

CASE REPORT

A rare case of leiomyosarcoma of the inferior vena cava in a 32-year-old patient

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Abstract

Leiomyosarcoma is a malignant tumor that develops from the smooth muscle constituents of the middle layer in veins, usually affecting the inferior vena cava (IVC); however, this disease is observed in older patients. In this report, we describe a rare case of a 32-year-old female patient who presented with leiomyosarcoma. Initially, the patient complained of moderate-intensity pain in the abdominal region of the right hypochondrium and nausea, worsening postprandial starting 6 days earlier. Abdominal ultrasonography showed a heterogeneous hypoechoic expansive formation, with lobulated contours and ill-defined limits in the projection of the IVC. Inferior vena cavography, showed obliteration of the vessel in almost all of its extensions. Magnetic resonance examination showed a large expansive lesion occupying the IVC. The lesion was resected, and the specimen was sent for anatomopathological analysis, which showed a leiomyosarcoma of the IVC. Patients with leiomyosarcoma have a poor prognosis, and early diagnosis and complete resection with free surgical margins are the only chance of long-term survival. The report stands out due to the appearance of this condition in a young patient with no family history of this disease or other predisposing carcinogenic pathologies.

KEYWORDS

abdominal neoplasm, abdominal pain, diagnostic imaging, leiomyosarcoma, radiology, vena cava

1 | INTRODUCTION

Leiomyosarcoma is a malignant tumor that develops in smooth muscle tissue. Its etiology is varied, and its prognosis is usually poor. Leiomyosarcoma is considered the most common tumor of the venous

system and the second most frequent retroperitoneum neoplasm in the elderly. It can develop in any vein, with the inferior vena cava (IVC) being the most affected vessel, accounting for 5%–10% of soft tissue sarcomas.^{1–3} The growth pattern of leiomyosarcomas can be either intraluminal (5% of cases), extraluminal (62% of cases) or combined

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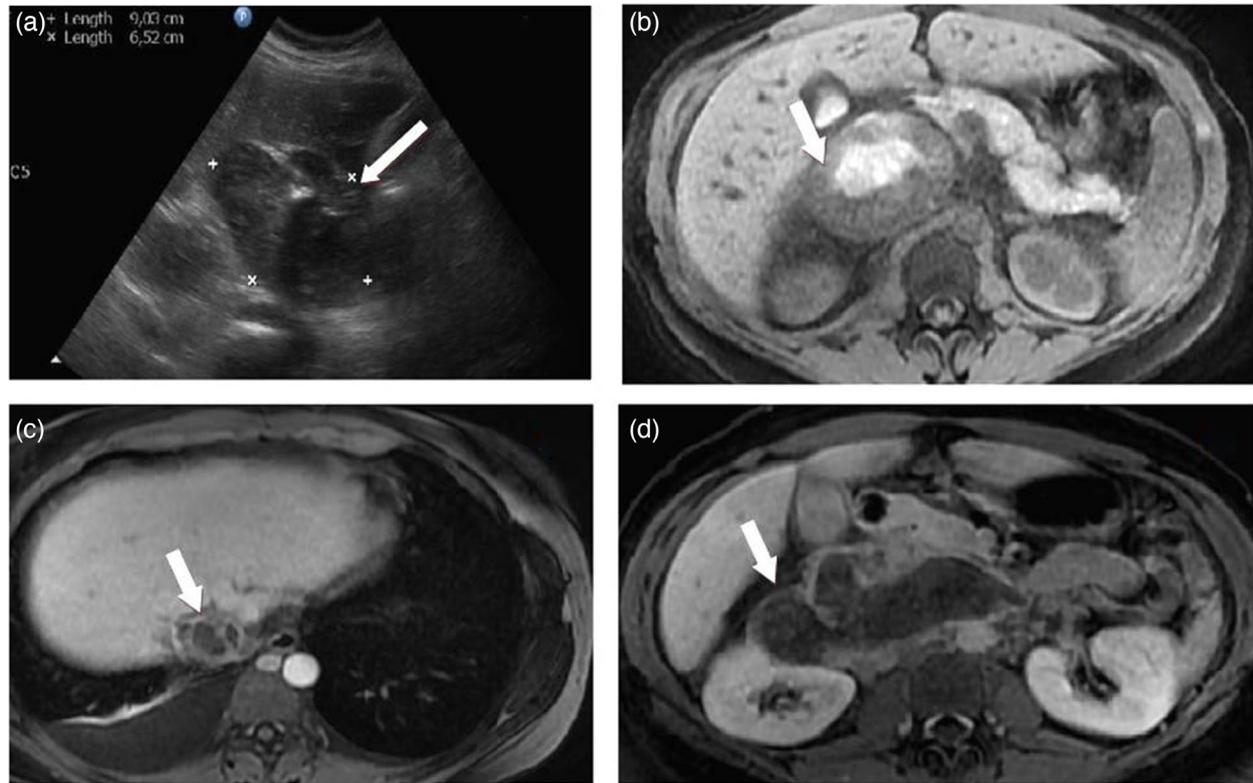


