Abstract:
Chondrosarcoma is the second common primary bone malignancy. The clear cell variant was recognized as a distinct entity only in 1976. It is a rare variant with a reported incidence of less than 2 percent among all variants of chondrosarcoma. It is a low grade malignant tumor with characteristic clinical, imaging and pathological features that are unique and different from other chondrosarcoma variants. It can often be erroneously confused with a benign bone tumor and managed accordingly with high rates of recurrence. Here we report a case of clear cell chondrosarcoma and discuss its unique and characteristic features.

Keyword: CLEAR CELL CHONDROSARCOMA

INTRODUCTION:
The clear cell variant of chondrosarcoma was first reported in 1976 by Unni et al [1] and accounts for around two percent of chondrosarcoma variants. It is a low grade malignant tumor with unique clinical, histopathological and imaging qualities that contrasts from other chondrosarcoma variants. The clear cell chondrosarcoma is often misdiagnosed as a benign bone tumor and managed accordingly with high rates of recurrence. It is common in young adults in 3rd to 5th decade. Clear cell chondrosarcoma involves the proximal end of the long bone in 75% of the cases (60% femur, 15% humerus), 15% around the knee (distal femur and proximal tibia). Rare sites include skull, spine, ribs, ulna, phalanges, etc. [4] Here we report a case of clear cell chondrosarcoma around the knee joint.

CASE REPORT
A 42 year old female presented in 2010 to our centre with pain and swelling of left distal femur and limitation of left knee movements with a previous history of curettage and bone cementing done for chondroblastoma of the distal femur 10 years back. A plain x-ray revealed a solitary, large expansile, predominantly eccentric lytic lesion with cortical thinning and no periosteal reaction of the left distal femur epiphyseal end with bone...
cement artifacts from the previous procedure. An image guided biopsy was done and was reported as Giant cell tumor. A diagnosis of benign bone tumor was made based on triple assessment. The patient defaulted treatment on recommendation of surgery. She again presented in June 2013 with worsening pain, limitation of movement and progressive swelling around the left knee. Clinical examination revealed a 12 X 8 cm circumferential bony hard swelling of left distal femur merging imperceptibly with bone. The swelling was warm with prominent dilated overlying veins. Active and passive left knee joint movements were restricted and painful. There was no evidence of pathological fracture. The skin over the swelling was normal. The patient had no distal neurovascular deficit or lymphadenopathy. X-ray and CT scan of the left knee confirmed an expansile intra osseous lesion of homogenous intermediate signal density with no calcification or periosteal reaction without soft tissue extension (figure 1). Radiological evaluation confirmed an involute joint space and the neurovascular structures to be uninvolved. A diagnosis of Giant cell tumour was made based on the clinical features, radiology and previous biopsy. She was treated with distal femur resection with Custom Mega Prosthesis reconstruction (figure 2). The resection was done through a medial utilitarian incision from mid thigh to mid leg which included previous scars. The tumour was found to be confined to distal femur. Knee joint, popliteal vessels and tibial nerves were free of tumour. The wound was closed in layers with suction drains and an above knee plaster of Paris slab given for immobilization. The immediate post operative period was uneventful. The patient was mobilized first with non weight bearing crutches and then with incremental weight bearing with knee braces for support. The patient was ambulatory at discharge. Gross histological examination revealed a yellowish grey tumour with distinct border areas of cartilage and necrosis. There was no new bone formation or cortical breach. Microscopy showed tumour composed of lobules of abundant clear cell chondrocytes with a vacuolated cytoplasm, mononuclear cells and giant cells with areas of cartilage formation and necrosis confirming a diagnosis of clear cell chondrosarcoma (figure 3). Circumferential and proximal margins were free of tumour and adequate. A post operative metastatic evaluation was done and was normal. She had one episode of prosthesis related wound infection in January 2014 which subsided with parenteral antibiotics. The patient is on regular follow up and physical examination with surveillance imaging confirm no recurrences. She has good functional recovery and is fully ambulant unaided.

