ORIGINAL RESEARCH PAPER

Surgical Oncology

KEY WORDS:
Retroperitoneal sarcoma, Leiomyosarcoma, En bloc resection, Surgical oncology, Giant tumor

COMPREHENSIVE MANAGEMENT OF GIANT RETROPERITONEAL LEIOMYOSARCOMA

Dr. Ashok Galande	MBBS ,MS General Surgery , Mchonco -surgery
Dr. Navin Kasliwal	MBBS, MS Generalsurgery ,fellowship (Surgicaloncology)
Dr. Urvee Parekh	MBBS , MS General Surgery Junior Resident – Third Year
Dr. Anish Adak	MBBS , MS General Surgery Junior Resident-second Year

ABSTRACT

Retroperitoneal leiomyosarcomas are rare malignant mesenchymal tumors that often remain clinically silent until they attain significant size. They account for a small proportion of soft tissue sarcomas and are associated with diagnostic and therapeutic challenges due to their proximity to vital structures. We report the case of a 58-year-old male presenting with a progressively enlarging abdominal mass. Imaging revealed a large retroperitoneal tumor measuring approximately 27 × 16 × 14 cm, displacing adjacent organs without clear vascular invasion. Despite initial concerns regarding operability, the patient underwent successful en bloc resection of a massive tumor measuring approximately 30 × 25 cm and weighing 8–9 kg. Histopathological examination confirmed a low-grade leiomyosarcoma with spindle cell morphology and immunohistochemical positivity for SMA and Desmin. The postoperative period was uneventful, and the patient is currently undergoing adjuvant radiotherapy. This case highlights the importance of detailed imaging, multidisciplinary planning, and aggressive surgical management in achieving favorable outcomes, even in giant retroperitoneal tumors.

INTRODUCTION

Retroperitoneal sarcomas are rare tumors, accounting for approximately 10–20% of all soft tissue sarcomas. Leiomyosarcomas arise from smooth muscle cells and are known for their aggressive behavior and late presentation due to the expansive nature of the retroperitoneal space. Patients often remain asymptomatic until the tumor grows large enough to exert pressure on surrounding organs. Early diagnosis using imaging modalities and confirmation via histopathology are crucial. Complete surgical excision with negative margins remains the cornerstone of treatment.

Case Report

A 58-year-old male presented with a progressively enlarging abdominal lump in the right lower quadrant for three months. The onset was insidious, with no associated bowel or urinary complaints. There was no significant past or family history.

On clinical examination, a firm, non-tender, irregular mass measuring approximately 20 × 10 cm was noted in the right lumbar region. Contrast-enhanced CT (CECT) scan revealed a large heterogeneous soft tissue mass measuring 27 × 16 × 14 cm with focal calcifications. The mass was indenting the liver superiorly, extending up to the L4–L5 level inferiorly, and displacing the right kidney medially without invasion. It also displaced the ascending colon anteriorly and psoas muscle posteriorly. No vascular invasion was noted.

A core biopsy suggested a spindle cell neoplasm. Due to the size and complexity, the tumor was initially considered inoperable.

Surgical Management

The patient underwent exploratory laparotomy. Intraoperatively, a large encapsulated retroperitoneal mass measuring approximately 30 × 25 cm was identified. The tumor was adherent to the right kidney and ureter, extending into the subhepatic space and involving the diaphragm. Medially, it was encasing the inferior vena cava and was highly vascular.

Despite these challenges, complete en bloc resection of the tumor was achieved. The excised mass weighed approximately 8–9 kg.

The postoperative period was uneventful, and the patient was discharged on postoperative day 10. He is currently receiving adjuvant radiotherapy.

Histopathological Findings

Gross examination revealed three globular masses, the largest measuring 28 × 17 cm, with a firm whitish-yellow cut surface.

Microscopy showed spindle-shaped cells withnuclei and eosinophilic cytoplasm arranged in fascicles with fibrocollagenous stroma. Occasional bizarre cells were noted, with no significant necrosis or high mitotic activity. Immunohistochemistry showed:

- SMA: Positive
- Desmin: Positive
- S100: Negative
- Cytokeratin: Negative
- C-Kit: Negative

Findings were consistent with low-grade leiomyosarcoma (Grade 2).

DISCUSSION

RPL are slow growing tumors, diagnosed incidentally in asymptomatic patients or with nonspecific symptoms [3], usually secondary to the mass effect of the large tumor, such as early satiety or abdominal distension. Imaging modalities such as CECT are essential for assessing the extent of the tumor, its relation to surrounding organs, and the presence of metastases. Histopathology and immunohistochemistry remain the gold standard for diagnosis, helping to differentiate leiomyosarcomas from other spindle cell neoplasms such as gastrointestinal stromal tumors or malignant peripheral nerve sheath tumors.

