

Large Retroperitoneal Leiomyosarcoma

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Abstract

Retroperitoneal sarcomas, such as leiomyosarcoma, often invade or displace vital organs in the abdominal cavity and exhibit an aggressive clinical course. Complete surgical resection of the tumor with en-bloc resection of adjacent adherent organs is the cornerstone in management of retroperitoneal leiomyosarcoma.

Keywords

- ▶ leiomyosarcoma
- ▶ sarcoma
- ▶ sarcoma retroperitoneal

Introduction

Retroperitoneal sarcomas are neoplasms of mesodermal origin derived from adipose tissue representing 10 to 14% of all soft tissue sarcomas¹ and 0.07 to 0.2% of all neoplasia.² In a population-based series from the Surveillance, Epidemiology, and End Results database, the average annual incidence of retroperitoneal sarcomas was ~2.7 cases per million population.³ Anatomically, the retroperitoneal space is the space posterior to the peritoneal cavity, anterior to paraspinous muscles, with the superior border being the diaphragm and inferiorly the pelvic diaphragm. Having said this, it is a relatively large space whereby retroperitoneal sarcomas can grow asymptotically up until a mass effect develops. Hence, patients usually present late with symptoms like vague abdominal discomfort, increasing abdominal girth, and lump abdomen. Diagnosis is usually made on the basis of computed tomography (CT) scans of the abdomen and pelvis. Surgery with en-bloc resection of the tumor and adherent nearby structures is the gold standard of treatment. Herein, we present a case of 59-year-old male patient presenting for surgical management of 19 x 15 x 14 cm retroperitoneal leiomyosarcoma.

Case Report

A 59-year-old male patient, known to have hypertension controlled by medication with no previous abdominal surgeries or any remarkable family history, presented with the complaint of increased abdominal girth and nonspecific abdominal discomfort, 8 months prior to presentation. Patient denied any weight loss, fatigue, and change bowel habits. On physical examination, his abdomen was non tender, distended, and hard to palpation. Blood workup including a complete blood count, electrolytes, creatinine and liver function tests was done and turned out to be normal. Hence, a colonoscopy and CT scan abdomen pelvis with intravenous contrast (IV) contrast were scheduled. Colonoscopy was normal. However, the CT scan of the abdomen and pelvis with IV contrast showed a well-defined large lobulated heterogeneous mass originating from the left retroperitoneum (orange arrow) (▶ Fig. 1). The mass measures 19x15x14 cm in its largest craniocaudally, transverse, and anteroposterior axis, respectively. It shows multiple hypoattenuating central regions compatible with necrosis (blue arrow) (▶ Figs. 1 and 2) with few tiny calcifications. It is responsible for lateral displacement of the spleen and left descending colon. The tumor

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invades the perirenal fat on the medial aspect of the left kidney (red arrow) (►Fig. 3), and shows mild compression on the left renal pelvis (green arrow) (►Fig. 4) with no evidence of significant hydronephrosis. There is also mass effect on the left renal arteries (yellow arrow) (►Fig. 5) with no stenosis. Patient was scheduled for surgical excision of the mass with an R0 resection as the aim. Surgery was performed with a generous midline incision from the xiphoid process to the pubic bone with extension to the left subcostal area. The tumor was exposed (►Fig. 6), displacing the left colon laterally (►Fig. 7), with no clear cleavage plane with the left colon mesentery in between. We opted for en-bloc resection of the tumor, left adrenal, left kidney, and wedge resection of the mesentery of the left colon. Pathology turned out to be retroperitoneal leiomyosarcoma, with intermediate grade, invading the perirenal fat and the medial aspect of the kidney, resection margins free with intact fibrous pseudocapsule. Sections showed a neoplasm composed of spindle shaped

