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Case Report

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

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Tumors of the inferior vena cava are rare, and most of these are leiomyosarcomas. They are most frequent in the sixth decade of life, with a female predominance. We present a 45-year-old male patient with a tumor involving the entire extent of the inferior vena cava. Computerized tomography revealed a heterogeneously enhancing mass with marked expansion of the inferior vena cava with extraluminal extension. Computerized tomography–guided biopsy of the extraluminal component showed features of a fasciculated spindle cell tumor positive for vimentin, smooth muscle actin, and calponin and negative for S-100 antigen. We discuss the clinical presentation imaging findings and review the literature.

Key Terms: Leiomyosarcoma – Inferior vena cava – Computerized tomography – Magnetic resonance imaging

Leiomyosarcomas of the inferior vena cava (IVC) are rare slow-growing mesenchymal tumors, with ~218 cases reported in literature.[1] The tumor is most frequently seen in the sixth decade of life, with a female predominance.[2] The clinical presentation varies with the location and the pattern of growth (extraluminal versus intraluminal).[1,3] Slow growth, delay in diagnosis, and juxtaposed vital structures usually limit the extent of surgical resection.[4] We present a case report of a 45-year-old male patient with leiomyosarcoma involving the entire IVC, with review of literature.

Case Report

A 45-year-old male patient presented with dull abdominal pain and distension of the abdomen that was 4 months in duration. There was no history of fever or bladder and bowel complaints. There was no lower limb edema. Clinical examination revealed presence of ascites. An ascitic tap was performed, which revealed straw-colored fluid and protein of 3.9 g%. Cytology revealed few mesothelial cells. Gram stain and acid-fast bacillus stain were negative. Hemogram, liver function tests, and renal function tests were within normal limits. Ultrasonography of the abdomen revealed a lobulated mass in the retroperitoneum medial to the hilum of the right kidney. Contrast enhanced computerized tomography (CECT) of the chest and abdomen revealed a heterogeneously enhancing mass within the IVC extending from confluence of the iliac veins to the right atrium. The mass was expanding the IVC with dis-

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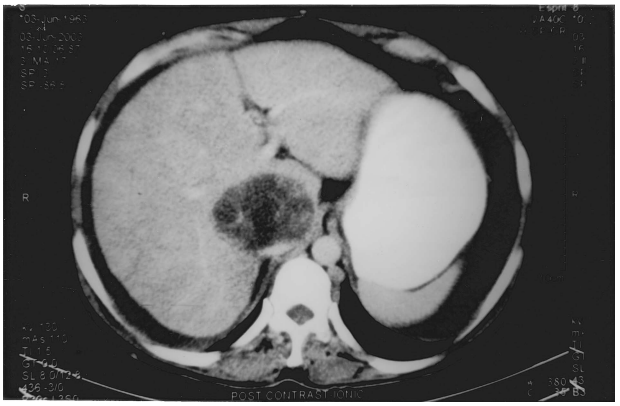

Fig. 1 Contrast enhanced CT scan shows a large well-defined heterogeneously enhancing, mass lesion causing marked expansion of the lumen of the IVC.

F1 placement of the adjacent structures (Fig. 1). Extraluminal extension of the mass was seen at the level of
F2 the renal veins with no definite capsule (Fig. 2). There was no calcification within the mass. Azygous and hemiazygous veins were enlarged with multiple retroperitoneal collaterals. The liver showed mottled enhancement, and the mass was seen obstructing the
AQ: A hepatic venous outflow. IVC gram revealed obstruction of the IVC with multiple retroperitoneal collat-
F3 erals (Fig. 3). A transvenous biopsy could not be attempted because of technical reasons. A CECT-guided biopsy of the extraluminal part of the tumor revealed features of a fasciculated spindle cell tumor with necrosis. Immunohistochemical reactions of the tumor cells were positive for vimentin, smooth muscle actin (SMA), and calponin and negative for S-100 antigen. Based on the location of the tumor and morphological and immunohistochemical findings, a diagnosis of intermediate grade leiomyosarcoma of the IVC was made. In view of the extent of tumor involvement, the patient was offered palliative radiotherapy. The patient subsequently died from the disease 6 months later.

