

CASE REPORT

A rare case report of paratesticular leiomyosarcoma

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Abstract

A leiomyosarcoma (LMS) is a malignant mesenchymal tumor of soft tissue that rarely presents in the scrotal region. This case report describes a primary paratesticular LMS originating in the dartos layer of the scrotum in a symptomatic 37-year-old male with a history of hypertension, generalized seizure disorder post-excision of meningioma, and an extensive family history of cancer. Primary paratesticular LMSs in the superficial dartos layer are exceedingly rare. Given the rarity of this particular type of LMS, this case report includes a discussion on its prevalence and incidence. The rarity of this tumor also warrants a review of the literature and related cases to inform the treatment plan and enhance understanding of these oncologic conditions.

Keywords: Paratesticular leiomyosarcoma; Testicular leiomyosarcoma; Paratesticular cancer; Testicular cancer; Leiomyosarcoma; Sarcoma

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1. Background

Testicular tumors are a common cause of cancer in young males, typically occurring between the ages of 15 – 45, and they are generally highly curable. In the United States, the mean age at diagnosis for testicular cancer is reported to be around 33 years.^{1,2} Among the various subtypes of testicular cancer, leiomyosarcomas (LMSs) are exceedingly rare, accounting for <1% of case.³

LMS is a type of cancer originating from smooth muscles or their precursor mesenchymal stem cells that can develop virtually anywhere in the body. It is a subtype of soft-tissue sarcoma, which arises from mesenchymal tissues derived from the embryonic mesoderm. In the context of the male genital system, testicular LMS can be classified into two types: paratesticular and intratesticular, with paratesticular LMS being the more common tumor. The term “paratesticular” encompasses structures within the scrotum that are not of testicular origin,^{4,5} including the epididymis, spermatic cord, tunica vaginalis, and associated supporting structures. Due to the complex anatomy and variable locations of these structures, LMS in the paratesticular region can present with a wide range of malignancies. The majority of male genitourinary sarcomas originate in the spermatic cord. The peak incidence of LMS occurs in men between the ages of 50 – 60 years and often presents with nonspecific symptoms.^{4,5} This cancer can metastasize through local extension, hematogenous routes, and lymphatic spread, with local spread being the most common.^{6,7} Hematogenous metastases commonly involve

the liver and lungs, while lymphatic spread affects the external iliac, hypogastric, para-aortic, and common iliac lymph nodes.^{6,7} Long-term follow-up is essential because, despite a low risk of recurrence within the first 5 years post-treatment, there is a documented risk of relapse up to 15 years later.^{7,8}

In this case report, we document the unique medical journey of a 37-year-old male who presented with a 2-year history of a gradually growing mass, which was determined to be LMS originating from the dartos layer of the scrotum. This rare form of cancer affecting the testicular tissue is not widely documented in medical literature. The report delves into the scarcity of case studies related to testicular LMS, providing an in-depth exploration of existing reports. Furthermore, it comprehensively reviews available treatment options and seeks to uncover any discernible patterns in the presentation of this uncommon condition.

2. Case presentation

A 37-year-old male with a medical history of hypertension, a personal history of meningioma (with post-excision seizure disorder), and an extensive familial history of cancer (including breast, pancreatic, esophageal, lung, and parotid gland cancers) presented with a scrotal mass. Initially, the mass was non-tender, and an ultrasound revealed a hypoechoic, heterogeneous, and slightly lobulated mass measuring $2.0 \times 1.8 \times 1.0$ cm between the testes. At that time, the mass was diagnosed as a hydrocele by a urologist, and the patient was subsequently lost to follow-up. Two years later, the patient returned to the clinic, reporting that the scrotal mass had increased in size and was now causing pain. He was then scheduled for a wide excision of the right scrotal wall.

