GIANT CELL TUMOR OF DISTAL END FEMUR: A CHALLENGE IN TREATMENT

SURAJ BAJRACHARYA, GURU PRASAD KHANAL, PRAVIN NEPAL, BIKRAM PRASAD SHRESTHA, MAHIPAL SINGH

ABSTRACT

We present a case of malignant giant cell tumor of distal end of right femur treated with resection of the tumor mass en block with acute docking of proximal and distal end and fixed with long K-nail across knee from femur to tibia. After complete consolidation/ union of the ends, removal of K nail was done followed by corticotomy along with distraction osteogenesis with the help of Ilizarov ring fixator. The length was achieved with this process. The end result was very good in this case. We reviewed the treatment options for malignant giant cell tumor of femoral distal end and the challenges in its treatment.

Keywords: *Giant cell. Ilizarov's technique. Distraction osteogenesis.*

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INTRODUCTION

Giant cell tumor of the bone is a relatively uncommon tumor. It is characterized by the presence of multinucleated giant cells. The tumor is usually regarded as benign. In most patients, giant cell tumors have an indolent course, but tumors recur locally in as many as 50% of cases. Metastasis to the lungs may occur.

Cooper first reported giant cell tumors in the 18th century. In 1940, Jaffe and Lichtenstein defined giant cell tumor more strictly to distinguish it from other tumors. Giant cell tumor usually occurs de novo but also may occur as a rare complication of Paget disease of the bone. Giant cell tumor of the bone accounts for 4-5% of primary bone tumors and 18.2% of benign bone tumors. The incidence is increased in patients with Paget disease of the bone, in which giant cell tumor is a rare neoplastic complication. Giant cell tumor is a rare complication compared with Paget sarcoma, which has an incidence of sarcomatous change of <5%.

A slight female predominance is noted; approximately 50-57% of cases involve female patients. Typically, giant cell tumors occur in skeletally mature patients aged 20-40 years. The incidence peaks in those aged 20-30 years. Giant cell tumors are much less common in children; the rate is 5.7% in skeletally immature patients. Vertebral tumors tend to occur in younger patients; 29% of these tumors occur in those aged 0-20 years. Multicentric giant cell tumors also occur in a younger group, with a peak incidence in those aged 10-20 years. Multicentric tumors involve less than 1% of patients.

CASE PRESENTATION

A 25 years old young man attended at Out Patient Department of Orthopaedics with the chief complains of progressive swelling of the right knee joint for last eight months without preceding history of trauma to the knee joint. He did not give history of fever, chest pain, other joint swelling. He had not taken any kind of treatment prior to presenting at our OPD. On examination there was diffuse swelling of the knee joint more on the femoral condyle region circumferentially. The popliteal fossa was obliterated. There was jog of motion of the right knee joint. The distal neurovascular status was intact.

On plain radiographs (Figure 1), there was diffuse expansible radiolucent lytic lesion with cortical breach on medial lateral as well as posterior aspect of the lateral and medial condyles of femur Histopathological examination confirmed the diagnosis of giant tumor. Through midline approach to knee joint, resection of the distal of femur about 5 cm from the margin of the tumor and resection of upper end of tibia was done. Acute docking was done and fixed with Long K nail across the femur and tibia with cancellous bone graft (Figure 2a). After 9 months of follow up, there was sound union, therefore Long K nail was removed. Corticotomy was done on the upper end of tibia and fixed with ring Ilizarov across the corticotomy site (Figure 2b). Distraction osteogenesis was done to increase the length of the right lower limb. There was increase in 7.8 cm in length of the limb by the end of fourteenth month of follow up. The fixator construct was removed on eighteenth month of its application and was kept on non weight bearing crutch walking for 3 more months. The patient was able to walk without support twenty first month of follow up (Figure 2c).

