Scholars Journal of Applied Medical Sciences
Abbreviated Key Title: Sch J App Med Sci
ISSN 2347-954X (Print) | ISSN 2320-6691 (Online)
Journal homepage: www.saspublishers.com

OPEN ACCESS

Orthopedic Surgery

Extra-Bone Ewing Sarcoma about a Case and Review of the Literature

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| **Received:** 13.02.2019 | **Accepted:** 27.02.2019 | **Published:** 30.03.2019
DOI: 10.36347/sjams.2019.v07i03.013

Abstract **Case Report**

Ewing's soft tissue sarcoma is a rare condition that affects young adults. Primary neuroectodermal tumors (PNET) and soft tissue Ewing's sarcoma have long been considered as two distinct anatomo-clinical entities. More recently, it has been found that Ewing's sarcoma is only the most undifferentiated form of PNET. The diagnosis is essentially cytogenetics and immunohistochemistry. Given the absence of specific clinical and radiological signs, it seems necessary to include it in the differential diagnosis of any primary tumor of the soft tissues. The treatment is based on the combination of surgery, chemotherapy and radiotherapy. The high-dose multidrug therapy associated with hematopoietic stem cell transplantation significantly improves the long-term prognosis. We report, in this, work an observation of a patient who presents a Ewing sarcoma extra bone in the right leg.
Keywords: Ewing's soft, tumors, immunohistochemistry.

INTRODUCTION

Soft tissue Ewing's sarcoma (SEPM) was first described in 1969 by Tefft *et al*. It is a rare sarcoma; whose localization is variable. It originates in non-bone supporting tissues. In the absence of specific clinical and radiological signs, the diagnosis remains dependent on anatomopathological examination, particularly cytogenetics and molecular biology. SEPM has a poor prognosis. The mortality rate can reach 30 to 50%, it is related to the metastatic spread, mainly a pulmonary one.

OBSERVATIONS

This is a 25-year-old patient, with no significant pathological history, presenting for a year and a half a swelling in front of the upper 1/3 of the right leg of rapid evolution, painful, with no inflammatory sign opposite.

The clinical examination revealed a voluminous swelling of the upper 1/3 of the right leg. The knee joint was free. The somatic examination was normal, and no lymphadenopathy was individualized. The biological assessment was normal without inflammatory syndrome. The standard X-ray showed opacity next to the upper 1/3 of the right leg. The assessment was completed by an MRI showing a heterogeneous tumor process in hypersignal occupying the upper 1/3 of the right leg (Figure 1 and 2). A biopsy was performed, and the diagnosis of Ewing's sarcoma was made. The extension balance is negative. Initial multidrug therapy with cyclophosphamide and Adriamycin was initiated to reduce tumor size, and two months later the patient was rejected by an external way (Figure 3 and 4).

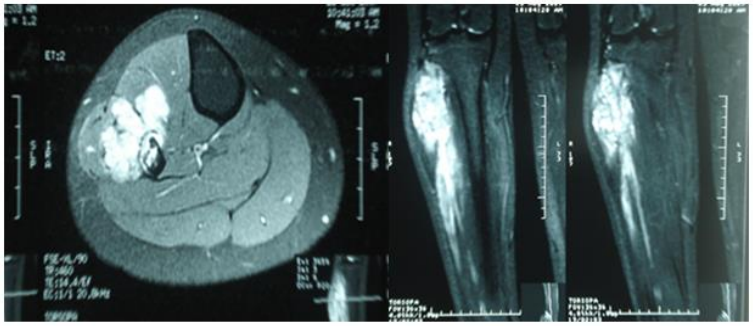

Fig-1 and 2: MRI of the right leg shows on an axial and coronal section the tumor at the external side of the leg in hypersignal T 2

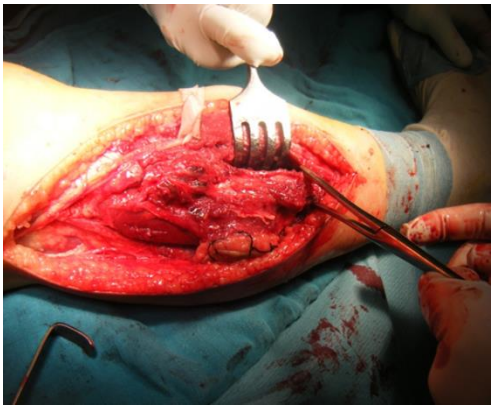

Fig-3: Intraoperative view of the tumor during surgical resection.

