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Splenic Angiosarcoma – An Unusual Cause of Bleeding Gastrointestinal Tract

Y F A Chung, I Busmanis, G S Hong, K C Soo

ABSTRACT

Splenic angiosarcoma is a rare malignant vascular tumour with about 100 reported cases to date. The presentation of splenic angiosarcoma is highly variable, frequently causing diagnostic difficulty. It usually presents with splenomegaly, abdominal pain and occasionally with a microangiopathic type of anaemia. Here we report an additional case of primary angiosarcoma of the spleen presenting as a problem of bleeding from the gastrointestinal tract.

Keywords: splenic angiosarcoma, gastrointestinal haemorrhage

INTRODUCTION

Primary angiosarcoma of the spleen is rare, with only about 100 reported cases to date[1-5]. It was first described in 1879 by T Langhans[6]. It occurs almost equally in females and males with the peak incidence in the sixth decade. The symptomatology is highly variable, often posing difficult diagnostic problems and it is usually discovered at autopsy.

CASE REPORT

Mr OLH, a 54-year-old Chinese gentleman was admitted on 29 December 1994 for passing of melena for 2 days. He was pale but there was no abdominal mass. Per rectal examination revealed stale melena. Initial full blood count picture showed the haemoglobin to be 3.8 g/dL, total white count 9.6 x 10^9/L and the platelets 25 x 10^9/L. Blood film showed normocytes with few microcytes. The clotting profile and liver function test were normal. Serum iron was 4 μmol/L and total iron binding capacity was 46 μmol/L. Bone marrow biopsy showed a regenerating marrow with high reticulocytes count of 2.2%. Gastroscopy revealed non-erosive duodenitis. Eight units of blood were transfused. The bleeding source was then assumed to be from the duodenitis.

He was readmitted on 12 July 1995 with complaints of lethargy, shortness of breath and fainting spells. He had mild hepatomegaly and splenomegaly. Investigations showed the haemoglobin to be 3.7 g/dL and the platelets at 61 x 10^9/L. The blood film revealed a mixed picture of normocytes and hypochromic microcytes. A repeat serum iron and total iron binding capacity revealed results of 4 and 62 μmol/L respectively. Upper and lower gastrointestinal endoscopy did not reveal any bleeding sites. Small bowel series and Meckel's scan did not show any abnormality in the mid-gut. Red blood cell radionuclide tagged scan also did not reveal any bleeding points. Repeat colonoscopy was normal. He was transfused a total of 22 units of blood and was discharged as requested, with a haemoglobin of 9.7 g/dL. While in hospital, his platelet counts fluctuated between 26 and 61 x 10^9/L.

He was admitted again on 15 August 1995 for passing of melena for 2 days which was associated with epigastric discomfort. Angiogram (selective gastroduodenal, superior mesenteric and inferior mesenteric arteries) failed to show any active bleeding site. Repeat red blood cell radionuclide tagged scan showed evidence of active bleeding from the distal small bowel. He underwent a laparotomy on the same day. Intraoperative enteroscopy via an enterotomy and colonoscopy did not reveal any bleeding sites. Intraoperative gastroscopy revealed bleeding gastric erosions. A gastrotomy and underrunning of the erosions was done. Intraoperatively, the liver and spleen were noted to be enlarged with dilated vascular channels interpreted as haemangiomatas. He was subsequently discharged.

He was readmitted on 9 September 1995 complaining of passage of melena. Imaging of the abdomen was done to identify the extent of the haemangiomata.

CT scan revealed multiple low attenuation nodules in the liver and spleen with the largest in the spleen (Fig 1). There were also multiple similar lesions in the lung, lumbar vertebra and enlarged intra-abdominal lymph nodes.

In an attempt to control the thrombocytopenia, splenectomy was performed on 27 September 1995.

The splenic mass measured 20 x 14 x 12 cm, weighed 1.05 kg, and was composed predominantly of solid dark red-brown tissue, including some large dilated vascular spaces filled with thrombus, and yellow areas of necrosis (Fig 2). Histologically, focally necrotic tumour consisted of innumerable closely packed thin-walled vascular channels, with a surrounding dense spindle cell population showing

Department of General Surgery
Singapore General Hospital
Outram Road
Singapore 169608

