Report of a Rare Case of Childhood *Paratesticular Rhabdomyosarcoma* (PT-RMS) and Literature Review

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Abstract

Paratesticular rhabdomyosarcoma (PT-RMS) is a rare, aggressive soft tissue malignancy affecting children and adolescents. Early diagnosis and multimodal treatment are crucial to reduce recurrence and improve survival. We report a 6-year-4-month-old male with advanced PT-RMS presenting with a bleeding scrotal mass, severe malnutrition, and anemia. The patient had previously received 25 chemotherapy cycles and experienced relapse, requiring reinitiation of chemotherapy with vincristine, actinomycin C, and cyclophosphamide. Imaging revealed an extensive fixed mass involving the scrotum, penis, bladder wall, and enlarged inguinal and para-iliac lymph nodes. Multidisciplinary management included supportive care, wide excision, and planned bilateral inguinal lymph node dissection. This case highlights the diagnostic and therapeutic challenges of advanced PT-RMS, emphasizing the role of proper imaging, radical surgery with inguinal lymphadenectomy, and adjuvant chemotherapy to optimize local control and prevent metastasis. Effective management of pediatric PT-RMS requires timely diagnosis, complete surgical resection with lymph node evaluation, and adequate adjuvant therapy to improve outcomes and minimize recurrence risk.

Keyword: *Chemotherapy, Inguinal Lymph Node Dissection, Paratesticular Rhabdomyosarcoma, Pediatric Oncology.*

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A. INTRODUCTION

The malignant soft tissue sarcoma known as rhabdomyosarcoma (RMS) is thought to develop from primitive mesenchymal cells, which normally undergo skeletal tissue differentiation. These tumors, however, can develop in any anatomical location and in different kinds of tissue. Just 1% of soft tissue sarcomas are identified in adults, with RMS being the most frequent in youngsters. There is still much to learn about the etiology and risk factors. Rhabdomyosarcoma can be linked to hereditary disorders; however, the majority of cases are spontaneous (Kaseb et al., 2025; Amer et al., 2019; Li et al., 2021).

The most prevalent soft tissue sarcoma in children and adolescents is rhabdomyosarcoma, which affects 4-7 out of every million 15-year-olds annually (Mejía-Salas et al., 2017). According to the study, Dr. Cipto Mangunkusumo General Hospital in Jakarta, Indonesia, it has 44 RMS instances in 8 years, while Dharmais Cancer Hospital has 11 RMS cases in 4 years (Reniarti et al., 2020).

The embryonic mesenchyme that develops into skeletal or striated muscle is the source of RMS. As a result, RMS may develop anyplace in the human body where skeletal muscle is present or absent, including the bile ducts, bladder, etc. Not all of the symptoms manifest as a lump. The primary site of the tumor determines the

clinical manifestations, with most symptoms relating to the masses compression of nearby structures and possibly resulting in organ-specific complaints (Reniarti et al., 2020). The head, neck, and genitourinary tract are where it most commonly occurs (Ihsaan et al., 2022).

Paratesticular embryonal rhabdomyosarcoma (RMS) is a rare and aggressive tumor that accounts for approximately 7% of all RMS cases. It originates from the mesenchymal tissues of the testis, epididymis, spermatic cord, and surrounding structures, and most commonly affects children, adolescents, and young adults. Clinically, it typically presents as a painless scrotal mass (Kura et al., 2025; Aydin et al., 2020). Serum testicular tumor markers are typically not increased, making preoperative detection of paratesticular rhabdomyosarcoma difficult. Moreover, ultrasonography results are frequently non-specific (Kura et al., 2025).

B. CASE PRESENTATION

A 6-year-4-month-old male child presented to the hospital with complaints of bleeding from a scrotal mass that began one day prior to admission. The initial bleeding episode was profuse, saturating approximately one gauze pad, and was temporarily controlled using a compression dressing. Currently, the bleeding persists in small intermittent amounts. The mass has progressively increased in size, accompanied by itching and purulent discharge. Additionally, the patient experienced a single episode of epistaxis that resolved spontaneously. There were no associated symptoms such as nausea, vomiting, fever, cough, shortness of breath, seizures, or altered consciousness. Bowel movements and urination were within normal limits.

The patient was diagnosed with paratesticular rhabdomyosarcoma at the age of 3. He had undergone 25 cycles of chemotherapy but experienced a relapse. As a result, chemotherapy was restarted from the beginning of the rhabdomyosarcoma protocol. He had recently completed his 9th cycle on January 28, 2025, and was scheduled to receive the 13th cycle on February 20, 2025. He is the third child of a mother with a G5P5A0 obstetric history and was born via spontaneous vaginal delivery at a midwife clinic. The birth weight was 3.1 kg and the infant cried immediately after birth. The child had incomplete immunization, with the last dose received at 6 months of age. Growth was previously appropriate for age until the illness onset.

