Clinical Case Report

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Alveolar paratesticular rhabdomyosarcoma mimicing epididymitis

Case report and literature review

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Abstract

Rationale: Most patients with paratesticular rhabdomyosarcoma may typically present as a unilateral, painless palpable scrotum mass. However, only a few cases of RMS presenting as painful edema of the scrotum mimicing epididymitis. We herein report an unusual case of alveolar paratesticular rhabdomyosarcoma misdiagnosed as epididymitis.

Patient concerns: A 19-year-old adolescent, presented to urologist with painful swelling of the scrotum on the left side over the preceding several days. Antibiotics were administered by physician for two months and the pain improved, but the swelling did not fade.

Diagnoses: Alveolar praratesticular rhabdomyosarcoma.

Interventions: A left, soft tissue mass in the scrotum without definite metastasis or lymphadenopathy was confirmed by computed tomography(CT) and magnetic resonance imaging. A radical left orchiectomy via the inguinal approach was performed successfully.

Outcome: The patient received 8 cycles of adjuvant chemotherapy, the patient remains recurrence- and metastasis-free at 13 months after surgery.

Lessons: When paratesticular RMS is presenting with symptoms of epididymitis, this malignant tumor is usually overlooked. When patients complain of painful scrotal swelling, RMS arise from paratesticular tissue should be considered.

Abbreviations: CT = computed tomography, MRI = magnetic resonance imaging, PET/CT = positron emission tomography-computed tomography, RMS = rhabdomyosarcoma.

Keywords: alveolar paratesticular rhabdomyosarcoma, epididymitis, rhabdomyosarcoma

1. Introduction

Rhabdomyosarcoma (RMS) is the most common pediatric sarcoma, while it is relatively rare in young adults.[1] RMS is responsible for about 6.5% of malignancy in patients aged <15 years.[2] Paratesticular RMS arises from the epididymis, testicular envelopes, spermatic cord, and only represents 7–10% of genitourinary RMS tumors.[3] Paratesticular RMS usually presents as painless and rapidly growing masses in the scrotum or inguinal canal. However, to our knowledge, only a few previous cases had involved symptoms of epididymitis.[4,5] We herein report a rare case of alveolar paratesticular RMS present with painful scrotal swelling symptoms misdiagnosed as epididymitis.

2. Case report

A 19-year-old young man with no significant previous medical history, presented to urologist and complained that he had pain and left scrotal swelling over the preceding several days. Physical examination found an irregular, enlarged, mild tender mass in the cauda epididymis, with warm scrotal skin, and a diameter of $4 \times 3 \times 1.5\,cm^3$. The bilateral inguinal lymph nodes were unpalpable. No significant findings were detected on the other physical examinations. A scrotal ultrasonography revealed an inhomogeneous echoes swelling of the left epididymis ($32 \times 21\,mm^2$) with abundant flow of color Doppler signals, suspicious for epididymitis. Therefore, antibiotics were administered by physician for 2 months and the pain improved, but the swelling did not fade.

Then, the patient presented to the Department of Urology of the first hospital of Quanzhou affiliated to Fujian Medical University with scrotal swelling. He was hospitalized for further

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Informed patient consent was obtained for publication of this case report.

The committee of human research at Lishui Central Hospital approved the study. The authors have no conflicts of interest to disclose.

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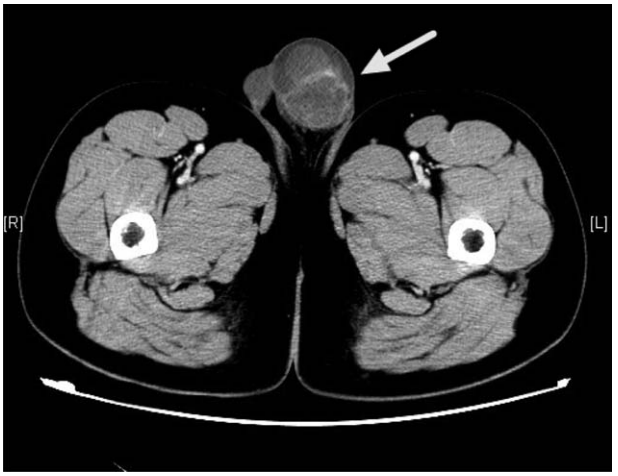

Figure 1. The contrast enhanced computed tomography scan revealed an epididymal mass co-exist with hydroceles in the scrotum (arrow).

