Journal of Surgery
Markeviciute V, et al. J Surg 7: 1538
www.doi.org/10.29011/2575-9760.001538
www.gavinpublishers.com
Case Report

Surgical Management of Pediatric Osteosarcoma: Case Reports

V. Markeviciute[1*], J. Ramonaite[2], E. Cekanauskas[3], M. Stravinskas[1]

[1]Department of Orthopaedics and Traumatology, Hospital of Lithuanian University of Health Sciences, Kaunas 50161, Lithuania

[2]Lithuanian University of Health Sciences, A. Mickevičiaus g. 9, Kaunas 44307, Lithuania

[3]Department of Paediatric Surgery, Hospital of Lithuanian University of Health Sciences, Kaunas 50161, Lithuania

[*]**Corresponding author:** Vetra Markeviciute, Department of Orthopaedics and Traumatology, Hospital of Lithuanian University of Health Sciences, Kaunas 50161, Lithuania

Citation: Markeviciute V, Ramonaite J, Cekanauskas E, Stravinskas M (2022) Surgical Management of Pediatric Osteosarcoma: Case Reports. J Surg 7: 1538. DOI: 10.29011/2575-9760.001538

Received Date: 20 July, 2022; **Accepted Date:** 26 July, 2022; **Published Date:** 29 July, 2022

Abstract

Background: Osteosarcoma is a primary malignant bone tumor most commonly presenting in children and young adults. Osteosarcoma has a peak incidence occurring during the ages of 10-14 years old. The first peak of osteosarcoma correlates with the increased proliferation of bone during the pubertal growth period. Tumor develops from primitive mesenchymal stem cells and had tendency for appearing in bones with rapid growth plate. Symptoms of osteosarcoma may be present from weeks to months and most commonly, the presenting symptom is bone pain, particularly with activity. After clinical manifestation and correct diagnosis, the next step is treatment to remove the tumors with chemotherapy and surgery. Complete surgical excision is important to ensure an optimum outcome. The main surgical challenge in children is how to reconstruct the limb after removal of the tumor.

Materials & Methods: Retrospective case reports analysis of patients treated for primary osteosarcoma in Hospital of Lithuanian University of Health Sciences Kauno klinikos.

Results: In this review, we summarized treatment of osteosarcoma with focus on different methods of surgery and present two cases from the Hospital of Lithuanian University of Health Sciences Kauno klinikos.

Conclusion: The optimal choice of surgical reconstruction following extremity osteosarcoma resection in pediatric population remains an enigma. However, the use of patient-specific instruments show good functional outcomes and patient acceptance. If possible, tumor resection through the epiphyseal line should be performed, thus preserving the function of the knee joint. the disease is rare, each patient's case must be evaluated, and the treatment strategy adopted individually.

Keywords: Endoprtohesis; Osteosarcoma; Patient-specific instruments

Introduction

Osteosarcoma is a primary malignant bone tumor most commonly presenting in children and young adults. Osteosarcoma has a peak incidence occurring during the ages of 10-14 years old. The first peak of osteosarcoma correlates with the increased proliferation of bone during the pubertal growth period [1]. Tumor develops from primitive mesenchymal stem cells and had tendency for appearing in bones with rapid growth plate. Osteosarcoma most often occur in the metaphyses of the lower extremity tubular bones. The population affected is predominantly children and young adults and incidence rate of 4.7 per million people in children and adolescents and causes 8.9% of cancer-related deaths in children mostly because of pulmonary metastatic disease [2]. The etiology of osteosarcoma remains obscure but there is an association between the quick bone growth happening throughout the puberty and osteosarcoma progression [3]. Symptoms of osteosarcoma may be present from weeks to months and most

commonly, the presenting symptom is bone pain, particularly with activity [4]. After clinical manifestation and correct diagnosis, the next step is treatment to remove the tumors with chemotherapy and surgery. Complete surgical excision is important to ensure an optimum outcome. The main surgical challenge in children is how to reconstruct the limb after removal of the tumor. In this review, we summarized treatment of osteosarcoma with focus on different methods of surgery, describe how patient-specific instruments are used to perform the resections of osteosarcomas. In this review, we summarize treatment of osteosarcoma with focus on different methods of surgery and present two cases from the orthopedic traumatology clinic in Hospital of Lithuanian University of Health Sciences (HLUHS). Both patients were the treated by the EURAMOS1 protocol MAP scheme. Three of the active drugs used include cisplatin, doxorubicin, and high-dose methotrexate; this combination (MAP), given preoperatively and postoperatively, is widely used for the treatment of osteosarcoma. Both patients got neoadjuvant and adjuvant chemotherapy. The patients are followed-up according to the hospital approved sarcoma protocol. Local magnetic resonance imaging and computerized tomography scans are performed, as well as whole body scans - initially every 6 months.

