Giant haemangioma of the liver with haemangio-dudenal fistula: the first reported case in literature

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ABSTRACT

Giant liver haemangiomas are usually asymptomatic with normal liver function, which makes the course long and uneventful. The most commonly reported complications of giant haemangiomas are rupture with intraperitoneal haemorrhage that is either traumatic or non-traumatic, consumption coagulopathy, Budd–Chiari syndrome and congestive heart failure. We describe the first reported complications of a giant liver haemangioma as a fistula between the haemangioma and the gastrointestinal tract.

KEYWORDS

Giant haemangioma – Liver resection – Duodenal fistula

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A haemangioma is a congenital vascular malformation and considered to be the most common benign tumour of the liver with a frequency of 0.4–7.3%.¹ It has a mesenchymal origin composed of masses of blood vessels with atypical or irregular arrangement and size. These tumours most frequently affect female patients (80%) and adults in the fourth and fifth decade of life.²

Liver haemangiomas are usually asymptomatic with normal liver function, which makes the course long and uneventful. As a result of this asymptomatic character, the size of the haemangioma varies greatly when discovered.³ Adam *et al* defined giant haemangiomas as >4cm in diameter.⁴ Giant liver haemangiomas can present with abdominal pain, fullness, mass, and complications such as platelet consumption coagulopathy (Kasabach–Merritt syndrome) and intraperitoneal haemorrhage due to rupture.⁵

There is considerable debate regarding the ideal management of giant liver haemangiomas. Some authors recommend long-term follow-up of these lesions, proposing that most require no treatment.^{6,7} Others recommend surgical intervention based on symptoms of the patient, increase in size, uncertain diagnosis or higher risk of complications.^{8,9} The most commonly reported complications of giant haemangiomas are rupture with intraperitoneal haemorrhage that is either traumatic or non-traumatic, consumption coagulopathy, Budd–Chiari syndrome and congestive heart failure.^{3,10,11}

Case History

A 40-year-old woman presented to our hospital with right hypochondrial pain and fullness as well as severe pallor. Three years previously, she had one session of transcatheter

arterial embolisation in a private clinic for a giant haemangioma involving the left hepatic lobe. She neglected herself with no follow-up.

The patient came to our hospital seeking medical advice for this persistent pain and pallor. On physical examination, she was very pale with no history of haematemesis or melena and the abdomen was markedly distended with an isolated anterior mass, which was mobile and slightly painful.

Ultrasonography showed a huge (about 20cm) hyperechoic hepatic mass in the left lobe with areas of necrosis inside it. Computed tomography (CT) showed a huge left hepatic haemangioma measuring 20cm × 17cm × 20cm, with a lot of areas of necrosis and thrombosis involving segments 2, 3 and 4. It was extruding from the lower surface of the liver with an air filled cavity inside the haemangioma. Another small, well defined haemangioma measuring 2cm × 2.5cm was detected in segment 6 (Fig 1).

Laboratory investigations revealed a haemoglobin level of 5mg/dl and a haematocrit level of 15%. Other liver function tests were normal, as were alpha-fetoprotein levels.

A central venous line and two wide bore cannulas were applied urgently, and four units of packed red blood cells were given. After 48 hours haemoglobin was 9.5mg/dl with 28% haematocrit.

As we were worried about the air filled cavity in the haemangioma, CT was performed again with upper contrast study. Surprisingly, the cavity was filled with contrast, indicating a fistula between the haemangioma and the gastrointestinal tract (Fig 2). Upper endoscopy was excluded owing to the possibility of rupture of the haemangioma caused by air insufflation during the endoscopy.

At laparotomy, a huge liver haemangioma was found involving the left lobe. It was adherent to the first part of

Figure 1 Helical computed tomography of the abdomen with intravenous contrast showing a large hypodense mass in the left hepatic lobe with a peripheral nodular enhancement pattern with an air filled cavity (arrow) in the centre of the mass

the duodenum. A small gastrotomy was performed in the prepyloric region and a finger was introduced through it to the first part of the duodenum. A fistulous track (about 2cm in diameter) was found between the duodenum and the haemangioma (Fig 3), and a finger was introduced into the centre of the haemangioma.

The diagnosis was confirmed as a huge hepatic haemangioma involving the whole left lobe with a fistula connecting

Figure 2 Helical computed tomography of the abdomen with oral contrast showing presence of contrast in the air filled cavity (arrow) inside the haemangioma

Figure 3 Postoperative specimen showing left lobe of the liver with huge liver mass (white arrows) with an opening in the centre of the mass at the site of the fistulous track (blue arrow) between haemangioma and duodenum

the haemangioma with the first part of the duodenum. This was the cause of the occult chronic bleeding and the patient's severe anaemia.

A left hepatectomy was performed with excision of the fistulous track in the first part of the duodenum and repair. Our patient had a smooth postoperative period and received another two units of packed red blood cells to replace the blood lost during the operation.

The patient was discharged on the eighth postoperative day with a haemoglobin level of 11.5mg/dl. She was followed up for nine months and was doing well.

Discussion

Haemangioma is the most common benign liver tumour in autopsy series. Its size varies widely owing to the common asymptomatic course. Giant haemangiomas (>4cm) are now detected more frequently with increased use of imaging modalities.² Although most of these lesions required no intervention during the long follow-up periods, intervention is still needed for symptomatic or complicated giant haemangiomas.⁹

Hepatic haemangiomas are mainly asymptomatic although they can present with right hypochondrial pain either due to distension of Glisson's capsule or focal infarction and necrosis. Giant haemangiomas are more liable to rupture with intraperitoneal haemorrhage, platelet sequestration or consumption with increased bleeding tendency (Kasabach–Merritt syndrome), Budd–Chiari syndrome, biliary colic, obstructive jaundice or gastric outlet obstruction.^{1,12} Although hepatic artery embolisation shows controversial results, it is considered to be the second most widely reported method after surgery for treatment of symptomatic giant haemangiomas.¹¹

No previous reports in the literature have mentioned a fistula between a giant haemangioma and the gastrointestinal

tract. Ours is the first such case described in the literature. We believe that repeated attacks of haemorrhage, thrombosis, necrosis and infarction in the haemangioma may have caused localised perihepatitis, which in turn could have caused the fistula.

In our case, stabilisation and optimisation of the patient's general condition was our first priority to enable her to withstand surgical intervention. The air filled cavity confused us as that indicated continuity of the haemangioma to the gut but CT with upper contrast confirmed this as the air cavity filled with contrast. We would not recommend upper endoscopy owing to the possibility of rupture of the haemangioma caused by air insufflation during the procedure.

Conclusions

There is a possibility of fistula formation between giant haemangiomas and the gastrointestinal tract (in our case, the first part of the duodenum). This should be suspected when there is presence of air inside the haemangioma. CT with upper contrast is a safe and effective modality for diagnosis once suspected. In our case, surgical intervention was considered to be the best management to remove the haemangioma and cut off the fistula.

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