Case Report

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Recurrent paratesticular liposarcoma: a case report

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Introduction and importance: Paratesticular liposarcoma (PTL) is a rare condition, with fewer than 200 cases reported worldwide. It is a malignant tumor that originates from fat tissue with high risk of recurrence. Herein, the authors present a contralateral recurrence of a treated PTL. To the best of the authors' knowledge, in the current literature, there are few cases reported with recurrent PTL.

Case presentation: The authors report the case of a 62-year-old man who presented with a rapidly growing painless right hemiscrotal swelling. Clinical and radiographic evidence suggested the presence of two paratesticular tumors. The patient underwent a radical orchidectomy with resection of the two tumors through an inguinal approach. The histologic examination revealed a sclerosing, well-differentiated liposarcoma. The decision of the multidisciplinary consultation meeting was not to do adjuvant treatment. A follow-up of 12 months showed recurrence of the contralateral scrotum revealed by an FDG-PET/scan.

Clinical discussion: PTL, a rare spermatic cord tumor, affects adults aged 50–60, often presenting with scrotal swelling. Diagnosis involves ultrasound, computed tomography, and magnetic resonance imaging. Surgical intervention, including radical orchiectomy and adjuvant radiotherapy, is common for management, while the role of chemotherapy is inconclusive. High-grade subtypes carry a higher recurrence risk.

Conclusion: PTL is often misdiagnosed preoperatively. It is typically managed through radical orchidectomy, which includes wide excision and high ligation to ensure free surgical margins and avoid recurrence. The role of adjuvant therapy remains debatable. Despite a generally favorable prognosis, long-term follow-up is crucial because of the elevated risk of recurrence.

Keywords: paratesticular liposarcoma, rare tumor, recurrence, scrotal masses

Introduction

Paratesticular sarcomas are tumors of mesenchymal origin that may arise from the spermatic cord, inguinal canal, testicular tunic, or epididymis. The incidence of soft tissue sarcoma is 5 per 100 000 per year, 5% of which affects the genitourinary tract and accounts for ~2% of all genitourinary malignancies[1,2]. Liposarcoma primarily affects adults, typically peaking between the ages of 40 and 60, with a slightly higher prevalence in men[3].

Liposarcoma of paratesticular tissues is a rare tumor that represents 3–7% of all liposarcomas[4] and 7–10% of all intrascrotal tumors[2]. The majority of intrascrotal liposarcomas arise from the spermatic cord (76%), with fewer cases originating from the testicular tunics (20%) and epididymis (4%). However, accurately determining the precise origin poses challenges due to the size of the tumor and its degree of adhesion[3].

Well-differentiated liposarcomas (WDLs) and dedifferentiated liposarcomas (DDLs) are the predominant histological variations, constituting more than 60% of all liposarcoma cases[5].

It was first reported in 1845, and ~200 cases have been reported to date in English literature[6].

We report a case of paratesticular liposarcoma (PTL) in which a patient underwent radical treatment and subsequently developed a recurrent contralateral tumor. This case report has been reported in line with SCARE 2023 Criteria[7].

HIGHLIGHTS

- Paratesticular liposarcoma is a rare condition.
- The therapeutic approach for this pathology is mainly surgery.
- The role of adjuvant therapy remains debatable.
- Long-term follow-up is crucial because of the elevated likelihood of recurrence.

Case presentation

A 62-year-old man with a history of hypertension presented with a painless right hemiscrotal swelling that had significantly increased in size over the previous year.

The patient had no prior history of local trauma and no significant findings were documented in the past medical and surgical records.

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Physical examination

On local examination, his right scrotum was enlarged with a negative transillumination test. Palpation revealed a unilateral, large, firm, painless, mobile right hemiscrotal mass measuring ~6 × 4 cm. It had poorly defined borders and was inseparable from the spermatic cord and the right testis. The left testicle and epididymis were palpable without any detectable mass. No palpable lymphadenopathy was noted.

Laboratory examination

The results of routine serum and urine laboratory investigations, including the concentrations of beta-human chorionic gonadotropin (beta-hCG), lactate dehydrogenase (LDH), and carcinoembryonic antigen (CEA), were all within the normal ranges.

