Radio-histopathological Presentation and Multidisciplinary Treatment of Adolescent Paratesticular Rhabdomyosarcoma: A Case Report

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Abstract
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Keywords
Rhabdomyosarcoma, Paratesticular mass, Pediatric tumor, Adolescent tumor

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Conflict of Interest Statement
The authors state there are no conflicts of interest.

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CASE REPORT

Radio-histopathological Presentation and Multidisciplinary Treatment of Adolescent Paratesticular Rhabdomyosarcoma: A Case Report

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Abstract

Paratesticular rhabdomyosarcoma (PRMS) is a rare condition predominantly affecting the pediatric and adolescent population. In this case report, we discuss a 17-year-old male with a slow-growing, painless scrotal mass, ultimately diagnosed as embryonal PRMS — the most common and favorable rhabdomyosarcoma subtype. The report underscores the typical clinico-radiological presentation of PRMS and the pivotal part of histopathological evaluation in establishing a definitive diagnosis. Multidisciplinary intervention including surgery and chemoradiotherapy resulted in total remission. Therefore, this report underlines the significance of prompt diagnosis and comprehensive management in ensuring a favorable prognosis of PRMS.

Keywords: Rhabdomyosarcoma, Paratesticular mass, Pediatric tumor, Adolescent tumor

1. Introduction

Rhabdomyosarcoma (RMS) is a rare yet aggressive tumor. Paratesticular rhabdomyosarcoma (PRMS) is the third most prevalent variant of urogenital RMS, accounting for approximately 7% of cases, following those of the prostate and bladder. Among the three histological subtypes of RMS, the embryonal variant is the most prevalent. PRMS generally has a favorable prognosis, attributable to its superficial location, which enables complete surgical resection if diagnosed promptly. However, delay in diagnosis has been associated with increased mortality and morbidity rates. Therefore, early diagnosis and subsequent management are crucial for achieving positive outcomes. In this report, we detail the case of an adolescent diagnosed with PRMS, who underwent a successful multidisciplinary treatment regimen involving surgery, chemotherapy, and radiotherapy, leading to complete remission. This case underscores the significance of timely intervention and a comprehensive approach to managing this tumor.

2. Case presentation

A 17-year-old Caucasian male presented with a small, painless, left-sided scrotal mass that he noticed four months ago. The mass showed minimal change in size over this period. The patient had no significant medical or family history and denied experiencing urinary symptoms, fever, night sweats, weight loss, or breathing difficulties.

The initial ultrasound (US) with Doppler of the left scrotum revealed a 1.7-cm solid paratesticular ovoid solid mass superior and slightly medial to the left testicle and adjacent to the left epididymis. The mass had heterogenous echotexture and demonstrated prominent internal vascularity on Doppler imaging (Fig. 1A and B). No intratesticular mass was observed. Tumor markers including AFP, HCG, and LDH were within normal limits.
The patient underwent trans-scrotal excision of the paratesticular mass, and pathological examination revealed a high-grade malignant neoplasm composed of small round blue cells in a myxoid background along with broad fibrous bands (Fig. 2A). Some tumor cells exhibited abundant eosinophilic cytoplasm and eccentrically placed nuclei, indicating rhabdomyoblasts, characteristic of RMS (Fig. 2B). The tumor was immunohistochemically positive for myogenin, Myo-D1, and CT-1, and focally positive for desmin, MSA, and CD-10. However, it tested negative for SMA, AE1/3, CAM 5.2, LCA, CD30, synaptophysin, chromogranin, and PLAP. The final diagnosis was embryonal paratesticular rhabdomyosarcoma, measuring 2.5 cm and extending to the tissue edges, classified as Intergroup Rhabdomyosarcoma Study Group IIA (IRSG-IIA). Because the initial surgery did not include regional lymph node sampling, lymph node involvement was not factored into the IRSG staging.

Postsurgical CT and PET scans with contrast showed the recently resected left paratesticular
mass, with soft tissue thickening along the left scrotum and a small amount of FDG activity (SUV 4), possibly representing postsurgical changes or residual tumor. Additionally, borderline prominent left external iliac chain lymph nodes were observed.

