Leiomyosarcoma of the inferior vena cava

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

References