Y F A Chung, MBBS, FRCS (Edin)
Registrar

G S Hong, MBBS, MSc (Lond), FRCS (Glas)
Consultant

K C Soo, MBBS, MD, FRACS
Clinical Associate Professor and Head

Department of Pathology
Singapore General Hospital

I Busmanis, MBBS, FRCPA
Consultant

Correspondence to:
Dr G S Hong

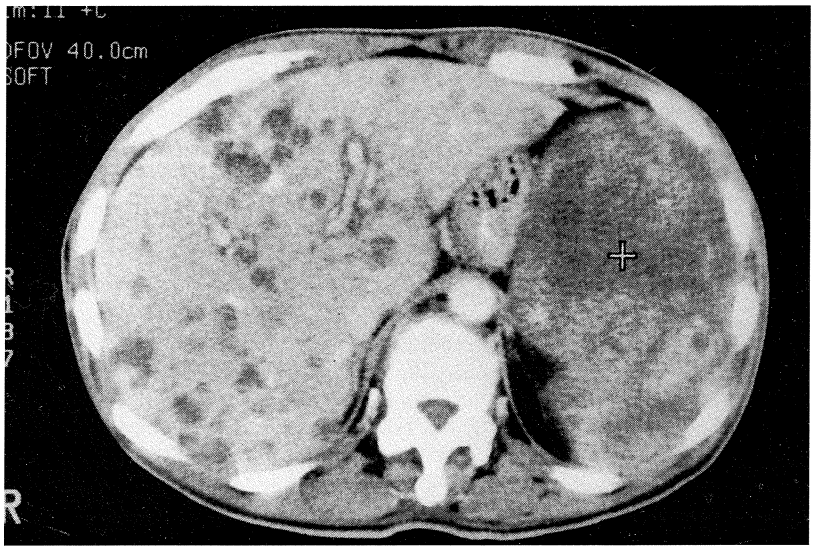

Fig 1 – CT scan showing angiosarcoma of the spleen with liver secondaries.

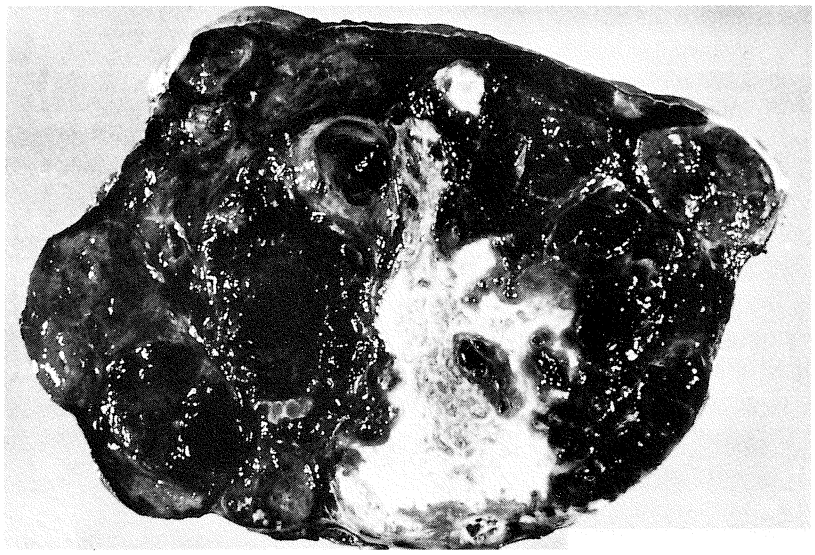

Fig 2 – Dark red haemorrhagic cut surface of spleen showing pale irregular foci of necrosis.

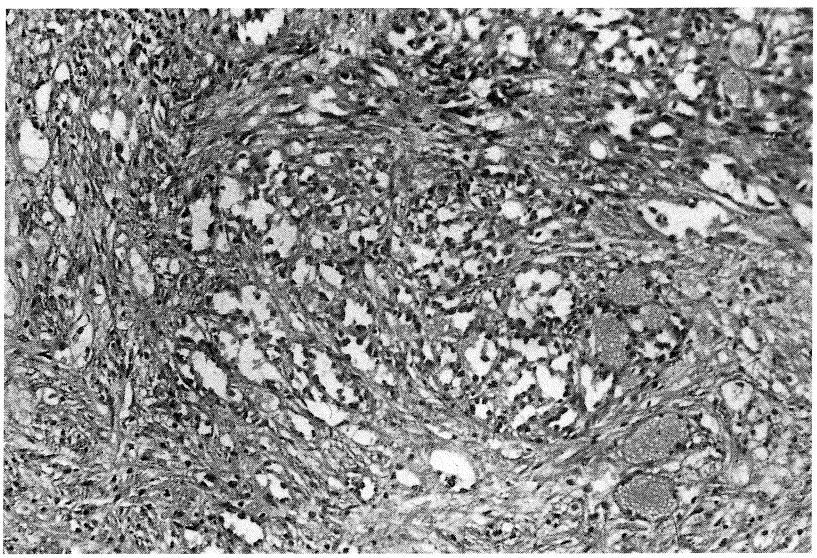

Fig 3 – Angiosarcoma composed of closely packed vascular channels lined by atypical endothelial cells (H & E stain x 400).

occasional mitoses. The blood vessels were lined by atypical endothelial cells with enlarged hyperchromatic nuclei (Fig 3). No residual normal spleen was identifiable.

