Global Journal of Medical Research: I
Surgeries and Cardiovascular System
Volume 18 Issue 3 Version 1.0 Year 2018
Type: Double Blind Peer Reviewed International Research Journal
Publisher: Global Journals
Online ISSN: 2249-4618 & Print ISSN: 0975-5888

Multi-Recurrent Primary Leiomyosarcoma of the Seminal Vesicle: A Surgical Challenge

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Abstract- *Background:* Rare, primitive tumors of the seminal vesicle are often a poor prognosis. Moreover, the physiopathology remains misunderstood. Tumors are frequently classified as carcinomas and to a lesser extent as sarcomas. We present a challenging case of multi-recurrent primary leiomyosarcoma of the seminal vesicle surgically treated.

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Keywords: *leiomyosarcoma, grade 1, seminal vesicle, recurrent.*

GJMR-I Classification: *FOR Code: NLMC Code: WP 460*

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Multi-Recurrent Primary Leiomyosarcoma of the Seminal Vesicle: A Surgical Challenge

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One year later, follow-up showed on MRI a mass infiltrating the right side of the prostate's base and the bladder neck with two new nodular formations on the internal iliac chain.

A robot-assisted laparoscopic radical prostatectomy with tumor mass excision and extended lymph node dissection was performed and histopathological analysis of the specimen determined recurrence of leiomyosarcoma, grade 1 according to the FNCLCC.

Conclusion: Primary leiomyosarcomas of the seminal vesicle are exceedingly rare, and data on optimal treatment are lacking. Most of the time, a personalized treatment is proposed to the patient according to his characteristics and that of the tumor. This is a rare case of relapsing primary grade I leiomyosarcoma of the seminal vesicle. In the literature, cystoprostatectomy appears to be the treatment of choice for those tumors. This is the only case published in the literature of a recurrent primary grade I leiomyosarcoma of the seminal vesicle with up to 72-month follow-up. Early diagnosis and treatment are essential to improve the prognosis of this disease. Multimodal treatment should be discussed in a multidisciplinary approach.

Keywords: leiomyosarcoma, grade 1, seminal vesicle, recurrent.

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I. Introduction

Seminal vesicle is frequently involved by a contiguous spread of locally advanced malignancies from adjacent organs. However, primary neoplasms of the seminal vesicle are rare with primary adenocarcinoma being the most common (1). Primary leiomyosarcomas of the seminal vesicle are exceedingly rare with only eight cases published in the literature (2).

In addition, the lack of long-term monitoring data explains the fact that we do not have information on the best treatment. Most of the time, a personalized treatment is proposed to the patient according to his characteristics and that of the tumor. Therefore the bloc resection is the cornerstone of management. Here, we report a challenging case of multi-recurrent primary leiomyosarcoma of the seminal vesicle with a follow-up of more than six years, and we review the literature. We discuss as well the potential causes of recurrence and available treatment options.

II. Case Presentation

Back in 2011, the patient, a 58-year-old male, was referred to our department for an incidental right para-prostatic mass on an abdomen-pelvis computed tomography performed for unspecific abdominal pain. The patient had a history of high blood pressure, burnout, and a stable thoracic aortic aneurysm. To note, the patient had no LUTS and no family history of prostate cancer. Physical exam was unremarkable

