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Embryonal rhabdomyosarcoma of the testis in a 17-year-old male

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ABSTRACT

Primary rhabdomyosarcoma of the testis is an exceptionally rare and highly malignant sarcoma. To date, there are only 23 reported cases in the literature. We report a 17-year-old male patient presented with massive scrotal swelling that had been progressively enlarging over seven-months. Scrotal ultrasound and contrast-enhanced CT revealed a 10 × 10cm left testicular heterogeneously enhancing mass that extends into the spermatic cord. A left inguinal orchiectomy was performed, and histopathological examinations showed findings consistent with Rhabdomyosarcoma, Embryonic-type. Primary embryonal testicular rhabdomyosarcoma has poor prognosis, particularly for adolescents, and tumour size greater than 10cm. Early diagnosis and radical orchiectomy improve the prognosis.

1. Introduction

Primary rhabdomyosarcoma (RMS) of the testis is an extremely rare and highly malignant sarcoma. Rhabdomyosarcoma represents approximately 3% of adult soft tissue tumours and is the most prevalent soft tissue sarcoma in children and adolescents, with an incidence rate of 4.5 per 1 million individuals.[1] One-fifth of urinary system rhabdomyosarcomas arise in the testis or epididymis, the third most common soft tissue sarcoma. It is derived from striated muscle cells and mesenchymal cells that have differentiated into rhabdomyosarcoma.

Primary embryonal testicular rhabdomyosarcoma is exceptionally rare. To date, there are only 23 reported cases in the existing body of the literature.[2] We present a 17-year-old male patient with Primary testicular embryonal rhabdomyosarcoma successfully managed with radical inguinal orchiectomy.

2. Case

A 17-year-old male patient came to our urology outpatient clinic complaining of massive left scrotal swelling, which was gradually enlarging over seven months. The mass was painless, but he experienced discomfort from the mass. The mass was hard and painless on physical examination, with no translumination (Fig. 1). A scrotal ultrasound revealed a mass lesion with heterogeneous echogenicity and central necrotic areas measuring approximately 13 × 10 cm. In the Dopper examination, low-velocity vascular structures were observed in the central part of the lesion. Tumour markers were within normal limits (β-hCG: 2.0, AFP: 1.29), except LDH showed a mild elevation of about 269 (normal range: 0–248). Other blood investigations were unremarkable.

A contrast-enhanced abdominopelvic CT scan revealed an enlarged left testis (10 × 10 cm), which is a hypodense heterogeneously enhancing mass that extends into the spermatic cord to the inguinal canal (Fig. 2). No metastatic lesions were detected on the chest and abdominopelvic CT scan. A left inguinal orchiectomy was performed, and the specimen was extracted with caution to the proximal cord for high ligations (Fig. 3). During the operation, no significant enlarged inguinal lymph nodes were seen.

Macroscopic findings of histopathological examinations showed testis measuring 15 × 12 × 10 cm, epididymis measuring 7 × 2 × 0,5 cm, and mass measuring 14 × 9x8 cm, with a cut surface of grey-white focal hemorrhagic and semisolid lobulated tissue. The section shows primitive mesenchymal cells that show variable degrees of skeletal muscle differentiation and both hypocellularity and hypercellular areas with a loose, myxoid stroma with Sheets of small, stellate, spindled or round cells with scant or deeply eosinophilic cytoplasm and eccentric, small oval nuclei (Fig. 4). These findings are consistent with Rhabdomyosarcoma, Embryonic-type, and invading the epididymis, negative for malignancy for spermatic cord side (stage pT2).