FIGURE 1 Frame (a) Ultrasound image of hypoechoic expansive formation, heterogeneous with lobulated contours and ill-defined limits, in the projection of the inferior vena cava. (b–d) Magnetic resonance imaging showed a large expansive lesion occupying the inferior vena cava, its intrahepatic segment, and the superior vena cava, extending to the renal veins and displacing the head of the pancreas, kidney, and adrenal to the right of intrahepatic bile ducts, ascites, and bilateral pleural effusion

(33% of cases). Regarding prevalence, this disease occurs in both males and females, with a higher incidence in the fifth and sixth decade of life.^{4,5–10}

According to the location, leiomyosarcomas of the IVC can be classified into three categories: (i) lower segment, which extends from the common iliac veins in the lower vena cava to the region close to the renal vessels (34% of leiomyosarcomas occur in this segment); (ii) infra-renal or middle segment, which occurs between the renal and hepatic veins (42% of leiomyosarcomas cases occur in this segment); and (iii) the upper segment, which extends from the hepatic veins to the right atrium of the heart (24% of leiomyosarcomas occur in these segments).^{11–14}

Involvement of the upper segment might manifest as Budd–Chiari syndrome, middle segment involvement might manifest as nephrotic syndrome, and involvement of the lower segment might manifest with edema of the lower extremities. The most common symptom is mild, non-specific, abdominal pain, usually preceding diagnosis by months or even years.⁴ The differential diagnoses included lymphoma, gastrointestinal stromal tumor, primary retroperitoneal leiomyosarcoma, and leiomyosarcoma of the IVC.

Leiomyosarcomas are slow-growing tumors and, often present with insidious and non-specific symptoms, such as abdominal pain, weight loss, palpable abdominal mass, weakness, fever, anorexia, vomiting, and night sweats, due to their retroperitoneal location.^{11,12,14,15} Uncommon complications include pulmonary embolism, anemia, depletion of

clotting factors, and cardiac arrhythmias. Due to this disease, additional metastatic invasion to adjacent organs, such as the kidneys and liver, can occur. In addition, distant tissues, such as the brain, can also be compromised, because of portosystemic anastomoses, mainly the lumbar veins.¹¹

2 | CASE REPORT

A 32-year-old female patient, previously healthy, went to the emergency department of a hospital in the city of Dourados, Mato Grosso do Sul, Brazil, presenting with moderate-intensity abdominal pain and nausea that started 6 days before the date of her entry and worsened postprandially. On physical examination, the patient had a distended abdomen, which was painful on palpation, with a palpable round abdominal mass, with hardened consistency in the right hypochondrium.

Total abdominal ultrasonography showed a gallbladder with thickened walls with free peripheral fluid, dilation of the intrahepatic bile ducts, and ascites. In addition, heterogeneous hypoechoic expansive formation, heterogeneous with lobulated contours and ill-defined limits, in the projection of the IVC (Figure 1) was shown.