Upon examination, the patient was alert (compos mentis) with a body weight of 12.7 kg and a height of 106 cm, indicating severe malnutrition (height for age: -2.55 SD; BMI for age: -4.69; MUAC: 11.5 cm, classified as severe wasting). Vital signs were within normal limits: blood pressure 106/67 mmHg, respiratory rate 20x per minute, heart rate 82 bpm, and temperature 36.6°C. The abdomen was flat and soft with active bowel sounds and no guarding.

Urological examination revealed no palpable kidneys or tenderness in the flank regions. The suprapubic region was non-distended and non-tender. A carcinomatous mass was noted in the scrotum, extending to the penis, suprapubic area, and left

inguinal region. The mass measured approximately 10 × 8 × 6 cm, was fixed, easily bled, and was tender to palpation. Both testes could not be evaluated due to tumor infiltration.

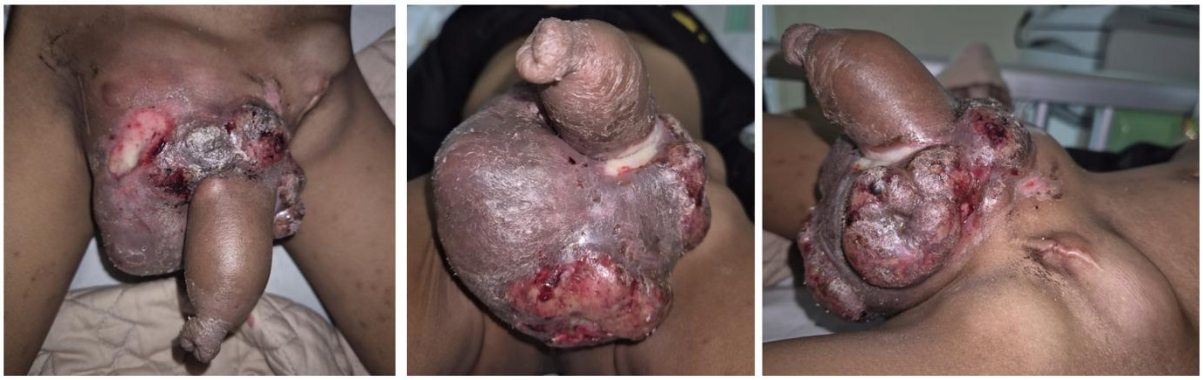

Figure 1 Clinical Photo

Scrotal ultrasound showed bilateral hydronephrosis and infiltration of the posterolateral bladder wall. MRI of the pelvis with contrast revealed an inhomogeneous, irregular mass measuring approximately 10 × 8 cm in the bilateral scrotal region extending into the subcutaneous anterior inguinal areas. The mass infiltrated both testes and the dorsal penis. Multiple enlarged para-iliac and inguinal lymph nodes were identified, the largest measuring 3 × 2 cm. There was also evidence of bilateral pelvocaliceal dilatation. The urinary bladder appeared irregular with a visible Foley catheter balloon inside and a suspected blood clot without vascularization on color Doppler. The prostate appeared homogeneous with a volume of 86.8 cc based on TAUS.

Based on clinical, imaging, and treatment history, the diagnosis was fixed and unresectable testicular rhabdomyosarcoma stage cT4N1M0, currently on the 9th chemotherapy cycle. Complicating conditions included severe malnutrition, stunted growth, underimmunization, anemia related to chronic disease, and hypokalemia. The patient was planned for general condition optimization in coordination with the Pediatric Department, followed by wide excision of the scrotal mass and bilateral inguinal lymph node dissection (ILND).

a b c

Figure 2 Scrotal USG (a) Right Scrotum; (b) Inferior Part of Testicle; (c) Left Scrotum

