

Case Report

Surgical management of chondromyxoid fibroma at meta-diaphyseal junction of proximal tibia: a case report

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ABSTRACT

Chondromyxoid fibroma (CMF) is a rare, benign tumour of the bone. It represents less than 1% of all bone tumours. We present a case of a 20-year-old female who came with chief complaints of pain and swelling in right leg for 3 months. Radiograph showed features suggestive of large multiloculated osteolytic, radiolucent lesion with thinned out cortices in right proximal tibia. MRI showed exophytic lesion and features suggestive of osteosarcoma. USG guided biopsy reported as chondromyxoid fibroma. Patient underwent curettage of tumour followed by reconstruction with allograft and stabilized with internal fixation. Final histopathological report showed features suggestive of chondromyxoid fibroma. Patient made uneventful recovery with no signs of recurrence and good functional range of motion (ROM) of the knee joint after 2 years of follow up. Histopathology is the gold standard and mandatory for confirmation of the diagnosis. Curettage and filling the defect with polymethyl methacrylate (PMMA) or bone graft is the mainstay treatment of choice.

Keywords: Chondromyxoid fibroma, Bone tumour, Curettage, Polymethyl methacrylate, Allograft

INTRODUCTION

Chondromyxoid fibroma (CMF) is a rare benign tumour apparently derived from cartilage-forming connective tissue. CMF accounts for less than 1% of all bone tumours.¹ Previously it was considered to be myxoma or myxomatous variant of giant cell tumour.² CMF may occur at any age, more commonly affecting second to third decade of life, with slight male predominance.^{3,4} About two-third of all cases arises from tubular long bones most commonly metaphyseal region with approximately one-third of them arising from tibia.^{1,3} The proximal tibia is the most common site of involvement followed by distal femur.⁴ Clinically pain is the most common presenting symptom associated with localized slow growing mass.² On radiography appears as a well circumscribed lesion with a rim of sclerosis.⁵ There is no specific management protocol for chondromyxoid fibroma due to limited published articles and research. Treatment consists of resection or extended curettage with bone grafting. Bone grafting and/ or PMMA reduces the risk of recurrence.

Radiation therapy is not indicated except for very rare surgically inaccessible tumour.¹ Wide resection is associated with a lower risk of recurrence than is intralesional curettage but results in a potentially larger functional deficit.⁴ Local recurrence varies from 3-22%. Malignant transformation of CMF is rare and seen in 1-2% of cases. Distant metastases have not been reported.³

CASE REPORT

A 20-year-old, female presented to OPD with complaints of Pain in right leg for 3 months, which was insidious in onset, gradually progressive, dull aching type associated with swelling at proximal aspect of right leg. There was no history of fever, weight loss, morning stiffness, loss of appetite, evening rise of temperature. On examination there was no local rise of temperature, a solitary mass of 10×8 cm present at anterolateral aspect of right proximal leg, skin over the swelling appears normal, diffuse indistinguishable margins, tender with smooth surface and hard in consistency. Radiograph (Figure 1) revealed a

large multiloculated osteolytic, radiolucent lesion with thinned out cortices present over proximal one-third of right tibia at meta-diaphyseal junction, sparing epiphysis. MRI (Figure 2) showed exophytic mass lesion in the posterior aspect of proximal tibia. STIR hyperintense marrow edema was seen in proximal tibia and was reported as features suggestive of osteosarcoma. In order to confirm our diagnosis, we went ahead with USG guided biopsy (Figure 3) for the tibial mass reported as benign bone tumour suggesting features of chondromyxoid fibroma.



Figure 1: X-ray showing large multiloculated osteolytic lesion in proximal metaphyseal region of tibia.

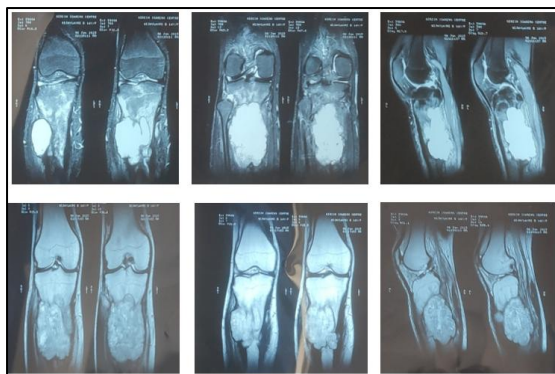


Figure 2: MRI showing exophytic growth in proximal tibia.

