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Case Report

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EMBRYONAL PARATESTICULAR RHABDOMYOSARCOMA: CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Paratesticular rhabdomyosarcoma (RMS) is a rare type of RMS that represents only 7% of all RMS cases originating from the mesenchymal tissues of the spermatic cord, epididymis, testis, and testicular tunics. We report an 17-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 an uncommon malignant tumor in children and young adults that arises in the scrotal area and is not of germ cell origin. While localized forms of the disease have a favorable prognosis, metastatic tumors exhibit a very poor outcome. The current standard of care for treating paratesticular RMS involves a combination of surgery, chemotherapy, and radiation therapy. Even though tumor markers such as b-human chorionic gonadotropin (b-HCG), alpha-fetoprotein (AFP), and lactate dehydrogenase (LDH) were not elevated, scrotal ultrasonography indicated the presence of a paratesticular lesion. In one case, a patient who underwent radical orchiectomy for paratesticular sarcoma experienced local recurrence one year later. Both patients were treated with either radical inguinal orchiectomy or resection of recurrent tumors along with nerve-sparing retroperitoneal lymph node dissection. Histologic examination showed that both patients had embryonal RMS (eRMS) without lymph node metastasis. We emphasize the importance of involving a multidisciplinary team in the detection of paratesticular RMS and the use of preoperative ultrasound-guided needle biopsy (UNB) for rapid and accurate diagnosis.

INTRODUCTION

Paratesticular rhabdomyosarcoma (RMS) is a rare and highly malignant tumor that develops from the mesenchymal tissue in the spermatic cord and epididymis. It results from abnormal proliferation of rhabdomyoblasts that can occur in any part of the body with embryonic mesenchymal tissue.[1]

Typically, patients present with painless scrotal swelling and the embryonal RMS histological subtype is most common and associated with a favorable prognosis.[2]

The first reported case of sarcoma in the spermatic cord dates back to 1845 when Lesauvage described it. Since then, there have been only a few cases reported in the literature, especially in adults. Treatment for these cases has evolved over the decades with the use of combined modalities.[3]

We report the clinical case of a child with an embryonal paratesticular rhabdomyosarcoma and we will proceed with a literature review.

CASE REPORT

We report a case of an 17-year-old man who presented with a painless right scrotal mass that had evolved over four months. An ultrasound revealed a huge right epididymis with hydrocele pushing back the right testicle (Figure 1).

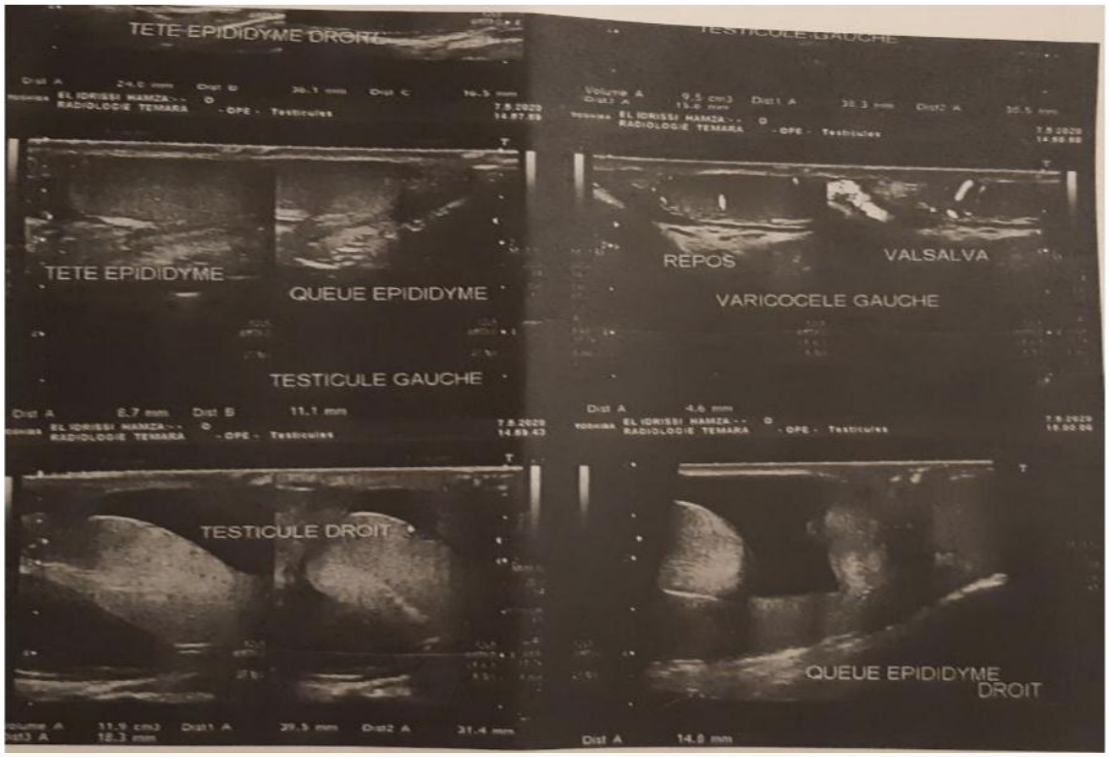

Figure 1: A scrotal ultrasound revealed a huge right epididymis with hydrocele.

