Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiation Oncology

Leiomyosarcoma of Prostate: Case Report and Literature Review

S. Chabli[1*], E. Mayoussi[1], R. Ousalm[1], M. Mekouar[2], B. Boutakioute[2], M. Ouali Idrissi[2], N. Cherif Idrissi Gannouni[2], M. Darfaoui[1], A. Elomrani[1], M. Khouchani[1]

[1]Department of Radiation Oncology, Mohammed VI University Hospital, Marrakesh, Morocco
[2]Department of Radiology Arrazi, Mohammed VI University Hospital Center, Marrakesh, Morocco

DOI: 10.36347/sjmcr.2023.v11i09.008 | **Received:** 17.07.2023 | **Accepted:** 24.08.2023 | **Published:** 04.09.2023

***Corresponding author:** S. Chabli
Department of Radiation Oncology, Mohammed VI University Hospital, Marrakesh, Morocco

Abstract **Case Report**

Prostate sarcomas originate from the mesenchymal tissues including smooth muscle, fibromuscular stroma, paraganglia, nerves, and blood vessels. They account for less than 0.1% of all prostate tumors and often present with obstructive symptoms. It is an aggressive malignancy with a high risk of metastasis and a poor prognosis that poses unique diagnostic and treatment challenges. Histopathological examination is essential for definitive diagnosis, and can be performed at an early stage using guided transrectal prostate biopsy. Since prostate specific antigen levels are generally normal, digital rectal examinations are extremely important. We report on a 51-year-old man who was diagnosed with leiomyosarcoma based on the histopathology of his transurethral prostatectomy specimen.

Keywords: Prostate sarcomas, fibromuscular stroma, metastasis, leiomyosarcoma.

INTRODUCTION

Sarcomas are a rare and heterogenous group of tumors of mesenchymal cell origin, accounting for less than 1% of adult malignancies [1]. The World Health Organization Classification of Tumors broadly divides sarcomas into two groups: soft tissue sarcomas (including fat, muscle, nerve, nerve sheath and blood vessels) and bone sarcomas [2]. Pelvic sarcomas are uncommon in these groups, representing 5% of soft tissue sarcomas and 10–15% of bone sarcomas [3-5].

Soft tissue sarcomas are rare and histologically heterogeneous mesenchymal tumors, usually with poor outcome. They represent 1-1.5% of all malignancies in adults. Approximately 10-15% of soft tissue sarcomas arise from the retroperitoneum [6]. Histologically, the most common types are liposarcoma, fibrosarcoma, rhabdomyosarcoma, leiomyosarcoma and angiosarcoma [7].

Leiomyosarcomas (LMS) are malignant lesions that develop from the smooth muscles of blood vessels, visceral organs and uterus. It is the predominant sarcoma originating from the large blood vessels. LMS is one of the more common types of soft tissue sarcoma to develop in adults. It should not be confused with leiomyoma, which is a benign tumor originating from the same tissue. LMS is an extremely rare type of cancer and can be very unpredictable. It can remain dormant for long periods of time and recur after years [8].

Here we present a unique case of prostate leiomyosarcoma and a literature review regarding the clinical and pathological features, diagnostic modalities, therapeutic aspects and prognosis of this rare entity.

CASE REPORT

Fifty-one-year-old male patient performed an urological consultation for moderate lower urinary tract symptoms, in particular nocturia and frequency associated to a complain of abdominal distension for 8 months. He reported no hematuria, weight loss, or perineal pain. The patient reported no history of tobacco use, exposure to hazardous chemicals, and had no family history of genitourinary cancers. On clinical examination, the presence of a hard hypogastric mass, fixed relative to the deep plane. On the digital rectal examination, the prostate was enlarged, nodular, integrating with the bladder base. Serum PSA was 1.6 ng/ml. A prostate biopsy was performed, the histopathologic examination demonstrated atypical cytology consistent with high-grade leiomyosarcoma (Figure 1). Immunohistochemical analysis revealed positive staining for vimentin, smooth muscle actin, desmin (partial), cytokeratin, smooth muscle myosin,

muscle specific actin, and Ki-67 (40% expression) (Figure 2).

