VIDEO OF ORTHOPAEDIC TECHNIQUE

Limb Salvage by Distraction Osteogenesis for Distal Tibial Osteosarcoma in a Young Child: A Case Report

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Under the effective chemotherapy protocol, physeal distraction could be used as an effective limb salvage in the growing children with lower-limb malignant metaphyseal bone tumours and could actively prevent leg length discrepancy or malalignment that profoundly affects function. In this paper, we report a 7-year-old boy with right distal tibial osteosarcoma but not yet invaded the epiphysis, which underwent surgical treatment after effective chemotherapy. The surgical procedure included two phases: epiphysiostasis (physeal distraction) and en-bloc resection of tumor and distraction osteogenesis. In the first phase, after osteotomy of the proximal fibula was performed, two Ilizarov rings were attached to the proximal tibia and one Ilizarov ring was applied to the distal tibia. At same time, U-shape ring as an important external fixator was been used in the axis of rotation of the ankle joint in order to balance the stress from the surrounding tissues, increase distracting stabilization and prevent the deformity or contracture of ankle joint. In the second phase, the tumor bone (5.7 cm) was en bloc resected by diaphyseal osteotomy. Distraction osteogenesis was commenced 1 week postoperatively at a rate of 1 mm twice a day. The patient was reviewed by X-ray intermittently. The new bone was formed and the entire bone defect was covered in four months after the operation. Doxorubicin and cisplatin as the neoadjuvant chemotherapy protocol are effective to osteosarcoma and chemotherapy process did not adversely affect the union. Through this technique, the right distal tibial epiphysis was reserved successfully. At the last follow-up, there are no local recurrence or metastasis and we achieve to prevent leg length discrepancy or malalignment that profoundly affects function.

Key words: Bone tumor; Children; External fixation; Physeal distraction

Introduction

In children and adolescents, 75% of malignant bone tumors are located close to the growth plate¹. Limb-sparing surgery in young children is challenging because of the expected future longitudinal and radial growth of the unaffected limb, which can lead to limb length discrepancy and malalignment that affects function profoundly. In cases of malignant metaphyseal bone tumors in the lower limb that have not invaded the epiphysis, physeal distraction is an effective form of limb salvage that can prevent subsequent limb-length discrepancy and deformity. Cañadell *et al.* were the first to describe this procedure and apply it in practice. The Cañadell technique usually includes two phases: separation of the epiphysis from the tumor-bearing metaphysis and en bloc resection of the tumor².

Most malignant bone tumors affect the proximal end of the tibia or humerus, or the distal end of the femur; they rarely occur in the distal tibia. This report describes a surgical procedure for limb salvage in a child with a distal tibia osteosarcoma, the procedure comprising physeal distraction, en bloc resection of the tumor and reconstruction of the bony defect by Ilizarov ring fixation.

Case Report

A 7-year-old boy presented with right distal tibial intermittent pain for 2 months. Examination revealed slight local swelling without superficial erythema; the movement of the right ankle joint was normal. The patient had recently noticed a gradual increase in the area of sensitivity along

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with swelling after exercise. Anteroposterior and lateral radiographs of the right tibia showed an osteolytic lesion containing periosteal proliferation and Codman's triangle in the distal third of the tibia. A CT scan showed an area of flocculent and spongiform destruction in the tibia, suggesting a diagnosis of bone tumor that had been progressively destroying normal cortical bone. MRI showed that the lesion in the distal tibia had low signals on T1WI images and high signals on T2WI/fat-(inhibition) images. The tumor appeared to be in contact with the physis, but because no signals were transgressing or crossing the adjacent physis, surgery was considered indicated on the basis of MRI findings. Histologic examination of a CT-guided needle biopsy showed that the tumor had the characteristics of osteosarcoma.

The systemic treatment of resectable osteosarcoma usually consists of preoperative chemotherapy, definitive surgery and adjuvant chemotherapy. This patient received preoperative chemotherapy with two cycles of doxorubicin 30 mg/m² on days 1–3 and cisplatin 120 mg/m² on day 1 in our department. The patient's main symptom, pain, was relieved by the preoperative chemotherapy and positron emission tomography-CT showed that the maximum standardized uptake value (SUV-max) had declined significantly from 16.1 (pre-chemotherapy SUV, SUV1) to 8.2 (post-chemotherapy SUV, SUV2) in the distal tibia (SUV2/SUV1 = 0.51). According to the PET-CT scan, the range of focal hypermetabolism and volume of the tumor had reduced markedly. A doxorubicin and cisplatin neoadjuvant chemotherapy regimen is very effective and safe in patients with osteosarcoma.

