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Case Report

Paratesticular leiomyosarcoma with metastasis to the extremities and lungs ☆

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ABSTRACT

Paratesticular leiomyosarcoma is a rare malignant tumor deriving from the smooth muscle of structures surrounding the testes, including the epididymis or scrotum. With few cases of genitourinary soft tissue sarcomas reported in the literature, little is known about progression, management, and treatment. Herein, we report a case of metastatic paratesticular leiomyosarcoma in a 47-year-old male with no past urological history. The patient initially presented with a firm, painless right scrotal mass, and ultimately developed soft tissue and pulmonary metastases.

Introduction

Paratesticular sarcomas are rare clinical findings of the genitourinary tract that are believed to derive from smooth muscle structures [1]. A common histological subtype includes leiomyosarcoma (LMS), which presents as a mass distinct from the testes and can be accompanied by a hydrocele [1]. Like other malignant neoplasms, paratesticular LMS has the ability to directly invade surrounding tissue or metastasize to distant parts of the body via lymphatic or vascular routes [1]. Scrotal recurrence is another characteristic of the condition, with rates reaching up to 37% [1]. This case illustrates the progression of paratesticular LMS and its ability to metastasize.

Case presentation

A 47-year-old male with no significant past medical history presented with a right scrotal mass. The patient noted the mass increased in size over time, but denied dysuria, incontinence, straining during urination, and associated symptoms. He had no known family history of urological malignancy. On physical examination, a painless, firm 5.9 cm mass was palpated near the right testis. An ultrasound (US) image of the scrotum demonstrated a heterogeneous extratesticular mass on the right scrotum, the borders of which were distinct from the testicle and epididymis, and a reactive hydrocele (Figs. 1 and 2).

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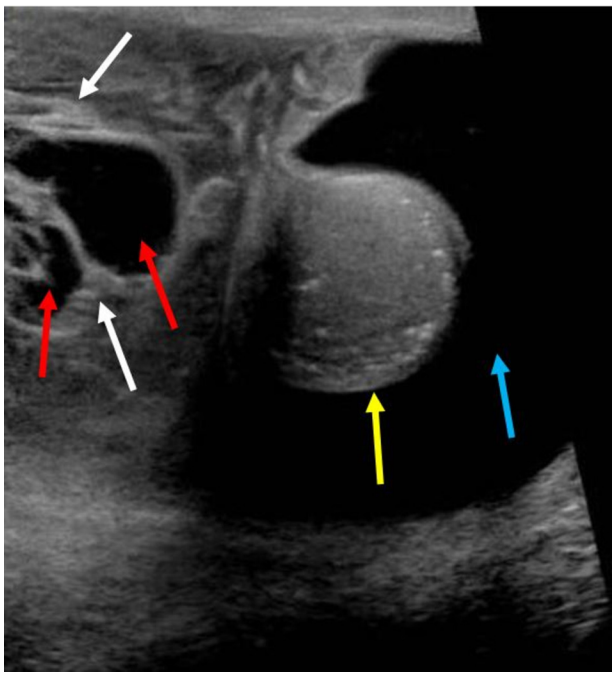

Fig. 1 – Transverse ultrasound image through the scrotum shows a partially imaged extratesticular mass (white arrows), area of central necrosis (red arrows), right testis (yellow arrow), and partially imaged right hydrocele (blue arrow).

Two weeks after the initial ultrasound, the patient underwent a right radical orchiectomy. Resection of the mass exhibited a paratesticular LMS measuring 9.7 × 7.0 × 6.5 cm. The tumor involved the spermatic cord base and the hilum of the testis, extending to the tunica albuginea and vaginalis. Invasion of the epididymis and testes was not present. Vascular invasion was present within the testicular vein, near the spermatic cord margin, but absent from the spermatic cord margin. The tumor was classified as pT2, according to the American Joint Committee of Cancer, and the final diagnosis was a 10 cm T2 Grade 3 LMS.