Imaging examination

Scrotal ultrasound showed a 6-cm heterogeneous bilobed lesion in the right scrotum. Scrotal MRI showed two oval, well-encapsulated paratesticular masses, each measuring 4 cm in low signal intensity on T1, high signal intensity on T2, high signal intensity on diffusion-weighted imaging (DWI) with a low apparent diffusion coefficient (ADC), and low enhancement (Fig. 1). We suspected this to be a remodeled lipoma or a teratoma.

Treatment

Under general anesthesia, the patient underwent a radical right orchidectomy through an inguinal approach. The exploration revealed two cord-dependent masses, distinct from the testis. The spermatic cord was dissociated up to the inner ring and sutured. The two masses as well as the right testicle and all right inguinal canal contents up to the deep inguinal ring were resected. The two tumors, each measuring 6 cm, presented as soft yellow-gray and well-encapsulated masses connected to the spermatic cord (Fig. 2).

Pathological analysis

The macroscopic examination revealed a piece of orchidectomy measuring 18 × 10.5 × 2 cm comprising a testicle measuring 7.5 × 7.5 × 1 cm, covered with albuginea. The epididymis measured 5 × 1.5 cm. The spermatic cord measured 9 × 3 × 1 cm and extended into two masses of similar appearance, finely encapsulated, with a bilobed surface. Both measured 6 × 5 × 2 cm and were connected to the spermatic cord. On the cut surface, they were yellowish containing white septal bands. No necrotic or hemorrhagic foci were seen. On histopathological examination, the two masses consisted of a tumor proliferation made up of lobules of mature adipocytes, with variable size, separated through thick sclera-hyaline fibrous partitions comprising small, scattered spindle-shaped cells with atypical mitotic nuclei. The mitotic index was estimated at 2 mitoses/10 high-power fields. Multivacuolated lipoblasts with hyperchromatic nuclei were also seen. The stroma was dense containing abundant strands of collagen and an inflammatory infiltrate of moderate abundance associating lymphocytes and eosinophils. This proliferation also included focal areas of cartilaginous differentiation. The tumor was surrounded by a thin fibrous capsule focally showing a break-in and vascular invasion. The diagnosis of sclerosing WDL was established (Fig. 3).

Outcome and follow-up

The patient's postoperative course was uneventful. No complications following surgery were reported. The patient was discharged on the third postoperative day, with no need for additional adjuvant therapy.

Follow-up with a CT scan of the thorax, abdomen, and pelvis after 6 months was recommended to detect any local recurrence or metastasis. There were no signs of local recurrence or distant metastasis.

However, the FDG-PET/scan performed after 6 months revealed a focal hypermetabolic area in the left scrotal region with a maximum standardized uptake value (SUV max) of 7.75 (Fig. 4).

After a multidisciplinary meeting, we recommended that the patient undergo additional radiotherapy; however, the patient refused treatment. Therefore, we decided to closely monitor the patient's evolution. As of the latest examination, conducted a week prior to manuscript submission and more than 18 months

