

AN 18 YEAR-OLD MAN WITH PARATESTICULAR RHABDOMYOSARCOMA: A CASE REPORT**S. Razine¹, S. Najem¹, S. Harrak¹, S. Lemsanes¹, K. Benchekroun¹, S. Sninate², S. Lkhoyaali¹, S. Boutayeb¹, B. Ghissassi¹, H. Errihani¹**¹Department of Oncology, National Institute of Oncology, Rabat, Morocco.²Department of Radiology, National Institute of Oncology, Rabat, Morocco.***Corresponding Author: S. Razine**

Department of Oncology, National Institute of Oncology, Rabat, Morocco.

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ABSTRACT

Paratesticular rhabdomyosarcoma (RMS) accounts for only 7% of all the RMS cases arising from the mesenchymal tissues of the spermatic cord, epididymis, testis and testicular tunics. We report an 18-year-old man presenting with painless and rapidly growing mass in the scrotum. Radical inguinal orchiectomy was performed. A histological examination of the excised tissue revealed an embryonic rhabdomyosarcoma. In addition, the CT scans showed Intra-abdominal lymph node metastasis and pulmonary metastases. The patient had three sessions of chemotherapy with vincristine, actinomycin C and cyclophosphamide with failure and disease progression. Paratesticular rhabdomyosarcoma (RMS) is a rare nongerm cell intrascrotal malignant tumor in children and young adult, Localized forms have a good prognosis whereas metastatic tumors show very poor results. The treatment of paratesticular rhabdomyosarcoma has evolved over several decades; the current standard of care is multimodal treatment including surgery, chemotherapy, and radiation. Whereas the data showed no upregulation of tumor markers such as b-human chorionic gonadotropin (b-HCG), alpha-fetoprotein (AFP), and lactate dehydrogenase (LDH), scrotal ultrasonography indicated the existence of paratesticular lesion. There was local recurrence in one patient who underwent radical orchiectomy for the sarcoma one year ago The two patients underwent radical inguinal orchiectomy or resection of the recurrent tumors with nerve-sparing retroperitoneal lymph node dissection. Histologic examination revealed embryonal RMS (eRMS) without lymph node metastasis. We highlight the importance of multi-disciplinary participation for paratesticular RMS detection and preoperative ultrasound-guided needle biopsy (UNB) for rapid confirmatory diagnosis.

INTRODUCTION

Paratesticular rhabdomyosarcoma (RMS) is a rare highly malignant mesenchymal neoplasm arising from spermatic cord and epididymis. RSM results from the abnormal proliferation of rhabdomyoblasts, which can grow in any part of the body that contains embryonic mesenchymal tissue. It usually seems unilateral, painless, scrotal swelling mass.^[1]

The first documented case of spermatic cord sarcoma was described by Lesauvage in 1845. Since this date, few cases were reported in the literature especially in adult. The treatment of these cases has evolved over the past decades because of the use of combined modality therapy.^[2]

CASE REPORT

We report a case of an 18-year-old man who presented with a painless right scrotal mass that had evolved over five months.

An ultrasound revealed a huge right epididymis with hydrocele pushing back the right testicle (Figure 1).

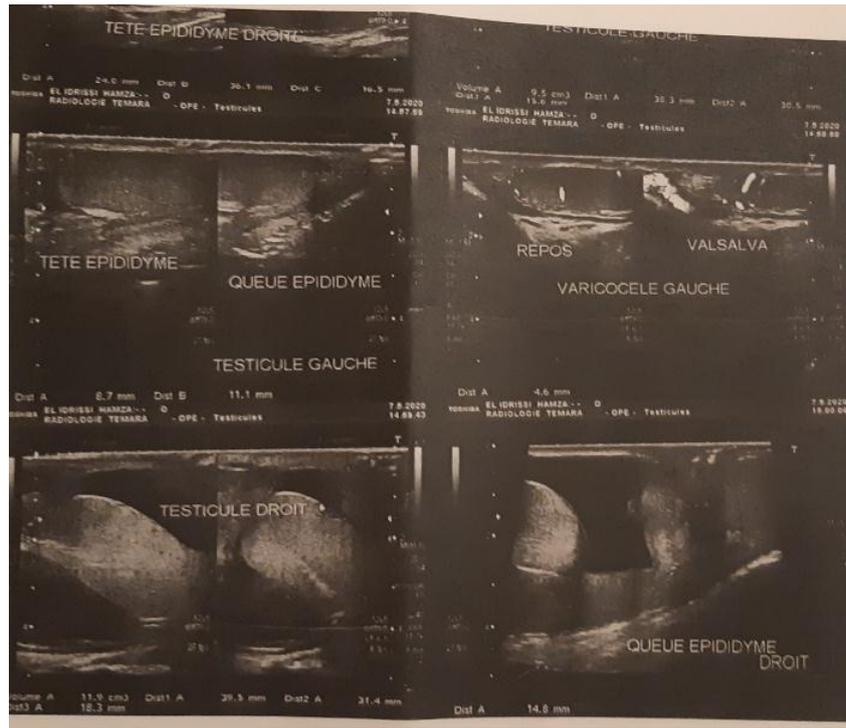


Figure 1: An ultrasound revealed a huge right epididymis with hydrocele.

A computed tomography (CT) scan of his thorax, abdomen and pelvis showed Intra-abdominal lymph node metastasis and pulmonary metastasis.