Microscopically, the tumor is composed of pleomorphic spindle cells with eosinophilic cytoplasm forming fascicles intersecting at right angles (Fig. 3). There are large areas of necrosis, marked cytologic atypia, and brisk mitotic activity (Figs. 4 and 5). Immunohistochemical stains show the tumor cells are strongly and diffusely positive for desmin, and negative for myoD1 and myogenin, supporting their smooth muscle lineage (Fig. 6). The patient had serology tests, which were all within normal limits. Alpha-fetoprotein measured 2.6 ng/mL (0.0-8.3 ng/mL), lactate dehydrogenase measured 174 IU/L (121-224 IU/L), and beta-human chorionic gonadotropin measured <1 mIU/mL (0-3 mIU/mL).

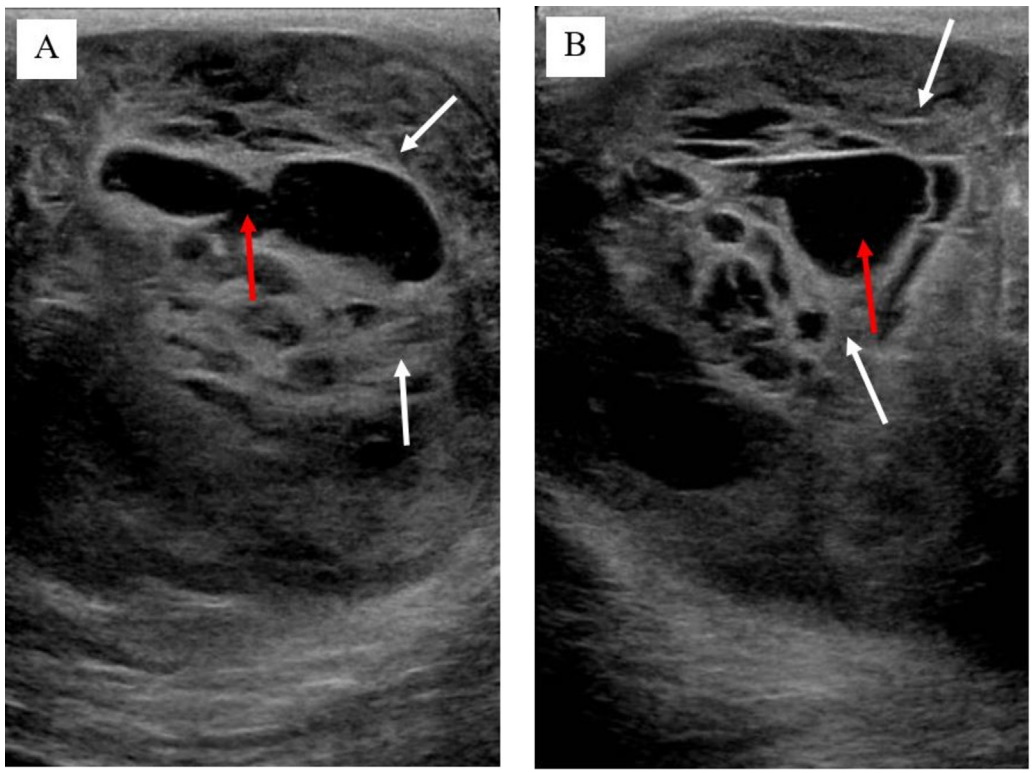

Fig. 2 – Longitudinal (A) and transverse (B) ultrasound image through the scrotum shows an extratesticular mass (white arrows) and areas of central necrosis (red arrows).

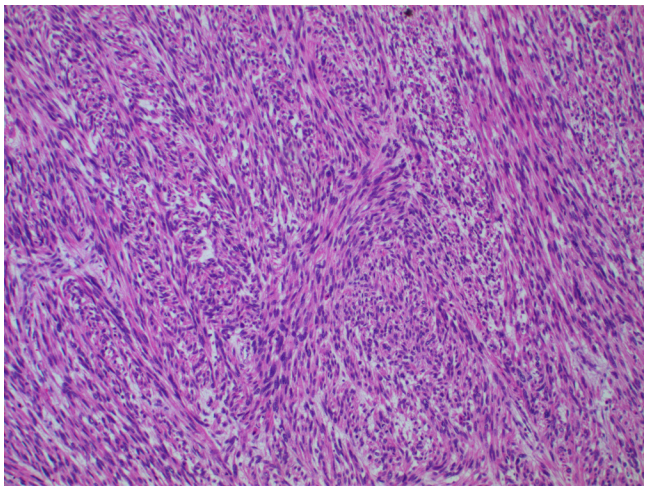

Fig. 3 – H&E (10×) shows pleomorphic spindle cells with eosinophilic cytoplasm forming fascicles intersecting at right angles.

