Scientific Research Publishing

Open Journal of Gastroenterology, 2025, 15(2), 57-67
https://www.scirp.org/journal/ojgas
ISSN Online: 2163-9469
ISSN Print: 2163-9450

Case Report: Leiomyosarcoma Originating from the Gonadal Vein

Vithiarithy Chey[1,2], Neang Nov[1], Koung Tep[3], Vitou Leang[1], Reaksmey Chhay[1,4], Dimanche Chhit[1,2], Tharuom Ny[1,2], Keoseyla Unn[1]

[1]Faculty of Medicine, University of Health Sciences, Phnom Penh, Cambodia
[2]Gastroenterology Department, Khmer Soviet Friendship Hospital, Phnom Penh, Cambodia
[3]International University, Phnom Penh, Cambodia
[4]Thoracoabdominal Surgery Department, Khmer Soviet Friendship Hospital, Phnom Penh, Cambodia
Email: novserey@gmail.com

How to cite this paper: Chey, V., Nov, N., Tep, K., Leang, V., Chhay, R., Chhit, D., Ny, T. and Unn, K. (2025) Case Report: Leiomyosarcoma Originating from the Gonadal Vein. *Open Journal of Gastroenterology*, **15**, 57-67.
https://doi.org/10.4236/ojgas.2025.152006

Received: November 10, 2024
Accepted: February 23, 2025
Published: February 26, 2025

Abstract

Leiomyosarcoma of the gonadal vein is an exceedingly rare entity, representing a small subset of smooth muscle tumors that more commonly arise in the retroperitoneum, uterus, and blood vessels. To date, fewer than 10 cases of gonadal vein leiomyosarcoma have been reported in the literature, highlighting its rarity and the limited understanding of its clinical behavior. These tumors are often diagnosed incidentally or present with nonspecific symptoms, such as abdominal pain or a palpable mass, which can complicate early detection. The proximity of gonadal vein leiomyosarcomas to critical structures, such as the ureter, renal vessels, and surrounding organs, introduces unique diagnostic and surgical challenges. Previous reports have underscored the importance of advanced imaging techniques, including CT and MRI, in delineating the tumor's anatomical relationships and guiding surgical planning. This case, involving a leiomyosarcoma closely associated with the patient's left ureter, provides an opportunity to build on existing knowledge by addressing the clinical presentation, diagnostic approach, treatment pathway, and long-term follow-up strategies required for optimal management. By presenting this detailed review, we aim to contribute valuable insights into the diagnosis and management of this rare malignancy.

Keywords

Leiomyosarcoma, Gonadal Vein, Rare Tumor, Smooth Muscle Tumor, Diagnostic Challenges, Multidisciplinary Management

1. Introduction

Sarcomas are a heterogeneous group of tumors originating from mesenchymal

tissues, including bone, cartilage, muscle, and other connective tissues. Based on histopathological characteristics and primary tissue type, approximately 80% of sarcomas are classified as soft tissue sarcomas (STS), 15% as bone sarcomas, and 5% as gastrointestinal stromal tumors (GISTs) [1]. Leiomyosarcoma, a common subtype of STS, accounts for about 10% to 20% of all sarcomas [2] [3].

Leiomyosarcoma (LMS) is a rare malignant tumor of mesenchymal origin, specifically arising from smooth muscle cells [4]. Retroperitoneal LMS constitutes 50% of all soft tissue LMS cases [5]. While LMS can appear in hollow organs made up of smooth muscle, such as the stomach, bladder, uterus, blood vessels, and intestines, it is most commonly found in the uterus and abdomen [6]. Primary vascular LMS is very rare, comprising less than 2% of all LMS, with the vena cava being the most commonly affected vessel, accounting for 60% of vessel-involved LMS cases. LMS originating from the gonadal vein is particularly rare, with only about a dozen cases reported in the literature [7].

We present a case of this rare neoplasm in a patient managed through a multidisciplinary approach involving close collaboration. The primary treatment was surgical resection, followed by adjuvant therapy based on histology and immunohistopathology results.

