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Primary Intratesticular Leiomyosarcoma: A Case Report and Literature Review

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ABSTRACT

Background: Primary intratesticular leiomyosarcomas are rare neoplasms. Diagnosis is established after radical inguinal orchiectomy through histological and immunohistochemical analysis. According to the literature, testicular leiomyosarcoma may exhibit indolent behavior and can potentially be cured with early detection and surgical intervention.

Case Report: A 63-year-old man presented with painless enlargement of the right testis. High inguinal orchiectomy was performed. Histopathological evaluation of the excised specimen demonstrated low-grade leiomyosarcoma.

Conclusion: The clinical, radiological, and histopathological characteristics of this tumor are described, along with an overview of current therapeutic approaches.

Keywords: Leiomyosarcoma, orchiectomy, testicular mass, testis.

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INTRODUCTION

Leiomyosarcoma of the testis is a rare tumor, with the first case reported by Yachia and Auslaender in 1989. Since then, approximately 39 cases have been documented.[1] In younger patients, this neoplasm has been associated with prior radiotherapy and anabolic steroid use. The clinical management is similar to that of other testicular masses, with painless scrotal swelling being the most common clinical presentation.[2] For diagnostic evaluation, ultrasonography effectively identifies a hypoechoic mass with or without calcification, whereas cross-sectional imaging is necessary to assess for metastatic disease. Definitive diagnosis is achieved after radical inguinal orchiectomy through histological and immunohistochemical analysis. The literature indicates that testicular leiomyosarcoma may demonstrate indolent behavior and can potentially be cured with early diagnosis and surgical intervention.

CASE REPORT

A 63-year-old man was referred to the urology clinic for evaluation of lower urinary tract symptoms. His medical history included hypertension, managed with enalapril, and previous surgeries, including right inguinal hernia repair in 2022 and right testicular sperm extraction (TESE) in 1987.