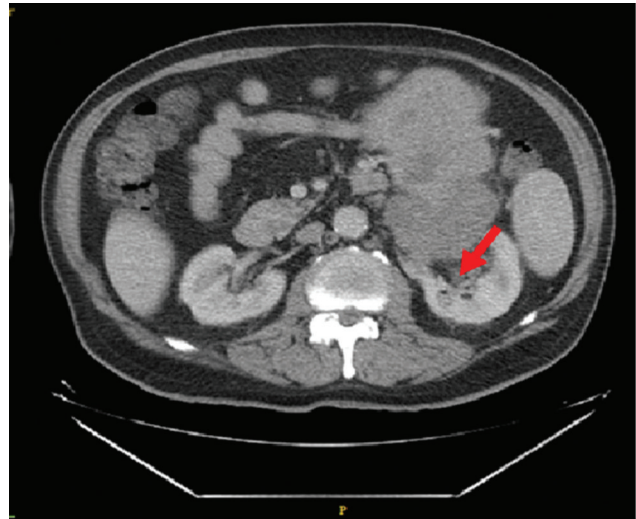


Fig. 3 The tumor invades the perirenal fat on the medial aspect of the left kidney (red arrow).

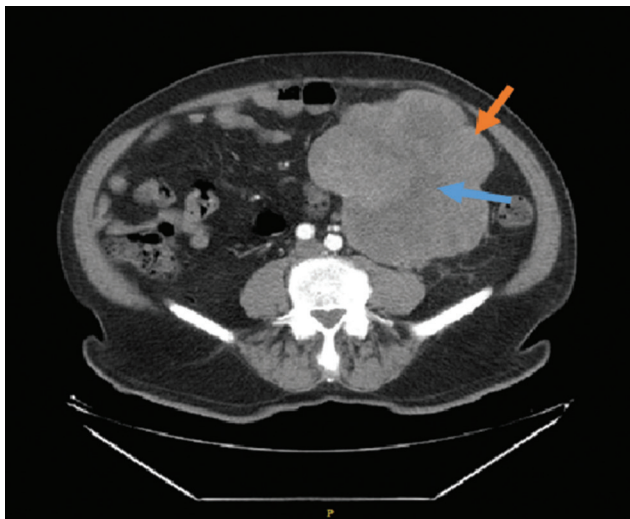


Fig. 1 Well-defined large lobulated heterogeneous mass originating from the left retroperitoneum (orange arrow). Blue arrow indicates central necrosis of the tumor.



Fig. 4 Mild compression on the left renal pelvis (green arrow).

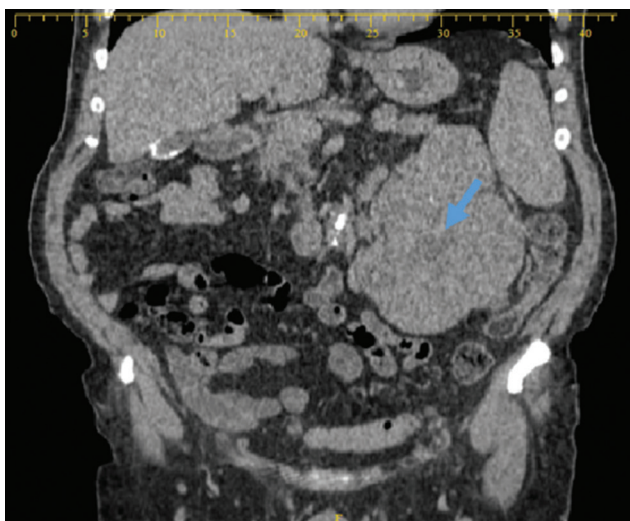


Fig. 2 Multiple hypoattenuating central regions compatible with necrosis (blue arrow).

