



Case Report

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Leiomyosarcoma of the Scrotum: A Case Report

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Leiomyosarcoma (LMS) within the scrotum is a rare malignant genitourinary tumor in adults. Due to its rarity, a consensus on a clear approach to diagnosis and treatment guidelines has not yet been reached. We present a case of a paratesticular LMS in a 64-year-old man with a painless swelling of the right scrotum. Scrotal ultrasound and pelvic MRI were performed and revealed an extratesticular mass with a heterogeneous appearance. The patient underwent a right radical orchiectomy, and the mass was confirmed to be LMS, originating from the right spermatic cord. Preoperative diagnosis of LMS is challenging since it has overlapping clinical and imaging features with other sarcomas. MRI is a valuable imaging modality for the definitive characterization of the tumor. To provide timely and appropriate treatment, the correlation of the patient's clinical information and imaging findings is necessary.

Keywords: Leiomyosarcoma; Paratesticular tumor; Paratesticular leiomyosarcoma; Magnetic resonance imaging

INTRODUCTION

Genitourinary sarcomas are extraordinarily rare tumors found in adults and account for less than 5% of all soft-tissue sarcomas [1]. Leiomyosarcoma (LMS) is the second most common histologic subtype of sarcoma within the scrotum and represents about 20% of all intrascrotal sarcomas, following liposarcoma [2]. LMS is a malignant mesenchymal tumor arising from smooth muscles, and more than 75% of intrascrotal LMS are located at the spermatic cord [3]. Because of its rarity, there is no firm evidence of a clear approach to diagnosis and treatment guidelines. This report outlines a case of LMS in the scrotum, presenting as a painless mass lesion, and reviews the radiological findings using ultrasound (US) and MRI.

CASE REPORT

A 64-year-old man presented to our institution with a painless swelling of the right scrotum. On physical examination, a firm and non-tender mass was found in the superior portion of the right scrotum. The patient had no previous medical or surgical history, and tu-

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mor markers such as β -human chorionic gonadotropin (β -hCG), alpha-fetoprotein (AFP), and lactate dehydrogenase (LDH) were within normal ranges. Scrotal US was performed as the primary radiologic examination, and it revealed a well-circumscribed heterogeneous echoic mass within the right inguinal canal. A normal right testis and epididymis were located below, away from the mass, so it was considered to be of paratesticular origin. Color Doppler US imaging showed vascular flow within the tumor (Fig. 1). For evaluation of the tumor component, a pelvic MRI with contrast enhancement was conducted using a 3-T scanner. T2-weighted and T1-weighted images showed a 6.0 cm $\times$ 4.8 cm $\times$ 5.0 cm sized, well-defined, low signal intensity mass without definite necrosis or cystic degeneration

located at the right spermatic cord. A T2-weighted fat-suppressed image showed that there was no fat component within the tumor. The tumor showed marked enhancement and diffusion restriction was not definite (Fig. 2). The bilateral testes and epididymides were unremarkable. These findings suggested the possibility of a paratesticular malignant sarcoma, such as rhabdomyosarcoma or LMS, originating from the right spermatic cord. The patient underwent a right radical orchiectomy under spinal anesthesia. A gross examination of the resected specimen showed a 5 cm-sized, well-demarcated, solid mass located 3 cm apart from the resection margin of the right spermatic cord. The cut surface of the mass showed a relatively uniform grayish-white appearance with the presence