2. Case Report

A 57-year-old woman presented at Khmer Soviet Friendship Hospital (KSFH) with intermittent pain in her right flank. She reported detecting an abdominal mass seven months earlier, along with a 12 kg weight loss and chronic constipation.

Lasting several months. Initially, she managed her intermittent abdominal symptoms at a private clinic, where an ultrasound identified an abdominal mass, and she was given medication to manage her symptoms. Despite this, her abdominal pain persisted, and she was unable to pass stool for four days. With no improvement, she sought care at KSFH's Gastroenterology Department. At KSFH, she underwent a colonoscopy, abdominal ultrasound, and CT scan, which revealed a mesenteric tumor encasing the left ureter. An ultrasound-guided biopsy showed spindle cell proliferation, suggesting a gastrointestinal stromal tumor (GIST) or a similar tumor type. After discussing the histological results, the patient opted for surgical treatment in KSFH's thoracoabdominal surgery department.

Clinical Examination

The patient appeared well, and alert, with normal conjunctiva color and no signs of anemia. An abdominal exam showed distension in the epigastric region and a non-tender, immobile mass with irregular borders in the left upper quadrant and flank. No additional nodules or hepatomegaly were detected. The abdomen was dull to percussion, with reduced bowel sounds in the left upper quadrant and normal sounds elsewhere. Laboratory tests, along with chest X-ray, abdominal X-ray, and colonoscopy, showed no significant abnormalities.

Abdominal Ultrasound

A hypoechoic, heterogeneous mass with polylobed contours was found on the

left side, measuring 116 mm × 94 mm × 115 mm. Conclusion: likely a tumor in the left flank (mesenteric or possibly left colon).

Abdominal CT scan with Contrast

A large, contrast-enhancing, heterogeneous mass was observed on the left paramedian side of the intra-abdominal cavity, measuring 110 mm × 129 mm × 128 mm, closely associated with the left ureter. There was fluid in the Douglas pouch but no signs of intestinal-colonic obstruction. The liver, pancreas, biliary system, spleen, adrenal glands and kidneys were normal. No significant lymph node enlargement or bone abnormalities were found. CT Conclusion: a probable retroperitoneal tumor located in the left para-median cavity near the left ureter, suggesting an ultrasound-guided biopsy would be beneficial (Figure 1, Figure 2).

Ultrasound-Guided Biopsy and Histology

Biopsy analysis revealed tumor proliferation in the submucosa, composed of spindle cells with eosinophilic or occasionally vacuolated cytoplasm, arranged in lobules or palisades and sometimes dissociated by skeinoid fibers. Moderate anisokaryosis and occasional mitoses were observed in the biopsy samples.

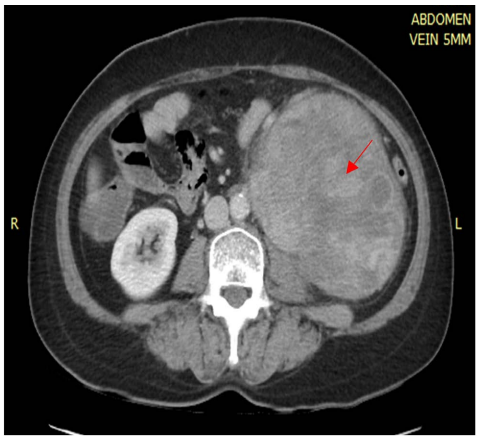


Figure 1. Retroperitoneal tumor Arrows: mass in the left para median intra-abdominal cavity.

