

CLINICAL CASE

THE MULTIDISCIPLINARY APPROACH IN A CASE OF PEDIATRIC EWING'S SARCOMA

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Abstract

Ewing's Sarcoma is a rare, malignant osseous tumor arising from undifferentiated cells originating from the neural crest; however it may also develop in the soft tissue. The initial evaluation of a patient who is suspected to be suffering from Ewing's Sarcoma entails taking a thorough medical history along with performing a detailed physical examination. Our patient was diagnosed with Ewing's Sarcoma upon seeking medical attention due to the sudden appearance of pain in his left hip. The medical staff conducted a series of investigations including X-rays, MRI, CT and 3D-CT scans of the inferior limb and pelvis in order to yield the diagnosis of Ewing's Sarcoma; however, the definitive diagnosis was confirmed only by the biopsy performed before the surgery for modular endoprosthesation. The patient received preoperative chemotherapy (in order to reduce the tumor dimension) according to the EuroEwing 2008 Protocol. After the seventh course of chemotherapy, surgery was performed, consisting of in-block resection of the proximal left femur and hip joint, followed by the insertion of a modular femoral endoprosthesis. Postoperative follow-up, however necessitated another six courses of chemotherapy.

Keywords: *Ewing's sarcoma, chemotherapy, EuroEwing 2008, block resection, modular endoprosthesis*

Introduction

Ewing's Sarcoma is a malignant osseous tumor, composed of small caliber round cells, with a strong metastatic potential. It affects 1-9 children per one million cases, appearing most commonly between the ages of 5 to 30 years, with an increased incidence between 12 and 18 years. Oftentimes, clinical symptoms are uncharacteristic, however, seldomly there may be swelling of the soft tissues accompanied by local inflammation. The most common symptom is pain; other symptoms are dependent on the organs in proximity to the tumor [1,3].

Case presentation

We present the case of a 10-year-old male patient of proper height and weight for his age, diagnosed with Ewing's Sarcoma in April 2015, who was admitted to the Pediatric Orthopaedics Department of the Emergency Hospital for Children "Maria Sklodowska Curie" in June 2015 for the confirmation of the probability diagnosis and proper treatment.

The symptoms began, suddenly, in January 2015 with the appearance of pain in the left hip. In February 2015, the pain worsened, as our patient began to experience difficulty in walking on his left leg. In this regard, an MRI scan of his

left leg and pelvis was performed which revealed a left intertrochanteric mass of 5.5 cm, invading into the cortical bone and extending into the regional soft tissue extension (Figure 1).



Figure 1 - Pelvis MRI. Intertrochanteric tumor, mostly diaphyseal

After this imagistic diagnosis, in April 2015 he is sent to the Pediatric Orthopaedics Department of the Emergency Hospital for Children "Maria Sklodowska Curie", Bucharest. For elucidation of diagnosis, a thoracic CT scan is performed which does not reveal any metastasis. CT and 3D-CT examinations showed a heterogeneous intertrochanteric fracture in the left femur (Figure 2).

In April 2015, for the diagnosis certainty, an open biopsy was performed. Regarding the TNM staging, the tumor was a stage I, T1N0M0 and an Enneking stage IIB, G2T2M0[2]. Preoperative diagnosis of Ewing's sarcoma in the left femoral neck (intertrochanteric) was suggested by the imagistic investigations, the diagnosis certainty being given by the histopathological examination which was followed by immunohistochemistry.

The patient had six preoperative chemotherapy cycles. The first round of preoperative chemotherapy according to the EuroEWING 2008 Protocol consisted of the following: Vincristine 2mg/day, Ifosfamide 4000 mg/day, Doxorubicin 27 mg/day, Etoposide 200 mg/day, Adjuvants: Mensa, Osetron, Filgrastinum.

Chemotherapy effects:

Postoperative chemotherapy induced aplastic anemia, { WBC 110 / mmc , Hb : 8.4 g / dL , platelets : 11000 / mmc } requiring two units of platelets. Following the appearance of fever,

blood cultures were harvested and empiric antibiotic therapy was initiated. Klebsiella pneumonia was isolated on culture media.

