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A case report of resection arthroplasty for Giant cell tumor of distal femur with megaprosthesis

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Abstract
Giant cell tumors (GCT) are neoplasms of mesenchymal stromal cells with varied manifestations. Giant cell tumor is considered to be of benign nature but 3% of giant cell tumors are primarily malignant or will undergo malignant transformation and metastasize. The World Health Organisation has classified GCT as "an aggressive, potentially malignant lesion", which means that its evolution based on its histological features is unpredictable. Although there are studies available mentioning various treatment modalities, their advantages and disadvantages, definitive criteria were not laid down regarding treatment. We consider worthwhile reporting a case of distal femoral aggressive GCT treated by enbloc excision and reconstruction with custom mega prosthesis with good functional outcome.

Keywords: GCT, Distal femur, Megaprosthesis

Introduction
Giant cell tumor of bone was first described by Cooper and Travers in 1818 [1]. It is one of the commonest tumor that is presented to Orthopaedic surgeon. Giant-cell tumor of the bone accounts for 4-5% of primary bone tumors and about 20% of benign bone tumors [2] with unusually high prevalence in southern India and China, where GCT represents 20% of all primary bone tumors [3].
There is slight female predominance with a female-to-male ratio of 1.3-1.5:1 [6]. GCT occur most commonly in the third decade of life; less than 5% of GCT occur in patients who are skeletally immature [4, 5]. The natural history of GCT varies widely and can range from local bony destruction to local metastasis, metastasis to the lung, metastasis to lymph nodes (rare), or malignant transformation (rare) [4, 6, 7, 8, 9].
Most lesions develop in epiphysis of long bones (75%–90%), with the majority of cases (50%–65%) occurring about the knee, but tumor can extend to metaphysis as well as diaphysis [14]. The three most common locations are the distal femur, proximal tibia, and distal radius, respectively [10, 11]. GCT may occur in association with Paget disease, most commonly in the skull, facial bones, pelvis, and spine [12].
Pain and swelling are the most common presenting features with increased risk for pathological fractures. Plain radiograph usually shows eccentric radiolucent lytic lesion of epiphysis with thinned out cortex. MRI required to show extent of soft tissue involvement with well circumscribed homogenous signal intensity [13]. Diagnosis is confirmed by histopathogical examination. Grossly, these lesions are characteristically chocolate brown, soft, spongy, and friable with blood filled cavities. Microscopically, multinucleated giant cell is the characteristic cell type with background network of stromal mononuclear cells [15].

Case Report
A case of 31 year old male presented with gradual onset of pain and swelling of right knee since 2 years. Pain was dull aching, nonradiating associated with weight bearing on right lower limb without any constitutional symptoms. There was no recent history of trauma or similar complaints in other joints. On examination, patient was conscious, oriented to time, place and person and vitally stable. There was 15*12cm solid mass with hard consistency over right distal femur without local rise of temperature or tenderness. Overlying skin was freely mobile withour any scar, sinus or dilated veins.

Knee flexion was restricted to 0-30' and there was no distal neurovascular deficiet.

Plain radiograph shows expansile, lytic lesion involving distal femoral epiphyseal and metaphyseal area without any periosteal reaction. Patient had his previous xrays of knee which shows initial small, eccentric, radiolucent lytic lesion on medial aspect of distal femur without any periosteal reaction which progressed gradually to involve whole distal femur (predominantly on medial aspect) with thinned out cortex and surrounding soft tissue involvement. MRI of right knee suggests large extensively lobulated expansile heterogenous enhancing lesion involving metadiaphyseal region and extending to epiphysis of lower end of right femur upto the articular surface with breech in the articular cortex of distal femur and extension of lesion into the joint cavity. There is associated thinning and break at medial and inferior aspect of right medial femoral condyle. Lesion measures approximately 15*14*11cm with large central necrotic area within lesion and extends medially and anteriorly and displaces quadriceps tendon anteriorly and crossing the knee joint. FNAC was carried out to confirm the diagnosis. The patient was screened for metastasis with computed tomography of the brain and chest, ultrasonography of the abdomen and pelvis and there was no obvious evidence of any secondaries.