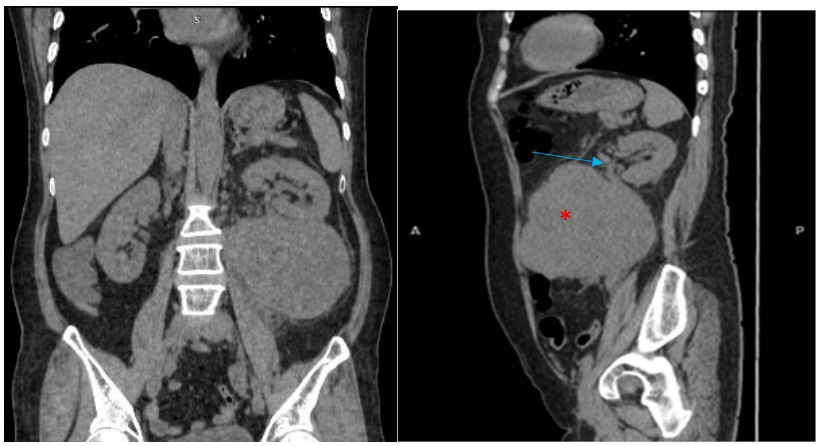


Figure 2. Retroperitoneal tumor Arrows: left ureter; *mass in the left para median intra-abdominal cavity in close contact with the left ureter.

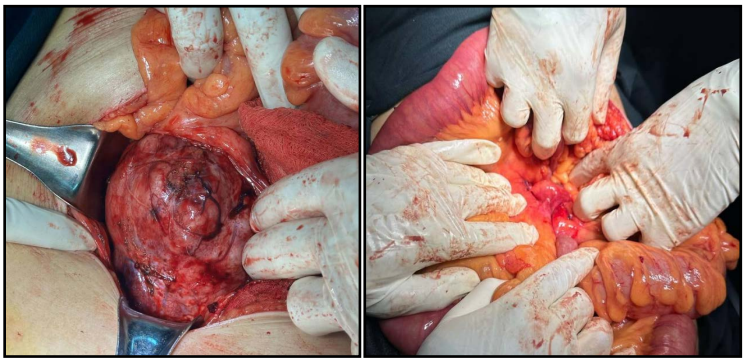
Conclusion

Histology findings suggested a retroperitoneal stromal tumor (such as GIST). Immunohistochemistry was recommended to confirm the diagnosis.

Surgical Management

During surgery, exploration revealed a tumor on the left side of the retroperitoneum, measuring approximately 12 cm × 13 cm × 13 cm. The tumor extended posteriorly to the transverse mesocolon, the superior mesenteric artery, and the jejunum mesentery along the left para-aortic area, and anteriorly near the left ureter without any adhesion. There was no evidence of distant metastasis.

Tumor resection was performed by first applying traction to the transverse colon, moving it anteriorly and superiorly. The tumor was found to originate from the left gonadal vein, and dissection across the transverse mesocolon was conducted. The tumor was carefully separated from the retroperitoneum, avoiding contact with the left ureter. The left gonadal vein and surrounding vessels were ligated and dissected, and hemostasis was confirmed. The tumor specimen was then removed from the surgical field. A limited wedge resection of the retroperitoneum was conducted, encompassing the tumor along with a sufficient negative margin of surrounding tissue. Preoperative diagnosis classified it as a resectable retroperitoneal tumor (gonadal vein), with staging T3N0M0 and a macroscopic R0 resection was achieved (Figure 3, Figure 4).



(a) (b)

Figure 3. Intraoperation presentation image (a) before tumor resection, (b) after tumor resection.

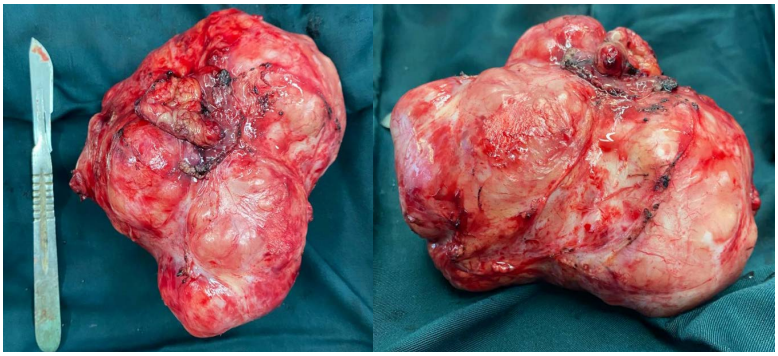
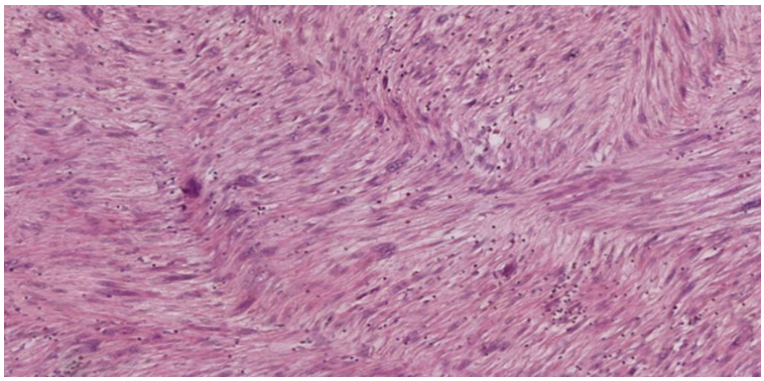


