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TESTICULAR EMBRYONAL RHABDOMYOSARCOMA IN ADOLESCENCE: FROM RADICAL ORCHIECTOMY TO CATASTROPHIC LOCAL RECURRENCE – AN UROLOGICAL PERSPECTIVE ON THERAPEUTIC CHALLENGES AND FATAL OUTCOME

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ABSTRACT

Background: Testicular rhabdomyosarcoma (RMS) is an aggressive soft tissue sarcoma, exceptionally rare in late adolescence, presenting significant diagnostic and therapeutic challenges distinct from common germ cell tumors. This report discusses the urological management of a metastatic embryonal RMS with a focus on surgical dilemmas and local control. **Case Presentation:** A 17-year-old male presented with a 16 cm painful left scrotal mass. Initial tumor markers were negative. CT imaging revealed pulmonary metastases and renal cystic lesions (Bosniak IV). He underwent left radical inguinal orchiectomy. Pathology confirmed embryonal RMS with deep dermal infiltration. Adjuvant VAC chemotherapy (Vincristine, Actinomycin-D, Cyclophosphamide) was initiated but complicated by severe septic shock and scrotal necrosis requiring multiple debridements. Despite initial systemic response, the patient developed extensive local recurrence infiltrating the perineum and contralateral scrotum, alongside progression of bone and pulmonary metastases. Surgical excision of the recurrence was performed for palliation, but the patient rapidly deteriorated due to pancytopenia and extensive metastatic burden, succumbing to the disease 11 months after diagnosis. **Conclusion:** This case illustrates the "therapeutic collision" between the need for aggressive local control and the limitations imposed by systemic metastatic disease. Radical orchiectomy remains the cornerstone of diagnosis and local control, yet scrotal skin involvement portends a dismal prognosis. Early recognition, multidisciplinary management, and balancing surgical morbidity with quality of life are critical in these catastrophic presentations.

KEYWORDS: Embryonal rhabdomyosarcoma; Testicular neoplasm; Radical orchiectomy; Adolescent urology; Local recurrence; Palliative care.

1. INTRODUCTION

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children but represents a distinct rarity in late adolescence and young adulthood. Paratesticular RMS accounts for approximately 7% of all RMS cases, typically affecting young children (bimodal peak: 1-5 years and 15-19 years).[1,2] While the prognosis for localized disease in children is generally favorable with multimodal therapy, outcomes for adolescents, particularly those with metastatic disease (Stage IV) or unfavorable histology like the embryonal subtype in older patients, remain poor.[3,4] From a urological perspective, the presentation of a scrotal mass in a 17-year-old predominantly raises suspicion of a germ cell tumor (GCT). The absence of elevated serum tumor markers (AFP, βHCG) creates a diagnostic blind spot, often resolved only after radical orchiectomy.[5] The management becomes exponentially more complex when the disease presents with massive local bulk, scrotal skin infiltration, and distant metastases.[6]

We present a fatal case of metastatic testicular embryonal rhabdomyosarcoma in a 17-year-old adolescent. This report emphasizes the urological

challenges: the surgical management of the primary tumor, the catastrophic complication of local recurrence requiring salvage surgery, the failure of fertility preservation, and the ethical dilemma of aggressive local control in the face of terminal systemic progression.

2. CASE PRESENTATION

Patient Profile and Clinical Presentation

A 17-year-old male, with a family history of breast cancer (sister), presented with a 3-month history of a progressively enlarging, painful left scrotal mass. He had no history of cryptorchidism or prior trauma. Physical examination revealed a large, hard, fixed left scrotal mass measuring approximately 16x10 cm, with associated inflammatory changes of the scrotal skin. There was no palpable inguinal lymphadenopathy initially.

Diagnostic Workup

Scrotal ultrasound confirmed a heterogeneous, solid intrascrotal process, highly suspicious for malignancy. Serum tumor markers were notably non-contributory for a classic non-seminomatous germ cell tumor: βHCG <10 UI/L, AFP 0.93 ng/mL (later 2.13), and LDH 232 U/L. A staging CT scan (June 2023) revealed a 9x9 cm left testicular mass and, critically, multiple bilateral pulmonary nodules (13mm right, 6mm left) consistent with metastatic disease. Incidental findings included a complex renal cyst (Bosniak IV) on the left kidney, which added to the oncological burden but was secondary to the primary scrotal pathology.