Surgical Procedure

The surgical procedure consisted of the following two phases.

Phase 1: Epiphysiolytic (Physal Distraction) under General Anesthesia

First, osteotomy of the proximal fibula was performed via a lateral approach at the level of the middle of the lower leg. Secondly, two Ilizarov rings of appropriate size were attached to the proximal tibia and fixed by two tensioned Kirschner wires. After this, in a key part of the procedure, two Kirschner wires were inserted into the epiphysis under imaging and tensioned to an Ilizarov ring. Third, in order to strengthen the stability of the Ilizarov ring and prevent deformity of ankle joint, a U-shaped ring was added to the foot. The calcaneus and metatarsal bones were fixed to the U-shaped ring by two and one Kirschner wires, respectively. Finally, the rings were connected by distraction rods. The area of distraction of the left distal tibia became painful on postoperative Day 8; by which time there was radiographic evidence of division of the physis.

Phase 2: En bloc Resection of Tumor and Distraction Osteogenesis (Bone Transport)

First, the 5.7 cm bone tumor was resected en bloc by diaphyseal osteotomy via an anterior approach to the distal tibia, achieving a wide margin. Although the prior MRI had not detected tumor in the epiphysis, the resected tumor was sent immediately for intraoperative histological examination, which showed absence of tumor at the edges of the resected segment. Lengthening of the transport segment was commenced 1 week postoperatively at a rate of 1 mm twice daily. The patient was reviewed by X-ray every month for the first 6 months. New bone had formed and the entire bone defect been covered by 4 months after the operation. The distraction index was 17.9 days/cm, the maturation index 26.7 days/cm and the external fixation index 44.8 days/cm. Two years after the operation, the patient had no leg length discrepancy and no problems with the activities of daily living. When the regenerated bone had consolidated as indicated by callus formation on at least three sides of the bone, the fixator was removed completely and activities of daily living encouraged. At the 2-year follow-up, the patient remained symptom- and disease-free with good radiological evidence of consolidation of bone.

Discussion

Physal distraction, a type of distractive technique that is achieved by external fixation, has a number of advantages in limb-saving surgery in children. In tumor surgery, physal distraction may provide a safe margin of excision. First, epiphysis preservation with preoperative physal distraction may provide a safe margin of resection, thus minimizing the risk of tumor recurrence. Second, physal distraction enables preservation of the epiphysis, which in turn enables the bones of children and adolescents to grow and thus lengthen the affected limb. Third, limb-saving by epiphyseal preservation has the advantage of preserving joint function. Finally, when compared with other epiphyseal-sparing procedures such as transepiphyseal resection or multiplanar osteotomy, physal distraction has the advantage of greater intraoperative safety.

To achieve satisfactory clinical results, the indications and contraindications for physal distraction must be understood. The key factor is whether the tumor has crossed the physis. San-Julian *et al.*, have published a summary of the MRI-based indications for tumor resection with physal distraction with the aim of preserving the epiphysis³. Physal distraction is indicated for pediatric bone sarcomas located in the metaphysis. The physal cartilage has to be open and the tumor must not have transgressed the physis. If the tumor has crossed the physis, or is in contact with all of the physis, or the patient has presented with a pathological fracture, preservation of the epiphysis is contraindicated. Partial contact between the tumor and physis (contact area <50% of

the physis) is a relative indication for physeal distraction. San-Julian *et al.* reported promising results from physeal distraction, even when the tumor was in close contact with the physis. However, Cañadell *et al.* recommend intraoperative checking of histological findings. If tumor cells are found in the physeal margin of the resection, transepiphyseal or epiphyseal resection is indicated². Our patient with osteosarcoma that was in partial contact with the physis according to MRI had relative indications for physeal distraction according to San-Julian *et al.*'s criteria. We therefore followed Cañadell *et al.*'s suggestion; the intraoperative histology was negative.