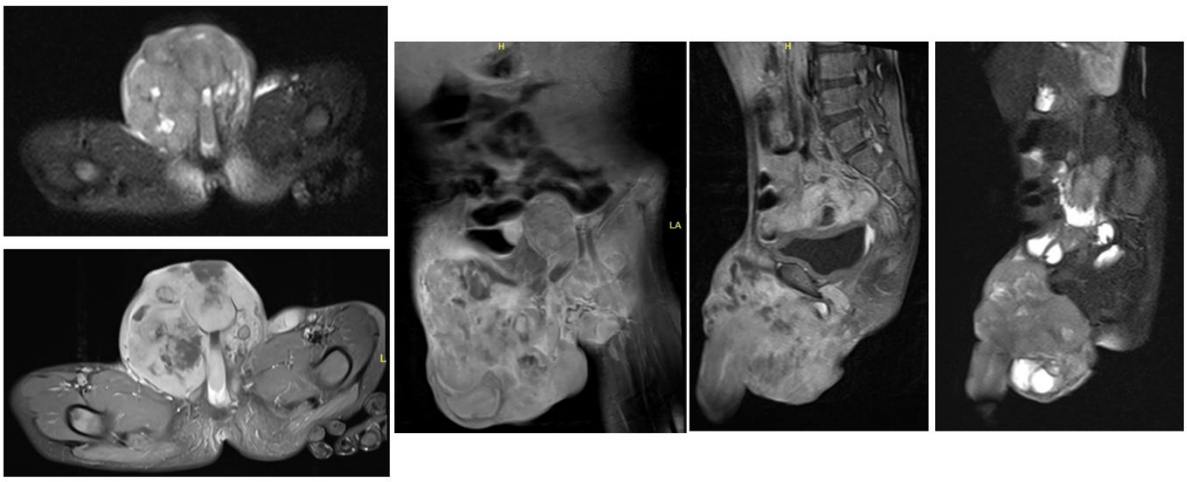

Figure 3 Pelvic MRI with Contrast.

C. RESULTS AND DISCUSSION

Primitive mesenchymal cells with variable levels of skeletal muscle development are the source of rhabdomyosarcoma. It is more common in the genitourinary tract, extremities, head, and neck. The majority of rhabdomyosarcoma cases are intermittent and lack known risk factors. Nonetheless, there are a few known hereditary and non-genetic risk factors for rhabdomyosarcoma (Kura et al., 2025; Yi et al., 2016).

Although the intratesticular position of urogenital lesions is uncommon, the paratesticular location is the most common. Young adults are diagnosed with it less commonly than children and adolescents. Children between the ages of 1 and 5 have the highest incidence of paratesticular rhabdomyosarcoma. The age of sixteen marks the second peak incidence. Usually, a painless unilateral scrotal enlargement is the initial sign of intrascrotal embryonal rhabdomyosarcoma. However, in this case, the children have a bilateral paratesticular rhabdomyosarcoma.

In previous studies, it was reported that patients experienced pain along with accelerated tumor growth, while scrotal ultrasound scans showed areas of no flow on Doppler interrogation and anemia, possibly due to intra-tumor hemorrhage and infarction (Kura et al., 2017). This is similar to what our patient experienced; the examination results showed bleeding in the mass. The patient was also diagnosed with anemia and severe malnutrition.

The clinical signs of paratesticular RMS are non-specific when compared to other intra-scrotal malignancies. Ultrasonography (US) can be a helpful diagnostic tool when physical examination of tumor location is challenging. According to the scrotal US data, the lesions were heterogeneous echogenic masses that spread to the inguinal area and scrotum. According to earlier research, US of the scrotum and its contents has a sensitivity of > 95% and can accurately differentiate between testicles and scrotal masses. Although MRI and CT scans are both capable of precisely determining the mass's location, size, and metastases, they are not suitable for use as confirmatory diagnostic procedures. Lesions like leiomyosarcoma, liposarcoma, and

fibrosarcoma are included in the differential diagnosis of paratesticular RMS. Since these tumors lack imaging characteristics, postoperative pathology is necessary for confirmation diagnosis (Yi et al., 2016; Graiouid et al., 2019; Secil et al., 2017). Regional lymph nodes that ascend from the spermatic cord into the ipsilateral retroperitoneum and ultimately reach the renal arteries receive the drainage of PT-RMS. The scrotum empties into the inguinal nodes, while the epididymis empties into the external iliac and ipsilateral pelvic nodes. These locations' nodes are likewise regarded as regional nodes. Metastases are lymph node metastases that occur above the renal vessels (Rogers et al., 2021).

Usually, the testicular tunica is directly invaded, and metastases form through hematogenous or lymphatic pathways. Rhabdomyosarcoma frequently spreads to the brain, liver, omentum, lung, bone marrow, and lymph nodes (Aydin et al., 2020). Similarly with this case, an inhomogeneous, uneven mass of roughly 10 × 8 cm was found in the contralateral scrotal region of the pelvis on a contrast-enhanced MRI, spreading into the subcutaneous anterior inguinal areas. Both the dorsal penis and the testes were penetrated by the bulk. There were several enlarged inguinal and para-iliac lymph nodes found, the largest of which was 3 × 2 cm.

