

# Leiomyosarcoma of the retrohepatic vena cava: Report of a case treated with resection and reconstruction with polytetrafluoroethylene vascular graft

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## ABSTRACT

Leiomyosarcoma of the vena cava is a rare malignant tumor. A 61-year-old woman was admitted with right upper quadrant abdominal pain. Computed tomography revealed a retrohepatic vena cava tumor originating 2 cm below the confluence of the hepatic veins and ending 2 cm above the renal veins. The tumor was resected with 1 cm clear surgical margins, without requiring liver resection. Polytetrafluoroethylene vascular graft was used for reconstruction of the vena cava. Now 32 months postoperatively, there has been no recurrence or metastasis. Radical resection with negative surgical margins is the best curative therapy for leiomyosarcoma. Polytetrafluoroethylene vascular graft can be used in extensive tumors located at the vena cava.

**Keywords:** Leiomyosarcoma, retrohepatic vena cava, surgical treatment

## INTRODUCTION

Soft tissue sarcomas are an uncommon and heterogeneous group of tumors which make up less than 1% of adult malignancies and 10% of pediatric malignancies. In addition, less than 5% of soft tissue malignancies are leiomyosarcomas in adults. Vascular leiomyosarcoma originates from the muscular layer of the wall of major blood vessels (1, 2). Although vascular leiomyosarcoma is rarely seen in clinical practice, the vena cava is the most common site, with more than 50% of all vascular leiomyosarcomas presenting in the inferior vena cava (3). Since first described by Perl and Virchow in 1871, only approximately 450 cases have been reported in the literature (4, 5).

Here we present a case of leiomyosarcoma located at the retrohepatic vena cava, treated by total resection of the tumor and reconstruction with polytetrafluoroethylene vascular graft.

## CASE PRESENTATION

A 61-year-old female was admitted to the hospital with right upper quadrant abdominal pain. Past medical history was significant only for type 2 diabetes mellitus. Abdominal computed tomography (CT) revealed cholelithiasis as well as a retrohepatic vena cava tumor measuring 3 x 3.5 x 4 cm (Figure 1a, b). There were no other lesions or metastasis in the abdomen or on thoracic CT. Blood tests and tumor markers were normal. Surgery was performed via upper midline laparotomy using a right subcostal incision. The liver was completely mobilized. The tumor was located 2 cm below the confluence of the hepatic veins and ended 2 cm above the renal veins. The caudate lobe of the liver was displaced anteriorly by tumor; there was no tumor invasion in the liver (Figure 2a). There was no metastasis to the lymph nodes or to other abdominal organs. The suprahepatic and infrahepatic vena cava were totally clamped 2 cm from the tumor margins. The renal veins were left open. *En bloc* tumor resection was performed with 1-1.5 cm clear surgical margins. Liver resection was not required. A polytetrafluoroethylene vascular graft (24 mm diameter, 9 cm length) was used for reconstruction of the vena cava (Figure 2b). Cholecystectomy was also performed. Anticoagulant treatment was started on the first postoperative day, and the patient was discharged from the hospital 9 days after surgery on anticoagulant treatment. Pathological findings revealed a leiomyosarcoma 7 cm in length originating from the retrohepatic vena cava, with negative surgical margins (Figure 2c). Desmin, SMA and h-caldesmon were strongly positive on immunohistochemical staining. The tumor was classified histologically as grade 2 according to the Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grading system. Written informed consent was obtained from the patient. The patient is now 32 months after surgery, with no recurrence or metastasis reported (Figure 3a, b).

## DISCUSSION

Although first described in 1871 by Perl and with the first surgical resection performed in 1928 by Mechior, leiomyosarcoma of the inferior vena cava (IVC) remains very rare (1). Due to a lack of prospective