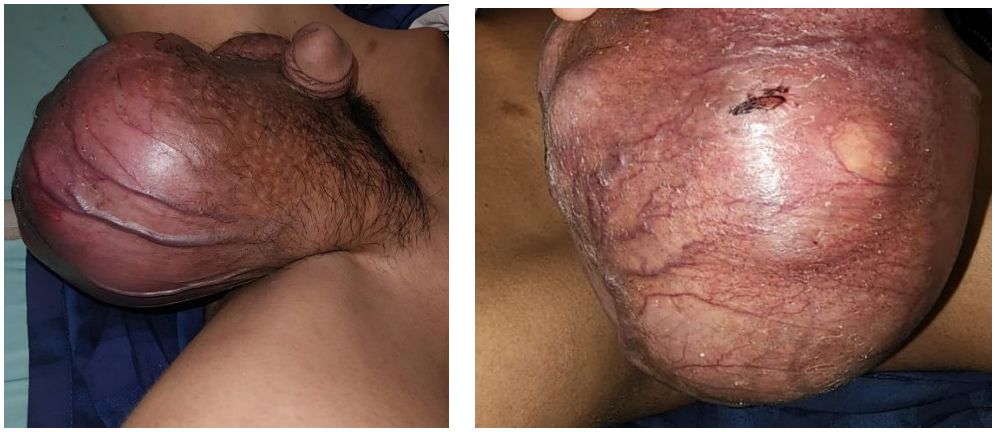

Figure 1: Image showing a left large scrotal mass.

Surgical Management: Phase I

Given the symptomatic nature of the mass and the need for definitive histological diagnosis, the patient underwent a left radical inguinal orchiectomy on July 17, 2023. Intraoperative findings were significant for a tumor adherent to the scrotal wall. The procedure involved high ligation of the spermatic cord at the internal inguinal ring.

Histopathological examination revealed a poorly differentiated malignant proliferation, largely necrotic, arising from paratesticular tissues and engulfing an atrophic testis. Crucially, microscopic analysis showed deep focal infiltration of the scrotal dermis by tumor cells. Immunohistochemistry (IHC) confirmed the diagnosis of **Embryonal Rhabdomyosarcoma**. Semen analysis performed for fertility preservation revealed azoospermia, precluding sperm banking.

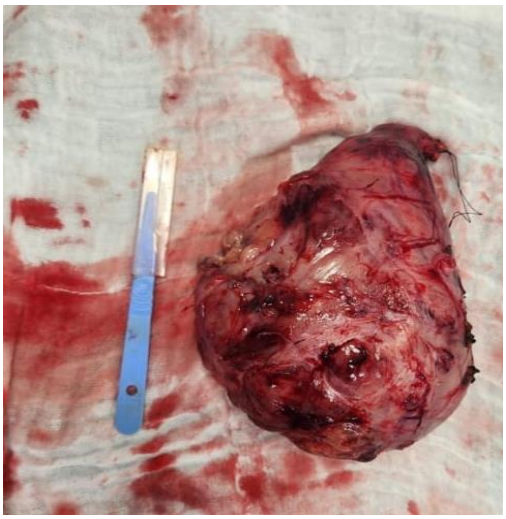

Figure 2: Radical left inguinal orchidectomy specimen with high ligation of the spermatic cord.

Systemic Therapy and Complications

The patient was started on the VAC protocol (Vincristine, Doxorubicin/Adriamycin, Cyclophosphamide) in September 2023. The first cycle was complicated by severe febrile neutropenia (PNN 890/mm³) and septic shock, necessitating transfer to the Intensive Care Unit. Locally, the patient developed extensive scrotal necrosis and gangrene extending to the perineum and inguinal region, likely exacerbated by the rapid tumor lysis and compromised skin vascularity from tumor infiltration. This required aggressive supportive care with broad-spectrum antibiotics (Ceftriaxone, Metronidazole, Aminoglycosides, Vancomycin) and multiple surgical debridements in consultation with plastic surgery.

