

Giant cavernous haemangioma of the spleen presenting as massive splenomegaly and treated by partial splenectomy

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ABSTRACT

Cavernous haemangioma is a rare disorder of the spleen with fewer than 100 cases reported. Only rarely do they attain large sizes. A 36-year-old woman presented with a six-month history of pain in the left hypochondrium and a massive splenomegaly. Ultrasonography, Doppler studies, and computed tomography could not distinguish between a haemangioma and a secondary deposit. Magnetic resonance imaging showed characteristic features of splenic haemangioma with central fibrosis, thrombosis and haemorrhage. Partial splenectomy was done. Intraoperative imprint cytology was negative for malignant cells. Histopathology showed cavernous haemangioma with areas of infarction necrosis. It is extremely rare to have such a massive solitary splenic haemangioma presenting as a giant splenomegaly. Preoperative investigations are often inconclusive and may not distinguish between haemangioma and metastases. Magnetic resonance imaging is the most reliable imaging method. Haemangiomas are treated only when they are symptomatic or very large with an increased risk of haemorrhage. Partial splenectomy is the treatment of choice.

Keywords: cavernous haemangioma of spleen, giant splenomegaly, haemangioma, partial splenectomy, splenic haemangioma

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INTRODUCTION

Cavernous haemangioma is a rare disorder of the spleen with fewer than 100 cases reported in literature.⁽¹⁾ Nonetheless, it is the most common primary neoplasm of the spleen.^(2,3) They are usually less than 2 cm in size, and only rarely attain a large size.⁽⁴⁾ Splenic haemangiomas are usually asymptomatic and discovered incidentally.⁽¹⁾ They are not treated unless they are symptomatic or very

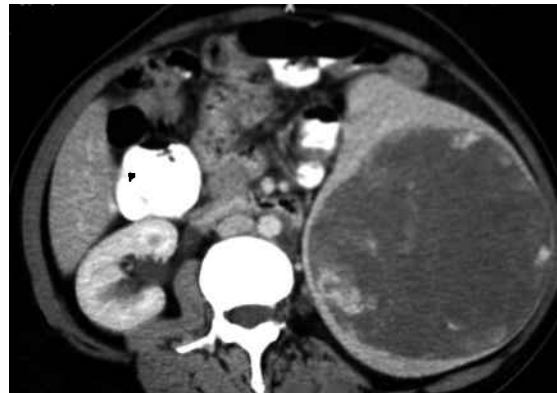


Fig. 1 Contrast-enhanced CT image of the splenic lesion shows peripheral enhancement.



Fig. 2 Contrast-enhanced coronal T1-W MR image shows massive splenomegaly due to a large, solid, heterogeneous mass lesion involving the lower pole of the spleen.

large with an increased risk of haemorrhage.⁽⁴⁾ We report a rare case of a giant, solitary cavernous haemangioma of the spleen presenting as a massive splenomegaly, and provide a review of the literature.

CASE REPORT

A 36-year-old woman presented to the surgical outpatient department, Lady Hardinge Medical College, New Delhi, with occasional pain in the left upper abdomen for the

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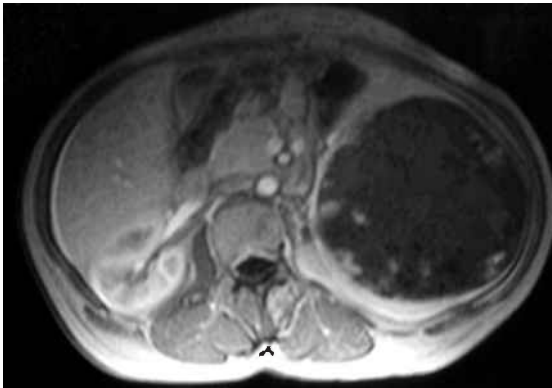


Fig. 3 Contrast-enhanced axial T1-W MR image shows peripheral enhancement of the splenic haemangioma.

last six months, with progressively increasing dragging sensation in the left side of her abdomen. There was no history of fever, weight loss, or anorexia. The general physical examination was unremarkable. On abdominal examination, the spleen was grossly enlarged, extending almost up to the umbilicus, with a smooth surface and firm consistency. Blood counts and serum chemistry were within normal limits.

Ultrasonography (US) of the abdomen showed a large heterogeneous mass, measuring 12.2 cm × 10.6 cm, located in the left lumbar region, indenting the left kidney, probably arising from the spleen. The liver was normal and there was no free fluid in abdomen. Contrast-enhanced computed tomography (CECT) of the abdomen showed a large, heterogeneous, predominantly cystic lesion 10.6 cm × 11.2 cm in the lower pole of the spleen (Fig. 1), suggestive of a haemangioma or a hydatid cyst. Hydatid serology was negative. Doppler US showed a 10 cm × 11 cm size round, well-defined, mixed echoic space-occupying lesion seen at the lower pole of the spleen, with echoes and cystic areas within it. Prominent vessels were seen at the periphery of the mass showing high velocity external flow with scanty vascularity at the centre, suggestive of a haemangioma. However, with the mass having mixed echogenic and large necrotic components, the possibility of malignancy could not be ruled out. Magnetic resonance (MR) imaging of the abdomen showed an enlarged spleen with a solid heterogeneous mass lesion, measuring 10 cm × 12 cm, involving the lower half of the spleen (Figs. 2 & 3). The lesion was hypointense on T1-weighted images and showed peripheral hyperintensity on T2-weighted images with central hypointense areas, suggestive of fibrosis, thrombosis, and haemorrhage. Contrast-enhanced MR imaging showed early peripheral nodular enhancement with multiple non-enhancing areas characteristic of a splenic haemangioma.

