

# A case of giant leiomyosarcoma of the inferior vena cava with liver metastases: A surgical challenge

Yuki Tsuchiya, Tomoaki Ito, Mutsumi Sakurada, Tomoyuki Kushida, Hajime Orita, Hiroshi Maekawa, Miki Yamano, Ryo Wada, Koichi Sato

## ABSTRACT

**Introduction:** Leiomyosarcoma of the inferior vena cava is a rare tumor that is hard to diagnose because there are no definitive symptoms. **Case Report:** A 72-year-old Japanese female presented with edema in both legs. Enhanced computed tomography scan of the abdomen revealed a heterogeneous tumor measuring 10×11×15 cm, which exerted pressure on the inferior vena cava. We preoperatively diagnosed the tumor as leiomyosarcoma in the middle region of the inferior vena cava. During resection of the tumor, the right and left renal veins and inferior vena cava were clamped for 32 minutes. A 90-mm elliptical resection was taken from the inferior vena cava and replaced with a 20-mm straight vascular graft. Pathological examination indicated leiomyosarcoma of the inferior vena cava and liver metastasis. The patient was put on anticoagulant drugs and discharged on the 14th postoperative day. **Conclusion:** We report a rare case of giant leiomyosarcoma of the inferior vena cava with liver metastasis.

**Keywords:** Inferior vena cava, Leiomyosarcoma, Liver metastasis

Yuki Tsuchiya<sup>1</sup>, Tomoaki Ito<sup>1</sup>, Mutsumi Sakurada<sup>1</sup>, Tomoyuki Kushida<sup>1</sup>, Hajime Orita<sup>1</sup>, Hiroshi Maekawa<sup>1</sup>, Miki Yamano<sup>2</sup>, Ryo Wada<sup>2</sup>, Koichi Sato<sup>1</sup>

**Affiliations:** <sup>1</sup>Department of Surgery, Juntendo University Shizuoka Hospital, Shizuoka, Japan; <sup>2</sup>Department of Pathology, Juntendo University Shizuoka Hospital, Shizuoka, Japan.

**Corresponding Author:** Hajime Orita, Department of Surgery, Juntendo University Shizuoka Hospital, Shizuoka, Japan; E-mail: oriori@juntendo.ac.jp

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## INTRODUCTION

Primary leiomyosarcoma of vascular origin is a rare tumor that arises most commonly from the inferior vena cava (IVC). Less than 200 cases of IVC leiomyosarcoma have been reported since 1871 [1]. Leiomyosarcomas generally originate from the smooth muscles and are malignant with a slow growth pattern. There are no definitive symptoms, but typical symptoms include abdominal or back pain and edema in leg [2]. The recommended treatment is surgical resection. There is no evidence of long-term survival benefit from chemotherapy and radiation therapy.

This report describes a rare case of resection of giant leiomyosarcoma of IVC with liver metastasis.

## CASE REPORT

A 72-year-old Japanese female was admitted to our hospital with treatment-resistant edema in both legs. She had been prescribed diuretic drugs two months earlier at another clinic. On physical examination, abdominal palpation showed that a fist-sized mass in the right upper

quadrant was hard. She had had a benign ovarian tumor and received an operation when she was 42 years old. A laboratory examination was normal. Tumor markers were normal. Enhanced computed tomography scan of abdomen revealed a heterogeneous tumor, measuring 10×11×15 cm, which exerted pressure on inferior vena cava (Figure 1A). Magnetic resonance imaging scan of this tumor revealed contrasting weak high intensity on the T1-weighted image and weak high intensity on the T2-weighted image (Figure 1B). There was no metastasis. Abdominal angiography showed that the tumor was connected by branch vessels to the three feeding arteries, the proper hepatic, superior mesenteric and lumbar arteries. GIST or lymphoma thinks as a differential diagnosis. These diseases do not infiltrate the blood vessel. The tumor was diagnosed as a leiomyosarcoma in the middle region of IVC, because this tumor was connected with inferior vena cava revealed hypervascular.

