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Chondromyxoid fibroma of Ulna: A case report

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Abstract
Chondromyxoid fibroma is a rare benign cartilaginous tumour accounting to less than 1% of bone tumours. Most commonly seen in lower extremity involving tibia. Chondromyxoid fibroma of ulna is rare. We report a rare case of chondromyxoid fibroma of proximal ulna in a 34-year-old female who presented with dull aching pain over left elbow. Planned for staged procedure. After confirming the diagnosis with needle biopsy, an extended curettage with bone grafting and plate stabilization was done.

Keywords: Chondromyxoid fibroma, radiologic appearance, histopathological, pathophysiology

Introduction
Chondromyxoid fibroma is a benign cartilaginous tumor of young adults, arising in proximal or distal parts of long bones, showing zonal architecture. Local recurrence is common; malignant transformation is extremely rare. Patients presents with pain, often of long duration. Characteristic radiologic appearance is eccentric lytic lesion with sharp, sclerosed and scalloped intramedullary edges. Histologically, zonal architecture comprised of lobules of myxoid to chondroid tissue with intervening spindle and multinucleated giant cells, showing variable coarse calcifications. Pathophysiology - upregulation of glutamate receptor gene GRM1 coding region of chromosome 6 through recombination with several partner genes in up to 90% of cases. Diagnosis requires integration of radiological and histopathological findings.

Case Report
A 34 year old female presented with left elbow pain of 6 months duration. It was a dull aching type of pain. There was no history of trauma. On examination, diffuse tenderness was felt over the proximal aspect of ulna. No swelling was palpable. No local rise of temperature and no neurological deficits. Elbow range of movements was normal.
Radiographs of left elbow revealed an expansile lytic bone lesion at metaphyseal region with multiple incomplete septations. No cortical break. No involvement of articular surface. MRI of left elbow revealed T2W hyperintense and T1W hypointense expansile lytic lesion with multiple incomplete septation - suggestive of simple bone cyst. Routine biochemical investigations showed ESR of 8, serum alkaline phosphatase was 80 IU/L. Other blood tests within normal limit. No features of hyperparathyroidism. After proper counselling, planned for staged procedure. Needle biopsy was done and specimen was sent for histopathological examination.