Follow-Up

One year after the right radical orchiectomy, the patient reported a marble-sized lump located on the right side of the scrotum, as well as a mass on the left upper arm and left thigh. On physical examination, the superior portion of the patient's scrotum displayed a palpable, solid, fixed mass approximately 3.5-4.0 cm in size. The left arm revealed a small superficial mass stretching from the distal left anterior to mid bicep. A small superficial mass on the anteromedial portion of the proximal left thigh was also observed. The patient denied pain, swelling, and associated symptoms.

Superficial US of the left upper extremity demonstrated a 2.2 × 2.1 × 1.5 cm heterogeneous solid vascular mass displacing the neighboring musculature (Fig. 7). US of the left lower extremity showed a 2.6 × 1.4 × 1.2 cm heterogeneous hypoechoic solid vascular mass displacing the neighboring musculature. The sonographic features were comparable to the mass in the left upper arm. CT CAP with contrast exhibited solid nodules in the right scrotum and right inguinal region, indicating local recurrence (Fig. 8). Solid bilateral pulmonary nodules were present in the right middle and left lower lobes, suggestive of metastases (Fig. 9).

PET CT revealed hypermetabolic masses in the right groin, left thigh, and left elbow in addition to hypermetabolic pulmonary nodules. Left upper extremity biopsy was performed percutaneously and revealed metastatic tumor morphologically identical to the patient's paratesticular LMS. The metastatic tumor was positive for desmin by immunostain, and the Ki67 proliferation index was up to 30%. These findings indicate lymph node involvement and distant metastatic disease.

Discussion

Leiomyosarcomas are a type of soft tissue sarcoma that most commonly arise from abdominopelvic organs, including the testes and its surroundings [2]. Paratesticular LMS are rare clinical entities that are characterized by their presence within the scrotum with a clear origin from the epididymis or spermatic cord [3]. Additionally, they have the ability to metastasize by means of the blood or lymphatic system [1]. Incidence is highest in males during the 6th and 7th decades [4]. Patients can present with a painless mass, noticeable on physical examination, followed by hydrocele on sonography.

Diagnosis requires both histological findings and imaging techniques, such as sonography, MRI, and CT. Sonography is a key imaging method used for patients presenting with a primary extratesticular mass, typically being characterized by general grey-scale sonographic features [5]. It also has a sensitivity of up to 100% to differentiate between intratesticular and extratesticular masses [5]. MR imaging allows for the determination of vascularity and fat content of the tu-

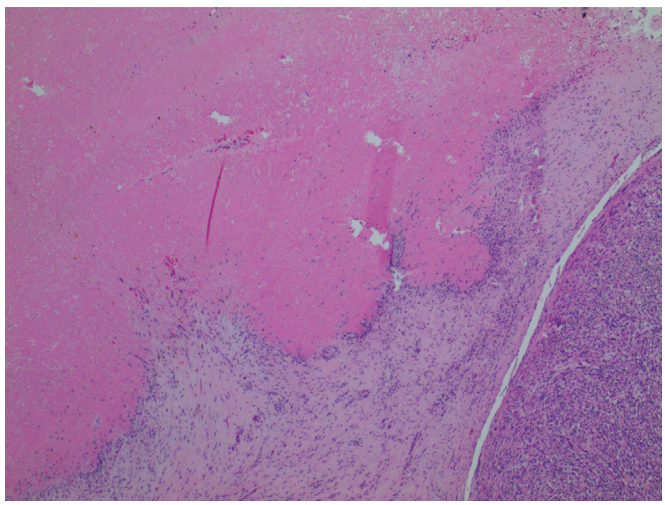

Fig. 4 – H&E (10×) showing large area of necrosis (pink geographic area).