Magnetic resonance imaging showed a large expansive lesion occupying the IVC, intrahepatic segment, and superior vena cava, extending to the renal veins and displacing the head of the pancreas, kidney, and

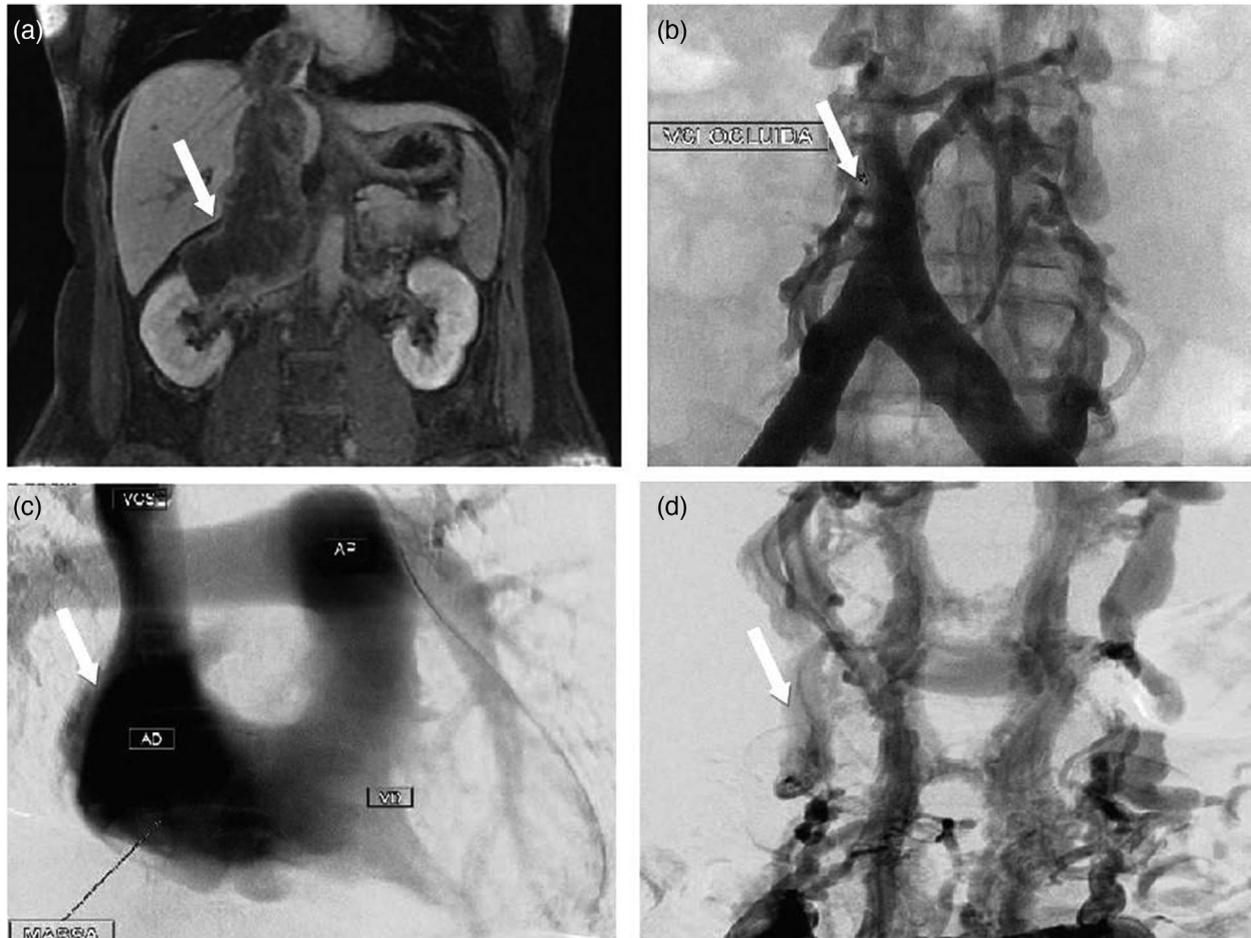


FIGURE 2 (a) Magnetic resonance imaging showed expansive formation with discreet irregular gadolinium enhancement, involving the right renal vein and inferior vena cava, extending to the right atrium. (b–d) Cavography showing the inferior vena cava obliterated in almost all of its extension, the right superior hepatic vein with subocclusive intraluminal mass, and collateral circulation through the paravertebral venous plexus

adrenal to the right of the intrahepatic bile ducts. Furthermore, ascites and bilateral pleural effusion were observed (Figures 1 and 2)

Inferior vena cavography showed obliteration of the IVC in almost all of its extension, a subocclusive intraluminal mass in the right superior hepatic vein, and collateral circulation through the paravertebral venous plexus (Figure 2). In the study of the right cardiac chambers, an atrial intraluminal mass projecting from the IVC was identified.