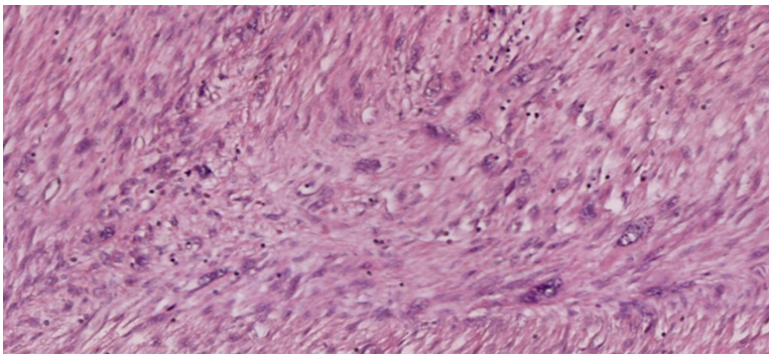
Figure 4. The specimen removed from the surgical field arrows left gonadal vein.

Histopathology

Histological and immunohistochemical analysis of the excised tumor confirmed leiomyosarcoma, measuring 15 cm, with 11 mitoses per 10 high-power fields. Evidence of coagulative tumor cell necrosis was present, with no lymphovascular invasion. Margins were confirmed clear, and immunohistochemistry showed that CD117 and Anti-Dog-1 were negative, Smooth Muscle Actin (SMA) was positive, and anti-Desmin showed partial positivity in the tumor cells (Figures 5-8).

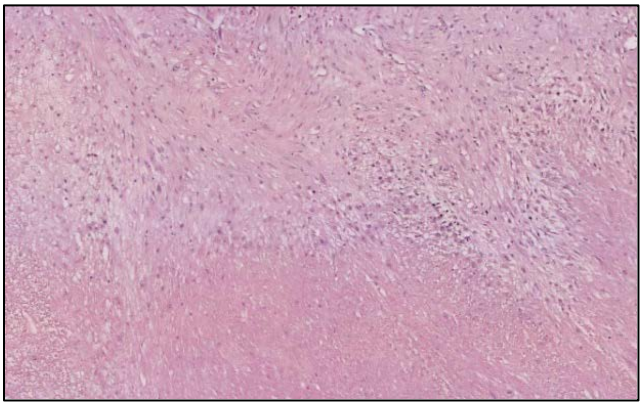


(a)

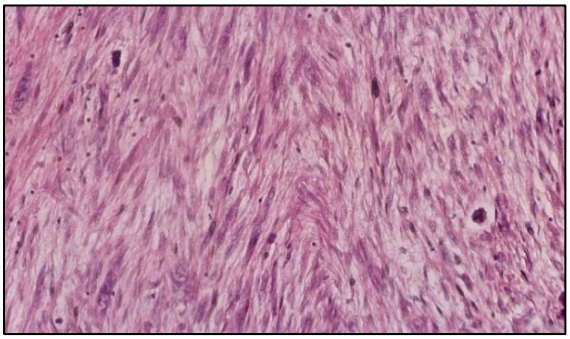


(b)

Figure 5. (a) and (b): High power view showing the tumor cells exhibit severe nuclear atypia with large hyperchromatic nuclei and prominent nucleoli.