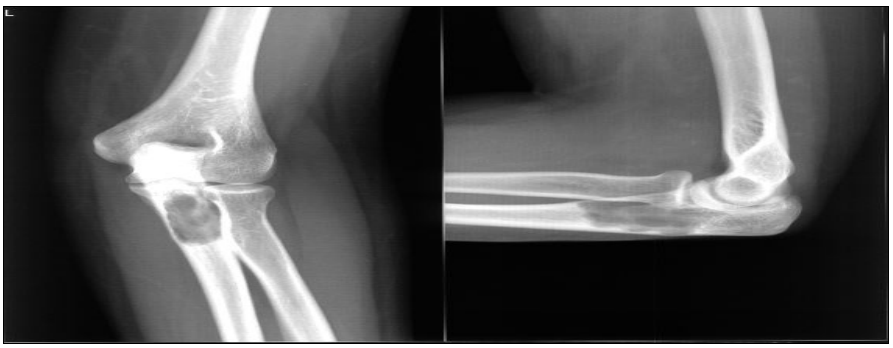

Fig 1: Pre-operative X- ray

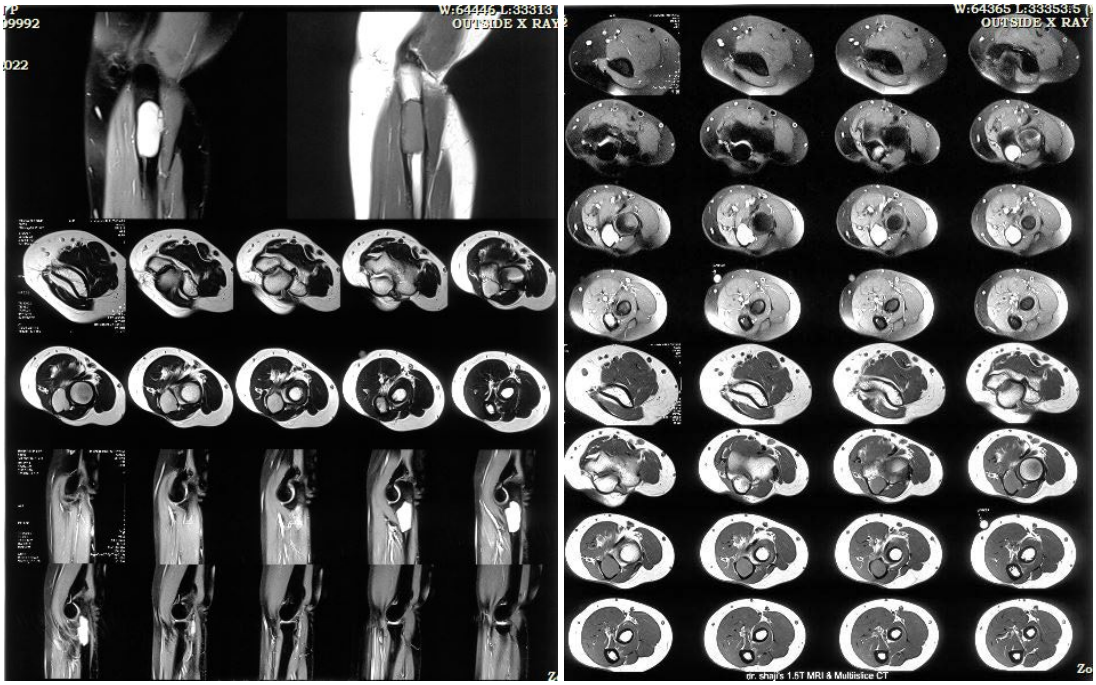

Fig 2: MRI of elbow joint showing expansile lytic lesion with multiple incomplete septations

On microscopic examination, section showed fragments of neoplasm composed of spindle to stellate cells arranged into vague lobules with central hypocellular and peripheral cellular areas. Spindle cells are embedded in myxohyaline stroma. Focal areas of calcifications noted in lobules. No atypia or increased mitosis seen. So, final diagnosis offered was chondromyxoid fibroma ulna.

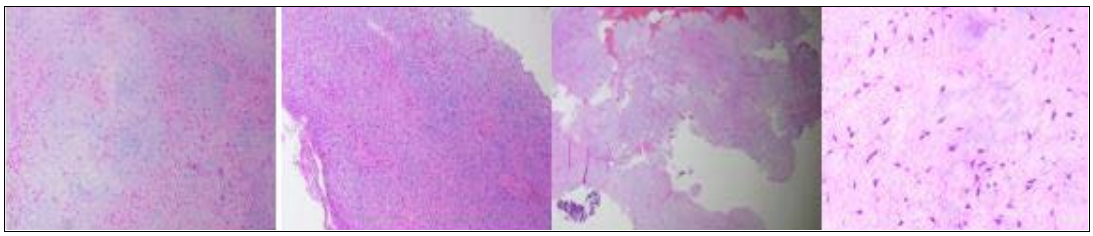

Fig 3: Histopathological image

Extended curettage with bone grafting and plate stabilization was done. An above elbow POP slab was applied and elbow range of movements exercises were started after one month. Patient was followed up for 6 months. Patient had a good range of painless functional movements and was able to carry out her day today activity without any assistance.

