

Solitary Hepatic Lymphangioma

A Rare Benign Tumour: A Case Report

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Hepatic lymphangiomas are extremely rare; moreover cystic lymphangiomas usually arise in areas such as the neck and axilla, where loose connective tissue allows the expansion of lymphatic channels. The case of a 65-year old male is described, who presented with a solitary lymphangioma in the liver. The lesion was discovered incidentally and due to diagnostic uncertainty was removed surgically. A short review of histology, clinical presentation and preoperative diagnostic difficulties of hepatic lymphangiomas is given.

KEY WORDS: Liver lymphangioma cystic hygroma

INTRODUCTION

Lymphangiomas are probably congenital malformations of the lymphatic system, composed of dilated endothelial—lined spaces of varying sizes, containing lymph¹. In 95% of the cases, lymphangiomas are located in the neck or the axilla while the remaining 5% are scattered throughout the body². Hepatic lymphangiomas (HL) are very uncommon and after the first description by Ziegler in 1892³, only a few cases have been documented^{4–15}. They are usually observed in children or adolescents¹⁶, and liver involvement, if any, is usually part of diffuse lymphangiomatosis of multiple organs, including spleen, kidneys, bones, gastrointestinal tract, mesentery, mediastinum, lungs, pleura, pericardium and soft tissues^{2–4,15–18}. This report describes the case of a solitary HL in an adult, the presenting symptoms were at first attributed to chronic calculous cholecystitis.

CASE REPORT

A 64-year old male, with a history of heterozygous- β -thalassaemia, presented, complaining of colicky epigastric and right hypochondrial pain. His upper right abdomen was slightly tender, and a soft mass was

palpable in the epigastrium. Total serum bilirubin was 3.2 mg/dl, with the conjugated fraction 2.5 ml/dl. Hepatic biochemistry, α -fetoprotein, carcinoembryonic antigen and anti-echinococcal antibodies, were normal. Hepatic ultrasound (U/S) and computerized-tomography (CT-scan) examination, demonstrated a cystic, thick-walled mass, with a solid component (Figure 1). At laparotomy, a soft, well circumscribed hepatic tumour, was discovered in segment III. The gallbladder contained multiple stones, and no other pathology was detected in the abdomen. The tumour was removed by an atypical hepatic resection. Cholecystectomy was performed and an intraoperative cholangiogram revealed a normal biliary tree. The patient was discharged on the 7th postoperative day, and two years later, he is in good health, with normal liver biochemistry, except for a slightly elevated bilirubin. On macroscopic examination, the tumour was a sharply defined, partially encapsulated solitary cyst, 4 cm in greatest diameter and located in the liver parenchyma. The cyst had a thick (8 mm), sponge-like, gray-white wall and was filled with chylous-like watery fluid, admixed with a small amount of blood. Microscopically, the wall of the cyst consisted of a meshwork of large, empty lymphatic channels, lined with flattened endothelial cells, resting on a loose myxoid connective tissue stroma (Figure 2). The liver parenchyma surrounding

