

A REVIEW OF THE MODERN TRENDS IN THE SURGICAL TREATMENT OF MALIGNANT BONE TUMORS IN CHILDHOOD

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Abstract - Osteogenic sarcoma is a primary bone tumor characterized by malignant cells producing osteoid matrix. Ewing's sarcoma is a rare highly malignant bone tumor and second in frequency after osteosarcoma in children. Current treatment of both entities is multidisciplinary and consists of preoperative chemotherapy (sometimes in combination with radiation therapy), surgery to remove the tumor, and postoperative chemotherapy. Surgical treatment takes the main place for local control with excision of the tumor from the affected limb, rotational plasty and limb salvage operations with subsequent reconstructions. Limb salvage surgeries allow limb preservation and functionality, and with long-term follow-up the survival rate could reach 90% in the first 10 years.

Key words: Osteosarcoma, Ewing's sarcoma, limb salvage surgeries.

1 Introduction

Osteogenic sarcoma is a primary bone tumor characterized by malignant cells producing osteoid matrix and is the second most common malignant bone tumor after myeloma (Fig. 1). It occurs with a frequency of approximately 6.5% of all primary bone tumors and tumor-like diseases and 20% of all primary malignant bone tumors^(1,2,3,5,6,8,14). Its population incidence is 2 cases per 1,000,000 people per year. It affects childhood and adolescence with the highest peak in the second decade - 50% of cases occur in it. Males are affected twice as often^(1,2,5,8,14).

Ewing's sarcoma is a rare highly malignant bone tumor, occurring at a frequency of 4-12% of all primary bone tumors and second in frequency after osteosarcoma in children. It was described by James Ewing in 1921 as "diffuse endothelioma of bone" and belongs to the group of primary neuroepithelial tumors, along with primitive neuroectodermal tumors (PNET), Askin's tumor, and the extrasosseous form of Ewing's sarcoma.

It can be found in the age from 5 to 50 years, but it is a tumor of the second decade (more than 50% of patients are between the ages of 10 and 19), twice as often affects the male sex. The preferred location is the pelvis and long tubular bones, with a primary diaphyseal location. Strong malignancy, characteristic histological findings and high radiosensitivity assign a special place to the neoplasm in bone pathology and give it great social importance ^(1,2,3,5,14,16,17).

Current treatment of Ewing's sarcoma is also multidisciplinary and consists of pre-operative chemotherapy (sometimes in combination with radiation therapy), surgery to remove the tumor, and postoperative chemotherapy. Surgical treatment takes the main place for local control with excision of the tumor from the affected limb, rotational plasty and limb salvage operations with subsequent reconstructions.

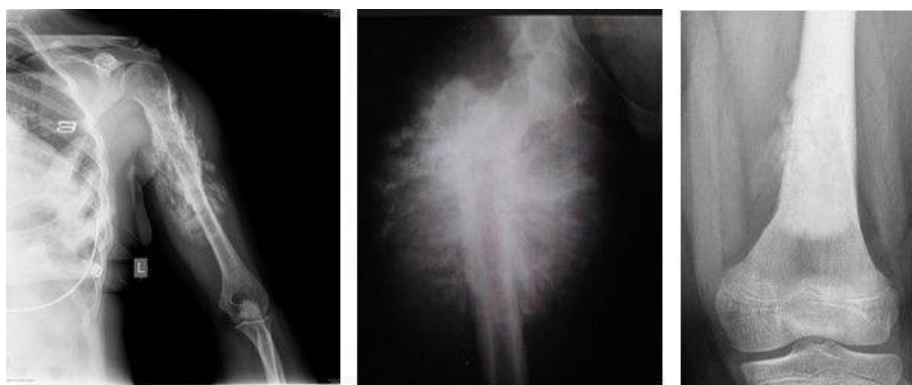


Fig. 1. – Conventional osteosarcoma involving the humeral shaft, proximal femur, and distal femur.

2 Materials and methods

Modern multimodal treatment of osteosarcoma allows limb salvage operations to be performed using modern modular prostheses - shoulder, elbow, hip, knee, ankle. Optimal tumor resection and a functional limb with increased patient survival are the goals of modern orthopedic oncology. Of utmost importance, before starting treatment, is the establishment of an absolutely accurate diagnosis, and this is where the biopsy comes

in. It must be performed by an experienced surgical oncologist who is part of the team that finally completed the operation (Fig. 2).



Fig. 2. – Cicatrix from a biopsy performed, which is excised when performing local control of the disease.

After making the correct diagnosis, preoperative chemotherapy is carried out, which aims to reduce the size of the tumor and reduce the risk of its dissemination. During this period, periodic control is carried out for influencing the tumor by MRI, CT of the lungs, PET/CT, blood analysis. Preoperative planning is mandatory and determines the exact bone resection and the appropriate size of the endoprosthesis with its constituent modules if reconstruction is performed by endoprosthesis (Fig. 3). After the resection of the bone (fig. 4), together with the tumor, comes the turn of the limb salvage surgery.