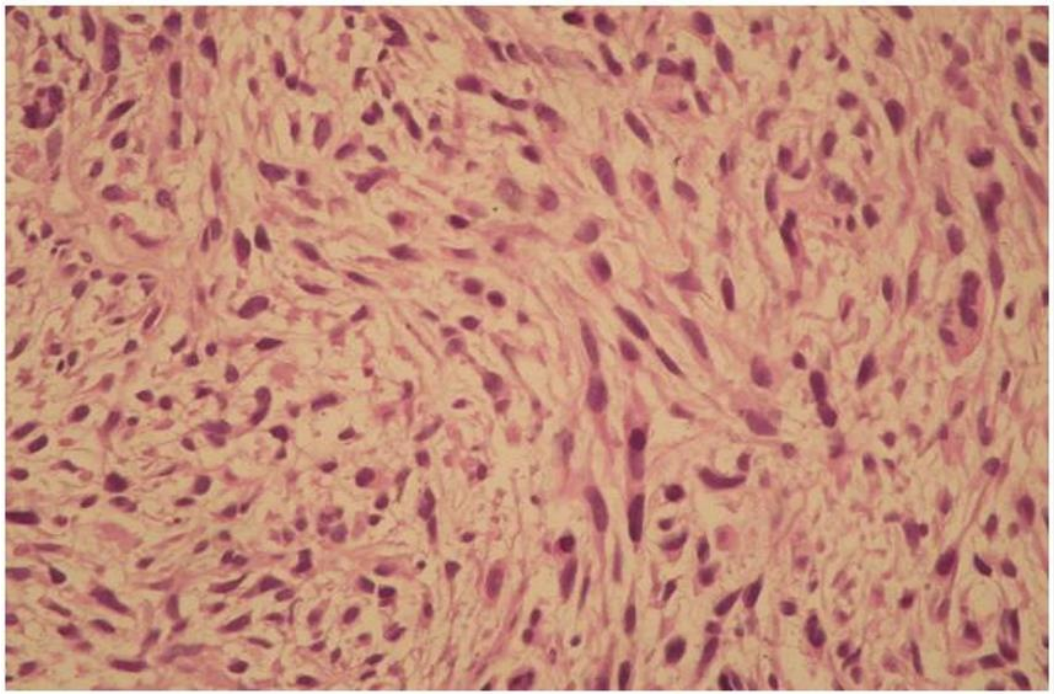

Figure 1: Leiomyosarcoma composed of dominant population of neoplasic spindle cells

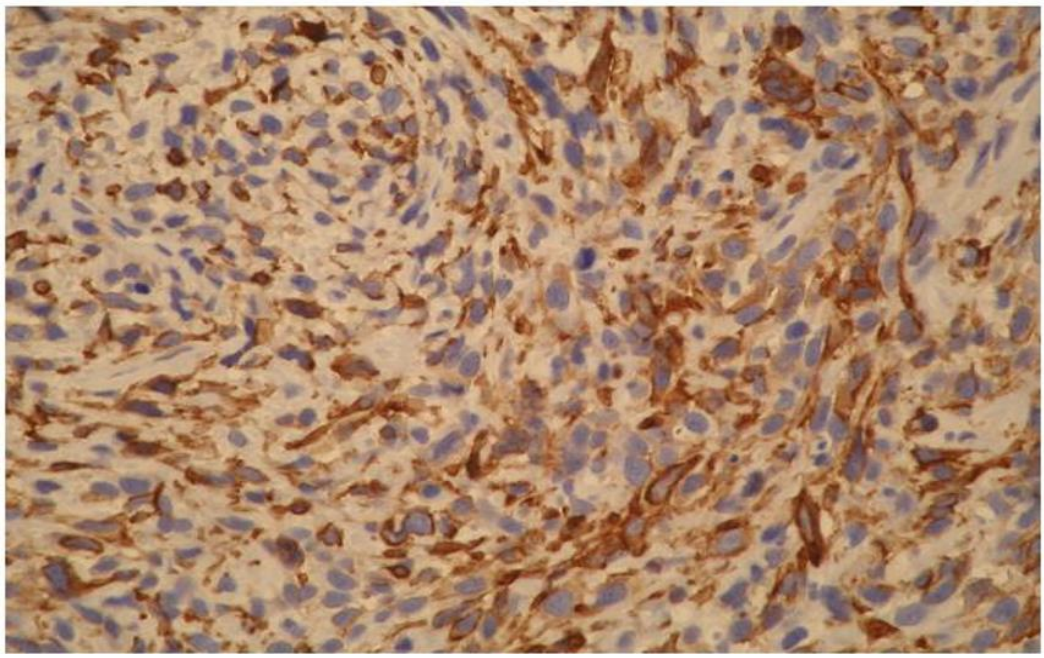

Figure 2: Immunohistochemistry demonstrates that tumor cells express smooth muscle actin

The pelvic MRI revealed presence of a solid-cystic centro-pelvic mass measuring 14x15x17cm polylobed, whose soft tissue portions are T1 hyposignal, T2 heterogeneous hypersignal and intensely enhanced after contrast injection with large areas of necrosis. The tumor appears to be centered on the prostate and seminal vesicles without being clearly individualized (Figure 3). Below, it fills the entire pelvic cavity, infiltrating the right levator ani muscle and extending into the homolateral ischioanal fossa. Above, it scallops the bladder, pushing it up with loss of the separating fat interface in places. It exerts a mass effect on both ureteral meats, with major bilateral upstream ureterohydronephrosis. Anteriorly, it is close to the anterior wall of the abdomen, with loss of the separating fat border. Posteriorly, it is in contact with the rectosigmoid hinge, which is laminated against the lumbosacral spine without any bone lysis (Figure 4).