nevertheless a slight asymmetry was noticed on digital rectal examination, but no nodule was palpable. The prostate specific antigen level was two ng/ml. To further characterize this mass, a multiplanar MRI was performed. MRI confirmed the origin of the mass from right seminal vesicle with no extension toward adjacent organ. A fluorodeoxyglucose positron emission tomography confirmed the presence of a 2.8 cm mass originating from the right seminal vesicle with a SUVmax of 74 and a transrectal ultrasound-guided biopsy was performed on October 2011. The biopsy revealed the presence of a spindle cell tumor with uniform cigar-shaped and slightly ovoid centrally located nuclei with an abundant granular eosinophilic cytoplasm. The immunohistochemical analysis shows a positive reaction for desmin, caldesmon and smooth muscle actin (SMA). The most likely diagnosed was benign leiomyoma. The patient has had surgical removal of the right seminal vesicle by robot-assisted laparoscopy. Pathological examination showed a grade I leiomyosarcoma of the seminal vesicle with negative margins. These results were further confirmed by anatomopathological revision of the slides by a pathology expert at a referral center in Massachusetts General Hospital-Boston USA. The patient did not receive adjuvant therapy based on a multidisciplinary decision and was closely monitored with both a multiplanar MRI and a fluorodeoxyglucose positron emission tomography. Thirty months after surgery, he presented evidence of recurrence on both imaging. He underwent a re-excision of the right peri-ureteral nodule and benefited from a right iliac lymph node dissection. The pathological examination revealed a grade I leiomyosarcoma of the right peri-ureteral nodule and the harvested lymph nodes were free of tumor. One year later, follow-up showed another evidence of recurrence. MRI showed a 17 mm mass infiltrating the right side of the prostatic base and protruding inside the bladder neck as well as the appearance of two new nodular formations on the internal iliac chain measuring 11 and 14 mm, respectively (figure 1-3). No distant metastases were seen on fluorodeoxyglucose positron emission tomography. A urethra-cystoscopy did not report any bladder endoluminal lesion but the protrusion of a slightly more right domed prostatic lobe. The measured PSA level was 1.03 ng/ml. After the Oncologic Multidisciplinary Committee approval, a robotic-assisted laparoscopic radical prostatectomy with tumor mass excision was performed. Intra-operatively, peritoneal and para-vesical and iliac nodules were present. Histopathological analysis of the specimen determined recurrence or metastasis of granulocyte leiomyosarcoma, grade 1 according to the FNCLCC (The French Federation of Comprehensive Cancer Centers), known in the patient (Figure 5).

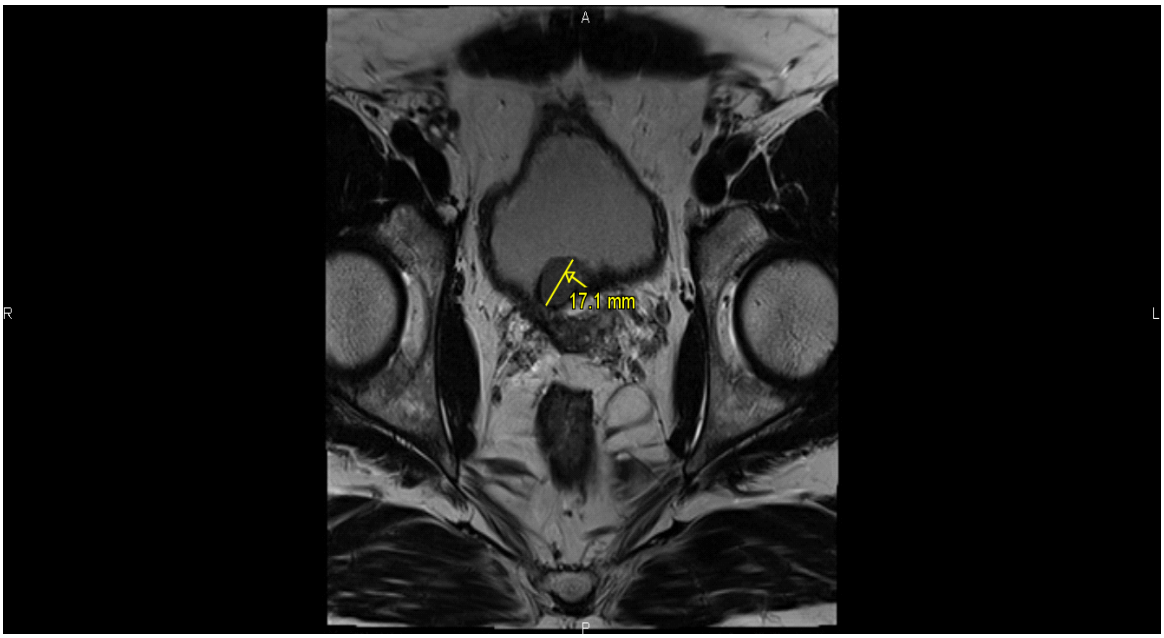

Fig. 1: The Resonance Confirms a Nodular Formation at the Level of the Bladder Floor just Opposite the Central Gland about 17 mm in Diameter (Cross Cut)