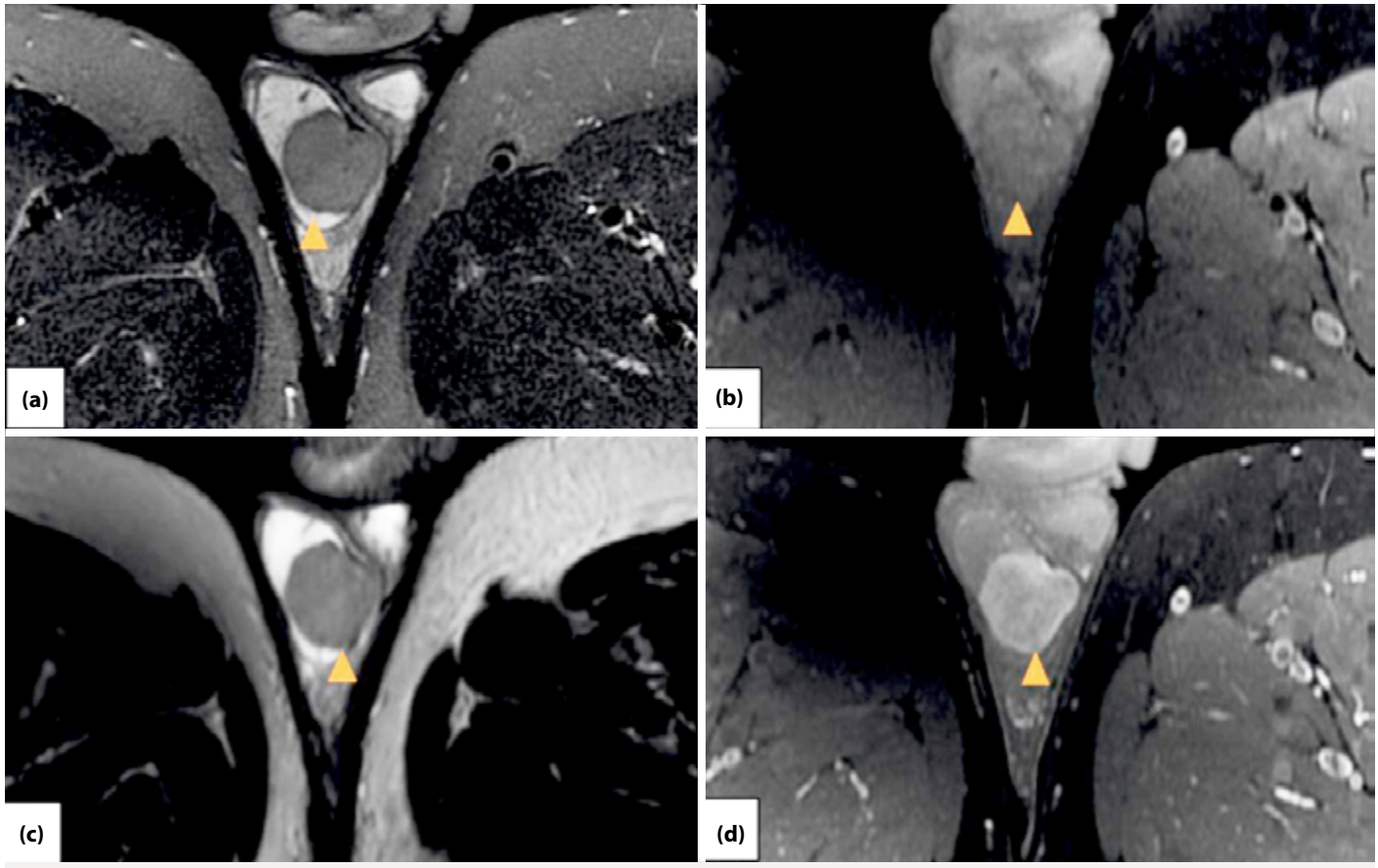

Figure 1. Pelvic MRI findings of the tumor. **(a)** Axial T2-weighted image. **(b)** Axial T1-weighted image. **(c)** Axial T2-weighted fat-suppressed image. **(d)** Axial T1-weighted contrast-enhanced image.

Physical examination revealed that the right testis was solid, irregular, and atrophic, whereas the left testis was also atrophic but regular and soft. Testicular tumor markers, including alpha-fetoprotein, lactate dehydrogenase, and beta-human chorionic gonadotropin, were within normal limits. Scrotal Doppler ultrasonography (USG) was performed as the initial radiological assessment and identified a well-circumscribed, heterogeneous lesion measuring 26×24 mm in the right testis. Doppler imaging demonstrated an isohypoechoic lesion with central vascularity relative to the testicular parenchyma. Millimetric echogenic foci consistent with microlithiasis were observed in the parenchyma of both testes, more prominently on the left. The left testis was reduced in size, but no intratesticular solid or cystic mass was identified.

One month after USG, contrast-enhanced pelvic magnetic resonance imaging (MRI) identified a hypointense mass in the right testis on axial T2-weighted images (Fig. 1a), characterized by regular borders, necrosis, and cystic degeneration. The mass appeared isointense on axial T1-weighted images (Fig. 1b). Axial T2-weighted fat-suppressed images (Fig. 1c) confirmed the absence of a fat component within the tumor. Significant contrast enhancement was observed on axial T1-weighted contrast-enhanced fat-suppressed images (Fig. 1d). Axial diffusion-weighted imaging (b=800 sec/mm^2) and apparent diffusion coefficient analysis revealed no definite restricted diffusion within the tumor. Thoracoabdominal computed tomography (CT) scans did not detect any additional primary or metastatic lesions. Based on the imaging findings, a clinical diagnosis of testicular malignancy was established. The patient subsequently underwent high inguinal orchiectomy under spinal anesthesia. No complications were observed during the postoperative period.

