E-ISSN: 2707-8353
P-ISSN: 2707-8345
IJCRO 2023; 5(1): 86-89
www.orthocasereports.com
Received: 08-01-2023
Accepted: 17-02-2023

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Oncological and functional outcome in a rare case of clear cell chondrosarcoma of the proximal femur

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DOI: https://doi.org/10.22271/27078345.2023.v5.i1b.149

Abstract
Chondrosarcomas are the second most common bone sarcomas, accounting for less than 0.5% of all cancers. The treatment plan can be improved by analyzing clinical characteristics and treatment patterns. The rarity and slow-growing nature of this tumour often lead to prolonged symptoms and also initial misdiagnosis with avascular necrosis of the femur, benign bone lesions like chondroblastoma. The initial reports of this patient suggested avascular necrosis of the femur head. His pain became severe and further investigations were done, MRI confirmed the diagnosis of Chondrosarcoma. An image-guided core needle biopsy suggested Clear Cell Chondrosarcoma (CCC). Wide local resection with negative margins forms the mainstay of treatment since intralesional procedures predispose to a high local recurrence rate. A prolonged follow-up is required since late local recurrences of tumour and distant metastases are common.

Keywords: Oncological and functional outcome, cell chondrosarcoma, proximal femur

1. Introduction
Chondrosarcomas accounts for less than 0.5% of all cancers [2]. Primary or conventional chondrosarcoma arises in preexisting normal bone and is distinguished from the rarer secondary tumours, which occur in a preexisting enchondroma or osteochondroma [3] and are common in the metaphysis. Unlike conventional chondrosarcoma, CCC have a predilection for the epiphysis of long bones and can be mistaken for benign entities. Tumour may extend into the metaphysis and the diaphysis, commonly occurring around the third to fourth decade of life and more frequently in males [3]. It was first described by Unni *et al*. 1976 [4], The tumour usually involved the proximal femur or humerus. Roentgenographically, the lesion was usually well defined and indistinguishable from chondroblastoma. Serial roentgenograms occasionally showed malignant progression of the lesion. Histologically, benign giant cells and cells with clear cytoplasm were always present, and areas of conventional chondrosarcoma were seen in about half their cases. Excision usually resulted in recurrence; resection seemed to be curative in their case series. We report a case of CCC of the proximal femur which was misdiagnosed as avascular necrosis of femur head elsewhere and treated. Patient-reported to us later and diagnosed as Clear cell chondrosarcoma and treated with wide excision of left proximal femur and reconstruction with Bipolar tumour mega prosthesis of left hip (uncemented), we report the Oncological and Functional Outcome of this case.

2. Aims and objective
This is a report of a case of Clear cell chondrosarcoma proximal femur in a middle-aged man who was initially misdiagnosed to have avascular necrosis of the femur head and was eventually treated with Limb salvage surgery and Tumour Megaprosthesis.

3. Case presentation
A 50-year-old gentleman, presented with a history of pain in the left hip and difficulty in walking for more than 2 years duration. His initial reports suggested avascular necrosis of the femur head. His pain became severe and further investigations were done. MRI indicated a T1 hypointense and T2, STIR Hyperintense lesion involving the left femur head and neck with the cortical breach. An image-guided core needle biopsy suggested Clear cell chondrosarcoma. Whole-body FDG PET Ct scan showed no evidence of metastases.

Tumour dimension was 7.5 x 6.8 x 4 cm. The family was counselled about the feasibility of limb salvage surgery and he subsequently underwent wide excision of left proximal femur and reconstruction with Bipolar tumour mega prosthesis of the left hip (uncemented).

Post excision histopathology evaluation confirmed clear cell variant and negative margins. Started physiotherapy from postoperative day 1, non-weight bearing mobilization on the left side (for 6 weeks) with walker support.

At 5 years of follow up, he is disease-free and mobilizing without support, doing all daily activities including riding a two-wheeler. His left hip range is full and able to climb up and down stairs and walk for more than 3 to 4 km.

