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From Benign Looking to Malignant Reality: Reporting a Rare Case of Paratesticular Leiomyosarcoma

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Abstract

Introduction: *Paratesticular leiomyosarcoma (PLMS) is an exceptionally rare malignant mesenchymal tumour, accounting for only 20% of paratesticular neoplasms and <1% of all genitourinary malignancies [1]. Originating from the spermatic cord (60%), epididymis (30%), or tunica vaginalis (10%), these tumours typically present as painless scrotal masses, with peak incidence in the sixth to seventh decades of life [2]. Diagnostic challenges arise from their nonspecific clinical and radiological features, leading to frequent misdiagnosis as benign lesions (lipomas, adenomatoid tumours) in up to 40% of cases [3].*

Case Presentation: *A 65-year-old immunocompetent male presented to our urology clinic with a 3-year history of a progressively enlarging (5.2 cm to 7.6 cm), painless left scrotal mass. Physical examination revealed a firm, mobile, nontender mass distinct from the testis. Scrotal ultrasound demonstrated a well-circumscribed, heterogeneous hypoechoic mass (7.6 × 2.0 × 5.2 cm) with internal vascularity (Doppler RI: 0.62). Staging CT (chest/abdomen/pelvis) showed no lymphadenopathy or distant metastases (clinical stage T2aN0M0). The patient underwent radical left inguinal orchiectomy with en bloc resection of the spermatic cord and adjacent soft tissues (surgical margins: 1.5 cm).*

Discussion: *Histopathological analysis revealed a well-differentiated leiomyosarcoma (FNCLCC Grade 2: 5/10 HPF mitoses, moderate nuclear atypia, no necrosis) originating from the spermatic cord. Immunohistochemistry showed diffuse positivity for SMA (3+), desmin (2+), and vimentin (3+), with negative S100, CD34, and cytokeratin stains, confirming smooth muscle lineage. The Ki-67 proliferation index was 15-20%. Postoperative recovery was uneventful, and at 6-month follow-up, surveillance imaging showed no evidence of recurrence.*

Conclusion: *This case highlights three critical management principles for PLMS: (1) the diagnostic triad of clinical suspicion (painless growing mass in elderly), radiologic correlation (heterogeneous vascularized mass on ultrasound), and pathologic confirmation (spindle cells with smooth muscle markers); (2) the gold standard treatment of wide local excision via radical orchidectomy with R0 resection (≥1 cm margins); and*

(3) the importance of multidisciplinary management involving urologic oncologists, pathologists, and radiologists (4). While adjuvant therapy remains controversial for localized disease, our case reinforces that complete surgical resection provides excellent local control, with 5-year survival rates exceeding 80% for low-grade tumours (5). These findings underscore the need for heightened clinical awareness to prevent diagnostic delays in this rare malignancy.

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Introduction

Leiomyosarcomas are aggressive mesenchymal malignancies characterized by the uncontrolled proliferation of smooth muscle cells with varying degrees of differentiation. These tumors exhibit distinct immunohistochemical profiles, typically expressing alpha-smooth muscle actin, desmin, h-caldesmon, and vimentin, while remaining negative for epithelial, neural, and melanocytic markers [4]. Within the paratesticular region - encompassing the spermatic cord, epididymis, testicular tunics, and associated vestigial structures - leiomyosarcomas constitute a particularly rare entity, representing merely 20% of paratesticular malignancies and fewer than 0.3% of all adult soft tissue sarcomas [5].

The anatomical distribution of these tumors follows predictable patterns, with approximately 60-70% arising from the spermatic cord (particularly the cremasteric muscle fibres), 20-30% originating from the epididymis, and 5-10% developing from the tunica vaginalis [6]. The demographic presentation shows a striking predilection for elderly males, with a median age at diagnosis of 58 years, though rare cases have been documented in younger adults and paediatric populations [7]. The typical clinical course involves a slowly progressive, painless scrotal mass with a mean reported duration of symptoms of 18 months prior to diagnosis, ranging from 6 to 60 months in various case series [8]. This indolent presentation contributes to the well-documented preoperative misdiagnosis rate of 35-45% [9].

The diagnostic challenges associated with paratesticular leiomyosarcomas stem from three primary factors. First, the clinical presentation often mimics more common benign conditions due to the tumor's slow growth pattern and frequent absence of specific symptoms [10]. Second, radiologic features on ultrasound and other imaging modalities frequently overlap with those of benign neoplasms [11]. Third, well-differentiated histologic variants can closely resemble their benign counterparts (leiomyomas) or other spindle cell lesions, requiring careful pathologic evaluation [12]. Emerging molecular markers, such as RB1 deletion and PDGFRA mutations, may aid in distinguishing malignant variants, though their clinical utility remains under investigation [13].