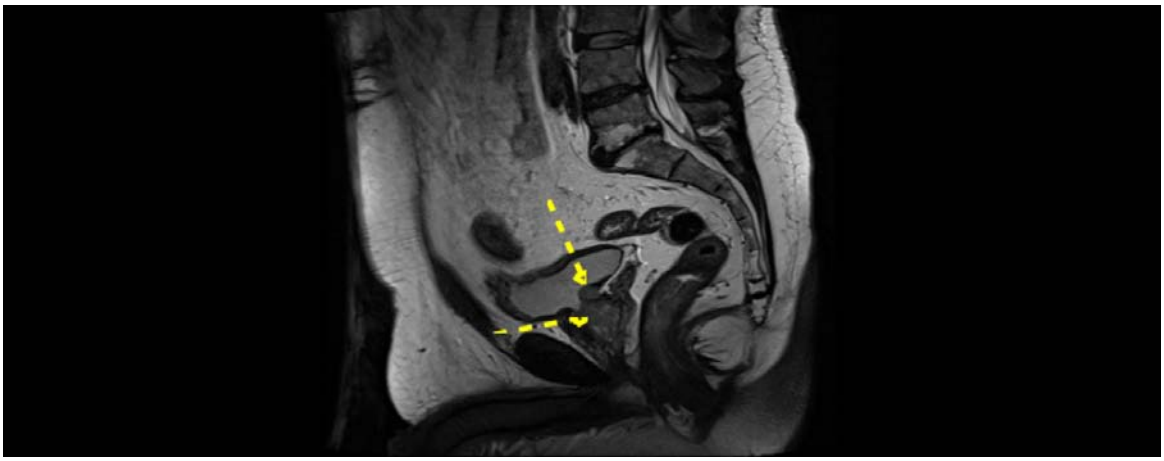

Fig. 2: The Resonance Confirms a Nodular Formation at the Level of the Bladder Floor just Opposite the Central Gland about 17 mm in Diameter (Sagittal Cut)

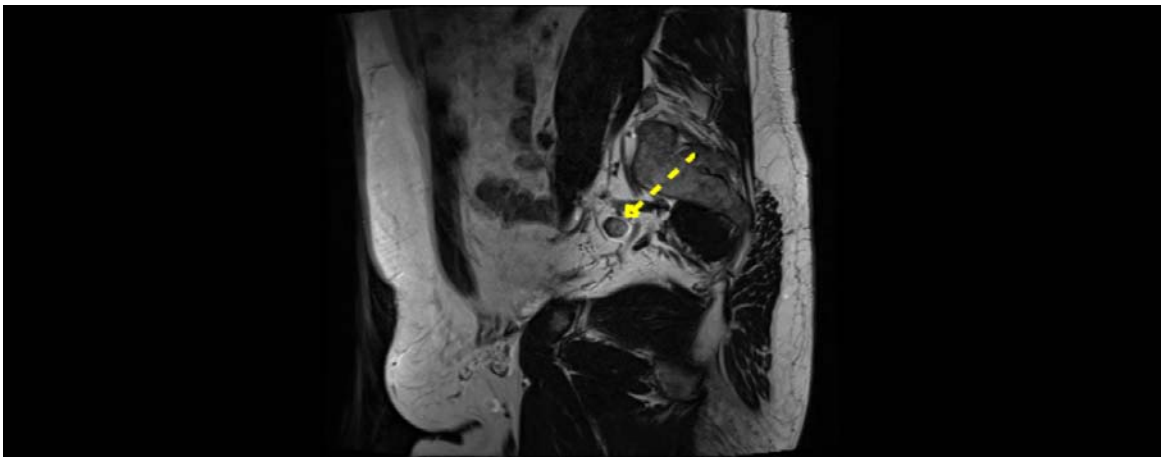

Fig. 3: The Resonance Confirms a New Nodular Formation on the Right Internal Iliac Chain (Sagittal Cut)