During surgery, a $7.4 \times 5.2 \times 4.5$ cm lobulated, pink-tan, and firm mass was excised. The procedure was complicated by a post-operative hematoma, which was subsequently evacuated. On sectioning, the mass appeared solid, white, and whorled, with central pink-tan softened foci. Immunohistochemical staining revealed that the tumor was strongly positive for h-caldesmon and smooth muscle actin, weakly positive for CAM5.2, desmin, and epithelial membrane antigen, and negative for p63, S100, and SOX-100. Histological analysis confirmed that the mass was a grade 2/high-grade LMS of the dartos layer of the scrotum (Figures 1 and 2), presented with ten mitoses/mm², 10% necrosis, and tumor presence at the surgical margin, indicating the necessity for further surgical intervention. Based on these findings, the patient was formally diagnosed with LMS of the paratesticular region.

Due to the rare nature of this diagnosis, the patient was referred to a sarcoma center of excellence for expert

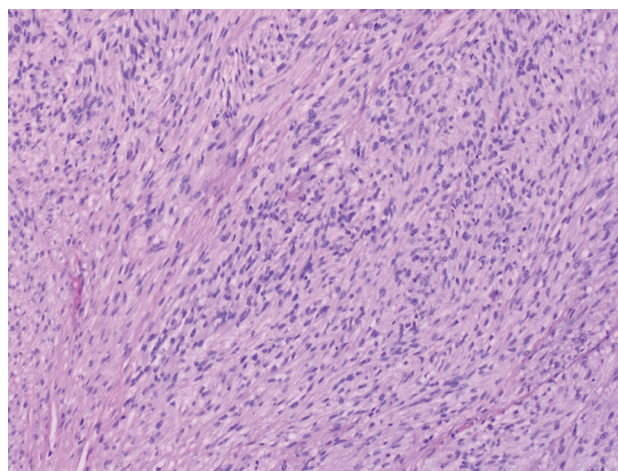


Figure 1. Low magnification of spindle cells characterizing leiomyosarcoma. Note: Magnification: $\times 10$.

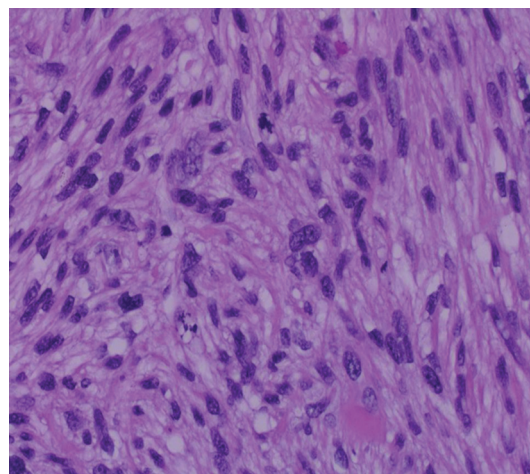


Figure 2. High magnification of spindle cells characterizing leiomyosarcoma in their mitotic phase. Note: Magnification: $\times 40$.

opinion and potential clinical trial recommendations. Following consultation, staging computed tomography (CT) scans were performed, which revealed no evidence of metastatic disease. A next-generation sequencing panel of 648 genes was conducted and did not identify any targetable mutations. Given the positive margins from the initial resection (as stated earlier), the patient underwent a wide local re-excision with intraoperative frozen sections, which were negative for residual disease. The patient is currently being followed for surveillance with CT scans of the chest, abdomen, and pelvis every 3 months. He has been doing well since the procedure.

3. Discussion

The present case report describes a primary LMS in the paratesticular region. Paratesticular sarcomas are exceedingly rare, occurring at a rate of $<5\%$ among all

sarcomas.⁹ According to the Memorial Sloan Kettering Center's adult sarcoma database, only 3% of sarcomas originate from the genitourinary system, with 44% of these being LMSs.¹⁰ Further, contributing to the rarity of this case is the location of the LMS within the dartos layer of the scrotum, which accounts for only 2% of paratesticular LMS cases.^{11,12} In general, paratesticular sarcomas are found in the epididymis and spermatic cord.^{4,5,13,14} The vast majority of paratesticular masses are benign, including lipomas, adenomatoid tumors, and leiomyomas.¹⁴ Malignant masses, which are less common, include liposarcomas, rhabdomyosarcomas, and LMSs, with LMSs being the most frequent.¹⁰