Fig. 3. – Preoperative planning of limb salvage surgery and reconstruction with MUTARS modular tumor endoprosthesis.

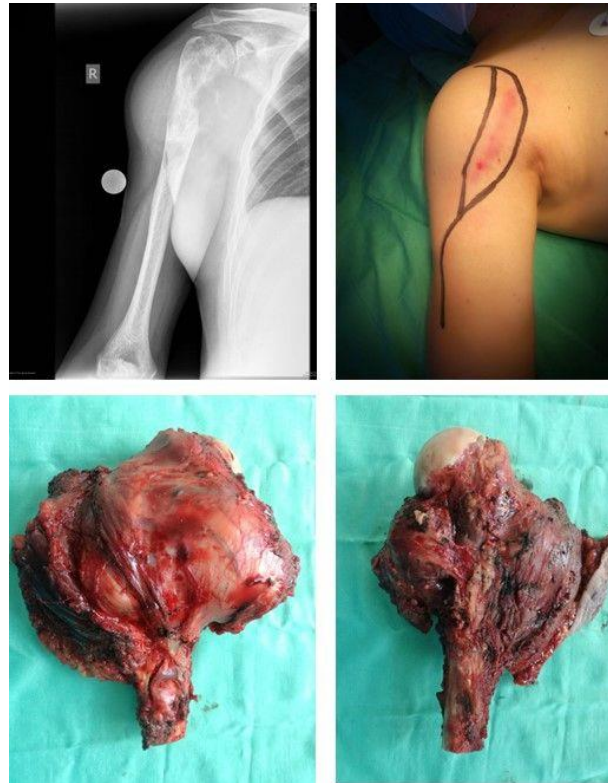


Fig. 4. - This 17-year-old patient with osteosarcoma of the proximal humerus underwent bone resection 20 cm distal to the humerus

But if the surgical procedure is carried out within sufficiently wide margins while preserving the limb and its functionality, it is possible that amputation will not be necessary⁽⁹⁾. Organ-sparing surgery within adequate resection margins can be achieved in many patients without compromising survival, but the risk of local recurrence can be significant, up to 19%^(1,2,10). Repair of large segmental defects after resection is a challenging task. The ideal reconstruction should be durable, compensate for the loss of growth of the affected limb in patients with incomplete growth, result in the function and appearance of the implants as close as possible to normal, be compatible with early rehabilitation, be cost-effective and easily accessible. Reconstruction with megaprotheses is a common method of reconstruction because it has a predictable functional outcome, allows for early rehabilitation, allows for intraoperative flexibility in the length of the reconstruction, is non-biological, and is unaffected by adjuvant chemotherapy. The main disadvantage of the megaprosthesis, however, is the vulnerability to wear and tear leading to loosening and/or fracture in the long term. In addition, tendon reinsertion to prostheses is another factor compromising functional outcome⁽¹¹⁾. For this reason, recently we use specially made for this purpose textile sleeves, with which

we "dress" the prostheses and attach the resected tendons to them (Fig. 5). Having an extendable ('growing') prosthesis minimizes the problem of limb length discrepancy in young children with significant residual growth, as they can be lengthened noninvasively⁽¹²⁾.

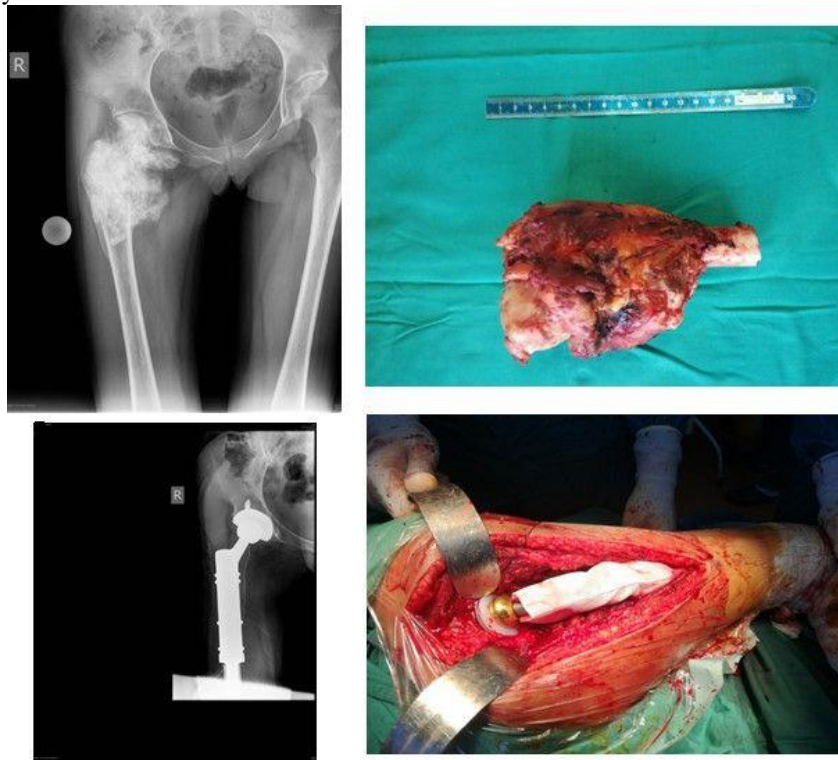


Fig. 5. - In this 17-year-old patient with an osteosarcoma in the proximal femur, 15 cm of bone was resected. Reconstruction was performed via MUTARS - proximal femur, using a "textile sheath" to reinsert the resected tendons.

