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CASE REPORT

Angiosarcoma of the Spleen with Spontaneous Rupture

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ABSTRACT

Primary splenic angiosarcoma is an extremely rare solid tumour. Splenic angiosarcoma is a highly aggressive malignancy associated with early metastases and poor prognosis. Most patients present with abdominal pain or a palpable abdominal mass. Spontaneous splenic rupture with fatal haemorrhage is a fairly common presentation and is reported in the literature in up to 30% of patients. This report is of a patient with spontaneous splenic rupture due to splenic angiosarcoma.

Key Words: *Hemoperitoneum; Splenic neoplasms; Splenic rupture; Splenomegaly; Tomography, X-ray computed*

中文摘要

自發性破裂的脾臟血管肉瘤

譚家盈、陳澤宗、呂沛欣、李醒芬

原發性脾血管肉瘤是一種極其罕見的實性腫瘤。脾血管肉瘤是一種高度侵襲性的惡性腫瘤，它會早期轉移，並且預後差。大多數患者因腹痛或可觸及的腹部包塊前來就診。自發性脾破裂引發致命性大出血是很常見的病徵，文獻報導有多達30%的患者會出現這情況。本文報告一宗因脾血管肉瘤而引致自發性脾破裂的病例。

INTRODUCTION

Primary splenic angiosarcoma is an extremely rare malignant tumour with high metastatic potential and poor prognosis despite treatment. However, splenic angiosarcoma is the most common primary malignant tumour of the spleen, affecting 0.14 to 0.25 per million people per year.[1] This neoplasm shows a slight male predominance, with no predilection for race, geographic location, or inheritability.[2] Different from hepatic angiosarcoma, splenic angiosarcoma shows no association with occupational exposure of chemicals such as vinyl chloride or arsenic. The mean age at presentation is at 50 to 60 years.[1,3] The clinical manifestations range from asymptomatic disease to splenic rupture with fatal haemorrhage. Splenic rupture occurs in 13 to 32% of patients as a presenting manifestation. Splenic rupture is the worst prognostic factor for survival because of increased risk of peritoneal dissemination of neoplastic tissue or vascular access for haematogenous spread. Splenectomy before organ rupture is associated with better survival rates.[4]

CASE REPORT

A 46-year-old woman presented to the Accident and

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Emergency Department at North District Hospital, Hong Kong, in September 2009 with sudden onset of left upper abdominal pain after a trivial trauma. She was in hypovolaemic shock with blood pressure measuring 70/40 mm Hg on arrival. At physical examination, the abdomen was distended with guarding and rebound tenderness. At presentation, blood tests showed normochromic normocytic anaemia with a haemoglobin level of 83 g/L (reference range, 120-150 g/L) and thrombocytopenia with a platelet count of 101 x 10^9 /L (reference range, 150-450 x 10^9 /L). Her blood clotting profile showed a minimally prolonged activated partial thromboplastin time (APTT) of 30.8 seconds (reference range, 25-40 seconds) and prothrombin time (PT) of 15.1 seconds (reference range, 10-13 seconds), with an international normalised ratio of 1.2.

After resuscitation, urgent contrast computed tomography (CT) of the abdomen and pelvis showed extensive haemoperitoneum with a sentinel clot at the perisplenic region (Figure 1a). The spleen was enlarged (polar length, 15 cm) and showed heterogeneous parenchyma. Contrast extravasation was seen at the dependent region posterior to the spleen (Figure 1b). The spleen showed heterogeneous enhancement with multiple small enhancing nodules (Figure 2a). Enhancement defects compatible with thrombus were present in the splenic vein (Figure 2b). Non-specific