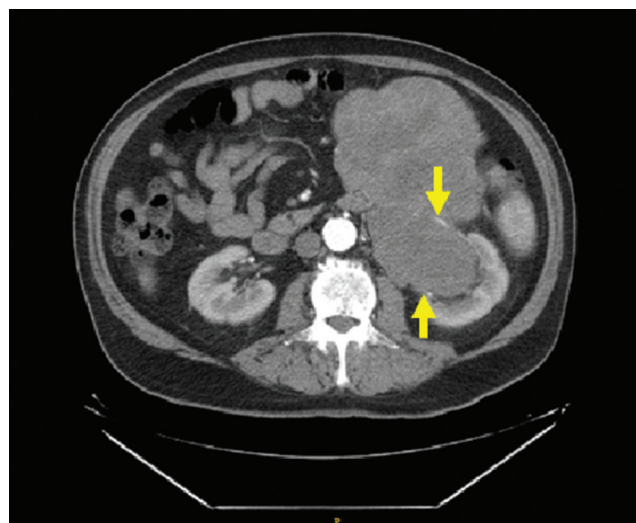


Fig. 5 Mass effect on the left renal arteries (yellow arrow).

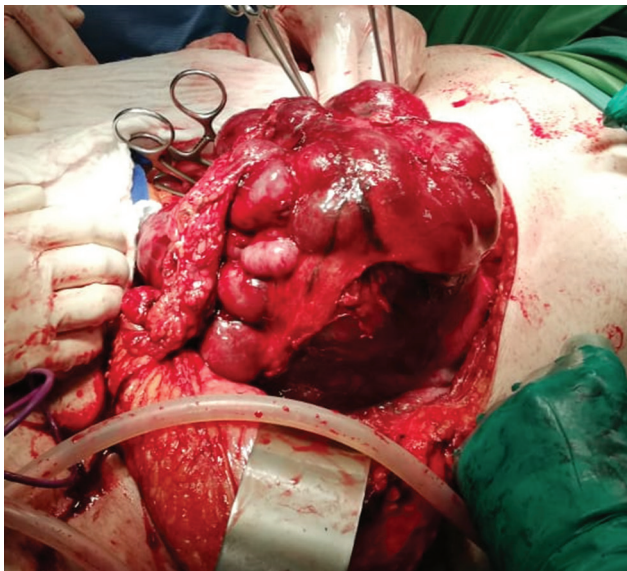


Fig. 6 Tumor exposed.

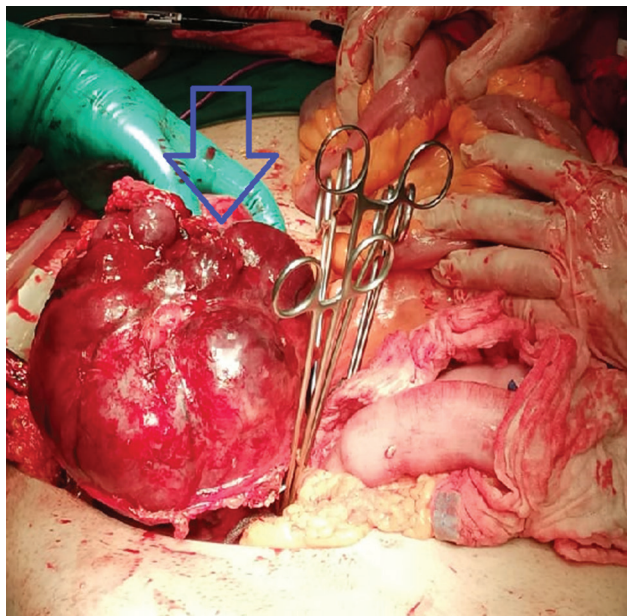


Fig. 7 Left colon displaced laterally.

cells that exhibit a fascicular growth pattern (►**Fig. 8**). Some of the cells show oval shaped nuclei with blunt ends, and minimal pleomorphism, while others exhibit much more pleomorphic hyperchromatic nuclei with marked variation in size and shape (►**Fig. 9**). The stroma is scant, and shows areas of tumor necrosis, corresponding to the grossly visible yellow scattered patches. The necrotic tissue, both grossly and microscopically, accounted for less than 50% of the tumor mass. Mitotic figures are abundant, with a count of 14 mitosis per 1.735 mm² (equivalent to 7 high-power field on our microscope). Also grossly and microscopically, the tumor invades the renal sinus fat of the lower pole of the kidney as well as the perirenal fat along the medial border of the kidney. The renal parenchyma itself, as well as the renal pelvis and ureter, was free of neoplasia. Immunohistochemistry