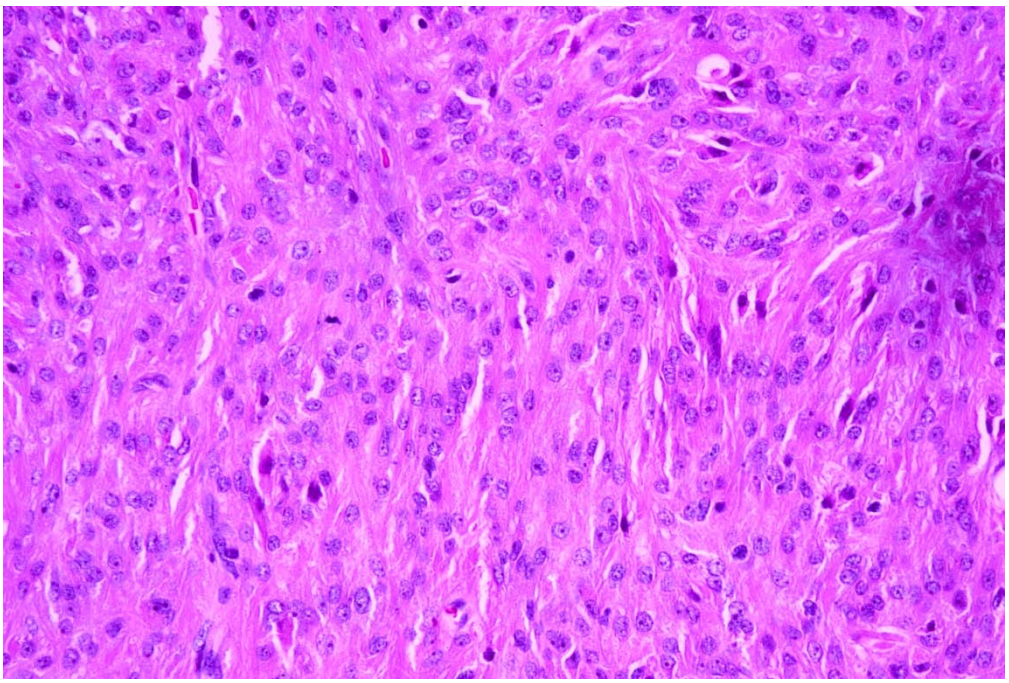

Fig. 4: Mixed Spindle and Epithelioid Tumoral Cells, with Abundant Eosinophilic Granular Cytoplasm, Moderate Atypia, and Low Mitotic Activity. (H & E X400)

III. Discussion

Primary tumor of the seminal vesicle is a rare entity and an exclusion diagnosis at the same time. According to Dalgaard and Giertsen (3), there must be no other demonstrable tumors present in the body because tumor invasion from adjacent organs or secondary localization is far more common that primary one. In our case, imaging, endoscopic work-up and transrectal ultrasound-guided biopsy of the prostate as well as the tumor showed no evidence of bladder and prostatic disease. Surgical resection of the lesion confirmed that a cleavage plan was present between the prostate and right the seminal vesicle where the tumor originates. Additionally, the PSA was mot high and the fluorodeoxyglucose positron emission tomography did not detect any other suspected localization in the body. Histopathological examination revealed grade 1 leiomyosarcoma originating from the seminal vesicle with a negative margin.

Primary malignant tumors of the seminal vesicle reported include carcinomas, sarcomas and an unusual group of tumors with mixed epithelial and stromal components (4). Although rare, carcinomas are more common than leiomyosarcomas (5).

A thorough review of the literature revealed eight cases of primary leiomyosarcoma of the seminal vesicle with a limited follow-up.

The table shows that primary leiomyosarcoma is more common in adults than in children, with the average age being 57 years. No specific risk factors have been identified.

Because tumor is asymptomatic, it is challenging to diagnose it. Most of the time, the discovery is fortuitous (by digital rectal examination, or radiologically) similarly to our case. Nevertheless, some patients report urinary and rectal symptoms often due to the size and extent of the tumor. No need to remember that no cases of hemospermia, hematuria or anejaculation have been reported.

Ultrasonography, CT, and MRI can all demonstrate the existence of a tumor mass although MRI being the most sensitive and specific in the pelvis. In this case, the image was strongly suggestive of a tumor originating from the right seminal vesicle. Tumor markers were negative. Our patient had a normal PSA.

The diagnosis is confirmed by histopathological examination of tissues collected by transrectal needle biopsy or by analysis of the surgical specimen. For our patient, a first diagnosis was made following the biopsies and was confirmed at a later stage by the analysis of the various operative pieces.

There are several staging of sarcomas. We used the FNCLCC classification because its performances are much better than the other classifications, regarding of reproducibility, performance and prognostic value (12, 13, 14). Grade 1 is considered to have a very low risk of recurrence and metastasis (12). The karyotype of our patient was normal so we could not integrate it into a familial leiomyosarcoma form, such as the hereditary leiomyomatosis and renal cell carcinoma (HLRCC) or Reed Syndrome in familial uterine leiomyosarcomas. The HLRCC syndrome is a rare autosomal-dominant condition caused by a mutation in the fumarate hydratase tumor suppressor gene.

The prognosis of seminal vesicle leiomyosarcoma is poor and unfavorable compared to other urological sarcomas from the bladder or para-testicular site (10). We could explain this by the delayed diagnosis due to the unusual form and paucisymptomatic character of this tumor, but also by the difficulty of complete surgical excision. Our case represents the one with interesting because it is the longest follow-up (72 months). We do not find any cases of recurrence for a grade 1 leiomyosarcoma in the literature. Nevertheless, patients with metastatic relapses have been reported suggesting that these sarcomas share prognostic features with other soft tissue sarcomas such as uterine leiomyosarcomas (15).

