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Case Report

Atypical chondroblastoma of the distal femur with exophytic mass: A rare presentation and diagnostic dilemma

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ABSTRACT

Chondroblastoma is a benign epiphyseal chondromatous lesion typically seen as a small, well-defined, eccentric epiphyseal lesion with thin rim of surrounding bone sclerosis. Soft tissue extension, metaphyseal involvement, periosteal reaction, cortex breach though not pathognomonic, are reported in literature. We present a case report of a patient with chondroblastoma showing atypical radiological presentation with exophytic mass in order to give reader a perspective on atypical radiological features and corroborating these finding intraoperatively and on histopathology.

1. Introduction

Chondroblastoma is an uncommon benign cartilaginous tumour, typically occurring in the second decade of life before skeletal maturity.[1,2] It is twice as common in males as is in females.[1] This tumour represents 1%–2% of all primary bone tumours and about 9% of all benign bone tumours.[1,3] The femur, humerus, and tibia harbour nearly 60 to 70% of these tumours.[1,2] Chondroblastoma is primarily an epiphyseal tumour but metaphyseal extension is also known. Radiographically, it presents as a well-defined eccentric lesion in the epiphysis of a long bone. The lesion is rimmed by a border of host bone sclerosis along with small punctate calcifications. Usually, the physis adjacent to lesion is open at the time of diagnosis. All these features when present are pathognomonic of chondroblastoma.[1,4,5] Infection, giant cell tumour, aneurysmal bone cyst[2] are a few common differentials for such lesions. Features such as soft tissue extension, metaphyseal involvement, periosteal reaction though not pathognomonic of chondroblastoma have been previously documented thereby adding aggressive lesions to our list of potential differential diagnoses. Furthermore, soft tissue extension as a exophytic mass in chondroblastoma is extremely rare and hence can prove to be a diagnostic challenge. An exophytic mass arising from the epiphysis and protruding towards the joint may also raise the suspicion of Trevor's disease (dysplasia epiphysealis hemimelica).[6] Here, we report a case of an unusual exophytic chondroblastoma with an atypical presentation proving to be a diagnostic dillemma.

2. Case Presentation

A 13-year-old boy presented with complaints of pain around posterior aspect of knee joint since 4 months aggravated since 15 days. Pain was dull aching in nature, aggravated on walking and squatting. There were no diurnal variations. No history of trauma, fever, significant weight loss was noted. On examination, tenderness was present over posterolateral aspect of the distal femur. There was no appreciable swelling but patient had 10 degrees of flexion deformity of

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knee with further range up to 140 degrees. Both medial and lateral hamstring tightness was present.

Anteroposterior and lateral radiographs of affected knee (Figure 1) showed a lytic lesion in lateral condyle of distal femur with some periosteal reaction. Magnetic resonance imaging (MRI) was than performed revealed well defined 1.6 x 1.8x 2.1 cm expansile partially exophytic cortical/subcortical lesion along the posterior inner lateral femoral physis and epiphysis with posterior soft tissue extension. Involvement of adjoining metaphysis and marked overlying cortical thinning was noted. The lesion showed T2 hyper-intensity and T1 hypo-intensity with heterogeneous moderate enhancement on post contrast images. Extensive perilesional marrow edema was seen (Figure 2). J needle biopsy was then performed and on histopathology, the lesion consisted of sheets of polygonal cell (chondroblasts), few giant cells. Islands of chondroid cartilage with lacelike calcification surrounding individual cells giving the classical "chicken wire" appearance characteristic of chondroblastoma was seen.(Figure 3).

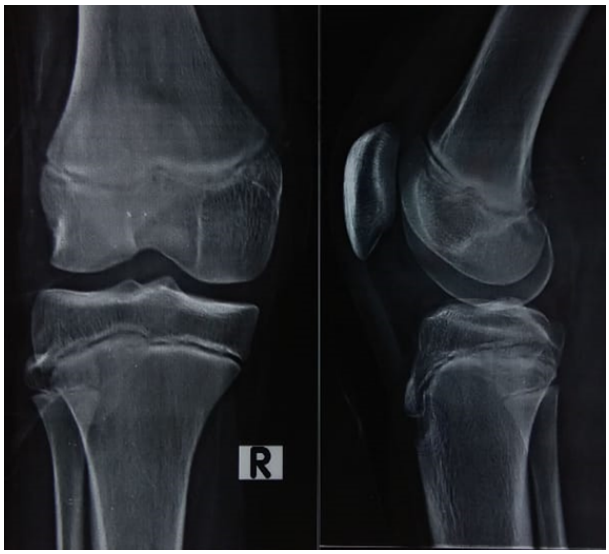

Fig. 1: Anteroposterior and lateral radiograph of knee showing lytic lesion on posterior aspect of lateral condyle with some periosteal reaction

