



Primary Low Grade Intratesticular Leiomyosarcoma: Case Report and Review of the Literature

Primer Düşük Dereceli İntratestiküler Leiomyosarkom: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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ABSTRACT

A 49-year-old male presented with a painful mass in the left scrotum. An inguinal orchiectomy was performed. Pathological examination revealed a well-differentiated leiomyosarcoma completely located inside the testicular parenchyma. We report this unusual case because primary leiomyosarcoma of the testis proper is extremely rare; our patient being the 19th case recorded thus far in the medical literature. It can lead to significant clinical and diagnostic difficulty due to its wide differential diagnosis and extreme rarity.

Key Words: Testis, Leiomyosarcoma, Sarcoma, Malignant mesenchymal tumor

ÖZ

49 yaşında erkek hasta sol skrotumda ağrılı şişlik ile başvurmuştur. İnguinal orşiektomi gerçekleştirilmiş, patolojik incelemede tamamen testis parankimi içine lokalize iyi diferansiye bir leiomyosarkom saptanmıştır. Primer intratestiküler leiomyosarkom son derece nadirdir ve hastamız şu ana kadar tıp literatüründe sunulan bu tanıya sahip 19. olgudur. Söz konusu antite, nadir olmasının yanı sıra, önemli klinik ve patolojik tanısal güçlüklerle yol açabilme potansiyeli nedeniyle sunulmaktadır.

Anahtar Sözcükler: Testis, Leiomyosarkom, Sarkom, Malign mezenkimal tümör

INTRODUCTION

Leiomyosarcoma (LMS) is a malignant mesenchymal neoplasm thought to have originated from smooth muscle. It accounts for approximately 7% of soft tissue malignancies. LMS's are generally seen in middle-aged and elderly patients, often located in the uterus or gastrointestinal organs and constitute the majority of sarcomas developing from large vessels in the retroperitoneum. Male genitourinary LMS's are seldom. Approximately one hundred paratesticular leiomyosarcomas have been reported in the literature (1). Primary intratesticular leiomyosarcoma is even more rare with only 18 cases known so far (2-18). Herein, we would like to document an additional case of primary intratesticular leiomyosarcoma with its clinical and pathological features.

CASE REPORT

A 49-year old white man presented with the complaint of left testicular enlargement and scrotal pain for 6 days. There was a tender hard mass palpated in the left testis during physical examination. Inguinal lymph nodes were not enlarged. Scrotal ultrasonography showed 35x33x28 mm,

hypoechoic, solid mass on the left testis while the right side was normal. Serum levels of LDH, α -fetoprotein and β -human chorionic gonadotropin were within the normal range. Thoracic and abdominal computed tomography scans did not reveal significant findings. A left radical orchiectomy with high ligation of the spermatic cord was performed.

Pathology: The testis was 5.5x3.5x3 cm in dimensions. When the organ was cut sagittally, a solid nodular mass, measuring 3.5 cm in diameter, was observed (Figure 1). Tumor was located in the center of the testis being unrelated to tunica albuginea, epididymis or spermatic cord. It was an unencapsulated but well-delineated rubbery lesion, homogeneous creamy white in color with fibrillary-whorled appearance. Hemorrhage or necrosis was not apparent grossly. The rest of the testis parenchyma and ductal structures were unremarkable. Under microscopical examination, the tumor was a mesenchymal neoplasm composed of spindle cells showing smooth muscle differentiation with characteristic cigar shaped nuclei and long eosinophilic cytoplasm (Figures 2, 3A-C). Neoplastic cells formed intersecting

fascicles. They displayed diffuse smooth muscle actin and desmin expression immunohistochemically (Figure 4A,B). Stains for pan-cytokeratin, inhibin, S-100, melan-A, HMB 45, CD34, CD68 and CD117 were negative. There was



Figure 1: A well-delineated intratesticular lesion displaying whorled appearance similar to leiomyomas.

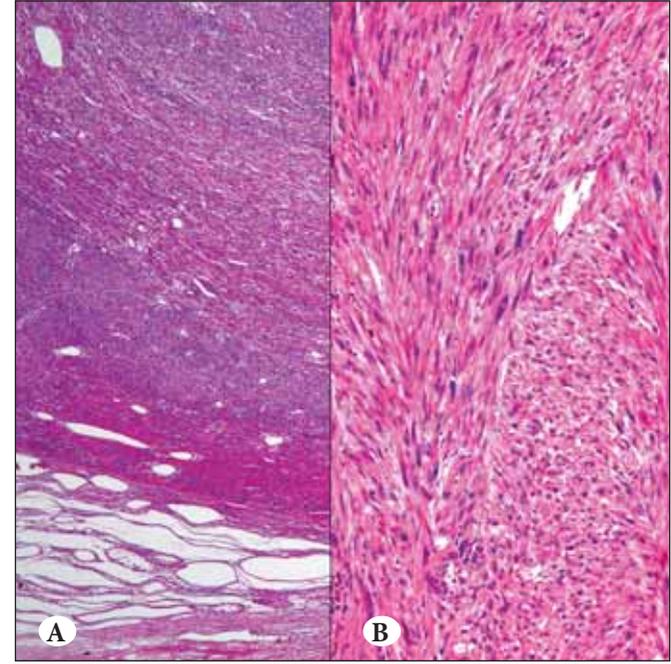


Figure 2: The neoplasm consisted of elongated cells forming intersecting fascicles. (A) H&E x40, (B) H&E x200.

