [5]. Other associated factors for a poorer prognosis are Age more than 60 years, Tumor over 5 cms in size, Distant metastasis and Local recurrence. Low grade tumors such as grade I and grade II, usually respond well to surgery and has good prognosis and reasonably well functional outcome.

4. Images

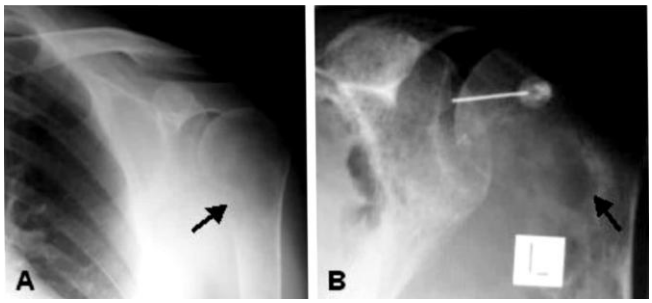

Image A & B: Roentgenography of Left shoulder shows -> Destructive lesion at metaphysis of the proximal humerus with cortical breach noted in the medial aspect of humeral surgical neck.

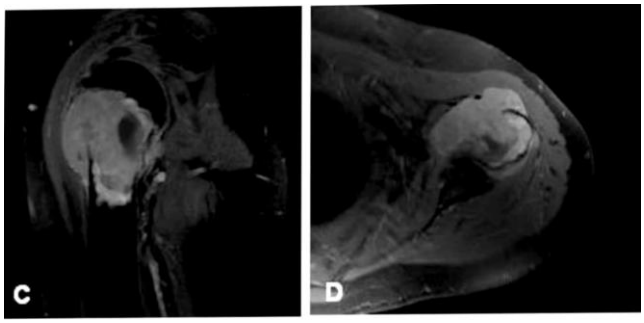

Image C & D: MRI of Left shoulder shows ->aggressive tumor mass in the proximal diaphysis of humerus involving the humeral head and adjacent soft tissues

MRI is the modality of choice for assessing soft tissue sarcomas, as it is best able to locally stage a tumour. These tumours are typically relatively well circumscribed, located within or adjacent to muscle, exerting a positive mass effect on surrounding structures due to their (usual) large size at presentation.

a) T1

- Intermediate (to low) signal intensity, similar to adjacent muscle [3,4]
- Heterogeneity if haemorrhage, calcification, necrosis, myxoid material present
- Prominent enhancement of solid components

b) T2

- Intermediate to high signal intensity
- Heterogeneity if haemorrhage, calcification, necrosis, myxoid material present

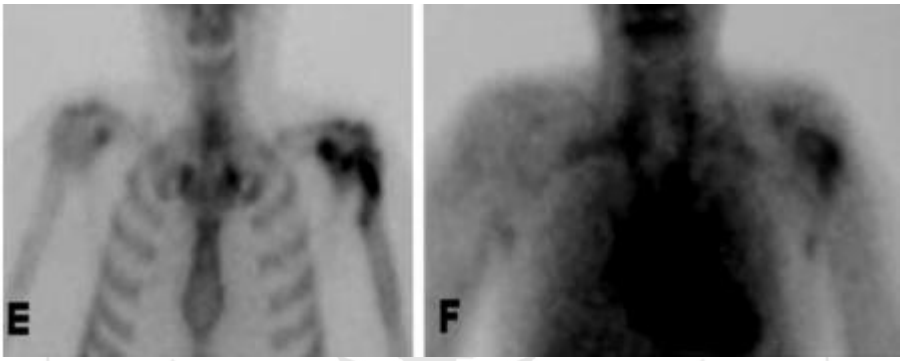

Image E & F: Bone scan shows ->Tumor involving Left Proximal Humerus without metastasis.

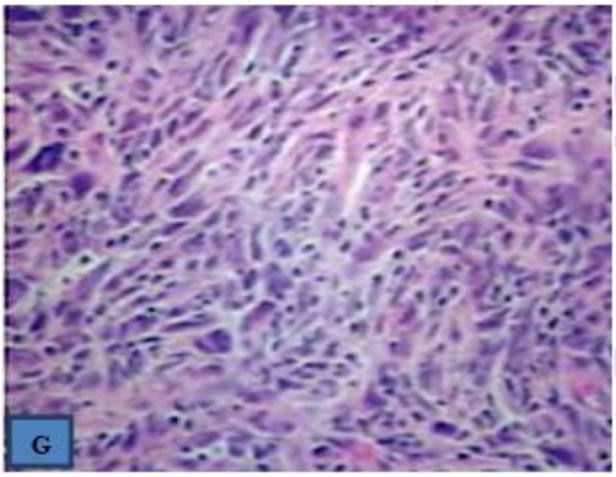

Image G: Microscopic image shows ->Tumor tissue composed of large bizarre multinucleated cells with foamy cytoplasm, with delicate capillary vasculature. In areas, there is spindle cells with storiform pattern along with mixed inflammatory cells. Scattered pleomorphic pseudo-lipoblasts are seen.

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