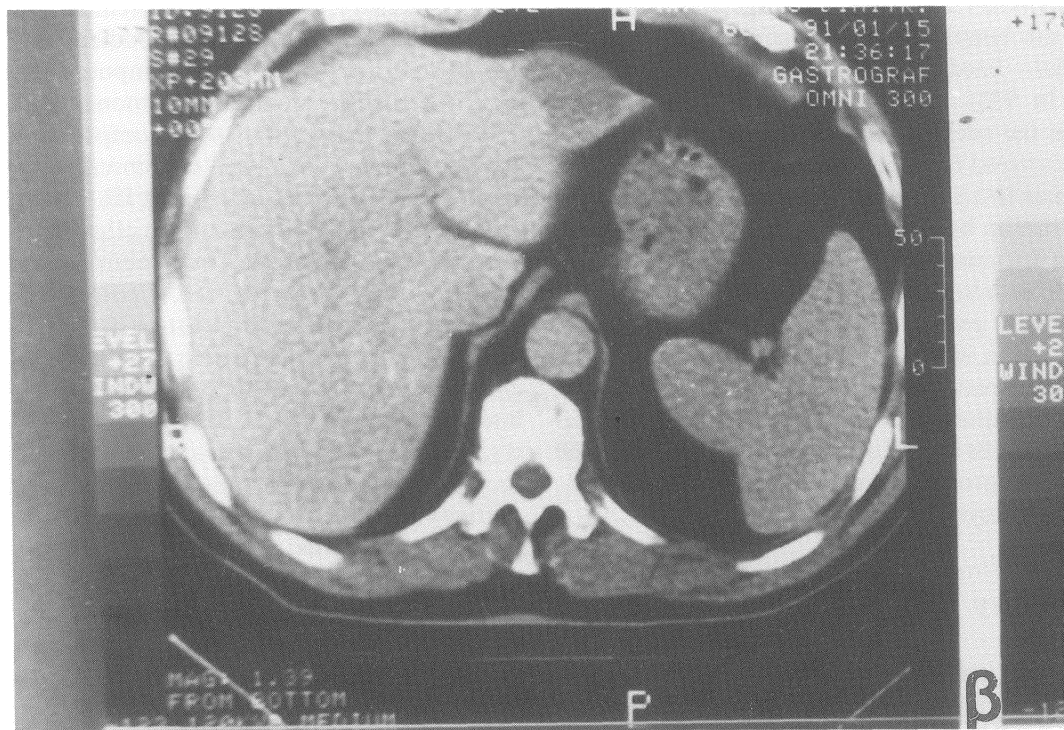
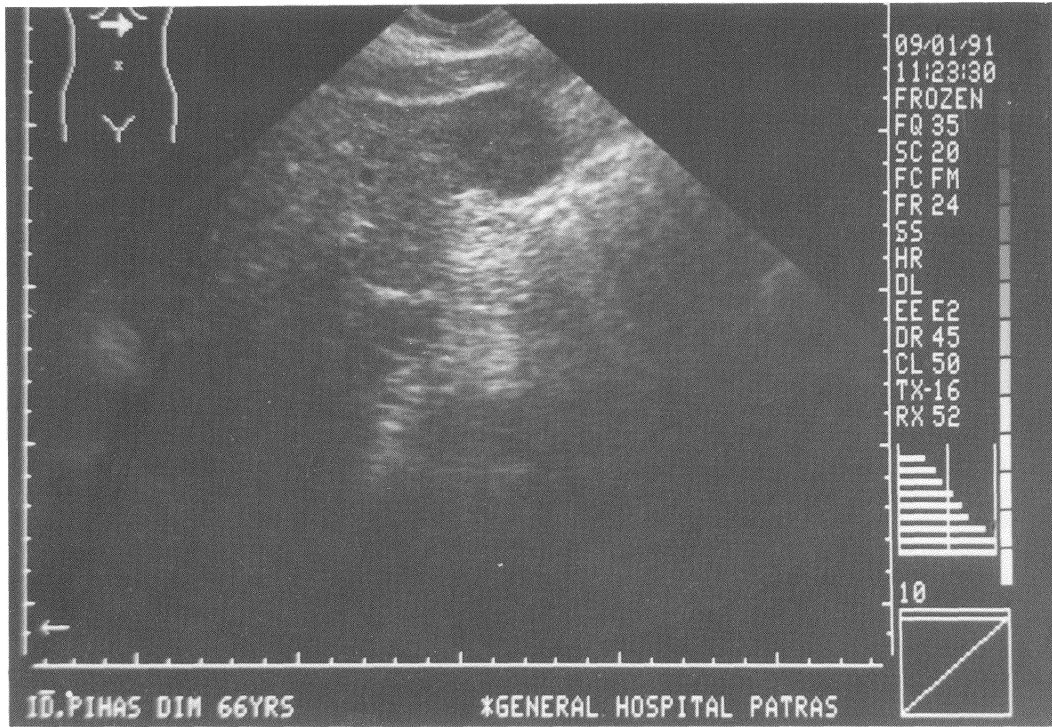


Figure 1 Ultrasound (a) and computerized tomography (b) of the liver. They revealed the lesion in the lateral segment of the left lobe.

