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Metastatic paratesticular rhabdomyosarcoma: A case report

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ABSTRACT

Paratesticular rhabdomyosarcoma (PRMS) is a rare and aggressive tumor in children and adolescents.

Clinically, it is often revealed by the accidental discovery of a large painless bursa.

The therapeutic strategy depends on the stage of the tumor and on the prognostic group according to the Intergroup Rhabdomyosarcoma Study (IRS) classification.

We report the case of a patient treated in our institution for a paratesticular rhabdomyosarcoma of the embryonic type discovered at a late stage in an adolescent, with the aim of confirming the fatal evolution of this pathology with "awful" metastatic potential.

1. Introduction

Paratesticular rhabdomyosarcoma is a rare and aggressive tumor. Paratesticular localization is the most frequent of urogenital lesions. Paratesticular rhabdomyosarcoma (PRMS) constitutes 7% of all cases of rhabdomyosarcoma (RMS) in adults[1]

Several forms are described and the embryonic variant is the most frequent. The prognosis is bad. The management is multidisciplinary combining surgery, chemotherapy and radiotherapy.

2. Case report

Mr. MY, 16 years old with no particular pathological history, consults for a right scrotal mass evolving for 6 months with weight loss of more than 10 kg. Clinical examination revealed a cachectic patient with a painless right scrotal mass on palpation that extended towards the course of the spermatic cord, with right pleural effusion syndrome. The examination of the lymph node areas was free.

The ultrasound showed a tissue mass in the right testicle.

The chest x-ray showed a pleurisy of great abundance on the right.

Tumor markers were normal (alpha-fetoprotein, beta-hCG) except for a slight increase in LDH (lacticodehydrogenase).

Thoraco-abdominopelvic computed tomography objectified the presence of a large heterogeneous right intrascrotal mass of irregular contours measuring 63 × 37 mm surrounded by a small scrotal effusion, retroperitoneal lymphadenopathy of 10–15 mm, right pleurisy of great abundance with pulmonary nodules bilateral metastatic (Fig. 1).

A right inguinal orchiectomy was performed. The testis measured 13 cm × 10 cm × 6 cm, and the spermatic cord, 5 cm × 1.5 cm. On gross cut, a greyish tumor occupied the entire testis and invaded the epididymis, spermatic cord and scrotal wall (Fig. 2).

Histological examination of the orchiectomy piece shows an embryonic RMS of the paratesticular region (Fig. 3).

The thoracic surgeons performed a pleural talcage with pleural biopsy at the same time of operation, the pathology of which demonstrated the metastatic origin.

The tumor is classified as stage IV according to the classification established by the Intergroup Rhabdomyosarcoma Study group (IRSG).

The patient died 15 days later, even before starting chemotherapy.

3. Discussion

RMS represent a distinct entity from soft tissue sarcomas, which develop from primitive mesenchymal cells characterized by varying degrees of skeletal muscle differentiation.[2] RMS is found mainly in children, adolescents and, to a lesser extent, in young adults. There are three histological types of RMS: pleomorphic, embryonic and alveolar. The embryonic type (97%), demonstrated in our patient, offers the best prognosis when it is localized in time.[3] In our case, it was observed at an advanced metastatic stage.

In a retrospective multicenter study, older age was shown to correlate with metastatic stage at presentation of embryonic RMS.[4] Older age was also correlated with under pejorative histological type (alveolar). Adulthood is a factor pre - dictif poor prognosis RMS[4]

RMSP can develop from the spermatic cord, epididymis, and testicular tunics, resulting in a painless scrotal mass.[1] The absence of pain

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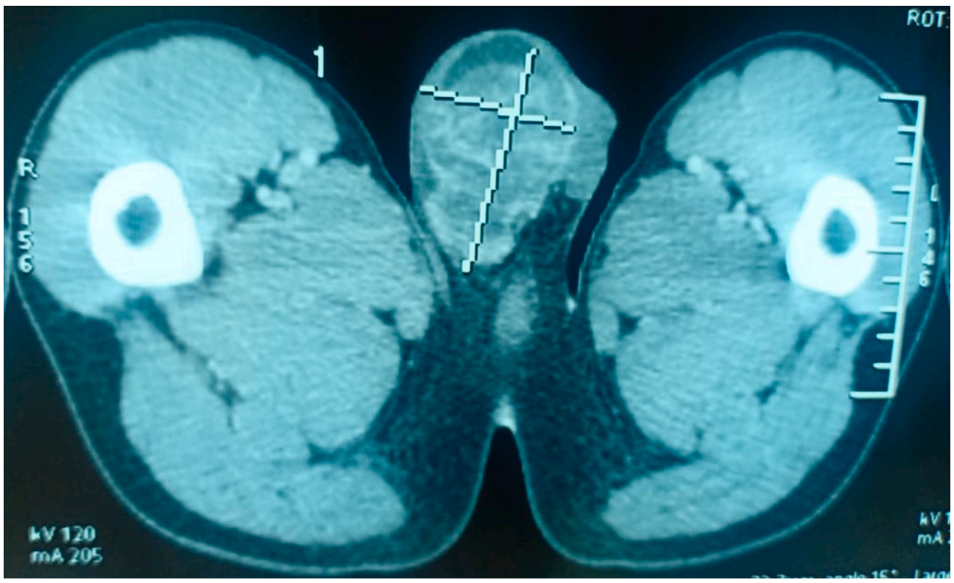