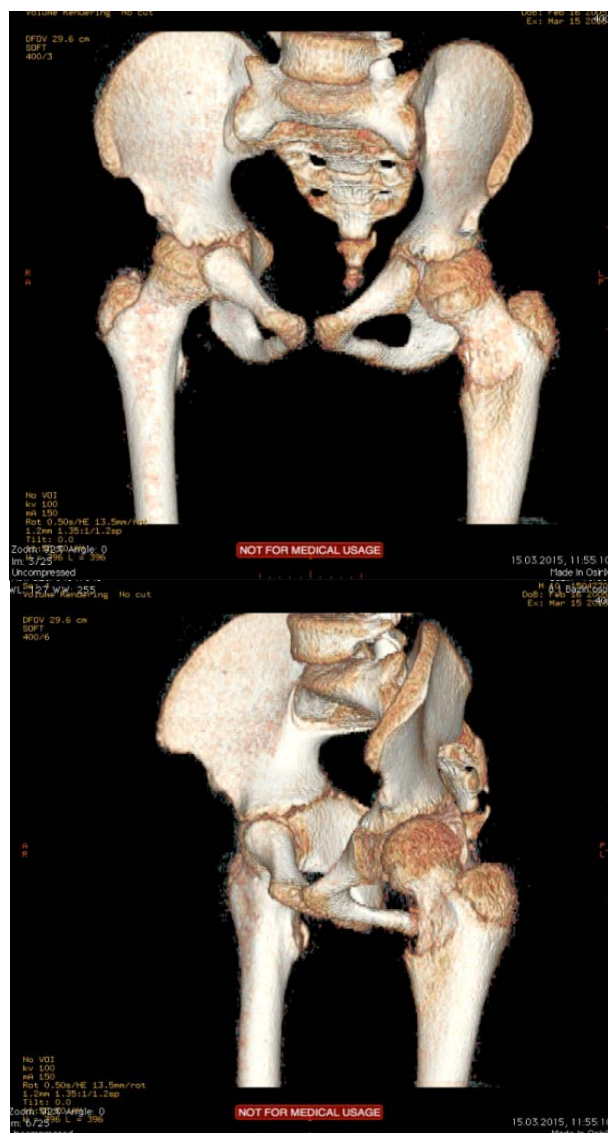


Figure 2 - 3D-CT exam illustrating the erosion of the femoral neck in the maximum resistance zone

The patient underwent a total of 6 cycles of VIDE chemotherapy, during which he required numerous blood product transfusions. All hemocultures were negative upon repetition. Normally, after completing the EWING Euro 2008 Protocol, the patient would have undergone surgery; however, due to the lack of an appropriate prosthesis, surgery was postponed and chemotherapy was continued, this time according to the VAI Regimen consisting of Vincristine 1.95 mg/day, Ifosfamide 3900 mg/day, Actinomycin, Cosmegen 1 mg of adjuvant Mesna and Osetron.

Imaging after chemotherapy described a shrinkage of the intraosseous lesion and adjacent periosteal reaction, as well as a reduction in the degree of soft tissue infiltration and absence of metastases (Figure 3). This evaluation helped us to convert the tumor from II B Enneking stage to II A.



Figure 3 - MRI after chemotherapy: tumor volume diminished



Figure 4 - Proximal extremity of the left femur with Ewing's sarcoma resected with the neighboring tissue

The patient is transferred to the Pediatric Orthopaedic Ward for surgery. Under general anesthesia, an anterolateral Smith-Petersen approach is used and the block left proximal

femur is resected along with the left hip joint and a left proximal femoral and coxofemoral joint modular endoprosthesis is inserted. Intraoperative frozen sections confirm the presence of malignant lesions on the resection limits. No intraoperative complications and accidents occurred.



Figure 5 - Longitudinal section resected showing a modified osseous structure on the femoral neck and intertrochanteric

To avoid the thromboembolic complication, anticoagulants were administered for 30 days.



Figure 6 - The difference of length between legs, the left one being longer

Postoperative follow-up was favorable, with progressive mobilization since the 3rd postoperative day.

Topographic examination showed a difference of length between legs with the shortness of the contralateral femur of 26 mm (Figure 6). The higher length of the operated leg

is done to annihilate the side effect of endoprothesation, so that when the boy is aged 22, the discrepancy between the left and right leg is not bigger than 0.5 cm, after Paley's scheme [3].

The patient was discharged from the Orthopedic Ward and transferred to the Oncology Ward to continue chemotherapy. The oncologist prescribed a 6-course VAI chemotherapy regimen from the EuroEwing 2008 Protocol. Interdisciplinary assessment and follow-up is imperative; oncologic re-evaluation was scheduled for the 21st postoperative day, followed by orthopaedic follow-up 6 months after surgery, which is planned to comprise clinical and imaging investigations.

Discussions

Prolonging the preoperative treatment with chemotherapy for 3 weeks was done so that we can use the most performant endoprosthesis that can be used in the case of a malignant bone tumor, to preserve the function of the pelvitrochanteric muscles.

The efficiency of the preoperative treatment indicated by EuroEwing 2008 Protocol can enable the change from Enneking stages IB or IIB to IA or IIA. When the tumor is

intracompartmental limited, the en bloc resection is efficient.

To diminish the length differences between legs, the patient will wear until the equalisation a plantar sustainer and plantar uplifting of 3 cm, thus the static and dynamic balance being ensured.

Conclusions

Timely diagnosis of malignant femoral tumor increases the chance of using an endoprosthesis which keeps the abduction and adduction of the legs. If patients are found in Enneking stage I, the en bloc resection and the modular endoprosthetic can be the only ways to cure them.

References

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