Case Report

Rare Occurrence of Paratesticular Rhabdomyosarcoma in Adulthood: A Case Report

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Abstract

Paratesticular rhabdomyosarcoma is an extremely rare malignant tumour that primarily affects pediatric patients but is uncommon in adults. Prompt diagnosis and treatment are essential to prevent metastasis and improve prognosis. A 28-year-old male was presented with a painless right scrotal swelling. Physical examination revealed a palpable mass in the right scrotal region, raising suspicion of a malignant tumour. Ultrasonography showed a hypoechoic mass originating from the right epididymis. The patient underwent a high inguinal orchidectomy, and histopathological examination confirmed the diagnosis of paratesticular rhabdomyosarcoma. Staging computed tomography revealed no evidence of metastasis. The patient received chemotherapy based on risk stratification. Paratesticular rhabdomyosarcoma carries a poor prognosis in advanced stages, highlighting the importance of early diagnosis. Surgery remains the mainstay of treatment and the role of adjuvant therapy in adults is not well established. Understanding the clinical manifestations, diagnosis and treatment of this rare malignancy can aid in improved management and outcomes

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Introduction

Primary testicular tumours are the most common solid malignant tumours in men aged 20 to 35 years. Germ cell cancers account for over 90% of all testicular cancers, including seminomas and nonseminomas such as choriocarcinoma, yolk sac tumours and teratomas. However, for paratesticular tumours, the majority of cases are benign such as lipoma, leiomyoma, haemangioma, with only around 30% being malignant, such as mesotheliomas and rhabdomyosarcomas (1).

Rhabdomyosarcoma is a rare form of malignant tumour that originates from mesenchymal cells and belongs to the skeletal muscle lineage. It is more commonly found among the pediatric age group, with the majority of cases occurring before the age of ten (2). Paratesticular rhabdomyosarcoma is extremely rare in adults, making up less than 1% of adult malignancies, with an estimated incidence of 1 in 20 million males per year. In this case, we aimed to describe the clinical manifestations, diagnosis, treatment and prognosis of paratesticular rhabdomyosarcoma in our patient, with the goal of enhancing the understanding of this uncommon malignancy.

Case Report

A 28-year-old male, otherwise healthy with no comorbidities, presented with a painless right scrotal swelling that had been presented for 2 months. The swelling was slow-growing, and the patient described a heavy sensation while walking. He denied experiencing constitutional symptoms. Physical examination revealed a fit and young man with a palpable, hard mass in the right scrotal region. The distinction between the testis and paratesticular

structures was clinically indistinguishable, with a negative cough impulse and no transillumination. No abdominal mass was palpable, and the respiratory and cardiovascular systems appeared normal. Routine baseline blood investigations and urine full examination microscopic examination (UFEME) yielded unremarkable results. Alpha-Fetoprotein (AFP) and Beta-Human Chorionic Gonadotropin (B-HCG) levels were within the normal range.

An ultrasonography assessment revealed a heterogeneous hypoechoic mass originating from the body of the right epididymis, measuring 3.5 x 4.7 x 5.2 cm (AP x W x CC). Minimal vascularity was observed within the mass, and it had a clear plane with the adjacent right testis (Fig. 1). There was no invasion of the overlying skin or extension into the scrotal neck. A negative cough impulse was demonstrated, and both testes were normal in size and echogenicity. Minimal hydrocele was present bilaterally. These findings raised the possibility of an epididymal mass, possibly malignant in nature.

After thorough clinical consultation, the patient was scheduled for a high inguinal orchidectomy. Due to his young age and desire to conceive in the future, a referral for sperm banking was made prior to the surgery. The operation proceeded without complications, and the patient was discharged on the first day after the surgery. A follow-up clinic visit two weeks later revealed a healed wound, and the final histopathology report confirmed a diagnosis of right paratesticular rhabdomyosarcoma.

The histopathological examination of the right orchidectomy specimen included the spermatic cord (70 x 15 mm) and the testis (50 x 20 x 30 mm) (Fig. 2), with a combined weight of 147 grams. The tunica vaginalis was partially cut open, exposing a tumour in the paratesticular area. Upon sectioning, the tumour measured 75 x 55 x 55 mm and was located 75 mm from the spermatic cord resection margin. The tumour displayed a smooth multilobulated outer surface and a homogenous nodular cut surface with a whorling appearance. No necrotic areas or hemorrhage were observed. There was no invasion through the tunica vaginalis, adjacent testicular parenchyma, or spermatic cord.

