CASE REPORT

Rupture of splenic angiosarcoma: a rare cause of spontaneous haemoperitoneum

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SUMMARY

Primary splenic angiosarcoma, a very rare mesenchymal tumour of endothelial cell origin, comprises 2.6% of all cases of angiosarcoma and 10% of all primitive splenic tumours. Clinical presentation is usually unspecific, with abdominal pain and anaemia. Rupture is a rare complication and should prompt emergency splenectomy. Prognosis is usually poor because of liver, lung or bone metastases. We describe the case of an 80-year-old woman admitted to the emergency room with syncope, hypotension and vomiting. She stabilised after fluid resuscitation. Investigations showed anaemia, a large, heterogeneous spleen and free fluid in the abdominal cavity. She underwent emergency splenectomy. Pathology revealed primary splenic angiosarcoma. The postoperative period was complicated by respiratory failure but the patient made an otherwise uneventful course and was discharged 2 weeks after surgery. Six months after the operation she remains free of disease with no adjuvant treatment.

BACKGROUND

Spontaneous splenic rupture is a rare cause of haemoperitoneum and is usually caused by infections and tumours.[1] Splenic angiosarcoma is a rare primary splenic tumour and has been described as an extremely rare cause of rupture.[2–4] We present the case of a patient with acute abdomen and shock, in which only a moderately enlarged and heterogeneous spleen was found as probable cause. In the absence of a history of trauma, splenic rupture is a rare event but the treating clinician should nevertheless follow the principles of immediate management of intra-abdominal bleeding: resuscitation, confirmation of probable cause and definitive treatment.

CASE PRESENTATION

An 80-year-old woman was admitted to the emergency room after an episode of syncope, hypotension and vomiting. In the emergency room she was initially tachycardic and hypotensive but her haemodynamic status improved after fluid resuscitation with a fluid challenge of 500 ml of saline. She denied a recent history of trauma and her medical history included bilateral hip replacement 6 years earlier.

On clinical examination the patient had slightly discoloured skin and the abdomen was slightly distended and tender.

ECG and cardiac enzymes were normal. Complete blood count revealed anaemia (haemogobin 8.2 g/dl) and arterial blood gases were normal; except for elevated lactates (2.79 mmol/l).

Ultrasonography showed moderate amount of free fluid in the right and left paracolic gutters, with bowel gas precluding evaluation of solid organs or retroperitoneum. CT was performed at this stage and showed an enlarged, irregular and heterogeneous spleen with moderate amount of high attenuation fluid in the abdominal cavity (figures 1 and 2).

DIFFERENTIAL DIAGNOSIS

On an otherwise healthy elderly patient with syncope and haemodynamic collapse, the first diagnosis could be acute myocardial infarction or pulmonary embolism. The finding of abdominal tenderness on clinical examination would lead to consider other possible aetiologies: ruptured aortic aneurysm, mesenteric ischaemia, perforated peptic ulcer, severe acute pancreatitis and ruptured liver tumour. The finding of free fluid in the ultrasonography is suggestive of acute abdomen, but does not define its precise aetiology.

The CT findings of an enlarged, heterogeneous spleen with free fluid suggested spontaneous splenic rupture. Differential diagnosis includes infections, haematological disorders and malignancies.[1 5] However, occult splenic rupture after mild unknown trauma is another possibility.

TREATMENT

Oxygen was administered, two large-bore peripheral venous catheters were placed and the patient

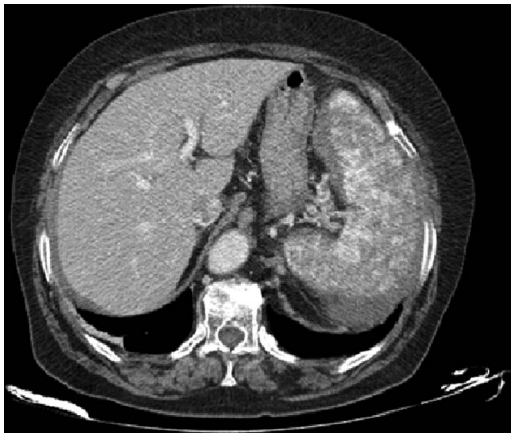

Figure 1 CT showing enlarged, irregularly shaped and heterogeneous spleen with free intra-abdominal fluid.

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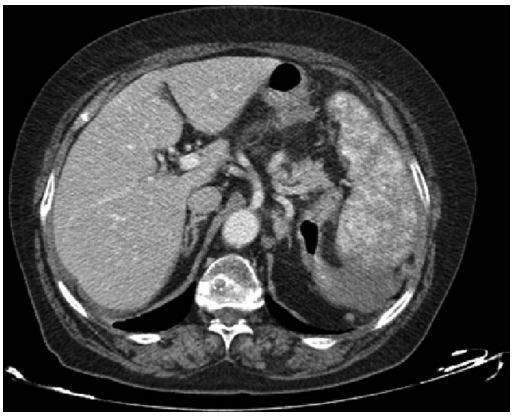

Figure 2 CT showing enlarged, irregularly shaped and heterogeneous spleen with free intra-abdominal fluid.

underwent exploratory laparotomy. Moderate haemoperitoneum and an enlarged spleen with spontaneous rupture and on-going bleeding were confirmed; splenectomy was performed. Intraoperatively the patient received 4 units of packed red blood cells.

