Urology Research, 2025, Volume 3, Issue 4
https://ojs.bbwpublisher.com/index.php/UR
Online ISSN: 2981-8230
Print ISSN: 3083-4910

Primary Leiomyosarcoma of the Inferior Vena Cava: A Case Report and Literature Review

Xiang Li, Shuai Ruan, Jinhan Zou, Ao Xu, Tenghong Huang, Bozi Zhou, Xi Guo

Department of Urology, The First Affiliated Hospital of Hunan Normal University, Hunan Provincial People's Hospital, Changsha 410005, Hunan, China.

Abstract: *Objective*: To investigate the diagnosis and treatment methods for primary leiomyosarcoma of the inferior vena cava. *Methods*: The clinical course of a patient with primary inferior vena cava leiomyosarcoma admitted to our department in September 2022 was retrospectively analyzed. Combined with a literature review, the clinical characteristics of inferior vena cava leiomyosarcoma were summarized. *Results*: The patient recovered smoothly after radical resection. However, follow-up examination at six months postoperatively revealed local tumor recurrence and hepatic metastasis. Following a multidisciplinary discussion, the patient promptly commenced treatment at the oncology center with epirubicin (50 mg/m^2) and ifosfamide (2500 mg/m^2). Nevertheless, severe side effects, such as hepatotoxicity, hindered timely drug administration, and the patient was lost to follow-up after the third cycle of chemotherapy. *Conclusion*: Radical surgical resection remains the only potentially curative treatment for patients with inferior vena cava leiomyosarcoma. However, for tumor recurrence and metastasis, a comprehensive treatment approach incorporating chemotherapy may be more effective in improving patient survival rates.

Keywords: Inferior vena cava leiomyosarcoma; Surgical procedures; Vascular reconstruction; Chemotherapy

Online publication: Dec 31, 2025

1. Introduction

Primary leiomyosarcoma of the inferior vena cava (PIVCLMS) is a rare malignant tumor originating from the smooth muscle of the venous wall. This disease was first reported by Perl in 1871 [1]. Radical surgical resection is the only potentially curative treatment for patients with PIVCLMS. However, the prognosis for these patients is generally poor due to the high malignant potential of the tumor itself, frequent delays in diagnosis, and the technical challenges associated with the surgical procedure [2]. This paper reports the diagnosis and treatment process of one case of PIVCLMS admitted to our department.

2. Clinical data

The patient was a 56-year-old female admitted due to upper abdominal pain persisting for three months. Her

medical history included uterine fibroids, for which she had undergone a hysterectomy. Physical examination revealed mild tenderness in the right upper abdomen. The abdomen was soft, no masses were palpable, and there was no edema in the lower extremities. Laboratory tests showed no significant abnormalities, and multiple tumor markers were within normal ranges. Abdominal ultrasound indicated a widened lumen in the retrohepatic segment of the inferior vena cava (maximum diameter approximately 31 mm), within which a heterogeneous, predominantly hypoechoic mass measuring about 68 × 32 × 30 mm was detected. The mass had relatively clear borders and exhibited longitudinal growth along the inferior vena cava. Whole abdominal CT and inferior vena cava CT venography (CTV) revealed an ovoid, slightly hypodense lesion protruding extraluminally from the suprarenal segment of the inferior vena cava. On its largest cross-section, it measured approximately 44 × 41 mm. Contrast-enhanced CT showed marked heterogeneous enhancement with small patchy areas of necrotic tissue exhibiting less enhancement. The lesion also involved the right renal vein as shown in **Figure 1a**. MRI demonstrated a mass both within and anterior to the inferior vena cava, appearing hyperintense on T2-weighted imaging (T2WI), hypointense on T1-weighted imaging (T1WI), and showing no signal change on fat-suppressed T1WI. The largest cross-sectional dimension was approximately 51 × 32 mm, involving a segment of the inferior vena cava about 59 mm in length. Additionally, an ovoid, slightly hyperintense focus on T2WI with relatively marked enhancement was found in segment IV of the liver (see **Figure 1b**). Radionuclide renal dynamic imaging indicated good bilateral renal function.