The patient was explored by a left subcostal incision

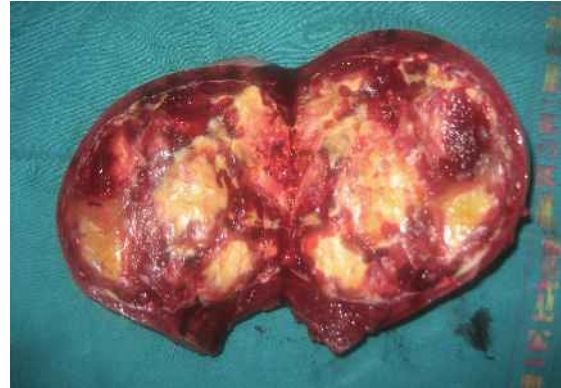


Fig. 4 Photograph of the cut surface of the splenic lesion after partial splenectomy.

under general anaesthesia. Peroperatively, there was a giant splenomegaly with a large solid lesion confined to the lower half of the spleen. Partial splenectomy was done after ligating the segmental blood supply to the lower half of the spleen. The resected specimen was bisected (Fig. 4), and imprint cytology was sent, but did not show any evidence of malignant cells. Postoperative recovery of the patient was uneventful. Histopathology of the partial splenectomy specimen showed a large cavernous haemangioma occupying almost the entire partial splenectomy specimen with large areas of infarction necrosis.

DISCUSSION

Although unusual, haemangioma is the most common primary splenic neoplasm. They are usually less than 2 cm in size.⁽⁴⁾ Haemangioma presenting as giant splenomegaly is very rare. Haemangiomas may be single or multiple (splenic haemangiomatosis), or may be part of generalised angiomatosis when haemangiomas involve multiple organs, especially the liver and skeleton.^(2,5) Cavernous haemangioma is an unencapsulated mass of dilated endothelial-lined vascular channels filled with slow-flowing blood.⁽⁶⁾ Haemangioma of the spleen has a silent clinical picture; the lack of symptoms has been attributed to the very slow growth of the tumour.⁽⁷⁾ However, large lesions causing significant splenomegaly may lead to fullness and left upper quadrant discomfort. Spontaneous rupture with haemorrhage is a risk with larger lesions,⁽⁸⁾ and rupture has been reported to occur in up to 25% of such cases reported.^(8,9) Kasabach-Merritt syndrome is a disease reported in patients with large haemangiomas. These patients develop anaemia, thrombocytopenia, and coagulopathy with multiple haemangiomas throughout the body, particularly in the liver and lung.⁽¹⁰⁾

US may show an inconsistent and nonspecific appearance of echogenicity and sharp margination,

sometimes with cystic regions. Unenhanced CT shows a low attenuation mass; but after injection of a contrast material, there is an increased attenuation of mass, which has been reported to be from periphery to the centre.^(4,11,12) MR imaging is more sensitive and specific than other imaging modalities in the diagnosis of splenic haemangioma. It can be used for imaging splenic lesions in which differential diagnosis is not reached by CT. They are typically hyperintense at T2-weighted MR imaging with a centripetal filling pattern after administration of gadopentetate dimeglumine. Hyperintense signal on T2-weighted images is due to long T2 relaxation times; this quality generally distinguishes it from solid neoplasms.⁽⁸⁾ However, regions of liquefied necrosis in solid tumours may also be hyperintense on T2-weighted images.⁽⁸⁾

The cut surface of a surgical specimen may be homogeneously solid or may demonstrate multiple cystic areas of varying sizes within the large tumour mass.⁽¹¹⁻¹³⁾ Microscopically, it is seen as an encapsulated mass of endothelial-lined vascular channels, filled with slow-flowing blood.⁽⁶⁾ Malignant transformation has been reported,⁽⁸⁾ but these cases may represent primary haemangiosarcoma. Haemangiomas are not treated unless symptomatic, or very large with increased risk of haemorrhage. The usual reported treatment is splenectomy. Recently, there have been reports of benign lesions being treated with partial splenectomy after ligating the segmental supply of the involved half of spleen.⁽¹⁴⁾ The advantage of partial splenectomy is the preservation of its immunological function. Overwhelming postsplenectomy sepsis, although rare, can be a fatal untoward outcome of splenectomy. When the spleen is absent, its immunological functions are generally compensated for, but the phagocytic clearance of bacteria, especially the encapsulated pneumococci and *Haemophilus influenzae*, are reduced, particularly if the host has a deficient concentration of opsonising antibodies.⁽¹⁾

Although unusual, haemangioma is nonetheless the most common primary splenic neoplasm. Haemangiomas

are usually less than 2 cm in size. It is extremely rare to have such a massive solitary lesion. Most splenic haemangiomas are discovered incidentally, and their clinical importance lies in differentiating them from other space-occupying lesions of the spleen (both solid and cystic), particularly from metastases. Preoperative diagnostic investigations are often inconclusive and rarely distinguish between haemangiomas and metastasis. MR imaging is the most reliable imaging method. When possible, partial splenectomy is the treatment of choice for isolated splenic lesions.

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