evaluation. Complete blood cell count showed mild leukocytosis. The tumor markers α- fetoprotein, lactate dehydrogenase, and β-human chorionic gonadotropin were normal. The other laboratory evaluations such as liver function, renal function, and coagulation function were also within the normal range. The second color Doppler reexamination showed the size of left extratesticular masses increased to 40×34 mm and was suspected of epididymal malignant tumor. The contrast enhanced computed tomography (CT) scan and magnetic resonance imaging (MRI) also revealed an epididymal mass co-exist with hydroceles in the scrotum (Figs. 1 and 2). A left, soft tissue mass in the scrotum without definite metastasis or lymphadenopathy was confirmed by CT and MRI. A radical left orchiectomy via the inguinal approach was performed successfully. During the operation, an irregular, hard tumor could see on the cauda epididymis (Fig. 3). Malignancy of the epididymis was identified by an intraoperative frozen-section biopsy. Histologic examination revealed deep staining small round cells with atypical nuclei

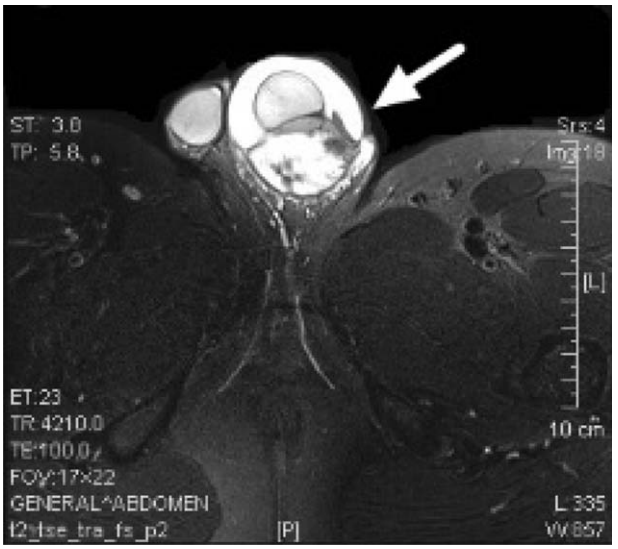

Figure 2. Magnetic resonance imaging revealed an epididymal mass co-exist with hydroceles in the scrotum.

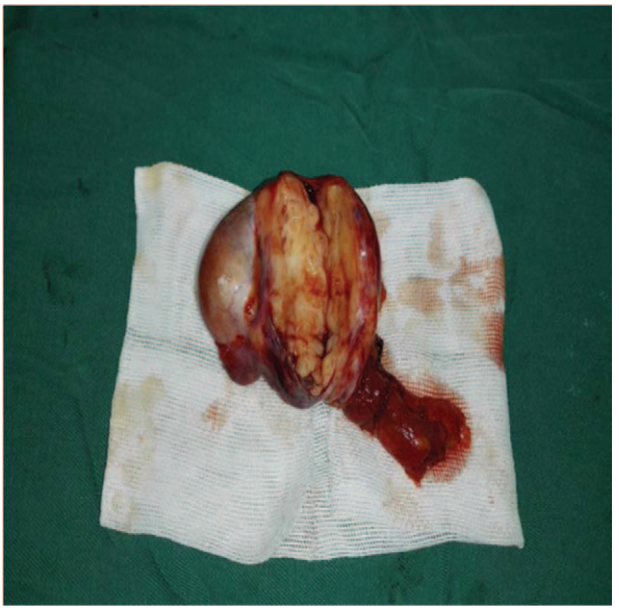

Figure 3. Grossly, the tumor is white-gray in color. The specimen is $4 \times 4 \times 3$ cm^3 in size.

and formed alveolar structures (Fig. 4). The testis white film had been invaded, while testicular parenchyma was not infringed and the surgical margin was negative. On immunohistochemical examination, the tumor cells were diffusely and strongly positive for MyoD1 and myogenin (Fig. 5). We conclusively diagnosed alveolar paratesticular RMS (pT2N0M0).[6]

On the postoperative period, the patient received 8 cycles of adjuvant chemotherapy (VAC regimen: vincristine 1.5 mg/m^2, actinomycin 1.5 mg/m^2, and cyclophosphamide 500 mg/m^2) in the Cancer Hospital of Fudan University. Over a 13-month follow-up period, there was no evidence of recurrence detected by positron emission tomography-computed tomography (PET/CT).

