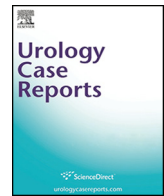




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En bloc removal of a large leiomyosarcoma arising from the inferior vena cava using retroperitoneal laparoscopic dissection of posterior vessels followed by cavectomy in a thoracoabdominal approach

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ABSTRACT

A 55-year-old Japanese woman had a large retroperitoneal tumor involving the inferior vena cava (IVC) in the right infrahepatic space. We performed *en bloc* tumor resection with the right kidney and ipsilateral adrenal gland. Because of the large tumor size, we used a retroperitoneal laparoscopic approach for the posterior dissection and performed renal artery ligation. Following open conversion by a thoracoabdominal anterior approach, the tumor was completely removed without major reconstruction. Pathological examination confirmed a leiomyosarcoma arising from the IVC. Retroperitoneal laparoscopic dissection of the posterior side enabled easy and safe surgery even for a large tumor involving the IVC.

Introduction

Retroperitoneal leiomyosarcoma (LMS) is the secondary major histological subtype of retroperitoneal soft tissue sarcoma (RSTS). It most commonly arises from the inferior vena cava (IVC), originating from smooth muscle cells of the vascular walls.¹ Studies of primary RSTS resection have demonstrated that the surgical margin status is a significant predictor of overall survival.² To achieve negative margins, *en bloc* resection of the tumor with retroperitoneal organs is recommended.³ Complete resection with negative margins remains challenging because of the frequent expansive appearance of IVC-LMS at initial diagnosis.² We herein report a case of a large IVC-LMS treated by the retroperitoneal laparoscopic posterior approach. After right renal artery ligation and tumor isolation, *en bloc* removal with right nephrectomy and adrenalectomy were safely performed with a pathologically negative margin. This approach was feasible even for a large retroperitoneal tumor involving the IVC. Sufficient mobilization of the tumor and IVC by the posterior approach provides complete resection with good tumor handling and safety.

Case presentation

A 55-year-old Japanese woman with no medical history was referred to her local hospital with suspected lower extremity exanthema. Computed tomography and magnetic resonance imaging demonstrated a 10-cm-diameter retroperitoneal mass in the right infrahepatic space (Fig. 1A–C). The tumor showed intraluminal extension to the IVC, and liver invasion was suspected. Intraluminal extension with complete obstruction of the segment between the infrahepatic and infrarenal IVC facilitated development of collateral circulation (Fig. 1A). Laboratory testing and MIBG scintigraphy showed no abnormalities. *En bloc* tumor resection including the segment between the infrahepatic and infrarenal IVC, right kidney, and adrenal gland was scheduled. Because of the complete IVC obstruction, adequate development of collateral circulation on preoperative imaging indicated that we could ligate the IVC without reconstruction. We considered that standard transabdominal open surgery might prevent access to the posterior vessels and renal artery because of the large tumor size. We therefore used a retroperitoneal laparoscopic approach until the posterior dissection, renal artery ligation, and IVC isolation.

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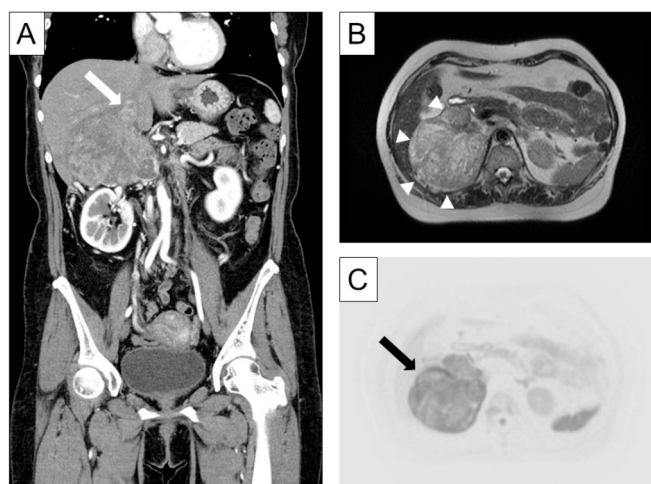


Fig. 1. A. Coronal contrast-enhanced computed tomography shows the heterogeneous retroperitoneal mass with extension to the infrahepatic inferior vena cava (white arrow). B. Axial T2-weighted magnetic resonance imaging shows that the mass involved the inferior vena cava with areas of high signal intensity (white square). C. Diffusion-weighted magnetic resonance imaging shows high signal intensity within the mass (black arrow).