Whereas the data showed no upregulation of tumor markers such as b-human chorionic gonadotropin (b-HCG), alpha-fetoprotein (AFP), and lactate dehydrogenase (LDH).

Radical inguinal orchiectomy was performed. A histological examination of the surgical specimen demonstrated tumoral proliferation with unorganized architecture in sheets of pleomorphic clear tumoral cytoplasmic cells and eosinophils with atypical nuclei with rhabdomyoblastic aspects were observed, the diagnosis of an embryonic rhabdomyosarcoma was confirmed.

Three chemotherapy sessions of vincristine 1.5 mg/m², actinomycin C 1.5mg/m² and cyclophosphamide 500 mg/m² were performed. Each chemotherapy session was conducted over five days, with a cycle of 21 days.

Failure after 3 cycles with disease progression, the decision was second line of chemotherapy with adriamycin and carboplatin.

DISCUSSION

Rhabdomyosarcoma (RMS) is the most common soft-tissue sarcoma of childhood. Its incidence is similar in Africo-American and Caucasian and appears to be lower in Asian populations. This histologic subtype develops with two peaks, the first at the age of 4 years and the

second at the age of 18 years.^[3] Among all cases of rhabdomyosarcoma, approximately 7% occurs in paratestis. Clinically paratesticular tumour presents as a hard painless inguino-scrotal swelling.^[4] A hydrocele can be occasionally present in adults explaining the frequent mis diagnostic of paratesticular rhabdomyosarcoma with hydrocele in this population.

According to the international RMS classification, the common histological subtypes are alveolar, embryonal, botryoid embryonal, spindle cell embryonal, and anaplastic.^[5] Embryonal RMS(eRMS) is the most frequent type, which accounts for 60% of the cases.

Thus, since it's uncommon in adults, few studies have reported paratesticular eRMS.^[6,7] Pathologically, the eRMS presents the existence of poorly differentiated cells and rhabdomyoblasts, with abundant eosinophilic cytoplasm indicating embryonal rhabdomyosarcoma. Cytogenetically, the eRMS is characterized by loss of heterozygosity on the short arm of chromosome 11.^[8,9] Whereas, electron microscopy and chromosome analysis are useful methods to improve the pathological diagnosis of the eRMS. They were not examined in our cases.

A testicular ultrasound is routinely performed for a scrotal mass. This imaging modality shows a mass with heterogeneous echogenicity and inguinoscrotal extension in 80% of cases.^[10] This allows the nature of the intrascrotal tissue mass to be determined and specifies the exact topography. Ultrasound is used to establish the differential diagnosis and eliminate diagnoses of simple cyst and varicocele.

Whereas both CT scan and MRI can accurately evaluate the location, size and metastasis of the mass, they cannot be used as confirmatory diagnostic tools. Differential diagnosis of paratesticular RMS includes lesions such as leiomyosarcoma, liposarcoma and fibrosarcoma. These tumors lack imaging features, thus confirmatory diagnosis relies on postoperative pathology.^[11-12]

In rhabdomyosarcoma, tumoral markers including alpha-fetoprotein, beta-human chorionic gonadotropin and carcinoembryonic antigen are usually normal. This was the case with our patient.

Radical orchidectomy by the inguinal route with first cord ligation remains the essential act for histological diagnosis and constitutes the first step of treatment regardless of the stage of the disease. Hemiscrotectomy associating inguinal treatment is indicated first in scrotal cases whenever local invasion or presence of lymph are clinically evidenced.^[13,14]

Retroperitoneal lymph node (RPLN) is found in approximately 25% of patients at presentation.^[1] The most appropriate supplementary therapy seems chemotherapy after orchidectomy without positive RPLN. If the RPLN is negative, additional therapy must be limited. Patient race, histology, and tumor laterality are not significant predictors of nodal metastasis.^[15] For further staging and treatment, highly selective RPLN dissection (RPLND) is needed after chemotherapy if the positive nodes are present. Radiotherapy is not recommended for patients with completely resected localized disease and is only recommended to the patients with locally advanced disease or nodal involvement on RPLND.^[16]

An alternative approach for patients with clinically enlarged retroperitoneal nodes is the administration of an adjuvant chemotherapy regimen (VAC or vincristine plus dactinomycin and ifosfamide (VAI)). The development of this adjuvant therapy has increased survival in patients with localized disease to approximately 60%.^[17] In the metastatic setting, many protocols of chemotherapy have been tried. VAC, IVA, and VIE protocols (V: vincristine, A: actinomycin, I: ifosfamide, E: etoposide, and C: cyclophosphamide) and better results were observed with VAC protocol.^[18-19] Radiotherapy is a complementary treatment of chemotherapy and surgery to eliminate residual foci and retroperitoneal lymph nodes.

Our patient benefited from an inguinal orchidectomy. Three sessions of chemotherapy were performed with failure.

CONCLUSION

Paratesticular RMS is rare and malign neoplasm in young adults with multimodality approach in diagnosis

as well as treatment. Preoperative ultrasound-guided needle biopsy is an available option for rapid diagnosis.

Extensive and thorough surgical excision is key to the treatment. Radiotherapy or retroperitoneal lymph node dissection used based on the tumor clinical stage. The adult chemotherapy regimen can refer to pediatric VAC regimen.

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