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PARATESTICULAR RHABDOMYOSARCOMA: WHAT MANAGEMENT?

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SUMMARY

Paratesticular rhabdomyosarcoma is a rare and aggressive mesenchymal tumor. It is an intrascrotal tumor, localized to the paratesticular structures: vaginal, epididymis or spermatic cord. Genito-urinary localizations are among the most frequent. In light of three observations and a review of the literature, the authors discuss the diagnostic and therapeutic modalities.

KEYWORDS: Tumor, rhabdomyosarcoma, management.

INTRODUCTION

Paratesticular rhamdomyosarcomas are malignant tumors developed from the embryonic mesenchyme. It is a rare entity accounting for approximately 7% of all rhabdomyosarcomas.[1] The age distribution is bimodal, with peaks from 1 to 5 years then to 16 years.[2] Multidisciplinary and aggressive management and localized diagnosis can improve the prognosis.

OBSERVATIONS

Case1: 19-year-old patient, with no particular pathological history, consults for a large painless purse evolving over the past year and gradually increasing in volume. Clinical examination finds a hard, polylobed, 10 cm diameter scrotal mass (figure 1). Ultrasound examination on admission shows a large tissue mass, pushing back both testicles. MRI shows a heterogeneous tissue process hyper intense in T2 on contact with the testes repressed without sign of invasion with presence of inguinal lymphadenopathy. The tumor markers were normal. A biopsy was performed objectifying a rhabdomyosarcoma. The patient received three neo adjuvant chemotherapy cures without clinical improvement: volume of the unchangeable mass. He benefited from an inguino-scrotal tumorectomy. Histological examination of the orchidectomy part shows an embryonic RMS with round cells of the par testicular region.

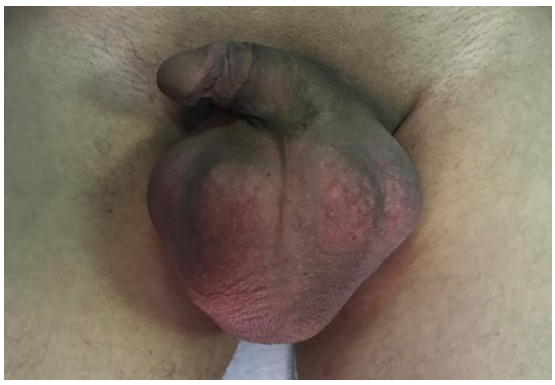

Figure 1: Clinical examination of the scrotal mass.

Case2: the second case is that of a young 20 year old, with no particular history, consults for a large painless mass evolving for about 6 months. Clinically, the scrotum was enlarged, with a mass that appeared to be independent of the testicles, hard in consistency, painless on palpation and without inflammatory signs. Ultrasound at admission shows a large intrascrotal tissue mass of heterogeneous structure, echogenic, with small areas of necrosis. The patient underwent a pelvic MRI which revealed a heterogeneous scrotal tissue mass infiltrating the scrotal skin, without invasion of the testicles (figure 2). The dosage of tumor markers was normal. The patient underwent an inguinal tumorectomy, radio-chemotherapy followed by lumbo-aortic dissection.

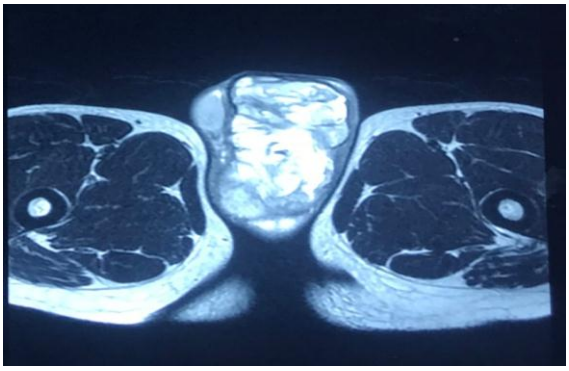

Figure 2: MRI objectifying a 12 cm multi-lobed scrotal mass.

Case3: Mr. A.O., 19 years old, with no particular pathological history, consults for a right scrotal swelling evolving for 8 months. Hormonal dosages were found to be normal. The ultrasound performed shows a 6 cm tissue mass, pushing back the right testicle. The orchiectomy showed that it was a paratesticular embryonic rhamdomyosarcoma. The surgical treatment was followed by 3 sessions of chemotherapy then of lumbo-aortic dissection.

DISCUSSION

Paratesticular rhabdomyosarcoma can occur at any age, especially in children and young adolescents. There are two peaks of incidence between the ages of 1 and 5 and in adolescence,[2] Paratesticular localization represents 7% of rhabdomyosarcomas, all localizations combined.[3] Epidemiologically, there is a male predominance with a sex ratio of 1.4 and differences in incidence according to ethnicity: Caucasian and African American ratio.[4] There are essentially three histological types of rhabdomyosarcoma: the embryonic type which represents 97% of the cases, the alveolar type and the pleomorphic type.[5] Clinically, paratesticular rhabdomyosarcomas have the same clinical symptoms as other tumors with intrascrotal development. It is often a large painless bursa, most often discovered by chance, sometimes with simple scrotal pain.[6] Physical examination often finds a hard mass, fast growing, rarely painful, which is seen on the left as on the right without predominance. There are no tumor markers that can help with the diagnosis, which is based only on the histological examination of the operating room. Ultrasound and especially thoracoabdominal CT, allow us to assess the extension in search of pelvic and lumboaortic lymphadenopathy and hepatic or pulmonary metastases.[7] The analysis of the orchidectomy part and the extension assessment allow the staging of the tumor according to the IRS. Treatment is surgical, combined with chemotherapy and / or radiotherapy. The orchidectomy is performed inguinally with the first and upper ligature of the spermatic cord. The benefit of systematic lymph node dissection has been very controversial since the publication of the SIOP. Para-aortic lymph node dissection does not seem necessary in localized forms. Due to the efficacy of multidrug therapy on micrometastatic lymph node invasion,[8] it is recommended not to perform lymphadenectomy in the absence of lymph node involvement revealed by CT. In this case, polychemotherapy makes it possible to eradicate occult metastases. It is indicated for all prognostic groups with a significant improvement in the probability of overall survival and progression-free survival. Radiation therapy, which is not systematic, is used as a therapeutic supplement on residual focus in advanced forms, on retroperitoneal nodes or on metastases, especially pulmonary. The IRS recommends irradiation of the lumboaortic and iliac chains for stage 2, 3 and 4 tumors at a rate of 40 to 50 Gy spread over five to six weeks.[9]

CONCLUSION

Paratesticular RMS is a rare tumor, it requires an early diagnosis and a precise extension assessment. The improvement of their prognosis depends above all on an early clinical and radiological diagnosis, on a precise assessment of extension and on a well codified treatment based on the combination of chemotherapy, radiotherapy and surgery.

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