Case Reports

Case 1

In September of 2020 17-year-old patient was admitted to the emergency department due to pain in the thigh and knee and instability that had occurred the day before. X-rays and Computerized Tomography (CT) scan were performed: destruction of the lower third of the femur was observed. Magnetic Resonance Imaging (MRI) showed a tumor of about 3 cm in the distal diaphysis of the left femur, covering almost the entire cross-section of the bone with tumor infiltration, in distal femoral diaphysis and metaphysis up to about 17 cm circular reaction of the periosteum - sarcoma was suspected. Biopsy was performed - osteoblastic osteosarcoma, G3. Positron Emission Tomography (PET) scan was done to assess the spread of disease - a highly metabolic left femoral tumor without clear regional or remote metastases was confirmed. Chemotherapy was initiated in November 2020. In January 2021, PET scan was repeated to evaluate the dynamics: a tumor was significantly decreased but metabolic activity was still observed, new metabolically active foci were not detected. Surgery was planned for 8 weeks, while the components of the special oncological prosthesis and the approval of the State Health Insurance Fund regarding the financing of the prosthesis were received.

In March 2021, surgery was performed – resection of the distal part of the femur (18x8x5,5 cm), while preserving the knee joint. The blood vessels and nerves of the thigh were preserved. Patient specific instruments - individualized cutting guides were made for resection of the femur. Resection was performed 3 mm above the epiphyseal line, without damaging ligaments of the knee. Femoral resection was performed 18,5 cm above the knee, removing the femoral fragment together with the tumor. "MUTARS " endoprosthesis was used. A special individual component of the femur was inserted into the distal femoral metaphysis-epiphysis, and then additionally fixed with screws. Cementless segmental endoprosthesis was implanted in the femur. The patient underwent plastic reconstructive surgery of the front thigh muscle group. Histological examination of the postoperative material confirmed the pre-operative clinical diagnosis - osteoblastic osteosarcoma pT3N0M0 G3 IIB. Orpha: 668. Necrosis - 20%. Postoperative chemotherapy was started after the surgery. At present, treatment of the patient is complete and there is no evidence to suspect relapse and metastasis. During surgery the tumor was resected through the epiphyseal line using patient-specific instruments, thus preserving the knee joint. 3 years after the surgery, the knee and leg functions are fully restored. The functional results are excellent, the evaluation according to the MSTS (Musculoskeletal Tumor Society Scoring System) is very high – 30 out of 30 points (Figure 1).