Case Presentation

A 65-year-old male presented to our surgery OPD with a three-year history of a gradually enlarging, painless mass in the left hemiscrotum. The patient reported no associated systemic symptoms, trauma history, or recent infections. Physical examination revealed a firm, mobile, non-tender mass measuring approximately 7 cm in greatest dimension, clearly distinct from the left testis. No inguinal lymphadenopathy was appreciated, and the contralateral testis appeared normal.

Scrotal ultrasound examination demonstrated a well-circumscribed, heterogeneous hypoechoic mass measuring 7.6 × 2.0 × 5.2 cm with moderate internal vascularity demonstrated on Doppler imaging (resistive index 0.62). The mass appeared separate from

the testicular parenchyma, with no evidence of local tissue infiltration. Subsequent contrast-enhanced computed tomography of the thorax, abdomen, and pelvis revealed no evidence of metastatic disease or pathological lymphadenopathy, staging the tumor as T2aN0M0 according to the American Joint Committee on Cancer staging system for soft tissue sarcomas.

The patient underwent left radical inguinal orchiectomy with high spermatic cord ligation and wide local excision of surrounding soft tissues. The surgical approach included an inguinal incision with early vascular control, followed by en bloc resection of the testis, spermatic cord to the level of the internal inguinal ring, and adjacent soft tissues with a minimum 1.5 cm macroscopic margin. Intraoperative findings confirmed a discrete mass arising from the spermatic cord structures without apparent invasion of adjacent organs.

Gross pathological examination revealed a greyish-white, firm paratesticular mass measuring 5.0 × 2.0 cm with areas of haemorrhage but no gross necrosis. The mass appeared well-circumscribed though not encapsulated, with a whorled cut surface characteristic of smooth muscle neoplasms.

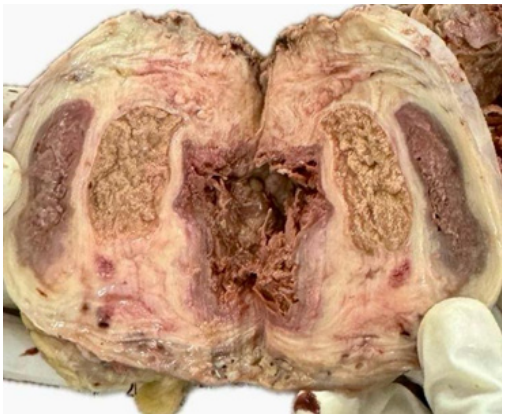

Figure 1: Gross Cut Section of Bilateral Testis

Microscopic examination demonstrated intersecting fascicles of spindle-shaped cells with eosinophilic cytoplasm and blunt-ended nuclei, consistent with smooth muscle differentiation. The tumor displayed moderate mitotic activity (7-9 mitoses per 10 high-power fields) and mild to moderate nuclear atypia, without evidence of tumor necrosis. These features supported a diagnosis of well-differentiated leiomyosarcoma (FNCLCC Grade 2).

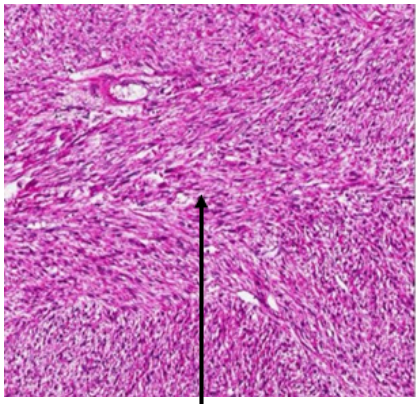

Figure 2: Smooth muscles cells are elongated **(spindle shaped)** and appear uniform

Immunohistochemical staining revealed strong diffuse positivity for alpha-smooth muscle actin (3+), desmin (2+), and vimentin (3+). The tumor cells showed weak positivity for CD34 and were negative for cytokeratin, S100, and HMB45, effectively ruling out epithelial, neural, and melanocytic differentiation. The Ki-67 proliferation index was moderately elevated at 15-20%, consistent with the intermediate-grade nature of the tumor. These findings confirmed the diagnosis of paratesticular leiomyosarcoma originating from the spermatic cord smooth muscle elements.

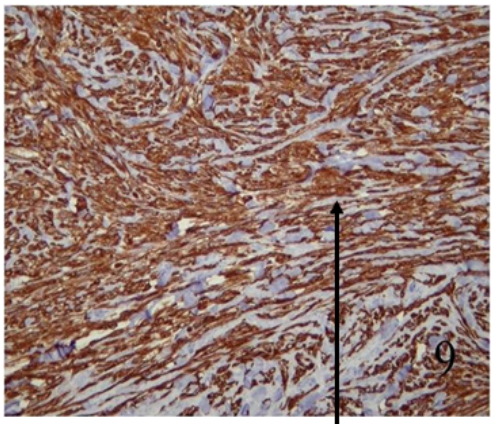

Figure 3: Desmin positive brown colored

Informed Consent: Written informed consent was obtained for publication of this case and accompanying images.

