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Case Study

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PARATESTICULAR EWING SARCOMA: ABOUT A CASE

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ABSTRACT

Extra-bone Ewing's Sarcoma (EUS) is an exceptional variant of Ewing's sarcoma affecting mainly men in their forties. We report a case of paratesticular extra-bone ewing sarcoma in a 19-year-old boy.

KEYWORDS: Paratesticular, Ewing Sarcoma Tumors, Treatment.

INTRODUCTION

Ewing's sarcoma (SE) is a rare malignant tumor that is most often primitively bony in children or young adults. It consists of round cells, most of them expressing CD99. It is characterized by a translocation of the EWSR1 gene (22; q12) with a gene of the ETS family encoding transcription factors. The extra-osseous variant is exceptional Ewing's sarcoma mainly affecting men in their forties. The management is multidisciplinary conbinant essentially surgery and chemotherapy.

OBSERVATION

Patient of 19 years, without particular pathological antecedents consults for a painful scrotal mass increasing progressively of volume and hindering the march. The clinical examination found a patient in good general condition, apyretic a hard scrotal mass polylobed 10 cm long major inflammatory axis, and inguinal lymphadenopathy.

The ultrasound performed shows a large heterogeneous echogenic mass poorly limited, lobulated 12 * 11 * 10.5 cm pushing testicles without evidence of invasion. The testes have normal size with regular contours without detectable lesions. And a heterogeneous hypoechogenic mass of 4 cm at the right spermatic cord.

MRI shows a hyperintense heterogeneous tissue process in T2 with intrascrotal development of 7.5 * 11.6 * 6.2 cm in contact with the testicles which appear to be repressed without signs of invasion with inguinal lymphadenopathy (figure 1).

Thoracoabdominopelvic tomography showed the presence of a 5.7 mm pulmonary nodule, primary pre-aortic and iliac lymph nodes, bilateral inguinal lymph nodes the largest is 11 mm.

Tumor markers were normal (HCG, AFP, LDH). A biopsy was performed demonstrating a morphological and immunohistochemical aspect in favor of ewing sarcoma.

The patient received a neoadjuvant chemotherapy according to the VAC protocol (vincristine, doxurubicin and cyclophosphamide) with a good clinical response: the tumor volume decreased by almost 8% and the appearance of a bulky ulceronecrotic mass. (Figure 2).

The patient has benefited from inguino-scrotal tumerectomy. Intraoperative exploration exploration finds an invasion of envelopes and cavernous bodies, testis and right spermatic cord.

The histological examination of the piece finds a lobulated tumor proliferation, the lobules are dissociated by hemorrhagic suffusions. The fundus is myxoid. The proliferation is made of round cells with reduced cytoplasm and irregular dense nucleus. The immunohistochemical study shows an intense and membrane expression of CD99.

The evolution was marked by the appearance of pulmonary metastases resulting in the death of the patient in the 6th month.

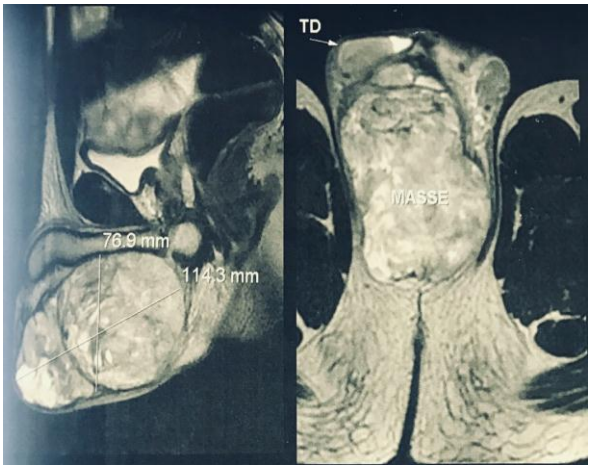

Figure 1: MRI, cross section showing the paratesticular mass.

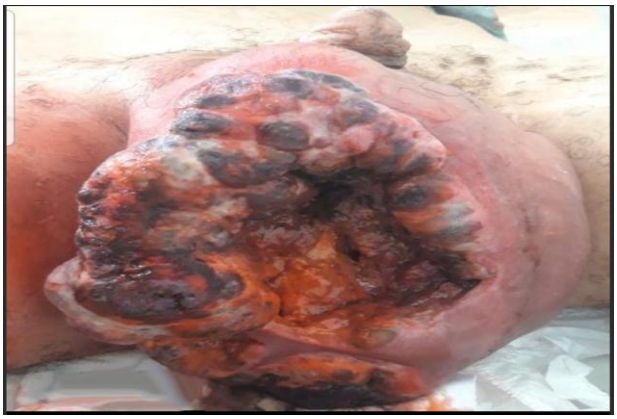

Figure 2: Scrotal mass 22 cm ulcero-necrotic.

DISCUSSION

The primary peripheral neuroectodermal tumors, along with the Askin tumor constitute, with Ewing's sarcoma and peripheral neuroepithelioma, the family of Ewing tumors. These tumors share the same histological, immunohistochemical and cytogenetic definition. This group of primary neuroectodermal tumors is characterized by the almost constant presence of a translocation involving chromosome 22, initially described in Ewing's bone sarcomas.[1,2]

It rarely develops from soft tissue or it will be classified as a separate entity called extra-bone Ewing's sarcoma. He is seen especially during the second decade of life with male predominance and predilection for Caucasian subjects. It mainly affects the soft tissues of the trunk and limbs.[3,4]

It is typically a bulky painful mass associated with multiple satellite nodules with sometimes visceral infiltration. Evolving in a context of cachexia and unexplained fever. It more rarely affects the pleura, ovaries or paratesticular region.[5]

The symptomatology of paratesticular localization is nonspecific and consists of the discovery of a scrotal mass or the appearance of testicular pain.[6] Computed tomography often shows a mass with areas of necrosis and haemorrhage. On MRI the tumor is hypersignal during T1 and T2 sequences.[7]

The histology intervenes to clarify the diagnosis by the presence of a proliferation of small nucleated round cells, with scant cytoplasm. The histological differential diagnosis arises with rhabdomyosarcoma and lymphoma, immunohistochemical examination is a valuable aid to separate Ewing's sarcoma from other small round-cell tumors with positive labeling by antiCD99 antibodies.[8]

The current treatment of these tumors is based on a multimodal treatment combining systemic chemotherapy, surgery and radiotherapy. Current chemotherapy regimens include combinations of vincristine, cyclophosphamide and doxorubicin often combined with ifosfamide and etoposide.[9] Surgical treatment is the essential time of treatment. Radical excision is extra-compartmental excision and has the lowest risk of recurrence.[10] These goals involve collaboration between radiologists, surgeons and anatomopathologists.

The prognosis remains bad because of the micro metastatic spread often present at the time of diagnosis which are mainly pulmonary. Ten-year survival differs from one series to another and depends on the therapeutic protocol. The combined use of surgery and / or chemotherapy and radiotherapy has improved long-term survival from less than 10% to almost 40%.[11]

CONCLUSION

The diagnosis of extra-patent ewing sarcoma is difficult because of its rarity and lack of specificity. It is based on morphological, immunohistochemical and cytogenetic criteria for anapathology. The treatment, although not yet standardized, remains modeled on that of Ewing bone sarcomas. . Multidisciplinary management involving extensive surgery, aggressive chemotherapy and possibly radiotherapy seems to improve survival.

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