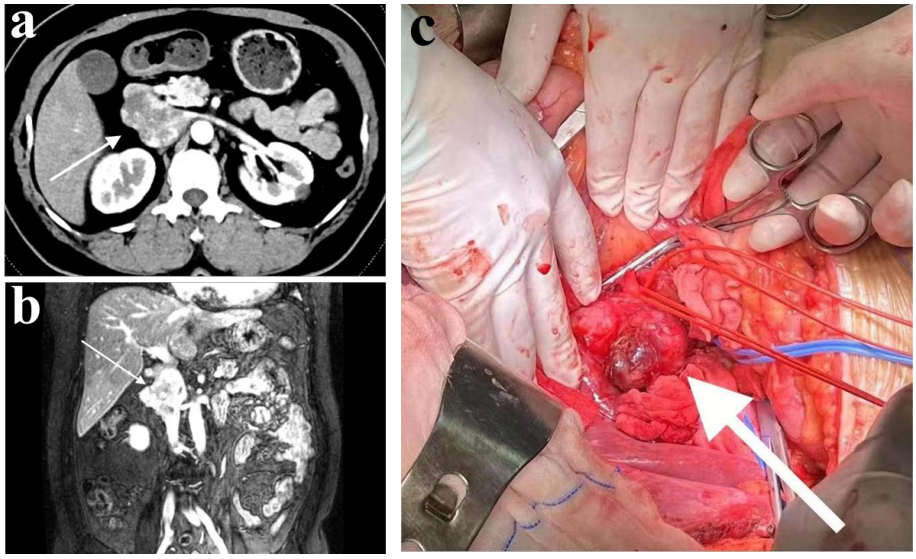

Figure 1. Preoperative imaging findings and gross appearance of the tumor.
(a) Preoperative CTV demonstrates a space-occupying lesion in the inferior vena cava, which exhibits heterogeneous enhancement on contrast-enhanced imaging. (b) MRI shows the lesion primarily involving segment II of the inferior vena cava, protruding from the lumen and extending superiorly to the inferior edge of the hepatic segment of the vessel. (c) Grossly, the tumor appears as an irregular, nodular mass with adhesion to surrounding tissues and a firm consistency.

The preoperative clinical diagnosis was inferior vena cava leiomyosarcoma (mixed intra- and extraluminal growth type). After excluding surgical contraindications, the patient underwent radical resection on September 22, 2022. Intraoperatively, the tumor appeared irregular in shape with lobulated growth, and part of the tumor tissue protruded significantly from the vessel lumen, measuring approximately 7 × 6 × 5 cm with a hard texture (see **Figure 1c**). During dissection of the mass, two nodular lesions, each about 10 mm in diameter, were identified in

the caudate lobe and the right posterior hepatic lobe, respectively, and were resected along with the primary tumor. An F18 Dacron prosthetic graft was used to reconstruct the inferior vena cava defect (refer **Figure 2**). The total operative time was 390 minutes, with a cumulative vascular ischemia time of 78 minutes, and the estimated blood loss was approximately 200 mL.

Postoperative pathological examination confirmed the diagnosis of moderately to poorly differentiated leiomyosarcoma with negative resection margins. The resected hepatic nodules also contained leiomyosarcoma. Immunohistochemical staining results were as follows: Desmin (+), SMA (+), Ki-67 (75% positive), S-100 (-), CD34 (-), CD117 (-).