**DISCUSSION:**
Chondrosarcoma is the second common primary bone malignancy after osteosarcoma. They account for 20 to 27 percent of primary malignant osseous neoplasms. Based on histology of these tumors several types have been described each with unique biological behavior, clinical presentation, radiological appearance and prognosis. Ninety percent are conventional chondrosarcomas which are low to intermediate grade tumors. These tumors are slow growing with a low metastatic potential. They are considered relatively refractory to chemotherapy and radiation therapy. It usually involves flat bones but can also occur in extra osseous tissues. High-grade chondrosarcomas, which include 5 to 10 percent of conventional
chondrosarcomas as well as some rare variants, have a high metastatic potential and a poor prognosis following resection alone. Some of the rare subtypes are more responsive to chemotherapy and radiation.\[6\] Chondrosarcoma exhibits considerable radiotherapy and chemotherapy resistance hence comprehensive oncological surgery is the only effective treatment modality. They are often large tumors at presentation and by virtue of involving the load bearing skeleton, resection mandates reconstructive procedures. Clear cell variant occurs in younger age group than conventional chondrosarcoma types (3rd to 5th decade), prefers proximal (epiphysis) ends of humerus and femur rather than flat bones. The most common presentation is long standing pain which is progressive. Other presentations include swelling, disabilities, pathological fractures, etc.\[3,4\] Radio graphically the lesion is almost always located in the epiphyseal area of long bones with or without extension to the metaphysis. Diaphyseal involvement is rare. They are usually radiolucent, expansile with calcification with or without a sclerotic rim and thinning of cortex. Soft tissue involvement is rare (less than 10%). CT scan is more sensitive in detecting cortical involvement and calcification. MRI shows a well demarcated geographic lesion with intermediate signal intensity on T1-weighted images, and higher signal intensity on T2-weighted images. Differential diagnoses include chondroblastoma, giant cell tumour, chondroma, chondromyxoid fibroma and cysts, etc. It lacks the characteristic ring and arc type of matrix calcification of conventional types, and presents as rather purely lytic lesion. \[3\] Serum alkaline phosphatase levels are often elevated at diagnosis and may provide a useful tumor marker.\[8\] Genetic mutations associated with clear cell variant are Chromosome 20 gain, 9 loss, 18q loss, mutations in PTHLH, PDGFIHH, Runt-related transcription factor2, matrix metalloproteinase 2 (MMP2), p53 over expression, etc. Immunohistochemistry for Collagen II, S 100 protein and presence of glycogen are useful in the diagnosis.\[5\] They are often misinterpreted as chondroblastoma or giant cell tumor based on the slow growth and benign appearance radiologically and histologically. The delay in diagnosis at an early stage also hampers treatment. Surgery is the treatment of choice. Intra-lesional excision (e.g. curettage, incomplete excision) yields an unacceptably high local recurrence (83-86%) and death rate (29 to 50%). Complete surgical removal of the tumour with a wide margin (e.g. en-bloc resection, primary amputation) has a lower local recurrence rate (less than 15%) lower death rates.\[1,3,8,9\] Radiotherapy and Chemotherapy are not useful as these tumours are not radio or chemo sensitive. Amputation is rarely required and most patients can be treated with en bloc resections with wide margins. It is a low grade tumor with minimal metastatic potential. As mentioned, tumours with incomplete resections and/or margin positive tumours tend to have a poor prognosis when compared with patients who are adequately treated. Long term follow up is mandatory as these tumours have shown to recur many years later. \[8,9\] Distant metastases are rare.

**CONCLUSION:**
Clear Cell Chondrosarcoma is a rare variant with benign looking clinical, radiological and histological features. It is often misdiagnosed as Chondroblastoma or Giant Cell tumour. It is a low grade tumor with minimal metastatic potential. En Bloc Resection with wide margins is the treatment
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of choice. Prognosis is relatively good but long term follow up is necessary.

Figure 1: X Ray and CT Scan

Figure 2: Distal Femur Resection and Custom Mega Prosthesis Reconstruction
Figure 3: HPE: Clear Cell Chondrosarcoma - H & E Staining

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