Patient was planned for resection of tumour and insertion of custom made megaprosthesis. Extended medial parapatellar approach which aids in vascular dissection and hence that the popliteal vessels can be separated and tumor dissection carried out. We used the technique of sleeve resection of quadriceps musculature. The main objective of this technique is to excise a sleeve of quadriceps musculature all around the tumor but retain the functioning rectus femoris tendon. The excision removes a portion of the vastus lateralis, medialis and intermedius, but preserves enough musculature to provide soft-tissue coverage for the prosthesis and retains adequate extension power. The custom mega prosthesis contains a femoral condylar component, a pivot pin, a thrust-bearing pad made of high molecular weight polyethylene and tibial component. Proximally, the prosthesis is angulated laterally by 6° to resemble the valgus angle of the lower limb. The function of the thrust-bearing pad is to impart a flexion of 150° between the femoral and tibial components. The ultra-high-molecular-weight polyethylene-bearing pad serves to relocate the load transmitted during weight-bearing. The rotating axis mechanism provides 3° of rotation between the femoral and tibial component.

Quadriceps strengthening exercises were started from the 2nd post-operative day. Patient was allowed to walk with the help of walker on the 3rd post-operative day. On the 15th post-operative day sutures were removed and the patient was discharged. Knee bending was started after 3 weeks. On follow-up after 2 months, patient was walking with a good range of flexion, without any support. There was no evidence of flap necrosis, prosthetic failure or peri prosthetic fractures.

Discussion

The management of juxta-articular giant cell tumors around the knee occurring in young patients continues to be one of the most challenging areas in orthopedic oncology [4, 16]. Enneking's and Campanacci's radiographic classifications and surgical staging are helpful in planning the initial surgical treatment, as a number of the active (Stage 2) lesions and most of the aggressive (Stage 3) lesions have a higher incidence of local recurrence (20-50%) when treated by curettage with or without bone grafting [4, 17, 18]. The use of methylmethacrylate cement has equivalent recurrence rates [19, 20]. The safety and advantages of additional adjuvant treatment of the bone bed with phenol or liquid nitrogen after tumor resection is questionable [21, 22]. Since the local behavior of giant cell tumors can be aggressive and they have a greater risk of local recurrence, some authors advocate en bloc resection and reconstruction for these Grade III lesions from the point of view of preventing local recurrence rate and preserving joint [23, 24]. Although it is the treatment of choice for these tumors, wide resection creates a problem for the reconstruction of large bone gaps. The reconstructive procedure has to be based on several considerations, such as durability of the surgical procedure, the oncological prognosis, restoration of the anatomy and function, and the needs of the patient [25]. Rotationplasty gives excellent functional results but the cosmetic outcome is a serious disadvantage of this procedure [26]. Resection arthrodesis achieves excellent stability but has the major drawback of lack of knee motion [27].

The use of mega prosthesis has become the method of choice after bone tumor resection at the knee [28]. It is the primary modality in the management of malignant bone tumors of lower limb [29]. Custom mega prosthesis has proved to be a simple, technically superior method of replacing the lost segment of the bone in benign aggressive lesions with pathological fractures and where disease progression has resulted in a clinical situation that prevents skeletal reconstruction after intralesional curettage [29, 30]. The advantages of custom mega prosthetic arthroplasty are cost-effectiveness, early resumption of knee function with unassisted ambulation and least rates of recurrence. The possible complications include flap necrosis, secondary infection, aseptic loosening fracture and breakage [31, 32].

Bone resection is not usually recommended because of its significant morbidity. It is only indicated in proximal radius and fibula and distal ulna, tubular bones of hand and foot, coccyx, sacrum and pelvic bones, also in situations in which their reconstruction is not possible as in some patterns of pathological fractures and massive involvement with an incomplete shell of cortex that is insufficient to contain cement [33, 34].