OUTCOME AND FOLLOW-UP

The spleen weighed 242 g; histopathological examination disclosed areas of spindled cells and ectatic vascular spaces lined by atypical endothelial cells with CD31 and CD34 expression consistent with the diagnosis of splenic angiosarcoma (figures 3 and 4).

In the postoperative period the patient developed respiratory failure and was admitted to the intensive care unit for mechanical ventilation. She was extubated on the tenth postoperative day and discharged 4 days later.

Because of her advanced age no adjuvant treatment was initiated and, 6 months after the operation, she does not present any clinical, biological or radiological signs of recurrent disease.

DISCUSSION

Splenic angiosarcoma is a rare form of visceral angiosarcoma, comprising 2.6% of all cases of angiosarcoma and 10% of all primitive splenic tumours.[2 6] It usually affects patients in the fifth and sixth decades of life and has no particular preference for either sex.[3 4 7] Risk factors are a previous history of irradiation or exposure to vinyl chloride of thorium dioxide.[2 8] Our

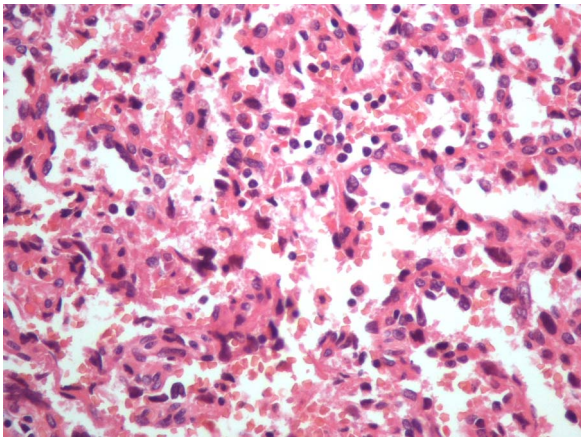

Figure 3 H&E staining showing areas of spindled cells and ectatic vascular spaces lined by atypical endothelial cells, with some mitosis evident (H&E ×100).

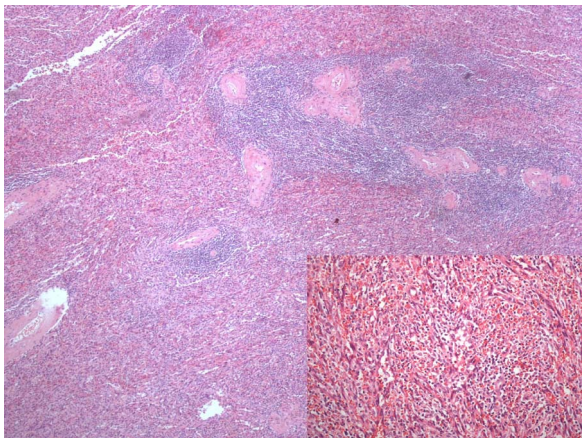

Figure 4 White pulp at the top and spindle cell proliferation (H&E ×100); inset (previous image ×200) pleomorphic spindle endothelial cells.

patient, however, gave no such history. The most frequent clinical presentation is abdominal pain, splenomegaly and systemic symptoms such as fever, weight loss and fatigue.[3 4 7] Blood tests usually show anaemia and thrombocytopenia.[3 4]

Rupture is a rare complication, with a reported incidence of 13–25%.[3 4] Diagnosis is usually by imaging, with CT revealing an enlarged spleen with heterogeneous masses, and simultaneous hyper and hypodense areas.[9] Complete parenchymal replacement by the tumour is another possibility[10 11] and distinction between spontaneous rupture of angiosarcoma and rupture of haematoma can sometimes be difficult.[12] In the case we report the spleen presented only a discrete enlargement.

The usual therapy of ruptured splenic angiosarcoma is emergency splenectomy.[3 4] Laparoscopic splenectomy has been described,[13] but is usually contraindicated in the setting of haemodynamic instability; as was the case in our patient. Splenic artery embolisation has been described as an alternative.[14]

Prognosis is usually poor, with overall survival less than 6 months. Recurrence usually occurs in the form of liver, bone or lung metastases.[3 4 7] Adjuvant therapy is controversial since there are no phase III trials and most reports are case series. However, paclitaxel seems a promising drug.[15] In the case we report the age of the patient contraindicated systemic therapy. On follow-up the patient is disease free 6 month after surgery.

In conclusion, this case illustrates an acute presentation of a rare tumour, in which successful management was owing to early recognition and management of intra-abdominal bleeding.

Learning points

- Splenic rupture is a rare cause of spontaneous haemoperitoneum and can be caused by primary or secondary splenic tumours.
- Management of a patient with shock in the emergency room should include simultaneous fluid resuscitation and aggressive search for the underlying cause. In suspected intra-abdominal bleeding investigation should me minimal and limited to assessing the origin of bleeding, either vascular or visceral.
- Splenic angiosarcoma is an aggressive disease and rupture, although rare, is life threatening.

Contributors All authors listed contributed in the article design, research, writing and reviewing.

Competing interests None.

Patient consent Obtained.

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