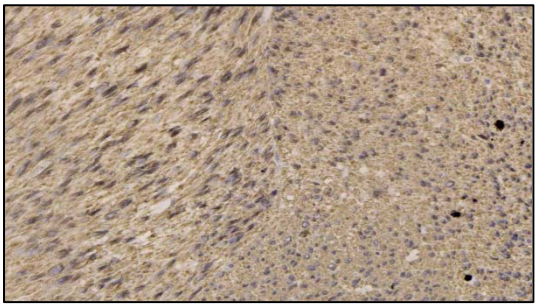


(a)

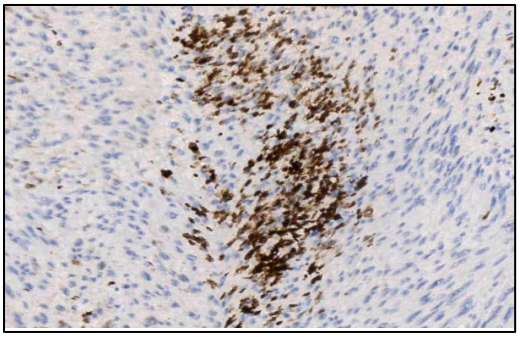


(b)

Figure 6. (a) Low power view of tumor cell necrosis with abrupt transition from viable tumor cells to necrotic cells. (b) High power view showing multiple atypical mitotic figure.

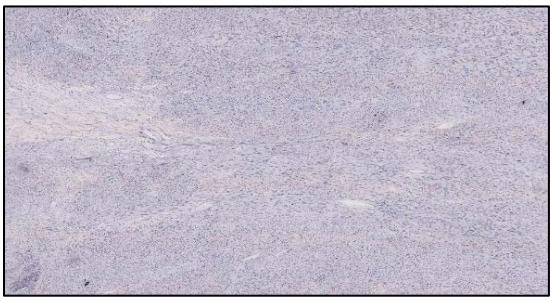


(a)

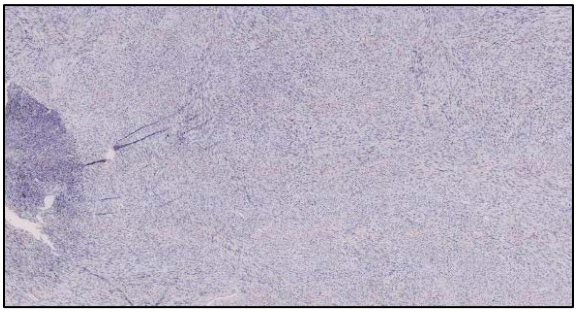


(b)

Figure 7. (a) High magnification showing antibody anti-SMA is diffuse positive for tumor cells. (b) Hight magnification showing antibody anti-Desmin is partially positive for the tumor cells.



(a)



(b)

Figure 8. (a) Low magnification showing antibody anti-C-kit (CD117) is negative for tumor cells. (b) Hight magnification showing antibody anti-Dog-1 is negative for tumor cells.

Discharge and Follow-Up

The patient began a regular diet on the second postoperative day and was discharged on the seventh. Given the tumor's high-grade nature, close follow-up with monitoring is essential. Imaging is critical for monitoring recurrence in retroperitoneal leiomyosarcoma cases, with abdominal-pelvic ultrasound, CT, or MRI recommended every 3 - 6 months for the first 2 - 3 years, every 6 months for the following 2 years, and annually afterward. Regular chest imaging, preferably with CT, is also advised.

3. Discussion

Incident

Soft-tissue sarcomas, including leiomyosarcoma (LMS), can originate in various anatomical locations, with the retroperitoneum being a significant site, accounting for 12% - 69% of cases. The incidence of LMS rises with age and peaks in the seventh decade of life. Retroperitoneal LMS and those arising from visceral blood vessels are more frequently seen in women, while LMS at other locations is more common in men. Leiomyosarcoma of the gonadal vein is exceptionally rare, with only ten cases reported in the literature [2] [4] [12].