A laparotomy was performed in the left lateral decubitus position. The tumor adhered to the gallbladder, liver and duodenum. First of all, we resected the gallbladder. We chose fractional excision of the mass from the IVC (Figure 2A) because of the size of the leiomyosarcoma. The proper hepatic, superior mesenteric and lumbar arteries were ligated. The tumor grew from the IVC, extending 5 cm above the renal vein. The right and left renal veins and IVC were clamped for 32 minutes. A 90 mm elliptical resection was taken from IVC and replaced with a 20-mm straight vascular graft (Figure 2B). A hepatic metastasis at the right lobe of the liver (Segment 6) measuring 2 cm was revealed and was resected (Figure 2C). Operative time was 375 minutes. The postoperative recovery was uneventful. The patient was put on anticoagulant drugs and discharged on the 14th postoperative day. In an examination of the resected specimen, a 15×17×11 cm primary tumor (Figure 3A) arose from the IVC (Figure 3B). Microscopically, the primary tumor and liver metastasis measuring 1.5 cm consisted of spindle cells with hypercellularity (Figure 4A). Immunohistochemical examination revealed that the tumor cells were positive for  $\alpha$ - smooth muscle actin, desmin, vimentin and caldesmon and negative for c-kit, CD34, S-100, estrogen receptor, progesterone receptor, and CAM5.2 (Figure 4B). Elastica van Gieson staining showed the tumor was connected with the internal elastic membrane of IVC (Figure 4C). The Ki-67 proliferation index of the tumor was 40%. Pathological diagnosis was leiomyosarcoma of inferior vena cava.

Postoperatively, the patient has been on follow-up for the past five months. Warfarin was chosen as anticoagulant. PTINR was controlled to keep it below two-fold. There has been no distant metastasis or local recurrence and CT scans show no occlusion of the vascular graft in IVC.

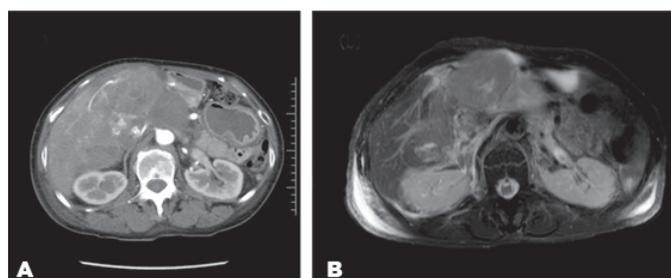


Figure 1: (A) Enhanced computed tomography of the abdomen revealed a heterogeneous tumor, 10×11×15 cm, which exerted pressure on the inferior vena cava, (B) Magnetic resonance imaging of this tumor revealed a contrasting weak high intensity on the T2-weighted image.



Figure 2: (A) Leiomyosarcoma grew on the inferior vena cava, (B) A 9-cm, elliptical resection of the IVC was replaced with a 20-mm straight vascular graft, (C) Liver metastasis was found on Segment 6 and resected.

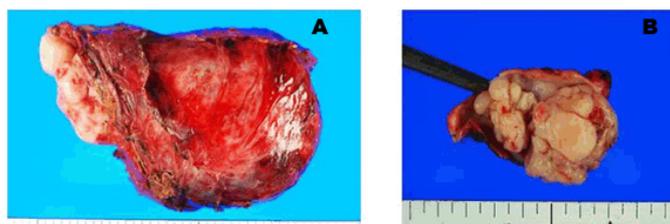


Figure 3: (A) A 15×17×11 cm tumor with IVC and (B) The tumor arose from the IVC

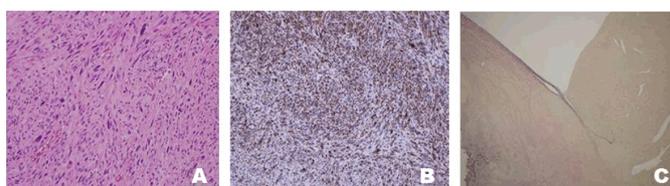


Figure 4: (A) Microscopic examination revealed that the tumor consisted of uniform and spindle cells (H&E stain, ×100). (B) The tumor cells showed positive for  $\alpha$ - smooth muscle actin (magnification: ×100), (C) Elastica van Gieson staining showed the tumor was connected with the internal elastic membrane of the IVC. Pathological diagnosis was leiomyosarcoma of the inferior vena cava (magnification: ×40)

## DISCUSSION

Soft tissue sarcomas represent less than 1% of all malignancies, and leiomyosarcomas constitute 6% of these tumors [3]. Vascular leiomyosarcomas constitute just 2% of all leiomyosarcomas [4]. Leiomyosarcomas originating in IVC are very rare, and are malignant with a slow growth pattern.