Bone stability during the distractive period depends on the stability of the external frame, which can be increased by using two rings instead of one for each bone segment. On the basis of two tibial and one epiphyseal ring, a U-shaped ring was applied to increase stability at the axis of rotation of the ankle joint and connected to the epiphyseal ring. The ring frame was linked by the distraction or fixation rods and the underlying bone stabilized by using transfixion wires and half pins. A U-shaped ring can be used to share and balance the stress from the surrounding tissues. At the same time, it promotes favorable results by augmenting or providing primary stabilization of the ankle joint and midfoot. The U-shaped ring was removed in a second procedure to preventing contracture of the ankle joint. In this second procedure, a Kirschner wire was run through the tibia and fibula and two half-pins were added to the proximal tibia and transport bone to facilitate distraction osteogenesis.

Since the incorporation of chemotherapy, multimodal treatment has improved survival to approximately 70% at 5 years with little improvement in the last two decades⁴. Neoadjuvant chemotherapy is currently an important element of the treatment of adults or children with osteosarcoma; it usually comprises four active agents, namely methotrexate with leucovorin rescue, doxorubicin, cisplatin and ifosfamide. However, the optimal combination of these drugs continues to be debated^{5,6}. A doxorubicin and cisplatin chemotherapy regimen has a long history and it has several advantages, including lower cost, more tolerable, moderately effective and good response in patients with osteosarcoma. When a good response has been identified by at least two of four radiological methods (plain radiography, MRI, angiography and ²⁰¹Ti scintigraphy) more than 90% tumor necrosis can be expected and a conservative rather than radical resection can be performed. We confirmed chemotherapeutic effects before operation by the above four methods and also by the change in SUV ratio (SCR index) of PET-CT examination. Bajpai *et al.* have reported that an SCR index of <0.48 indicates a good response to chemotherapy⁷. The SCR index in our patient was 0.51. Thus, a good chemotherapeutic response is a significant prerequisite for managing osteosarcoma and

evidence for such a response can include the following: reduction in pain; shrinkage and rigidity of any local mass; normal range of movement of the affected limb; reduction in SUV-max after chemotherapy compared with pre-chemotherapy values; sclerotic changes and clearly defined lesion margins visible on plain radiographs; and marked shrinkage of tumor, reduction of marrow edema, and marked shrinkage of any extension of the tumor into soft tissue on MRI.

Earlier reports have focused on the adverse effects of chemotherapy on bone transport. Ozaki *et al.* directly linked their poor results to adjuvant chemotherapy following tumor resection. Chemotherapy delays bone recovery, resulting in prolongation of external fixator time⁸. Cañadell *et al.* have also reported that chemotherapy has an adverse effect on bone consolidation⁹. Recent experimental studies have demonstrated that chemotherapeutic agents have no serious influence on callus tissue. Kapukaya *et al.* consider that the main cause of complications such as non-union and delayed union is the size of the transferred bone segment rather than the effect of chemotherapy or extension of the defect¹⁰. The most important factor influencing the duration of treatment is patient age, rather than chemotherapeutic agents. Tsuchiya *et al.* also reported that there was no significant difference in the duration of fixation or rates of healing when patients receiving postoperative chemotherapy were compared with those who did not receive chemotherapy after surgery; thus, they considered that chemotherapy has minimal impact on distraction osteogenesis and does not affect eventual bone healing¹¹. We reviewed the duration of distraction and fixation, postoperative chemotherapy and imaging information (X-ray) in our pediatric case at final follow-up. We also believe that postoperative chemotherapy for malignant bone tumors does not adversely affect achievement of union. However, given that this paper is only a case report, our data cannot clarify the relationship between chemotherapy and union.

Conclusions

External fixation technology can be safely used in limb salvage by physeal distraction in young children with distal tibial osteosarcomas and successfully combined with pre- or post-operative chemotherapy and the requisite fixation and limb lengthening. Postoperative chemotherapy for malignant bone tumors reportedly does not significantly negatively affect achievement of union and causes a minimal delay in achieving fixation and maturation in subjects undergoing distraction osteogenesis.

Video Image

Additional video images may be found in the online version of this article.

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