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Case Report

Pathology Section

Clear Cell Chondrosarcoma Masquerading as a Benign Lesion of Bone: A Rare Case Report

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ABSTRACT

Clear Cell Chondrosarcoma (CCC) accounts for about 2% of all chondrosarcoma subtypes and typically presents in individuals during their third to fourth decades of life. This type of tumour most commonly arises in the proximal epiphyseal region of the femur, followed by the humerus, tibia, and small bones. It is frequently observed in males and often manifests as a painful lesion that reduces mobility, leading to misdiagnosis as a benign condition. Histopathologically, the tumour is characterised by round to polygonal cells having clear to vacuolated cytoplasm, arranged in diffuse sheets and lobules, accompanied by reactive woven bone formation and rare mitotic activity. Due to high recurrence rates associated with conservative treatments such as curettage, wide excision of the affected bone followed by reconstruction is recommended. We report a rare case of CCC involving the proximal end of the femur in a 30-year-old male patient who presented with right hip pain for five months. Radiological investigations revealed a well-defined osteolytic lesion confirmed by MRI. Biopsy showed clear cells, reactive woven bone and a few multinucleated giant cells. Herein, we discuss the several differentials possible and discuss the challenges encountered during final diagnosis.

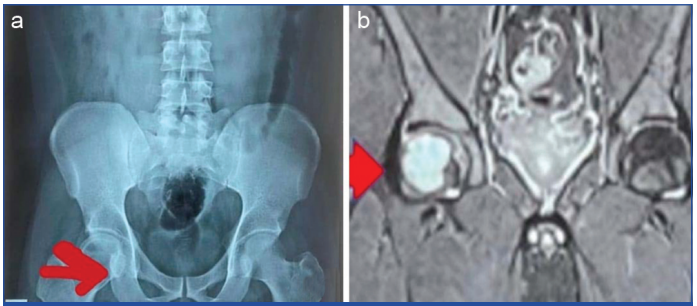
Keywords: Chondroblastoma, Epiphyses, Histopathology, Osteosarcoma

CASE REPORT

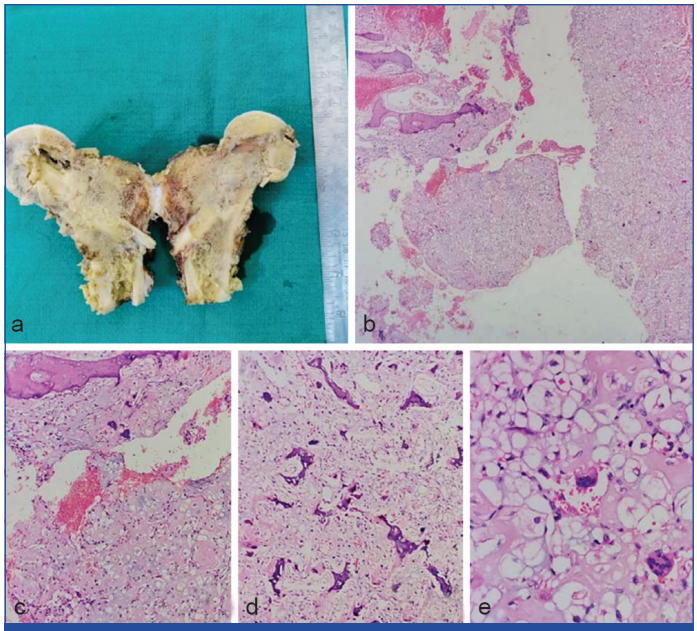
A 30-year-old male presented to the orthopaedics OPD with the chief complaints of right hip pain for five months and difficulty in walking for four months. The pain was insidious in onset, gradually progressive, and did not relieve despite taking analgesics and affected his daily routine activities. There was no history of trauma, fever, or any other comorbidities. Family history was unremarkable. His general and systemic examination showed no significant findings. However, there was mild tenderness and swelling at the right hip with reduced mobility at the knee and ankle joint. Sensory examination was intact. His haematological and biochemical investigations were within normal limits. Basic complete blood counts and biochemical tests like liver, renal function, thyroid function and lipid profile with serum calcium were also within normal limits. Plain X-ray of the right hip showed a well-defined osteolytic lesion over the medial aspect of the right femoral head [Table/Fig-1a]. Magnetic Resonance Imaging (MRI) of the right hip revealed an expansile, well-circumscribed T2 hyperintense lesion measuring 2×1.8×1.5 cm over the medial aspect of the right proximal femur with surrounding mild oedema [Table/Fig-1b]. There was no evidence of periosteal reaction or invasion into the adjacent soft tissues on radiology. With the clinical and radiological suspicion of malignancy, extensive curettage was done and a fibular graft was placed, following which the curettage tissue was sent for histopathological examination [Table/Fig-2a]. Multiple bits of greyish brown tissue structures, along with bony tissue received altogether, measuring 3×2.5×1.5 cm. Microsection showed tumour cells arranged in diffuse sheets and a lobular pattern [Table/Fig-2b]. Individual tumour cells were medium to large-sized, oval to polygonal in shape, having abundant amounts of clear cytoplasm with a sharply defined cytoplasmic border. Tumour cells had centrally placed round nuclei having vesicular chromatin, with few showing prominent nucleoli [Table/Fig-2c,e]. Mitosis was sparse. Reactive woven bone formation was seen surrounding the tumour cells [Table/Fig-2d]. There were numerous multinucleated giant cells scattered along with bony trabeculae within the tumour tissue [Table/Fig-2e]. With the differential diagnosis of chondroblastoma, giant cell tumour, and chondroblastic osteosarcoma, definitive excision of the proximal femoral head with femoral head reconstruction was done. An entire proximal femoral head was received, which showed a well-circumscribed greyish white lesion in the proximal epiphyseal region without any adjacent tissue invasion [Table/Fig-2a]. The final histopathology was confirmatory, hence a final diagnosis of CCC of the right femur was made. The patient on an 18-month follow-up postoperatively showed no signs and symptoms of recurrence, with the ability to walk and perform daily routine activities, with postoperative radiograph showing intact proximal femoral constructive plates [Table/Fig-3].

