Mesenchymal hamartoma of the liver – report of an unusual case

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Summary: An 8 year old girl presented with gross hepatomegaly, anaemia and pyrexia unresponsive to antibiotics. At laparotomy a cystic mass 19 cm in diameter was found arising from the liver. The cyst contained 2.4 litres of changed blood and an abscess cavity filled with thick pus was present in the cyst wall. Histological examination showed the appearances of a mesenchymal hamartoma of the liver, a rare diagnosis at this age. This case is unusual as mesenchymal hamartoma has not been previously reported presenting with pyrexia and anaemia as well as hepatomegaly.

Introduction

Mesenchymal hamartomas are benign masses which occur in or are attached to the liver. The term ‘mesenchymal hamartoma’ was introduced by Edmondson (1956) who realized that various cystic lesions of the liver, which had been described under a miscellany of names, were all variants of the same condition. These hamartomas are thought not to be neoplasms but to arise congenitally (Edmondson, 1956). They are rare. Stocker & Ishak (1983) managed to collect only 30 cases from the Armed Forces Institute of Pathology records and reviewed 42 previously published cases. Microscopically, they consist of connective tissue mixed with bile duct elements and hepatocytes.

Cysts are found in nearly all mesenchymal hamartomas. Some are formed by fluid accumulating in areas of degenerate mesenchyme whilst others are dilated bile ducts (Stocker & Ishak, 1983). The cysts usually vary from a few millimetres to several centimetres (Stocker & Ishak, 1983). Only three cases have been reported in which the biggest cyst was larger than 10 cm, one of 12 cm (Symmers & Ward-McQuaid, 1950), one of 14 cm (Stocker & Ishak, 1983) and one of 26 cm (Dooley et al., 1983).

Mesenchymal hamartomas usually present early in life and the median age at diagnosis is 10 months (Stocker & Ishak, 1983). Only four cases are known to have been diagnosed over the age of 5 years (Katzman, 1972; Grases et al., 1979; Stocker & Ishak, 1983; Dooley et al., 1983) and these were at 8, 10, 19 and 21 years respectively.

We present a case of mesenchymal hamartoma of the liver which was diagnosed in a girl of eight. The clinical presentation, simulating a liver abscess, has not been previously recorded.

Case history

An 8 year old girl was admitted to hospital with gross hepatomegaly and pyrexia. She was normally resident in Malta. At the age of 2 years her liver was found to be slightly enlarged (2 fingersbreadth below the right costal margin) and her liver function tests were normal except for a gamma-glutamyl transpeptidase level of 27 IU/l. She was clinically well and her parents were advised that no action was necessary.

She remained well for 6 years until 17 days before admission when she became febrile (96–102°F) with nausea and vomiting. Paracetamol and amoxycillin brought no improvement. On examination at that time she had gross hepatomegaly extending to the right iliac fossa and centrally to the umbilicus. There were no ascites, clinical signs of liver failure or infected skin lesions. She was continued on antibiotics for a further 4 days before being flown to the United Kingdom.

On admission she was clearly ill with a distended abdomen, a haemoglobin level of 6.3 g/dl, a white blood cell count of 17.0 x 10^3/l, and raised gamma glutamyl transpeptidase and alkaline phosphatase levels. Microbiological examination of blood, urine and faeces showed no pathogens. A monospot test was negative and complement fixing antibodies to herpes

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simplex, cytomegalovirus and toxoplasma were not raised. An ultrasound scan showed a very large midline mass of homogeneous reflectivity arising in the liver. After blood transfusion, she underwent laparotomy when a huge cystic mass was found occupying the whole of the right hypochondrium. No masses were present elsewhere; 2.4 litres of thick brown fluid, probably altered blood, were aspirated from the tumour and the collapsed mass was then separated from the right lobe of the liver and the diaphragm. Post-operative recovery was uncom-
plicated and her haematological and biochemical indices returned to normal. Microbiological examination of the fluid from the mass showed pus cells but no bacteria. There was no growth on culture but metron-
idazole and cefuroxime had been given pre-operative-
ly.

