

Total Femur Replacement Using a Humeral Reconstruction Component in a 36-month-Old Ewing Sarcoma Patient

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KEY WORDS: custom-made implant, endoprosthesis, Ewing sarcoma, malignant bone tumor, total femur replacement

IMAJ 2019; 21: 833–834

Ewing sarcoma is a primary malignant bone tumor usually occurring during childhood and adolescence. It is most often situated at the diaphysis of long bones, and treatment consists of a multimodal therapy involving chemotherapy, surgery, and/or radiation therapy [1,2]. Multi-agent chemotherapy regimens are very effective and patient survival is increasing; therefore, surgery aims at limb preservation using reconstructive techniques. Aside from biological, composite, and ablative procedures, modular endoprosthesis replacements of these large bony defects have become

the most common and accepted operations performed in adolescent and adult patients [3]. In infants and young children, however, biological reconstructions, rotationplasty, and amputation are still considered to be feasible therapeutic alternatives [4]. Efforts have been made to implement growing prostheses as a means of reconstruction for these patients [5]. However, because of differences in the anatomy of skeletally immature patients (small bone diameter and length, less soft tissue coverage, closer proximity to neurovascular bundles, and loss of growth plates resulting in limb length discrepancies), tumor specific endoprosthesis replacements have not been established as a standard reconstruction procedure in small children.

We describe the case of a 36-month-old girl who was referred to our hospital after initial diagnosis of Ewing sarcoma in

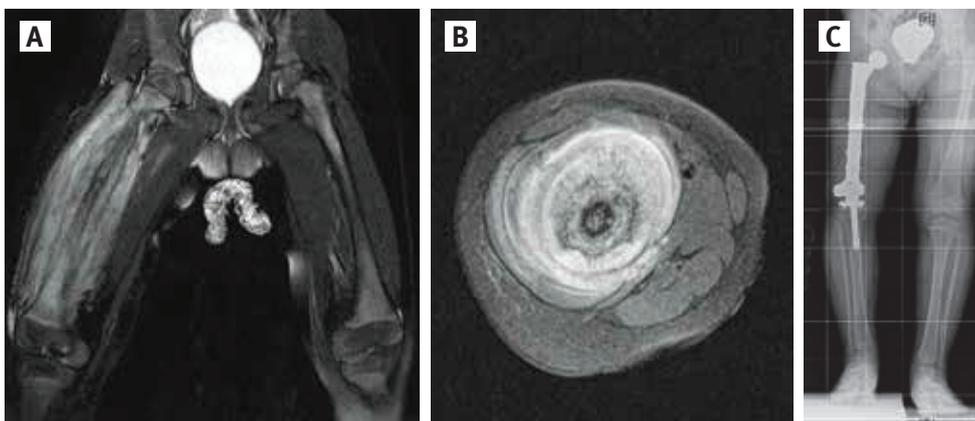
Russia. She was treated with wide tumor resection of the entire femoral bone and limb reconstruction using a humeral tumor endoprosthesis component to replace the femur, which was similar in size to her femoral bone, and a custom-made tibia implant. To the best of our knowledge, this is the first report of such a surgical procedure. The current follow-up was 30 months. Written informed consent was obtained from the patient’s mother (legal guardian) for publication of this case report and any accompanying images.

PATIENT DESCRIPTION

A 36-month-old Russian girl presented to our clinic after being diagnosed with Ewing sarcoma of the right total femur (Figure 1A, 1B). Staging was negative for distant metastases. After neoadjuvant chemotherapy was started at a local hospital in Russia, it was concluded according to the EURO-E.W.I.N.G 2008 protocol in our pediatric oncology department.

