A Rarest Presentation of Chondromyxoid Fibroma in Proximal Ulna

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Abstract: Chondromyxoid fibroma (CMF) is one of rarest benign tumours of cartilaginous origin. It accounts for less than 0.5% of bone tumours and less than 2% of benign tumours. It mainly affects the metaphysis of long bones of lower extremity, the proximal tibia, distal Femur being the most common location and rare in upper extremity (ulna bone). After taking informed consent from patient for this study, we present a case of chondromyxoid fibroma in a 12 year old female involving right proximal end of ulna which was treated with En bloc Excision and filling with bone grafting (PMMA) With excellent result on Follow up.

Keywords: Chondromyxoid fibroma, proximal ulna, benign tumor, En bloc Excision and Bone Grafting

1. Introduction

A rare and benign chondrogenic lesion characterized by variable amounts of chondroid, fibromatoid and myxoid elements. Most cases are characterized by GRM1 gene fusion or promoter swapping. It can be associated with a translocation at t(1;5)(p13;p13). It mostly occurs in patients who are 10 to 30 years old and presents more commonly in men than in women. In pathological study it is composed of a mixture of chondroid, myxoid, and fibrous tissue. The diagnosis of CMF depends on its characteristic histological appearance like a lobular pattern with stellate shaped cells in a myxoid or chondroid back ground. There are various treatment options for this condition such as, which include curettage alone, curettage with phenol, and en bloc resection with bone grafting (PMMA). Prognosis of this tumor depend on natural history and recurrence in CMF is not uncommon may occur in 20-30% of cases.

2. Materials and Methods

A 12 Yr Old Female patient presented with 3 months history of pain at the proximal end of right ulnar the pain was a dull aching mild intensity continuous an increasing in intensity and non radiating without any diurnal variation and aggravates on movement and relieves on rest. patient given history of edema at proximal end of right fore arm started gradually one and half month after the onset of pain.no history of trauma, weight loss, similar complaints in family. On general examination of patient is moderately built and nourished, afebrile, vitals stable. On inspection skin over lesion is normal, no stretched, no redness, scar, sinus.

Blood investigations shows Hb:13.5 g/dl, WBC 9, 700 cells/cumm, platelets 2.3 Lakhs /cumm, Neutrophils 45 %, Eosinophils 05%, lymphocytes 45%, monocytes 05%, Basophils 00%, CRP 6 mg/dl, S.ALP 99 U/L, S.Uric acid 2.5 mg/dl, s. creatinine 0.4. On palpation localized swelling of approx. 8cm x 5cm over right medial aspect of proximal ulna, no local rise in temperature, tenderness over swelling present, no distal neurological deficit seen. Range of movement of right elbow is non restricted.

X-ray right elbow shows osteolytic destruction of posterior cortexradiolucent lesion in metaphysis origin in proximal third of ulna with no periosteal reaction.

On radiological findings
On CT scan image findings:

Irregular expansile osteolytic lesion with narrow zone of transition with adjacent soft tissue component noted in proximal shaft of ulna (metaphyseal region) posteriorly, approximate size of lesion 38mm (length) x 26mm (AP) x 18mm (transverse) shown by arrow in the image.

**Intraoperative image of en bloc resection with bone grafting:** After obtaining written consent from the patient, en bloc excision with bone grafting done. Excised lesion sent for histopathological examination.
Image of bone graft used:

Synthetic bone graft made of hydroxyapatite, later it absorbs and replaced as natural bone heals over a few months time. The principles involved in successful bone grafts include osteoconduction, osteoinduction and osteogenesis.

Postoperative radiograph:

Postoperatively patient immobilized in acylindrical plaster cast followed by movement of joint at 3 to 4 weeks.
3. Discussion

Chondromyxoid fibroma (CMF) is a benign, locally aggressive tumor of cartilaginous origin and accounts for less than 0.5% of all bone tumors. In 1948, the tumor was first described by Jaffe and Lichtenstein as a lesion derived from cartilage-forming tissue and composed of various proportions of chondroid, fibrous, and myxoid tissues. The common site of the tumor is the metaphysis adjacent to the epiphyseal growth plate which indicates it arises from cartilage at these sites [4]. For establishing the diagnosis, a thorough clinical, radiological, and pathological examination is important as it might easily be misdiagnosed as other malignant tumors such as chondrosarcoma because of some pathological similarities.

Patients typically complain of pain and swelling at the site of the lesion. The pain is usually mild, intermittent, and a dull ache, no diurnal variation not associated with fever as seen in our case. On some occasions, the tumor may be asymptomatic and may present as an incidental finding on radiographic examination.

The radiographic picture shows an eccentrically located lesion in a large limb bones, as seen in our case. When the tumor involves a smaller tubular bone like the rib or the fibula, the radiographic appearance is not typical because the lesion may extend throughout the entire width of the affected bone, expanding both surfaces and often making diagnosis between fibrous dysplasia and chondroma difficult. In this case MRI useful to know extent of lesion in bone.

Diagnosis of CMF basically depends on its characteristic histological appearance. The typical histological features of CMF are a lobular pattern with stellate-shaped cells in a myxoid or chondroid background with hypocellular centres and hypercellular peripheries. The differential diagnosis of CMF includes chondroblastoma, chondrosarcoma, enchondroma, and aneurysmal bone cyst, but it is the salient histological features that distinguish these lesions.

The treatment options of CMF include simple curettage, curettage with phenol application, and en bloc resection with bone grafting. Curettage with phenol application followed by bone grafting has a very low rate of recurrence of seven percent. Further reduction in recurrence rate was observed when the lesion was treated with en bloc excision and bone grafting. So, our patient was treated by en bloc excision with bone grafting, which got incorporated very well, and the lesion has shown no signs of recurrence.

4. Conclusion

Chondromyxoid fibroma is an uncommon benign bone neoplasm, and uncommon in upper extremities than lower extremities. Sometimes no symptoms seen in patient and it accidentally finds in radiograph, so proper history of patient and stepwise diagnosis is required for this tumor for not to confuse with other tumors.

References


