



Retroperitoneal Leiomyosarcoma: A Case Report

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Abstract

Background: Retroperitoneal leiomyosarcoma is a rare, aggressive soft-tissue sarcoma arising from smooth muscle cells. Due to its deep location and slow, silent growth, it often presents late with large tumor burden and involvement of adjacent structures. Diagnosis requires a combination of imaging and definitive histopathological evaluation.

Case Presentation: We report the case of a 52-year-old female who presented with abdominal pain, fullness, vomiting, and a palpable left-sided abdominal mass. Clinical examination revealed a firm, ill-defined mass in the left lumbar region. Contrast-enhanced CT demonstrated a large retroperitoneal soft-tissue mass encasing the left ureter, resulting in hydronephrosis. Ultrasound-guided trucut biopsy suggested a spindle cell neoplasm with smooth-muscle differentiation. Following surgical excision, histopathology confirmed a high-grade (FNCLCC Grade III) leiomyosarcoma, showing interlacing fascicles of pleomorphic spindle cells, brisk mitoses, and areas of necrosis. Immunohistochemistry was positive for Vimentin, SMA, and Caldesmon, supporting smooth-muscle origin. PET-CT revealed high metabolic activity in the primary lesion and metastatic right cervical and supraclavicular lymph nodes. Given the tumor's extent, retroperitoneal mass excision with left nephrectomy was performed. Postoperatively, the patient was planned for Ifosfamide- and Adriamycin-based chemotherapy.

Conclusion: Retroperitoneal leiomyosarcoma is a challenging malignancy due to late presentation and anatomical complexity. Accurate tissue diagnosis and timely surgical intervention remain the cornerstones of management.

Keywords: Retroperitoneal, leiomyosarcoma, Diagnosis, nephrectomy

Introduction

Soft tissue sarcomas (STSs) are a rare and heterogeneous group of malignant tumors that arise from mesenchymal stem cells capable of differentiating into muscle, fat, fibrous, and vascular tissues. [1] They account for less than 1% of all malignancies. Among these, leiomyosarcoma (LMS) represents approximately 5%–10% of all soft tissue sarcomas. [1] STSs most commonly occur in the extremities; however, nearly 40% are found in intra-abdominal or retroperitoneal locations. [2, 3]

Retroperitoneal sarcomas comprise about 10%–15% of all soft tissue sarcomas, with leiomyosarcomas being one of the predominant histological subtypes.

[4] Approximately half of all leiomyosarcomas arise within the abdominal or retroperitoneal region, while the uterus remains the most frequent primary site. [5] Due to their deep-seated location and indolent growth, retroperitoneal leiomyosarcomas often present late, when they have reached a considerable size or begun to exert pressure on adjacent structures. [6] Cross-sectional imaging, particularly computed tomography (CT), plays a crucial role in the initial assessment and surgical planning of these tumors; however, histopathological evaluation remains the gold standard for diagnosis.

Here, we report a case of a 52-year-old female presenting with a large retroperitoneal leiomyosarcoma, highlighting the diagnostic challenges, imaging findings, and management considerations associated with this rare malignancy.

Case Presentation

A 52-year-old female presented with a 1-month history of abdominal pain and fullness, accompanied by episodes of vomiting. The vomiting was non-bilious, non-projectile, contained food particles, and was typically followed by dull, aching abdominal pain. She also reported a 20-day history of a palpable lump on the left side of her abdomen. Loss of appetite was present; however, there was no significant loss of body weight.

Clinical Examination

On physical examination, a non-tender, smooth-surfaced mass was palpated in the left lumbar region, measuring approximately 7×4 cm. The lump had ill-defined margins, was hard in consistency, and was non-ballotable.

Radiological Assessment

CECT of the abdomen revealed a large, well-defined, lobulated, solid soft-tissue density mass measuring $9.5 \times 10.2 \times 10.5$ cm arising from the retroperitoneal mesentery of the left iliac fossa, just below the lower pole of the left kidney. Mild adjacent fat stranding was noted. The lesion abutted the lower pole of the left kidney and the anterior surface of the left psoas muscle while maintaining intervening fat planes. Medially, it encased the proximal and mid-left ureter, causing complete luminal obstruction with resultant upstream hydroureteronephrosis and loss of surrounding fat planes.

Figure 1:



Biopsy and Histopathology

An ultrasound-guided trucut biopsy of the retroperitoneal mass initially demonstrated features of a spindle cell neoplasm with smooth-muscle differentiation. Subsequent histopathological examination of the excised specimen revealed a unifocal retroperitoneal tumor measuring $15 \times 12 \times 8$ cm, with both the kidney and ureter free of tumor infiltration. Microscopy showed interlacing fascicles

of spindle cells with marked nuclear pleomorphism, abundant eosinophilic cytoplasm, brisk mitotic activity, and areas of coagulative necrosis, consistent with a Classic (Conventional) Leiomyosarcoma, Grade III (FNCLCC). Immunohistochemistry demonstrated strong positivity for Vimentin, Smooth Muscle Actin (SMA), and Caldesmon, while CD117 was negative. The combined morphological and

immunohistochemical findings confirmed the diagnosis of high-grade Leiomyosarcoma.