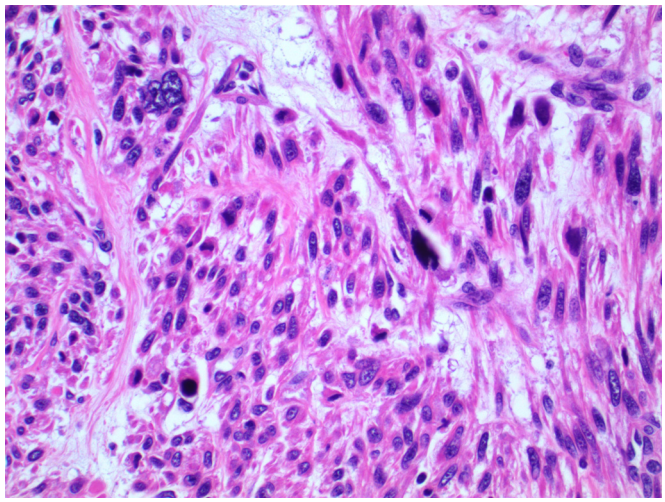

Fig. 5 – H&E (40×) displaying marked cytologic atypia and frequent mitotic figures.

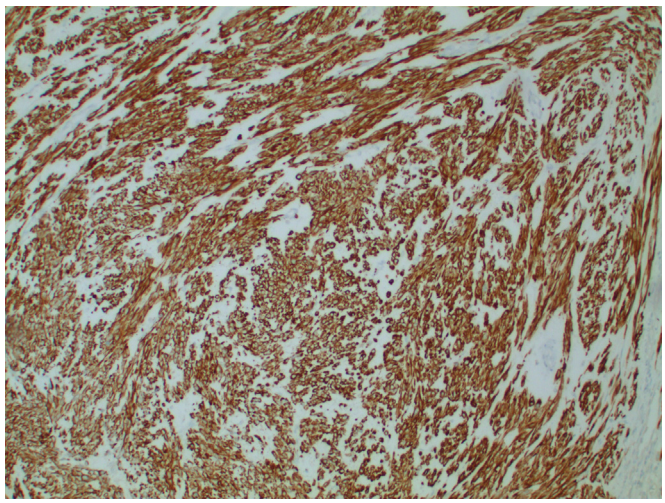

Fig. 6 – Desmin immunostain (10×) shows the tumor is strongly and diffusely positive for desmin, and negative for myoD1 and myogenin (not shown) by immunohistochemistry, supporting their smooth muscle lineage.

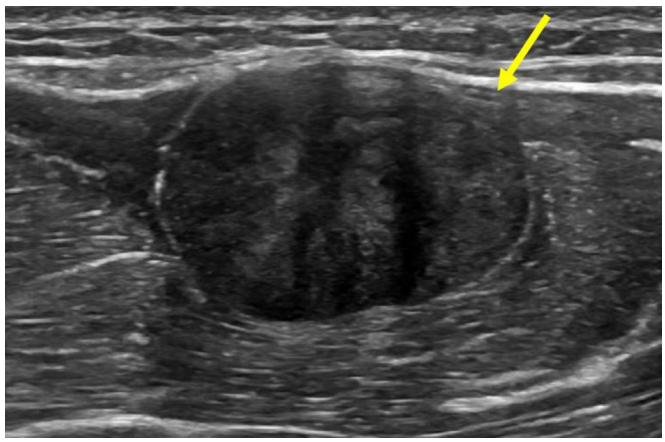

Fig. 7 – Long-axis superficial ultrasound image of the left upper arm shows a soft tissue mass (yellow arrow).