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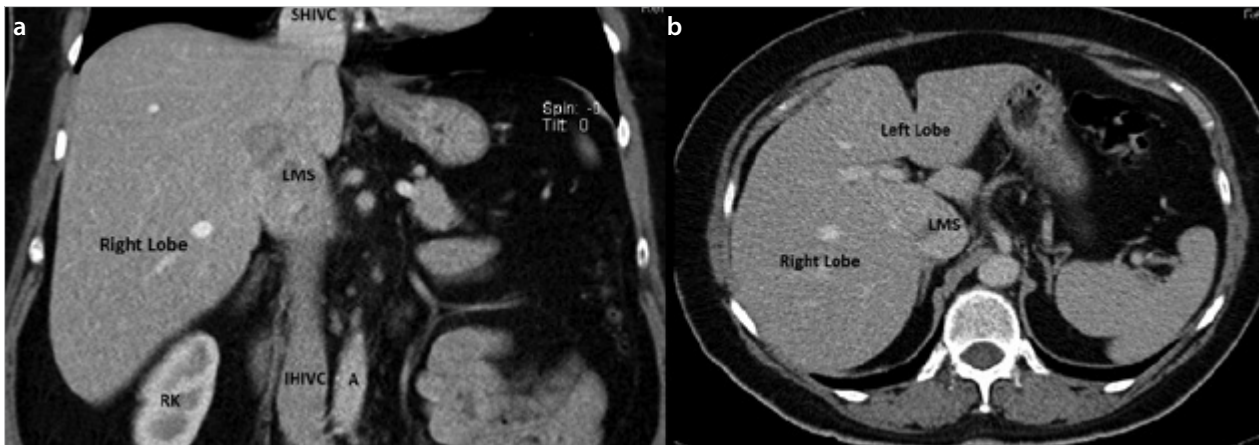


Figure 1. a, b. Preoperative CT images of retrohepatic IVC leiomyosarcoma displacing the caudate lobe of the liver anteriorly. (a) Coronal CT image of the leiomyosarcoma. (b) Horizontal CT image of the leiomyosarcoma  
LMS : leiomyosarcoma; IHIVC: infrahepatic inferior vena cava; SHIVC: suprahepatic inferior vena cava; A: aorta; RK: right kidney

randomized trials, there may never be a uniform multimodality approach to this rare malignancy (6). Mingoli et al. (7) reported an international registry of 218 cases in 1996, and Laskin et al. (3) reported the largest single center experience of 40 cases in 2010. Several centers have reported their experience, commonly in case reports, with a smaller number of patients (2, 5, 8, 9).