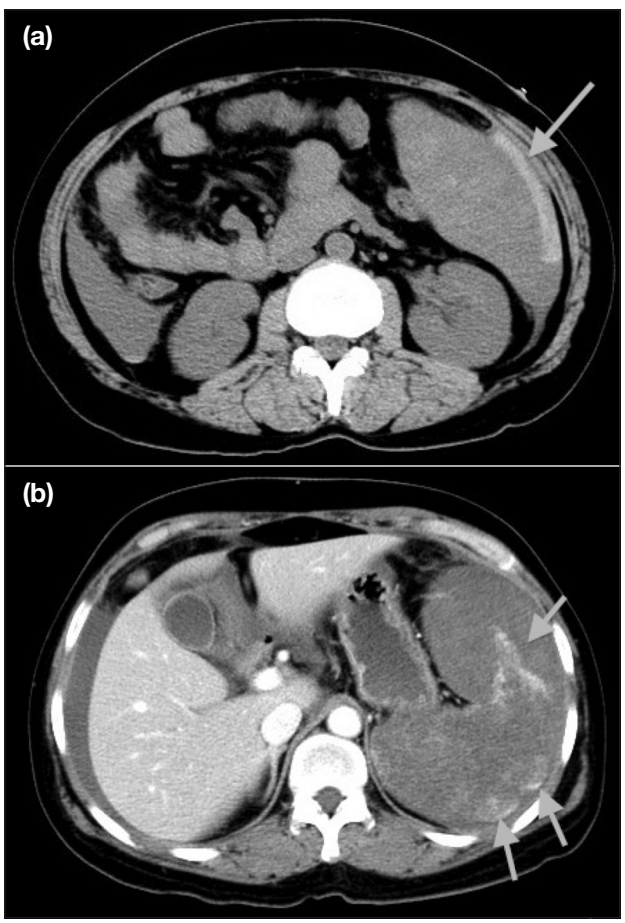

Figure 1. Computed tomography scans of the abdomen and pelvis showing (a) high attenuation fluid at the prehepatic space suggestive of haemoperitoneum with a sentinel clot at the perisplenic region (arrow); and (b) gross haemoperitoneum and contrast extravasation at the dependent region of the spleen (arrows).