Clinical Features

The clinical presentation of LMS depends on its size, location, and spread. In its early stages, LMS is often asymptomatic but can present with nonspecific symptoms such as fatigue, fever, weight loss, malaise, nausea, and vomiting. Pain is an uncommon feature, although swelling and a palpable mass are frequently observed. Retroperitoneal LMS, in particular, can grow to a substantial size before symptoms emerge, with nonspecific abdominal pain and the presence of a palpable mass being the most common findings. Imaging studies often identify large retroperitoneal tumors that compress or invade adjacent structures, potentially leading to metastases [13] [14] (**Table 1**).

In our case, a 57-year-old woman presented with abdominal pain and an abdominal mass, which aligns with the typical presentation of retroperitoneal LMS. Previous studies have highlighted the variability of symptoms in these tumors:

Noa de la Fuente *et al.* (2019) reported cases of chronic diarrhea, while Takuro Hirano *et al.* (2019) described patients with abdominal discomfort or no symptoms at all. These examples emphasize the broad spectrum of clinical presentations associated with LMS [9] [10].

Table 1. Summary table of retroperitoneal mass cases.

Author/Year	Basic Clinical Characteristics	Diagnostic Modality	Differential Diagnosis	Multidisciplinary Term	NeoTreatment	Histologic	Follow-Up
Vithiarithy CHEY *et al.*, 2024	57F; Abdominal pain, abdominal mass, weight loss; Left retroperitoneal mass (Left Gonadal vein)	CT, biopsy	GIST	Yes	No Surgical resection	G1	Follow-up
María Elena López-Ruiz, 2016 [7]	67F; Abdominal discomfort and pelvic mass; Left ovary (Left ovarian vein) 130 × 126 × 770 mm^3	Ultrasound	None	No	No Surgical resection + Chemotherapy (Adriamycin)	High-grade leiomyosarcoma	17-month survival, recurrence, and disseminated metastasis
Kazuomi Suzuki *et al.*, 2019 [8]	69F; Right abdominal pain; Right retroperitoneal mass (Right ovarian vein) 50 mm	Ultrasound, CT, MRI	Malignant lymphoma, leiomyoma, gastrointestinal stromal tumor	No	No Surgical resection and chemotherapy (doxorubicin + olaratumab)	-	5-month multiple lung metastases
Takuro Hirano *et al.*, 2019 [9]	71F; No symptoms; Right retroperitoneal mass (Right ovarian vein) 55 mm	Ultrasound, CT, MRI	Desmoid tumor, leiomyoma, LMS, and malignant mesothelioma	No	No En bloc excision	-	No signs of local recurrence or metastasis in 6 months
Noa de la Fuente *et al.*, 2019 [10]	51F; Chronic diarrhea; Right Gonadal vein 65 × 60 × 90 mm^3	CT, MRI	Neurogenic tumor, Mesodermal tumor	Yes	No En bloc resection	High-grade leiomyosarcoma (G2)	3-year survival, no recurrence
Giusy Carmen Imbriani *et al.*, 2023 [11]	44F; Abdominal pain, abdominal mass, hydronephrosis; Left retroperitoneal (Left gonadal vein) 119 × 90 × 110 mm^3	CT, biopsy	None	Yes	Yes Radical En bloc resection	High-grade leiomyosarcoma (G2)	-

LMS: Leiomyosarcoma; GIST: Gastrointestinal Stromal Tumor; CT: Computed Tomography Scan; MRI: Magnetic Resonance Imaging; G: Grade.

Diagnostic Challenges

Diagnosing LMS requires a combination of clinical evaluation, imaging studies such as CT, MRI, and PET scans, and tissue biopsy for histopathological confirmation. In some cases, fine-needle aspiration may aid in the diagnosis, while immunohistochemical testing is particularly valuable in distinguishing LMS from other tumors [6] [13] [15].