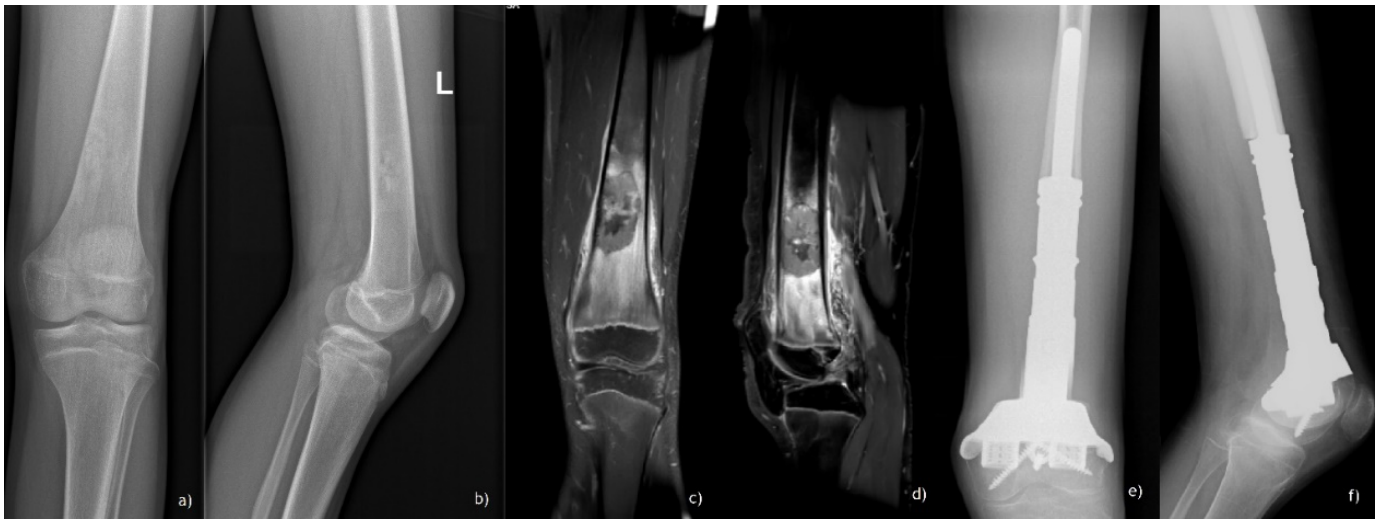

Figure 1: a,b) X-rays before treatment: destruction of the lower third of the femur. **C,d)** MRI: tumor of about 3 cm in the left distal diaphysis of the femur, covering almost the entire cross-section of the bone with tumor infiltration, in distal femoral diaphysis and metaphysis up to about 17 cm circular reaction of the periosteum. **E,f)** X-rays after femur resection and endoprothesis surgery

Case 2

In August 2017, a 16-year-old girl was consulted for left leg pain that lasted for 1 month. No history of trauma.

Solid, painful mass was palpated near the knee joint and the distal lateral part of the femur. No damage to the meniscus or ligaments were observed and movements were not limited. Knee X-ray was performed - bone tumor was suspected. Femoral MRI - the distal side of the left femur at about 20cm high - altered, uneven structures, destructive changes which included distal diaphysis, metaphysis, and almost the entire epiphysis. In the dorsolateral part of the distal metaphysis, in 9th cm of length, a 1,6 cm thick extraosseous tumor component was detected, without invasion to muscle or joint. The lesions were suspected to be osteosarcoma. Same month tumor biopsy results were received - femoral osteosarcoma, G3. In thoracic / abdominal CT no tumor process was seen. PET / CT confirmed left femoral osteosarcoma without clear regional or distal metastases.

In September 2017, chemotherapy was started. Wide tumor resection with left knee and partial femoral prosthesis implantation using MUTARS endoprosthesis was performed in November 2017. Histological examination of the postoperative material confirmed the pre-operative clinical diagnosis - osteoblastic osteosarcoma pT2N0M0 G3 IIB. Necrosis - 20%. Postoperative chemotherapy was started after the surgery. 2019-03-05 MRI was performed on an outpatient basis - examination of the lower 2/3 of the thigh and the upper 1/3 of the calf - on the right side of the femur prosthesis, a stretch of homogenous fluid of up to ~1,6 cm remains visible, extending upwards ~20 cm from the joint, communicating with the knee joint cavity at the bottom, no clear signs of tumor recurrence are visible. Punction results - seroma (about 300 ml of fluid was obtained). At present, treatment of the patient is complete and there is no evidence to suspect relapse and metastasis. The functional results are excellent, the evaluation according to the MSTS (Musculoskeletal Tumor Society Scoring System) is very high - 28 out of 30 points. The course of the second clinical case was more complicated, longer rehabilitation was required until the maximum leg function was restored, therefore it is recommended to always perform knee joint-preserving surgeries whenever possible. Especially in children, performing resections through the epiphyseal line (Figure 2).