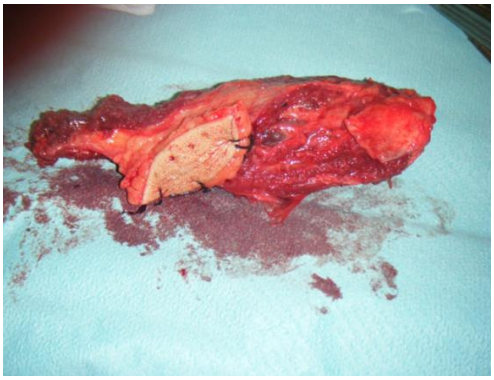

Fig-4: The surgical specimen including the scar of the biopsy

DISCUSSION

Ewing's sarcoma is a rare tumor that mainly affects young adults and affects both men and women [1]. For most authors, pain is the most common sign [1-3], it is often located at the tumor site but can be projected and does not give way to the usual analgesics. The swelling exists almost constantly, it is of varied volume. The radiological signs are, at the beginning, minimal and can escape the interpretation; the radiological aspect is very varied and is not always a characteristic.

Computed tomography gives more complete information about the bone, its environment, as well as the anatomical structures of the surrounding parts. MRI is currently the most effective examination, it replaces or supplements CT [3], and it allows better characterization of tumor elements and their environment.

The diagnosis of certainty is based on anatomopathological examination [4], which can be performed either with the trocar or with a surgical approach. Macroscopically, the tumor is often whitish, multi-lobulated, friable infiltrating and destroying all the plans in the area. Microscopically, these are small cells resembling other sarcomas with a mesenchymal ultrastructure.

The confirmatory diagnosis is essentially cytogenetics and immunohistochemistry, particularly thanks to in situ hybridization by immunofluorescence. 80% of the PNET / EES have a translocation (11,22) (q24, q12) involving the EWS gene (Ewing's sarcoma gene) and 5% to 10% a translocation (21,22) (q22, q12).

The prognostic factors are based on the tumor volume, the initial site of the tumor, the age of the patient, the existence of metastases at the time of diagnosis, the histological response to chemotherapy. Importance of the response to neoadjuvant chemotherapy: This is the most important prognostic factor [8]. A good response to primary chemotherapy is defined by a tumor residue <10%, that histopathological examination of the tumor after chemotherapy shows that less than 10% of cancer cells remain in the tumor tumor. Tumor Necrosis: The 5-year survival is 80% in the absence of tumor necrosis and falls to 50% when there is one. It is observed in 50% of sarcomas [7], however, there is no consensus on how to evaluate it.

The treatment is based on chemotherapy first, in order to have a volumetric regression of the tumor; this chemotherapy is based on polychemotherapy to potentiate their effects without increasing their toxicity. Since the use of intensive chemotherapy, the 5-year survival is close to 50 to 60% [5, 6, 9]. Surgery is an act both diagnostic and therapeutic, its goal is to perform a satisfactory oncological intervention passing in any point in healthy tissue, and it can be either conservative or radical.

Radiation therapy will be considered in the presence of metastatic Ewing's sarcoma, in case of certain inoperable tumor sites, or in cases of incomplete resection [5,6]. Currently, there is no consensus in the literature on the modalities and timing of Ewing's sarcoma monitoring.

Metastases are possible and the target organs are the lung, the skeleton, the liver, the brain, the peritoneum and the ganglia. The prognosis is a priori bad with a possibility of local recurrence or late metastases [6].

CONCLUSION

Ewing's sarcoma is a malignant tumor that develops frequently in bone tissue and rarely in the soft tissues. The positive diagnosis remains difficult even on the anatomopathological level. The therapeutic management is heavy and combines a local treatment based on surgery and / or radiotherapy to chemotherapy. Monitoring is clinical and radiological.

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