Complete resection of RPL with intact capsule is the cornerstone of the management of this entity. En bloc resection of tumor with adjacent organ resection, most commonly the kidney and colon, is frequently needed to achieve complete resection [4]. It is the most important feature to improve the overall survival rate. Studies have

shown that the median survival of patients with complete resection was 103 months versus 18 months with incomplete resection, similar to non-operative treatment outcome [2]

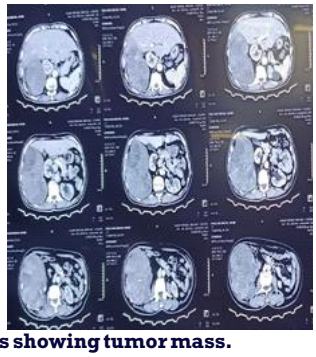
The potential benefit of neoadjuvant radiation includes decreasing tumor size, improving resectability, and improving local control. No studies have definitely demonstrated a benefit to preoperative radiation for RPL [5]. Some studies reported that the 5-year disease-free survival was 37% with an overall survival of 56% in patients treated with a dose of 50.4 Gy [5].

In patients undergoing an R0 resection, postoperative radiotherapy should be given to patients with high-grade tumors, extremely large tumors, and those with close margins. After an R1 resection, the NCCN recommends postoperative radiotherapy if neoadjuvant therapy was not given and a boost of 10–16 Gy if preoperative radiotherapy was given [5]. In case of locoregional recurrence (20–75% of patients, even if R0), surgery still remains the mainstay of treatment in the absence of distant metastatic disease [4].It is often more complicated. Multiple studies have also shown that the metastatic progression of the disease is associated with significantly lower rates for complete resection (70%). In case of unresectability, radiation therapy (if unifocal recurrence) or systemic therapy can be proposed [4].

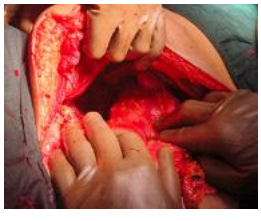
Despite successful surgical resection, retroperitoneal leiomyosarcomas have a high recurrence rate, and long-term outcomes depend on tumor grade and completeness of excision. Studies indicate a 5-year survival rate of 50–60% for retroperitoneal leiomyosarcomas. This case underscores the importance of a multidisciplinary approach involving surgeons, oncologists, and radiologists to optimize patient outcomes. The need for adjuvant therapies, particularly in high-grade tumors, is an area requiring further research.

CONCLUSION

Retroperitoneal leiomyosarcomas require a comprehensive and multidisciplinary approach. Even large tumors previously deemed inoperable can be successfully managed with meticulous surgical planning. Early diagnosis, complete excision, and appropriate adjuvant therapy are essential to improve patient outcomes.



CECT findings showing tumor mass.



1.Intraopertive extension of tumor upto liver
2.Specimen

REFERENCES

1. Choi JH, Ro JY. Retroperitoneal Sarcomas: An Update on the Diagnostic Pathology Approach. Diagnostics (Basel, Switzerland) [Internet]. 2020 Aug [cited 2024 Nov 27];10(9):642. Available from: https://pmc.ncbi.nlm.nih.gov/articles/PMC7555595/
2. Dull BZ, Smith B, Tefera G. Surgical management of retroperitoneal leiomyosarcoma arising from the inferior vena cava. Weber S J Gastrointest Surg 2013;17:2166–71. [DOI] [PMC free article] [PubMed] [Google Scholar]
3. Matsuoka T, Tsujioka H, Matsuoka S, Sorano S, et al. Retroperitoneal leiomyosarcoma: a case report. Eur J Gynaecol Oncol 2017;38:150–2. [PubMed] [Google Scholar]
4. Olimpiadi Y, Song S, Hu JS, Matcuk GR, et al. Contemporary management of retroperitoneal soft tissue sarcomas. Curr Oncol Rep 2015;17:39. [DOI] [PubMed] [Google Scholar]
5. Von Mehren M, Kane JM, Agulnik M, Bui MM, et al. Soft tissue sarcoma, version 2.2022, NCCN clinical practice guidelines in oncology. J Natl Compr Cancer Netw JNCCN 2022;20:815–33
6. Marko J, Wolfman DJ. Retroperitoneal Leiomyosarcoma From the Radiologic Pathology Archives. Radiogr a Rev Publ Radiol Soc North Am Inc [Internet]. 2018 Sep 1 [cited 2024 Nov 27];38(5):1403–20. Available from: https://pmc.ncbi.nlm.nih.gov/articles/PMC6166742/
7. Sun JN, Yang R, Jiang XL, Zhang F, Zhao HW. Giant retroperitonealliposarcoma with multiple organ involvement: a case report and literature review. BMC Nephrol [Internet]. 2024 Dec 1 [cited 2024 Nov 27];25(1):281. Available from: https://pmc.ncbi.nlm.nih.gov/articles/PMC11363381/