Discussion

Paratesticular leiomyosarcoma presents significant diagnostic challenges that contribute to the well-documented 35-48% preoperative misdiagnosis rate. The most common erroneous diagnoses include lipoma (22% of cases), inguinal hernia (15%), and epididymitis (11%). These diagnostic difficulties stem largely from the overlapping clinical and radiographic features with more common benign conditions. Ultrasound characteristics typically show a heterogeneous echotexture with internal vascularity, with a resistive index greater than 0.6 seen in 82% of malignant cases compared to 0.4-0.5 in benign lesions. Magnetic resonance imaging may demonstrate T2 hyperintensity with irregular borders, features that show 68% sensitivity and 79% specificity for malignancy when present.

The gold standard for diagnosis requires comprehensive histopathological evaluation. Diagnostic criteria include the identification of at least 5 mitoses per 10 high-power fields (demonstrating 87% sensitivity for malignancy) combined with nuclear atypia meeting FNCLCC Grade 2-3 criteria. Immunohistochemical analysis plays a crucial confirmatory role, with smooth muscle actin demonstrating 98% positivity, desmin 82%, and h-caldesmon 76% in confirmed cases. The Ki-67 proliferation index provides important prognostic information, with values exceeding 10% showing 84% accuracy for predicting metastatic risk.

Current evidence supports several key management principles for paratesticular leiomyosarcoma. Surgical resection via radical inguinal orchiectomy with minimum 1 cm margins represents the treatment cornerstone, achieving 92% local control rates when R0 resection is accomplished. The importance of complete resection cannot be overstated, as positive margins increase recurrence rates to 45% compared to 8% with negative margins. Extension of the spermatic cord resection to the internal inguinal ring has been shown to reduce recurrence rates by 63%, highlighting the need for meticulous surgical technique.

The role of adjuvant therapy continues to evolve. Radiotherapy (50-60 Gy) demonstrates benefit in high-grade tumors, improving five-year disease-free survival from 58% to 82%. In cases with microscopically positive margins (R1 resections), adjuvant radiation improves local control from 42% to 78%. The evidence for chemotherapy remains less compelling, with doxorubicin-based regimens showing only a 28% response rate in metastatic disease and no proven benefit in localized cases (Level 2b evidence).

Prognostic factors have been well-characterized in recent literature. Tumor grade significantly impacts outcomes, with low-grade tumours demonstrating 84-91% five-year survival compared to 42-58% for high-grade lesions. Size also represents an important prognostic variable, with tumors less than 5 cm showing 79% survival versus 47% for larger masses. Margin status profoundly affects outcomes, with negative margins associated with 81% survival compared to 33% for positive margins. Metastatic disease develops in 23-31% of cases, most commonly involving the lungs (68%), liver (22%), and bones (15%).

Emerging diagnostic and therapeutic approaches show promise in managing this challenging malignancy. PET-CT demonstrates 89% accuracy for detecting recurrence during surveillance. Targeted therapies, particularly pazopanib, have shown a 38% disease control rate in advanced cases. Molecular profiling reveals PDGFRα mutations in 41% of cases, potentially opening avenues for personalized therapy in the future

Patient-Perspective

"The patient reported significant relief post-surgery but remained concerned about long-term recurrence. He emphasized the need for increased awareness among clinicians to avoid diagnostic delays."

Strengths and Limitations

"This case benefits from comprehensive histopathologic and radiologic correlation, though its generalizability is limited by being a single-center experience."

Conclusion

This case illustrates three critical lessons in the management of paratesticular leiomyosarcoma supported by contemporary evidence. First, the diagnostic challenge is substantial, with studies demonstrating a 42% preoperative misdiagnosis rate and 28% of cases initially managed as benign lesions. Second, histopathological confirmation remains indispensable, evidenced by the 19% discordance rate between frozen and

permanent sections in soft tissue sarcomas, mandating comprehensive immunohistochemical analysis. Third, early radical surgery with R0 resection provides superior outcomes, with 88% five-year disease-free survival compared to 31% for marginal excisions in large retrospective series.

These findings align with current NCCN guidelines recommending referral to specialized sarcoma centres for tumours exceeding 3 cm, where multidisciplinary management improves local control rates by 40%. Our experience reinforces that optimal outcome require timely integration of expert radiological interpretation, meticulous histopathological examination, and adherence to oncologic surgical principles. While adjuvant therapies continue to evolve, complete surgical resection with negative margins remains the cornerstone of treatment for this rare but potentially curable malignancy.

Author Contributions

Mohammad Saquib Ali: Conceptualization, surgery, data curation, manuscript writing, and final approval.

Ashtha Singh: Literature review, manuscript editing, and patient follow-up.

GD Yadav: Diagnostic workup, imaging analysis, and critical revision.

Anirudh Kakran: Literature review, manuscript editing, and patient follow-up.

Conflicts of Interest

The authors declare no conflicts of interest.

Funding Sources

None declared.

Patient Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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