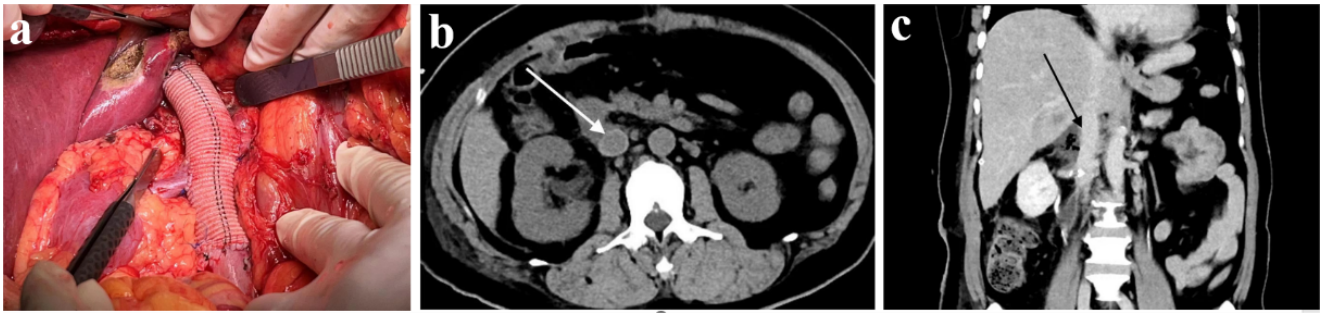

Figure 2. Intraoperative reconstruction of the inferior vena cava and postoperative follow-up findings. Following complete resection of the tumor, the inferior vena cava was reconstructed using a prosthetic graft (a). Postoperative follow-up CT demonstrates patency of the inferior vena cava (b, c).

The patient recovered smoothly during the postoperative hospital stay and was discharged on the 11th day after surgery. However, a follow-up CT scan six months postoperatively revealed local tumor recurrence and hepatic metastasis **Figure 3**). Following a multidisciplinary expert discussion, the patient promptly commenced treatment at the oncology center with epirubicin (50 mg/m^2) and ifosfamide (2500 mg/m^2). Nevertheless, severe side effects, such as hepatotoxicity, hindered timely administration of the chemotherapy, and the patient was lost to follow-up after the third cycle.

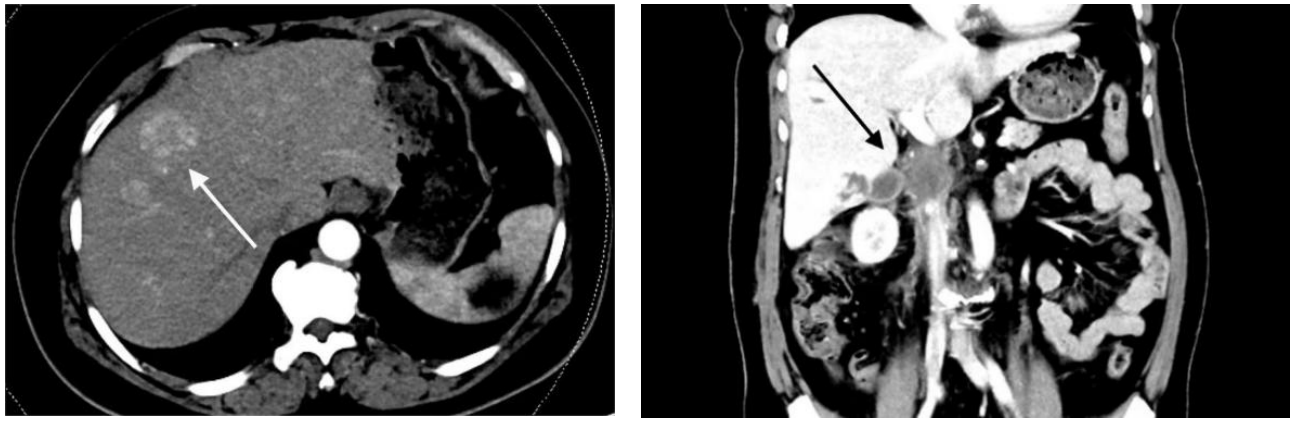

Figure 3. Local tumor recurrence and hepatic metastasis detected during follow-up examination at six months postoperatively.

3. Discussion

Primary leiomyosarcoma of the inferior vena cava (PIVCLMS) commonly occurs in middle-aged and elderly

women, with approximately 80% of patients being women aged 50–60 years. The prognosis is generally poor, with a high rate of recurrence [3]. Due to its occult location and slow growth, PIVCLMS is often not detected early, and clinical symptoms in its mid-to-late stages are also non-specific. Clinically, the inferior vena cava is segmented based on its anatomical relationship to the renal veins and hepatic veins. Leiomyosarcomas most frequently occur in segment II of the inferior vena cava. The majority of patients present with non-specific abdominal or back pain. Precisely because of these atypical symptoms, coupled with the significantly higher incidence in women compared to men, consulting physicians may initially consider gynecological conditions rather than vascular diseases, which can impede an accurate diagnosis.