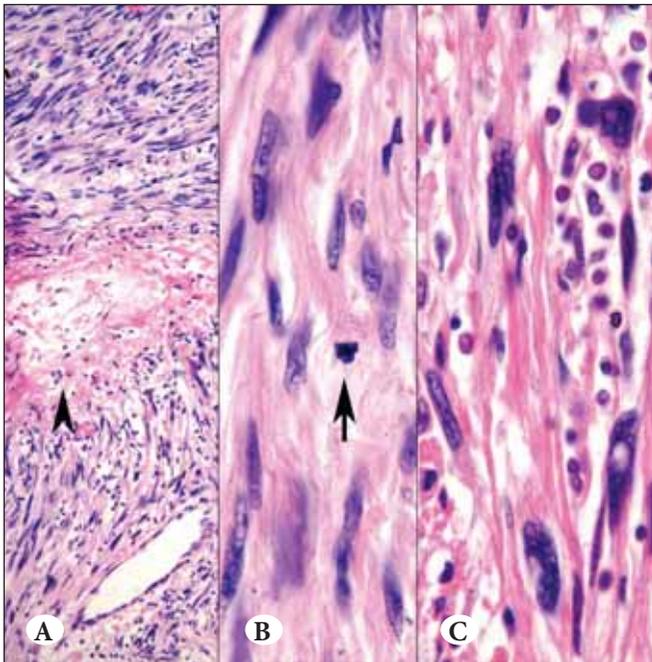


Figure 3: Smooth muscle tumor displaying features of malignancy. (A) Area of necrosis depicted by arrow-head (H&E x200), (B) Mitotic figure pointed by arrow (H&E x400), (C) Cellular pleomorphism (H&E x1000).

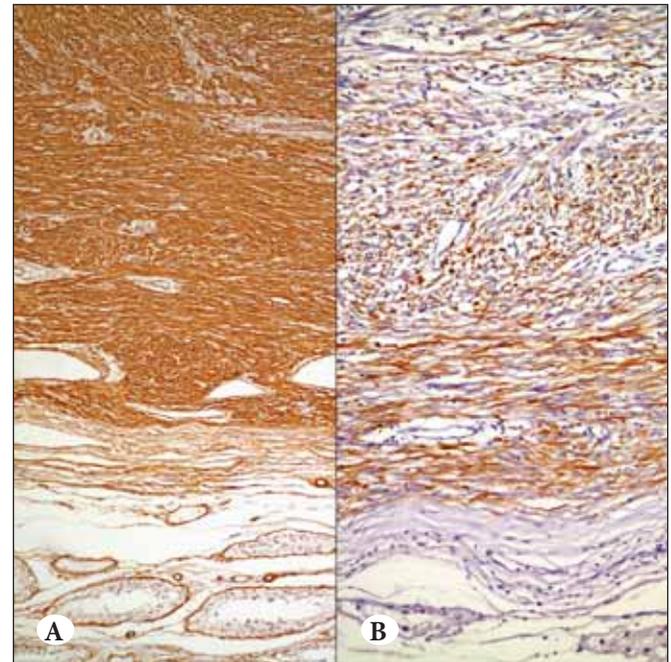


Figure 4: Neoplastic cells expressed smooth muscle actin and desmin diffusely. (A) Immunohistochemistry, anti-SMA Ab x100, (B) Immunohistochemistry, anti-desmin Ab x200.

leiomyosarcoma. There was no extratesticular tissue involvement; the rete testis, epididymis and spermatic cord were normal. All surgical margins were free for the tumor.

Follow-up: The patient did not receive any adjuvant therapy. He has been put on follow-up with abdominal USG and chest x-rays. He has no evidence of disease for 24 months after the operation.

DISCUSSION

Primary testicular mesenchymal neoplasms are distinctly rare. Sarcomas constitute only about 1% of all malignant tumors of the organ. Leiomyosarcoma (LMS) is the third most frequent primary soft tissue sarcoma after malignant fibrous histiocytoma and liposarcoma in testis. However, almost all testis LMS's are paratesticular, which originate from the spermatic cord, epididymis or scrotum. LMS arising in the testis-proper is extremely uncommon, with only 18 cases reported so far (Table I). It is thought to derive from the smooth muscle elements of the testicular parenchyma, such as blood vessels or contractile cells of

the seminiferous tubules (2,8). In their etiology, external radiation therapy (10), high dose anabolic steroid use (4) and chronic inflammation (6) have been associated with intratesticular leiomyosarcoma. At the end of detailed investigation and questioning, no risk factor could be identified in our patient.

