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Splenomegaly

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A 64-year-old man presented with left-sided abdominal pain. He also complained of pain in his right shoulder and pelvis. A liposarcoma had been excised from his left sole six years earlier followed by local radiotherapy. There was no history of exposure to carcinogens. On clinical examination he had splenomegaly but no other significant findings. His blood profile showed a normochromic normocytic anaemia with normal white cells and platelets. Liver function tests and coagulation profile were normal. Ultrasound examination confirmed the splenomegaly. A computed tomography (CT) scan of the abdomen is shown in the figure. A bone scan was suggestive of metastatic disease.

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Figure CT scan of the abdomen

Questions

- 1 What is the abnormality shown?
- 2 How may the diagnosis be established?

Answers

QUESTION 1

There are multiple filling defects in the spleen of variable density and two focal lesions in the liver. Appearances are suggestive of angiosarcoma of the spleen with liver metastases.

QUESTION 2

Fine needle biopsy of the spleen has been reported but there is risk of rupture. Splenectomy provides a tissue diagnosis and prevents spontaneous rupture.

Percutaneous biopsy showed the presence of angiosarcoma. At laparotomy a grossly enlarged partly solid, partly cystic spleen was confirmed in addition to two focal nodules in the liver. Splenectomy was performed and the liver lesions biopsied. The splenic tissue was replaced by tumour deposits composed of vascular spaces and lined by atypical endothelial cells. Immunostaining for vascular markers (Factor VIII antigen and Qbend 10) was positive as was Vimentin. Cytokeratin staining (Cam 5.2) was negative. Histology of the liver lesions was similar, both lesions having the characteristics of angiosarcoma. The previously excised sarcoma from the sole was re-examined with confirmation of the original histology.

Discussion

Splenic angiosarcoma is a rare aggressive neoplasm with a poor prognosis. Metastases

occur at an early stage, especially to liver and bone.¹ The spleen may rupture and a presentation with abdominal pain may signify subcapsular haemorrhage. This may indicate, as in the present case, the need for splenectomy.¹ In the few reported series there appears to be a survival advantage to those undergoing splenectomy electively rather than acutely for haemorrhage.²

Biopsy of these lesions has the potential danger of blood loss and splenic rupture. Unfortunately routine imaging with ultrasound and CT has no specific diagnostic features. Enhanced magnetic resonance imaging may be the best available method but is not entirely specific.³

Haemangiosarcoma of the spleen is not associated with exposure to carcinogens unlike the same lesion in the liver. However, a concomitant malignancy was present in three of the 40 patients reviewed by Falk *et al*¹ and another had previously undergone chemotherapy for lymphoma. There have been no previous reports of an associated soft tissue liposarcoma. The metachronous presence of these two rare tumours would suggest there may be a causal environmental or genetic link.

Final diagnosis

Metachronous primary sarcomas

Keywords: angiosarcoma, liposarcoma, spleen

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