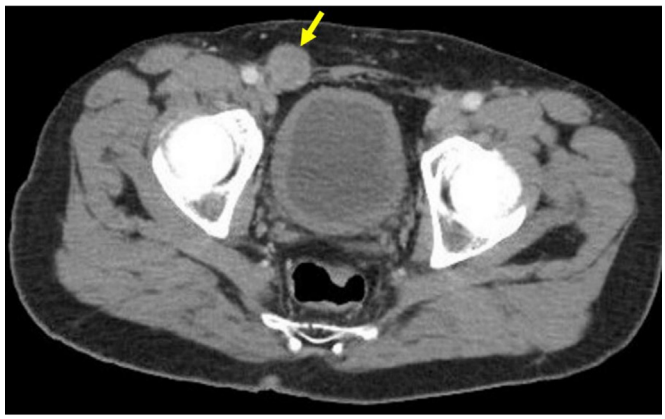

Fig. 8 – Axial IV contrast-enhanced CT image through the pelvis shows a soft tissue nodule in the right inguinal region compatible with lymph node metastasis (yellow arrow).

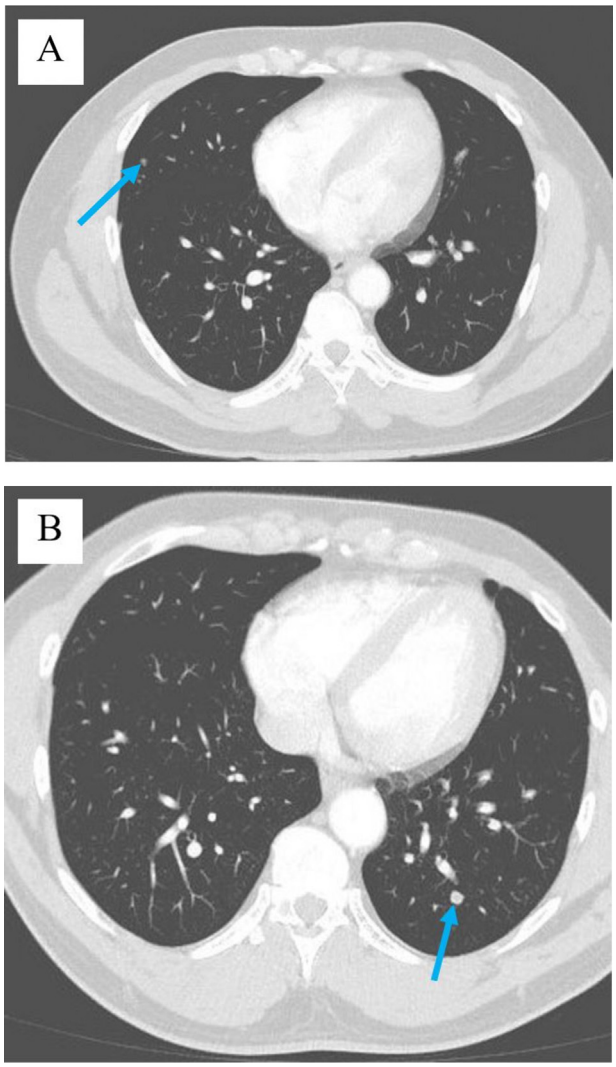

Fig. 9 – Axial IV contrast-enhanced CT image through the chest shows subcentimeter pulmonary nodules in the right middle lobe and in the left lower lobe (blue arrows).

mor [5]. Histological examination of paratesticular LMS has revealed atypical forms, coagulative necrosis of tumor cells, and immunohistochemistry positive for desmin [1]. Differential diagnoses include hydrocele, inguinoscrotal hernia, benign leiomyoma, rhabdomyosarcoma, and various testicular malignancies [6]. Radical orchiectomy is the typical operation performed, followed by adjuvant radiotherapy to decrease the rate of local recurrence [6]. Prognosis of the condition depends predominantly on the tumor grade [4].

Conclusion

Due to of its uncommon presence in the population, paratesticular LMS presents challenges in treatment, as standard of care is not well established [1]. While radical orchiectomy is typically performed, the metastatic capability of the sarcoma can lead to recurrence in the regional area [1]. Adjuvant radiotherapy can be considered as it has been observed to decrease the rate of local recurrence [1]. In male patients presenting with scrotal mass, sarcoma such as paratesticular LMS should be considered as part of the differential diagnosis. While paratesticular LMS is rare, early detection can potentially alter clinical outcomes and impact rates of recurrence and metastases. Characteristic imaging followed by biopsy to establish pathology diagnosis on histology early is crucial for timely treatment and favorable prognosis.

Patient consent

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

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