Fig 1: X-ray of the pelvis shows an osteolytic, expansile lesion in proximal femoral epiphysis with no calcification, the sharp interface between tumour and surrounding bone with no sclerotic rim. The overlying cortex is intact, with no periosteal reaction and no soft tissue extension

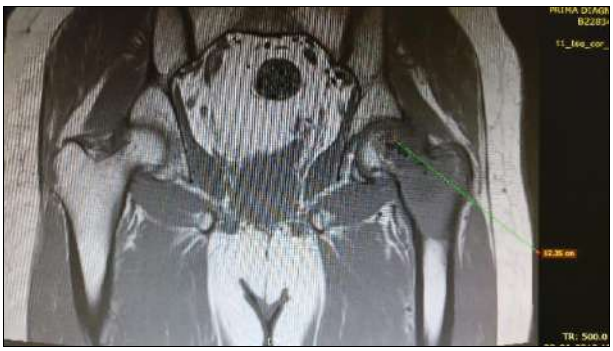

Fig 2: MRI of the pelvis shows a Large expansile lytic lesion (7.5 x 6.8 x 4 cm) in the left proximal femur demonstrating chondroid matrix and endosteal scalloping with a possible cortical breach - is suggestive of a primary cartilaginous tumour

- Alkaline phosphatase & Calcium levels were within the normal range.
- As none of the radiological investigations were confirmatory, the patient was subjected to CT guided biopsy.

Histopathology

1. Gross morphology

Specimen of 2 grey white tissue bits each measuring 0.5 cm

2. Microscopic description

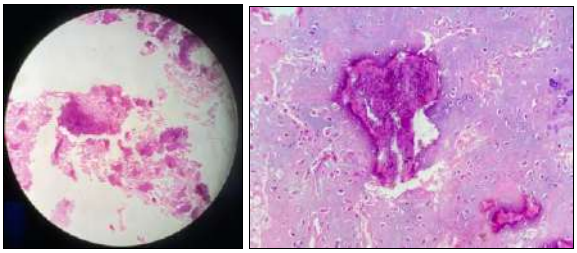

Fig 3: Microscopic description

- Lobules of cartilaginous tissue were noted.
- Multiple brownish tissues

The section shows blood admixed tumour fragments composed of round to oval and atypical spindle cells lying in a chondroid matrix with some areas showing maintained lobulated appearance. Also seen admixed larger polygonal cells with an atypical pleomorphic hyperchromatic nucleus and clear cytoplasm with intervening thin wall blood vessels.

- CT thorax didn't show any metastasis to the lungs.
- X-pert MTB/RIF assay (GeneXpert) was negative.
- The lesion being juxta-articular with no cortical breach or soft tissue involvement patient was planned for Wide excision + Bipolar uncemented mega prosthesis left hip.

4. Procedure

Procedure performed under spinal anaesthesia. Postero-lateral approach to hip was done. Intraoperatively there was no evidence of pathological fracture or soft tissue extension. The femur is reamed up to 14 & 14 sized uncemented implant is used, bipolar with proximal femur construct including 65mm trochanter, 20mm coupling, 15mm neck and 10mm collar.

Construct assembled with 27 COCR head & 47 bipolar cup. Anterior cortex longitudinal split was noted during implantation of the uncemented stem which was stable and encirclage wiring done.

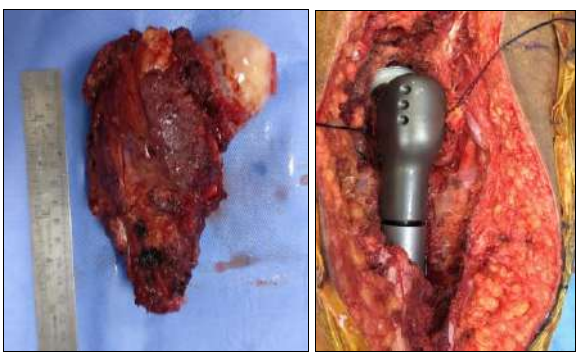

Fig 4: The excised specimen was sent for histopathological examination. The postoperative period was uneventful.

