

Extraskelatal Ewing's Sarcoma of the Leg: A Rare Case Report

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Article History

Received: 05.05.2018

Accepted: 16.05.2018

Published: 30.05.2018

DOI:

10.36347/sjmcr.2018.v06i05.013



Abstract: Ewing's sarcoma is a rare tumor that mainly affects young adults, and affects both men and women. Extraskelatal Ewing's sarcoma is rarer, since only about 150 cases had been written in 1988. The authors report in this work a case of a patient who presents an extraskelatal Ewing's sarcoma in the right leg.

Keywords: Ewing's sarcoma, Extraskelatal, leg.

INTRODUCTION

Extraskelatal Ewing's sarcoma is a rare tumor; it mainly affects the young adult. The authors report in this work a case of a patient who presents an extraskelatal Ewing's sarcoma in the right leg.

CASE REPORT

A 25-year-old patient, with no significant pathological history, presented for a year a swelling in 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.

Standard radiography showed opacity in the soft tissues of the upper 1/3 of the right leg. The assessment was completed by an MRI which showed a heterogeneous tumor process in hypersignal occupying the upper 1/3 of the right leg (Figure A and B). A biopsy was performed and the diagnosis of Ewing's sarcoma was confirmed. The extension assessment was negative.

Initial multidrug therapy with cyclophosphamide and adriamycin was initiated to decrease the size of the tumor, and two months later the resection of the tumor was done (Figure C and D).

DISCUSSION

Ewing's sarcoma is a rare tumor that mainly affects young adults, and affects both men and women [1]. Extraskelatal Ewing's sarcoma is rarer, since only about 150 cases had been written in 1988. For most authors pain is the most common sign of call [1-3], it is often located at the tumor site but can be projected, and does not give in to the usual analgesics. The swelling exists almost constantly, but the volume is variable. The radiological signs are, at the beginning, minimal and can escape the interpretation; the radiological aspect is very variable and is not always specific. CT scan gives

more complete information of the bone, its environment, as well as the anatomical structures of the neighborhood tissues.

MRI is currently the most effective examination, it replaces or supplements CT [3], it allows better characterization of tumor elements and their environment. The diagnosis of certainty is based on the anatomopathological examination [4], which can be performed either by trocar or by a surgical approach. Macroscopically, the tumor is often whitish, multi-lobulated, infiltrating and destroying all the plans of a region. Microscopically, they are small cells resembling other sarcomas with a mesenchymal-type ultrastructure.

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.

The treatment is based on chemotherapy first, in order to have a volumetric regression of the tumor; this chemotherapy is based on multidrug therapy to potentiate their effects without increasing their toxicity. Since the use of intensive chemotherapy, the 5-year survival is close to 50-60% [5,6].

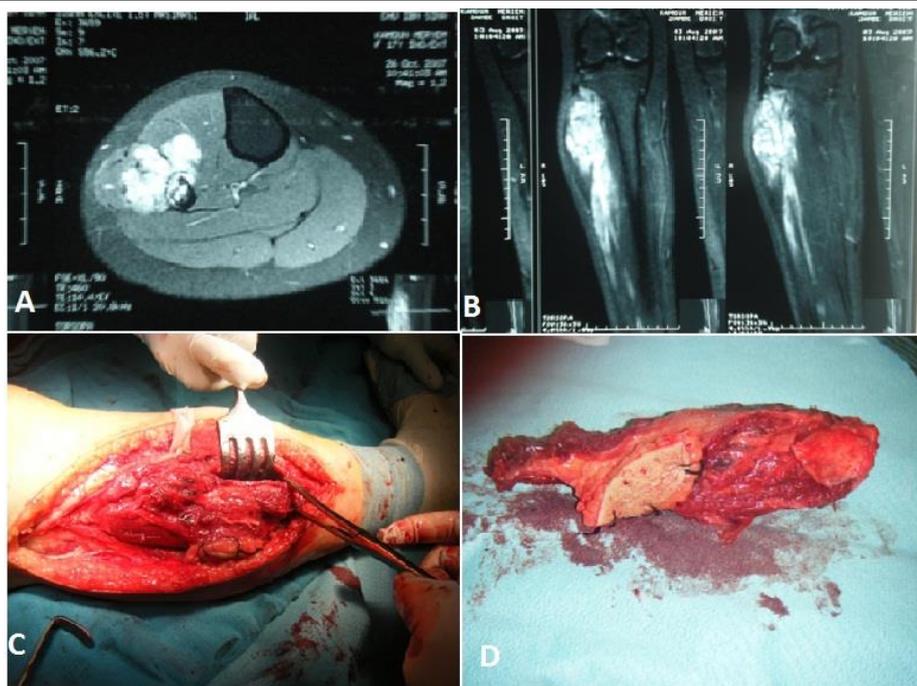


Fig-A: MRI of the right leg: tumor on the external side of the leg. B: MRI of the right leg: tumor on the external side of the leg. C: peroperative appearance of the tumor D: the resected specimen

Surgery is an act both diagnostic and therapeutic, its ultimate goal is to perform a satisfactory oncological intervention passing in any point in healthy tissue, 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. Most of time the prognosis is 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 in terms of anatomical pathology. The therapeutic management is heavy and combines a local treatment based on surgery and/or radiotherapy and chemotherapy. Surveillance is clinical and radiological.

DECLARATION

The authors declare that they have no conflict of interest.

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