DISCUSSION

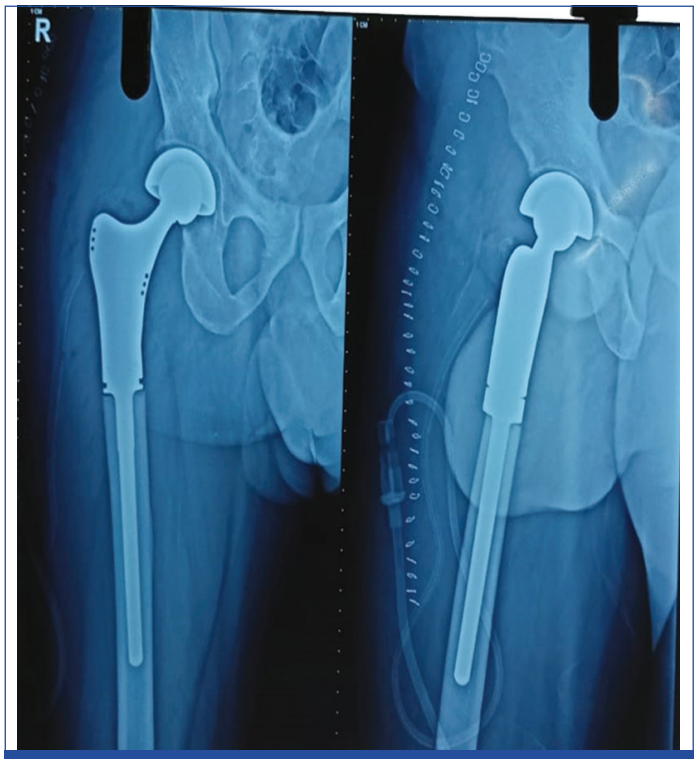
Chondrosarcoma, a primary malignant tumour of cartilage, accounts for approximately 20-25% of all primary malignant bone tumours [1]. CCC, a rare variant, constitutes about 2% of all chondrosarcoma types [2]. CCC typically occurs in long bones or the pelvis, with the proximal end of the femur being the most common site, followed by the humerus, tibia, and other small bones [3]. This subtype is most frequently observed in males during their third and fourth decades of life [2]. CCCs are slow-growing tumours with low potential for metastases and are often misdiagnosed as benign lesions, which complicates their diagnosis [4]. In this report, we present a rare case of CCC in a young adult, accompanied by detailed clinical, radiological, and histopathological investigations.



[Table/Fig-1]: a) Plain X-ray of the right hip showed a well-defined osteolytic lesion over the medial aspect of the right femoral head; b) Magnetic Resonance Imaging (MRI) of the right hip revealed an expansile, well-circumscribed T2 hyperintense lesion measuring 2×1.8×1.5 cm over the medial aspect of the right proximal femur with surrounding mild oedema.



