

## CASE REPORT

# Leiomyosarcoma of the spermatic cord

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## ABSTRACT

Leiomyosarcoma of spermatic cord is very rare. The patient is 65-year-old man who presented with a tumor in scrotum. Excision of tumor was performed and it showed a solid tumor measuring 4 cm × 3 cm × 3 cm in left spermatic cord. Histologically, it showed proliferation of hypercellular atypical spindle cells with hyperchromatic nuclei. The tumor cells were arranged with fascicles of cigar-shaped spindled cells with scant acidophilic cytoplasm. The tumor was not encapsulated, and mild invasive features into surrounding tissue were noted. The mitotic index was 17; 17 mitotic figures were seen in 10 high-power-fields. Atypical mitosis was also seen. Immunohistochemically, tumor cells were positive for vimentin,  $\alpha$ -smooth muscle actin, h-caldesmon, desmin (focal), p53, and Ki-67 (labeling index = 34%). No other tumors were identified in the body, and the patient further treated with auxiliary local radiation and chemotherapy (cisplatin and doxorubicin). He is now healthy and free from tumors in the body 15 months after the operation.

**Key Words:** Spermatic cord, Leiomyosarcoma, Histopathology, Immunohistochemistry

## 1. INTRODUCTION

Smooth muscle tumors are classified into the following three types: leiomyoma, smooth muscle tumor of uncertain malignant potential, and leiomyosarcoma.<sup>[1]</sup> Leiomyoma contains various histological variants such as cellular leiomyoma.<sup>[1-3]</sup> Smooth muscle tumors of the peritesticular areas are very rare.<sup>[2]</sup> Adenomatoid tumor, mesothelioma, melanotic neuroectodermal tumor, desmoplastic small round cell tumor, and mesenchymal tumor including smooth muscle tumors occur in peritesticular areas.<sup>[2]</sup>

Leiomyosarcoma of spermatic cord is very rare; a PubMed search revealed about 100 cases in the world literature and only 43 cases in the English literature. The most recent study also reported that there are only about 115 cases of leiomyosarcoma of the spermatic cord in the world literature.<sup>[4-7]</sup> This pathological entity in spermatic cord was first

reported by Wessel in 1953.<sup>[8]</sup> All of these reported cases of leiomyosarcoma in spermatic cord are case reports, and there have been no comprehensive studies using many cases. In addition, there have been relatively a small number of case reports with an immunohistochemical studies in leiomyosarcoma of the spermatic cord.<sup>[4,9]</sup>

## 2. CASE REPORT

A 65-year-old man noticed a tumor in scrotum. A blood laboratory test revealed no significant changes. Tumor markers were within normal ranges. Scrotal palpation and echo images revealed an intrascrotal tumor measuring 4.3 cm × 3.4 cm × 3.2 cm, and whole body CT identified no tumors other than the scrotal tumor. The first choice of clinical diagnosis was seminoma. Resection of tumor with wide margins were performed, and it showed a solid, well-defined tumor measuring 4 cm × 3 cm × 3 cm in left spermatic cord (see

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Figure 1). Histologically, the tumor of spermatic cord was composed totally of hypercellular atypical spindle cells with hyperchromatic nuclei (see Figure 2). No areas of epithelioid differentiation or myxoid differentiation were recognized. The tumor cells were arranged in fascicles of cigar-shaped spindled cells with scanty relatively acidophilic cytoplasm (see Figure 2). Nucleoli and possible apoptotic figures were often seen (see Figure 3). Giant cells with bizarre nuclei were recognized in a small number. The tumor was not encapsulated, and mild invasive features into surrounding tissue were recognized. There were many mitotic figures. The mitotic index was 17; 17 mitotic figures were seen in 10 high-power-fields (HPF). A few atypical mitotic figures were also seen. No necrotic areas were recognized. Collagen histochemical stains revealed a few collagen fibers in the tumor. There were no obvious features of lymphatic or vascular permeation. The surgical margins were negative for tumor cells.