In this case, imaging revealed a left retroperitoneal tumor measuring 110 mm × 129 mm × 128 mm, encasing the left ureter but without distant metastasis. The tumor was specifically associated with the left gonadal vein. Similar findings have been documented in previous studies, including those by Noa de la Fuente *et al.* and Kazuomi Suzuki *et al.*, which noted tumor involvement in the retroperitoneal

space and gonadal veins. These findings highlight the critical role of advanced imaging and histological analysis in diagnosing retroperitoneal tumors [8] [9].

Histopathological Findings

Histologically, LMS is identified by nuclear atypia, high mitotic activity (exceeding 5 - 10 mitoses per high-power field), and areas of tumor necrosis. The classic histological pattern includes spindle-shaped cells arranged in interlacing fascicles, resembling smooth muscle tissue, with elongated, hyperchromatic nuclei and abundant eosinophilic cytoplasm [2]. Immunohistochemical markers such as SMA, Desmin, and h-Caldesmon are typically positive, while CD117 (KIT), a marker for gastrointestinal stromal tumors, is negative [1].

In our case, the postoperative histopathological analysis revealed a well-encapsulated tumor composed of spindle cells arranged in short fascicles. The tumor cells were uniform in size and shape, with elongated nuclei, moderate pale eosinophilic cytoplasm, 11 mitoses per 10 HPF, and areas of coagulative necrosis. There was no evidence of lymphovascular invasion, and the surgical margins were clear. Immunohistochemical analysis confirmed diffuse positivity for SMA and Desmin, while CD117 and Dog-1 were negative. These findings were consistent with a Grade 1 LMS based on FNCLCC grading criteria.

Therapeutic and Prognostic Considerations

The primary treatment for retroperitoneal LMS is complete surgical resection with negative margins, which is essential for reducing local recurrence, improving outcomes, and preventing distant metastasis [2] [12].

The prognosis, however, is often poor due to the tumor's high metastatic potential and the nonspecific symptoms that delay diagnosis. Retroperitoneal LMS is particularly challenging, as more than 50% of patients experience disease recurrence even after complete macroscopic resection. En bloc resection with clear margins offers the best outcomes, with a 5-year survival rate ranging from 33% to 68% [4] [10].

In our case, the patient underwent surgical resection with R0 margins, indicating complete removal of the tumor with clear margins, which is associated with a better prognosis. Treatment strategies for LMS are highly individualized. For example, Giusy Carmen Imbriani *et al.* (2023) described the use of neoadjuvant therapy before surgery, while Kazuomi Suzuki *et al.* (2019) reported cases where patients underwent en bloc resections or chemotherapy after surgery. These variations reflect the need for treatment plans tailored to the patient's clinical condition and tumor characteristics [8] [11].

Postoperative monitoring is crucial for detecting recurrences. According to the National Comprehensive Cancer Network (NCCN) guidelines, regular imaging of the abdomen and pelvis with CT or MRI is recommended every 3 - 6 months for the first 2 - 3 years, every 6 months for the next 2 years, and annually thereafter. Chest imaging, preferably with CT, is also advised [12] [13]. In this case, the low-grade nature of the tumor and the absence of metastasis indicate a favorable prognosis. However, long-term follow-up is essential. Previous studies have reported

varied outcomes: María Elena López-Ruiz (2016) documented early recurrences and metastases, while Kazuomi Suzuki *et al.* (2019) observed stable conditions post-surgery. These findings underscore the need for personalized follow-up strategies to optimize patient outcomes.

4. Conclusion

Leiomyosarcoma of the gonadal vein is an exceptionally rare type of mesenchymal tumor that typically presents as a retroperitoneal mass. The cornerstone of effective treatment is complete surgical excision with clear microscopic margins. However, achieving this can be challenging due to the tumor's location and potential involvement of surrounding structures. To optimize outcomes, a multidisciplinary team approach is essential, facilitating comprehensive treatment planning to reduce the risks of local recurrence, improve patient functionality, and minimize the likelihood of distant metastasis.

