CASE REPORT

Successful management of a childhood osteosarcoma with epiphysiolysis and distraction osteogenesis

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ABSTRACT

In an 11-year-old boy with osteosarcoma in the proximal tibia (type III), 2 cycles of dia chemotherapy (cisplatin, ifosfamide, doxorubicin) were administered preoperatively while epiphysiolysis was performed. Clinical response was determined to be complete by radiography and histopathology. Marginal excision was then performed with preservation of the proximal tibial epiphysis. Metaphyseal reconstruction was performed using distraction osteogenesis. Six cycles of dia chemotherapy were administered postoperatively. Twenty months later, the patient had developed no complications and experienced full bone healing, with no limb discrepancy.

In selected adolescent patients with osteosarcoma, in whom the tumour is in full contact with the epiphysis, epiphysial preservation by epiphysiolysis and reconstruction by distraction osteogenesis can provide an excellent outcome, resulting in a stable reconstruction that functionally restores the native limb.

KEY WORDS

Osteosarcoma, epiphysis preservation, epiphysiolysis, distraction osteogenesis, limb function, quality of life

1. INTRODUCTION

In malignant bone tumours affecting children and adolescents, 75% are located close to the growth plate1. In children, treatment of juxtaarticular osteosarcomas near the knee with epiphysis preservation and biologic reconstruction using distraction osteogenesis can yield excellent functional results2. However, if all the tumour is in full contact with the epiphysis (type III), physeal distraction is contraindicated3.

Here, we describe a case of osteosarcoma in the proximal tibia (type III) that was successfully managed with epiphysiolysis and distraction osteogenesis. Related papers are also reviewed.

2. OSTEOSARCOMA MANAGEMENT

An 11-year-old boy presented in 2012 with pain in his right knee during running. He was referred to our orthopedic department after a radiograph showed an abnormal shadow.

This patient’s past medical history and family history included nothing of note. Physical examination revealed right knee edema and warmth. Serum phosphorus was 1.92 mmol/L (normal: 0.9–1.9 mmol/L), and serum alkaline phosphatase was 186 IU/L (normal: 45–125 mmol/L). Radiography revealed sclerotic changes and a periosteal reaction in the lateral part of the proximal tibia.

On magnetic resonance imaging (MRI), the lesion had homogeneous low signal intensity on T1-weighted images and heterogeneous high signal intensity on T2-weighted images, which demonstrated full contact between tumour and epiphysis. Angiography showed a tumour stain, and 99mTc scintigraphy demonstrated increased uptake in the proximal tibia.

A puncture biopsy was performed, and the tumour was histologically diagnosed as an osteosarcoma (Figure 1).

We administered 2 cycles of preoperative chemotherapy consisting of cisplatin 120 mg/m² (day 1), doxorubicin 30 mg/m² (days 1–3), and ifosfamide 2 g/m² (days 1–5)4. Consequently, the chief complaint of pain was relieved, and by angiography and 99mTc uptake, the tumour stain disappeared. Serum phosphorus and alkaline phosphatase concentrations declined to 1.06 mmol/L and 113 IU/L respectively. The tumour response was judged to be complete.

The surgical procedure consisted of two phases:

- In phase 1, two pins were inserted into the epiphysis and another two into the diaphysis 10 cm from the tumour. An external monolateral fixator with a 5-cm T-shaped piece for the epiphysial pins, strong enough for achieving epiphysiolysis, was attached5. Distraction began in the operating room and was continued at a rate of 1–1.5 mm...
daily until 1–2 cm of distraction was achieved. Distraction was maintained for the next 15 days, with the patient’s report of referred pain indicating rupture of the growth plate. Radiography showed disruption of the physis.

- In phase 2, a transepiphyseal resection was performed. En bloc resection of the tumour left a wide margin, but without exposing the metaphyseal surface of the physis (Figure 2). In the resulting surgical specimen, the histologic response to preoperative chemotherapy was classified as grade 4 (no viable cells).

Reconstruction involved bone transport using distraction osteogenesis. The metaphyseal defect was 7 cm. A medial gastrocnemius flap was transferred anteriorly, covering the bone defect. The patellar tendon then was reattached to the transferred gastrocnemius flap. A bone cylinder taken from the diaphysis was fixed using wires. Tibial alignment was not affected by the external fixation. Dressings were typically changed twice weekly. Active movement of the knee was initiated as bleeding and pain eased.

One week after the operation, the bone cylinder was applied to transport bone toward the residual epiphysis at a rate of 1 mm daily. Knee-motion exercises were started 3 weeks after the operation because the patellar tendon was reattached to the gastrocnemius flap. After 1 year, the external fixation was removed, and use of weight-bearing crutches was instituted. Six cycles of postoperative chemotherapy (same protocol as was used preoperatively) were given.

We evaluated the functional results and quality of life of this patient by physical examination, radiographs, and a questionnaire based on recommendations from the U.S. Musculoskeletal Tumor Society. Follow-up studies included clinical examination and imaging with plain radiography, computed tomography, and MRI.

Follow-up at 1 month, 3 months, 6 months, 9 months, 12 months, and 20 months showed no local recurrence in the epiphyseal region, no infection, no limb length discrepancy, and no other complication. The Musculoskeletal Tumor Society questionnaire scores at those visits were 26, 28, 27, 29, 31, and 32 respectively (Figure 3). The quality-of-life scores were 3, 3, 4, 4, 5, and 5 respectively. After 20 months, lung imaging by computed tomography showed no metastasis, and radiography showed full bone union (Figure 4).

This patient is still being followed.