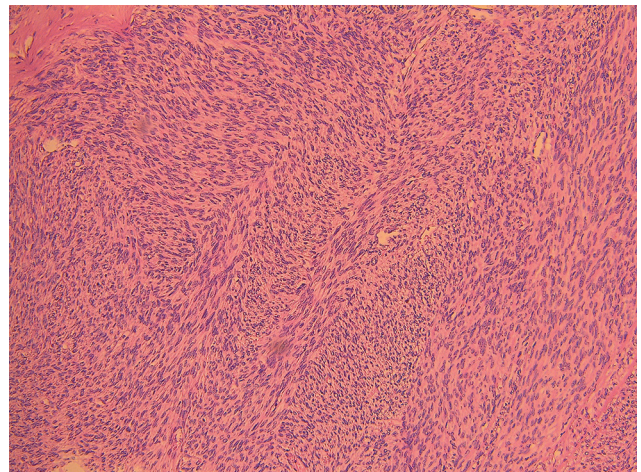


Fig. 8 Fascicular growth pattern of hypercellular spindle tumor cells.

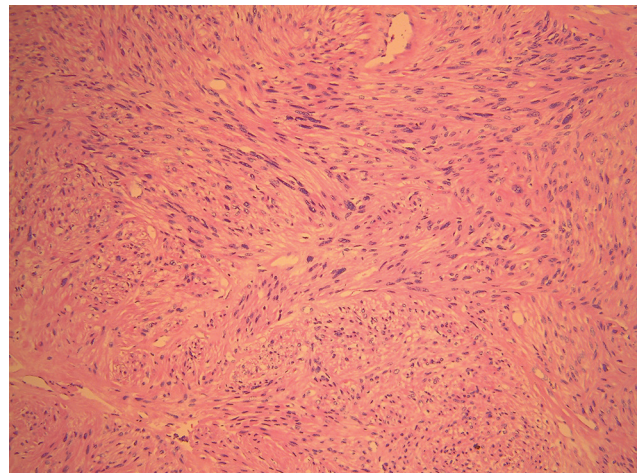


Fig. 9 Pleomorphic tumor cells, showing marked variation in size and shape.

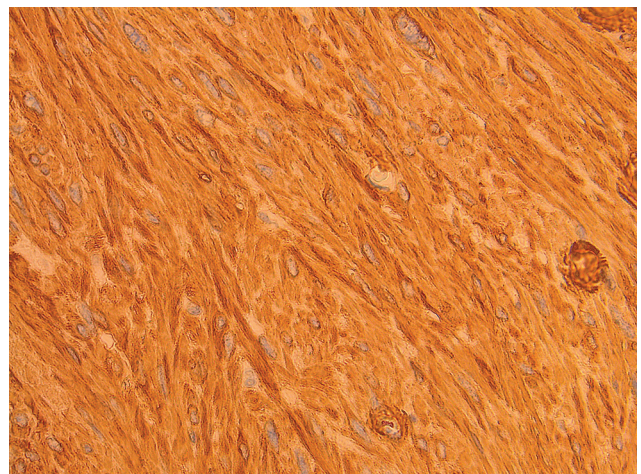


Fig. 10 Smooth muscle actin strong diffuse positivity.

done showed strong diffuse positivity for smooth muscle actin (►**Fig. 10**), desmin, and H-Caldesmon (►**Fig. 11**). S-100 and CD117 were negative.

