

# Successful Treatment of a Recurrent Leiomyosarcoma of the Inferior Vena Cava

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**Summary.** A patient with a recurrent leiomyosarcoma of the inferior vena cava (IVC) was treated with aggressive surgery. The tumor originated from the inferior vena cava at the confluence of the renal veins and extended into the right atrium. The tumor was resected utilizing a cardiopulmonary bypass. It recurred in the right hepatic vein and right kidney 4 years after the initial operation. A right hepatectomy, right nephrectomy, and partial resection of the inferior vena cava was performed to treat the recurrence. The patient presently shows no evidence of recurrent disease 6 years after the initial operation. Aggressive surgery should thus be employed in the treatment of leiomyosarcomas of the inferior vena cava even in patients with tumor recurrence.

**Key words**—leiomyosarcoma, inferior vena cava, recurrence, hepatectomy, nephrectomy.

## INTRODUCTION

Leiomyosarcoma of the inferior vena cava (IVC) is a rare tumor which is associated with a poor prognosis and a high rate of recurrence.<sup>1)</sup> Patients with involvement of the upper segment of the IVC have a much higher mortality because it is more difficult to ensure complete tumor resection. In recent years, aggressive surgery using a cardiopulmonary bypass has been employed in the treatment of this disease.<sup>2)</sup> Still, recurrences are frequent. Surgical resection is the treatment of choice, even in patients with recurrent disease.

## CASE REPORT

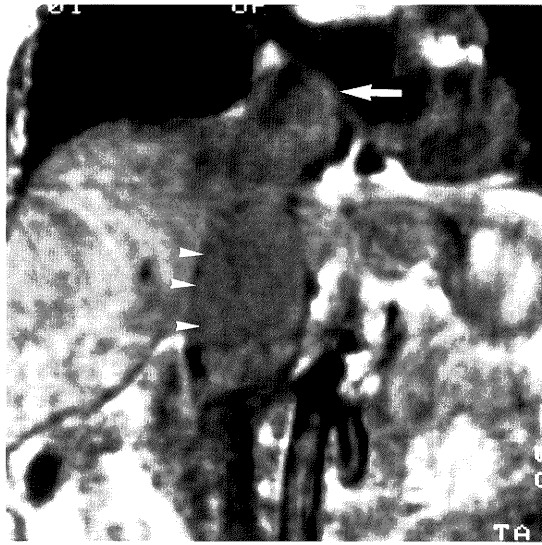
A 55-year-old woman was admitted with the chief complaint of fever and abdominal pain. No underlying liver disease including portal hypertension was recognized. She was found to have a subhepatic tumor, and tumor thrombus in the infrahepatic IVC extending into the right atrium (Fig. 1). Computed tomography (CT) and magnetic resonance imaging (MRI) revealed abutting the caudate lobe of the liver. Celiac arteriography demonstrated that the blood supply to the tumor originated from the hepatic artery. Venography showed a tumor thrombus at the confluence of the IVC and the renal veins and well-developed collaterals (Fig. 2). The differential diagnosis included hepatocellular carcinoma and a retroperitoneal tumor originating from the adrenal gland or IVC.

A midline incision was made from the manubrium to the umbilicus. The tumor was identified as a leiomyosarcoma originating from the wall of the IVC (Fig. 3a). It was resected from the suprahepatic and infrahepatic IVC, utilizing a cardiopulmonary bypass. Since the intraatrial component of the tumor was larger than the lumen of the IVC, the mass was divided at the orifice of the right atrium. Histologic examination of the resected specimens revealed leiomyosarcoma.

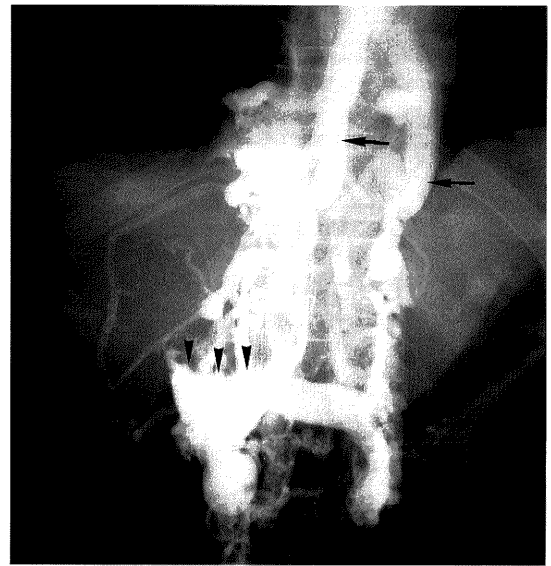
Tumor recurrence was identified 4 years after the initial operation on routine follow-up CT. MRI revealed tumors in the right hepatic vein, and in the hilum of the right kidney (Fig. 4). These tumors were discontinuous. There was no tumor thrombus in the IVC.