No standard or ideal treatment for seminal vesicle sarcoma has been established given the limited data in the literature. Nevertheless, through this case, we could conclude that Primary leiomyosarcomas is managed radically by surgery. Two surgical options were proposed: cystoprostatectomy with extended lymphadenectomy, or vesiculo-prostatectomy without cystectomy. There is insufficient data to clearly establish the superiority of one choice over the other. We opted for a vesiculo-prostatectomy because the patient is relatively young, the tumor had a grade 1, and the bladder was free from any lesion. Adjuvant radiotherapy may be used in the case of positive margins even though its role has not yet been clearly established (2,11).

Concerning adjuvant chemotherapy, its role in soft tissue sarcoma remains uncertain (16). The most widely used molecules are mesna, doxorubicin, ifosfamide, dacarbazine in combination (11), but some authors have used other anthracycline-based chemotherapy, gemcitabine and docetaxel, as seen with leiomyosarcomas from other sites (17).

IV. Conclusion

This case presents our experience with a multi-recurrent grade 1 primary leiomyosarcoma of the right seminal vesicle that until present revealed no signs of distant metastases, yet poses a surgical predicament. In this report we aim to support the critical significance of regular follow-ups of patients with grade 1 leiomyosarcoma, and aggressive local treatment in an attempt not to compromise long term oncologic outcome.

Table 1: Describing the Characteristics of the Nine Cases of Seminal Vesicle Leiomyosarcoma Reported in the Literature.

Author	Country	Age (Yrs)	Clinical Symptoms	Left / Right	Biopsy	Grade	Patho-logical Margins	Tum or Size	Adju-vent Treat-ment	Follow Up
Schned (6)	USA	69	None (RE)	Left	Yes	Inter-mediate	R0	3,5 Cm	No	14 Months: No Relapse, Alive
Wang (7)	USA	NA	None (RE)	NA	NA	High	R1	NA	No	24 Months: No Relapse, Alive
Wang (7)	USA	NA	None (RE)	NA	NA	High	R1	NA	No	29 Months: Metastasis (Lung), Alive with Disease (Doxorubicin)
Amirkhan (8)	USA	68	Rectal and Pain	Right	Yes	High	R0	10 Cm	No	13 Months: No Relapse, Alive
Muentene (9)	Switzerland	64	Urinary	Left	No	High	R0	8 Cm	No	24 Months: Metastasis (Kidney), Alive with Disease (CT)
Upreti (10)	India	46	Urinary and Pain	Right	Yes	NA	NA	NA	No	6 Months: No Relapse, Alive
Agrawal (11)	India	37	Urinary And Rectal	Right	Yes	Inter-mediate	NA	15 Cm	CT (MAID) + RT	20 Months: No Relapse, Alive
Cauvin (2)	France	59	Rectal	Right	Yes	Inter-mediate	R1	8 Cm	RT	29 Months: Metastases (Subcutaneous, Lung, Liver: MAI); 51 Months: Alive with Disease (Gemcitabine-Docetaxel)
Our Case	Belgium	58	Unspecific Abdominal Pain (CT)	Right	Yes	Low	R0	2,8 Cm	No	75 Mounts: Alive, Relapse + Metastases: Peritoneal Nodules

RE: rectal examination, NA: not available, CT: chemotherapy, MAID: mesna + doxorubicin + ifosfamide + dacarbazine, RT: radiation therapy.

Abbreviations:

PET-CT: Positron emission tomography computed tomography.
MRI: Magnetic Resonance Imaging.
FNCLCC: The French Federation of Comprehensive Cancer Centers.
LUTS: Lower urinary tract symptoms.
SUVmax: Standardized Uptake Value.
SMA: Smooth muscle acting.
PSA: Prostate-specific antigen.

Consent for Publication:

We obtained the written informed consent of the patient for the publication of this case report and accompanying images.

Availability of Data and Material:

All data and material are available.

Declaration of Interest:

The authors declare that they have no conflicts of interest in relation to this article.

Funding:
The authors have no funding.

Authors Contributions:
A.S: Wrote the article.
F.A, G.A, W.O, S.R, A.B, N.S, FX.O, S.S, T.G, E.H, K.L, T.R, A.P: Have read and corrected the article

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