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Department of Orthopaedics B P Koirala Institute of Health Sciences
Correspondences to: Department of Orthopaedics B P Koirala Institute of Health Sciences Dharan, Nepal, E-mail: drsurajbajra@yahoo.com

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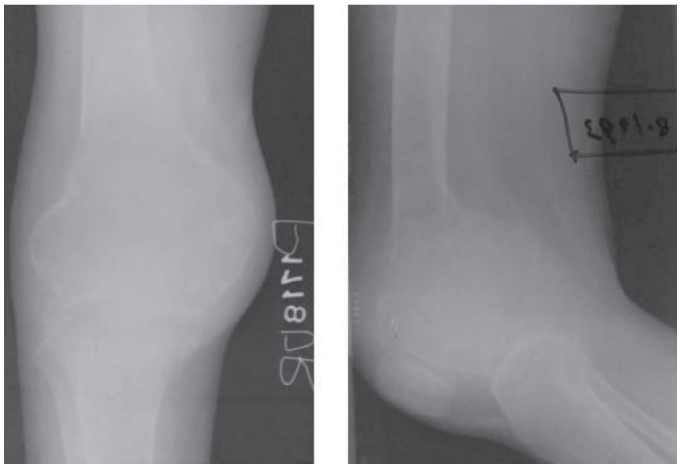

Figure 1 – *A showing malignant giant cell tumor of distal end of femur*

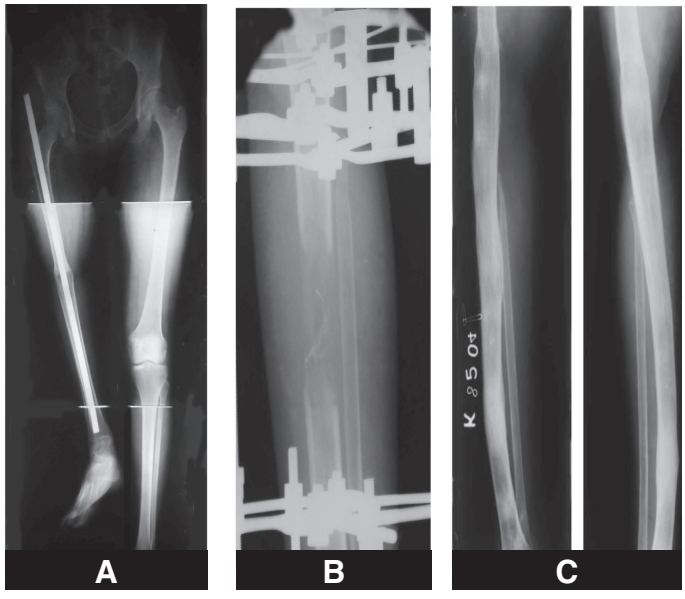

Figure 2 – *B Showing (a) Consolidated Arthodesis of Right knee joint after resection of tumor of distal end of femur(b)Lengthening of limb after removal of K nail with Distraction osteogenesis by Ilizarov technique(c)Lengthened Right lower limb- an end result*

REVIEW OF LITERATURE AND DISCUSSION

Muscolo et al.[1] reviewed the results of bone-allograft reconstruction after the resection of giant-cell tumor close to the knee in fifty-two patients (fifty-five allografts), who had been followed for a mean of seven years (range, two to twenty-four years). One giant-cell tumor was graded as stage 1; twenty, as stage 2; and thirty-one, as stage 3. Three reconstructions were repeated transplants that were done after the failure of a previous transplant. Ten allograft reconstructions were intercalary and were combined with an arthrodesis of the knee, and forty-five were osteoarticular. Major complications included infection (after three reconstructions), resorption of the graft (six), collapse of the articular surface (two), fracture (two), and recurrence (one). According to the criteria described by Mankin et al. for functional analysis, forty-two (76 per cent) of the extremities had a result that was considered to be excellent or good. Radiographic evaluation according to the system of the Musculoskeletal Tumor Society showed a mean score of 72 per cent for osteoarticular reconstructions, and of 86 per cent for intercalary reconstructions.[1]