Patient Consent Declaration

The authors confirm that informed consent was obtained from the patient for the publication of clinical details and images. The patient was assured that identifying information, including her name and initials, would remain confidential and not be disclosed.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Damerell, V., Pepper, M.S. and Prince, S. (2021) Molecular Mechanisms Underpinning Sarcomas and Implications for Current and Future Therapy. *Signal Transduction and Targeted Therapy*, **6**, Article No. 246. https://doi.org/10.1038/s41392-021-00647-8

[2] Menon, G., Mangla, A. and Yadav, U. (2024) Leiomyosarcoma. StatPearls Publishing. https://www.ncbi.nlm.nih.gov/books/NBK551667

[3] Serrano, C. and George, S. (2013) Leiomyosarcoma. *Hematology/Oncology Clinics of North America*, **27**, 957-974. https://doi.org/10.1016/j.hoc.2013.07.002

[4] Matsuzono, T., Chan, C.Y. and Chan, M.Y. (2015) Gonadal Vein Leiomyosarcoma: A Case Report with Radiological Findings. *Intractable & Rare Diseases Research*, **4**, 152-154. https://doi.org/10.5582/irdr.2015.01016

[5] Bharti, J.N., Dey, B., Desai, P., Gupta, R., Khurana, N. and Gandhi, G. (2014) Primary Leiomyosarcoma of Peritoneal Cavity. *Rare Tumors*, **6**, 22-23. https://doi.org/10.4081/rt.2014.5165

[6] (2020) Leiomyosarcoma—NCI. https://www.cancer.gov

[7] López-Ruiz, M.E., Yébenes, L., Berjón, A. and Hardisson, D. (2016) Primary Leiomyosarcoma of the Ovarian Vein: Case Report and Literature Review. *International Journal of Surgical Pathology*, **25**, 339-343. https://doi.org/10.1177/1066896916679888

[8] Suzuki, K., Taniguchi, K., Ito, S., Serizawa, A. and Yamamoto, M. (2019) Leiomyo-

sarcoma Arising from the Right Ovarian Vein. *Journal of Surgical Case Reports*, **2019**, rjz302. https://doi.org/10.1093/jscr/rjz302

[9] Hirano, T., Okumura, H., Maeda, S., Shimada, M., Imakiire, A., Makisumi, K., *et al.* (2019) Vascular Leiomyosarcoma Originating from the Right Ovarian Vein: A Case Report and Literature Review. *Surgical Case Reports*, **5**, 1-6. https://doi.org/10.1186/s40792-019-0679-5

[10] de la Fuente, N., Rodríguez Blanco, M., Cerdán, G., Moral, A. and Artigas, V. (2019) Leiomyosarcoma of the Right Gonadal Vein: Review of the Approach and Prognostic in a Rare Case. *Case Reports in Surgery*, **2019**, Article ID: 4042689. https://doi.org/10.1155/2019/4042689

[11] Imbriani, G.C., Guarro, G., Di Somma, M.D., Sarno, I., Ardito, R., Botte, M., Fabrizio, T. and Scala, D. (2023) Gonadal Vein Leiomyosarcoma, from Clinical Practice to a Literature Review: Surgical, Oncological and Histopathologic Correlation. *Acta Biomedica*: *Atenei Parmensis*, **94**, e2023129.

[12] Marko, J. and Wolfman, D.J. (2018) Retroperitoneal Leiomyosarcoma from the Radiologic Pathology Archives. *RadioGraphics*, **38**, 1403-1420. https://doi.org/10.1148/rg.2018180006

[13] NORD: Leiomyosarcoma—Symptoms, Causes, Treatment. https://rarediseases.org

[14] Mulat, B.B., Haider, T. and Guddati, H. (2022) S2651 Retroperitoneal Leiomyosarcoma: An Unusual Cause of Gastrointestinal Bleeding. *American Journal of Gastroenterology*, **117**, e1752-e1753. https://doi.org/10.14309/01.ajg.0000867244.66257.d8

[15] Devaud, N., Vornicova, O., Abdul Razak, A.R., Khalili, K., Demicco, E.G., Mitric, C., *et al.* (2022) Leiomyosarcoma: Current Clinical Management and Future Horizons. *Surgical Oncology Clinics of North America*, **31**, 527-546. https://doi.org/10.1016/j.soc.2022.03.011