After histopathological confirmation the patient underwent surgery in the form of extensive curettage by using high speed burr, where the entire exophytic mass was removed by posterolateral approach. (Figures 4 and 5) An above knee plaster of Paris (POP) slab was applied. Intraoperatively, a breach was found in the posterior cortex from which the exophytic mass was protruding out. Need of bone grafting was not deemed necessary. Postoperative radiograph of knee confirmed the removal of the lesion. After 2 weeks postoperatively, sutures and slab were removed and knee range of motion (ROM) was initiated without weight bearing. Full weight bearing allowed after 4 weeks.

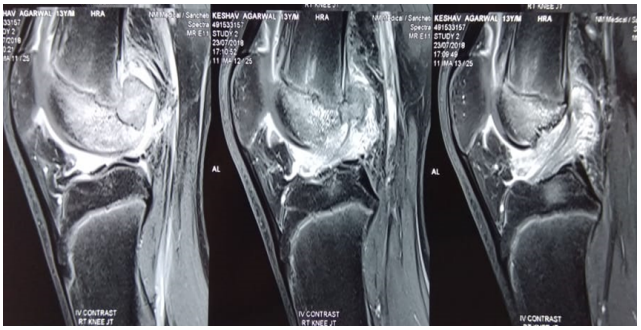

Fig. 2: MRI showing partial exophytic cortical/subcortical lesion along the posterior surface of lateral femoral condyle with extra osseous soft tissue extension

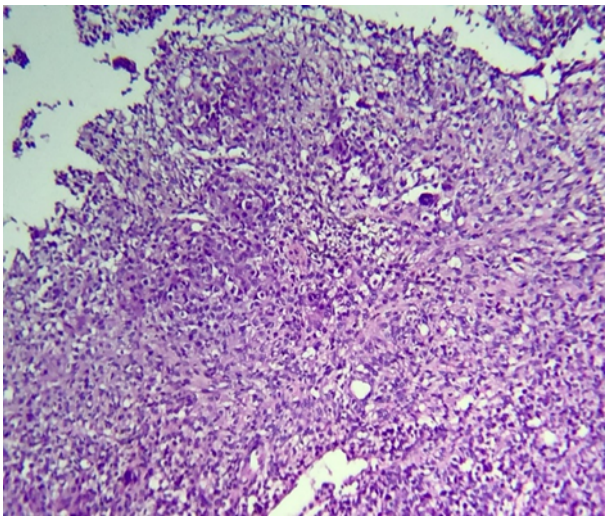

Fig. 3: Histopathology examination showing sheets of polygonal cell (chondroblasts), few giant cells, island of chondroid cartilage with lacelike calcification surrounding individual cells confirming chondroblastoma

3. Discussion

Chondroblastoma was first described in detail by Codman in 1931. He called the lesion an epiphyseal chondromatous giant cell tumour.[7] Further Jaffe and Lichtenstein in 1942 renamed the Codman tumour as a benign chondroblastoma to emphasize the chondroblastic genesis of the lesion and to distinguish it from the classic giant cell tumour of bone.[8] Typically, the chondroblastoma lesion seen as a small, well-defined, eccentric epiphyseal lesion. Sixty per cent of the lesions have a thin rim of surrounding sclerosis.[1] Flocculent calcific densities produced a mottled lesion in many of the cases.[4] Larger lesions commonly extend into the adjacent metaphysis and can erode or expand the overlying cortex. The metaphyseal portion of the tumour may not show the characteristic sclerotic rim on X-ray.[1,2] Lesions are usually

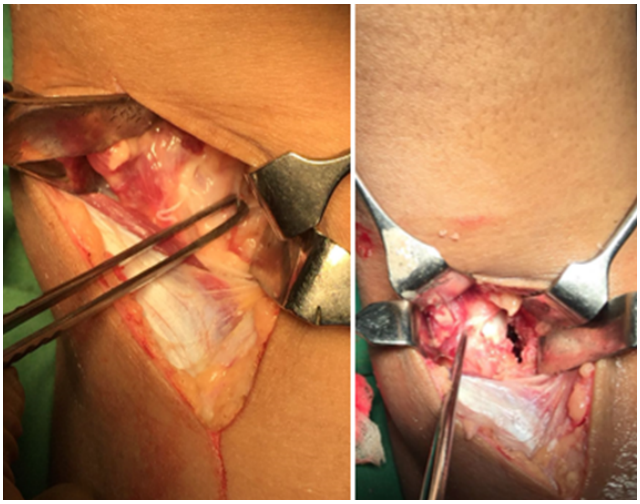

Fig. 4: Intraoperative images showing exophytic mass perforating posterior cortex of lateral condyle femur and cavity after curettage of lesion