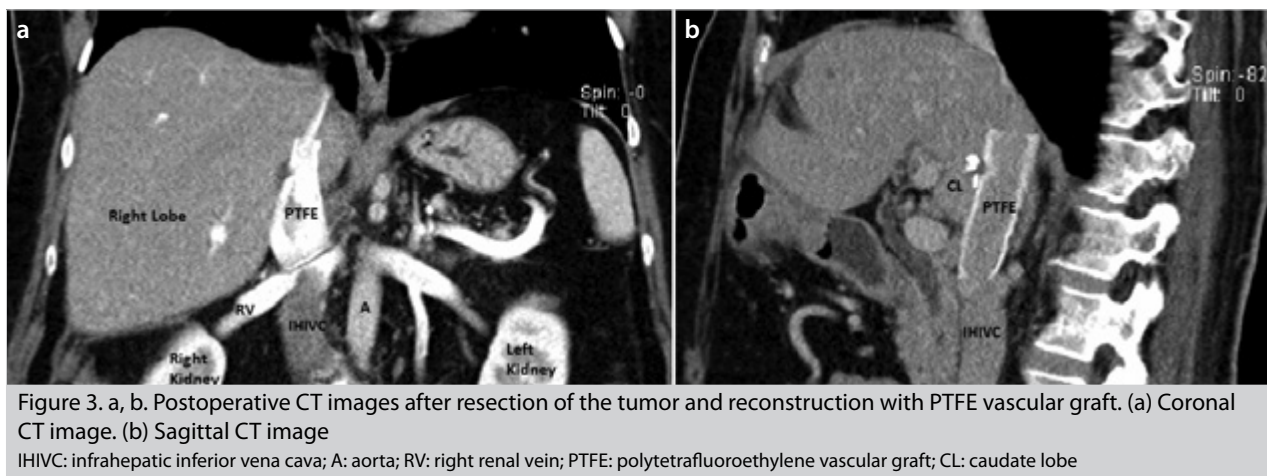
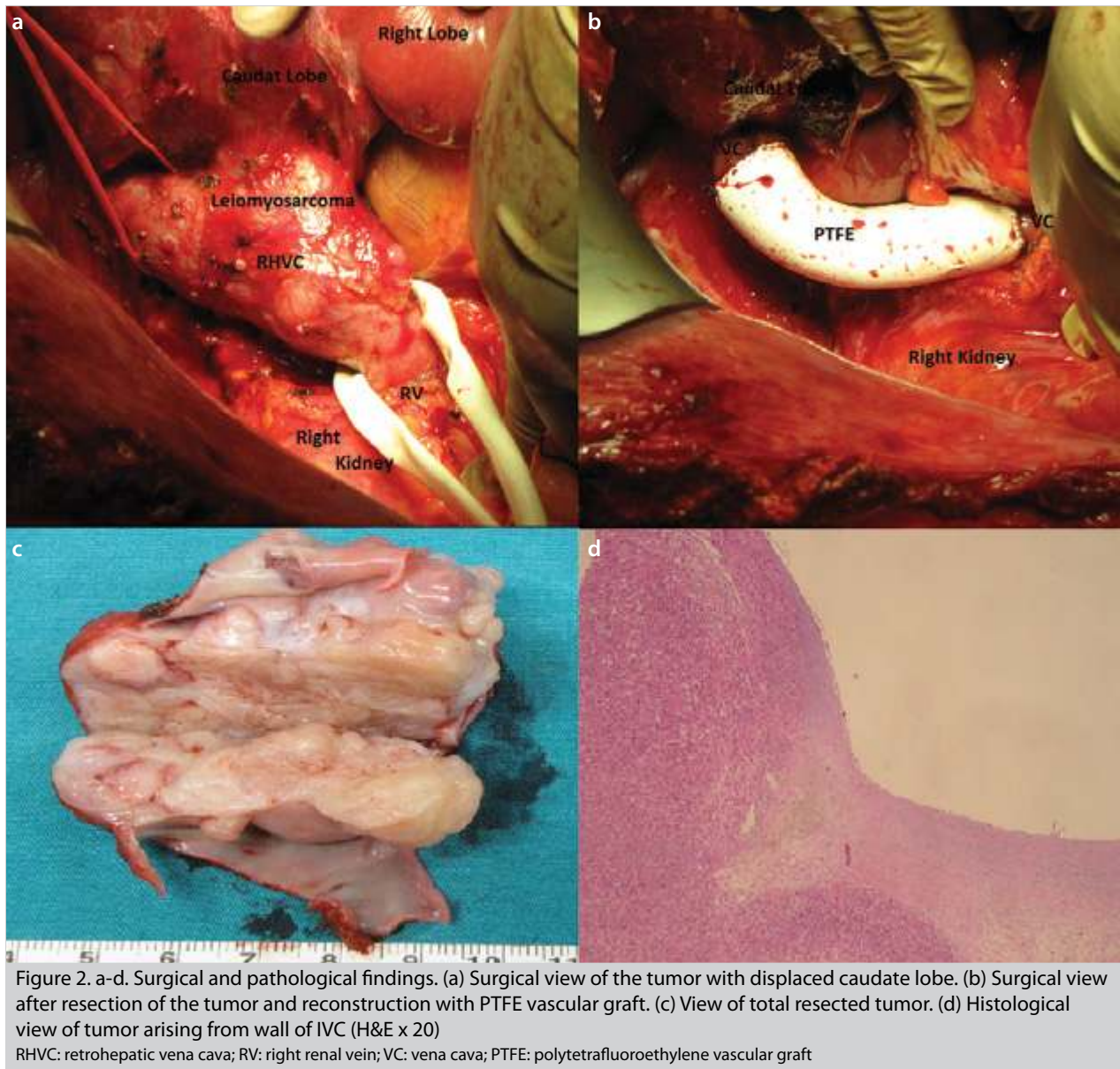
According to the limited recent experience, leiomyosarcoma of the IVC appears to be more common in females than males, with a ratio of 3:1 in the 5<sup>th</sup> to 6<sup>th</sup> decade of life. The single most common presenting symptoms is pain, as seen in our patient. Additional symptoms due to occlusion of the vessels could occur according to tumor size and location (3, 6). The presentation depends on its location on the IVC, which is divided into three levels by relationship with the hepatic and renal veins. Level 1 (lower), located below the renal veins and involving the IVC, is seen in 36% of cases. Typical presenting symptoms include lower extremity edema, deep venous thrombosis and palpable mass with abdominal pain. Level 2 (middle), from the renal veins to the hepatic veins, is involved in 44% of cases. Nephrotic syndrome and renal hypertension, possibly with abdominal pain, are presenting symptoms. Level 3 (upper), from the hepatic veins to the right atrium, is seen in 20% of cases. Presenting symptoms include weight loss, nausea, Budd-Chiari syndrome and cardiac arrhythmias (8). Leiomyosarcoma of the IVC is often misdiagnosed as an abscess cavity in the liver, a primary hepatic malignancy (when present at level 2), or as a thrombus in the IVC, as in our patient. According to CT, the second diagnosis in our patient was caudate lobe malignancy of the liver (Figure 1a, b). In an early stage diagnosis can be incidental, as in our patient. Our patient was evaluated due to right upper abdominal quadrant pain, which can be confused with symptoms of cholelithiasis. Cholelithiasis and the IVC tumor were seen on abdominal ultrasound, and the patient underwent CT scan.