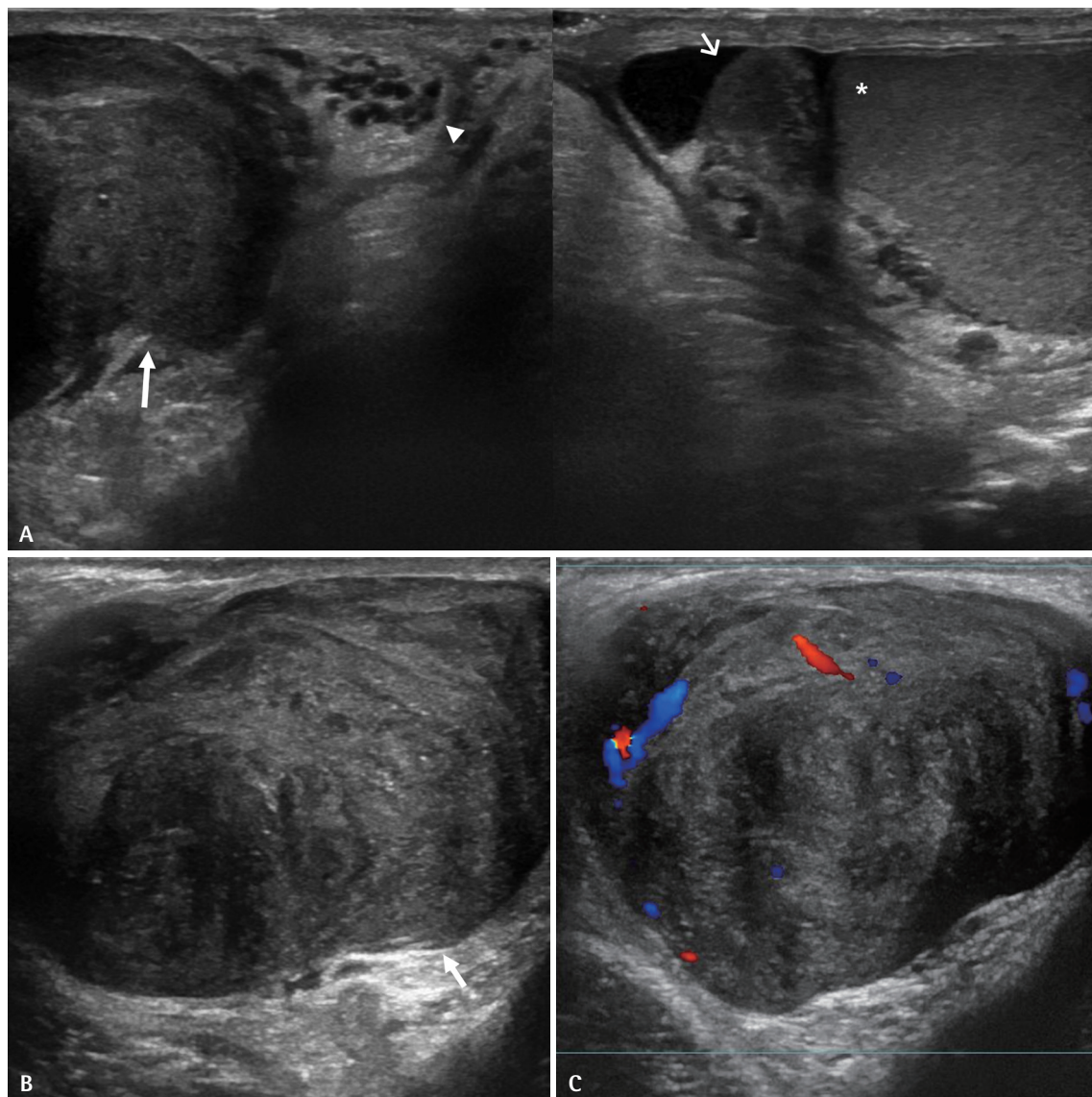


Fig. 1. Ultrasound (US) imaging of scrotum. Grayscale US images (A and B) show a well-circumscribed heterogeneous echoic mass (thick arrow) within the right inguinal canal, which is located superior to the normal right spermatic cord (arrowhead), testis (asterisk), and epididymis (thin arrow). Color Doppler US image (C) shows vascular flow within the tumor.

of some myxoid components. Microscopically, the tumor was a spindle cell neoplasm with marked nuclear pleomorphism and atypical mitosis. Immunohistochemistry confirmed positive desmin and smooth muscle actin (SMA) stains and a high Ki-67 expression, which suggested high proliferative activity

of the tumor (Fig. 3). These pathologic findings were consistent with LMS derived from the spermatic cord. A fluorodeoxyglucose (FDG) positron emission tomography/CT (FDG-PET/CT) showed no evidence of distant metastasis, and the patient underwent adjuvant radiation therapy.