3. DISCUSSION

3.1 Physeal Distraction

In tumours of the extremities or axial skeleton, the indications for limb salvage are the potential to achieve optimal surgical margins, moderate (or less) soft-tissue extension, no compromise to neurovascular bundles, absence of metastasis (or metastases that are responsive to curative treatment), and a patient that is in good clinical condition, free of infection, and compliant to treatment. The limb salvage procedure should not
delay adjuvant therapy; the reconstruction should be permanent and not be associated with a large number of local complications requiring secondary procedures and frequent hospitalizations; and the resulting limb function should approach that obtained by amputation. Optimal surgical margins around a malignant bone tumour are 3 cm of normal bone and 1 cm of normal soft tissue.

In most cases, especially those involving young patients, a 3–5 cm “safe” margin in osteosarcoma means loss of the adjacent joint when the tumour is in the metaphysis, usually leading to limb-length discrepancy. However, by definition, the wide margin is considered to be a layer of normal tissue—as opposed to reactive or inflammatory tissue—surrounding the tumour. Thus, a safe margin in the context of a metaphyseal tumour can be obtained without sacrificing the epiphysis. As described by Canadell and colleagues, physeal distraction in tumours that do not cross the growth plate provides a safe margin, while avertting loss of the epiphysis. They believe that the growth cartilage itself provides a dependable margin of safety and that the 3-cm to 5-cm margin suggested by most authors is unnecessary. The absence of anastomoses between epiphyseal and metaphyseal vessels means that, in cases in which imaging determines that the epiphysis has not been affected by the tumour, it is possible to conserve the epiphysis and the joint while resecting the tumour. This approach is made possible by physeal distraction according to Canadell’s technique. In tumour surgery, physeal distraction allows for preservation of the epiphysis in the growing bone and can provide a safe margin of excision. It is not used for bone lengthening, but is the first part of a tumour resection that allows for separation of the epiphysis from the tumour-bearing metaphysis.

The indications for physeal distraction in pediatric bone sarcomas are a tumour location in the metaphysis, an open physeis, and a tumour that has not transgressed the physeis. If the tumour is in contact with part of the physeis, physeal distraction can be tried. In evaluating physeal tumour involvement, MRI is the imaging method of choice, dividing tumours near the epiphysis into three types. As described by San-Julian, if the entire physeis is affected, the possibility of tumour cells passing the physeis is great, and so physeal distraction is contraindicated. Crucial factors are a positive response to chemotherapy, accurate preoperative assessment of tumour extension to the epiphysis by MRI, and appropriate reconstruction techniques. However, the imaging techniques currently in use, particularly MRI, can safely and accurately define tumour limits in patients who have a metaphyseal osteosarcoma growing toward the epiphysis in the knee joint. After their experience of 13 patients with a high-grade metaphyseal osteosarcoma, Muscolo et al. concluded that preservation of the epiphysis in high-grade metaphyseal osteosarcoma at the knee is an alternative in carefully selected patients in which the tumours are fully in contact with the growth plate. The reason is that MRI showed a sensitivity of 100% in detecting epiphyseal extension of osteosarcoma, with only a few false positives having been reported. Thus, MRI is a sensitive indicator of epiphyseal involvement by osteosarcoma, but its specificity is lower, sometimes overestimating the extent of the tumour.

Based on the foregoing considerations, we treated our patient (a defined type III) with physeal distraction before partial preservation of the epiphysis.

### 3.2 Distraction Osteogenesis

An ideal reconstruction should demonstrate biologic affinity, resistance to infection, and sufficient biomechanical strength and durability. Reconstructive procedures include distraction osteogenesis, autografted autogenous bone supplemented with a vascularized fibular graft, and frozen allograft. All of the relevant studies describe various complications, but no recurrences at the retained epiphysis.

Distraction osteogenesis, which was established by Ilizarov and Lediaev in the 1960s, harnesses the bone’s intrinsic regeneration potential by performing an osteotomy followed by bone distraction as a suitable approach to repairing large segmental bone defects. The advantages of the approach are the possibility to...
correct deformities\textsuperscript{18}, the small area of the associated soft-tissue defects, and immediate mobilization. Its disadvantages are the long duration of the external osteosynthesis materials, the frequency of pin-tract infection, and the pain accompanying transport\textsuperscript{19}.

Yokoyama \textit{et al.}\textsuperscript{16} used free vascularized fibular grafts to treat 4 patients with tibial bone defects and distraction osteogenesis to treat another 4. The mean defect length in the vascularized bone graft group was 7.3 cm; in the distraction group, it was 4.6 cm. The union rate of the distraction group was 100%; in the vascularized graft group, it was 75%. External fixation has been used safely and successfully during postoperative chemotherapy for fixation and distraction osteogenesis\textsuperscript{20}. Postoperative chemotherapy for malignant bone tumours has been reported not to adversely affect the ability to achieve union or cause hypertrophy, and to have a minimal effect on distraction osteogenesis\textsuperscript{20}.

Distraction osteogenesis avoids some complications, but might lead to others that require appropriate management: for example, deep infection, leg-length discrepancy and deformity, and even fracture\textsuperscript{21}. Fortunately, we encountered no such complications during our patient’s treatment.

The critical conditions for the success of the technique are a lesion no more than 15 cm in length, the availability of at least 0.5 cm of subchondral bone and sufficient remaining metaphyseal cortex after excision of the tumour, and a good response to chemotherapy\textsuperscript{2}.

4. CONCLUSIONS

In selected adolescent patients with osteosarcoma in which the tumour is in full contact with the epiphysis, epiphyseal preservation by epiphyseolysis and reconstruction by distraction osteogenesis can provide an excellent outcome, resulting in a stable reconstruction that functionally restores the native limb.

5. CONFLICT OF INTEREST DISCLOSURES

The authors have no financial conflicts of interest to declare.

6. REFERENCES


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