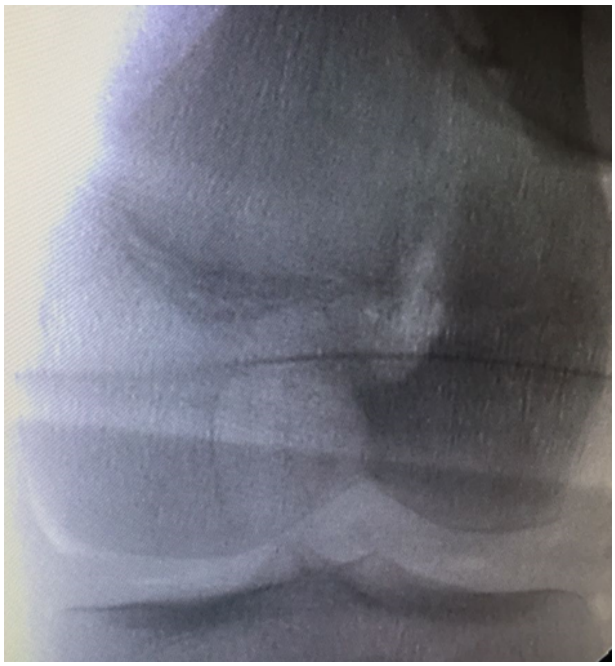

Fig. 5: Intra op C arm image showing removal of mass and hollow at lesion site

spherical or oval if cortex is not damaged. Once cortical extension occurs, either the tumour takes the shape of the cortex or retains its oval shape by expanding the cortex.[1] Cortical destruction is unusual, occurring in only 10% of cases.[9] Usually the margins of the lesions were smooth but some have scalloped margins.[1]

McLeod et al. in their study of 72 chondroblastoma lesions had reported that ten per cent were somewhat lobulated and five lesions presented with a "ballooned" or "blown-out" type of cortical appearance[1] but no further comprehensive radiological description or images were provided. Other Rare manifestations included extension into the adjacent joint, active periosteal new bone, pathologic fracture, trabeculations, cortical absence, and sclerosis beyond the confines of the tumour.[1,2] Dahlim et al in his study of 125 cases reported two chondroblastoma lesions with involvement of upper tibial epiphyses extending to become predominantly extraosseous lesions.[2] However detailed radiological descriptions were limited. Few authors reported aggressive chondroblastomas and primary pulmonary metastasis but these were a pathological rarity.[10,11]

Recently Karkhur et al reported a case of intra-articular chondroblastoma arising from proximal tibia in a 16-year-old boy and growing into the knee joint mimicking an intra-articular osteochondroma.[6] Dhanda et al reported a case of a patient with chondroblastoma showing atypical radiological presentation mimicking chondrosarcoma.[12] Gebert et al reported Chondroblastoma of the acromion mimicking fibrous dysplasia.[13] In our case features such as exophytic growth with cortex abutment and other radiographic features were unusual for a Chondroblastoma and thus can potentially misguide the treatment

4. Conclusion

Atypical presentations of chodroblastoma lesions can radiologically mimic aggressive tumours. Exophytic growth, cortex breach, soft tissue extension, metaphyseal involvement, periosteal reaction as presented in our case pose a challenge to reach an apt diagnosis. In the presence of above mentioned features one needs a high index of suspicion on imaging to correctly diagnose a chondroblastoma. Since the number of such cases reported in literature is few and radiological descriptions are limited these lesions may be misdiagnosed as more aggressive tumours. Incorrect diagnosis of malignancy may lead to inadvertent radical surgical management. Undoubtedly as a rule histopathological evaluation with biopsy is essential prior to definitive surgical management.

4.1. Clinical Message

Chondroblastoma can present with various atypical features inconsistent with classical clinical and radiological descriptions. It should be kept as a differential diagnosis even in seemingly aggressive lesions which otherwise point to a malignant disease.

Pre operative biopsy and histopathological diagnosis is of utmost importance before planning definitive management / surgery.

Consent : The patient has given their informed consent for surgery and case report to be published

All the authors meet each of the authorship requirements as stated by Editorial Board and all authors have read and concur with the content in final manuscript.

5. Competing Interests

The authors declare that they have no competing interests.

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