3. Discussion

The RMS is a malignant soft-tissue tumor originated from striated muscle cells or mesenchymal cells differentiated from striated muscle cells. It is the most common soft-tissue sarcoma in children. According to the international classification of RMS, the most common histologic types of RMS are botryoid embryonal, embryonal, spindle cell embryonal, anaplastic, and alveolar.[7] The alveolar RMS can arise from the extremities, head and neck, genitourinary tract, retroperitoneum, and orbit.[8] The alveolar histotype occurs less frequently than embryonal RMS and comprises about 20% of all pediatric RMS.[9] However, paratesticular RMS is relatively rare and accounts for about 7% of genitourinary RMS.[9] Our case had an alveolar paratesticular RMS, which is very rare, as only few case of alveolar paratesticular RMS had been published.[10]

The typical clinical manifestation for paratesticular RMS is a painless epididymal mass, or nonspecific symptoms, such as decreased appetite, fatigue, inguinal lymphadenopathy, and weight loss. A paratesticular RMS can cause pain when it oppresses the nerve. However, pain is extremely uncommon and present in only 7% of the cases, whereas a hydrocele may be also a rare presentation.[11–13] When paratesticular RMS present with

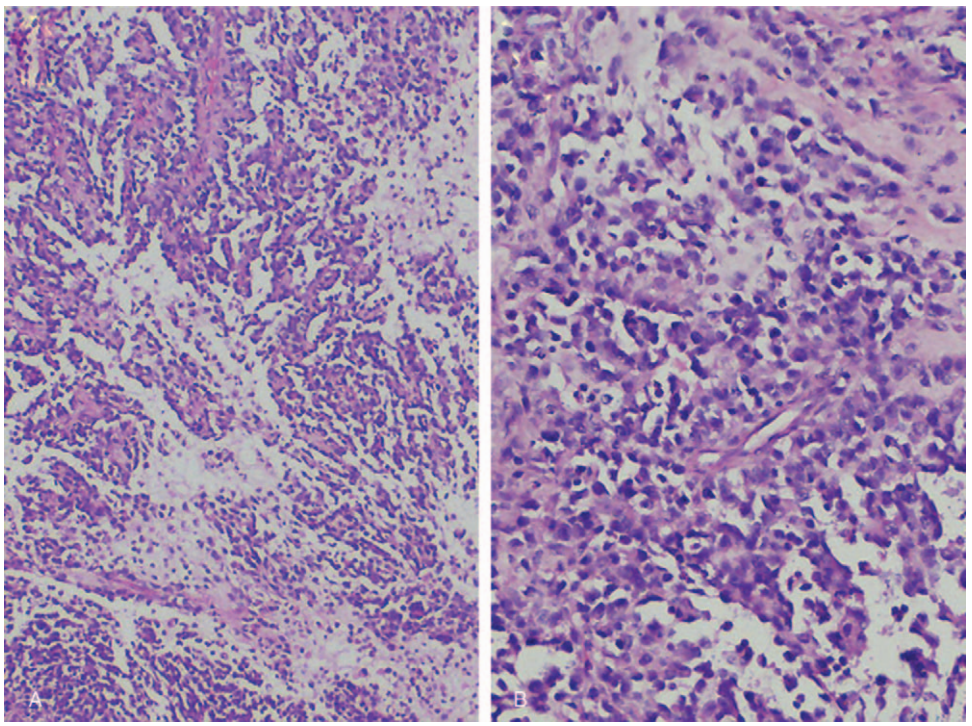

Figure 4. Hematoxylin and eosin staining of rhabdomyosarcoma. (A) Deep staining small round cells with atypical nuclei and formed alveolar structures (magnification, ×100). (B) Showed undifferentiated primitive mesenchymal cells and early differentiated rhabdomyocytes (magnification, ×200).

painful unilateral scrotal swelling symptoms, it often leads to a misdiagnosis of epididymitis. As in our case, the patient first presented with painful scrotum and warm scrotal skin symptom. Moreover, antibiotic treatment could relieve the scrotal pain. As a result, the diagnosis of paratesticular RMS was delayed. The gradual enlargement of the unilateral scrotum, which could not relieved by antibiotics, made us to suspect paratesticular malignancy. Therefore, the paratesticular RMS mimicing symptom of epididymitis leads to the delayed diagnosis in our patient.

