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Primary Leiomyosarcoma of the Scrotum in Bujumbura: A Case Report and Literature Review

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Abstract

Background: Leiomyosarcoma of the scrotum is an exceedingly rare malignant tumor of mesenchymal origin, typically arising from the smooth muscle of the spermatic cord, epididymis, or dartos layer. Its diagnosis remains challenging due to its clinical similarities to other intrascrotal masses. **Case Presentation:** We report the case of a 59-year-old man with a painless, progressive right scrotal mass, managed at the Bujumbura Central Polyclinic. Ultrasound revealed a well-defined, vascularized paratesticular mass. Surgical excision through right scrototomy was performed with preservation of the testis. Histological analysis confirmed a high-grade leiomyosarcoma (grade III for the tumor grading system of the National Federation of Cancer Centers (NFCC)) with negative margins. A thoraco-abdomino-pelvic (TAP) computed tomography (CT) scan performed three months postoperatively was normal. No adjuvant therapy was given. Clinical and imaging follow-up showed no recurrence or metastasis at three months. **Discussion:** Paratesticular leiomyosarcoma represents a small subset of genitourinary sarcomas. Scrotal leiomyosarcoma is particularly rare, and its clinical course and prognosis remain poorly defined. Histological confirmation is essential, and surgical excision with negative margins remains the cornerstone of treatment. Orchiectomy is the gold standard for the spermatic cord, intratesticular and epididymal forms, while lumpectomy maybe appropriate for purely scrotal tumors. The role of radio-

therapy and chemotherapy remains uncertain in non-metastatic disease. Close clinico-radiological follow-up is recommended due to the risk of recurrence. **Conclusion:** This case highlights the diagnostic and therapeutic challenges of primary scrotal leiomyosarcoma, especially in resource-limited settings. Early surgical management with histopathological confirmation is essential for favorable outcomes.

Keywords

Paratesticular Tumor, Leiomyosarcoma, Soft Tissue Sarcoma

1. Introduction

Paratesticular leiomyosarcoma is a malignant soft-tissue tumor arising from undifferentiated smooth muscle cells, most commonly originating from the spermatic cord and the epididymis [1]. It is very rare. Malignant spindle cell neoplasms have low frequency and are commonly sarcomas. Leiomyosarcoma accounts for 5% - 10% of sarcoma [2]. Genito-urinary sarcomas are exceedingly rare, representing less than 5% of all soft-tissue sarcomas [3]. Leiomyosarcoma (LMS) is the second most common histological subtype of intrascrotal sarcoma after liposarcoma, accounting for 20% of cases [4]. LMS is a malignant mesenchymal tumor arising from smooth muscles; Over 75% of intrascrotal LMS arise from the spermatic cord [5]. Leiomyosarcomas (LMS) of the testes can be classified into paratesticular (about 217 reported cases) and intratesticular. Up to 80% of paratesticular LMS arise from the soft tissue of the spermatic cord, whereas 20% originate from the epididymis or darts of the scrotum [6]. LMS are further classified as cutaneous LMS that derive from the arrector pili muscle of the hair follicle or the dartos muscle of the genital skin and subcutaneous LMS that originates from the smooth muscle of the genital organ or the vascular muscle layer of subcutaneous tissue [7] [8]. Leiomyosarcoma of the testis is an uncommon tumor which may develop after radiotherapy, chronic inflammation, or anabolic steroids use for long time. However, in the absence of risk factors, it is rarely observed [9]. The diagnosis is always based on histological examination. Complete and radical local excision with negative margins is crucial for optimal outcomes [8]. We describe the case of a patient presenting with a primary leiomyosarcoma of the right scrotum without notable risk factors managed at the Bujumbura Central Polyclinic with preservation of the homolateral testis.

2. Case Presentation

A 59-year-old man presented with a painless right scrotal swelling first noticed in January 2024. The mass had enlarged progressively, particularly from October 2024, prompting the patient to consult. He denied scrotal pain, redness, dysuria, or urethral discharge. Family history was unremarkable. On physical examina-

tion, a visible right scrotal mass was observed above the testis. It was firm, non-transilluminable, independent of the testis, and no inguinal lymphadenopathy was present. The left scrotum was normal and the rest of the physical examination was unremarkable.

Laboratory tests, including complete blood count, renal function, urinalysis and Prostate-specific antigen (PSA), were normal. Tumor markers, including beta-human chorionic gonadotropin (β-hCG), alpha-fetoprotein (AFP) and lactate dehydrogenase (LDH) were within normal limits. Ultrasound revealed a heterogeneous right paratesticular mass, vascularized on Doppler with a capsule, measuring 5.1 × 5.7 × 6.5 cm, or 100 cc. The right testis was normal and well vascularized; the right epididymis was slightly hypoechoic and enlarged (12 mm at the head) (**Figure 1**). Right inguinal lymphadenopathy with irregular borders was noted. The left testis and epididymis were normal.