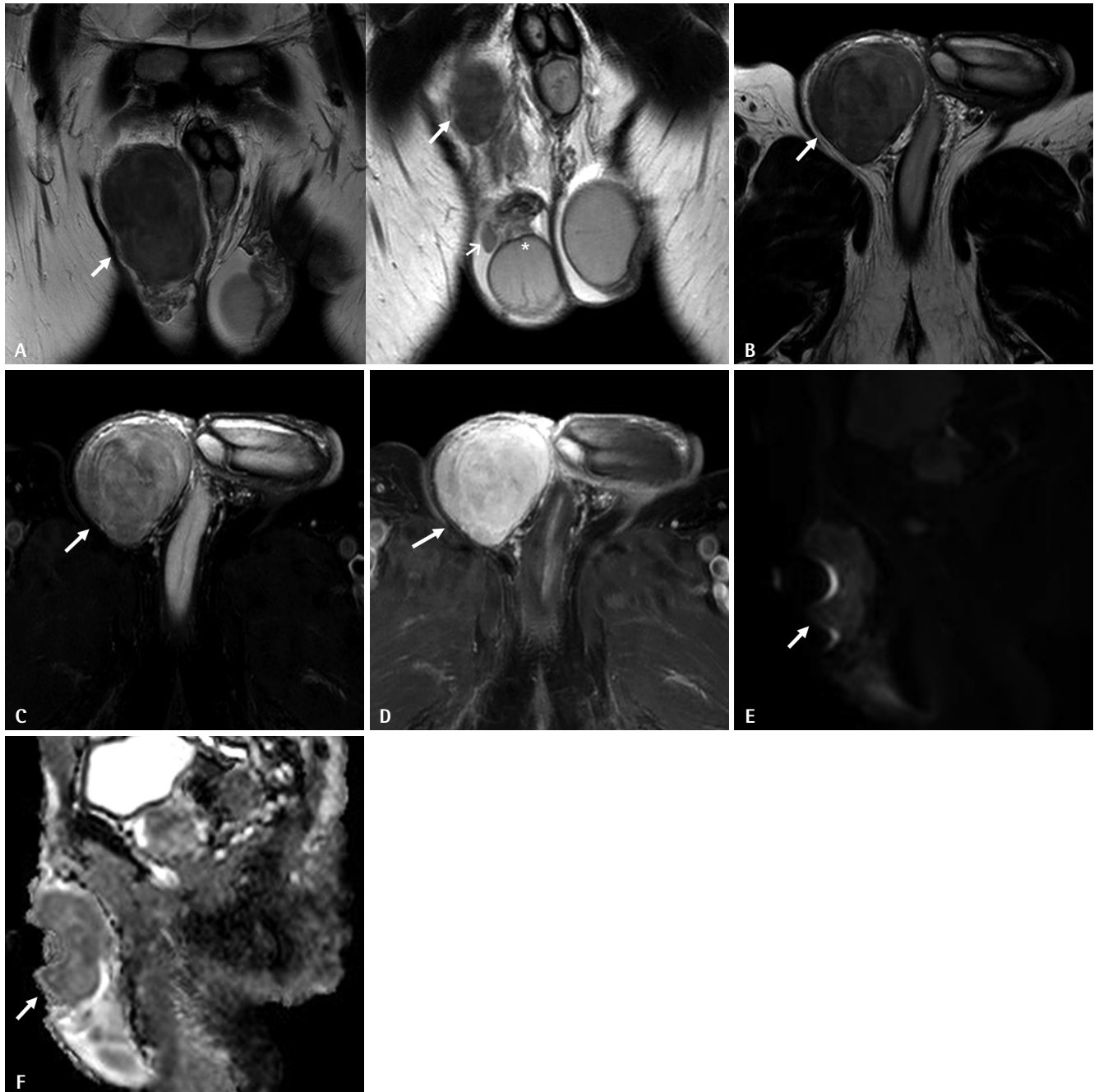


Fig. 2. Pelvic MRI findings. Coronal T2-weighted images (A) and axial T1-weighted image (B) show a 6.0 × 4.8 × 5.0 cm sized, well-defined, low signal intensity mass (arrows) without definite necrosis or cystic degeneration that is located in the superior aspect of the right testis (asterisk) and epididymis (thin arrow) within the right inguinal canal. Axial T2-weighted fat-suppressed image (C) shows no fat component within the tumor. The tumor shows marked enhancement on an axial T1-weighted contrast-enhanced image with fat suppression (D). Sagittal diffusion-weighted image ($b = 800 \text{ sec/mm}^2$) (E) and apparent diffusion coefficient (F) show no definite restricted diffusion within tumor (arrow).