Foukas et al.[2] reported the use of contained impacted morsellized allograft to revise an aseptically loose, massive distal femoral cemented endoprosthetic replacement in a 27-year-old Caucasian woman. The prosthesis was inserted 4 years earlier, following neoadjuvant chemotherapy and resection of a distal femoral high grade osteosarcoma. Impaction grafting was used to restore bone stock and maintain femoral length. The patient remains disease free with excellent function, at one year after revision with no evidence of loosening and maintenance of bone stock. They believe it is the first time this technique has been used in revision of a distal femoral endoprosthetic replacement.

Tumor endoprosthesis first have been used for replacement of pathologic lesions of the proximal and distal femur or humerus. Since the development of segmental defect prosthesis and modular tumor endoprosthesis it is possible to replace neoplastic lesions of long bones with diaphyseal parts. Review of Lembcke et al[3] clinic experience involving 46 cases of prosthetic replacement and only one case with a second procedure showed that this is a more viable concept than open reduction and internal fixation with methylmethacrylate supplementation.

Extensive local excisions of skeletal tumors in the knee region create reconstruction problems with several alternative solutions. Custom-made endoprostheses now compete with joint homografts and fusion with autogenous bone-grafts. Artificial fusion utilizing an extra long Kuntscher-nail and acrylic cement as a spacer is a fourth possibility with the advantages of immediate ambulation and weight-bearing, presented in two cases by Persson and Rydholm.[4] The expected disadvantages of loosening in long time survivors can be taken care of using one of the above-mentioned alternatives. This revision can be made after completion of adjunctive chemotherapy or later when called for by the occurrence of pain or instability.

Extensive osteoarticular allografts have been used for knee reconstruction, but because of their composite nature and the technical difficulty of the procedure, complication and failure rates have been high. There are few records of long-term results in the literature. In Vicas et al.[5] report, a 19-year-old man with a large aggressive giant cell tumor of the left distal femur was treated in 1976 by en bloc resection, massive femoral allografting and ligamentous reconstruction. Follow-up after 18 years showed no recurrence of the tumor, excellent incorporation of the graft and good knee function, which allowed the patient to work 9 hours a day on his feet without pain.

The current state of the art of prosthetic joint replacement permits sizeable segments of the appendicular skeleton to be resected and replaced with prosthetic components which are secured with methylmethacrylate cement. Occasionally it is necessary to resect a rather sizeable area of pathologic bone and to provide for some type of temporary fixation until a specially fabricated prosthetic component can be made available. Under such circumstances it is necessary to maintain length of the involved extremity, and provide for skeletal stability to the area of resection. Although external fixation offers a reasonable option, Volz et al.[6] advocate the employment of an internal type

of fixation in the form of titanium mesh reinforced with methylmethacrylate cement which seems more desirable. With this technique, the potential problem of pin track infection is avoided while space suitable to the dimensions of the prosthetic implant can be preserved.

Robert et al.[7] have described an operation for the conservative surgical treatment of benign giant-cell tumors in the distal end of the femur, in cases in which the articular cartilages of the knee joint are intact. The described method permits removal of the tumor, and yet retains mobility of the knee joint and the full weight-bearing function of the leg. The procedure consists of obliterating the cavity by telescoping the fragments of bone, after removal of the tumor by curettage and chemical cauterization. To date the patients (all women) have accepted the shortening incurred, and have declined their offers to equalize length by operative shortening of the normal limb.