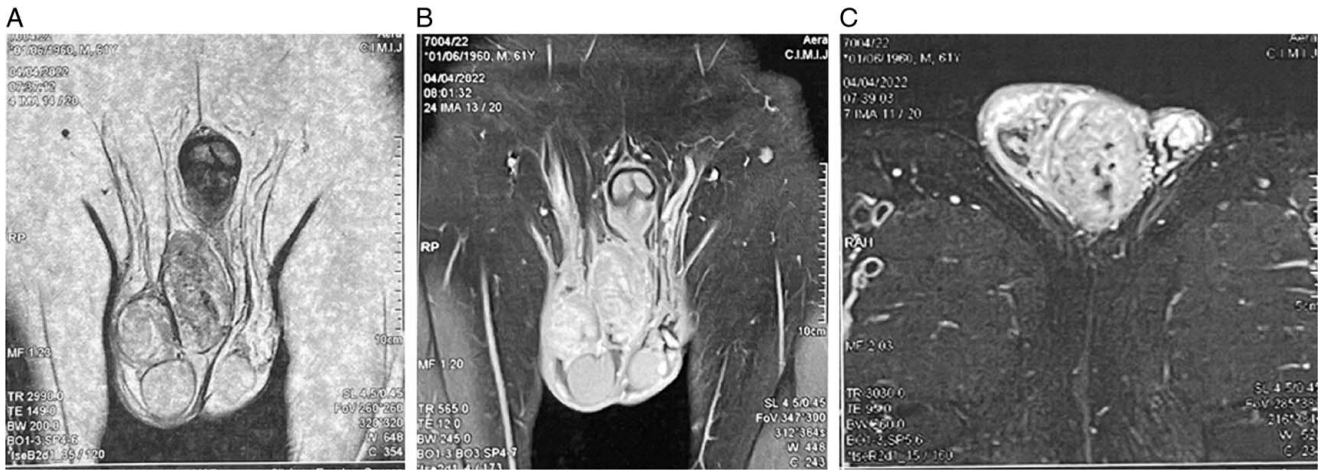

Figure 1. MRI imaging of the scrotum. (A) Coronal T2-weighted sequence. (B) Coronal T1-weighted fat saturated post gadolinium sequence. (C) Axial T2-weighted fat saturated sequence.

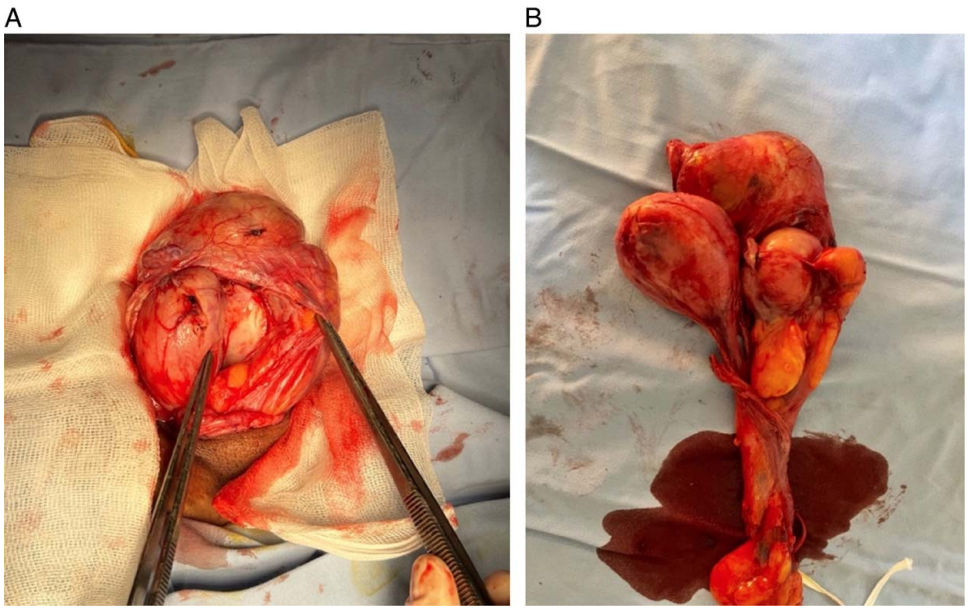

Figure 2. (A) Surgical exploration of the paratesticular tumor. (B) En bloc resection of the right testis and the two tumors.