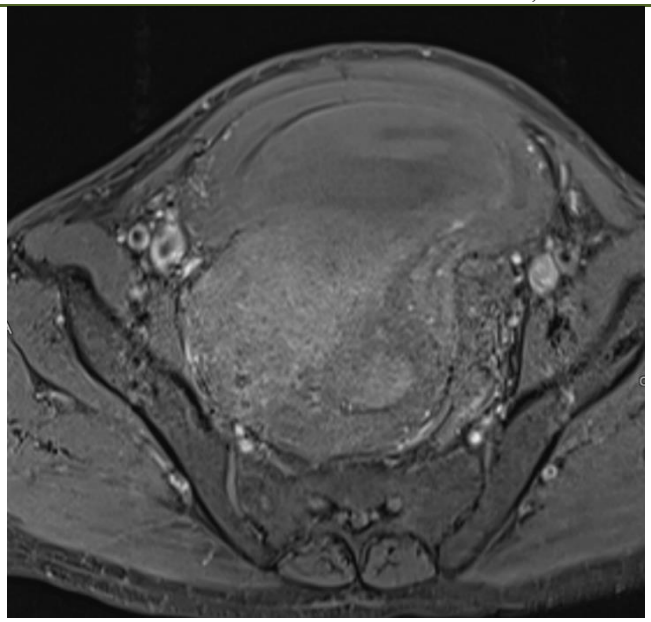

Figure 3: a) Abdominopelvic MRI: T1 axial after contrast injection

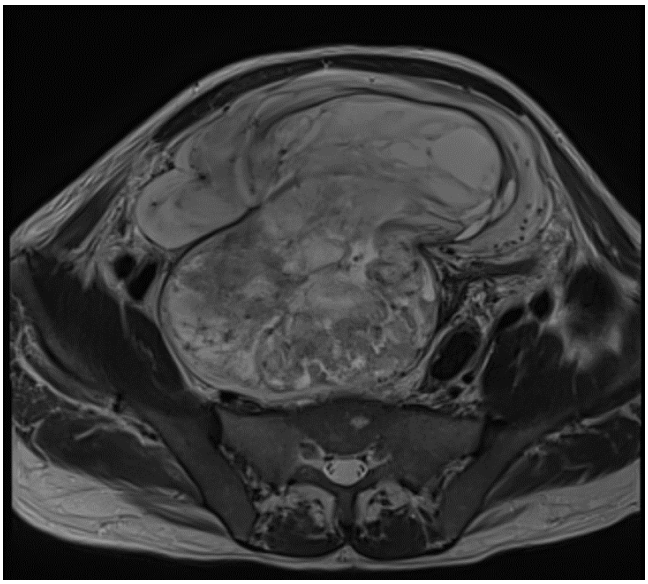

Figure 3: b) Abdominopelvic MRI: T2 axial

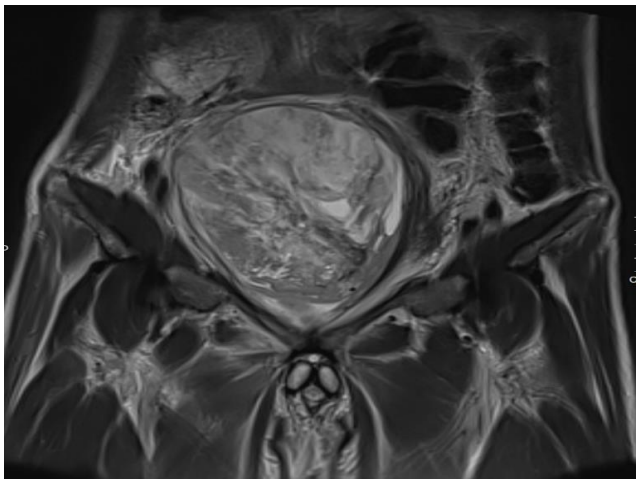

Figure 4: a) Pelvic MRI: T2 coronal

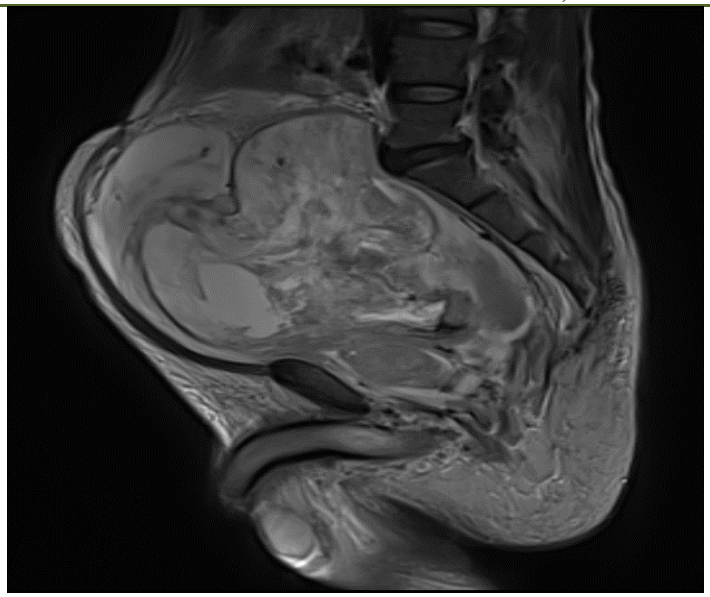

Figure 4: b) Pelvic MRI: T2 sagittal

Computed tomography (CT) of the chest and the brain was negative for metastatic disease. In view of the disease infiltrating into the surrounding structures and the expected morbidity, surgery was ruled out. In a multidisciplinary setting, the decision was to start with adriamycin-ifosfamide based chemotherapy and evaluate the tumour's progress. He received 6 cycles of ifosfamide (1,8g/m2) and adriamycine (60 mg/m2). A CT Scan done at his follow up at six months revealed a discreet increase in size of the centropelvic mass, with persistent local infiltration and major bilateral ureterohydronephrosis.

DISCUSSION

Prostate sarcoma is a rare malignant genitourinary cancer. According to different histological types, it can be divided into prostate leiomyosarcoma, rhabdomyosarcoma, fibrosarcoma and spindle cell sarcoma [7]. Patients range in age from 41 to 78 years at presentation with a mean age of 61 years. The lack of early specific symptoms results in more advanced disease at presentation. In fact, up to a third of patients have demonstrable metastases at presentation, usually to the lung, and sometimes to the liver as well [9, 10]. Individuals most commonly present with signs and symptoms of urinary obstruction. Additional associated symptoms include perineal pain, hematuria, burning on ejaculation, constipation and weight loss. Leiomyosarcoma can also present as an exophytic hypogastric mass of the prostate infiltrating the rectum or a perineal mass.