The postoperative was uneventful, and the patient was discharged

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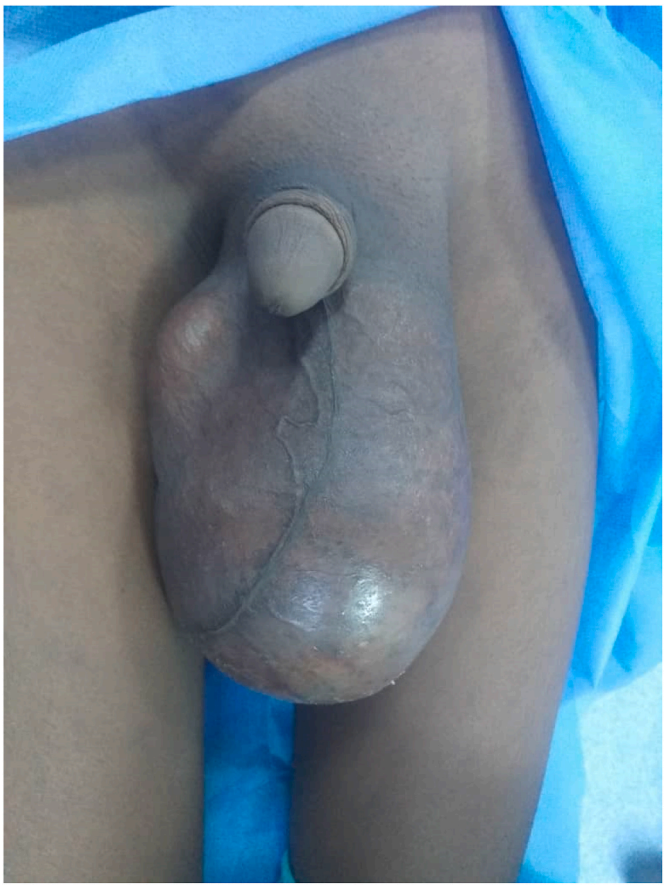

Fig. 1. Huge scrotal swelling.

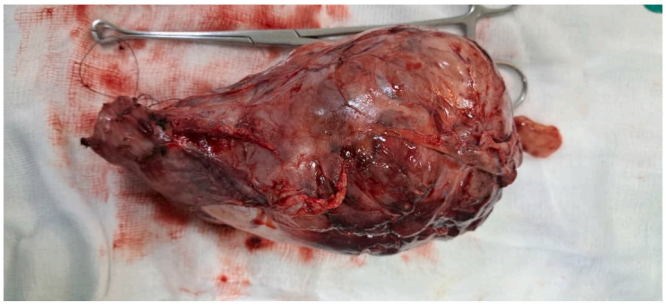

Fig. 2. Extracted specimen.

with antibiotics and analgesics. A consultation with an expert medical oncologist suggested no need for chemotherapy despite a negative metastasis check-up. A retroperitoneal lymph node dissection and inguinal lymph node dissections did not proceed due to their increased morbidity. A six-month follow-up with contrast-enhanced CT of the chest and abdominopelvic was planned for early detection of recurrence.

3. Discussion

Rhabdomyosarcoma is a mesenchymal tumour in various body parts such as the head, neck, genitourinary, and extremities. WHO reported different histological subtypes, namely alveolar, embryonal, pleomorphic and spindle types of RMS. The embryonal subtype is the most prevalent in children, accounting for 70% of childhood RMS.[3]

Primary intratesticular rhabdomyosarcoma (PITRMS) is an uncommon tumour found inside the scrotum, particularly in the tunica vaginalis, epididymis, or spermatic cord. There are two theories about the

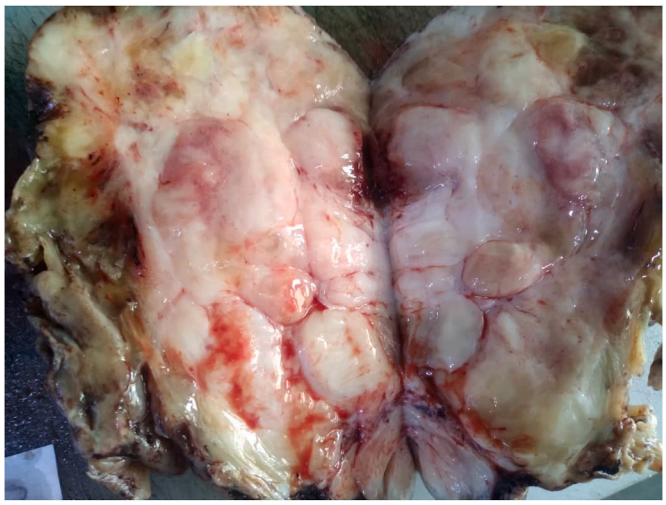

Fig. 3. Cutting surface of the tumour.