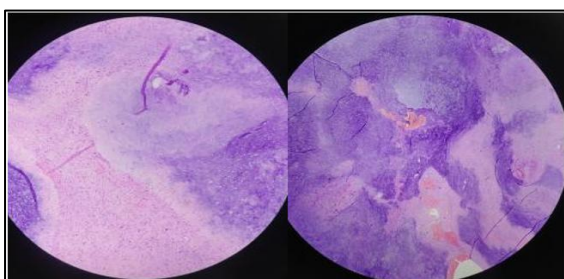


Figure 3: Microscopy showing hypocellular chondromyxoid fragments with the embedded stellate cells amidst which are a few moderately cellular fibromyxoid fragments having the spindled cells depicting a bland nucleus.

Surgical procedure

Under spinal and epidural anaesthesia, patient placed in supine position on a radiolucent table. Under aseptic precaution, parts painted and draped. Tourniquet inflated, a 14 cm skin incision taken curving the incision anteriorly over Gerdy tubercle and then extend it distally via antero-lateral approach.

Incision deepened as there is no internervous plane in this approach. Fascia overlying the tibialis anterior muscle incised and the muscle belly mobilized from the lateral aspect of the tibial shaft. Tumour visualized appeared grey-white to grey-brown colours (Figure 4a). Curettage was done followed by high-speed burr to clear tumour from its cavity (Figure 4b). The sample was sent for histopathology. The defect was reconstructed with allograft (Figure 4c) and stabilized with 4.5 mm proximal tibial plate and screws (Figure 4d) under fluoroscopic guidance. Drain placed and incision closed in layers.

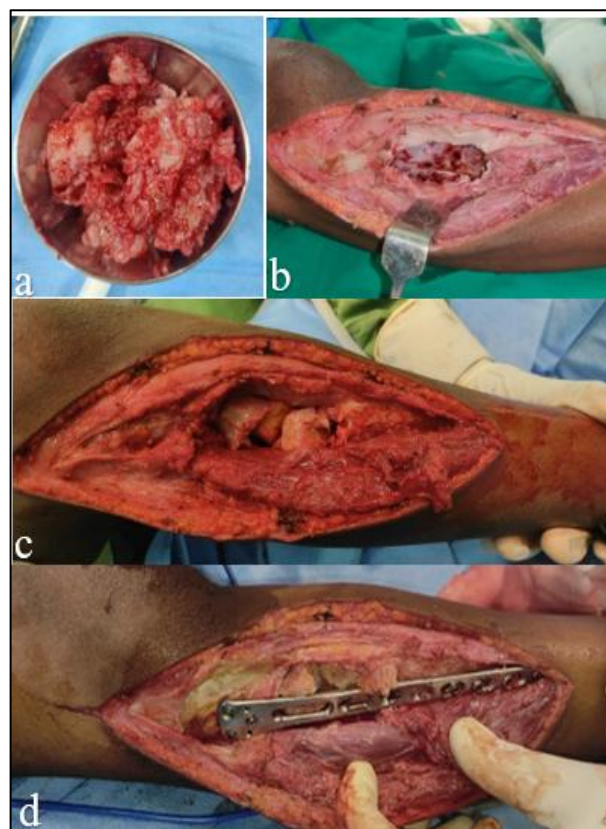


Figure 4: intra operative findings; (a) excised tumour appears grey white to grey brown; (b) defect in proximal tibia after tumour excision; (c) defect filled with allograft; (d) 4.5 mm plate fixation.

Post op

Final histopathology (Figure 5) report showed features suggestive of chondromyxoid fibroma. Post operative radiograph showed no remnant tumour (Figure 6a). Drain was removed on POD 3 and started on non-weight bearing

mobilisation with support. Complete suture removal was done after 3 weeks (Figure 6c) and knee ROM was started. After serial post operative radiographs monthly, she was advised partial weight bearing after 3 months and gradually progressed to full weight bearing without support. On follow up, patient is currently pain-free and knee ROM 0-110 degree (Figure 7). She's able to carry out activities of daily living independently. There's no signs of recurrence and good functional range of motion (ROM) of the knee joint after 2 years of follow up.