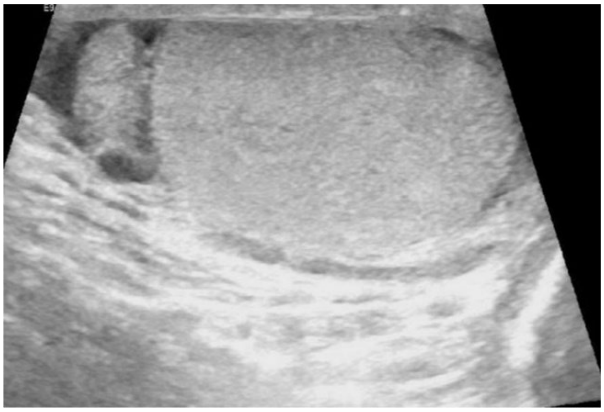
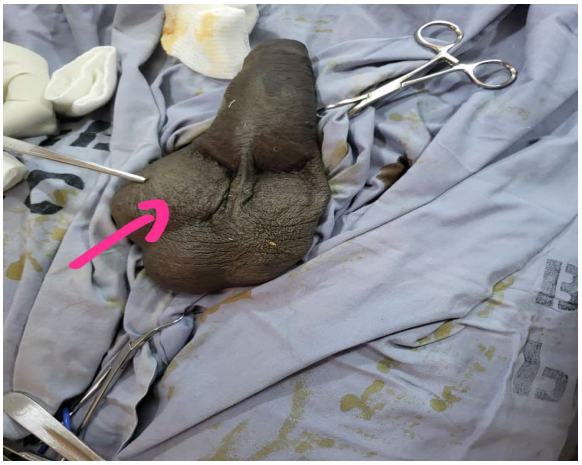


Figure 1. Scrotal ultrasound demonstrating right paratesticular mass.

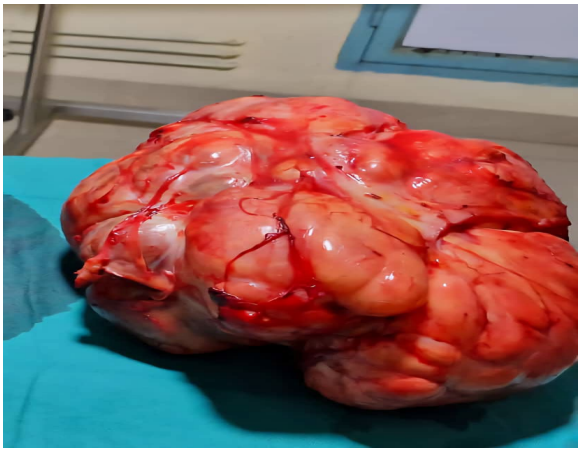
In May 2025, the patient underwent right lumpectomy via transverse scrototomy. Operative findings revealed a well-defined, solid paratesticular mass above a normal-sized testis, without vascular compromise. The mass was excised completely while preserving the testis and spermatic cord (**Figure 2(a)** and **Figure 2(b)**). This finding, along with the absence of local spread, warranted the testis-sparing approach, which was crucial to preserving function and avoiding a radical orchidectomy.

The post-operative course was straightforward, and the patient was discharged the day after surgery. At one-month follow-up, the wound was healed and the patient was asymptomatic.

Macroscopic examination of the specimen revealed a whitish tumor-like tissue with necrotic-hemorrhagic changes weighing 107 g and measuring 6 × 4 × 4 cm. Histopathology confirmed a high-grade leiomyosarcoma (NFCC grade III) with negative margins and no lymphovascular invasion. Immunohistochemistry was not performed. A thoraco-abdomino-pelvic CT scan, performed three months after surgery, revealed no evidence of metastatic disease or adenopathy. The patient did not receive adjuvant radiotherapy or chemotherapy.



(a)



(b)

Figure 2. (a) and (b) right suprascrotal swelling (arrow) seen on a preoperative image located above the right testis (3). Image of the mass after excision with clear margins (4).

3. Discussion

Paratesticular tumours account for less than 5% of intra-scrotal tumours, and approximately 30% are malignant, with sarcomas representing the majority [10]. Paratesticular tissues are derived from a combination of epithelial, mesothelial and mesenchymal cells and as a result the tumours form a heterogenous group which exhibit a broad range of behaviours and histological appearances [11] [12]. Around 30% of paratesticular tumours are malignant with sarcomas accounting for approximately 90%, however still comprising less than 5% of all sarcomas and 2% of urological malignancies [12]. Leiomyosarcoma is a rare disease entity not often encountered in the genito-urinary system. Leiomyosarcomas are the third smost common malignant tumors of soft-tissue (sarcoma) after malignant testicular fibrous histiocytoma and liposarcoma. Leiomyosarcomas arise from undifferentiated smooth muscle cells of mesenchymal origin. However, almost all testicular leiomyosarcomas are in fact paratesticular, originating from the spermatic cord, epididymis or scrotum [13]-[15]. Leiomyosarcoma of the scrotal wall is rare,