[Table/Fig-2]: a) Gross of entire proximal femoral head showing well delineated greyish white lesion with no adjacent tissue invasion; b) (HPE, 40x) Tumour cells arranged in diffuse sheets and lobular pattern; c) (HPE, 400x) Individual tumour cells having abundant, clear cytoplasm with centrally placed nucleus, vesicular chromatin and prominent nucleoli; d) (HPE, 100x) Reactive woven bone formation; e) (HPE, 400x) Numerous multinucleated giant cells seen.



[Table/Fig-3]: Postoperative radiograph shows proximal femoral head reconstructive plates.

Chondrosarcomas are the most common primary malignant tumours of cartilaginous origin seen in the elderly, with a male predominance [5]. They are the third most common primary malignancies amongst all the bone tumours after osteosarcoma and Ewing's sarcoma [5,6]. They tend to involve most commonly the epiphyses, followed by the epiphyseo-metaphyseal region and rarely may extend up to the diaphysis [5]. It constitutes only 2% of all variants of chondrosarcoma since the majority are conventional subtypes (90%) [2,4]. CCCs are most commonly seen in the third to fourth decades of life, with three times as likely seen in males than females [4,5]. CCC was first described by Unni KK et al., in 1976 as a distinct subtype of chondrosarcoma characterised by lobules of clear, polygonal cells. They commonly arise from the proximal end of the femur (60%), followed by the humerus and tibia and other small bones like talus, calcaneum [4] and others like skull, ribs, phalanges and spine. Localised bone pain is the initial symptom, which is gradually progressive and not relieved despite analgesics, as seen in this case. Radiologically, they are characterised by well-defined osteolytic lesions involving the proximal parts of the long bones in the early stages, with few stippling calcifications seen as radiodense lesions in the later stages. However, periosteal reaction is absent, negating the diagnosis of osteosarcoma [7]. MRI forms the investigation of choice, which shows a well-circumscribed, sharply defined hyperintense lesion of T2 weighted imaging [2,7], with variable surrounding oedema. These non-specific features point towards benign lesions such as chondroblastoma and giant cell tumour [7]. Most of the time, CCC behaves as low-grade with rare de-differentiation into malignancies [8]. Gross show firm, sometimes cystic lesion with gritty sensation on cutting the tissue structures. Histopathology is the gold standard for diagnosis, which shows tumour cells arranged in sheets and lobules. Individual tumour cells are medium to large, round to polygonal in shape, having abundant amounts of clear, vacuolated to slightly eosinophilic cytoplasm, a centrally placed nucleus and prominent nucleoli. The clear cytoplasm in CCC is attributable to abundant intracytoplasmic glycogen, which can be confirmed using PAS staining (both with and without diastase digestion). The cells typically express S100, D2-40, vimentin, and osteonectin [5,7,8]. There may also be the presence of a few osteoclastic giant cells, hence ruling out GCT and short bony trabeculae, indicating heavily calcified areas surrounding the cartilaginous matrix, with rare instances of tumour osteoid. Additionally, focal areas of cystic formation and hemorrhage may be observed. Due to their low-grade nature, CCCs typically exhibit no mitotic activity, necrosis, and low cellularity. Chondroblastoma is ruled out due to absence of chondroid matrix and chondroblastic cells exhibiting small grooved nuclei. In the elderly, metastatic clear cell Renal Cell Carcinoma (RCC) show rich capillarisation in addition to the clear cell morphology with PAX8 positivity [8]. Cytogenetics shows loss or structural aberrations of chromosome 9 and gain of chromosome 20 with diploid or near-diploid complements predominantly seen in CCCs [9]. Behjati S et al., study has shown the H3F3B pLys36Met mutation in CCC in one case, which is a specific driver mutation of chondroblastoma, hence suggesting a pathogenetic relation between the two tumours [10]. Wide surgical excision with reconstruction forms the mainstay of management, as conservative treatment shows higher recurrence rates [7,8]. Serum Alkaline Phosphatase (ALP) and other biochemical markers are non-specific for diagnosis; however, ALP can be used during follow-up postoperatively for early recognition of recurrence and metastasis [3]. In cases of inoperable lesions and the elderly, high-dose ionising radiotherapy may be done for local control [8].

CONCLUSION(S)

The epiphyseal tumours of long bones in young adults radiologically mimic numerous benign lesions like chondroblastoma or GCT; hence, the possibility of CCC in a young adult should be kept in mind despite its rarity. Clinical details, site of involvement, radiological investigations and histopathological correlation are always necessary to arrive at a final diagnosis for adequate management.

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