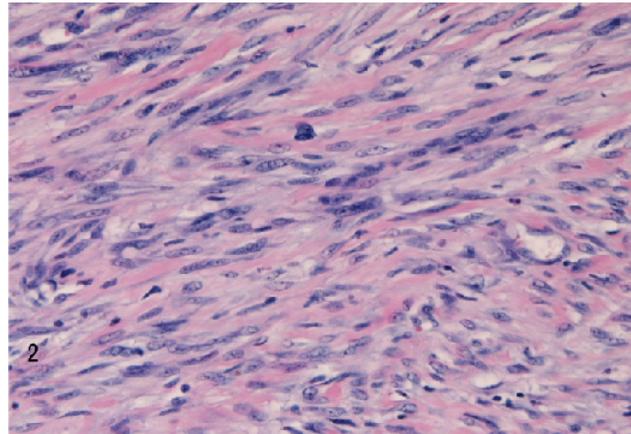


**Figure 1.** Gross pathologic findings of the resected left testis and left spermatic cord. A solid round tumor (left, arrow) measuring 4 cm × 3 cm × 3 cm is present in the left spermatic cord. The tumor was cut, and the cut surface shows solid white tumor. The tumor is well-defined and round.

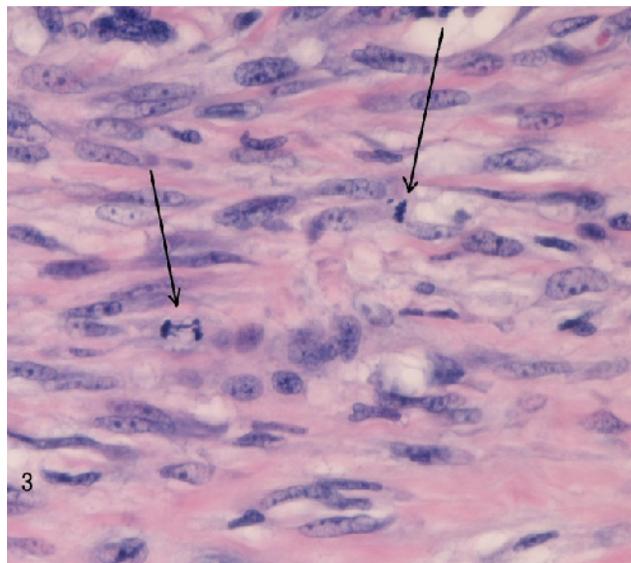
An immunohistochemical analysis was performed using Envision method.<sup>[10-13]</sup> The tumor cells were positive for vimentin,  $\alpha$ -smooth muscle actin, h-caldesmon (see Figure 4), desmin, p53, and Ki-67 antigen (labeling index = 34%). The tumor cells were negative for pancytokeratin AE1/3, pancytokeratin CAM5.2, KIT (CD117), CD34, S100 protein, and myoglobin. A pathological diagnosis of low-grade leiomyosarcoma of left spermatic cord was made by the author.

Postoperative various imaging modalities also revealed no tumors or lymphadenopathy in the body. The tumor was

in stage 1. Because no standard treatment regimen was available leiomyosarcoma of spermatic cord, the patient was treated according to NCI cancer treatment of uterine leiomyosarcoma at stage 1. Besides, the patient was treated with auxiliary local radiation (30 gray) and chemotherapy (cisplatin and doxorubicin). He is now healthy and free from tumors 15 months after the operation.



**Figure 2.** Histologic findings of the spermatic cord tumor. Low power view. The tumor is composed of cellular atypical spindle cells with hyperchromatic nuclei. HE, ×40.

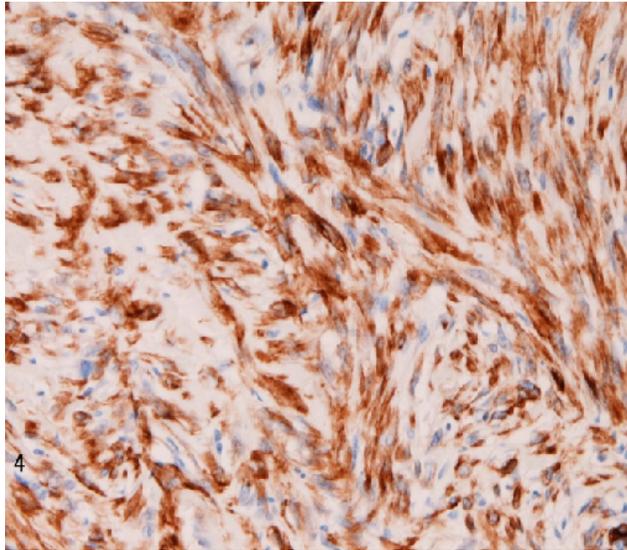


**Figure 3.** High power view. The tumor is composed of cigar-shaped atypical spindled cells with scant cytoplasm. Most tumor cells have prominent nucleoli. Relatively giant cells with bizarre nuclei are seen. Abundant mitotic figures (arrows) are seen in the tumor. HE, ×400.