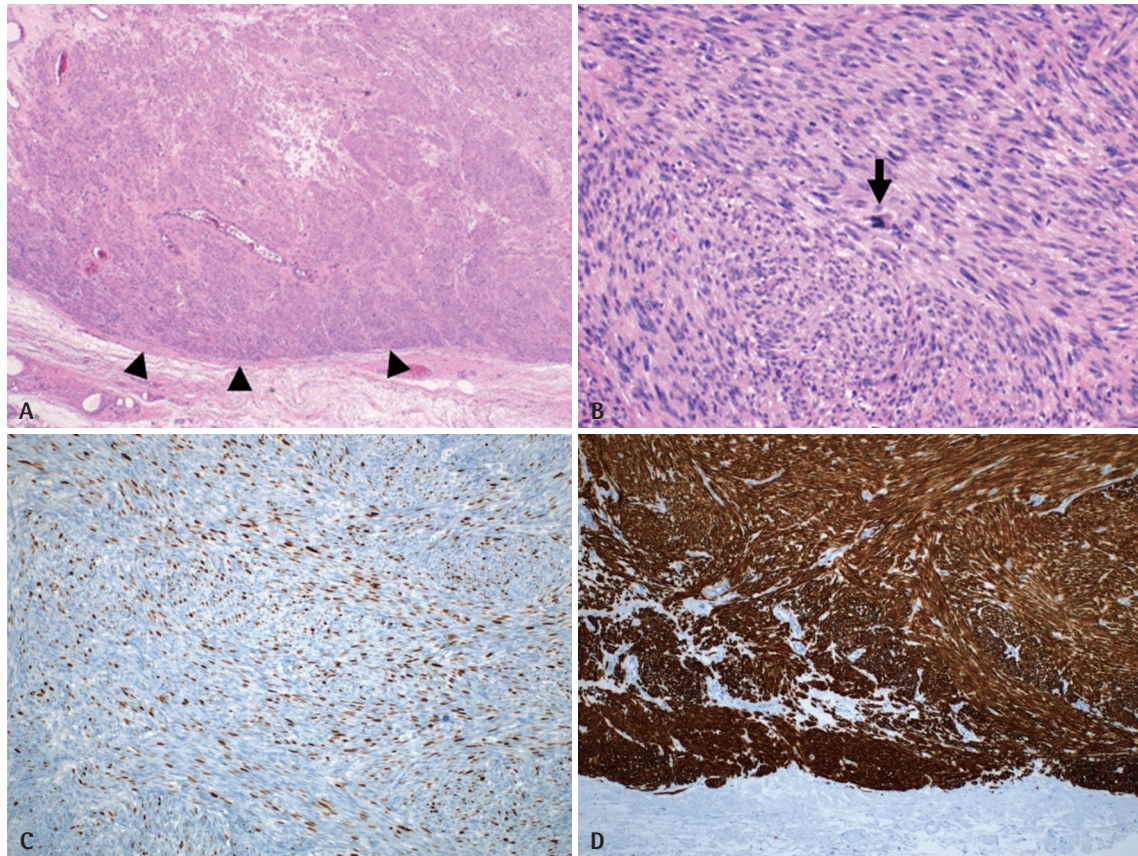


Fig. 3. Pathological finding. A and B: Photomicrography shows a well-defined mass (A, arrowheads) and atypical spindle cells with mitosis (B, arrow) (hematoxylin and eosin staining; A: $\times 40$, B: $\times 200$). C and D: The tumor shows high Ki-67 expression (C, $\times 100$), suggesting high proliferative activity, and the tumor is positive for desmin immunohistochemical stain (D, $\times 40$).

DISCUSSION

Masses within the scrotum can originate from the testes or paratesticular structures. Most intrascrotal masses in adults are of testicular origin, however, paratesticular masses are rarer and account for only 2%–3% of all scrotal masses [4]. The paratesticular area is composed of the spermatic cord (vas deferens, testicular and cremasteric arteries, arteries of the vas deferens, pampiniform plexus of veins, lymphatic vessels of the testis, and nerves), the scrotum (testicular tunica, dartos muscle, and scrotal subcutis), and the epididymides or vestigial remnants (appendices epididymis and testis) [5]. LMS is a malignant soft tissue tumor that derives from any tissue containing smooth muscle or multi-potential mesenchymal cells [6]. Paratesticular LMS most commonly arises from undifferentiated mesenchymal cells of the cremasteric muscle and vas deferens. Less frequently, the epididymal type derives from the smooth muscle that surrounds the basement membrane of the epididymis canal. The scrotal type of paratesticular LMS originates from the dartos layer [7]. Lymphatic metastasis is the most common mode of spread. The lymphatic drainage of the

two former types is through the retroperitoneal lymph nodes, and the latter type drains through the inguinal, external, and internal iliac nodes [7]. Hematogenous metastasis then follows and is mainly pulmonary or hepatic [6].