Leiomyosarcoma of the inferior vena cava typically originates from the tunica media of the vessel wall. In its early stages, it is confined to the venous wall, subsequently invading adjacent tissues extraluminally or expanding intraluminally in a thrombus-like manner. Its growth patterns are classified as intraluminal, extraluminal, or mixed [4]. If the tumor exhibits intraluminal growth, preoperative diagnosis is often straightforward. However, when the growth pattern is extraluminal or mixed, and particularly when the tumor is large and causes displacement of surrounding vessels by compression, it can be difficult to determine its exact origin preoperatively. Computed tomography angiography (CTV) clearly demonstrates the tumor's size, morphology, extent of invasion, and its relationship with adjacent vessels and organs. The lesion primarily grows longitudinally along the course of the inferior vena cava. On non-contrast CT, it appears as a soft tissue density, showing heterogeneous enhancement after contrast administration. Large lesions may exhibit central necrotic areas, and the vena cava may display filling defects or local non-opacification. Magnetic resonance imaging (MRI) typically reveals a slightly hypointense signal on T1-weighted imaging and iso- to hyperintense mixed signals on T2-weighted imaging. MRI can display the entire tumor mass and the extent of collateral circulation formation. However, definitive diagnosis still requires histopathological examination, which provides conclusive evidence of the tumor's nature and aids in formulating an appropriate postoperative treatment plan when necessary.

Although extensive research has been conducted on treatment strategies for PIVCLMS, radical surgical resection appears to be the only potentially curative approach [2]. However, the complex adjacent anatomy of the inferior vena cava, and the potential for tumor extension into the right atrium, pose significant clinical challenges. Furthermore, management of the inferior vena cava following tumor resection is crucial and depends on factors such as tumor location and involvement of hepatic or renal veins. Unfortunately, even radical resection does not guarantee long-term survival, with reported 5-year and 10-year survival rates of approximately 40–70% and 20–40%, respectively [3,5].

For PIVCLMS in segment III, the inferior vena cava in this segment has a rich collateral circulation. If the common iliac vein bifurcation is not involved, venous return from the lower limbs can be ensured via anastomoses between the internal iliac veins and the pelvic venous plexus. In such cases, direct ligation of the inferior vena cava is associated with almost no complications such as lower limb edema [6]. Simple ligation can not only eliminate the need for complex vascular reconstruction procedures, thereby shortening operative time, but also avoid a series of complications associated with vascular reconstruction and the use of prosthetic grafts. For segment I PIVCLMS, when the tumor involves the hepatic veins or right atrium, patients often present with massive ascites due to Budd-Chiari syndrome, frequently losing the opportunity for surgical intervention. Even if surgery is performed, it must be conducted under conditions of hypothermia and cardiopulmonary bypass with cardiac arrest. Regarding segment II PIVCLMS, which often involves the renal veins, treatment becomes more complex when the tumor involves the right renal vein. Due to the short length of the right renal vein, which primarily receives drainage

from the right ureteric vein and lacks sufficient collateral circulation, preserving renal venous outflow during tumor resection is challenging. Renal vein ligation, or even nephrectomy, was previously often unavoidable in the treatment of inferior vena cava leiomyosarcoma, consequently leading to frequent occurrences of renal failure. Therefore, once a decrease in urine output is detected postoperatively, early plasma filtration should be initiated [7]. Existing literature suggests that performing a venorenal ostioplasty (VRO) during tumor resection can preserve part of the connection between the renal vein and the inferior vena cava. This technique helps prevent stenosis during anastomosis with an inferior vena cava graft and is an effective method for reconstructing renal outflow, with reported patency rates reaching 90% [8].