Microscopically, the tumour tissue showed neoplastic cells arranged in patternless sheets, with areas demonstrating alternating hyper and hypocellular zones and myxoid areas. The neoplastic cells exhibited pleomorphic spindle to ovoid hyperchromatic nucleoli, with occasional conspicuous nucleoli. Abundant rhabdomyoblastic differentiation, perivascular accentuation by the tumour cells, and occasional cytoplasmic cross-striations (strap cells) were observed. Mitoses were brisk, with a count of 81 mitoses per 10 high-power fields (HPF). No necrosis was present. The tumour enveloped the epididymis, but there was no invasion of the tunica vaginalis, vas deferens, or testicular parenchyma. Lymphovascular invasion was not identified, and the spermatic cord resection margin was clear. Immunohistochemistry studies revealed that the neoplastic cells were positive

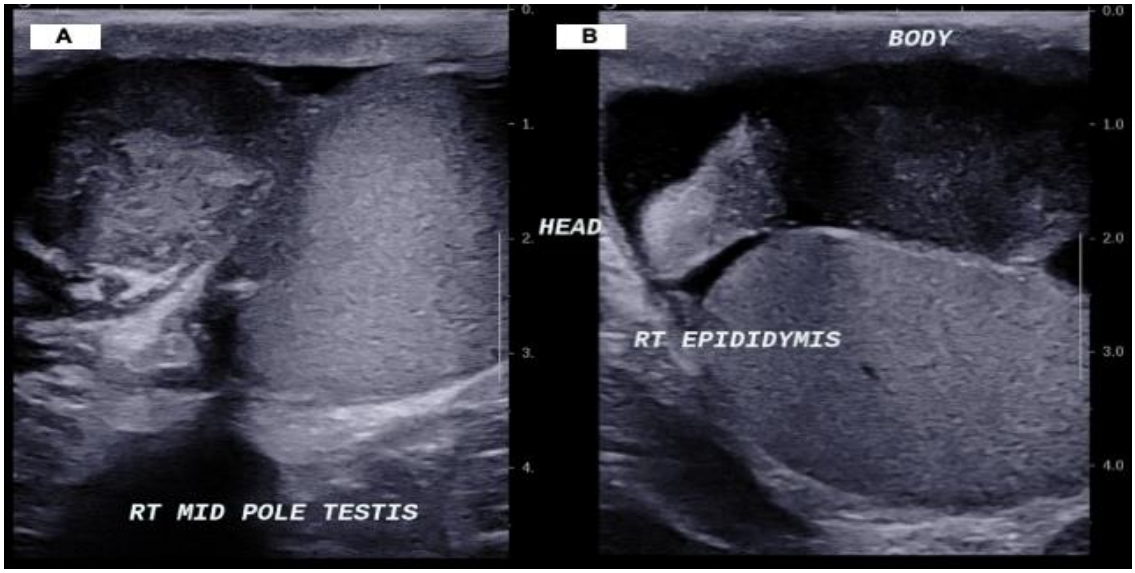

FIGURE 1: (A) and (B) showing mass arising from the right epididymis

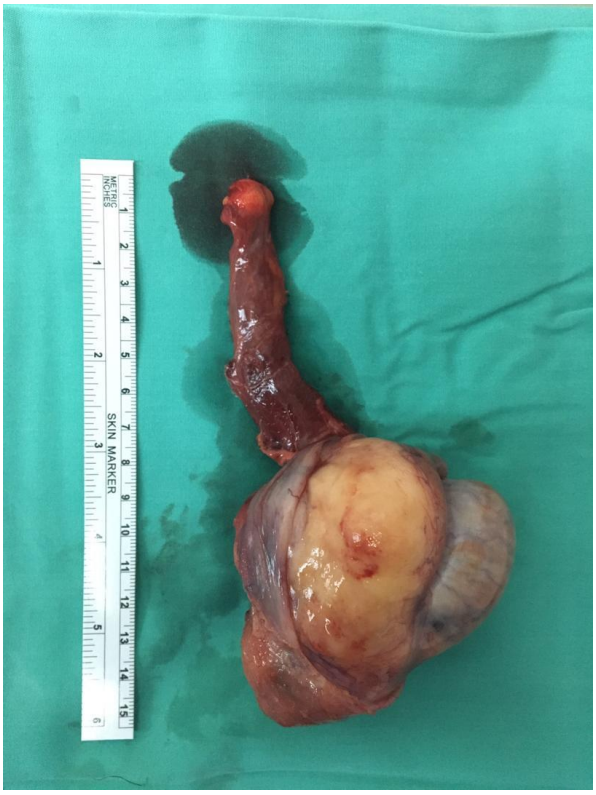

FIGURE 2: Intraoperative image showing the paratesticular tumour removed with the normal testis

for desmin and Myo-D1, while they were negative for CKAE1/AE3, S100, SMA and AFP. Overall, the findings were consistent with paratesticular rhabdomyosarcoma, FNCLCC grade 3.

Staging computed tomography (CT) was performed during this period, which did not reveal any evidence of distant metastasis. The case was subsequently discussed in a Multidisciplinary Team meeting involving the urologist, pathologist, radiologist and oncologist. It was decided to initiate chemotherapy based on the patient's risk stratification, with radiotherapy deemed unnecessary. The patient received nine cycles of chemotherapy over a 25-week period, with one cycle administered every three weeks. Ifosfamide was administered for four cycles, while vincristine and actinomycin-D were given in all nine cycles.