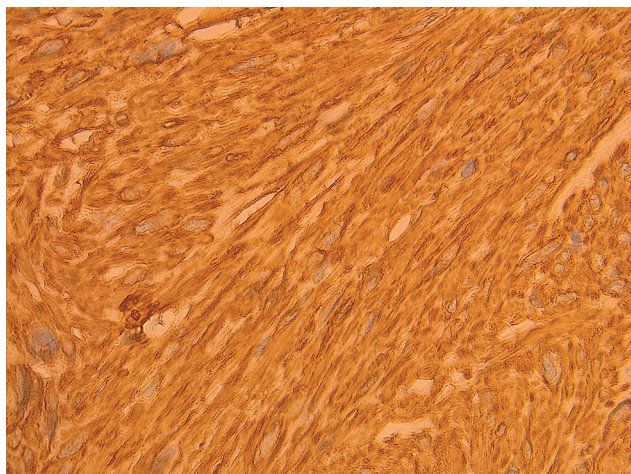


Fig. 11 H-Caldesmon strong diffuse positivity.

Patient had a smooth postoperative course and was discharged home 6 days after surgery. On 1-year follow-up, the patient is disease free as documented by imaging with no complaint.

Discussion

Retroperitoneal sarcomas are typically clinically silent given the anatomically large retroperitoneum. Most cases are diagnosed incidentally in an asymptomatic or minimally symptomatic patients. Symptoms usually develop when the retroperitoneal tumor starts to produce a mass effect on surrounding structures. Gastrointestinal symptoms include early satiety, obstruction, bleeding, and increase in abdominal girth. On the other hand, the tumor can locally invade and compress the retroperitoneal neurovascular bundle resulting in lower extremity edema and neurologic or musculoskeletal symptoms. Rarely, leiomyosarcomas present with paraneoplastic hypoglycemia, which is usually secondary to tumor production of insulin-like growth factor 2.⁴

Clinically, leiomyosarcomas differ from liposarcoma by their tendency to metastasize either to the lung or to the liver with the lung being a more frequent site for metastasis. This may be diagnosed at the time of presentation and in some occasions soon after the resection is done; hence, there is need of a closer follow-up and more aggressive approach.

Surgery with en-bloc resection of the tumor and adherent nearby structures, with intact capsule, remains the gold standard in surgical management of retroperitoneal sarcomas. Moreover, only a complete surgery has been proven to improve the overall survival in patients diagnosed with retroperitoneal sarcomas, with the most important variable being en-bloc resection of the tumor with adherent nearby structures. Aforementioned, studies have shown that the median survival of patients with complete resection was 103 months versus 18 months with incomplete resection, similar to when no surgery was done at all.¹ For tumors more than 10 cm in dimensions, complete R0 resection can be achieved in 70% of

cases; however, multiorgan resection is deemed necessary in 50% of cases to achieve R0 resection.⁵ Hence, the resection of a retroperitoneal sarcoma of remarkable size is a challenge due to multiple factors, the anatomical site, absence of an anatomically evident vascular, and lymphatic pedicles making it hard to obtain safe margin, adhesences to nearby organs, and the great vessels. In our case, the tumor was among the largest reported in literature adherent to the left kidney and mesentery of the left colon; hence, curative surgery necessitates the en-bloc excision of the left retroperitoneal tumor, left adrenal gland, left kidney, left ureter, and a wedge resection of the mesentery of the left colon. In instances where an R0 resection cannot be achieved, there is a 100% chance of recurrence and recurrence is considered the primary cause of death.⁶ Therefore, an aggressive surgical behavior is mandated, with the resection of the structures and viscera adjacent to the pathological process in an attempt to obtain an R0 resection. The use of chemotherapy is limited to metastatic disease as palliation, along with radiotherapy in nonoperable cases or incomplete resection.^{7,8} In summary, the ability to achieve R0 resection along with the histopathologic grade will solely determine the disease prognosis. The above-mentioned two factors are determined during surgery and are very hard to elucidate preoperatively.

Conclusion

The saying that one size fits all does not apply to retroperitoneal sarcomas. Approach to each patient should be individualized. However, complete surgical resection of the tumor with en-bloc resection of adjacent adherent organs remains the cornerstone in the management of retroperitoneal leiomyosarcoma.

Authors' Contributions

All authors contributed equally to writing this article.

Note

Informed consent was obtained from the patient to publish the case and all associated images.

Conflict of Interest

None declared.

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