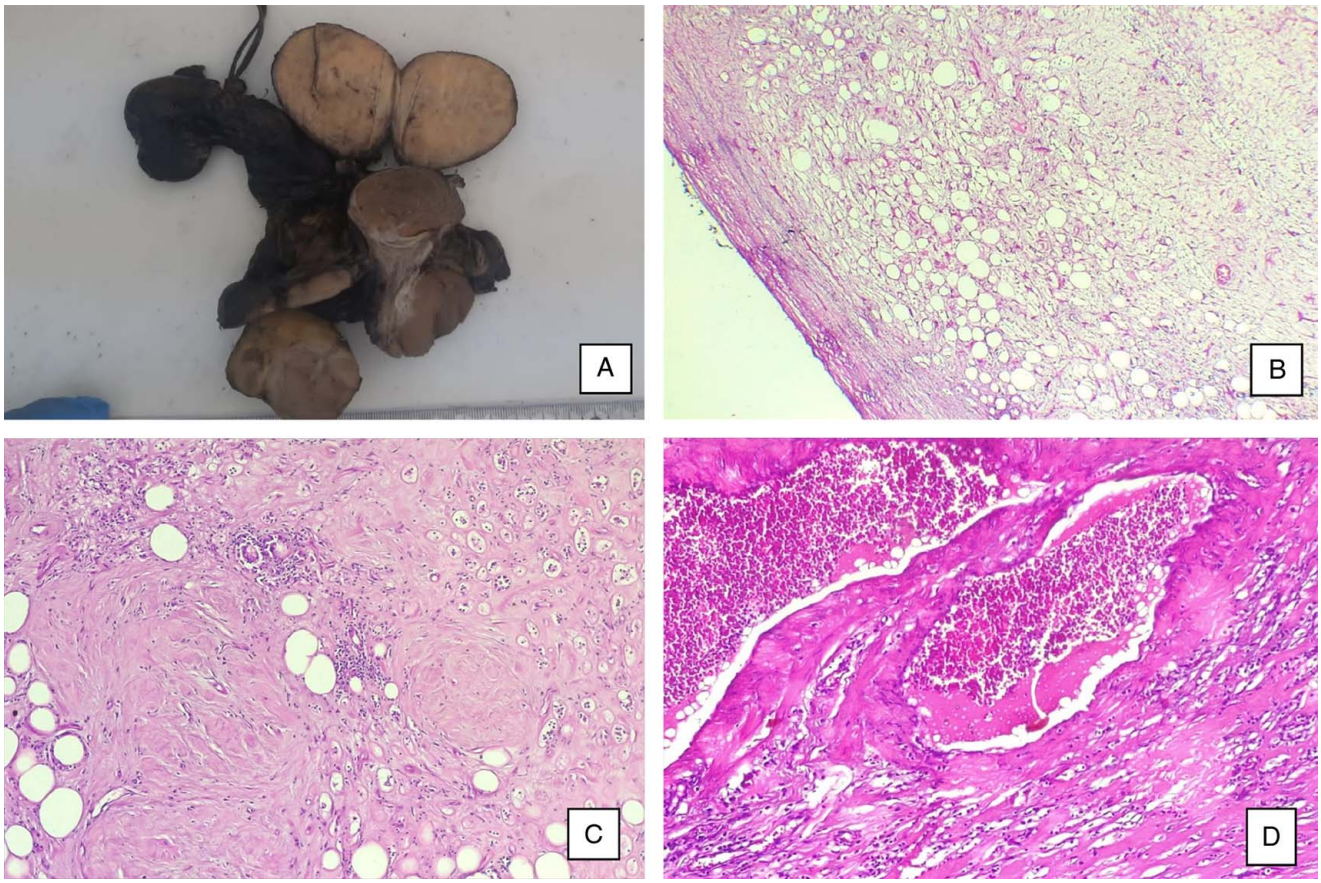

Figure 3. Histopathological examination of a paratesticular sclerosing well-differentiated liposarcoma. (A) Cut section of the piece of orchidectomy showing two masses with a yellowish appearance. No necrotic or hemorrhagic foci are noted. (B) Microscopic examination of the mass revealing an encapsulated proliferation consisting of mature adipocytes with variable size accompanied by spindle-shaped cells (hematoxylin and eosin, HE X40). (C) Dense abundant collagenous stroma with focal areas of cartilaginous differentiation (HE X100). (D) Blood vessels invaded by the tumor (HE X100).

