

# Leiomyosarcoma of the inferior vena cava

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## ABSTRACT

Primary vascular leiomyosarcoma is a rare tumor, which arises mainly from the inferior vena cava. Clinical signs are non-specific. Systemic metastasis occurs in the late stage. Imaging with color Doppler ultrasonography, contrast-enhanced computed tomography, or magnetic resonance imaging can significantly contribute to the diagnosis. We present a case of leiomyosarcoma of the inferior vena cava that extended into the right renal vein and the right iliac vein, as it is a rare case and emphasize the significance of the imaging methods in its diagnosis.

*Key words:* • leiomyosarcoma • inferior vena cava

**P** rimary vascular leiomyosarcoma is a rare tumor that arises mainly from the inferior vena cava (IVC) (1). It has 3 main growth patterns (2); 62% of cases demonstrate extraluminal, 5% intraluminal, and 33% extra- and intraluminal growth patterns. It is a slow-growing progressive tumor (3). Systemic metastasis occurs in the late stage. It is usually fatal because the presentation of symptoms occurs late (4). Imaging methods play an important role in the demonstration of the origin, extension, and early diagnosis of these tumors. We present a rare case of intraluminal leiomyosarcoma, which extended along the IVC into the right renal vein and right iliac vein, with ultrasonography (US), Doppler US, computed tomography (CT), and magnetic resonance imaging (MRI) findings to emphasize the significance of imaging methods in its diagnosis.

## Case report

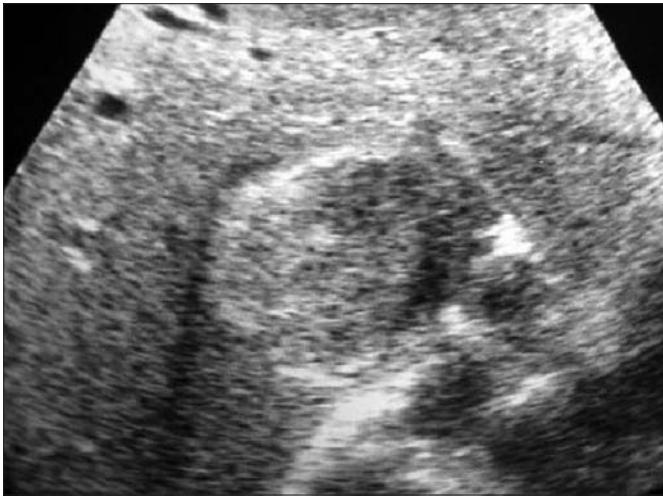
A 62-year-old female patient presented with epigastric and right upper quadrant pain, weight loss, and loss of appetite. Physical examination and laboratory findings were within normal limits. She had no history of abdominal or pelvic surgery. As the abdominal US of the patient demonstrated a mass in the IVC, Doppler US, abdominopelvic CT, and MRI examinations were performed.

Abdominal US demonstrated a heterogeneous soft tissue mass dilating the IVC (Fig. 1). The right hepatic vein was partially occluded (Fig. 2). Doppler US demonstrated peripheral low-resistance arterial flow in the intraluminal mass (Fig. 3). Contrast-enhanced abdominopelvic helical CT demonstrated central hypodense areas due to necrosis, and peripheral enhancement of the mass (Fig. 4). Extension of the tumor to the right renal vein and caudal right iliac vein was also demonstrated in CT (Fig. 5). A hypodense area of hypoperfusion secondary to the tumor thrombus in the right hepatic vein was detected in the right lobe of the liver.

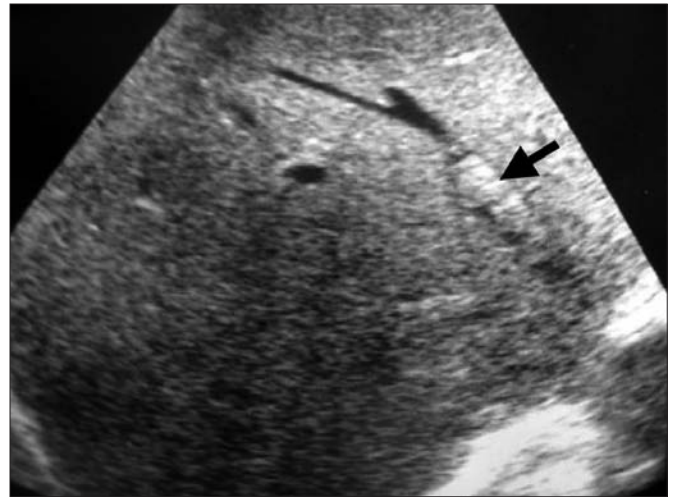
Abdominopelvic MRI clearly demonstrated the mass extending along and dilating the IVC on T2-weighted fat-saturated images (Fig. 6). Contrast-enhanced FLASH T1-weighted images demonstrated peripheral enhancement of the mass that was similar to CT findings (Fig. 7). Infiltration to surrounding tissues was not detected. Abdominopelvic CT and MRI examinations did not demonstrate prominent collateral venous circulation. Chest CT demonstrated multiple nodular lesions with smooth contours in the lung parenchyma that were consistent with metastasis. Tru-cut biopsy of the lung lesion was performed with an 18G needle under CT guidance which was diagnosed as leiomyosarcoma metastasis. This also confirmed that the mass in the IVC was primary leiomyosarcoma.