Disease Progression and Local Recurrence

Initial re-evaluation in December 2023 showed an 80% clinical improvement in the local wound and stable metastatic disease on CT. However, the respite was brief. By March 2024, PET-CT imaging demonstrated a hypermetabolic left scrotal lesion, persistent pulmonary nodules, and new osteolytic bone lesions with pleural thickening. An MRI in May 2024 confirmed a massive local recurrence: a residual scrotal mass with skin infiltration extending to the contralateral side, associated with left inguinal lymphadenopathy and extensive bone metastases to the bilateral proximal femurs, left iliac bone, and sacrum.

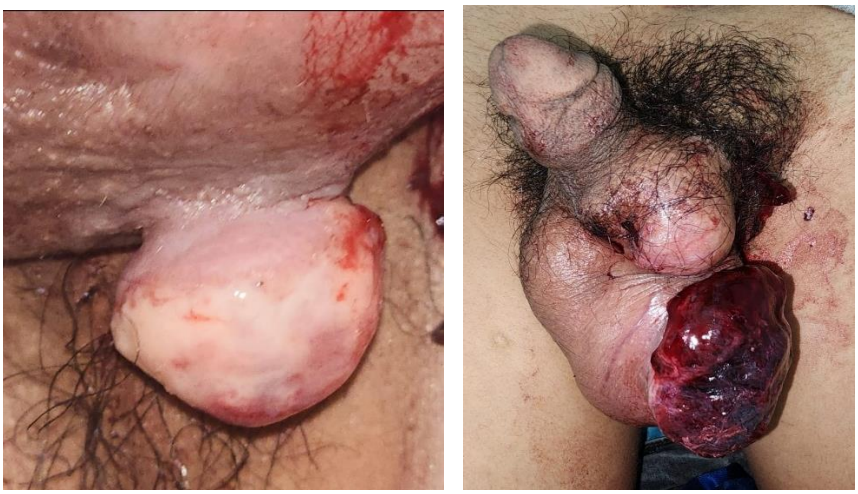

Figure 3: Hemorrhagic residual scrotal mass with skin infiltration extending to the contralateral side.

Surgical Management: Phase II (Salvage)

Despite the metastatic progression, the local recurrence caused significant pain, fungating wounds, and quality of life deterioration. A multidisciplinary decision was made to perform a salvage excision of the local recurrence in May 2024. The postoperative course was complicated by delayed wound healing and persistent serosanguinous discharge.

Terminal Phase

The chemotherapy regimen was switched to Etoposide and Carboplatin due to disease progression. In June 2024, the patient developed severe bicytopenia (Hemoglobin 4.9 g/dL, Platelets 6000/mm³), requiring massive transfusion support. On June 27, 2024, he presented with acute chest pain and dyspnea with elevated D-dimers (9520 ng/mL). CT angiography ruled out pulmonary embolism but revealed the "balloon release" appearance of extensive pulmonary metastases. The patient was declared beyond therapeutic resources on June 28, 2024, and transferred to palliative care, succumbing to the disease shortly thereafter.

Table 2: Timeline of Clinical Management.

Date	Clinical Event	Intervention / Outcome
May 2023	Initial Presentation	Painful scrotal mass (16cm). Markers negative.
June 2023	Staging	CT: Pulmonary metastases + Renal cyst Bosniak IV.
July 2023	Surgery	**Left Radical Inguinal Orchiectomy.** Path: Embryonal RMS.
Sept 2023	Chemotherapy C1	VAC Protocol initiated.
Oct 2023	Complication	**Septic shock & Scrotal Necrosis.** ICU admission, debridement.
Dec 2023	Re-evaluation	Clinical improvement. Stable disease on CT.
Mar 2024	Progression	PET-CT: Hypermetabolic local recurrence + Bone mets.
May 2024	Local Recurrence	**Salvage Excision** of fungating scrotal mass.
June 2024	Terminal Phase	Severe pancytopenia, massive mets. Palliative care.