Fig. 1. Abdominal CT scan, cross section showing the paratesticular mass.

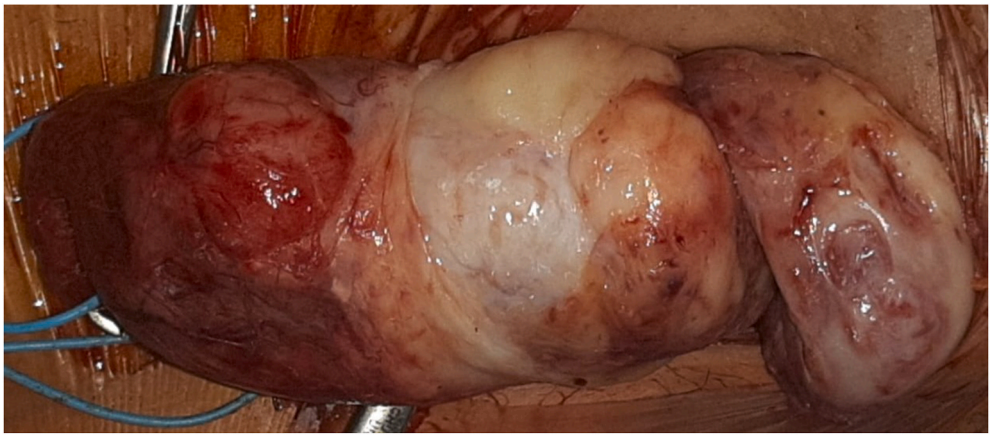

Fig. 2. Orchidectomy operative part.

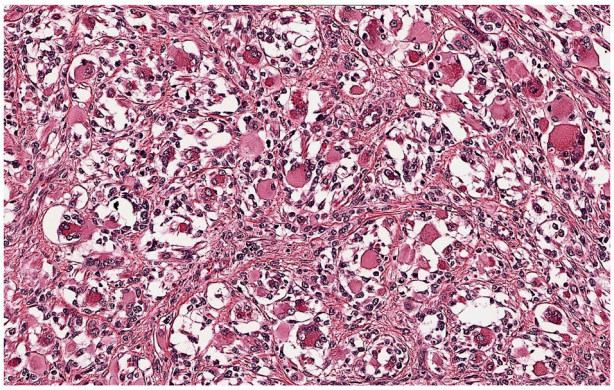

Fig. 3. Microphotography showing a diffuse proliferation made of atypical round cells. A rhabdoid morphology is sometimes observed.

could explain the delay in consultation in the case of our patient. However, the superficial location of the mass would promote early diagnosis and thus the chances of curative surgery. Thus, an ultrasound performed at an early stage would have made it possible to diagnose the scrotal mass in our patient in time. However, the high sensitivity of this examination remains limited by the experience of the radiologist who performs it.[5]

The assay of testicular tumor markers is systematically indicated in the presence of a testicular mass, but these markers are normal in the event of RMS.[3]

Different treatments are available to them (inguinal orchiectomy, radiotherapy, retroperitoneal lymph node dissection, chemotherapy), depending on the stage of the disease according to the post-surgical classification of the IRSG (Intergroup Rhabdomyosarcoma Study Group).[1]

The most widely used adjuvant chemotherapy regimen is VAcdC, which consists of vincristine, actinomycin D, and cyclophosphamide. This is due to the activity of multidrug resistance proteins (*MDR muti-drug resistance*) in tumor cells.[1]

However, RMSs show resistance to multidrug therapy. The prognosis in advanced stages is still grim.[2]

4. Conclusion

Paratesticular RMS is a rare tumor, found especially in children and young adults, but with extremely serious metastatic potential. It requires early diagnosis and a thoraco-abdominal-pelvic extension assessment.

The treatment is currently well codified combining surgery, multidrug therapy and radiotherapy.

Declaration of competing interest

The authors declare no conflict of interest.

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