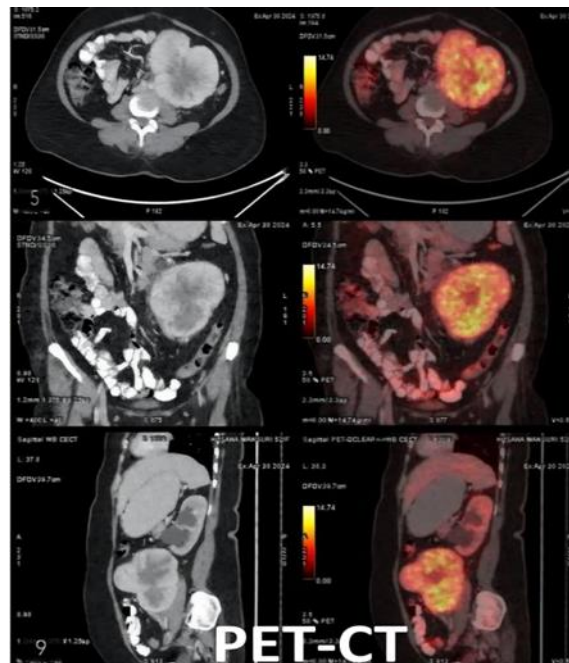
Figure 2:



PET-CT Findings

PET-CT demonstrated increased metabolic activity in the lobulated enhancing lesion (measuring 10.9×10.4 cm) located inferior to the left kidney. The mass encased the proximal and mid ureter, causing obstruction and consequent hydroureteronephrosis. The lesion was abutting the psoas muscle. Additionally, metabolically active right level IV cervical and right supraclavicular lymph nodes were noted suggestive of metastatic involvement.

Figure 3:



Surgical Findings

Exploratory laparotomy revealed a vascular mass measuring $10 \times 10 \times 10$ cm in the abdominal cavity with the mesentery of the large bowel adherent to it. The mass was encasing the left ureter and was closely abutting the left kidney, aorta, inferior vena cava, and left psoas muscle.

Given the extent of involvement, retroperitoneal mass excision along with left-sided nephrectomy was performed.

Management and Follow-up

The patient was advised to undergo Ifosfamide and Adriamycin based chemotherapy as part of postoperative management and to address the metastatic lymphadenopathy identified on PET-CT.

Discussion

Retroperitoneal leiomyosarcomas are typically slow-growing tumors and are often discovered incidentally, especially in asymptomatic individuals. [7] When symptoms do occur, they tend to be nonspecific and usually arise from the mass effect of the enlarging tumor, such as abdominal distension or early satiety. Rarely, these tumors may present with paraneoplastic hypoglycemia due to excess insulin-like growth factor 2 production. [8]

The mainstay of treatment is complete surgical excision with an intact capsule. Achieving clear margins often requires en bloc resection of the tumor along with adjacent organs most commonly the kidney or colon. Complete resection is the most significant factor influencing survival. Evidence shows that patients who undergo complete tumor removal have a markedly better median survival (103 months) compared to those with incomplete resection, whose outcomes are similar to non-operative management (18 months). [9]

In our case, women with a short history of abdominal pain, fullness, vomiting, and a palpable lump on the left side of her abdomen. Imaging studies revealed a large retroperitoneal mass encasing the left ureter and causing hydroureteronephrosis. Biopsy followed by detailed histopathological examination confirmed a high-grade (Grade III) leiomyosarcoma, characterized by spindle cells, marked pleomorphism, brisk mitotic activity, and areas of necrosis. Immunohistochemistry supported the diagnosis with strong positivity for

SMA, Vimentin, and Caldesmon. PET-CT demonstrated high metabolic activity within the mass along with metastatic involvement of cervical and supraclavicular lymph nodes. Surgically, the tumor was found to involve adjacent mesenteric structures and completely encase the ureter, requiring en bloc excision of the mass along with a left nephrectomy. Postoperatively, the patient was advised to undergo chemotherapy with Ifosfamide and Adriamycin.

Similar findings were reported by Amine Hermi et al., [5] who described a 59-year-old male with a large retroperitoneal leiomyosarcoma, emphasizing that RPL is a rare but predominantly malignant tumor in which complete surgical resection with negative margins remains the most effective treatment, though often technically challenging. Their study strongly supports the need for a multidisciplinary approach to improve survival outcomes.

Another report by T. Matsuoka et al., [7] highlighted diagnostic difficulty, where a retroperitoneal leiomyosarcoma was initially mistaken for ovarian cancer. The tumor showed poor response to gemcitabine–docetaxel chemotherapy due to its aggressive nature, and the absence of complete tumor resection resulted in a poor prognosis. Overall this case reinforce that early recognition, accurate diagnosis, and complete surgical excision offer the best chance for favorable outcomes in retroperitoneal leiomyosarcoma.

Conclusion

Retroperitoneal leiomyosarcoma is an uncommon and aggressive soft tissue tumor that typically presents at an advanced stage because of its deep location and vague clinical symptoms. This case emphasizes the diagnostic complexity of such lesions, where imaging is essential for assessing tumor extent, yet confirmation ultimately depends on histopathology and immunohistochemistry. It also reinforces the value of a multidisciplinary approach and highlights the critical role of early recognition and prompt treatment in improving patient outcomes.

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