## Discussion

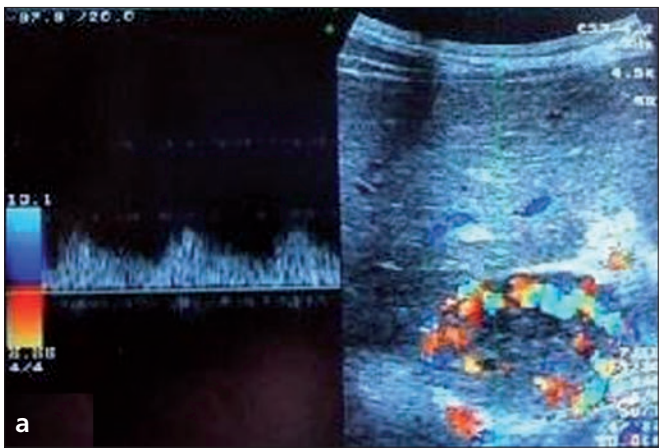
Primary vascular leiomyosarcoma is a rare tumor that originates from smooth muscle cells. More than 200 cases are reported in the literature



**Figure 1.** Abdominal ultrasonography examination demonstrates the heterogeneous, intraluminal soft tissue mass in the hepatic vein level dilating the inferior vena cava.



**Figure 2.** Abdominal ultrasonography examination demonstrates the right hepatic vein partially occluded by a tumoral thrombus (arrow).



**Figure 3. a, b.** Color Doppler ultrasonography examination in axial (a) and sagittal (b) planes demonstrates peripheral low-resistance flow in the intraluminal mass.

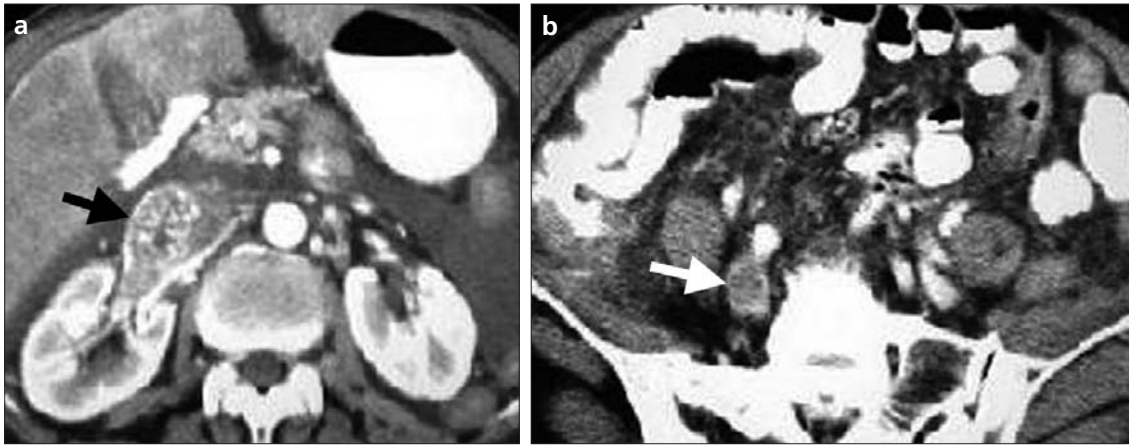


**Figure 4.** Contrast-enhanced axial CT image in the arterial phase demonstrates peripheral contrast enhancement and central necrosis of the mass.