Kapukaya et al.[8] applied callus distraction method to nine patients who were referred because of a bone tumor. All of the tumors were localized on the femur, and the histological diagnosis was two chondrosarcomas, one Ewing's sarcoma, three osteosarcomas, one giant cell bone tumor, and the remainder benign fibrous histiocytoma. The mean length of the defect after resection of the tumor was 11.5 (range 8-20) cm. Preoperative and postoperative chemotherapy were applied to patients with osteosarcoma and Ewing's sarcoma. The patients were followed up for 22 (range 15-30) months on average. The mean period of use of the external fixator was 12.5 (range 8-18) months. One patient suffered a tumor recurrence and died after 20 months. Complications included one deep infection, one skin invagination, and one premature consolidation and bone bridge in the defect area. All of the complications were successfully treated. Functional evaluation gave excellent results in four patients, good in three, and fair in two. They advocates that this method can be used without any need for massive autogenous bone graft in repairing defects of any length and diameter produced after excision of the lesion and thus can be considered as an alternative to other techniques.

Difficulties in local control of GCT of bones as well as high rate of local recurrence following initial surgery have led the investigators to use different surgical modalities for the treatment of GCT according to stage of the disease aiming at decreasing the rate of local recurrence with good functional and cosmetic results[9]. The use of physical adjuvants as cryosurgery and phenol in combination with curettage together with the use of bone cement and bone grafts to preserve shape and strength of the bone helps to achieve good results and limits the indications of resection and amputation. Bone cement compared to bone graft provides immediate support and allows for intensive curettage even of large tumors[10].

Cryosurgery extends the margin of simple curettage, making it biologically equivalent to wide intra-compartmental resection. Cryosurgery entails using a wide excision in situ but without the morbidity of en bloc resection and the need to sacrifice the joint with low rate of local recurrence[11]. Corticocancellous grafts are required to strengthen the subcortical bone; whereas fibular struts reconstitute the cortical defects[12].

In our study, different treatment modalities were used including curettage, curettage combined with adjuvant therapy, and resection and amputation. The highest rate of local recurrence was in cases treated by curettage alone where two of four cases (50%) had local recurrence within one year of treatment. This result was supported by Persson et al.[12] also reported 40% recurrence rate for GCT treated with curettage alone. With respect to curettage and bone cement, the reported rate of local recurrence by O'Donnell et al.[13] using curettage and bone cement was 33.3% that decreased to 16.6% when mechanical burr was used, so they recommend using the burr at the end of all procedures.

In addition, when the lesion reached the subchondral bone in weight bearing areas they put a layer of bone cement first under the subchondral plate to support it and then fill the rest of the cavity either by bone cement or bone graft. Use of barium-impregnated cement allows for early detection of the recurrence because of contrast between it and the bone[14]. In our study, we used curettage and bone cement in 7 patients. We did not use mechanical burr in any of our cases, and we did not use barium-impregnated cement in any of our cases. Local recurrence developed in 3 cases (40%). Curettage and cryosurgery with bone cement or bone graft were done in 22 patients with lowest rate of local recurrence occurring in 4 cases (18%). This result is in harmony with the results of other authors who reported recurrence rates lower than 15% in cases of GCT treated with curettage and cryosurgery.[7,12]

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[15,16].

Follow-up for three year revealed that two patients with GCT of distal femur and proximal tibia had recurrence out of sixteen patients with deferent sites of bone affection. They were treated primarily by bone resection. Amputation was reserved for massive recurrence and malignant transformation and it was done for 3 patients in our study in distal femur, proximal tibia and proximal radius.

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[17]. Radiotherapy can be used as an alternative to surgery in cases that cannot be treated with surgery or left with severe disfigurement after surgery[9,18]. In our study we did not use radiotherapy in the treatment of our patients.

CONCLUSION

The main primary treatment of GCT is surgery, the type of which depends on preoperative evaluation which includes clinical evaluation that involves the site and size of the tumor in relation to surrounding structures, together with plain X-ray, CT scan and/or MRI as indicated and tissue biopsy to define tumor grade. Curettage alone results in high rate of local recurrence. On the other hand, curettage and adjuvant cryosurgery using bone cement or bone grafts give low rate of local recurrence. Resection is recommended for stages IB and IIB, extremely large lesions, and in cases where resection results in no significant morbidity as proximal fibula and flat bones. Amputation is preserved for massive recurrences and malignant transformation.

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