Radiation therapy as adjuvant treatment is not routinely used because of concerns regarding efficacy of therapy as well as reports that mentioned sarcomatous change after radiotherapy [35]. Radiotherapy can be used as an alternative to surgery in cases that cannot be treated with surgery or left with severe disfigurement after surgery [36]. In our case we did not use radiotherapy in the treatment of our patient.

Conclusion

Distal femur endoprosthetic reconstruction with custom made megaprosthesis was shown to be a safe and reliable technique of reconstructing a large bony defect, created after resection of locally aggressive juxtaarticular tumor, providing good functional and oncologic outcomes.

There is no conflict of interest

Figure legends

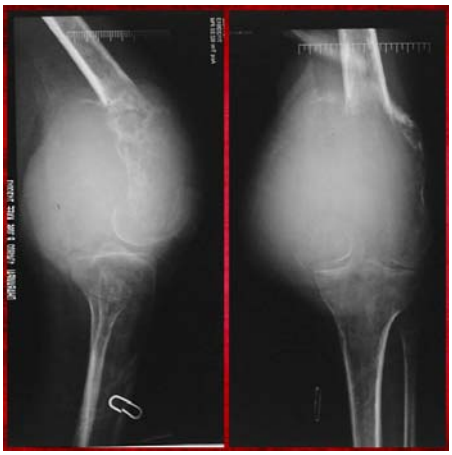

Fig 1: Anteroposterior and lateral view of right distal femur showing expansile, lytic Giant cell tumor

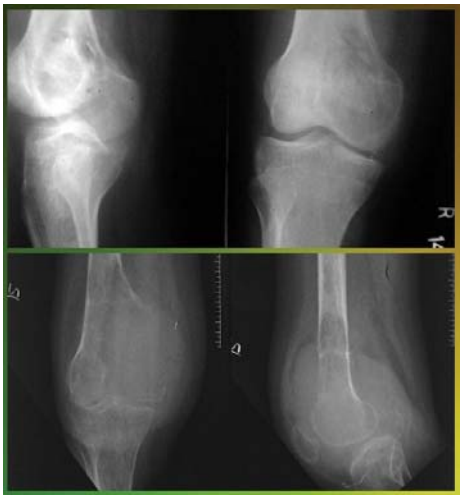

Fig 2: Previous Anteroposterior and lateral view of right distal femur showing small radiolucent, eccentric lytic lesion in medial femoral condyle which is progressing.

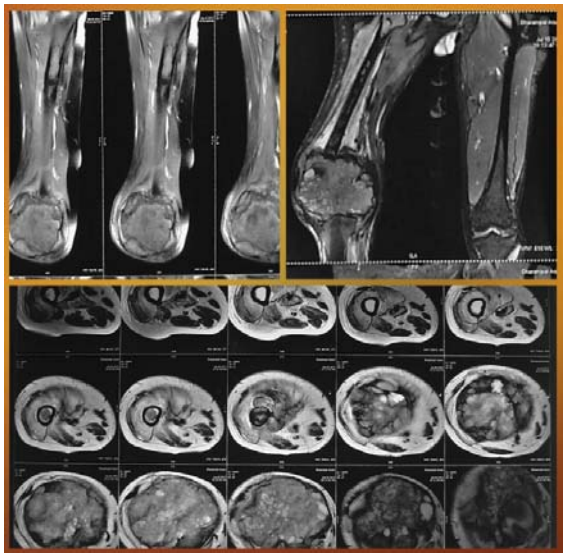

Fig 3: MRI images of distal femur GCT showing large tumor mass with central necrotic area