and its clinical significance and prognosis have not been well defined [16]. Leiomyosarcomas have three typical histological features: perpendicularly arranged fascicles of spindle cells with eosinophilic cytoplasm, hyperchromatic blunt-ended nuclei, and scattered paranuclear vacuoles. Immunohistochemical staining shows an expression of SMA, muscle-specific actin, and desmin in most leiomyosarcomas, and expression of S-100 protein, CD34, Ki-67, myogenin, and cytokeratin has also been reported in some cases [12]. They can present a diagnostic challenge as they are often clinically and radiologically indistinguishable from a testicular mass. When tumours are paratesticular, they most commonly arise from the spermatic cord where they originate from cremasteric muscle and the vas deferens. Less frequently, the epididymis is the origin, arising from the smooth muscle surrounding the basement membrane of the epididymis canal. The scrotal form occurs least frequently, from the dartos layer. This case demonstrates the most rare leiomyosarcoma subtype arising separately from the tissues highlighted above [10]. Clinical presentation is commonly as a painless, slowly enlarging mass with a discrete, nodular feel and may be associated with a hydrocele [17]. Age at presentation is most commonly between 50 and 70 years [18]. Our patient belongs to this category. Etiology of paratesticular LMS remains unclear though some authors have suggested that exposure to childhood radiation is a risk factor.

Ultrasound is the first-line imaging modality [19] and generally reveals a solid, heterogeneous, vascularized mass [20]; However, the accuracy of the scan is operator-dependent; in this case the patient required a repeat scan to confirm that the mass was paratesticular. Computed tomography can be used to assess the mass but is of most value of assessing for metastatic disease [15].

The diagnosis is made based on typical histological features of leiomyosarcoma, including spindle cells arranged in fascicular fashion, eosinophilic cytoplasm, blunt ended nuclei, cellular atypia and variable mitotic activity [21]. Immunohistochemical staining is frequently positive for smooth muscle actin and desmin and there may be expression of CD-34 and cytokeratin. Therefore, the tumor was classified as a grade III tumor using the NFCC grading system. Grading of the tumor based on histology is important to determine prognosis. High-grade tumors are more aggressive [1]. The lack of immunohistochemical analysis represents a notable diagnostic limitation in this case. Markers such as smooth muscle actin (SMA) and desmin would have provided definitive confirmation of the smooth muscle origin of the tumor, thereby ensuring greater diagnostic precision. Radical orchidectomy with high ligation of the spermatic cord is the standard treatment for these tumors [1] [22] [23].

For our patient, we opted for complete excision of the mass with preservation of the ipsilateral testis, as the mass was well-defined and the spermatic cord and testicular tissue could be preserved. High-grade tumors, such as the one observed in this case, are associated with a significantly higher risk of local recurrence and distant metastasis. The NFCC grading system classifies this tumor as grade III, reinforcing the need for close and rigorous clinical and radiological follow-up

post-surgery to monitor for potential recurrence or metastasis.

There is insufficient data on prophylactic lymph node dissection for relapse prevention. Although there are studies that may suggest that adjuvant radiotherapy may be effective in treating loco-regional microscopic disease, it has not shown survival benefit. Chemotherapy is controversial and has only been used in the case of metastatic disease [1] [5]. Although there is limited data about this tumor, some literature mentioned that five and 10-year disease-specific survival rates are 77% and 66% respectively [24]. Adjuvant therapies such as radiotherapy and chemotherapy are not commonly used in the absence of metastasis, recent guidelines on soft tissue sarcomas suggest that these therapies may be considered in high-risk cases to address potential microscopic disease. However, there is currently insufficient evidence to recommend routine adjuvant therapy for localized, non-metastatic paratesticular leiomyosarcoma. Regardless of this relatively good long-term survival, because recurrences are common, close follow-up is suggested for disease surveillance.

4. Conclusion

Paratesticular leiomyosarcoma is rare, with few cases of scrotal origin reported. Due to its rarity and nonspecific presentation, diagnosis is often delayed until histology. Surgical excision with negative margins remains the cornerstone of management. While orchiectomy is standard for spermatic cord and intratesticular forms, scrotal forms may be safely managed with lumpectomy. Adjuvant radiotherapy and chemotherapy are not routinely recommended. Vigilant clinical and radiological follow-up is essential given the risk of recurrence.

Author Contribution

RN contributed to study concept and design; SN drafted the manuscript. APN, JLB, DM, MAK, AM, SK, JCM & PB contributed to reviewing and finalizing the manuscript. All authors reviewed the manuscript for intellectual content and approved the submission.

Conflicts of Interest

We declare that we have no conflicts of interest.

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