



**ORIGINAL RESEARCH PAPER**

**Radiology**

**A RARE CASE REPORT ON LEIOMYOSARCOMA OF IVC**

**KEY WORDS:** Inferior vena cava, mesenchymal tumour, leiomyosarcoma of the inferior vena cava, computed tomography

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**ABSTRACT**

Leiomyosarcoma of the inferior vena cava is a rare tumour arising from the smooth muscle fibres of the media with a mean size at presentation generally around 12 cm (range 2–38 cm). This article comprises of role of CT (computed tomography) in evaluation of leiomyosarcoma of the inferior vena cava discovered incidentally and differentiating it from simple thrombus and/or other retroperitoneal tumours.

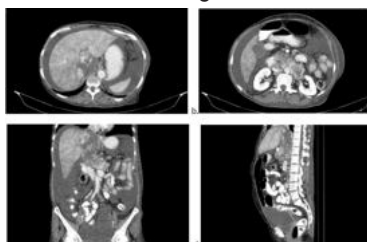
**Introduction:**

Leiomyosarcoma (LMS) of the inferior vena cava (IVC) is a very rare tumour (less than 1% of all malignant tumours) but is the most common primary IVC neoplasm. It is a mesenchymal tumour arising from the smooth muscle fibres of the media, more frequently in the sixth decade of life in women. The clinical and radiological features depend on its original site and on its pattern of growth (extra or intraluminal). Most of these tumours have a slow growth pattern and are located in the retroperitoneal space, so that symptoms are non-specific and occur later on. En bloc resection with negative margins is the gold standard treatment, but it is not always possible to carry out because the diagnosis is often delayed. Imaging plays an important role in preoperative staging of leiomyosarcoma. CT is useful for identifying, diagnosing and following up intravascular leiomyosarcoma.

**Case report:**

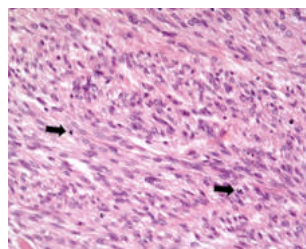
A 45 years old woman presented with 3 month history of epigastric pain, abdominal fullness and weight loss. At physical examination, the patient had epigastric and right upper quadrant pain and no mass was palpable at this location. Findings at electrocardiography and chest radiography were normal. Ultrasonography revealed hypoechoic rounded mass at the porta hepatis. For better characterization of the lesion CT abdomen and pelvis was performed.

CT showed the intrahepatic and infrahepatic part of IVC was irregularly distended and filled with approx. 38x47x128 mm sized complex heterogeneously enhancing intraluminal soft tissue density lesion. The intravascular tumour extended superiorly into the right atrium and inferiorly upto origin of bilateral renal veins. There was no calcification noted in the lesion. Lesion shows extraluminal extension in mid paraaortic region with associated significant fat stranding in adjacent space. No evidence of metastasis detected in other organ. There is presence of moderate to gross ascites was also noted.



**Figure: 1** Contrast enhanced CT axial images (a. & b.) showing irregularly dilated IVC with rounded heterogeneously enhancing intraluminal mass involving intra and infrahepatic portion of IVC with extraluminal extension in mid paraaortic region. A coronal (c.) and sagittal (d.) reformatted images showing longitudinal spread of the mass. Associated feature, moderate to gross ascites noted.

After USG guided biopsy, histopathological examination suggested a highly cellular spindle cell tumour, arising from the wall of the IVC. The tumour showed prominent mitotic activity (up to 40 mitotic figures/10 high power fields) and multiple foci of necrosis.



**Figure: 2** leiomyosarcoma with elongated cells some with larger and hyperchromatic nuclei.

**Discussion**

Leiomyosarcoma (LMS) of the IVC is the most common primary tumour of the IVC, although it is exceedingly rare, with only about 600 cases reported worldwide.

There is a 3:1 female predominance; hence, hormonal influence has been suggested, but not proven, as a risk factor. Leiomyosarcoma of the IVC is classified into three categories:

- Segment I: Infra-renal, presenting as lower extremity edema, right upper quadrant back, and flank, pain
- Segment II: Inter-renal and supra-renal, up to but not including the hepatic veins; presenting as abdominal and back pain, and sometimes renovascular hypertension.
- Segment III: Supra-hepatic, up into the right atrium; presenting with abdominal distention, nausea, vomiting, hepatomegaly, ascites, and jaundice.

Imaging plays an important role in preoperative staging of leiomyosarcoma, CT is useful for identifying, diagnosing and for follow up of intravascular leiomyosarcoma. the complex appearance of these tumours, CT is useful for differentiating a

neoplasm from simple thrombus. Leiomyosarcoma with extensive extravascular components are much more difficult to differentiate from retroperitoneal tumours that compresses or invade the inferior vena cava.

Although the imaging finding may strongly suggest a primary tumour of the inferior vena cava, biopsy is required, and USG guided biopsy is modality of choice for obtaining tissue for histological analysis.

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