Due to the possible residual tumor and regional lymph node involvement, a second surgery was performed, including orchietomy, hemi-scrotectomy, and ipsilateral retroperitoneal lymph node (RPLN) dissection. The other testicle was temporarily transposed to the abdomen during chemotherapy and radiation for fertility preservation. Following the second surgery, the patient received chemotherapy and radiation treatment. The chemotherapy regimen included vincristine, actinomycin, and cyclophosphamide. Six-month and one-year follow-up MRI scans with contrast showed no evidence of recurrence.

3. Discussion

RMS arises from primitive mesenchymal cells and exhibits varying degrees of skeletal muscle differentiation. PRMS, particularly, originates from the parenchymal tissue of the spermatic cord, epididymis, and testicular tunics, resulting in the formation of a painless scrotal mass. There are three histological types of RMS: pleomorphic, embryonal, and alveolar, with the embryonal type being the most prevalent (84%). The embryonal type also offers the best prognosis compared to the other histological types. PRMS is commonly observed in children and adolescents, presenting as a rapidly growing and painless scrotal mass. The median age at diagnosis is around 7 years. Fortunately, due to its superficial location, PRMS can be diagnosed early, enabling complete surgical resection and leading to a very favorable prognosis, as demonstrated in our case. About 26–71% of patients with PRMS show positive regional lymph nodes. Metastasis is less frequent and associated with poor prognosis. The most common site of metastasis is lung, followed by bone. Older age has been correlated with metastatic stage at presentation, alveolar histological subtype, and poor prognosis. Testicular tumor markers are usually normal in PRMS.

Scrotal US plays a crucial role in the early detection of scrotal masses, although its sensitivity depends on the experience of the radiologist. On US, PRMS is a lobulated paratesticular mass with variable echogenicity and increased blood flow compared to the adjacent testis. The US appearance can mimic acute epididymitis if the mass is within or abutting the epididymis; young patient age and gradual (rather than acute) symptoms help differentiate PRMS from epididymitis. Following US, staging cross-sectional imaging of the retroperitoneum, such as CT or MRI, is advised for detecting enlarged RPLNs, particularly when they exceed 1 cm. Nerve-sparing RPLN sampling is recommended for patients aged ≥10 years, irrespective of tumor size or radiological lymph node involvement.

Historically, embryonal RMS has been characterized by immature rhabdomyoblasts along with spindle cells arranged in fascicles and whorls. Cytologically, embryonal RMS displays spindle cells with centrally placed nuclei and well-defined cytoplasm, along with large fragments of grass-like cytoplasmic processes, sometimes exhibiting cross-striations. Other differentials for spindle cell neoplasms include fibrosarcoma, solitary fibrous tumor, malignant peripheral nerve sheath tumor, malignant fibrous histiocytoma, and inflammatory myofibroblastic tumor. Given the broad range of differential diagnoses for spindle cell tumors, immunohistochemistry is used to confirm the definitive diagnosis of RMS. In immunohistochemistry, RMS typically expresses myogenic markers such as desmin, myoglobin, MyoD1, and myogenin, as demonstrated in our case.

Treatment options for PRMS include inguinal orchietomy, radiotherapy, retroperitoneal lymph node dissection, and chemotherapy, depending on the disease stage, classified according to the post-surgical classification of the IRSG. The VAcdC regimen (vincristine, actinomycin D, and cyclophosphamide) is the most widely used adjuvant chemotherapy due to its effectiveness against multidrug-resistant proteins in tumor cells. RMSs have demonstrated resistance to multidrug therapies.

4. Conclusion

PRMS is a rare yet aggressive tumor, originating from the parenchymal tissues of the scrotal structures. Timely diagnosis through scrotal US and staging cross-sectional imaging are essential for determining the treatment approach, thus ensuring a good prognosis. Nerve-sparing RPLN sampling is also recommended for all patients aged ≥10 years. The histopathological examination, including immunohistochemistry, plays a pivotal role in confirming the diagnosis of PRMS and distinguishing it from other spindle cell tumors. Surgical resection combined with chemoradiotherapy is the mainstay of treatment, with the VAcdC regimen being the preferred choice due to its activity against multidrug-resistant proteins.
Conflicts of interest

The authors state there are no conflicts of interest.

References