Level 2 IVC leiomyosarcomas have a more favorable prognosis than Level 1 or Level 3 tumors as pain leads to an early presentation (3). Mingoli et al. (7) reported in the international registry and analysis of a worldwide series of 218 patient that Level 2 tumors had better 5- and 10- year survival rates (56.5% and 47.3%) than Level 1 tumors (37.8% and 14.2%) ( $p < 0.002$ ).

In Level 3 tumors, IVC involvement, lower limb edema, Budd-Chiari syndrome, intraluminal tumor growth and occlusion of the IVC were reported as associated risk factors for death in this study (7).

Surgical resection with negative surgical margins has been shown to be the most important treatment, and is the only treatment which improves survival. Two thirds of IVC leiomyosarcoma patients present with a localized tumor that can be resected. Debulking surgery may be performed for palliation. After resection, options for IVC reconstruction include placement of a synthetic graft, primary repair and patch repair. Mitotic rate and tumor grade and size have been suggested as other pathologic prognostic factors. Intravascular growth may be associated with early pulmonary metastasis. There is limited experience with neoadjuvant and adjuvant therapy for IVC leiomyosarcoma. Some studies have reported that adjuvant radiation therapy may be of benefit in local tumor control after incomplete resection or in high grade tumors (1, 7-9). In the few studies of the management of recurrent IVC leiomyosarcoma, surgery appears to be the only option if the risk of morbidity is low for local recurrence and isolated metastasis. Adjuvant chemotherapy and radiation therapy has not been proven but may be an option instead of surgery or following surgery (8, 9). In this case, we performed a total resection of the retrohepatic vena cava (Level 2 IVC) with negative surgical margins and reconstructed the vena cava with a polytetrafluoroethylene vascular graft (Figure 2b). Our patient was not treated with postoperative adjuvant therapy and has not developed recurrence or metastasis 32 months after surgery.

Leiomyosarcoma is a malignant tumor of mesenchymal origin with a differentiation toward smooth muscle morphology. Its histological appearance is composed of spindle shaped cells with eosinophilic cytoplasm with muscular striation and cigar-shaped round nuclei. Immunohistochemical staining for contractile fiber proteins such as actin, desmin and h-caldesmon can verify the diagnosis (1). Standard hematoxylin-eosin staining (Figure 2d) and immunohistochemical staining for smooth muscle markers was performed in our case. Desmin, SMA and h-caldesmon were strongly positive on immunohistochemical staining.



Some authors have recommended long-term postoperative anticoagulation after surgery to protect from thrombosis, but others have not (10). We used heparin during the initial postoperative period in our patient and continued oral warfarin therapy for one year after surgery.

#### CONCLUSION

Leiomyosarcoma of the retrohepatic IVC is a rare malignancy in which symptoms develop according to tumor location and size. Surgical treatment with negative surgical margins is the only treatment for survival. Patients undergoing treatment



at a center experienced in liver resection and transplantation have the benefit of a surgical team familiar with hepatocaval anatomy, and as a result, a better opportunity for safe radical resection of retrohepatic IVC leiomyosarcoma.

**Informed Consent:** Written informed consent was obtained from patient who participated in this case.

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