However, limb salvage should be performed in a patient only if the surgeon is sufficiently confident that wide surgical excision of the tumor is feasible and that the expected function of the limb after limb salvage surgery will be superior to prosthetics after amputation or disarticulation. The operation is contraindicated in case of fungal or severe bacterial infection, tumor infiltration of large neurovascular structures, pathological fracture not affected by neoadjuvant chemotherapy⁽¹³⁾. Biological reconstruction is used for arthrodesis, reconstruction with inserted grafts, osteoarticular allografts. They offer the advantage of good tendon attachment for optimum function, especially at sites such as the proximal tibia, proximal femur, proximal humerus. However, the availability of allografts is increasingly limited, and problems with infection, graft fracture (Fig. 6), nonunion, and osteoarthritis are a cause for concern^(14,15).



Fig. 6 - Diaphyseal osteosarcoma located in the femur in a 14-year-old girl, responded very well to preoperative chemotherapy. A 21 cm resection of the diaphysis was performed and subsequent reconstruction with an inserted frozen diaphyseal allograft. 12 months postoperatively there is consolidation of the implant in the proximal and distal end. 15 months postoperatively, in case of trauma, fracture of the graft occurs.



Fig. 7 a,b,c,d - A 16-year-old patient with osteogenic sarcoma in the proximal humerus (a) who underwent alternative treatment instead of preoperative chemotherapy. This necessitated an operation immediately after the diagnosis (b) - resection of 14 cm of the humerus and reconstruction with MUTARS - proximal humerus. Post-operatively, the patient again turned to alternative methods of treatment and 8 months later - postoperatively, he presented with lung metastases and unfortunately with inoperable local recurrence (d).

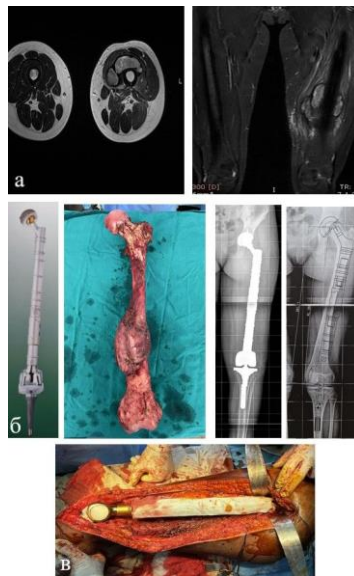


Fig. 8 a, b, c - Extrasosseous form of Ewing's sarcoma in a 12-year-old female patient. The operative procedure includes total excision of the femur and replacement with MUTARS – total femur (b, c). Currently - 24 months postoperatively there is no local recurrence and dissemination of the disease. The patient has normal motor activity, moves without aids.

3 Discussion

Patients treated with either limb salvage or amputation have similar survival, local recurrences, and complications, but better function is achieved in patients treated with limb salvage versus amputation. Local recurrences and complications are more common in limb-salvage patients. Optimal tumor resection and a functional residual limb with increased patient survival are the goals of modern orthopedic oncology⁽¹²⁾. Recent studies have surprisingly shown that the long-term psychological outcome of limb salvage surgery patients is the same as that of amputations⁽¹⁸⁾. It has been established conclusively that the survival of patients with osteosarcoma is not adversely affected by the choice of limb salvage surgery as a surgical treatment versus amputation⁽¹⁰⁾. Modern multimodal treatment of osteosarcoma and Ewing's sarcoma allows organ-preserving operations to be performed using modern modular prostheses. The type of reconstructive procedure is based on the anatomical location of the sarcoma, the potential for growth, the degree of bone and muscle resection required to remove the tumor, the skill of the surgical team. Close and active cooperation between the surgical and oncological team, periodic control of the effect of chemotherapy with a view to the preoperative preparation of the patient, follow-up throughout the treatment period and long afterwards are required. It is important that surgical treatment is carried out in specialized healthcare facilities by a team with experience in this area of bone pathology and reconstructive surgery. Certain cases require the work of a mixed team - vascular surgeon, neurosurgeon, in pelvic localizations - abdominal surgeon, gynecologist, urologist. Except for shoulder joint reconstructions, where movements remain significantly limited, the others allow patients to lead a fulfilling lifestyle.

4 Conclusion

Surgical treatment of osteogenic sarcoma and Ewing's sarcoma is part of the modern multidisciplinary approach to these diseases, which allows to preserve the limb and its functionality, and with long-term follow-up to reach a survival rate of over 90% in the first 10 years.

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