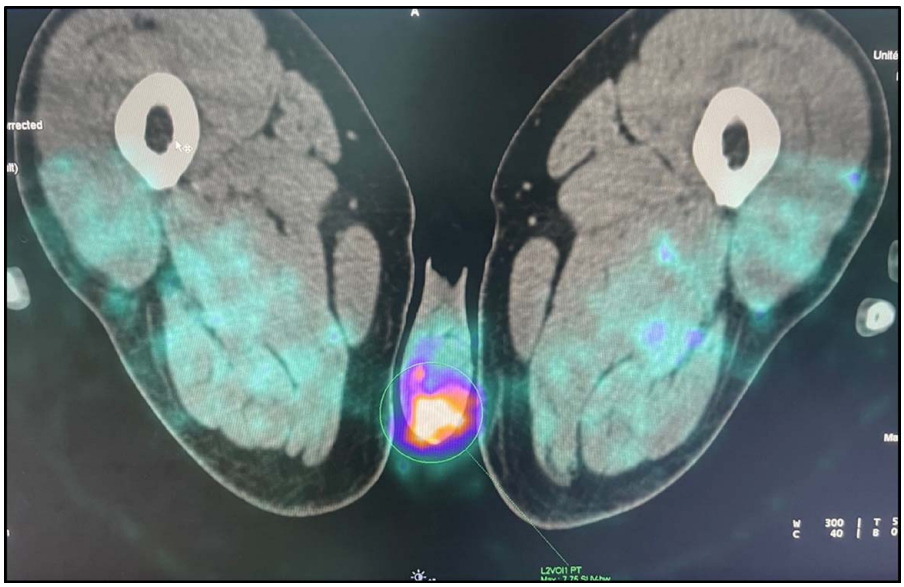

Figure 4. Staging fluorodeoxyglucose positron emission tomography findings: focal hypermetabolic area in the left hemiscrotum.

after the surgery, there has been no clinical recurrence, and CT scans revealed no regional soft tissue shadows. Ongoing monitoring and follow-up assessments are currently being conducted to evaluate patients' clinical status.

Discussion

PTL is a distinct subgroup of tumors, accounting for 7–10% of all intrascrotal neoplasms. These sarcomas arise from mesodermal tissue. Predominantly, they manifest within the spermatic cord, constituting the primary site of origin in ~90% of cases[6]. However, determining the precise anatomical source can be complicated because of the substantial tumor size and degree of adhesion.

The pathogenic mechanisms underlying PTL remain poorly understood due to its rarity and limited research into its etiology.

While data on the exact pathogenic mechanism are limited, it is known that they form *de novo* from connective tissue surrounding the testis, epididymis, and the spermatic cord rather than by the malignant transformation of a preexisting lipoma[8].

Most often, these tumors affect adults between the ages of 50 and 60[9].

The typical clinical presentation of these tumors often involves inguinal edema or the presence of a unilateral scrotal mass, which may be associated with pain in 10–15% of cases and occasionally accompanied by hydrocele[10].

Owing to the non-specific nature of these manifestations, it is crucial to differentiate them preoperatively from conditions such as inguinal hernia, hematocele, hydrocele, lipoma, epididymitis, orchiepididymitis, tuberculosis, and malignant testicular lesions. Generally, these tumors remain asymptomatic for many years, with symptoms varying from 1 week to 5 years[8,11].

In our case, the patient was a 62-year-old man whose manifestations began a year prior to his consultation, in line with the average age range cited in the literature. His symptoms were initially represented by right hemiscrotal swelling, which had significantly increased in size over 1 year.

In most cases, inguinal and scrotal masses are assessed using ultrasound to distinguish between intratesticular and extratesticular locations as well as to differentiate solid masses from cystic masses[8]. Typically, on ultrasound, these masses are represented by a hypervascular and heterogeneous appearance with hyperechoic areas, which depends on the amount of intratumoral adipose tissue present. Although ultrasound imaging can provide valuable information about the size, location, and consistency of masses, it lacks distinctive features to reliably distinguish between benign and malignant lesions, particularly when confronted with small tumors or those exhibiting a homogeneous fat pattern, making it challenging to differentiate between a lipoma and a WDL[12,13].

While early studies suggested that extratesticular lesions demonstrating hyperechogenicity relative to the testis on sonography were indicative of benign entities such as spermatic cord lipoma and fibrous pseudotumor, recent scientific investigations have documented instances of hyperechoic extratesticular malignancies, challenging the reliability of echogenicity as a preoperative discriminator[14].

CT is more useful for diagnosing liposarcoma, allowing for differentiation of tissue characteristics, morphological features, and precise assessment of tumor location. Furthermore, it provides insights into the extent of tumor infiltration into neighboring tissues. Nevertheless, there are no pathognomonic features that can unequivocally distinguish benign from malignant masses. MRI serves as an indispensable modality, providing crucial data for accurate tumor localization and delineation of its anatomical extension[15].