The diagnosis is established preoperative in only 10% cases because there are no definitive symptoms [5]. Symptoms and signs can include abdominal pain (66%), abdominal mass (48%), edema in leg (39%), and weight loss (30%). Other nonspecific symptoms are fever, weakness, anorexia, and dyspnea [6].

There is a perioperative mortality of 4%, and 42% of patients die of the disease itself [7]. Thus it is important to diagnose it as early as possible. Computed tomography scan is a useful screening test to diagnose IVC tumors. Fat suppression and gadolinium enhancement increases the quality of MRI images and helps in assessing retroperitoneal tumors [8].

In this case, the patient noticed edema in leg four months before presenting but she did not have any pain. Thus the tumor was in an advanced stage at diagnosis. We were able to preoperatively diagnose the tumor as leiomyosarcoma of the IVC. Enhanced computed tomography scan of the abdomen and magnetic resonance imaging scan of the T2-weighted image were effective for a diagnosis.

Inferior vena cava leiomyosarcomas are categorized according to the three regions in which they occur. Most IVC leiomyosarcomas (58%) occur in the middle region of the IVC between the hepatic and renal veins. The lower region accounts for 44% of cases and the upper region only 4% [9]. We classified this case as middle region. Patients with this type of IVC leiomyosarcoma often experience abdominal pain (76.8%), while those with the upper type often show edema in leg (78%) and the lower type does not have characteristic symptoms. Radical tumor resection was associated with better five-year and ten-year survival rates (49.4% and 29.5% respectively). Patients with tumors which arose from the middle segment fared better (56.7% and 47.3%) than those with lower segment tumors (37.8% and 14.2%). Resectability of the tumor is the prime factor affecting prognosis [10].

Surgical resection has been recognized as the only effective treatment for patients with IVC leiomyosarcoma. The tumor recurs in over 50% of patients who undergo radical resection, and the five-year survival rate is between 31% and 62% [11]. Postoperative therapy and the management of recurrence are difficult because of the absence of evidence for their effectiveness in patients. Radiation has been used in both the neoadjuvant and adjuvant settings, and some authors believe that it may help with the local control of the disease [12].

A final diagnosis is made by means of pathological and immunohistochemical methods. The pathognomonic indicators of leiomyosarcoma are spindle-shaped tumor cells with positive markers for smooth muscle cells, vimentin, muscle actin, alpha-smooth muscle actin, and desmin [13]. Our case showed  $\alpha$ -SMA(+), desmin(+), and vimentin(+). A liver mass was found to have tissues similar to leiomyosarcoma. This was diagnosed as metastasis.

Previous studies have reported that leiomyosarcomas originating in the IVC show the following characteristics:

- 1) The tumor shows a significant intraluminal component,
- 2) The part of IVC showing extraluminal growth requires resection to achieve adequate tumor excision, and
- 3) Surgical data indicates no other source of origin for the sarcoma [14]. Our case fulfilled these three criteria

## CONCLUSION

We diagnosed a very rare giant leiomyosarcoma of the inferior vena cava (IVC) with liver metastasis and excised it completely using a vascular graft. We must consider leiomyosarcoma when there is tumor growth between the hepatic and renal veins which presses against the IVC.

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## Author Contributions

Yuki Tsuchiya – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Tomoaki Ito – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Mutsumi Sakurada – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Tomoyuki Kushida – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Hajime Orita – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Hiroshi Maekawa – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Miki Yamano – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Ryo Wada – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Koichi Sato – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

## Guarantor

The corresponding author is the guarantor of submission.

## Conflict of Interest

Authors declare no conflict of interest.

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