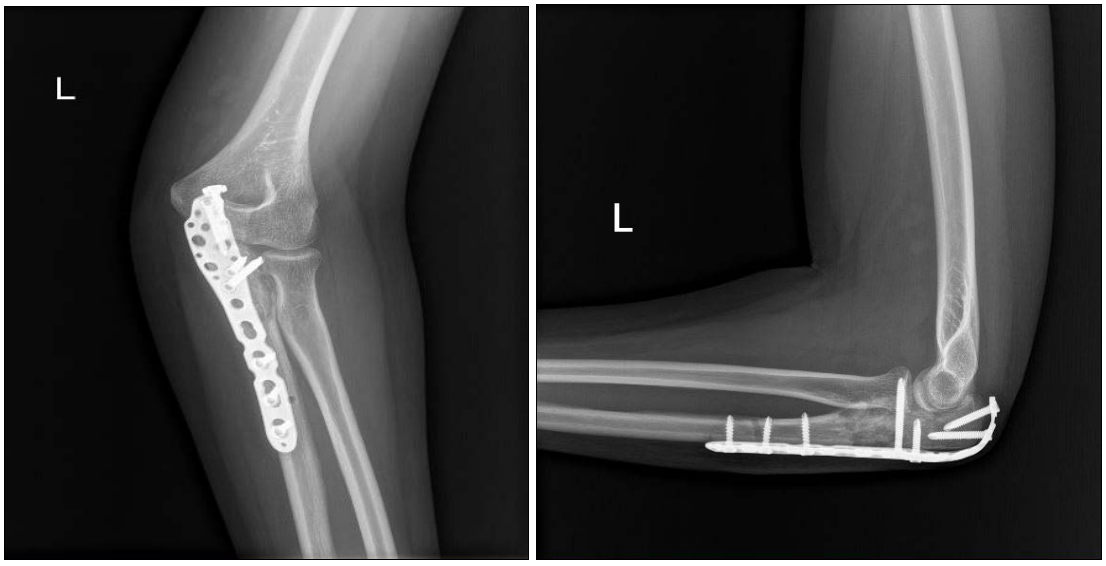

Fig 4: Immediate post op x-ray

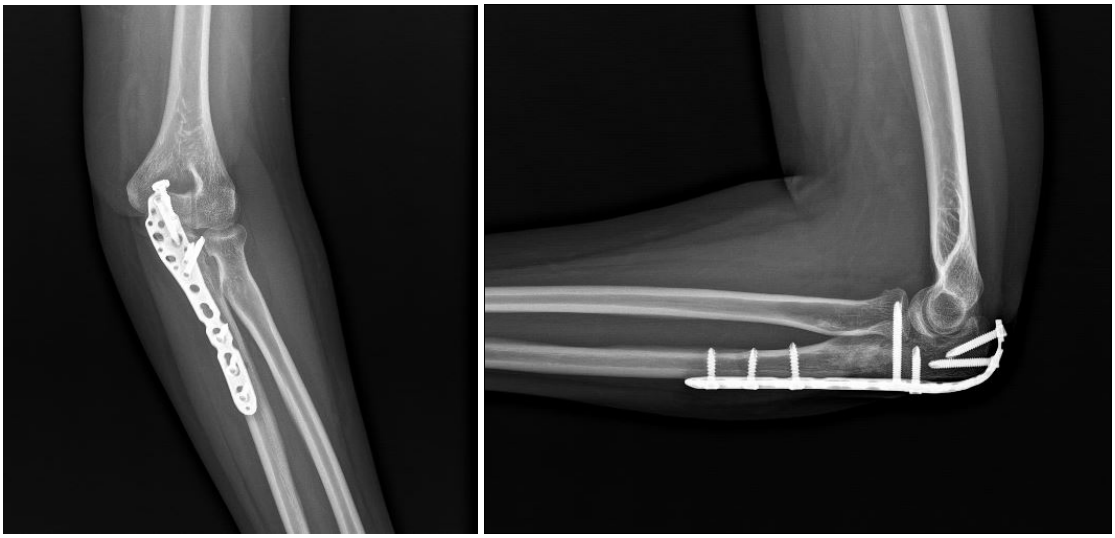

Fig 5: X-ray - 6 months follow up

Discussion

Chondromyxoid Fibroma (CMF) is a benign and extremely rare tumour accounting for less than 1% of benign and malignant bone tumours. It is a benign cartilaginous tumour. CMF predominantly affects adolescents and young adults in the second or third decade of life. CMF is often confused with other radiological and pathological mimickers. Histological evaluation remains a key in diagnosing this rare pathology.