Another factor that adds to the rarity of this case is the age of the patient. At 37 years old, the patient is considerably younger than the typical age of diagnosis for this type of cancer. Population-based analyses of genitourinary sarcomas demonstrate that the median age of diagnosis for paratesticular and scrotal sarcomas is between 50 and 60 years.^{4,13,15} A review of additional case reports reveals that most patients with paratesticular LMS are diagnosed between their 20s and 80s, with a higher prevalence in males older than 50 years.^{2,7,8,16-19}

Based on our case and others reported worldwide (identified using MeSH terms such as "leiomyosarcoma," "testicular leiomyosarcoma," "paratesticular leiomyosarcoma," "testicular cancer," "leiomyosarcoma," "sarcoma," "testicular neoplasm," and "orchiectomy"), testicular LMSs appear to have a lengthy indolent phase followed by a growth phase. In our patient, the indolent phase lasted nearly 2 years, during which the mass was initially thought to be a hydrocele. It was only when the mass began to increase in size and cause pain a few months prior that the patient returned to the clinic. Other cases of testicular LMS in the literature appear to follow a similar pattern: an initial hydrocele presentation, followed by an indolent phase, and then a growth phase that prompts the patient's follow-up visit.^{4,13,20,21} The growth of these testicular masses should prompt clinicians to conduct further investigations to assess whether the mass is benign or malignant.

The initial diagnostic approach recommended for further investigation, and the approach used in this case is ultrasound. Ultrasound has been described as the most efficacious diagnostic approach for the initial identification of testicular LMS and other cord/scrotal abnormalities.^{22,23} It has a sensitivity of 95 – 100% in distinguishing between testicular and paratesticular lesions.²² On ultrasound, testicular LMSs typically appear as hypoechoic masses, though echogenicity may vary.^{24,25} Ultrasound may also help distinguish between a benign leiomyoma and a malignant LMS, as benign leiomyomas are generally

located in the inguinal part of the spermatic cord, whereas LMSs are usually found in the paratesticular region near the scrotal skin.^{24,25} While some have recommended magnetic resonance imaging and CT imaging for staging and aiding in diagnosis, histology remains essential for a definitive diagnosis, making these imaging modalities not strictly necessary.^{21,24-26} A thorough clinical examination should be performed in combination with a testicular ultrasound to rule out other differential diagnoses for scrotal enlargement, such as simple hydrocele, complex hydrocele, inguinal hernia, epididymal cysts, spermatoceles, and chronic epididymitis.²³⁻²⁶ It is also important to perform CT scans of the chest, abdomen, and pelvis to rule out metastatic sites. If CT scans reveal lymphadenopathy, it may change the surgical approach and impact prognostic indicators. LMS is known to spread in three ways: locally, through the bloodstream to the liver and lungs, and through the lymphatic system to external iliac, hypogastric, para-aortic, and common iliac lymph nodes. Of these modes of spread, local spread is, by far, the most common.^{6,7}

The information obtained from the ultrasound, such as mass consistency and location, helps guide future surgical intervention and assess the potential for malignancy.⁹ However, it is nearly impossible to definitively determine the type of tumor present solely through ultrasound; thus, a biopsy is required. The treatment of choice and definitive diagnostic tool for all testicular masses, particularly sarcomas, is complete resection with histologically negative margins.¹⁰ The vast majority of documented paratesticular sarcomas are resectable.¹² According to the literature, the standard treatment is radical orchiectomy with high ligation of the spermatic cord. Due to anatomical constraints, local recurrence rates are reported to be between 30% and 50%. If the scrotal skin is involved, hemiscrotoectomy is recommended.^{2,7,8,16-19,28,29} In our case, the patient underwent a right scrotal wide wall excision, followed by a subsequent repeat surgery with intraoperative freezing (to ensure negative margins), which has proven sufficient thus far.^{27,28} The role of adjuvant therapy has not been rigorously studied, but factors such as histologic grade, tumor size, and positive margins are utilized to determine when adjuvant therapy is appropriate. In patients with narrow margins on repeat excision, radiation therapy is recommended; however, the involvement of radiation and chemotherapy is determined on a case-by-case basis.¹⁰ In a study involving 21 patients, 5-year disease-free survival rate was found to be 58% with surgery alone and 100% with the addition of radiation therapy. A separate study with 18 patients found similar results. While chemotherapy remains controversial, a case study reported no recurrence or metastasis following nine cycles of ifosfamide and adriamycin.^{2,7,8,16-19}