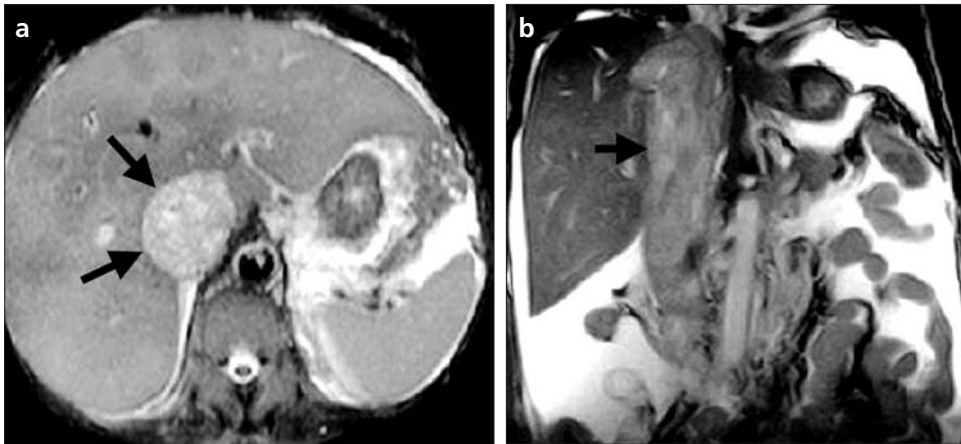
in the IVC. Upper segment involvement continues to the level of the renal veins and lower segment involvement is of the infrarenal portion. Tumors are most frequently located in the middle section of the IVC. The percentages of upper, middle, and lower section involvement are 24%, 42%, and 34%, respectively (8). Symptoms vary according to the dimension, growth pattern, and localization of the tumor. Upper section involvement might present with Budd-Chiari syndrome, middle section involvement with nephrotic syndrome, and infrarenal involvement with lower extremity edema (2, 9, 10). Patients generally present with non-specific symptoms, such as exhaustion, abdominal pain, and weight loss. Our patient presented with epigastric pain, and weight loss, although her tumor involved the middle and lower portions of the IVC.

(5). Although it is a rare tumor, it is the most common primary tumor of the IVC (6). It is most commonly detected in women in their sixth decade (7).

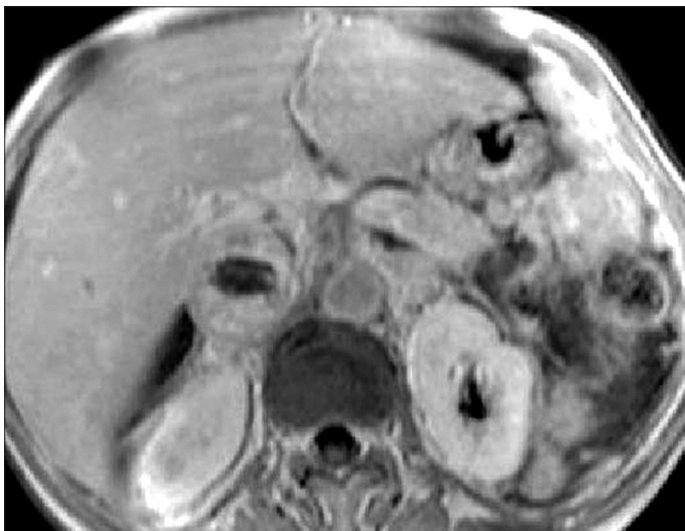
Vascular leiomyosarcoma might develop in all veins, but is usually located in the IVC. These lesions are classified into 3 groups according to localization



**Figure 5. a, b.** Contrast-enhanced CT images in the arterial phase (a, b) demonstrate the extension of the mass into the right renal vein (arrow, a) and right iliac vein (arrow, b).



**Figure 6. a, b.** Axial (a) and coronal (b) HASTE T2-weighted fat-saturated MR images demonstrate the tumoral mass (arrows) extending and dilating the inferior vena cava, starting from the hepatic vein level.



**Figure 7.** Axial contrast-enhanced FLASH T1-weighted MR image demonstrates peripheral intense enhancement of the mass.

vascularization and luminal dilatation were clearly demonstrated in all 3 of the imaging modalities employed in our case, and a blood thrombus was excluded with these findings.

Another condition that should be excluded in the differential diagnosis is intravascular leiomyomatosis (11). These lesions also present with an intravascular soft tissue mass and vascular dilatation, but they are even rarer than leiomyosarcomas. Extraluminal extension of leiomyosarcoma might be confused with masses of adjacent organs, such as the liver, uterus, or ovaries (12, 13).

Prognosis depends on tumor location and the existence of metastasis (14). Leiomyosarcoma of the IVC does not respond well to chemotherapy or radiotherapy. Surgical resection is the only choice of treatment if possible (15).

In conclusion, imaging features play an important role in the early diagnosis and evaluation of tumor extension of leiomyosarcoma located in the IVC. Contrast-enhanced CT, MRI, and color

Cross-sectional imaging methods play an important role in the early diagnosis and evaluation of the resectability of the tumor. Heterogeneous contrast enhancement is a characteristic feature in CT and is important in differentiating

the tumor mass from a blood thrombus. Tumor vascularity can be similarly demonstrated with Doppler US and contrast-enhanced MRI. Furthermore, dilatation of the IVC is an important finding of a tumor thrombus. Tumoral

Doppler US are especially important in the differential diagnosis of an intravascular mass and a blood thrombus.

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