Macroscopic examination of the resected specimen demonstrated a 3×2.2 cm gray-white solid tumor confined to the testis (Fig. 2). Microscopic examination revealed atrophic testicular tissue surrounding the mesenchymal tumor (Fig. 3a). The tumor was composed of spindle cells arranged in intersecting bundles, with increased cellularity, low mitotic activity, and mild to moderate pleomorphism

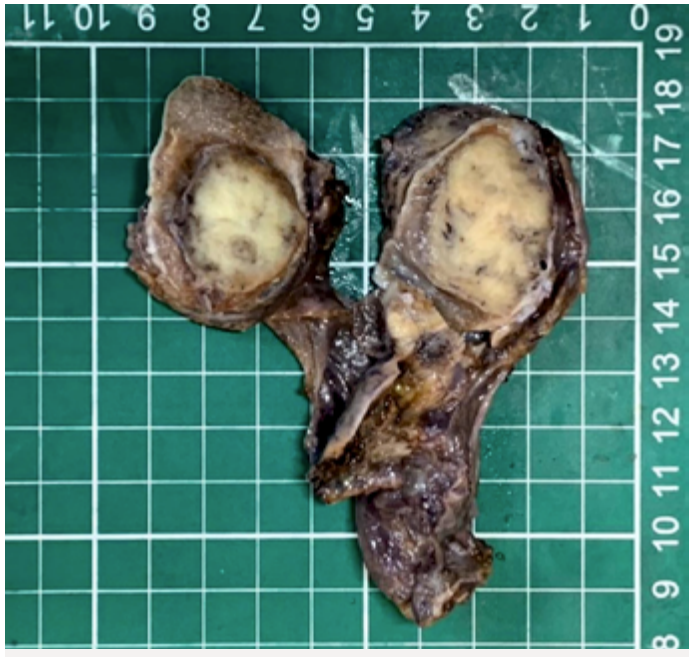

Figure 2. Macroscopic image of the intratesticular tumor mass.

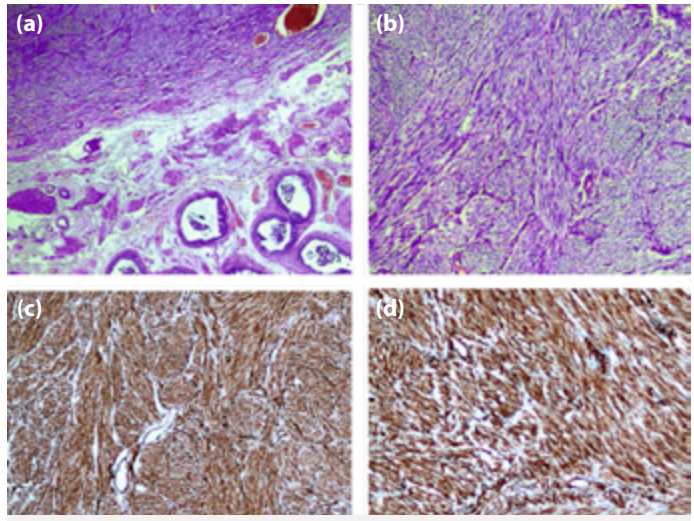

Figure 3. (a) Atrophic testicular tissue and epididymis surrounding the tumor (H&E, ×40). **(b)** Tumor composed of spindle cells forming intersecting smooth muscle bundles (H&E, ×200). **(c)** Diffuse positive staining for SMA in the tumor (×200). **(d)** Diffuse positive staining for desmin in the tumor (×400).

(Fig. 3b). Necrosis and lymphovascular invasion were absent. According to the FNCLCC grading system, the tumor had a total score of 3 (differentiation: 2, mitotic count: 1, necrosis: 0), corresponding to grade 1.

Immunohistochemical analysis demonstrated diffuse positivity of the tumor cells for vimentin, desmin, smooth muscle actin (SMA), and caldesmon. The tumor cells were negative for cytokeratin AE1/AE3, myogenin, S100, SOX10, inhibin, HMB45, Melan-A (MART1), PLAP, CD34, and CD117 (Fig. 3c, d). The Ki-67 proliferation index was low (3–5%).

