Case 10366

Leiomyosarcoma of the inferior vena cava

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Section: Abdominal imaging
Area of Interest: Veins / Vena cava
Procedure: Diagnostic procedure
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 62 years, male

Clinical History:

A 62-year-old man with no remarkable past medical history presented with four months of progressive asthenia, weight loss and epigastric pain. Laboratory analysis revealed no anomalies.

Imaging Findings:

Abdominal ultrasound (not available) revealed a heterogeneous soft tissue epigastric mass of uncertain origin. Contrast enhanced abdominal CT demonstrated a heterogeneously enhancing mass involving the inferior vena cava from the suprarenal segment up to the hepatic portion (Fig. 1a, b). The liver presented a mosaic perfusion pattern probably secondary to obstruction of suprahepatic inferior vena cava (IVC) confluence (Fig. 2). The mass exhibited an expansive growth with intraluminal and transparietal components (Fig. 3a, b). Although there were no clear signs of adjacent organs invasion, no cleavage plane between the mass and the gastric antrum was observed either (Fig. 4).

There was also a small (1.8cm) exophytic cortical-based mass in the middle third of the right renal parenchyma (Fig. 3b). The right renal vein had no filling defect (Fig. 5).

A surgical biopsy was performed and revealed a moderately differentiated leiomyosarcoma.

Discussion:

Leiomyosarcomas of the IVC are very rare tumours arising from the smooth muscle vein wall [1]. The majority of the tumours occur in women in the 5th or 6th decade of life. Presenting clinical characteristics are directly related to the pattern of tumour growth. Upper section involvement (suprarenal portion) might present with Budd-Chiari syndrome as in our case, middle section involvement (renal portion) with nephrotic syndrome, and lower involvement (infrarenal portion) with lower extremity oedema [2].

Cross-sectional imaging methods play an important role in the diagnosis and evaluation of the resectability of the tumour. Heterogeneous contrast enhancement is a characteristic feature in CT and MRI and is important in differentiating the tumour mass from a blood thrombus. Also, lumen dilatation is characteristic of tumoural thrombus. In our case, tumoural vascularization and luminal dilatation were clearly present [2].

Tumours such as renal cell carcinomas, adrenal carcinomas, hepatocellular carcinoma or testicular tumours can secondarily invade IVC and should be considered as differential diagnosis. In our case, there was a small mass in the right kidney that could confound the diagnosis. However, it was highly unlikely that such a small mass could originate a tumoural thrombus so large without involving the renal vein [3].

Another condition that should be included in the differential diagnosis is intravascular leiomyomatosis. Although it
presents in a very similar way as leiomyosarcoma, it is even rarer [1].

Despite advancing imaging technology, a biopsy is still indicated for formal diagnosis. Histopathology of leiomyosarcoma reveals spindle tumour cells, which are positive for markers of smooth muscle activity including vimentin, muscle actin, alpha-smooth muscle actin, and desmin [3].

Aggressive surgical management combined with adjuvant therapy offers the best treatment for patients with leiomyosarcoma of the IVC.
The overall prognosis remains poor with a mean survival of 2 years but ranging from a few weeks to eight years [3].

**Differential Diagnosis List:** Leiomyosarcoma of the inferior vena cava, Leiomyosarcoma of the inferior vena cava, Renal cell carcinoma with tumoural thrombus, Intravascular leiomyomatosis

**Final Diagnosis:** Leiomyosarcoma of the inferior vena cava

**References:**


Description: Coronal section demonstrating an heterogeneously enhancing mass involving the inferior vena cava from the suprarenal segment up to the hepatic portion. Origin: Pinto J, Department of Radiology, Hospital de S. Sebastião, Santa Maria da Feira, Portugal
Description: Sagittal section demonstrating an heterogeneously enhancing mass involving the inferior vena cava from the suprarenal segment up to the hepatic portion. Origin: Pinto J, Department of Radiology, Hospital de S. Sebastião, Santa Maria da Feira, Portugal
**Figure 2**

*Description:* Inferior vena cava mass exhibiting an intraluminal growth. *Origin:* Pinto J, Department of Radiology, Hospital de S. Sebatião, Santa Maria da Feira, Portugal.
Description: Inferior vena cava mass exhibiting a transparietal growth. Right renal parenchyma exhibiting a small (1.8cm) exophytic cortical based mass. Origin: Pinto J, Department of Radiology, Hospital de S. Sebatlão, Santa Maria da Feira, Portugal.
Description: No cleavage plane between the inferior vena cava mass and the gastric antrum. Origin: Pinto J, Department of Radiology, Hospital de S. Sebatião, Santa Maria da Feira, Portugal.
**Description:** Right renal vein with no filling defect. **Origin:** Pinto J, Department of Radiology, Hospital de S. Sabatião, Santa Maria da Feira, Portugal.
Description: Budd-Chiari-syndrome presenting with hepatic mosaic perfusion pattern due to obstruction of the suprahepatic IVC confluence by the mass. Origin: Pinto J, Department of Radiology, Hospital S. Sebastião, Portugal