The characteristic features of all reported intratesticular leiomyosarcomas, including our case have been summarized in Table I. All patients, except an 8-month infant, were adults with a wide age range from 19 to 77 years, more than 50% being above 40. Tumor size varied from 1.7 cm to 23 cm in the largest diameter (mean 80.5 mm, when including our case). There is no side predilection (9 right versus 9 left, one unknown). Its clinical presentation does not differ from that of other testicular malignancies. Diagnosis is achieved by histologic and immunohistochemical findings showing smooth muscle differentiation. Almost all of the patients presented as stage I disease, and the majority had good prognosis without recurrence or distant metastasis although the follow-up periods are short. They can be considered

Table I: Intratesticular leiomyosarcomas reported in the literature including the present case

Cases	Age	Laterality	Largest dimension of tumor (mm)	Follow-up	Outcome
Yachia , et al. (2)	55	R	45	2 yr	No R/M
Washecka , et al. (3)					
Case 1	47	R	48	6 yr	No R/M
Case 2	40	R	40	6 yr	No R/M
Froehner, et al. (4)	32	R	17	6,5 yr	No R/M
Hachi, et al. (5)	70	NS	230	14 mo	Pulmonary metastasis, death
Ali, et al. (6)	65	R	120	1 yr	No R/M
Sattary, et al. (7)	27	L	45	2,5 yr	No R/M
Singh , et al. (8)	26	L	26	NA	No R/M
Wakhlu , et al. (9)	8 / 12	L	230	1 yr	No R/M
Canales, et al. (10)	30	R	40	0,5 yr	No R/M
Takizawa, et al. (11)	76	L	74	1 yr	No R/M
Borges, et al. (12)	19	L	70	16 mo	Retroperitoneal metastasis
Kumar, et al. (13)	65	R	85	0,5 yr	No R/M
Yoshimine, et al. (14)	73	L	200	9 mo	Lung, para-aortic lymph nodes, spleen, muscle, subcutaneous tissue and vertebra metastasis
Raspollini, et al. (15)	77	L	40	1 yr	No R/M
Tobe, et al. (16)	71	R	NA	7 mo	No R/M
Labanaris, et al. (17)	73	R	35	28 mo	No R/M
Giridhar, et al. (18)	55	L	70	11 mo	Soft tissue and bone metastasis
Current case	49	L	35	24 mo	No R/M

R: Right testis; L: Left testis; NS: Not specified; yr: Year(s); mo: Months; R/M: Recurrence and/or metastasis; NA: Data not available.

as tumors of low malignant potential. Nevertheless, the biological behavior of these tumors is not easy to predict and four patients developed metastatic disease (5,12,14,18). High tumor grade is the common feature among the metastasizing sarcomas. All four have been stated as having high mitotic rate although in only 2 cases numeric counts have been indicated (2/10 hpf (14) and 8/10 hpf (18)). Two out of four were notably bulky, 20 cm (14) and 23 cm (5) in the largest diameter. Yet, many of the non-metastasizing indolent intratesticular leiomyosarcomas have also been reported as high-grade lesions with numerous mitotic figures (e.g. as much as 40-50/10 hpf in the tumor reported by Kumar et al. (13)). The accumulated cases constitute such a small number that it is difficult to reach a conclusion about their behavior by morphology alone. According to Folpe and Weiss, any mitotic activity in a deeply seated smooth muscle tumor with nuclear atypia should be considered as a marker for potential malignant behavior (1). Radical orchiectomy followed by surveillance is the treatment of choice in today's practice. Our patient, who had a low grade and a small tumor, treated as such has been under follow-up for 24 months without the stigmata of recurrent disease or distant spread.

When these tumors are intratesticular, their pathologic diagnosis can be intricate. The other more common malignancies must be considered and excluded from the differential. It is known that somatic malignancies including sarcomas may arise within teratomas. Additionally Yolk sac tumors and spermatocytic seminomas may have a sarcomatous component. Spindle cell morphology can be observed in some sex cord stromal tumors as well. Through tumor sampling during pathologic examination to show up all elements of the tumor is critical to avoid misdiagnosis.

Gross and microscopic features of intratesticular LMS are identical to those encountered elsewhere. Like our case, they are solid, firm, white masses on the cut surface. Under the microscope, neoplastic cells are spindled with elongated blunt ended nuclei and variably eosinophilic cytoplasm. They form interlacing bundles and sweeping sheets. Positive immunohistochemical staining with smooth muscle actin and desmin support the smooth muscle origin. Cytologic pleomorphism, mitosis and foci of necrosis, and invasion into surrounding tissues are the features of malignancy.

Herein, we have documented the 19th case of primary intratesticular leiomyosarcoma. We believe that such rare cases must be reported in the literature, so that sufficient data will accumulate in time, which may clarify the disease process and guide correct management of patients in the future.

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