The surgical options of amputation and rotation-plasty were refused by the patient’s parents who consented for their daughter to undergo an experimental treatment aiming at limb salvage using a tumor specific endoprosthesis replacement. To achieve wide tumor margins, a total femur resection was scheduled for the patient. With the femur only measuring approximately 22 cm in length, standard femoral prosthetic implants could not be used in reconstruction because of their size and a lack of soft tissue coverage. Therefore, custom-made prosthetic reconstructions using a distal

Figure 1. Per-treatment magnetic resonance imaging cross sections of patient’s right femur demonstrating all bone involvement **[A]** Sagittal T2 image **[B]** Coronal T2 image **[C]** X-ray demonstrating leg length discrepancy with no signs of loosening or local recurrence



humerus replacement component and a custom-made tibia plateau with a polished stem connected by a fixed-hinge joint were manufactured.

The operation was performed under general anesthesia using a lateral approach. Preservation of the major vessels and nerve proved to be possible and the total femur bone was resected performing intra-articular resections of both the hip and knee joint. After resection of the articulating face of the tibial plateau, the custom-made prosthesis was implanted without any complications.

A pathological examination found clear tumor margins and no vital tumor cells, corresponding with a regression grade 1 according to the Salzer-Kuntschik grade classification. Intra- and postoperative antibiotic prophylaxis was administered to minimize the risk of bacterial infection of the prosthesis. Following a wound-healing period of 20 days, chemotherapy was continued and the complete course was administered over the following months.

The patient completed her follow-up examinations in our out-patient clinic. At 30 months follow-up she could bear full weight without any walking aids. Motoric development was age-appropriate compared to her peers. The patient performed complex motion sequences such as slight running. The surgical wound healed completely and showed no signs of infection. The patient's range of motion in the knee joint was extension/flexion 0/90°. The current leg length discrepancy was 4.4 cm and the patient used compensating shoe lifts

(Figure 1C). Local MRI showed no sign of local recurrence and staging studies showed no hint of distant metastases.

COMMENT

The treatment of Ewing sarcoma and other primary bone sarcomas in infants and young child remains a challenge for orthopedic surgeons. Primary amputation and rotationplasty should be considered as the first choice of surgery in cases where an entire femoral resection needs to be performed for oncological safety. However, amputation and rotationplasty are rarely accepted among parents and patients, and the patients have an onset of sociopsychiatric disorders in the course of their life. Moreover, a patient's motoric development is severely disrupted at an early age.

In our case, we were able to show that endoprosthetic reconstruction is surgically possible using custom-made and humerus component implants (initial length achieved and prosthetic femoral diameter was similar to that of a healthy bone at the patient's age). Functional outcome proved to be satisfactory in this reported case at a short-term follow-up allowing the patient to interact as normally as possible with her peers.

However, our patient will grow and have increasing leg discrepancy and disability. One way to minimize the resulting leg discrepancy is the use of polished stems to preserve growth plate function despite the implantation of an endoprosthesis. Therefore, we propose that limb salvage surgery with humeral components or

custom-made endoprostheses is a viable treatment option in young children when ablative or psychologically challenging procedures are the only alternatives and patient acceptance for these treatments is poor. Additional evaluation of long-term follow-up in terms of leg preservation and functional outcome are necessary to determine whether endoprosthetic replacements will become a standard procedure in infants and younger children. Furthermore, a patient's agreement to multiple operations during their adolescence is necessary.

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Capsule

The ABCs of brain tumor prognosis

Meningiomas are brain tumors that are typically benign and curable by surgery. In about 20% of cases, however, the tumors recur, and then patient prognosis is poor. Tumor histopathology is currently used to predict which patients might benefit from more aggressive treatment, but this method can be inaccurate. **Patel** et al. used clinical, gene expression, and sequencing data to classify meningiomas from 140 patients. Their analysis identified three distinct groups of

tumors (A, B, and C), with type C being the most likely to recur. Type C tumors were characterized by altered activity of a protein complex that controls cell cycle progression. Importantly, this molecular classification system was a better predictor of prognosis than the method currently used in clinics.

Proc Natl Acad Sci USA 2019; 116: 21715

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