A definitive diagnosis of testicular LMS can only be made through histological analysis following excision. Histological examination of testicular LMS typically reveals spindle-shaped smooth muscle cells with eosinophilic cytoplasm, hyperchromatic blunt-ended nuclei, and scattered paranuclear vacuoles.^{20,30} This examination is followed by immunohistochemical studies, which, in sarcomas, generally show positivity for smooth muscle cell markers such as h-caldesmon, desmin, muscle-specific actin, and smooth muscle actin, and tend to be negative for S100 and vimentin, although some tumors may express these markers as well.^{7,17,20,31} A common feature of most genitourinary sarcomas is their tendency to present at a high histologic grade.¹⁰ In the Memorial Sloan Kettering Cancer Center report, 86% of the cases were found to be high grade.^{10,32}

In addition to histologic and immunohistochemical analyses, characteristics of the gross tumor, such as size and location, can assist in determining prognosis. Current reports suggest that tumors measuring 7 cm or larger are associated with a higher risk of being classified as stage 2 or higher, which, in turn, increases the risk of metastasis. However, a case report of an 88-year-old male with a history of prostate adenocarcinoma, who presented with tender swelling of the testicle and a 3.5 × 2.6 cm paratesticular mass that showed no recurrence, indicates that a history of cancer in the genitourinary region does not necessarily predispose one to a higher risk of recurrence.^{2,7,8,17-19} The Memorial Sloan Kettering Cancer Center report found that 56% of histologically high-grade genitourinary sarcomas were larger than 5 cm.^{10,32} There does not appear to be a predilection for one testicle over the other, and tumor sizes at presentation range from 2 – 20 cm.^{2,7,8,16-19} Despite this information on gross tumor characteristics, the present lack of comprehensive surveillance data on testicular LMS incidence makes it difficult to extrapolate grading/staging of the tumor to reliable 5-year overall survival rates.

The rarity of testicular LMS also means the risk factors are not well understood. LMS has been linked with various genetic syndromes, such as hereditary retinoblastoma and Li-Fraumeni syndrome. While no definitive causative factor has been identified, previous radiotherapy, trauma, chronic inflammation, anabolic steroid use, Epstein-Barr virus infection, and previous organ transplantation are speculated to be potential risk factors.³³⁻³⁶ In our case, the patient did not exhibit any of these risk factors. Although our patient had a personal history of meningioma, it was treated solely with excision and did not involve radiotherapy.

This rarity has also created uncertainty regarding appropriate follow-up schedules. Due to the unknown nature of testicular LMSs, a strict follow-up regimen

should be maintained. Some studies have recommended semiannual CT scans in addition to physical examinations of the groin region, including scrotal and inguinal palpation.^{23,37,38} However, due to the multitude of known and unknown factors contributing to the recurrence rate, no definitive follow-up guidelines have been established.

4. Conclusion

This case report details the course of care for a 37-year-old male with primary testicular LMS of the dartos layer of the scrotum, accompanied by a brief literature review discussing similar cases. Primary testicular LMSs in the dartos layer of the scrotum are exceedingly rare, particularly at the patient's young age of 37, and are, therefore, not well documented in the literature. As a result, the incidence and prevalence of testicular cancers, sarcomas, and LMSs were discussed at length. This was followed by a discussion on the clinical presentation, risk factors, diagnosis, treatment, and prognosis of testicular LMS based on our case and other cases reported in the literature. Although case reports and literature reviews on the topic exist, further comprehensive analyses are necessary to develop a definitive understanding of ideal diagnostic protocols, treatment guidelines, recurrence rates, and prognosis.

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Conflict of interest

The authors declare that they have no competing interests.

Author contributions

Conceptualization: All authors

Investigation: Santosh Nimkar, Bianca Glass

Methodology: Santosh Nimkar

Writing – original draft: Santosh Nimkar

Writing – review & editing: All authors

Ethics approval and consent to participate

Not applicable.

Consent for publication

Verbal consent for publication has been obtained from the patient. In addition, all identifying information of the patient was concealed.

Availability of data

Not applicable.

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