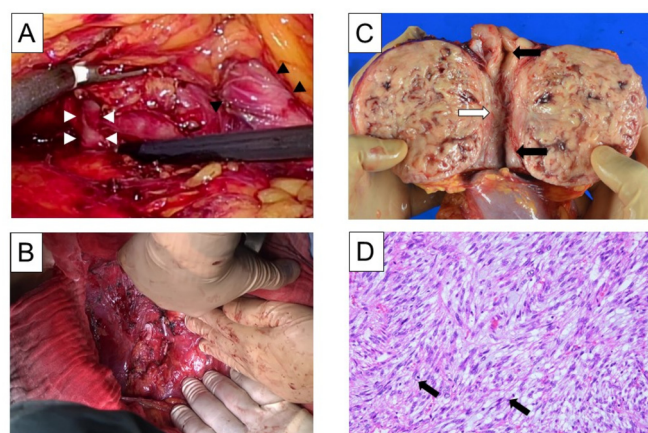


Fig. 2. A. Intraoperative image shows isolation of the right renal artery (white square) and infrarenal inferior vena cava (black square) by the retroperitoneal laparoscopic posterior approach. B. *En bloc* removal of the tumor with right nephrectomy was performed without major reconstruction. C. Gross appearance of the gray tumor shows localization from the inferior vena cava (white arrow) with the combination of an extravascular and intravascular (black arrow) growth pattern. D. Pathological examination shows interlacing bundle proliferation of spindle tumor cells. Many mitoses, including atypical mitosis, are present (hematoxylin–eosin, $\times 200$).

With the patient in the left full lateral position, pneumoperitoneum was created by open access. The retroperitoneal space was dissected, and the right renal artery was ligated using Hem-o-lok® clips (Fig. 2A). The surface of the IVC was resected and isolated at the caudal part of the tumor thrombosis. The patient was then moved to the hemilateral position. A thoracoabdominal incision was made via the seventh intercostal space on the mid-axillary line accompanied by a transrectal incision. After liver mobilization and Kocher's maneuver, the expanded IVC and contralateral renal vein were identified. The expansive tumor was easily mobilized with the wide dorsal space created by the initial laparoscopic procedure. Ultrasonography demonstrated the tumor tip in the infrahepatic IVC without direct liver invasion. After clamping these vessels, *en bloc* removal of the tumor with the right kidney, adrenal gland, and partial IVC was successfully performed without major reconstruction (Fig. 2B). The total operation time was 417

minutes, including 132 minutes for the laparoscopic procedure. The estimated blood loss was 700 ml without the need for a blood transfusion throughout the perioperative course. The patient recovered with no severe complications.

Macroscopic examination revealed a 9.0- \times 8.5- \times 8.0-cm well-circumscribed mass. The cut surface was gray, and focal hemorrhage was present. The tumor was located at the lumen of the IVC and extravascularly (Fig. 2C). Microscopically, spindle tumor cells with atypical nuclei proliferated in a fascicular pattern. Immunohistochemical findings were compatible with LMS arising from the IVC, with a pathologically negative margin. Mitosis was frequently seen (13 per 10 HPF), and the MIB-1 ratio was 36% (Fig. 2D). The patient was disease-free at the 6-month follow-up.

Discussion

LMS is a rare malignant neoplasm primarily composed of cells with smooth muscle degeneration and mainly occurs in the extremities, trunk wall, and body cavities. Retroperitoneal LMS is more common in women in their fifth to sixth decades of life.⁴ Only 5% of LMS directly arises from large blood vessels; the most common primary site is the IVC.¹

For cure of RSTS, complete resection with negative margins should be achieved at the initial operation whenever possible.³ Achievement of negative margins requires *en bloc* resection of retroperitoneal organs around the tumor. Standard removal of IVC-LMS with right nephrectomy necessitates adequate exposure of the upper abdomen and retroperitoneum with a large incision. Liver mobilization and Kocher's maneuver allow for identification and dissection of tumors involving the IVC, right kidney, and renal blood vessels. Usually, these procedures are performed by an anterior approach. However, this is technically demanding because of the mass effect of the tumor with the expanded IVC. Moreover, mobilization of the tumor with the right kidney and IVC and division of the right renal artery between the aorta and IVC are complicated procedures. Ciancio et al.⁵ reported the availability of posterior ligation of the renal artery in patients with large renal cell carcinoma treated by right radical nephrectomy. When performed by a posterior approach after medial mobilization of the kidney, this technique facilitates easier access to the renal artery than does the anterior approach.⁵ Early ligation of the renal artery provides tumor shrinkage and decreased blood loss from pararenal collaterals. Furthermore, division of lumbar veins by this approach also allows *en bloc* mobilization of the tumor and IVC.⁵ In our case, we successfully identified the renal pedicle and IVC and divided the right renal artery laparoscopically by a posterior approach. Despite the extremely large size of the tumor, combining laparoscopic and open procedures allowed for its complete mobilization and dissection.

Conclusion

To our knowledge, this is the first case report of a retroperitoneal laparoscopic approach followed by a thoracoabdominal open approach for *en bloc* removal of a large IVC-LMS. We believe that better mobilization of the tumor and IVC using a laparoscopic approach can help to achieve complete dissection of large retroperitoneal tumors.

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Informed consent

The patient provided written informed consent for her case report to be published anonymously.

Conflicts of interest

The authors declare that they have no conflict of interest.

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