3. DISCUSSION

Diagnostic Challenges and the Role of Orchiectomy

Testicular RMS in adolescents is an aggressive entity that defies the standard algorithms for scrotal masses. Unlike the pediatric population where outcomes are favorable, adolescents often present with advanced disease.[7,8] In this case, the negative tumor markers were a crucial negative predictor, diverting the diagnosis away from GCTs but failing to positively identify RMS preoperatively. Radical inguinal orchiectomy serves as both the definitive diagnostic tool and the first step in local control.[9] However, the sheer size of the tumor (16 cm) and skin involvement in this patient compromised the oncological safety margin typically afforded by the inguinal approach, leading to microscopic cutaneous infiltration.

The Significance of Scrotal Skin Involvement

The involvement of scrotal skin changes the staging and prognosis dramatically. Scrotal violation —whether by tumor extension or previous trans-scrotal biopsy (not performed here)—opens lymphatic drainage pathways to the superficial inguinal nodes, unlike the standard retroperitoneal drainage of the testis.[10,11] In our patient, the deep dermal infiltration noted on pathology likely contributed to the rapid, fungating local recurrence and the subsequent appearance of inguinal lymphadenopathy. This necessitates a discussion on whether hemiscrotectomy should be performed upfront in massive tumors with skin fixation to improve local control.[12]

Fertility Preservation Failure

Azoospermia is a known but under-discussed complication in adolescents with large testicular masses. Mechanisms include contralateral testicular suppression via hormonal pathways, anti-sperm antibodies, or direct compression and temperature elevation in the scrotum.[19,20] The inability to bank sperm added a layer of psychological morbidity to the patient's care, underscoring the need for early reproductive counseling even in advanced disease.

Management of Catastrophic Local Recurrence

This case highlights a "therapeutic collision": the intersection of aggressive surgical oncology principles and the futility of systemic failure. The decision to perform salvage excision of the local recurrence in May 2024, despite the presence of widespread bone and lung metastases, was driven by palliative intent—to control pain, odor, and infection from the fungating mass.[21] While aggressive local control (e.g., retroperitoneal lymph node dissection) is standard in non-metastatic paratesticular RMS[22], its role in stage IV disease is less clear and must be weighed against surgical morbidity. In this case, the surgery provided temporary local hygiene but did not alter the trajectory of systemic collapse.

Chemotherapy and Toxicity

The VAC regimen remains the gold standard for RMS.[14,15] However, the toxicity profile in this patient was devastating, inducing neutropenic sepsis and exacerbating local tissue necrosis. The aggressive biology of the tumor, indicated by the rapid progression to bone metastases (uncommon in early-stage RMS)[25], suggests that standard protocols may be insufficient for high-risk adolescent embryonal RMS.

Table 1: Comparative Profile of Testicular Tumors in Adolescents.

Tumor Type	Age Peak	Tumor Markers	Primary Treatment	Prognosis (Metastatic)
Rhabdomyosarcoma	Bimodal (1-5y, 15-19y)	Negative	Orchiectomy + Chemo ± RT	Poor (20-30% 5-yr survival)
Germ Cell Tumor	15-35y	↑ AFP, ↑ βHCG, ↑ LDH	Orchiectomy + Chemo (BEP)	Good (>70-80% survival)
Stromal Tumors	Any age	Usually Negative	Orchiectomy + RPLND	Variable

4. CONCLUSION

The fatal outcome of this 17-year-old patient with testicular embryonal rhabdomyosarcoma underscores the aggressive nature of this malignancy in the adolescent population. Radical orchiectomy remains essential, but the presence of scrotal skin involvement serves as a grim prognostic marker for local recurrence. Urologists must maintain a high index of suspicion for non-GCT malignancies in marker-negative scrotal masses. The management of such cases requires a delicate balance between aggressive multimodal therapy to prolong survival and palliative surgical interventions to maintain dignity and quality of life in the face of incurable disease.

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