### 3. DISCUSSION

Smooth muscle tumors are defined as benign and malignant neoplasms demonstrating smooth muscle differentiation.<sup>[1]</sup> Histologically, smooth muscle differentiation is shown by

spindle-shaped cells with cigar-shaped blunt nuclei and relatively acidophilic cytoplasm,<sup>[1]</sup> although there is an epithelioid variant.<sup>[1-3]</sup> Immunohistochemical demonstration of muscle-specific actin (HHF-35),  $\alpha$ -smooth muscle actin, desmin, and h-caldesmon indicates smooth muscle differentiation.<sup>[1-3]</sup> The current case fulfills this definition.<sup>[1-3]</sup>



**Figure 4.** Immunohistochemical findings. The tumor cells were positive for h-caldesmon. Immunostaining,  $\times 200$ .

Smooth muscle tumors are currently classified into the following three groups based on biological behaviors: leiomyoma, smooth muscle tumors of uncertain malignant potential, and leiomyosarcoma.<sup>[1]</sup> The differential diagnosis and evaluation of malignancy of these categories is occasionally difficult.<sup>[1-3]</sup> This difficulty is due to the fact that malignant potentials varies from one to another and are different among smooth muscle tumors in organs bearing them.<sup>[1-3]</sup> For example, smooth muscle tumors of superficial parts tend to be benign, while those of deep locations malignant.

Several histological parameters are used to evaluate malignant potential in smooth muscle tumors.<sup>[1-3]</sup> They include tumor size, cellular atypia, hypercellularity, mitotic index, atypical mitosis, coagulative necrosis, location of tumor, and tumor invasion, and metastasis. To evaluate malignant potentials, general cumulative evaluation of these parameters is essential. The large tumor size tends to be malignant, while small tumor size benign. The cellular atypia also reflects malignant potentials. The most reliable features of malignant potential is mitotic index (MI), which is defined as the number of mitotic figures per 10 HPF. In general, MI < 5 suggests a benign nature, MI of 5-10 suggests lesions inter-

mediate, and MI > 10 suggests a malignancy. In addition, the presence of atypical mitotic figures strongly suggests a malignant potential. Coagulative necrosis is an important parameter; its presence strongly suggests a malignant nature. In general, superficial and deep locations suggest benign and malignant characteristics, respectively. The invasive features suggest a malignant potential. The presence of remote metastasis indicates a malignant character, except for intravascular benign leiomyoma and benign metastasizing leiomyoma.<sup>[1]</sup> Immunohistochemical demonstration of p53 and high Ki-67 (MIB-1) labeling index suggests a malignant potential.

The present case showed cellular atypia, hypercellularity, giant cells with bizarre nuclei, prominent nucleoli, high MI, atypical mitotic figures, and mild invasive features, all of which strongly suggest malignant nature. The small tumor size (4 cm  $\times$  3 cm  $\times$  3 cm), absent coagulative necrosis, relatively superficial location, and negative metastasis may be parameter against the diagnosis of leiomyosarcoma. However, general cumulative evaluation of the parameters in the case is strongly in favor of leiomyosarcoma. Thus, the pathological diagnosis of leiomyosarcoma in the case seems correct. In the present tumor, the several negative parameters suggest that the leiomyosarcoma is relatively of low-grade.

The present study reported a very rare case of low-grade leiomyosarcoma of spermatic cord with an immunohistochemical study. Only about 115 cases of spermatic cord leiomyosarcoma have been reported in the world literature, and only about 43 cases in the English literature.<sup>[4-9]</sup> Thus, the present case seems important, because the present case adds a case in the lists of spermatic cord leiomyosarcoma. Immunohistochemically, the present tumor was positive for vimentin,  $\alpha$ -smooth muscle actin, h-caldesmon, desmin (focal), p53, and Ki-67 antigen (labeling = 34%), but negative for pancytokeratin AE1/3, pancytokeratin CAM5.2, KIT (CD117), CD34, S100 protein, and myoglobin. Positive expression of vimentin implies a mesenchymal tumor. Positive  $\alpha$ -smooth muscle actin, h-caldesmon and desmin almost indicate a smooth muscle differentiation. Positive p53 may indicate mutations of p53 gene, and suggest that disturbed p53-pathway play a part in the pathogenesis. High Ki-67 labeling index indicates high cell proliferative activity. Negative KIT excludes sarcomatoid small cell carcinoma, sarcomatoid germ cell tumor, and sarcomatous malignant melanoma.<sup>[7]</sup> Negative KIT and CD34 demonstrate that this case is not extra-gastrointestinal tumor.