Serum PSA is typically within normal limits, attributable to the non epithelial origin of this disease. Abdominopelvic ultrasound would reveal hydronephrosis; thickening of the bladder wall or infiltration of the base of the bladder [11]. Transrectal prostate ultrasound is a useful technique to better assess the characteristics of the prostate. It can show heterogeneous hypoechogenic lesions in the prostate, an invasion of the capsule or an extension in the rectum, the pelvic sidewall, the seminal vesicle, or the ejaculatory duct [11, 12]. Thoracoabdomino-pelvic computed tomography or magnetic resonance imaging (MRI) can be used to assess the characteristics of the prostate lesion, its location, and its local extent. They can indicate whether or not there is lymphadenopathy or metastases in the abdomen or the thorax, and also allow the patient to be evaluated and followed up after treatment. Bone scintigraphy makes it possible to search for bone metastases.

Diagnosis is made by TRUS guided needle biopsy or transurethral resection in most patients, and less commonly by open surgical procedures [9, 10]. The vast majority of leiomyosarcomas are high-grade lesions with increased mitotic activity. Histologic features are fundamental in distinguishing this subtype of sarcoma. Leiomyosarcomas lack normal glands, as opposed to stromal sarcomas, or sarcomatoid carcinomas which often contain mixed malignant prostatic glands [14]. Regarding immunohistochemical profile of the tumor, most of the cases expressed vimentin, actin, progesterone receptor and CD34. S-100 and CD 117 are negative in all tumors.

Multimodality treatment regimens including surgery, radiotherapy and chemotherapy are recommended [9]. Radical retro-pubic prostatectomy is a good curative option for prostatic sarcoma. Among the different chemotherapy regimens, an anthracycline-based combination (doxorubicin or epirubicin) with alkylating agents (cyclophosphamide, ifosfamide) and vinca alkaloids (vinblastine or vincristine) or a platinum-based combination was used with mixed results [9, 12].