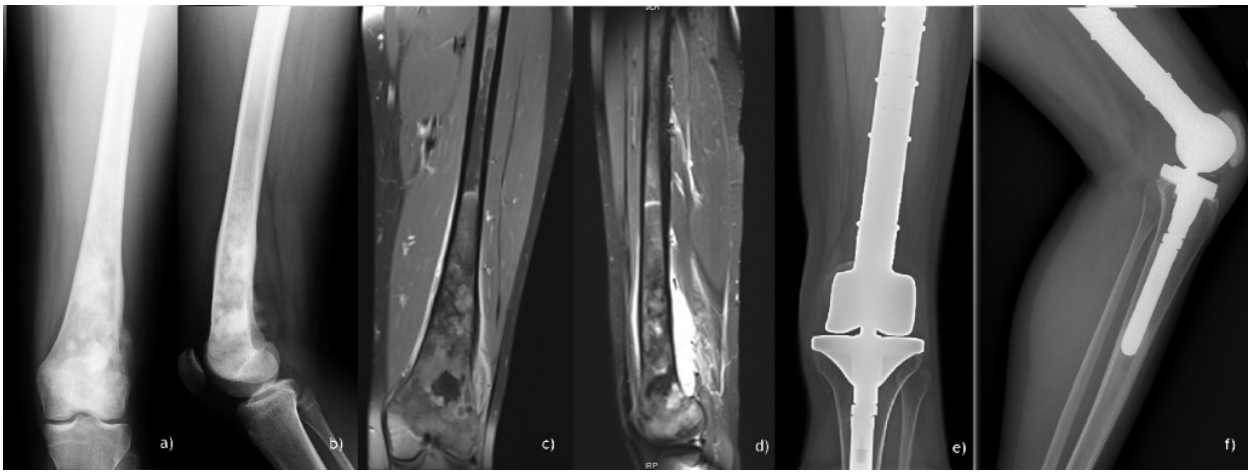

Figure 2: a,b) X-rays before treatment: destruction of the femur. **C,d)** MRI: the distal side of the left femur at about 20cm high. In the dorsolateral part of the distal metaphysis, in 9th cm of length, a 1,6 cm thick extraosseous tumor component was detected, without invasion to muscle or joint. **e,f)** X-rays after femur resection and endoprosthesis surgery

Discussion

Patients have symptoms for several months before a diagnosis is made. Pain is the most common symptom for osteosarcoma. Parents are often concerned that their child has incurred a sprain or growing pains but there may or may not be a reported history of traumatic injury [5]. The pain is constant and gets worse over the time. However, in the early stages, the tumor cannot be palpated and may not even show up on X-ray images [6]. The examination of a patient with suspected osteosarcoma begins with a full history, physical examination and X-ray imaging. Pain and swelling at the site of the primary tumor are usually noted in the history. Physical examination usually reveals a soft-tissue mass at the site of the primary tumor [7]. The current management strategy for newly diagnosed osteosarcoma includes neoadjuvant chemotherapy followed by surgical removal of the primary tumor, plus the addition of adjuvant chemotherapy after surgery. Neoadjuvant and adjuvant postoperative chemotherapy for patients in our hospital was administered according to the EURAMOS-1 protocol. The European and American Osteosarcoma Study (EURAMOS) collaboration, initiated by four internationally recognised study groups, was formed to improve outcomes in osteosarcoma [8]. The EURAMOS-1 collaboration agreed on a standard of care for osteosarcoma chemotherapy, in which there had been various approaches used. Accordingly, the three-drug combination with methotrexate, doxorubicin and cisplatin (MAP) following the previous COG trial was defined as standard chemotherapy [9]. The main goal of osteosarcoma surgery is the complete resection for primary tumor control. Children need to be operated differently from adults and treatment of skeletally immature children should consider any growth-related complications following tumor surgery [10]. Limb-length discrepancy is a major complication and extendable endoprostheses or distraction osteogenesis have been applied to address this problem [11,12]. One of the most important aspects to consider in children while using endoprosthesis for reconstruction is the expected limb length discrepancy. There are three different options based on estimated growth remaining at the resected physis when considering the right endoprosthesis for a growing child. When considering the choice of endoprosthesis, the growing child can be subdivided into three categories based on estimated growth remaining at the resected physis. The first, child approaching skeletal maturity with 2 cm or less of growth remaining. In this case, normal adult implants can be used with the residual limb length discrepancy being managed by an external shoe raise. The second group are the ones who are expected to have a limb length discrepancy between 2 and 6 cm. This group can be managed by using a growing endoprosthesis. The final group is one far from skeletal maturity with growth of more than 8 cm remaining. The only option is the use of growing endoprosthesis and also this group will require more than one growing prosthesis in addition to final conversion to an adult implant at skeletal maturity [13]. When the tumor is located close to the growth plates, transepiphyseal resection can save the epiphysis and is therefore advantageous in terms of preserving joint function. TER is an effective limb-salvage technique to treat malignant metaphyseal bone tumours in patients with osteosarcoma in the presence of open physes. When the tumor is not having contact with the physis, osteotomy is performimg through the epiphysis. When the tumor reaches the physis, an intraepiphyseal osteotomy is performed at least 3 mm from the physis. To strengthen the fixation between the epiphysis and the allograft, cannulated or cannulated screws