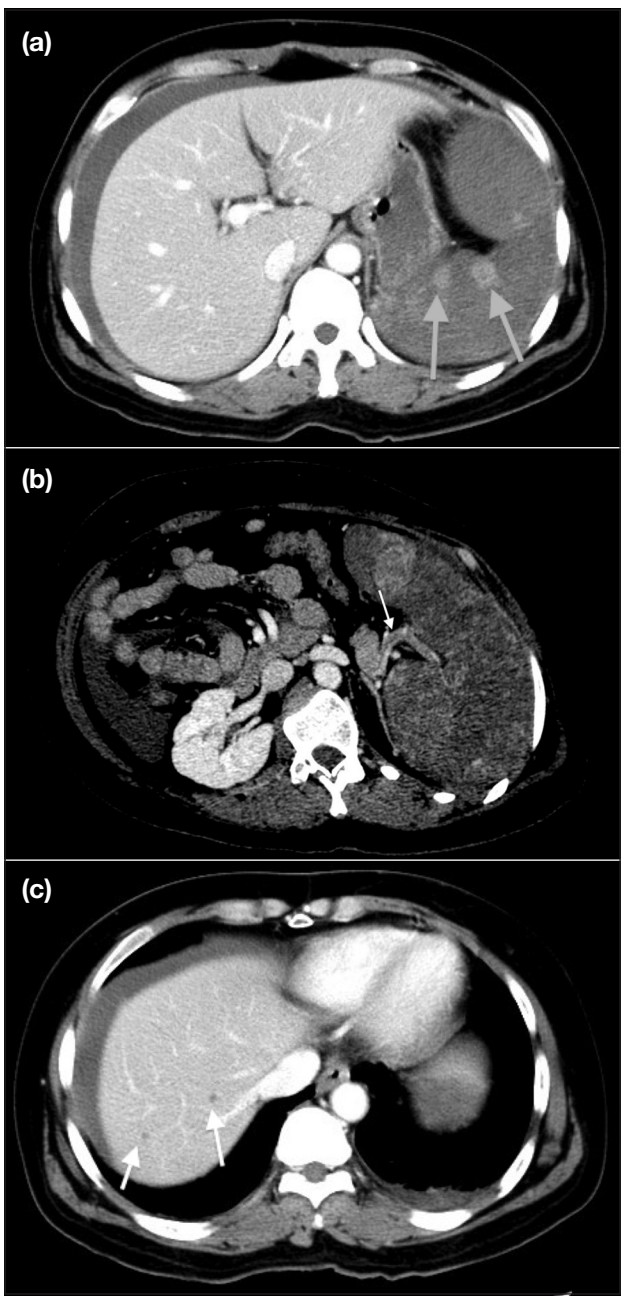

Figure 2. Computed tomography scans of the abdomen and pelvis showing (a) enhancing nodules within the enlarged spleen (arrows); (b) splenic vein thrombosis (arrow); and (c) non-specific hypodense lesions in the right lobe of the liver (arrows).

hypoenhancing lesions were present in both lobes of the liver (Figure 2c). A small amount of left pleural effusion was present.

The patient underwent emergency laparotomy, which revealed a massive haemoperitoneum with at least 4 litres of fresh blood. Intraoperative findings showed that the spleen was enlarged and congested. Active spurting of blood was present at the superior pole. Haemostasis and emergency splenectomy were performed. After surgery, her platelet count decreased to 14 x 10^9 /L.

The surgical specimen weighed 216 g and measured 15 x 6 x 5 cm. Capsular rupture measuring 4 x 2 cm was evident at the superior pole. Pathology examination of the spleen showed generalised congested and haemorrhagic splenic parenchyma and multiple nodules with haemorrhagic foci. The spleen tissues were nearly completely replaced by a high-grade tumour with immunohistochemical profiles compatible with a diagnosis of angiosarcoma of the spleen (positive for CD31 and CD34). No vascular thrombosis was detected in the resected specimen.

Postoperative fluorodeoxyglucose (FDG) positron emission tomography (PET) scan was negative. Although there was no evidence of an FDG-avid lesion on the PET scan, the patient completed a course of adjuvant chemotherapy (intravenous doxorubicin 50 mg/m^2 for 6 cycles) in view of the splenic rupture and high metastatic potential of splenic angiosarcoma. She is currently being followed up in the Department of Oncology.

Follow-up CT performed 9 months after the initial surgery showed that the liver was enlarged and infiltrated by numerous small hypoenhancing nodules. The lesions had increased in size and number compared with the preoperative CT (Figure 3). These lesions were compatible with metastatic infiltration of the liver.

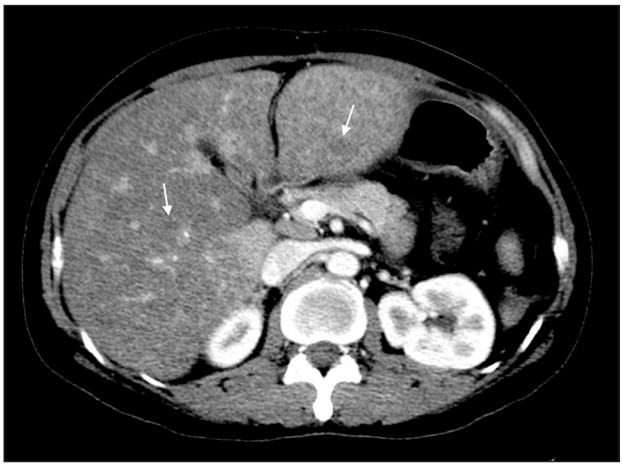

Figure 3. A follow-up computed tomography scan showing hepatomegaly infiltrated by multiple ill-defined nodules compatible with metastatic lesions. Some of the more discrete lesions are indicated by arrows.

DISCUSSION

Primary splenic angiosarcoma arises from the endothelial lining of the splenic blood vessels. Primary splenic angiosarcoma is the most common non-haematolymphoid malignant tumour of the spleen. Most patients have a very poor prognosis and die within 12 months of diagnosis.[1,3] Distant metastases occur most frequently in the liver (70% of cases) as in this patient, followed by the lungs, pleura, lymph nodes, bone, and brain.[1,3] The largest series reported that 67% of 12 patients had evidence of liver metastases at initial presentation.[5] In this patient, the non-specific hepatic lesions enlarged and were confirmed to be hepatic metastases at a follow-up scan after 9 months.