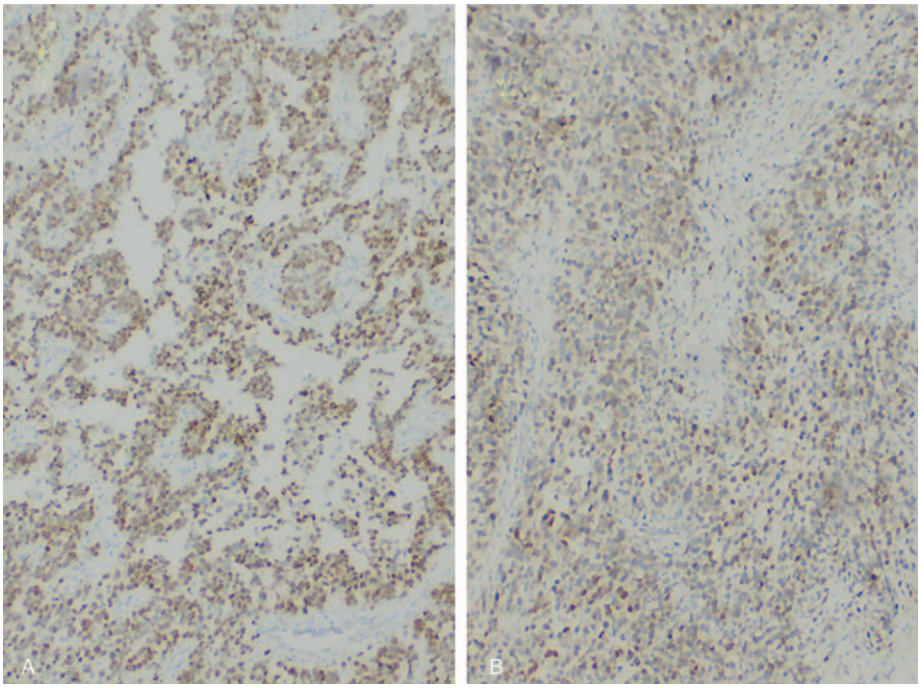

Figure 5. (A) Immunostaining revealed positive result of MyoD1 in the tumor cells (magnification, ×100). (B) Immunostaining revealed nuclear positivity for myogenin (magnification, ×100).

Scrotal color Doppler ultrasonography is the initial imaging modality for evaluating intrascrotal pathology, but color Doppler appearances of paratesticular RMS could lead to confusion with epididymitis, adenomatoid tumor, and leiomyoma.[14,15] In 1995, Wood and Dewbury[14] first reported the Doppler ultrasound features of paratesticular RMS in a 17-year-old boy. They described a highly reflective soft-tissue epididymal mass, with increased vascularity on color Doppler, which they first considered to be epididymitis. It was not diagnosed as paratesticular RMS until 6 months, after which progressive swelling of the scrotum occurred. In 2004, Mak et al[15] also reported paratesticular RMS in a 14-year-old boy could not distinguish from epididymitis at ultrasound. It illustrated that it may be difficult to differentiate RMS from epididymitis by color Doppler appearances. In the present case, our color Doppler ultrasonography also could not provide enough information for the primary lesion. In our opinion, further evaluation is very important in patients presenting with epididymitis symptom, especially when patients not respond well with antibiotic therapy.

The CT scan, MRI, or PET/CT is regularly used to evaluate distant metastases or make the diagnosis of alveolar RMS.[16,17] Both CT and MRI can be used to assess the site, dimensions, and any distant metastases of the tumor.[18] PET/CT can give accurate information about distant metastases of the malignancy. However, none of them is a confirmatory method. The definitive diagnosis of alveolar RMS requires histopathologic examination. The histologic appearance of alveolar RMS was characterized by aggregates of small, round tumor cells, separated by fibrous septa. Microscopically, alveolar RMS is highly cellular, composed of primitive cells with monomorphous round nuclei and formed alveolar structures. Immunostaining for myogenin and MyoD1 is essential for the differential diagnosis of RMS. Markers such as myoD1 and myoglobin can be used to determine alveolar RMS from other RMS tumors. Our immunostaining revealed positive result of MyoD1 and myogenin in the tumor cells, suggestive of malignant mesenchymal tumor. Electron microscopy and chromosome analysis may be helpful methods for improved pathologic diagnosis of alveolar RMS.[19]

According to the Intergroup Rhabdomyosarcoma Study Group (IRSG), surgicopathologic staging of RMS is predictive of outcome,[6] which also could guide the treatment. Some data indicate that staining for myogenin correlate with decreased survival.[20] Literature also reported that anatomic site was also a significant prognostic indicator.[21] Currently, primary paratesticular RMS is generally have a better prognosis and a higher survival rate in comparison with other RMSs.[22] In our case, the patient received 8 cycles of adjuvant chemotherapy, the patient remains recurrence and metastasis free at 13 months after surgery.

Finally, the present case serves to highlight one point worthy of notice: urologist should be aware of the possibility that scrotal pain and swelling could be a rare presentation of alveolar paratesticular RMS. As highly aggressive nature of alveolar paratesticular RMS, further evaluations is important in patients with symptoms of epididymitis, especially who not respond well to antibiotic therapy.

Author contributions

JL, JX, and W-HL contributed to the conception and design of the study. JX, W-FW, J-YZ, and Y-HL conducted the work and collected the data. W-HL drafted the manuscript. All authors approved the final version to be published.

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