The patient underwent a computed tomography (CT) scan of his thorax, abdomen, and pelvis which revealed the presence of intra-abdominal lymph node metastasis and pulmonary metastasis. Interestingly, the results of the tumor marker tests, including b-human chorionic gonadotropin (b- HCG), alpha-fetoprotein (AFP), and lactate dehydrogenase (LDH), did not show any upregulation. The patient then underwent radical inguinal orchiectomy, and a histological examination of the surgical specimen confirmed the diagnosis of an embryonic rhabdomyosarcoma, characterized by tumoral proliferation with unorganized architecture in sheets of pleomorphic clear tumoral cytoplasmic cells and eosinophils with atypical nuclei with rhabdomyoblastic aspects.

The patient received three chemotherapy sessions with a combination of vincristine, actinomycin C, and cyclophosphamide. Each chemotherapy session was conducted over five days, with a cycle of 21 days. However, after three cycles, the patient showed disease progression, and the treatment was deemed unsuccessful.

As a second-line treatment, the patient was started on a combination of adriamycin and carboplatin chemotherapy. The response to this treatment is yet to be determined.

DISCUSSION

Paratesticular rhabdomyosarcoma is a rare tumor. The embryonal subtype represents about 90% of paratesticular rhabdomyosarcomas.[4,5] The age of onset is characterized by two peaks of incidence, the first between 2 and 5 years old, and the second during adolescence.[5]

The clinical discovery of paratesticular rhabdomyosarcoma is often accidental, revealing an indolent intrascrotal mass.[6,4] In cases of acute scrotum, diagnosis can be difficult due to the presentation often being confused with testicular torsion.[6] Testicular ultrasound is performed as a first-line investigation.[6,8] It allows visualization of the intrascrotal mass, developed from the testicular envelopes. The testicle is usually intact.[6,7]

The distant spread occurs via lymphatic and hematogenous routes. Retroperitoneal lymph nodes are the first nodal relays.[7] Involvement of the lumbar-aortic lymph nodes is reported in 26% to 43% of cases. Lung, liver, and bone are the most frequent metastatic sites.[6]

The liver and bone are the most common sites of metastasis.[6] Thoraco-abdomino-pelvic CT scan helps to assess the distant extension of the disease. It allows exploration of the pelvic and lombo-aortic lymph node areas, as well as the search for liver and/or lung metastases.[6,9,10] Bone scintigraphy searches for secondary bone locations.[4]

MRI is useful in cases of significant locoregional extension, allowing for a better assessment of the tumor's relationship with pelvic organs.[8] FDG PET-CT (fluorodeoxyglucose positron emission tomography-computed tomography) is used in the staging of adult sarcomas, but its use is very limited in the pediatric population.[4]

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

The therapeutic strategy must be multidisciplinary and depends on the stage of the disease. Several classifications have been proposed to identify prognostic factors of the disease, the most commonly used being the IRS IV and the SIOP (International Society of Paediatric Oncology). Prognostic groups have been identified based on the quality of surgical resection and lymph node involvement. Treatment depends on the prognostic group, stage of the disease, and age at diagnosis. The paratesticular site is considered a site of favorable prognosis.[8]

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.[11,12]

Approximately 25% of patients with testicular cancer have retroperitoneal lymph node (RPLN) involvement at the time of diagnosis.[1]

Chemotherapy after orchidectomy is recommended as the most appropriate supplementary therapy if Retroperitoneal lymph node involvement is not detected. Additional therapy should be limited if RPLN involvement is not found. Patient factors such as race, histology, and tumor laterality are not reliable predictors of nodal metastasis.[13]

For further staging and treatment, a highly selective Retroperitoneal lymph node dissection (RPLND) is necessary after chemotherapy if positive nodes are present. Radiotherapy is not advised for patients with completely resected localized disease and is only recommended for those with locally advanced disease or nodal involvement after RPLND.[5]

For patients with retroperitoneal nodes that are clinically enlarged, an alternative approach is to use an adjuvant chemotherapy regimen such as VAC (vincristine, dactinomycin, and ifosfamide) or VAI (vincristine, actinomycin, and ifosfamide). This treatment has increased survival rates to around 60% for patients with localized disease.[14]

In the case of metastatic disease, various chemotherapy protocols have been tested, such as VAC, IVA, and VIE, but the VAC protocol has shown better results.[12-15]

Radiotherapy can also be used in conjunction with surgery and chemotherapy to eliminate any residual foci and retroperitoneal lymph nodes.