In the differential diagnosis of dedifferentiated liposarcoma with smooth muscle differentiation, microscopic examination did not reveal any fatty component. Immunohistochemical analysis was negative for CDK4 and MDM2. In addition, fluorescence in situ hybridization (FISH) analysis did not demonstrate CDK4 or MDM2 amplification. Other differential considerations included testicular teratoma with smooth muscle differentiation; however, no teratomatous component was identified. Based on the combined histopathological and immunohistochemical findings, the tumor was diagnosed as primary low-grade intratesticular leiomyosarcoma. The tumor infiltrated the tunica albuginea, with no involvement of the tunica vaginalis, epididymis, or spermatic cord. All surgical margins were tumor-free. The patient did not receive postoperative chemotherapy or radiotherapy. No metastasis or recurrence was observed during the 2-year follow-up period.

DISCUSSION

Leiomyosarcomas are malignant soft-tissue tumors that arise from tissues containing smooth muscle. Scrotal leiomyosarcomas are categorized as either paratesticular or intratesticular.[3] Paratesticular leiomyosarcomas are more common, with approximately 100 documented cases.[4] In contrast, intratesticular leiomyosarcoma, which develops from smooth muscle cells of the tunica propria, tunica albuginea, seminiferous tubules, or blood vessel walls, is rare and has been reported in only a few cases in the literature.[1]

Primary intratesticular leiomyosarcomas most commonly present between the fourth and seventh decades of life, with a mean age at diagnosis of 50 years. The precise etiology of testicular leiomyosarcoma is unknown; however, potential contributing factors include hormonal stimulation, such as prolonged anabolic steroid use, radiotherapy, and chronic inflammation.[5,6]

Scrotal ultrasonography is the primary radiological diagnostic tool. When ultrasonographic findings are inconclusive, MRI can effectively differentiate benign from malignant paratesticular tumors because of its superior soft-tissue contrast and high sensitivity to contrast enhancement.[7] Evaluation for distant metastases, including pulmonary and lymph node metastases, is performed using thoracoabdominopelvic CT.

Definitive diagnosis of intratesticular leiomyosarcoma depends on histopathological examination. Demonstrating the absence of germ cell elements within the tumor is essential for establishing the diagnosis of primary leiomyosarcoma.[8] Histological criteria for intratesticular leiomyosarcoma are limited in the literature. To date, 39 cases of intratesticular leiomyosarcoma have been reported.[1] Appendix 1 summarizes the clinical features, pathological findings, demographic characteristics, therapeutic approaches, and clinical outcomes of previously reported cases, as well as those of the current case. Diagnostic criteria for uterine spindle cell leiomyosarcoma, including nuclear atypia, mitotic count, and coagulative necrosis with nuclear debris, may also be applied to intratesticular leiomyosarcoma. Immunohistochemical studies are recommended to support the diagnosis of a primary intratesticular smooth muscle tumor.

High inguinal radical orchiectomy is the standard treatment for intratesticular leiomyosarcoma. Current literature suggests that adjuvant therapy, including chemotherapy and radiotherapy, is generally not recommended because of the low metastatic potential of these tumors.[9] Distant metastases have been reported in only 4 cases, all involving high-grade tumors. Although the role of adjuvant therapy in advanced disease remains poorly defined, chemotherapy is typically indicated in such cases.[10] The standard regimen for invasive or metastatic leiomyosarcoma includes doxorubicin or a combination of doxorubicin and ifosfamide.[2] In the present case, the tumor was low-grade, stage I, and confined to the testis, with no evidence of metastasis; therefore, adjuvant chemotherapy or radiotherapy was not administered after radical orchiectomy.

The prognosis of testicular leiomyosarcoma is generally more favorable than that of leiomyosarcomas arising in other organs. Follow-up every 6 months, including urogenital examination and CT imaging, is recommended.[7] Close monitoring is essential because late metastasis can occur even in low-grade tumors, and mortality due to pulmonary involvement has been reported up to 24 months after orchiectomy.[9]

CONCLUSION

Because of the rarity of this tumor, data on its clinical course, diagnostic criteria, prognostic factors, survival rates, and treatment options remain limited. Radical orchiectomy is considered the standard initial treatment. Nevertheless, optimal management strategies have not been well established because of insufficient research and the absence of formal guidelines. Existing studies indicate that early diagnosis and treatment are associated with a high cure rate.