Most paratesticular LMS presents as an enlarging but painless lump in the scrotum, as is the case with our patient. Complications, such as superimposed infection or bleeding from ulceration, can occasionally arise [8]. Patients range in age from 34–86 years, with a peak incidence occurring in the 60–70 year age group [3], and there is no sex predilection [5]. Preoperative diagnosis of paratesticular LMS is challenging, and a definitive diagnosis usually requires pathologic confirmation from a surgically removed specimen. The diagnostic workup typically includes: a serum tumor marker (AFP, β -HCG, and LDH), US, and CT or MRI. Most LMS patients show negative tumor markers, but one case report of paratesticular LMS presented a patient with a high β -HCG, which returned to normal six weeks after surgery [8]. US is the imaging modality of choice for diagnosing a scrotal lesion because it is inexpensive, easily accessible and straightforwardly determines the lesion's origin. US images show that the majority of the LMS appears to be of het-

erogeneous echogenicity, and calcifications are uncommon. Color Doppler imaging demonstrates an increased or minimal vascularity within the tumor. The size of the mass and the degree of differentiation of the mesenchymal components affect the tumor's image features. US images may not allow for the definitive characterization of the paratesticular mass. Most solid lesions, either benign or malignant, frequently have similar US findings, making it extremely difficult to exclude malignant tumors [7]. Where diagnosis using US is inconclusive, MRI is highly effective at discriminating between benign and malignant paratesticular tumors because of its high soft tissue contrast and high sensitivity for contrast enhancement. MRI can reveal a characteristic signal intensity, according to the types of tissue components such as fat, blood, and fibrotic or granulation tissue, which allows for a narrower differential diagnosis. LMS presents as solid lesions of an intermediate to low signal intensity on T2-weighted images and usually shows an avid and heterogeneous enhancement on gadolinium-enhanced T1-weighted images. The absence of areas with fat suppression also indicates the absence of fat within the lesion, which excludes the possibility of liposarcoma. MRI is more accurate than US in defining the location of the tumor, its relationship with paratesticular structures, and the local extent of disease, which are useful for preoperative planning. MRI is also independent of an operator's proficiency as compared to US.

Moreover, MRI can indicate the histologic grade of soft tissue sarcomas before biopsy [9]. Crombé et al. [9] identified that MRI features of heterogeneity, the amount of necrosis, and peritumoral enhancement of soft-tissue sarcomas were associated with high-grade tumors in a retrospective cohort of 121 patients, and its presence suggested a poor prognosis. CT is usually not performed to analyze the lesion itself but to evaluate the extent of the disease that is malignant. CT reveals a heterogeneous mass with cystic, solid, or calcified components and irregular and peripheral contrast enhancement. The ipsilateral spermatic cord shows edematous change with dilated vessels [7]. FDG-PET/CT is also helpful to see the nodal involvement or distant metastasis.

At gross examination, LMS is well demarcated with whitish lobulated masses that may contain some foci of hemorrhage and necrosis [5]. Microscopically, the majority of LMS displays perpendicularly oriented fascicles of spindle cells which contain elongated eosinophilic cytoplasm and blunt-ended nuclei [3,5]. The grading of paratesticular LMS is one prognostic factor, and it is based on the number of mitoses, the percentage of necrosis, and the severity of nuclear pleomorphism [7]. Immunohistochemistry plays a role in identifying its origin from the smooth muscle. The LMS components are positive for α SMA, desmin, and vimentin and negative for S-100 protein, keratin, and factor VIII-related antigen [10]. Due to the

rarity of LMS, the ideal treatment protocol is controversial. Radical orchiectomy with high ligation of the spermatic cord is considered to be the treatment of choice in this neoplasm. Locoregional recurrence is common, and achieving a negative margin is very important for reducing the possibility of recurrence, but this is rarely achieved during primary surgery. Therefore, when the negative surgical margin is not obtained, or the local recurrence is highly suspicious, adjuvant chemotherapy or radiation is usually needed [8].

In conclusion, paratesticular LMS is an uncommon malignant sarcoma in adults. Its diagnosis is challenging even when determining whether the paratesticular lesions are benign or malignant, given their overlapping imaging features. US is the initial imaging modality since it can determine the origin of the lesion, but its capacity is limited when making a single diagnosis. MRI is beneficial to narrow the differential diagnosis, clarify the tumor location, and anticipate the patient's prognosis in support of clinical decision-making. Herein, a rare paratesticular entity was assessed by US and subsequent MRI, ultimately emphasizing the importance of a preoperative MRI for a paratesticular scrotal mass. The correlation of the patient's clinical information and imaging findings can allow for a more accurate differential diagnosis, which results in timely and precise treatment.

Ethics Statement

The patient consented to report his medical condition and imaging findings by intraoperative magnetic resonance imaging.

Availability of Data and Material

The datasets generated or analyzed during the study are available from the corresponding author on reasonable request.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

Author Contributions

Conceptualization: Ji Yeong Hwang, Rae Rim Ryu. Formal analysis: Young Joong Kim. Validation: Dong Hyun Oh, Hyeong Chun Kim. Writing—original draft: Ji Yeong Hwang. Writing—review & editing: Keum Won Kim.

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