Compared to other RMS, paratesticular RMS typically has a better prognosis and a greater survival rate following radical tumor removal. It frequently affects children and young people.[13] According to earlier data, patients diagnosed at ages <10 and >10 had 5-year survival rates of 97 and 84%, respectively. In other words, maintaining and restoring fertility and other endocrine functioning is a critical concern for long-term survivors. In actuality, patients reproductive endocrine function is probably going to be negatively impacted in some way by surgery, chemotherapy, and radiation (Yi et al., 2016).

Radiation, chemotherapy, and surgery are examples of multidisciplinary treatment options that have significantly improved the prognosis for paratesticular rhabdomyosarcoma. The best course of treatment is chemotherapy after an orchiectomy. Multidisciplinary techniques are typically advised, although there is no set course of treatment. The primary surgical treatments are hemiscrotatectomy, radical orchidectomy, and high inguinal cord dissection combined with inguinal lymph node dissection (Sabbagh et al., 2023; Zhu et al., 2021). Imaging, such as a CT scan or lymphography, should be obtained prior to performing an inguinal lymphadenectomy (Aydin et al., 2020; Bouchikhi et al., 2013). In this case, patient treatment in this case is in line with several previous studies.

After surgery, chemotherapy is utilized as an adjuvant treatment. It can also be used as a neoadjuvant to downstage and make resectable big tumors in patients who have trouble resecting them. Since rhabdomyosarcoma is chemosensitive, chemotherapy is recommended for all patients. Patients with confined disease have a higher chance of survival when receiving chemotherapy. Adult patients are typically treated with regimens that were developed from clinical trials conducted on children (Chalouati et al., 2022). Numerous chemotherapy regimens have been used to treat rhabdomyosarcoma. Ifosfamide, vincristine, etoposide (IVE), ifosfamide, vincristine,

dactinomycin (IVA), or vincristine, dactinomycin, cyclophosphamide (VAC) are the most commonly utilized combinations (Chalouati et al., 2022).

In a study conducted by Chalouati et al. (2022) after six rounds of chemotherapy, a CT scan revealed a residual 5 cm lump in the paraaortic region, but no other indications of metastatic disease. There was no other hypermetabolic location seen on the patient's positron emission tomography (PET) CT scan, which revealed a 4 cm hypermetabolic mass at the level of the renal pelvis. Another study denote that the patient completed nine cycles of vincristine, actinomycin C, and cyclophosphamide chemotherapy, administered in four-day sessions every 21 days, with good clinical progress over four years of follow-up. This case highlights the importance of combining radical inguinal orchiectomy and lymph node dissection with adjuvant chemotherapy and/or radiotherapy to reduce recurrence or metastasis and improve prognosis in intratesticular rhabdomyosarcoma (Azizi et al., 2021).

Retroperitoneal lymphadenectomy is advised for adults with testicular cancer because of their increased risk of retroperitoneal illness, according to another piece of literature. Even in cases where the lymph nodes are clinically clear of disease, preoperative imaging examinations are essential for determining the extent of the disease. The function of retroperitoneal lymph node dissection (RPLND) in the accurate staging and initial management of non-metastatic rhabdomyosarcomas is a topic of continuous discussion, particularly when radiological tests reveal no indication of positive lymph nodes. RPLND, on the other hand, has been proposed to help with tumor debulking and stop lymph node regrowth in cases where nodes keep growing despite chemotherapy (Nhungo et al., 2024). In another study, by examining patients with PT RMS who were gathered from North America and Europe, the significance of carrying out an RPLN surgical lymph node examination was validated 7. Age and tumor size had a substantial impact on the estimated nodal involvement. Positive results were 3% for patients under 10 with tumors smaller than 5 cm, but 32% for those with tumors larger than 5 cm. Regardless of tumor size, nodal involvement was seen in about 32–35% of individuals aged ≥10 years. Regional (RPLN) disease failures were more common than local ones (42% vs 17%, respectively) (Rogers et al., 2021; Walterhouse et al., 2018).

While this patient received a wide excision of the mass, another study conducted that trans-scrotal excision of malignancies has a worse prognosis than an inguinal approach is unclear from conflicting data. Subsequent PRE specimens showed persistent tumor in 56% and 30% of cases following the initial trans-scrotal tumorectomy or trans-scrotal orchidectomy, respectively, and all required multiple procedures (Routh et al., 2020; Hammond et al., 2017; Rogers et al., 2020).

D. CONCLUSION

This case highlights the importance of comprehensive diagnostic workup, timely surgical intervention, and multidisciplinary care in managing advanced paratesticular rhabdomyosarcoma in pediatric patients. Wide excision combined with appropriate lymph node dissection remains the cornerstone of local control.

Awareness of the possibility of relapse and complications such as anemia and severe malnutrition should guide perioperative management to optimize outcomes and improve survival.

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