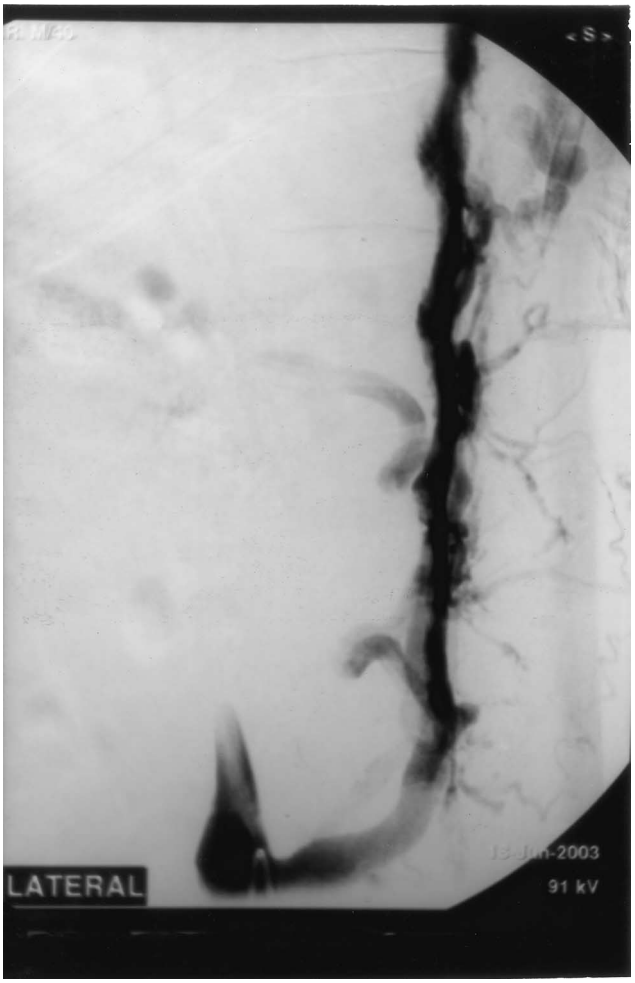

Fig. 3 IVC gram showing complete occlusion of the IVC lumen with extensive collaterals in the paravertebral region. AQ: B

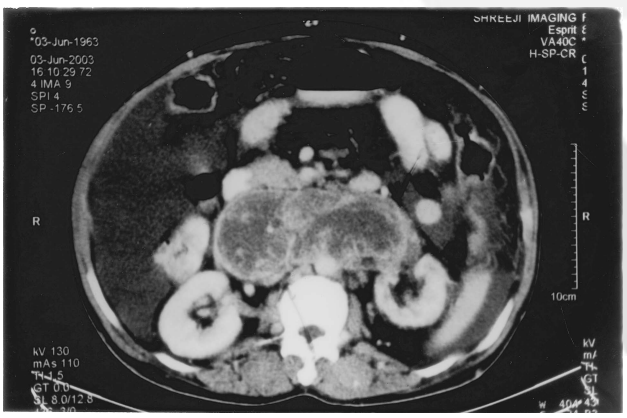

Fig. 2 Contrast enhanced CT scan shows a large, lobulated, heterogeneously enhancing, retroperitoneal mass arising from the IVC extending anterior to the aorta up to the left renal hilum.

Discussion

Tumors of the IVC are rare, and most are leiomyosarcomas. They are most frequently seen in the sixth decade of life with a female predominance.[2] Diagnosis is often not made until an advanced stage, because the symptoms are nonspecific, and they present late in the disease course. Symptoms vary

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according to the segment of the IVC involved, size, growth rate, and presence of thrombosis. Leiomyosarcoma of the IVC affects the middle segment (from the renal to the hepatic veins) in 42%, the lower segment (infrarenal IVC) in 34%, and the upper segment (from the hepatic veins to the right atrium) in 24% of patients.[2,5,6] In patients with tumors affecting the upper segment, symptoms include nausea, weight loss, Budd Chiari syndrome, and lower extremity edema. Cardiac symptoms, including arrhythmia, are encountered in patients with disease extending to the right atrium. Those affected by tumors involving the middle segment may present with epigastric and right upper quadrant pain. Disease of the middle segment may involve the kidney, resulting in renal vein thrombosis and nephrotic syndrome. Although uncommon, arterial hypertension may also result. Patients with disease involving the lower segment may experience right lower quadrant pain, back or flank pain, and leg edema.[7]