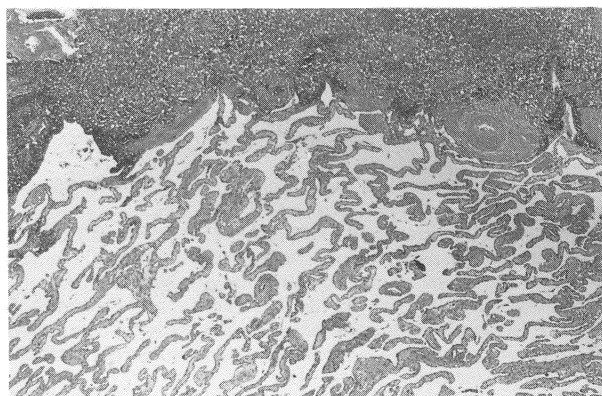


Figure 2 Liver lymphangioma: Freely anastomosing lymphatic spaces (HE \times 100).

the cyst, was compressed and hemorrhagic. The histologic diagnosis was that of a cavernous lymphangioma of the liver.

DISCUSSION

The majority of lymphangiomas seem to represent developmental lesions, occurring relatively early in life. They are generally considered as areas of localized lymphatic stasis, due to congenital blockage of the regional lymphatic drainage^{19,20}. Hepatic lymphangiomas are extremely rare tumours. Either as solitary hepatic tumours or as hepatic localization of diffuse lymphangiomatosis, no more than 42 cases have been reported in the literature⁴⁻¹⁵. Although there is some controversy regarding the hamartomatous, neoplastic, or lymphangiectatic nature of lymphangiomas, all authors agree that they are benign tumours without malignant potential⁴. Histologically they are composed of lymphatic spaces lined by attenuated endothelium and filled with proteinaceous fluid containing lymphocytes. Occasionally erythrocytes may also be present¹. Traditionally lymphangiomas are divided, depending upon the size of the lymphatic spaces, into three groups; capillary or simple, cavernous and cystic²¹. A combination of vascular anomalies, namely lymphangiomas and hemangiomas, can coexist²².

The morphology of the reported case of HL was atypical, with cavernous and cystic elements. Such cystic lymphangiomas are located almost exclusively in the neck where they are called "cystic hygromas"¹⁹. This lesion was either cystic from the beginning, or a typical cavernous lymphangioma that underwent central cystic degeneration.

The clinical presentation of HL is in general atypical^{1,15}, and pain, if any, is due to tension of the liver by the enlarging lesion^{1,15,16}. In our case, it is uncertain whether the patient's complaints were due to the lymphangioma or to coexisting cholecystitis.

The typical U/S, CT-scan and MRI appearance of HL, is that of a cystic or multicystic hepatic mass with internal septations²³, and it is difficult to differentiate it, from necrotic hepatic metastases, or hepatic hydatidosis. It has been suggested that, these investigations are used only for evaluation of the extrahepatic extent of the disease, in cases of diffuse lymphangiomatosis²⁴. In our patient, the preoperative investigations were unable to identify the nature of the hepatic mass. A diagnostic fine needle aspiration biopsy was not attempted, because of the possibility of hydatid disease and the need for an exploratory laparotomy was clear.

In conclusion, the discovery of an hepatic lesion, symptomatic or not, requires identification of the nature of the mass and a decision on further treatment²⁵. However, an accurate preoperative diagnosis is extremely difficult, and no imaging technique or tumour markers, can identify the nature of the hepatic mass that uncommonly happens to be a lymphangioma. Laparoscopy and puncture biopsies, have been suggested as the only way to achieve a reliable diagnosis of HL²⁶. If the diagnosis is obtained, since no malignant transformation of the tumour has been described for the asymptomatic or moderately symptomatic patients, no treatment is necessary. However, when the diagnosis is in doubt, or if the patient develops annoying symptomatology, surgical treatment by the most conservative hepatic resection is required. The prognosis following successful surgical removal is excellent.

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