can be inserted diagonally into the allograft [14]. The concept of patient-specific instruments used in clinical cases at our hospital was first introduced in the 1990s by Radermacher et al [15].

Conclusion

The optimal choice of surgical reconstruction following extremity osteosarcoma resection in pediatric population remains an enigma. However, the use of patient-specific instruments show good functional outcomes and patient acceptance. If possible, tumor resection through the epiphyseal line should be performed, thus preserving the function of the knee joint. The limited availability and high cost also limit the use of expanding endoprosthesis, particularly in low-income countries. Endoprosthesis provide a good option for reconstruction in the pediatric population but the long course of follow up and multiple procedures required need to be well understood by the child, parents and the treating surgeon. The disease is rare, each patient's case must be evaluated, and the treatment strategy adopted individually in a specialized orthopedic oncology center.

References

1. Belayneh R, Fourman MS, Bhogal S, Weiss KR (2021) Update on Osteosarcoma. Curr Oncol Rep 23.
2. Jafari F, Javdansirat S, Sanaie S, Naseri A, Shamekh A, et al. (2020) Osteosarcoma: A comprehensive review of management and treatment strategies. Ann Diagn Pathol 49: 151654.
3. Bielack SS, Hecker-Nolting S, Blattmann C, Kager L (2016) Advances in the management of osteosarcoma. F1000Res 5: 2767.
4. Lindsey BA, Markel JE, Kleinerman ES (2017) Osteosarcoma Overview. Rheumatol Ther 4: 25-43.
5. Prater S, McKeon B (2022) Osteosarcoma. [Updated 2021 Jun 4]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing 2022.
6. Taran SJ, Taran R, Malipatil NB (2017) Pediatric Osteosarcoma: An Updated Review. Indian J Med Paediatr Oncol 38: 33-43.
7. Petriceks AH, Salmi D (2019) Educational Case: Primary Osteosarcoma. Acad Pathol 2019.
8. Smeland S, Bielack SS, Whelan J, Bernstein M, Hogendoorn P, et al. (2019) Survival and prognosis with osteosarcoma: outcomes in more than 2000 patients in the EURAMOS-1 (European and American Osteosarcoma Study) cohort. Eur J Cancer 109: 36-50.
9. Hazewinkel AD, Lancia C, Anninga J, van de Sande M, Whelan J, et al. (2022) Disease progression in osteosarcoma: a multistate model for the EURAMOS-1 (European and American Osteosarcoma Study) randomised clinical trial. BMJ Open 12: e053083.
10. Kang S, Lee JS, Park J, Park S-S (2017) Staged lengthening and reconstruction for children with a leg-length discrepancy after excision of an osteosarcoma around the knee. Bone Joint J 99-B: 401-408.
11. Kang S, Lee JS, Park J, Park S-S (2017) Staged lengthening and reconstruction for children with a leg-length discrepancy after excision of an osteosarcoma around the knee. Bone Joint J 99-B: 401-408.
12. Pala E, Henderson ER, Calabrò T, Angelini A, Abati CN, et al. (2013) Survival of current production tumor endoprostheses: complications, functional results, and a comparative statistical analysis. J Surg Oncol 108: 403-408.
13. Groundland JS, Binitie O (2016) Reconstruction After Tumor Resection in the Growing Child. Orthop Clin North Am 47: 265-281.
14. Kim Y, Jang WY, Park JW, Park YK, Cho HS, et al. (2020) Transepiphyseal resection for osteosarcoma in patients with open physes using MRI assessment. Bone Joint J 102: 772-778.
15. Bellanova L, Paul L, Docquier PL (2013) Surgical guides (patient-specific instruments) for pediatric tibial bone sarcoma resection and allograft reconstruction. Sarcoma 2013: 787653.