Ethics Committee Approval: This is a single case report, and therefore ethics committee approval was not required in accordance with institutional policies.

Informed Consent: Written informed consent was obtained from the patient and relatives.

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Appendix 1. Summary of the 38 published case reports

Case no.	Authors	Age	Side	Clinical stage	Treatment	Follow-up (months)	Outcome
1	Yachia[1]	55	R	I	orchiectomy	24	survived
2	Pellice[2]	37	L	I	orchiectomy	24	survived
3	Washecka[3]	47	R	I	orchiectomy	49	survived
4	Washecka[3]	40	R	I	orchiectomy	42	survived
5	Froehner[4]	32	R	I	orchiectomy+LND	79	survived
6	Hachi[5]	70	L	I	orchiectomy	14	death (lung metastasis)
7	Ali[6]	65	R	I	orchiectomy	12	survived
8	Takizawa[7]	76	L	I	orchiectomy	12	survived
9	Canales[8]	30	R	I	orchiectomy	6	survived
10	Yoshimine[9]	73	L	III	orchiectomy+CTx	9	survived
11	Sattary[10]	27	L	I	orchiectomy	30	survived
12	Singh[11]	26	L	I	orchiectomy	not mentioned	survived
13	Wakhlu[12]	0,8	L	I	orchiectomy	12	survived
14	Borges[13]	19	L	I	orchiectomy+CTx+RT	16	survived (retroperitoneal recurrence)
15	Fadl-Elmula[14]	20	L	II	orchiectomy+CTx+RT	11	survived (cerebral metastasis+ local recurrence+paraaortic LN metastasis)
16	Joshi[15]	65	R	I	orchiectomy	6	survived
17	Raspollini[16]	77	L	I	orchiectomy	12	survived
18	Tobe[17]	71	R	I	orchiectomy	7	survived
19	Labanaris[18]	73	R	I	orchiectomy	28	survived
20	Giridhar[19]	55	L	I	orchiectomy+CTx	11	death (soft tissue and bone metastasis)
21	Komeya[20]	70	L	I	orchiectomy	34	survived (retroperitoneal metastasis)
22	Moona[21]	45	R	II	orchiectomy+CTx	not mentioned	survived (paraaortic LN metastasis)
23	Bakhshi[22]	60	R	I	orchiectomy+RT	12	survived
24	Bostanci[23]	68	L	I	orchiectomy	12	survived
25	Damle[24]	68	R	I	orchiectomy	6	survived
26	Hmida[25]	78	R	II	orchiectomy	24	survived
27	Rana[14]	50	L	I	orchiectomy	4	survived
28	Suvarchala[26]	19	L	I	orchiectomy	15	survived
29	Abdullazade[27]	49	L	I	orchiectomy	24	survived
30	González[28]	80	R	III	orchiectomy+CTx	3	death (liver, lung metastasis)
31	Oranusi[29]	61	R	I	orchiectomy	32	survived
32	Kammeyer[30]	3,5	R	I	orchiectomy	not mentioned	survived
33	Rajagopal[31]	70	R	not mentioned	orchiectomy+RT	not mentioned	survived (local recurrence)
34	Siraj[32]	27	L	I	orchiectomy	24	survived
35	Berg[33]	54	R	IV	orchiectomy+CTx	not mentioned	survived (pulmonary and subcutaneous metastasis)
36	Nazar[34]	53	L	I	orchiectomy	12	survived
37	Abdallah[14]	42	R	I	orchiectomy	12	survived
38	Joshi[15]	70	L	I	orchiectomy	3	survived
39	Current case	63	R	I	orchiectomy	24	survived

CTx: Chemotherapy; RT: Radiotherapy; LND: Lymph node dissection; LN: Lymph node.

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