## CONFLICTS OF INTEREST DISCLOSURE

The author declares no conflicts of interest.

**REFERENCES**

- [1] Hendrickson MR, Tavassoli FA, Kempson RL, et al. Mesenchymal tumours and related lesions. In: Tavassoli FA and Devilee P eds: World Health Organization Classification of Tumours. Pathology and genetics of tumours of the breast and female genital organs. Lyon: IARC Press; 2003. 254p.
- [2] Davis CJ, Sesterhenn IA, Woodward PJ, et al. Tumours of paratesticular structures. In: Eble JN, Sauter G, Epstein JI, Sesterhenn IA eds: World Health Organization Classification of Tumours. Pathology and genetics of tumours of the urinary system and male genital organs. Lyon: IARC Press; 2004. 267p.
- [3] Evans HL, Shipley J. Leiomyosarcoma. In: Fletcher CDM, Unni K, Mertens F eds. World Health Organization Classification of Tumours. Pathology and genetics of tumours of soft tissue and bone. Lyon: IARC Press; 2002. 131p.
- [4] Bozzini G, Albersen M, Romero Otero J, et al. Feasibility and safety of conservative surgery for the treatment of spermatic cord leiomyosarcoma. *Int J Surg.* 2015; 24: 81-4. PMID: 26578108. <http://dx.doi.org/10.1016/j.ijvsu.2015.11.012>
- [5] Dehghan A, Sami G, Eskandari N. Leiomyosarcoma of the Spermatic Cord in a Seventy-Five-Year-Old man. *Nephrourol Mon.* 2015; 7: e24308. <http://dx.doi.org/10.5812/numonthly.24308>
- [6] Soipi S, Vucic M, Ulamec M, et al. Leiomyosarcoma of the spermatic cord with scalp metastasis: case report and literature review. *Coll Antropol.* 2014; 38: 763-6. PMID: 25145020.
- [7] Kyratzi I, Lollis E, Antypa E, et al. Imaging features of a huge spermatic cord leiomyosarcoma: Review of the literature. *World J Radiol.* 2011; 28: 114-9. PMID: 21532872. <http://dx.doi.org/10.4329/wjrr.v3.i4.114>
- [8] Wessel HN. Leiomyosarcoma of the spermatic cord. *J Urol.* 1953; 69: 823-9. PMID: 13062396.
- [9] Dangle P, Basavaraj DR, Bhattarai S, et al. Leiomyosarcoma of the spermatic cord: case report and literature review. *Can Urol Assoc J.* 2007; 1: 55-8. PMID: 18542764.
- [10] Terada T. Pure sarcomatoid carcinoma of maxillary sinus and nasal cavity simulating malignant fibrous histiocytoma. *Am J Clin Pathol.* 2011; 135: 128-31. PMID: 21173134. <http://dx.doi.org/10.1309/AJCP4MEUHCJHU9X>
- [11] Terada T. Sarcomatoid squamous cell carcinoma of the vulva expressing smooth muscle actin and S100 protein. *Arch Gynecol Obstet.* 2011; 283: 1103-6. PMID: 20556408. <http://dx.doi.org/10.1007/s00404-010-1552-8>
- [12] Terada T. Sarcomatoid squamous cell carcinoma of the uterine cervix. *Arch Gynecol Obstet.* 2010; 282: 231-2. PMID: 20180127. <http://dx.doi.org/10.1007/s00404-010-1409-1>
- [13] Terada T. Sarcomatoid carcinoma of the lung presenting as a cutaneous metastasis. *J Cutan Pathol.* 2010; 37: 482-5. PMID: 19602061. <http://dx.doi.org/10.1111/j.1600-0560.2009.01292.x>