He was subsequently treated symptomatically and he expired at home on 27 October 1995.

DISCUSSION

The first reported case of splenic angiosarcoma with liver metastases was in a 30-year-old man who died 5 months after presenting with abdominal pain. Diagnosis was made at autopsy. Most of the cases reported since then had been discovered at autopsy and in the few instances of antemortem diagnosis, the patients were operated on for splenic rupture. The largest reported series consisted of 40 cases by Falk et al[5].

Splenic angiosarcoma commonly present with splenomegaly and left sided abdominal pain in 92% and 83% of cases respectively[5]. Systemic symptoms include weight loss (40%), fever (10%) and fatigue (5%). Haemoperitoneum from splenic rupture has been reported in about one third of cases[1,3,4]. Abnormal laboratory investigations are common with cytopenia in 91% of cases [commonest being anaemia, thrombocytopenia and pancytopenia], leucocytosis (38%) and thrombocytosis (3%)[5]. Deranged coagulation parameters (elevated prothrombin and/or partial thromboplastin times) may be present (8%). Diagnosis is often made at an advanced stage with the metastatic rate at 69% – 86%[1,3,5]. Metastases in the liver are common (80%), while metastases to other organs such as the lungs, lymph nodes, and bone occur in approximately one third of cases[1,3,5].

The diagnosis of primary splenic angiosarcoma was difficult in our case as it had an unusual presentation with bleeding gastrointestinal tract as the chief complaint. To our knowledge, only one other case of gastrointestinal haemorrhage has been previously reported[8]. This was due to angiosarcoma of the small intestine itself. Blix and Jacobson published a case of splenic angiosarcoma associated with consumptive coagulopathy but with no mention of gastrointestinal tract bleeding[7]. This mechanism was thought to be due to the consumption of platelets and fibrinogen during stasis within the tumour in the spleen.

Laboratory investigations revealed only thrombocytopenia and anaemia from the chronic blood loss in our case. In splenic angiosarcoma, the anaemia is typically normochromic/normocytic with bizarre cells consistent with those of microangiopathic haemolytic anaemia[1-3]. It is thought that there is direct specific damage by the irregular tumour vascular endothelium and fibrous strands. As a result, the features of haemolysis with large numbers of damaged red cells presenting as burr cells or schistocytes may be seen. In our patient, the primary angiosarcoma of the spleen caused thrombocytopenia which resulted in repeated bleeding from the gastrointestinal tract. Extensive gastrointestinal endoscopy and radiological

imaging failed to identify any bleeding source. Thus the chronic blood loss was reflected in a blood profile which was consistent with an iron deficiency anaemia and not the reported microangiopathic haemolytic anaemia.

Imaging remains the key to diagnosis. The two distinctive reported radiological patterns observed are: total tumour replacement of the spleen with preservation of normal contour or multiple nodular masses in the spleen[9]. The organ that contains the largest single tumour should be considered as the primary site of angiosarcoma. In our case, the spleen was the site of the largest mass of tumour, while the liver contained multiple nodules.

Histogenesis of splenic angiosarcoma is still uncertain. They are believed to be either malignant degenerations of haemangiomas or de novo tumours from cells of the splenic sinus wall[2]. Epidemiological factors of importance in hepatic angiosarcoma such as Thorotrast or vinyl chloride exposure, have not been implicated in splenic angiosarcoma. Macroscopically, the cut surface revealed replacement by numerous haemorrhagic nodules in most instances, although rarely, there may be diffuse involvement as in our case. Typically, the formation of vascular channels by neoplastic cells allows histological confirmation. In doubtful cases, ultrastructural techniques, reticulin and immunohistochemical staining of factor VIII-related antigen may be useful[3,5].

The prognosis of splenic angiosarcoma is uniformly poor with a short median survival of 6 months[5]. Therapeutic possibilities are limited. There has been no evidence of clinical benefit of chemotherapy. However upon diagnosis, splenectomy before rupture is advisable with significantly improved survival[1,5].

Diagnosis of splenic angiosarcoma demands a high index of suspicion and should be considered in patients with splenomegaly with microangiopathic anaemia but without evidence of lymphoma or leukemia. Despite this clinical awareness and improved imaging, the prognosis of splenic angiosarcoma will continue to be dismal because of its rarity and inherent aggressive metastatic potential.

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