Giant hamartoma of liver mimicking malignancy

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Summary: Two infants with mesenchymal hamartoma of the liver are reported. This lesion clinically simulates a malignant neoplasm very closely and awareness is essential to avoid unnecessary major hepatic resections. The cases were successfully treated with simple enucleation.

Introduction

Benign solid tumours of the liver are rare in infants and children compared to malignant lesions. Mesenchymal hamartoma of the liver^{1,2,3} is a rare benign neoplasm seen usually under one year of age. The tumour manifests itself as a rapidly growing upper abdominal mass closely mimicking a malignant lesion unless the treating clinician is specifically aware of the entity. We present two cases which were correctly diagnosed preoperatively and treated by simple enucleation, unlike most reported cases which were removed by hepatic resection.⁴

Case reports

Case 1

A 9 month old male baby was admitted with progressive abdominal distension of 25 days duration. The infant was anaemic and anicteric. The abdomen was protuberant. The liver was enlarged 10cm below the costal margin and a smooth, spherical, non-tender, ill-defined soft mass was palpable in the lower half of the enlarged liver. The alpha feto-protein (AFP) was negative. An ultrasound study showed a well differentiated lesion in the right lobe of liver composed of mixed echo pattern of solid and cystic areas without any infiltration into the surrounding hepatic

parenchyma. Fine needle aspiration biopsy was negative for malignant cells.

At laparotomy, a 15 × 10 cm sized, soft, greyish white tumour was found on the anterior and inferior aspect of the right lobe of liver. The tumour could be enucleated easily with blunt finger dissection after incising the capsule of tumour. The cavity was drained by a Malecot's catheter without attempting obliteration with sutures. The recovery was uneventful. The child was asymptomatic and thriving well at 18 months follow-up.

The excised specimen weighed 1230 g and was a soft, greyish white jelly-like mass with multiple cystic areas containing watery fluid and gelatinous material without any areas of haemorrhage or necrosis. The histology revealed a few cystic bile ducts, small islands of hepatocytes and large islands of loose mesenchymal tissue surrounded by a haphazard plexus of vascular channels.

Case 2

A 5 month old male was admitted with a rapidly enlarging abdominal lump of 10 days duration (Figure 1). He was not jaundiced. There was a 20 × 15 cm intra-abdominal, smooth, cystic and non-tender mass in the right side of upper abdomen in continuity with the liver. The liver function tests were within normal limits. Ultrasound examination revealed a large cystic mass with multiple septa in relation to the right lobe of liver (Figure 2).

At laparotomy, a well encapsulated greyish white lesion which was jelly-like with cystic spaces was found which could be enucleated easily after incising the capsule. Histologically, the cysts were

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Figure 1 Case 2 showing a large sized abdominal mass.

Figure 2 Case 2. Ultrasonic echogram using B-mode sector scan showing a well demarcated lesion in the right lobe of the liver. Mixed echo pattern with well defined septa and cystic areas may be seen.

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lined by flattened and cuboidal epithelium in myxoid stroma. The postoperative recovery was uneventful and the child was well at 8 months follow-up.

Discussion

Mesenchymal hamartomas are more common in boys under one year of age and may present as an asymptomatic abdominal mass.⁵ Because of the rarity of benign hepatic tumours and rapid growth, these lesions are invariably suspected to be malignant neoplasms preoperatively. The rapid increase has been attributed to the enlargement of cystic components of the tumour.⁶ Although the AFP was normal in Case 1, Ito *et al.*⁷ demonstrated markedly elevated levels in 5 out of 7 cases and suggested that AFP was produced by proliferating hepatocytes in the tumour.

An ultrasonogram shows well encapsulated tumour composed of solid and cystic areas without infiltration into the surrounding hepatic parenchyma or involving the vascular structures such as the inferior vena cava, and is diagnostic.² Morphologically these tumours are solitary, lobulated, cystic and almost invariably located near the lower surface of the right lobe of the liver. They may be pedunculated. Needle aspiration may yield colourless fluid or gelatinous material, especially in lesions having a predominant cystic component. The tumours are composed of a fibrous or myxoid stroma containing cysts and bile ducts.⁵

The treatment of choice is surgical. Total excision may be feasible with pedunculated tumours. In non-pedunculated lesions, if a definite diagnosis had been established at or before operation, simple enucleation is the easiest and most effective treatment and is preferable to major surgical procedures, like hepatic resections, as this tumour is completely benign and shows no recurrence after surgery.^{1,2,8} Here lies the importance of positive awareness of this type of tumour. Special caution should be exercised to avoid any active attempts to obliterate the dead space after enucleation of this tumour, because of the danger of picking up the intrahepatic bile ducts in the ligatures.⁹ The prognosis is good in all treated cases and recurrence is yet to be reported after excision.

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