Overall prognosis for prostate leiomyosarcoma is poor, and 50%–75% of patients die of cancer within 2–5 years [9, 14]. Prognosis is improved in patients with no evidence of distant metastases at initial presentation and in those with localized disease in whom complete resection can be achieved surgically with microscopically negative margins [9]. Adverse prognostic factors include metastatic disease at presentation and the presence of positive surgical margins after surgery. Overall survival is very poor, and it is estimated that the 1-, 3-, and 5-year survival rates are 68%, 34%, and 26%, respectively [14]. However, some studies estimate the 5-year survival to be anywhere from 0 to 60% [9].

CONCLUSION

Prostatic leiomyosarcoma poses a unique diagnostic challenge, as clinical presentation alone may not always be suggestive of underlying malignancy. This challenge is further exacerbated by its aggressive nature, high risk of metastasis, and difficulties with unclear treatment. Proper history and physical examination, differential diagnosis, and a multidisciplinary approach to patient care are the foundation for early detection and promoting improved survival.

REFERENCES

1. Siegel, R. L., Miller, K. D., Fuchs, H. E., & Jemal, A. (2021). Cancer statistics, 2021. *Ca Cancer J Clin*, *71*(1), 7-33. doi:10.3322/caac.21654
2. WHO classification of tumours of soft tissue and bone. 5th ed. IARC Press, Lyon, France 2020.
3. Puchner, S. E., Funovics, P. T., Böhler, C., Kaider, A., Stihsen, C., Hobusch, G. M., ... & Windhager, R. (2017). Oncological and surgical outcome after treatment of pelvic sarcomas. *PloS one*, *12*(2), e0172203.
4. Wirbel, R. J., Schulte, M., & Mutschler, W. E. (2001). Surgical treatment of pelvic sarcomas: oncologic and functional outcome. *Clinical Orthopaedics and Related Research (1976-2007)*, *390*, 190-205. doi:10.1097/00003086-200109000-00022
5. Keyzer-Dekker, C. M., Houtkamp, R. G., Peterse, J. L., & Van Coevorden, F. (2004). Adult pelvic sarcomas: a heterogeneous collection of sarcomas?. *Sarcoma*, *8*(1), 19-24.
6. Pacelli, F., Pio Tortorelli, A., Rosa, F., Papa, V., Bossola, M., Sanchez, A. M., ... & Doglietto, G. B. (2008). Retroperitoneal soft tissue sarcoma: prognostic factors and therapeutic approaches. *Tumori Journal*, *94*(4), 497-504.
7. Nathan, H., Raut, C. P., Thornton, K., Herman, J. M., Ahuja, N., Schulick, R. D., ... & Pawlik, T. M. (2009). Predictors of survival after resection of retroperitoneal sarcoma: a population-based analysis and critical appraisal of the AJCC staging system. *Annals of surgery*, *250*(6), 970-976.
8. Letessier, E., Hamy, A., Bailly, J., Paineau, J., & Visset, J. (1992, January). Leiomyosarcomas of the rectum. Amputation of the rectum or local resection?. In *Annales de Chirurgie* (Vol. 46, No. 5, pp. 442-444).
9. Sexton, W. J., Lance, R. E., Reyes, A. O., Pisters, P. W., TU, S. M., & Pisters, L. L. (2001). Adult prostate sarcoma: the MD Anderson Cancer Center experience. *The Journal of urology*, *166*(2), 521-525.
10. Cheville, J. C., Dundore, P. A., Nascimento, A. G., Meneses, M., Kleer, E., Farrow, G. M., & Bostwick, D. G. (1995). Leiomyosarcoma of the prostate. Report of 23 cases. *Cancer*, *76*(8), 1422-1427.
11. Sastri, G. J., Mohanty, S. K., Munshi, A., Ghosal, S., Sharma, S. C., & Patel, F. D. (2002). Leiomyosarcoma of the prostate-a case report. *Indian Journal of Urology*, *18*(2), 171-172.
12. Chen K. C., Lin C. M., & Hsieh T. S. (2000). Leiomyosarcoma of the prostate. *The Journal of Urology*, 11, 39–42.
13. Venyo, A. K. G. (2015). A review of the literature on primary leiomyosarcoma of the prostate gland. *Advances in Urology*, *2015*.
14. Vandoros, G. P., Manolidis, T., Karamouzis, M. V., Gkermpesi, M., Lambropoulou, M., Papatsoris, A. G., ... & Konstantinopoulos, P. A. (2008). Leiomyosarcoma of the prostate: case report and review of 54 previously published cases. *Sarcoma*, *2008*.