Our patient underwent an inguinal orchidectomy, followed by three sessions of chemotherapy that were ultimately unsuccessful.

CONCLUISON

Testicular rhabdomyosarcoma is a therapeutic and diagnostic emergency, and its management requires a multidisciplinary approach. Localized forms have a favorable prognosis, and a multimodal treatment approach can lead to excellent survival rates. Several therapeutic pathways are currently being explored, particularly targeted therapies, considering the specific molecular profile of rhabdomyosarcomas.

REFERENCE

1. Dangle PP, Correa A, Tennyson L, Gayed B, Reyes-Múgica M, Ost M.Current management of paratesticular rhabdomyosarcoma. Urol Oncol, 2016; 34: 84-92.
2. Ferrari A, Bisogno G, Casanova M, Meazza C, Piva L, Cecchetto G, et al. Paratesticular rhabdomyosarcoma: Report from the Italian and German Cooperative Group. *J Clin Oncol*, 2002; 20: 449–55. [PubMed] [Google Scholar]
3. Fagundes MA, Zietman AL, Althausen AF, Coen JJ, and Shipley WU. "The management of spermatic cord sarcoma". Cancer, 1996; 77(9): 1873–1876. PubMed | Google Scholar
4. Dasgupta R, Rodeberg DA. Update on rhabdomyosarcoma. Semin Pediatr Surg, 2012; 21: 68–78.
5. WuHY, SnyderHM, 3rd, WomerRB. Genitourinary rhabdomyosarcoma: which treatment, how much, and when? J Pediatr Urol, 2009; 5: 501–6.
6. Faure A, Diakité ML, Panait N, Chaumoître K, Rome A, Merrot T. Le rhabdo- myosarcome para-testiculaire de l'enfant : une urgence scrotale. Arch Pediatr, 2012; 19: 1340–4.
7. Reeves HM, MacLennan GT. Paratesticular rhabdomyosarcoma. J Urol, 2009; 182: 1578–9.
8. Sommelet D, Pinkerton R, Brunat-Mentigny M, Farsi F, Martel I, Philip T, et al. Standards, options and recommendations (SOR) for clinical care of rhabdo- myosarcoma (RMS) and other soft tissue sarcoma in children. Federation of the French cancer centers. French Society of Pediatric Oncology. Bull Cancer, 1998; 85: 1015–42. Zheng L, Tang H, Chen X, Yang H, Yang M. Paratesticular fetal-type rhabdo- myoma in a 12-year-old boy: a case report and literature review. Urology, 2013; 82: 1150–2.
9. Rodeberg D, Paidas C. Childhood rhabdomyosarcoma. Semin Pediatr Surg, 2006; 15: 57–66.
10. Kasmaoui E, Jira H, Alami M, Ghadouane M, Ameur A, Abbar M: Les rhabdomyosarcomes paratesticulaires: à propos de trois cas. Ann Urol, 2001; 35: 296–300.

11. Kourda N, El Atatb R, Derouicheb A, Bettaiba I, Baltagia S, Zermania R: Rhabdomyosarcome pléomorphe paratesticulaire de l'adulte: diagnostic et prise en charge. Cancer/Radiothérapie, 2007; 11: 280–283.
12. Alanee S, Holland B, Dynda D, Kamel O, Ganai S. Primary tumor size predicts pathologic findings in the retroperitoneal lymph nodes in patients with paratesticular rhabdomyosarcoma. Virchows Arch, 2014; 465: 697-701.
13. Wiener ES, Anderson JR, Ojimba JI, Lobe TE, Paidas C, Andrassy RJ, et al. Controversies in the management of paratesticular rhabdomyosarcoma: is staging retroperitoneal lymph node dissection necessary for adolescents with resected paratesticular rhabdomyosarcoma. Semin Pediatr Surg, 2001; 10(3): 146. PubMed | Google Scholar
14. Tazi K, Moudouni S, Koutani A, Ibn Attya, Hachimi M, and Lakrissa A. "Rhabdomyosarcome paratesticulaire de l'adulte jeune". Progrès en Urologie, 2000; 10(3): 469–472. PubMed |Google Scholar.