Fig 5: Three months follow-up x-ray images

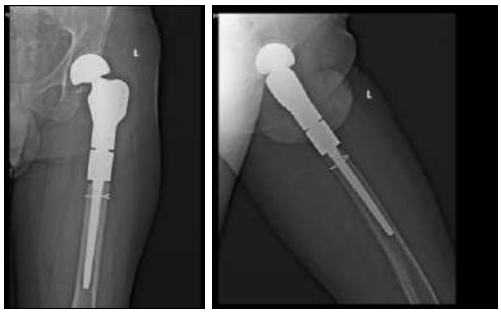

Fig 6: Five year follow-up x-ray images

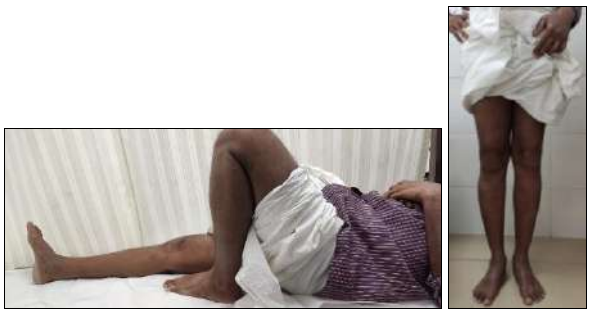

Fig 7: At 5 years of follow up, he was disease-free and mobilizing without support, doing all daily activities including riding a two-wheeler. His hip range is full and able to climb up and downstairs, squat

5. Discussion

Males are affected twice as common as females. Radiologically these tumours appear as expansile, radiolucent lesions in the epiphyseo-metaphyseal region with intact cortices. The proximal femur (60%) and proximal humerus (15%) are the most common locations for the tumour followed by the distal femur and proximal tibia (15%), and rarely involves other bones like the skull, spine, pelvis, phalanges and foot (10%) [5]. Though the age of occurrence ranges from 13 to 85 years, 95% of the tumour occurs in the age group of 5–25 years [6].

No periosteal new bone formation or matrix calcification is seen. Cartilage tumours generally exhibit matrix calcification in the form of rings or popcorn shape, clear cell chondrosarcoma is an exception [3].

Since the tumour is believed to arise from the chondrocytes in the secondary ossification centre, its predilection to the epiphysis or apophysis of long bones is the general rule. However, an extension of the tumour into the metaphysis and even diaphysis is not uncommon [7].

The diaphysis is rarely involved. Even though clear cell chondrosarcoma resembles chondroblastoma microscopical presence of clear cells makes them unique.

Most of the clear cells on electron microscopy show irregular-shaped nuclei with indentations [8]. Large dilated endoplasmic reticulum cisternae, bundles of actin-like filaments & a few glycogen particles. The presence of clear cells is the primary differentiating feature between giant cell tumour/aneurysmal bone cyst and clear cell chondrosarcoma.

Alkaline phosphatase (AP) has been described as a marker for primary and recurrent CCC. Ogose *et al*. [9] concluded that CCC produce AP and levels decline or normalize after excision. But alkaline phosphatase was normal in our case.

CCC is considered to be resistant to both radiotherapy and chemotherapy [10]. Despite the presence of a low malignant potential these tumours are not to be treated by curettage and grafting as it has a high recurrence rate. Wide excision and reconstruction is the treatment of choice.

6. Conclusions

Clear cell chondrosarcoma is a rare subtype of chondrosarcoma with a relatively low-grade malignant potential. A combination of clinical, radiological, and histopathological approach has to be used for appropriate diagnosis considering the wide spectrum of possible differential diagnoses as it is difficult to differentiate from other entities such as carcinoma metastases, chondroblastoma, giant cell tumour, and an aneurysmal bone cyst based on imaging alone [3].

Wide local resection with negative margins forms the mainstay of treatment since intralesional procedures predispose to a high local recurrence rate. A prolonged follow-up is recommended since late local recurrences and metastases are common.

7. Conflict of Interest

Not available

8. Financial Support

Not available

9. References

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How to Cite This Article
Porammanavar ST, Kulkarni A, Gopalan S, Srimath BS. Oncological and functional outcome in a rare case of clear cell chondrosarcoma of the proximal femur. International Journal of Case Reports in Orthopaedics. 2023;5(1):86-89.

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