The most common symptom of splenic angiosarcoma is upper abdominal pain, which has been reported in up to 67% of patients in a case series.[5] Spontaneous splenic rupture is a fairly common complication, and has been reported in up to 30% of patients.[1,3,6] Spontaneous rupture is the most serious manifestation of splenic angiosarcoma. Spontaneous rupture is the worst prognostic factor for survival because of increased risk of peritoneal dissemination with direct implantation or vascular access for haematogenous spread. Patients undergoing splenectomy before splenic rupture have been found to have better survival rates than those presenting with splenic rupture.[4]

It has been reported that splenic angiosarcoma is associated with consumptive coagulopathy or disseminated intravascular coagulopathy due to profound fibrinogenopenia and a lack of systemic fibrinolysis activation.[7] Together with thrombocytopenia, fibrinogenopenia leads to profuse intraperitoneal haemorrhage. Consumptive coagulopathy was not observed in this patient. However, the minimally prolonged APTT and PT and thrombocytopenia could have been due to splenomegaly and could partially account for the spontaneous splenic rupture and profuse bleeding.

CT scan is a useful imaging tool for assessment of splenic angiosarcoma. Splenomegaly is the most common finding, and the spleen is usually replaced or infiltrated by complex masses. The tumour masses commonly show heterogeneous enhancement with areas of central necrosis. Liver metastases are common. Ultrasonography has also been used for evaluation of suspected splenic angiosarcoma. Splenomegaly with heterogeneous echogenicity or hypoechoic areas suggestive of tumour necrosis are the common findings. For magnetic resonance imaging, splenomegaly is a common finding. The tumour usually shows decreased signal intensity with respect to normal splenic parenchyma on T1-weighted image. Occasionally, internal hyperintensity may be noted due to acute bleeding. On T2-weighted image, the tumour shows high signal intensity. Internal areas of decreased signal intensity may be evident due to the presence of old blood with haemosiderin deposits. With gadolinium administration, heterogeneous enhancement will be shown.[5] A recent case report demonstrated FDG uptake in a primary splenic angiosarcoma and in the surgical bed 5 months after splenectomy.[8] Hence, FDG PET is potentially a useful tool for tumour staging and detection of residual or recurrent disease.

The most common differential diagnoses for splenic angiosarcoma are metastatic lesions, lymphoma, haemangioma, littoral cell angioma, lymphangioma, haemangiopericytoma, and epithelioid vascular tumour. Lymphoma and metastatic lesions are the most common differential diagnoses and may mimic hypovascular angiosarcoma. For lymphoma, splenomegaly is found in up to 80% of patients.[6] There is usually evidence of diffuse homogeneous enhancement on CT with variable appearance including a solitary mass, multifocal lesions, or diffuse infiltration with organomegaly. Enlargement of the abdominal lymph nodes may help to distinguish lymphoma from splenic angiosarcoma as splenic angiosarcoma rarely metastasises to the abdominal lymph nodes. For metastatic lesions, almost one-half of all splenic metastases are from primary melanoma, followed by breast and lung carcinoma. Although metastatic splenic lesions could mimic a relatively avascular angiosarcoma, the diagnosis will be obvious if there is a known primary tumour.

In the CT image for this patient, splenic vein thrombosis was observed as small filling defects in a focal segment of the splenic vein anterior to the pancreatic tail, but this was too small to demonstrate any internal enhancement to suggest tumour thrombus. This could have been either a tumour thrombus or a thrombus caused by altered splenic haemodynamics. Splenic vein thrombosis was not documented in the pathology report for the resected spleen specimen. It is possible that the thrombosed segment was located away from the splenic hilum, which might not have been included in the resected specimen.

CONCLUSION

Splenic angiosarcoma is a rare solid tumour, although it is the most common primary malignant neoplasm of the spleen. Splenic angiosarcoma has a poor prognosis despite treatment. Spontaneous splenic rupture with fatal haemorrhage is a fairly common presentation, and significantly affects the prognosis.

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