Discussion

Two commonly found malignant paratesticular tumours are mesotheliomas and rhabdomyosarcomas. Mesotheliomas arise from the mesothelium covering the tunica vaginalis, tunica albuginea, epididymis or spermatic cord (3), while rhabdomyosarcoma is a rare malignant tumour that originates from mesenchymal cells of skeletal muscle lineage. Rhabdomyosarcoma is more common in pediatric age groups, with the majority of cases occurring before the age of ten (2). In adulthood, rhabdomyosarcoma comprises less than 1% of all malignancies (4). Most rhabdomyosarcomas tend to occur in extremities, while a small percentage originates from the mesenchymal tissue of the spermatic cord, epididymis, and testis (2). Early and accurate diagnosis of this rapidly spreading tumour is crucial for effective treatment. Rhabdomyosarcoma does not exhibit elevated levels of tumour markers such as AFP and B-HCG.

The confirmatory diagnosis of rhabdomyosarcoma is achieved through histopathological examination, which reveals several histological subtypes, including pleomorphic, alveolar, botryoidal, embryonal and spindle cell variants (5). Immunohistochemical staining, such as testing for Desmin and Myosin, is commonly used as an adjunct in diagnosing the disease. Embryonal type is the most common subtype in paratesticular rhabdomyosarcoma cases. However, in the presented case, molecular testing was not performed, so the specific subtype of rhabdomyosarcoma remains unknown. The embryonal and spindle cell subtypes of rhabdomyosarcoma are generally associated with a more favourable prognosis compared to the alveolar subtype (6).

Staging of soft tissue sarcomas, including rhabdomyosarcoma, utilises the TNM system to assess the extent of the tumour, lymph node involvement and presence of metastasis. Additionally, soft tissue sarcomas can be classified according to the grade of sarcoma using the French system (FNCLCC), which considers differentiation, mitotic count and tumour necrosis (7). Higher grade tumours indicate a more aggressive form of the disease and worse prognosis.

The recommended surgical approach for paratesticular rhabdomyosarcoma is a radical inguinal orchiectomy with high dissection and ligation of the cord structures, following the standard procedure for testicular or paratesticular malignancies. Transscrotal approaches, inguinal tumorectomy without orchiectomy, and inguinal tumour biopsy are not indicated as they carry a risk of scrotal contamination (8). Care must be taken not to breach the scrotal skin, as this can lead to seeding and metastasis of the tumour. If scrotal violation occurs, primary re-excision with hemiscrotectomy may be necessary. In this case report, our patient also underwent an inguinal orchiectomy with a high inguinal approach, adhering to the recommended surgical protocol.

There is no standardised treatment algorithm for adult paratesticular rhabdomyosarcoma. Complete resection of the primary tumour is the treatment of choice, while retroperitoneal lymph node dissection (RPLND) remains controversial (9) and unnecessary for localised disease due to the efficacy of computed tomography (CT) in detecting recurrence, the potential morbidity associated with RPLND, the low rate of retroperitoneal recurrence, and the presumed efficacy of chemotherapy in controlling of microscopic disease (10). Treatment is individualised based on thorough staging and histopathological evaluation. Chemotherapy, typically including the VAC regime (Vincristine, Actinomycin, and Cyclophosphamide), has been shown to be effective among pediatric patients. Radiotherapy is complementary and aims to eliminate residual foci and treat retroperitoneal lymph nodes. The role of adjuvant chemotherapy in adults is still poorly understood (9) but is believed to control retroperitoneal dissemination and minimise such spread (10). Due to the presumed efficacy of chemotherapy in controlling microscopic disease, prevention from disease spread, and its demonstrated effectiveness among pediatric patients, our patient received chemotherapy as recommended.

Paratesticular rhabdomyosarcoma in adulthood is an aggressive tumour that requires early diagnosis and prompt treatment to prevent metastasis. Localised disease carries a good prognosis and overall survival. Unfortunately, advanced paratesticular rhabdomyosarcoma has a poor prognosis, with a 1-year overall survival (OS) rate of 68% and a 5-year OS rate of 30% (4). In a study evaluating RMS at all sites in 2600 patients, adults with RMS experienced significantly worse prognosis than the children (11). Retroperitoneal lymph node (RPLN) involvement is an important prognostic factor, with a 5-year disease-free survival rate of 97% for patients without RPLN involvement and 42% for those with RPLN involvement (12). Other factors influencing survival rates include tumour histology, diameter, stage and location, patient age, response to chemotherapy and metastasis status.

Conclusion

Paratesticular rhabdomyosarcoma in adulthood is an aggressive tumour that requires early diagnosis and prompt treatment to prevent metastasis. Localised disease is associated with a favourable prognosis and overall survival. Therefore, surgery should be arranged as early as possible to obtain a histopathological evaluation, which helps in prognostication. While there is no standard proposed algorithm for the management of rhabdomyosarcoma, adjuvant therapy has been shown to improve survival rates.

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