IVC leiomyosarcomas respond only minimally to chemotherapy or radiation, and surgical resection is the only potentially curative treatment.[1,7,8] Correct early preoperative diagnosis is important because it might offer a better chance of complete resection and prolonged survival. Leiomyosarcomas of the IVC are best depicted by CECT and magnetic resonance imaging (MRI). The findings vary depending on the predominant pattern of growth, which may be intra- or extraluminal. In patients with extraluminal growth, CECT reveals a large, lobulated, heterogeneously enhancing, retroperitoneal mass with low-density regions of necrosis. On MRI, in patients with extraluminal growth, a mass of homogeneous intermediate signal intensity on T1-weighted images and heterogeneous intermediate to high signal intensity on T2-weighted images is seen.[2,3,5,6,9] In cases characterized by intraluminal growth, CECT shows filling defects within a markedly dilated IVC. On MRI, a tumor mass with homogeneous intermediate signal intensity is seen on T1-weighted images. T1-hyperintensities corresponding to thrombi may also be present. On T2-weighted images, the tumor appears hyperintense, and thrombus formation is associated with markedly high T2-hyperintensity.[2,3,5,6,9] Purely intraluminal tumors will have to be differentiated from bland thrombus. Findings that suggest neoplastic thrombus include enlargement of the vessel lumen, enhancement of the mass, and neovascularity.[2,3,5,6,9] Sometimes, IVC tumors with a large extraluminal component may present a diagnostic dilemma. Such growth will have to be differentiated from tumors of hepatic, adrenal, renal, or other retroperitoneal origin. Clues helpful in suggesting the origin are location immediately adjacent to IVC and displacement rather than invasion of surrounding organs.[2] Our patient on CECT had both intra- and extraluminal tumor growth with marked IVC dilatation and neovascularity. Venography is helpful; it is sensitive but not always specific.[2] Venography shows either an intraluminal filling defect or compression and deviation of the IVC, depending on whether there is intra- or extraluminal tumor growth. It is important for evaluation of collateral venous circulation and planning of venous reconstruction. A transvenous biopsy can also be obtained for histopathological diagnosis.[10] Other ways of obtaining preoperative histopathological diagnosis include ultrasonography or CECT-guided biopsy.

Complete surgical resection offers the only chance for cure.[1,7] The surgical management depends on the location and extent of the tumor. Infrarenal lesions can be completely resected, and the vena cava can be safely ligated. Small tumors can be managed with partial resection of the vena cava and lateral suture of the remaining vein for flow reconstruction. However, when the extent of tumor burden extends to the renal vessels or the hepatic veins, venous reconstruction is pivotal.[7,11] Successful IVC reconstruction using expanded polytetrafluoroethylene grafts and bovine pericardium have been reported.[7,11] Because the venous flow is much slower than arterial flow and veins are thinner than arteries, the principles for reconstruction of arterial vessels are not thoroughly valid for veins. Sutures should be placed near each other to avoid continuous bleeding after anastomosis. Additionally, the prosthesis should be smaller than the vessel; some authors state that the smaller the prosthesis, the better the result.[7]

Literature review reveals an increased risk of death with upper IVC segment involvement, lower limb edema, Budd Chiari syndrome, intraluminal tumor growth, and IVC occlusion.[1] Radical tumor resection was associated with better 5- and 10-year survival rates (49.4% and 29.5%).[1] Patients with tumors that arose from the middle segment fared better (56.7% and 47.3%) than those of the lower segment (37.8% and 14.2%).[1] No palpable abdominal mass and abdominal pain were associated, in patients who had radical surgery, with a better outcome and longer survival.[1] Patients with positive surgical margins fared significantly worse compared with those who underwent complete resection.[7,8] Radiation therapy diminished local recurrence and may improve median survival. Patients who re-

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ceived adjuvant combined chemotherapy and radiation lived longer than those who did not.[8]

In conclusion, leiomyosarcomas of the IVC are rare tumors. Nonspecific symptoms often delay diagnosis, with many tumors presenting in advanced stages. CECT scan and, if available, MRI are specific in diagnosing and distinguishing this tumor from other retroperitoneal tumors as well as nonneoplastic thrombus of IVC.[2,3,5,6,9] Despite having a tumor that originates from the IVC, patients with this tumor type can enjoy reasonably long-term survival if the tumor is diagnosed early and is completely excised. Survival of these patients is no worse than of patients with leiomyosarcomatosis lesions of other origin.[1,8] Aggressive surgical management combined with adjuvant therapy offers the best treatment for patients with leiomyosarcoma of the IVC.[1,7,8]

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