Following tumor resection, particularly after removal of segment II PIVCLMS, the question arises of whether reconstruction of the inferior vena cava and renal veins is necessary. If a patient's renal vein is involved, especially if chronically and completely occluded, a collateral venous circulation will form through shunt vessels [9]. Tumor resection without reconstructing the inferior vena cava or renal veins is a viable option for patients with well-developed collateral circulation. However, for patients with only partial renal vein occlusion and insufficient collateral circulation, simple surgical tumor removal is inadequate, and vascular reconstruction becomes essential [10]. In a series of cases reported by Yoshidome et al. where inferior vena cava reconstruction was not performed, 5 out of 10 patients underwent right nephrectomy due to venous outflow obstruction [11]. Reconstruction methods such as simple suture, patch repair, and graft replacement have all proven feasible to varying degrees, but the optimal management approach remains controversial [12]. In the case reported here, the inferior vena cava mass also involved the right renal vein. Therefore, reconstruction of both the inferior vena cava and the renal vein was performed using a Dacron prosthetic graft. This approach aligns with normal anatomical and physiological principles, successfully establishing a renal venous outflow tract, effectively preserving renal function. The patient recovered well on postoperative follow-up. Dacron material offers excellent mechanical properties and chemical stability, effectively resisting intra-abdominal pressure, demonstrating low thrombogenicity, and possessing notable advantages in longitudinal elasticity, allowing for better estimation of vascular defect length [13,14]. Practices regarding anticoagulant therapy after inferior vena cava reconstruction vary in the literature [15]. Given its high-flow nature, this patient did not receive postoperative anticoagulation, and no graft thrombosis was observed on follow-up examinations. Notably, thrombotic occlusion is often caused by graft deformation. During surgery, the liver is often rotated to facilitate anastomosis between the graft and the inferior vena cava, at which point the graft is patent. However, when the liver is returned to its normal anatomical position, the graft lumen can become twisted and deformed. Therefore, during inferior vena cava anastomosis, special attention should be paid to the degree of liver rotation to ensure the prosthetic graft and the inferior vena cava lie in the same plane.

Given the rarity of this disease, it is challenging to conduct large-scale controlled studies to evaluate the role of adjuvant chemotherapy in the treatment of inferior vena cava leiomyosarcoma. Currently, most treatment regimens are based on standard oncological principles and extrapolated from studies on soft tissue sarcomas. A meta-analysis indicates that combination therapy based on doxorubicin yields higher response rates and overall survival compared to doxorubicin monotherapy. Furthermore, anthracycline analogs can reduce certain side effects; for instance, epirubicin has lower cardiotoxicity while maintaining efficacy comparable to doxorubicin [16,17]. In this reported case, local recurrence and hepatic metastasis were detected during a follow-up examination six months postoperatively. Following a multidisciplinary discussion, the patient promptly commenced treatment at the oncology center with epirubicin (50 mg/m^2) and ifosfamide (2500 mg/m^2). Unfortunately, the patient was lost to follow-up after the third chemotherapy cycle due to adverse effects, which also implies that the efficacy

and outcomes of chemotherapy remain controversial. In Japan, when anthracycline-based chemotherapy fails, recently approved agents such as pazopanib, trabectedin, and eribulin are utilized as second-line or subsequent chemotherapy options [18].

4. Conclusion

In summary, primary inferior vena cava leiomyosarcoma is a rare disease. Due to its occult location, it is often diagnosed at an advanced stage. Early detection can significantly increase the feasibility of surgical intervention. Until more effective preoperative and postoperative adjuvant treatments are established, radical surgical resection remains the only potentially curative treatment option. Despite the challenges, ongoing research continues to explore multimodal strategies to improve outcomes for patients with this aggressive tumor.

Disclosure statement

The authors declare no conflict of interest.

References

[1] Nayyar R, Panda S, Saini A, et al., 2010, Leiomyosarcoma of Inferior Vena Cava Involving Bilateral Renal Veins: Surgical Challenges and Reconstruction with Upfront Saphenous Vein Interposition Graft for Left Renal Vein Outflow. Indian Journal of Urology, 26(3): 438–440.

[2] Cananzi F, Mussi C, Bordoni M, et al., 2016, Role of Surgery in the Multimodal Treatment of Primary and Recurrent Leiomyosarcoma of the Inferior Vena Cava. Journal of Surgical Oncology, 114(1): 44–49.

