Leiomyosarcoma: a rare but fatal tumour of the inferior vena cava

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Accepted 9 April 2017

DESCRIPTION

Leiomyosarcomas (LMS) of the inferior vena cava (IVC) are a rare entity. Signs and symptoms vary greatly, such as abdominal pain, weight loss/gain, fever, weakness, anorexia or vomiting. CT scan is the imaging modality of choice for evaluation of retroperitoneal LMS, but biopsy is necessary to establish the diagnosis.

A 62-year-old woman presented with intermittent, diffuse, dull and sharp abdominal pain radiating to the lower back, associated with nausea, bloating and constipation. She also had a 15-pound weight gain in 6 months. Prior work-up did not reveal any acute abnormalities and she received treatment for constipation.

Due to persistent symptoms, CT of the abdomen and pelvis was ordered and demonstrated heterogeneously enhancing mass of the IVC extending into the right atrium (figure 1A,B). CT of the chest was done and confirmed the extension of the tumour to the right atrium (figure 1C,D). Figure 2 shows the

Figure 2 Core biopsy of the mass shows pleomorphic tumour cells with hyperchromatic nuclei (A,B) and focal necrosis (A, arrow). Tumour cells are positive for desmin (C), caldesmon (D) and p53 (E). Ki67 shows proliferation index of 15%–20% (F).

Figure 1 CT of the abdomen and pelvis with and without contrast. (A) Heterogeneously enhancing mass lesion expanding the intrahepatic and suprarenal segments of the inferior vena cava (IVC). (B) Extension into the right atrium. It also demonstrates foci of intense arterial enhancement with cystic spaces of necrosis. (C) Heterogenous hypervascular tumour in the entire suprarenal IVC, and the upper margin of the tumour extends into the right atrium. (D) The tumour within the right atrium measures 4.3×3.4 cm and does not extend to the tricuspid valve.

biopsy that diagnosed a low-grade LMS. Vascular surgery recommended neoadjuvant chemotherapy with gemcitabine and docetaxel in preparation for surgical intervention. The patient expired after her first chemotherapy treatment.

Although rare but fatal, LMS is the most common primary malignancy of the IVC and presents with non-specific symptoms. CT scan can provide a detailed characterisation of the tumour. At the time of presentation, tumours are usually large and aggressive surgical management with wide resection is challenging, leaving a limited role of neoadjuvant chemotherapy.

Learning points

Rare disease with challenging surgical technique is associated with high mortality.¹
High level of suspicion of leiomyosarcomas is indicated when signs and symptoms of vascular obstruction are the presenting clinical picture.

Contributors AA, MT, CL and HA are responsible for study conception and design and analysis and interpretation of data. AA and MT are responsible for acquisition of data and drafting of the manuscript. CL and HA are responsible for critical revision.

Competing interests None declared.

Patient consent Consent obtained from next of kin.

Provenance and peer review Not commissioned; externally peer reviewed.

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To cite: Albaghdadi A, Teleb M, Liu C, et al. *BMJ Case Rep* Published Online First: [please include Day Month Year]. doi:10.1136/bcr-2017-220151

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