A curative right hepatic lobectomy and right nephrectomy were performed (Fig. 3b). Suspicious nodules were identified on the IVC, so partial resection of

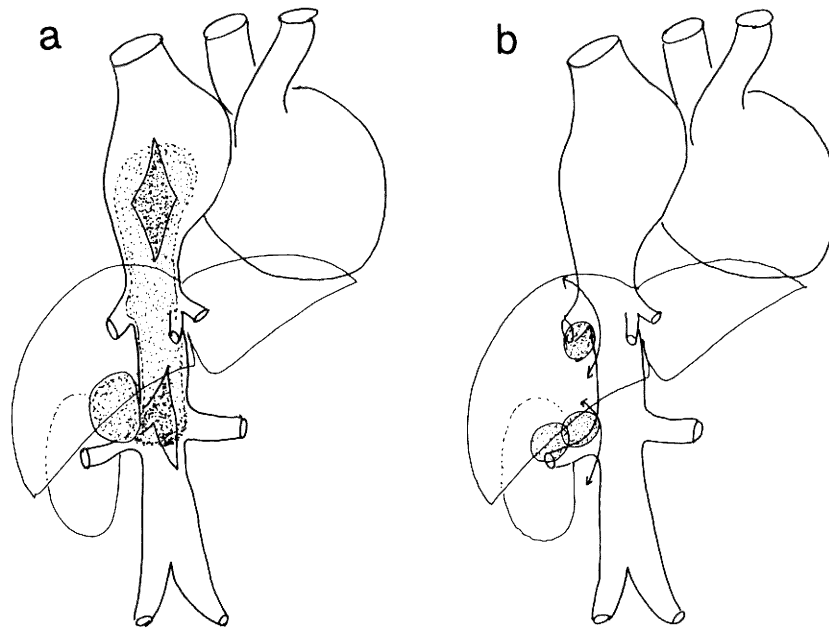
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**Fig. 1.** Magnetic resonance imaging (MRI) showing tumor thrombus extending from the infrahepatic inferior vena cava (*arrowheads*) to the right atrium (*arrow*).



**Fig. 2.** Venocavography demonstrating collateralization (*arrows*) and a filling defect in the inferior vena cava (*arrowheads*).



**Fig. 3.** **a.** Schematic diagram of the extent of tumor involvement at the initial operation, and **b.** schematic diagram of the extent of recurrent tumor involvement at the second operation.



Fig. 4. Magnetic resonance imaging (MRI) showing recurrent tumors in the right hepatic vein (black arrow) and right renal hilum (white arrows).

the medial segment of the IVC between hepatic and renal veins was carried out. Pathologic examination of the resected tissue revealed recurrent leiomyosarcoma in the hepatic vein and right kidney; these tumors recurred at the surgical stump of the initial operation. After surgery the left kidney and remnant liver functioned well. The patient was discharged on postoperative day 28, and is currently well 19 months after the second operation without any evidence of recurrent disease.

## DISCUSSION

In 1991, Mingoli et al. reported a series of 144 leiomyosarcomas of the IVC.<sup>1)</sup> Surgical resection was advocated to achieve long-term survival. In that series, there was a 5-year survival rate of 50% after curative resection. However, tumors located in the upper segment of the IVC were not readily resectable and were thus associated with a poor survival rate.

In recent years, an aggressive surgical approach to leiomyosarcomas of the upper segment of the IVC has been recommended, even in the patients in whom the tumor thrombus extends into the right atrium.<sup>2)</sup> In the case presented here, a cardiopulmonary bypass was used to facilitate tumor resection with partial resection of the IVC. The portion of the tumor thrombus which extended into the right atrium was divided for resection because it was larger than the orifice of the IVC. A cardiopulmonary bypass was necessary to complete the operation.

The rate of recurrence after resection of a leiomyosarcoma of the IVC is high.<sup>1-3)</sup> Unfortunately, factors which may predict these recurrences are unknown. In addition, the efficacy of adjuvant chemotherapy and radiation have yet to be delineated. Thus, complete surgical resection should always be considered, even in the treatment of recurrence. Mingoli et al. have reported reoperative resection in 5 of 40 patients who presented a recurrence. Until now, however, there have been no reports of a long-term survivor who underwent reoperative resection with a nephrectomy and partial hepatectomy.

Although curative resection of a leiomyosarcoma of the IVC may occasionally require hepatectomy or nephrectomy<sup>4)</sup> or both as in the initial operation of this case, judging from the mode of recurrence, hepatectomy is not usually performed when a cardiopulmonary bypass with systemic heparinization is required for tumor resection because of the attendant risk of bleeding from the cut surface of the liver.

The early detection of leiomyosarcoma of the IVC is usually difficult.<sup>5)</sup> Symptoms are uncharacteristic and vary considerably according to the site of venous obstruction by the enlarged tumor. MRI is the most useful modality to define tumor extension, and predict anatomical resectability. Since leiomyosarcomas are slow-growing tumors in most cases, close long-term follow-up is required. MRI may be used for early detection of recurrences.

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