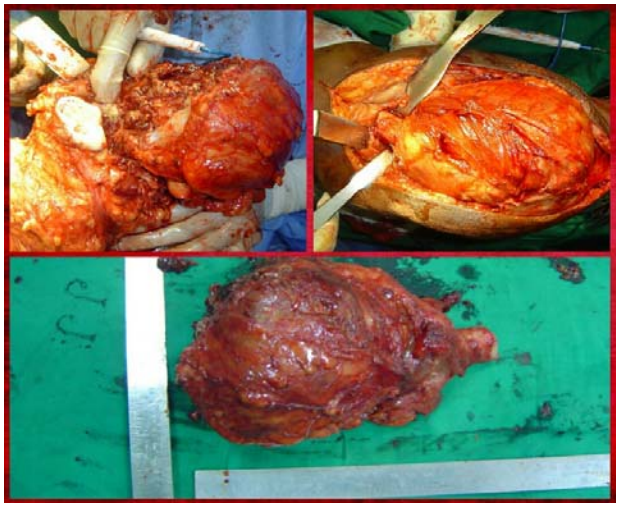

Fig 4: Medial parapatellar approach to right knee dissecting tumor area and excised specimen

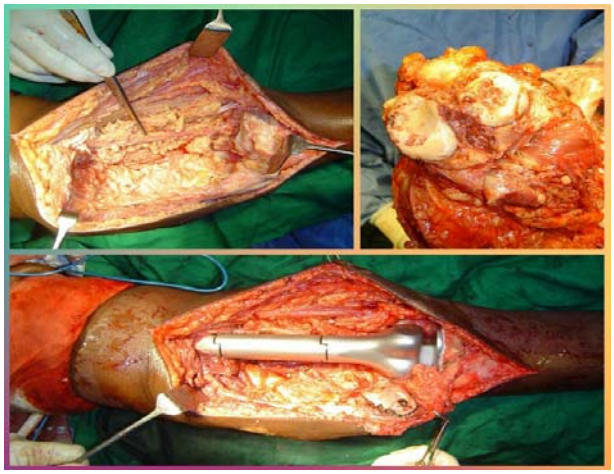

Fig 5: Intraoperative images of megaprosthesis insertion

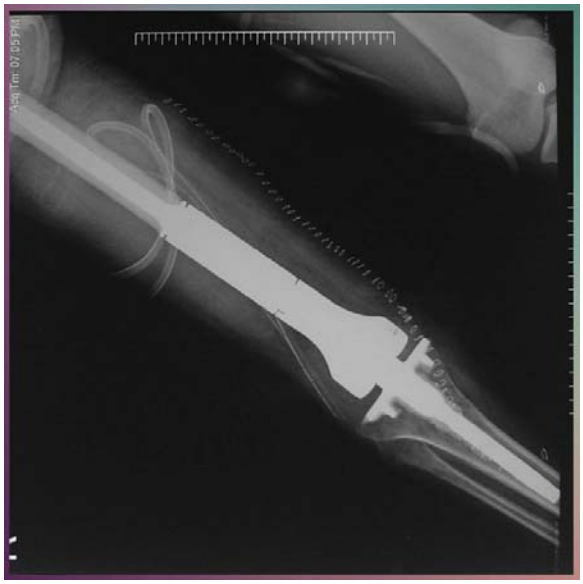

Fig 6: Post-operative xray showing megaprosthesis.

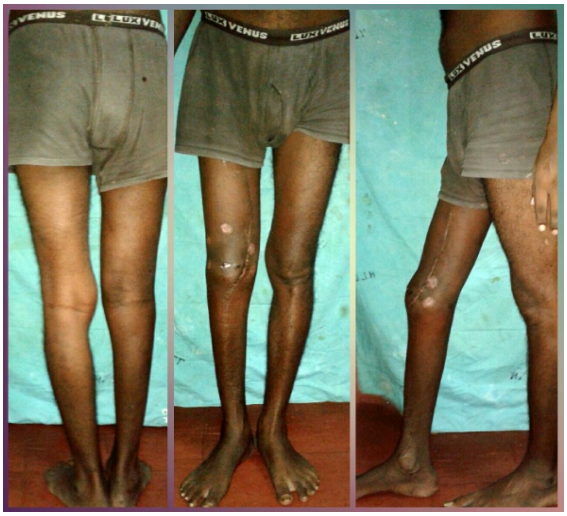

Fig 7: Post-operative clinical picture of patient.

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