CMF is most commonly seen in the lower extremity, particularly the proximal end of tibia. 95% cases of CMF are seen in long bones involving metaphyseal region. Less common sites are the sacrum, thoracic or lumbar spine and craniofacial bones [1]. In a study of 278 cases of CMF by Chen Tu Wu *et al*., 46.9% of cases involved long bones, 30.3% flat bones, 17.3% involved bones of hand & feet, and 15% skull and facial bones. Out of 46.9% long bone lesions, 55.4% involved tibia, 19.2% femur, 10.8% fibula and 3.1% radius [4].

The clinical presentation varies according to the area involved and is associated with long standing history of non-specific symptoms like pain and oedema. Usually CMF is slow growing tumour and detected incidentally on routine radiography. There is a long history of chronic local pain, swelling and oedema with palpable soft tissue mass and restricted movements in asymptomatic patients.

Radiologically these tumours are described as an eccentric, lobulated, expansile and lytic lesion with well-defined scalloped or lobulated margins with sclerotic bone formation. Radiographic calcification is noted in 10% of cases. Partial cortical erosion and septations are noted in few cases [2]. In present case MRI scan showed cortical erosion and septations in meta-epiphyseal region of proximal radius. Differential diagnosis on radiology are giant cell tumour, aneurysmal bone cyst, simple bone cyst, chondroblastoma and fibrous dysplasia [3]. In our case, based on MRI, the radiological diagnosis was simple bone cyst.

Grossly, the tumor is firm, grayish-white in color which may be lobulated or pseudo lobulated. Structure resembles that of fibrous tissue or hyaline cartilage. The lesion often thin the cortex and rarely destroy trabecular bone.

Classic microscopic appearance of CMF is mixture of fibrous, myxoid and cartilaginous elements with several nodules composed of rounded areas of myxoid or chondroid tissue. Stellate to spindle shaped cells are noted in the centre of lobule with characteristic high cell density towards the periphery of lobules. Multinucleated giant cells are noted in the periphery in few cases [5]. On microscopy indexed case showed fragments of neoplasm composed of spindle to stellate cells arranged into vague lobules with central hypocellular and peripheral cellular areas. Spindle cells are embedded in myxohyaline stroma. Focal areas of calcifications noted in lobules. No atypia or increased mitosis seen

Histopathological differential diagnosis of CMF is myxoid chondrosarcoma, Giant Cell Tumor (GCT) and chondroblastoma. Chondrosarcoma show lobules which are more distinct with presence of plump, bizarre cells with multiple nuclei. Presence of bubbly appearance of stroma with degenerative and liquefactive changes favours the diagnosis of myxoid chondrosarcoma over CMF. Chondroblastoma occur in skeletally immature and histologically show classic “chicken-wire” calcification. Multinucleated giant cells with nucleus identical to the background nucleus of the stromal cells are the major histologic characteristic of GCT. In CMF, the giant cells are few, which are located in the periphery.

Accepted treatment for CMF is surgical curettage with bone grafting but recurrence rate is more. Preferable treatment for CMF of long bones is enbloc resection with allograft and artificial bone grafts. Radiotherapy may be considered an option for unresectable tumours. In expendable bones such as fibula, en block resection is the ideal treatment of choice. Malignant conversion following en block resection is rare. In present case extended curettage with bone grafting and plate stabilization was done to minimize risk of recurrence.

Conclusion

Chondromyxoid fibroma is a rare benign aggressive cartilaginous tumour usually involving metaphysis of long bones. Radiological findings often mislead clinicians, as in our case it was reported as simple bone cyst. Histological evaluation remains a key in diagnosing this rare pathology. As recurrence rate of chondromyxoid fibroma is high, correct diagnosis and extended curettage is required with regular follow up. En block resection is curative for expendable bones like fibula.

Declaration of conflict of interest
The author (s) do not have any potential conflicts of interest with respect tthis manuscript.

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Declaration of informed consent
The author (s) hereby declare that there is no information (names, initials, hospital identification numbers or photographs) in the submitted manuscript that can be used to identify patients.

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