Tumor staging is based on histological examination, grading, and presence of metastases. According to the 2020 World Health Organization's (WHO) histological classification of sarcoma, liposarcoma can be divided into the following five categories: well-differentiated, dedifferentiated, myxoid, pleomorphic, and myxoid pleomorphic liposarcoma[16]. The first two subtypes have a high propensity for locoregional recurrence, while the latter subtypes are more likely to metastasize. Local recurrence rate

after conservative surgery alone reported in literature is up to 30%. The most common PTLs are WDL and DDL. High-grade subtypes (myxoid and pleomorphic) are rare, but are associated with a higher rate of recurrence and hematogenous metastasis to the lungs and bone[17]. The prognosis of liposarcoma depends significantly on its histological subtype.

Rodriguez *et al*. demonstrated that tumor grade, stage, and histologic type were identified as independent predictors of disease-specific survival. In their study, poor tumor differentiation was associated with a lower disease-specific survival time. Patients with distant stage and nodal involvement had worse disease-specific survival[18].

DDL is characterized by its increased aggressiveness and poorer prognosis compared to WDL, with a local recurrence rate estimated at ~40% and a metastatic rate ranging between 15% and 30%[19]. The majority of cases of PTL reported in the literature are WDL. In the present case, the pathological examination of the specimen revealed a sclerosing WDL.

Due to the rarity of PTLs, guidance on their management is primarily drawn from limited case series studies.

The primary surgical intervention for managing spermatic cord tumors is radical orchiectomy with high ligation of the spermatic cord, which ensures negative microscopic margins[20]. Tumor resection alone may not be adequate due to the likelihood of local recurrence, which can also contribute to the development of distant metastases, thereby impacting prognosis[21]. In cases of existing metastases, retroperitoneal lymph node dissection is typically performed[22].

Kamitani *et al*. demonstrated a significantly higher 3-year survival rate without recurrence (79.8%) in patients undergoing high inguinal orchiectomy compared to tumorectomy alone (54.1%). Notably, cases with negative margins exhibited an 88.6% 3-year survival rate, whereas those with positive margins had a rate of 54.2%[23].

Given the elevated risk of recurrence after surgery, there is compelling evidence supporting adjuvant radiotherapy for all spermatic cord tumors, irrespective of histological grade and type[12]. Several studies have indicated that postoperative radiation therapy can markedly decrease the recurrence rate in patients with high-grade tumors, particularly in those with features such as lymphatic invasion, positive margins, or relapses[24,25]. While data may not be extensive enough to firmly establish this, it is plausible that adjuvant radiation therapy could be advantageous for patients with dedifferentiated and pleomorphic liposarcoma, though its efficacy may be somewhat limited in cases of WDL[26].

In contrast, the role of chemotherapy in managing liposarcomas remains inconclusive according to the available literature[27]. Anthracyclines, typically doxorubicin combined with ifosfamide, are the standard chemotherapeutic agents with an overall effectiveness rate of 12%[28]. The round cell and pleomorphic subtypes are particularly sensitive to ifosfamide-based chemotherapy. However, DDL often exhibit chemoresistance, with less than one-third of patients showing a radiographic response[29]. Jones *et al*.[30] found that DDL had a response rate of only 25% to first-line chemotherapy.

Conclusion

PTL, a rare tumor with a painless scrotal mass, is often misdiagnosed preoperatively. Regardless of the tumor size, it is typically managed through standard treatment involving radical orchidectomy, which includes wide excision and high ligation to ensure free surgical margins and avoid recurrence. The role of adjuvant therapy remains debatable. Despite a generally favorable prognosis, diligent long-term follow-up is crucial because of the elevated likelihood of recurrence.

Ethical approval

This is a case report talking about a rare situation but derived from 'standard' clinical practice, so an ethics board approval was not required.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request.

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Author contribution

W.B.: writing – original draft; M.B.O.: writing and editing; I.O., W.H., and M.J.: supervision and reviewing; M.A. and O.B.: review and editing.

Conflicts of interest disclosure

The authors have no conflicts of interest to declare.

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