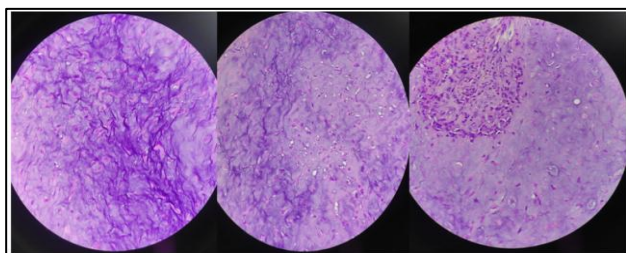


Figure 5: tumour cells arranged in lobules separated by mononuclear spindle cells. The lobules are composed of hypocellular centres and hypercellular periphery with tumour cells which are oval to spindle shaped having moderate amount of eosinophilic cytoplasm. The adjacent stroma is myxoid to chondroid. No evidence of atypia seen.



Figure 6: (a) Post operative X-ray; (b) post operative wound; (c) wound after suture removal.



Figure 7 (a-c): Follow up after 3 month shows Knee ROM 0-110 degree with no extensor lag.

DISCUSSION

CMF is a rare, benign and locally aggressive lobulated cartilaginous neoplasm with zonal architecture composed of chondroid, myxoid, and myofibroblastic areas.⁵ The tumour was first described by Jaffe and Lichtenstein in 1948 when they presented 8 cases and emphasized the danger mistaking this benign neoplasm for a malignant lesion, especially chondrosarcoma.¹

WHO describes CMF as a benign tumour characterized by lobules of spindle or stellate-shaped cell with abundant myxoid or chondroid intercellular material.³ The peak incidence was in the second and third decades of life. Patients in these two decades formed 50% of the entire group. Approximately 72% of the patients were younger than 40 years.⁶

CMF is commonly found in metaphyseal region of long bone. The most common site is the proximal tibia, estimated at 28%–52% of all lower extremity based on literature.⁷ The small bones of foot involvement are not uncommon.¹ Other sites of involvement include flat bones ilium, scapula and rarely ribs, sternum and clavicle. Most of the lesions are medullary in location (95%), but rarely subperiosteal, juxtacortical and intracortical occurrences are also known. CMF is associated with clonal abnormalities of chromosome 6.³

Occasionally, these tumours are asymptomatic incidental findings on radiography.¹ Radiographic features show geographic bone destruction, with either partial or complete erosion of the cortex.⁸ It may mimic non-ossifying fibroma by its bubbly appearance. Unlike other cartilaginous lesions, radiographic evidence of intralesional calcification usually is uncommon.⁴ They may appear as “bite-like” destruction. Cortex may be thinned out with mild expansion or break in the cortex with an extraosseous soft-tissue component.³

However approximately 10% of cases may show focal calcified matrix, more often detectable on CT.⁵ On MRI, CMF shows multilobulated pattern, low signal intensity on T1- weighted images and high signal intensity on T2-weighted images. With Contrast, central portion of lesion may show no enhancement due to the myxoid component.²

Gross features of CMF include well defined margins, grey or white tumour, cystic change, lack of obvious necrosis, and liquefaction. Tumour is typically well demarcated, multilobulated and scalloped margins.⁵ Microscopically, chondromyxoid fibroma appears lobulated. The center of the lobules contains loose myxoid tissue composed of stellate or spindle shaped cells, and the periphery contains a more cellular fibrous tissue. The background often appears chondroid, although distinct areas of hyaline cartilage are rare. Microscopic calcification may be present. The lesion may contain areas with atypical pleomorphic hyperchromatic nuclei, but this should not lead to the erroneous diagnosis of chondrosarcoma.^{3,4}

There are no major recommendations of CMF management due to the tumour being extremely rare. The management options include curettage and excision, with or without filling of the cavitary defect. Wide resection or en-bloc excision is probably the best method to avoid recurrence, but not all locations allow the mechanical imbalance these procedures can cause, so bone grafting is advised.⁹

With curettage alone, a 13-25% recurrence rate has been reported. The recurrence rate has been reduced with the use of allograft bone or polymethylmethacrylate.^{10,11} Gherlinzoni et al, recommend curettage with bone grafting as the first line of treatment. They found that the 80% recurrence rate in their series using curettage alone decreased to 7% with the addition of bone grafting. Till date, there has been no published study on metastases from CMF.¹² In the present case, we carried out tumour curettage and high-speed burr followed by filling the defect with bone graft. There is no sign of recurrence at 2 years post-operatively.

CONCLUSION

Chondromyxoid fibroma is a rare benign bone tumour. It is often misdiagnosed as malignant neoplasm. Histopathology is the gold standard and mandatory for confirmation of the diagnosis. Curettage and filling the defect with bone graft has shown good outcome and no recurrence.

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