Gross pathology

The resected surgical specimen consisted of a cystic mass measuring $19 \times 17 \times 11$ cm. The outer surface was covered by smooth grey serosa except where the mass had been dissected away from the liver. The wall of the mass contained an abscess cavity $15$ cm in maximum diameter filled with thick pus. This was separated from the main cyst by a thin membrane. The inner surface of the large cyst was smooth and light brown. The wall of the large cyst varied in thickness from 0.4 to 2.3 cm and was composed of leathery brown fibrous tissue with myxoid areas and small areas of cystic degeneration (Figure 1). In places the wall had been discoloured by haemorrhage. Where the abscess had formed, the underlying wall was covered by pale yellow necrotic tissue.

Light microscopy

Paraffin sections showed the wall of the mass to be predominantly composed of loose connective tissue, in places mixed with bile ducts and clusters of hepatocytes. The inner surface of the main cyst was lined by dense collagen with no evidence of an epithelial lining. The wall of the mass was richly vascularized with the blood vessels varying in calibre from capillaries to medium-sized arteries and veins. In some areas fluid had collected between the collagen fibres to create a loose network. Occasional mast cells were seen in and around these areas. Inflammatory cells were present scattered throughout the connective tissue and were mainly polymorphonuclear cells with some plasma cells and lymphocytes.

The bile duct component showed two patterns. Large, dilated and intricately branching spaces, lined by cuboidal or flattened epithelium, were seen in a very loose connective tissue with many areas of haemorrhage (Figure 2). These spaces were often partially encircled by incomplete rings of much smaller ducts, again with a cuboidal epithelium, set a small distance away from the large dilated duct (Figure 3). Small clusters of hepatocytes were seen in the wall, usually near the outer surface. No foci of extra-medullary haemopoiesis were found.

Figure 1  The cyst wall was of varying thickness. The left hand part of the cyst shows a ragged ulcerated surface (1) corresponding to the abscess. The membrane (dotted line) which separated this abscess from the main cyst cavity has been removed. Elsewhere the inner lining of the cyst is smooth (2). Myxoid areas and small foci of cystic degeneration are present in the cyst wall (3).

Figure 2  Section of the cyst wall showing a dilated and branching space which, unlike the main cyst, is lined by cuboidal or flattened bile duct epithelium. The surrounding connective tissue contains areas of haemorrhage. H & E $\times 25$. 
Where the abscess had been present, the underlying cyst wall was covered by a thick membrane composed of necrotic tissue and fibrin rich in polymorphonuclear cells. Underneath this necrotic tissue was a zone of hyperaemia with dilated vessels and foci of haemorrhage. There was no vasculitis and no bacteria were seen.

Discussion

The mass reported here contained an unusually large cystic cavity compared with previous cases of mesenchymal hamartoma. The distinctive histological features within the wall of this cavity are identical to those described in this condition (Stocker & Ishak, 1983). The age at presentation and the lack of an epithelial lining exclude biliary cystadenoma (Ishak et al., 1977) or solitary non-parasitic cyst of the liver (Ishak & Sharp, 1979). In addition, our case differs from the cystadenomas with mesenchymal stroma recently described by Wheeler & Edmondson (1985) with regard to the cyst lining and the cellularity of the stromal component. All their cases of cystadenoma with mesenchymal stroma presented in adults (mean age 42 years, range 19 to 67 years) and none had symptoms in childhood, which is very different from the case reported here.

The manner in which this girl presented with fever, nausea and vomiting, anaemia and hepatomegaly was highly unusual. Except for a few cases found incidentally at autopsy, previously reported cases have presented with progressive abdominal enlargement as the only symptom (Stocker & Ishak, 1983). In this case we assume that the pyrexia, nausea and vomiting were related to the abscess cavity present in the wall of the hamartoma. It is not known whether this abscess was due to an infective process. There was no history of infection at another site which could have seeded the hamartoma and organisms were not seen or cultured from the cyst fluid but the patient had received pre-operative antibiotics. Ischaemic necrosis cannot be excluded as the cause of the suppurrative change. The supposition that the pyrexia was related to the abscess is supported by the pyrexia settling quickly post-operatively.

Almost all mesenchymal hamartomas of the liver contain some cysts although these are usually less than a few centimetres in maximum diameter (Stock & Ishak, 1983). It has been suggested (Stock & Ishak, 1983) that the increase in size in mesenchymal hamartomas in infancy is due to enlargement of the cysts. Our case supports this view as mild hepatomegaly was documented at the age of 2 years and an unusually cystic hamartoma was found 6 years later.

In conclusion, our case expands the clinical spectrum of mesenchymal hamartoma of the liver. It emphasizes the need for early diagnosis and resection otherwise severe complications can ensue.

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References


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