[3] Wachtel H, Gupta M, Bartlett E, et al., 2015, Outcomes after Resection of Leiomyosarcomas of the Inferior Vena Cava: A Pooled Data Analysis of 377 Cases. Surgical Oncology, 24(1): 21–27.

[4] Alkhalili E, Greenbaum A, Langsfeld M, et al., 2016, Leiomyosarcoma of the Inferior Vena Cava: A Case Series and Review of the Literature. Annals of Vascular Surgery, 33: 245–251.

[5] Nooromid M, De Martino R, Squizzato F, et al., 2022, Surgical Resection and Graft Replacement for Primary Inferior Vena Cava Leiomyosarcoma: A Multicenter Experience. Journal of Vascular Surgery Venous and Lymphatic Disorders, 10(3): 617–625.

[6] Fiore M, Colombo C, Locati P, et al., 2012, Surgical Technique, Morbidity, and Outcome of Primary Retroperitoneal Sarcoma Involving Inferior Vena Cava. Annals of Surgical Oncology, 19(2): 511–518.

[7] Cho S, Marsh J, Geller D, et al., 2008, Surgical Management of Leiomyosarcoma of the Inferior Vena Cava. Journal of Gastrointestinal Surgery, 12(12): 2141–2148.

[8] Liu D, Ren H, Liu B, et al., 2018, Renal Function Preservation in Surgical Resection of Primary Inferior Vena Cava Leiomyosarcoma Involving the Renal Veins. European Journal of Vascular and Endovascular Surgery, 55(2): 229–239.

[9] Du Toit D, Warren B, 2002, Juxta-Renal Inferior Vena Cava Leiomyosarcoma. European Journal of Vascular and Endovascular Surgery, 23(1): 84–86.

[10] Araujo R, Gaujoux S, D'Albuquerque L, et al., 2014, End-to-End Renal Vein Anastomosis to Preserve Renal Venous Drainage Following Inferior Vena Cava Radical Resection due to Leiomyosarcoma. Annals of Vascular Surgery, 28(4): 1048–1051.

[11] Yoshidome H, Takeuchi D, Ito H, et al., 2005, Should the Inferior Vena Cava Be Reconstructed after Resection for Malignant Tumors? American Journal of Surgery, 189(4): 419–424.

[12] Illuminati G, Pizzardi G, Calio F, et al., 2016, Outcome of Inferior Vena Cava and Noncaval Venous Leiomyosarcomas. Surgery, 159(2): 613–620.

[13] Hollenbeck S, Grobmyer S, Kent K, et al., 2003, Surgical Treatment and Outcomes of Patients with Primary Inferior Vena Cava Leiomyosarcoma. Journal of the American College of Surgeons, 197(4): 575–579.

[14] Ghose J, Bhamre R, Mehta N, et al., 2018, Resection of the Inferior Vena Cava for Retroperitoneal Sarcoma: Six Cases and a Review of Literature. Indian Journal of Surgical Oncology, 9(4): 538–546.

[15] Ruiz C, Kalbaugh C, Browder S, et al., 2020, Operative Strategies for Inferior Vena Cava Repair in Oncologic Surgery. Journal of Vascular Surgery Venous and Lymphatic Disorders, 8(3): 396–404.

[16] Krikelis D, Judson I, 2010, Role of Chemotherapy in the Management of Soft Tissue Sarcomas. Expert Review of Anticancer Therapy, 10(2): 249–260.

[17] Zer A, Prince R, Amir E, et al., 2018, Multi-Agent Chemotherapy in Advanced Soft Tissue Sarcoma: A Systematic Review and Meta-Analysis. Cancer Treatment Reviews, 63: 71–78.

[18] Kobayashi H, Iwata S, Wakamatsu T, et al., 2020, Efficacy and Safety of Trabectedin for Patients with Unresectable and Relapsed Soft-Tissue Sarcoma in Japan: A Japanese Musculoskeletal Oncology Group Study. Cancer, 126(6): 1253–1263.