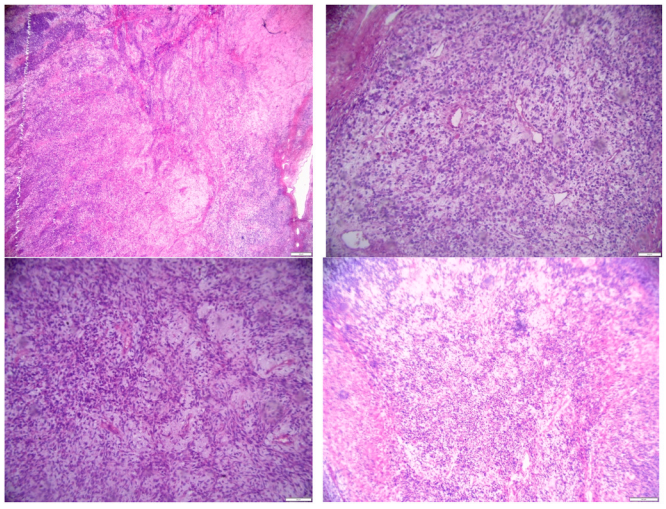

Fig. 4. Testicular malignancy with predominant small round blue cell features (200 ×), predominant spindle cell and myxoid features. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

origin of PITRMS. One suggests that it comes from dedifferentiated mesenchyme that can differentiate into rhabdomyolysis. The other theory proposes that it comes from embryonal muscle tissue displaced during early tissue development. However, it has yet to be proven to come from metaplasia of connective tissue or smooth muscle.[4]

Diagnosis of individuals with PITRMS is initiated with a thorough assessment of medical history and a comprehensive physical examination. The patients exhibit a gradual and painless enlargement of the scrotum that has been ongoing for a certain period. Other diagnostic modalities include abdominopelvic ultrasound, CT scan, MRI, histopathology and immunohistochemistry staining. In the present case, the ultrasound gives a clear description of the mass and the possibility of extension of the mass to adjacent areas, but it lacks specific RMS features. Other imaging modalities (CT and MRI) also need more specificity in revealing the nature of the tumour and details for distant metastasis.

Management of PITRMS patients remains controversial due to its rarity in nature and limited cases in the literature. A comprehensive treatment strategy that encompasses surgery and chemotherapy, with or without radiation, has been developed, considering risk stratification groups.[5] Performing radical inguinal orchiectomy (RIO) as the primary

therapy for ITRMS is necessary for all patients. In our case, the malignancy was confined to the epididymis and found negative for the spermatic cord side (stage pT2), and left **inguinal orchiectomy was performed.** Chemotherapy is often successful in treating RMS, and the standard chemotherapy regimen for IRSG treatment is VAC, which includes vincristine, actinomycin D, and cyclophosphamide.[5]

4. Conclusion

Primary embryonal testicular rhabdomyosarcoma has a poor prognosis, particularly for adolescents, and tumour size greater than 10cm. Early diagnosis and radical orchiectomy improve the prognosis. Adjuvant chemotherapy or retroperitoneal lymph node dissections are recommended, especially in the presence of positive lymph nodes or distant metastasis in radiological investigations.

Ethical approval

No approval from the Ethics Committee was necessary in case reports.

Patient consent

The parental informed consent for publication was obtained.

Conflict of interest and funding

The authors report no conflicts of interest and funding in this work.

CRediT authorship contribution statement

Abdikarin Ahmed Mohamed: Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Methodology, Formal analysis, Data curation, Conceptualization. **Aweis Abdullahi Sheik:** Writing – original draft, Validation, Supervision, Data curation, Conceptualization. **Mohamed Abdikarim Nur-Amin:** Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Formal analysis, Data curation. **Khaled Ali Mohamed:** Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Software, Formal analysis, Data curation, Conceptualization. **Abdikarim Hussein Mohamed:** Writing – review & editing, Writing – original draft, Visualization, Validation, Supervision, Investigation, Funding acquisition, Formal analysis, Data curation, Conceptualization.

References

1. Wu R, Liu X, Song Y, et al. Spontaneous rupture of rhabdomyosarcoma of the testis with unilateral ptosis: a case report and literature review. *Frontiers in Pediatrics*. 2022 Jun 28;10, 904275.
2. Zou J, Luo P, Chen T, Qian B. Embryonal rhabdomyosarcoma of the testis in a young male: a rare case report. *Asian J Surg*. 2023 Dec;3:S1015–S9584.
3. WHO Classification of Tumours Editorial Board. Soft tissue and bone tumours. In: *WHO Classification of Tumours Series*. Lyon (France): International Agency for Research on Cancer; 2020.
4. Yahaya JJ, Mremi A. Primary intratesticular rhabdomyosarcoma in children: a case report and review of the literature. *J Med Case Rep*. 2021 Jan 31;15(1):37. https://doi.org/10.1186/s13256-020-02599-z. PMID: 33516251; PMCID: PMC7847602.
5. Chen X, Zou C, Yang C, et al. Pleomorphic rhabdomyosarcoma of the spermatic cord and a secondary hydrocele testis: a case report. *World Journal of Clinical Cases*. 2020 Jun 6;8(12):2641.