A biopsy of this atrial mass was carried out. The differential diagnoses included lymphoma, primary leiomyosarcoma of the retroperitoneum, leiomyosarcoma of the IVC, and metastasis. An ultrasound-guided biopsy was carried out, which showed leiomyosarcoma according to the histological and immunohistochemical findings.

3 | DISCUSSION

Leiomyosarcoma of the IVC is a rare tumor and usually has a poor prognosis. However, with an aggressive surgical approach, long survival and eventual healing can be achieved, especially in cases where there is no metastasis.

Differentiating between an IVC sarcoma and another soft tissue retroperitoneal sarcoma that surrounds it can be difficult. The true sarcoma of the IVC derives from the smooth muscle cells of the vessel and its growth pattern can be intraluminal or extraluminal, and might involve neighboring structures. The imperceptible IVC signal at the point of greatest contact between the IVC and the retroperitoneal mass has a sensitivity of 75% and specificity of 100% for confirming IVC diagnosis.⁸

This tumor has a slow growth, which often delays the diagnosis and keeps patients asymptomatic for a long time. The discovery is made incidentally in 10.5% of patients and through autopsy in 33%.^{8,9} Clinical manifestations depend mainly on the IVC segment involved, the most common of which are: Budd–Chiari syndrome (upper segment), abdominal pain (middle portion), and palpable mass (lower portion). The tumor can also involve more than one segment, which generates a combination of signs and symptoms.^{9–11}

Imaging contributes significantly to diagnosing and treating leiomyosarcomas. Regarding diagnostic imaging tests, ultrasonography has low diagnostic accuracy, but it can be used to rule out hepatobiliary and pancreatic disorders, and to confirm the presence of the tumor mass.^{14,19} Computed tomography (CT) and magnetic

resonance imaging are the gold standards for detecting the tumor, establishing its growth pattern and its relationship with surrounding structures, and identifying the presence of vena cava obstruction. On CT, the tumor might be heterogeneous, well-defined, lobulated, and solid, with signs of hemorrhage and necrosis compressing the IVC, and might have mild-to-moderate contrast enhancement, usually peripheral enhancement between intraluminal growth and the wall of the pit.^{11,12,14,17-19}

CT has a sensitivity of 78% and specificity of 96% for the diagnosis of vena cava tumors. Furthermore, this test allows the use of intravenous contrast medium to assess vascularization and delimit the tumor mass.^{11,19} Alternatively, magnetic resonance imaging has 95% sensitivity and 100% specificity for the detection of tumors in the IVC, and provides information similar to CT; however, the sagittal section is more reliable in terms of the tumor extension and tumor thrombus.^{11,14,19}

Cavography allows the assessment of the intraluminal extent of the tumor, and the degree of collateral circulation. It has a sensitivity of 97%; however, the total obstruction of the IVC makes the examination not specific, even with its high sensitivity.^{11,19} Although the image might suggest a primary tumor of the IVC, a biopsy is necessary, and ultrasound-guided biopsy is the modality of choice for obtaining tissues for histological analysis.¹⁹

The location of the tumor and the extent of the local invasion are important factors in making treatment decisions. Surgical resection offers the only prospect of cure. There is no acceptable surgical therapy for tumors with superior localization of the IVC with hepatic vein involvement. Studies and randomized clinical trials in the literature showed that chemotherapy and radiotherapy have no benefits in survival. The present study reports a rare case, highlighting the importance of considering leiomyosarcoma as a differential diagnosis of retroperitoneal masses, given that surgical planning has an impact on patient survival.

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