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Giant Congenital Solitary Cyst of the Liver: Report of a Case

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Abstract Giant solitary nonparasitic cysts of the liver are rarely encountered in children, and establishing a preoperative diagnosis is usually difficult, especially when the cyst occupies the entire abdomen. We report herein the case of an 8-year-old girl found to have a giant congenital solitary cyst of the liver masquerading as an ovarian cyst.

Key words Congenital solitary cyst · Liver · Nonparasitic cyst · Ovarian cyst

Introduction

Congenital solitary nonparasitic cysts arising in the liver occur far less frequently in children than in adults. These cysts usually develop during the third to sixth decade of life, the mean age of patients being 48 years.[1] While improvements in imaging techniques such as ultrasonography and computed tomography (CT) have made diagnosis easier, in the case of very large cysts, even these modalities may be misleading. This report describes the case of a large solitary cyst of the liver masquerading as an ovarian cyst.

Case Report

An 8-year-old girl was referred to our hospital for investigation of progressive abdominal distension over the previous 3 years. She had a history of occasional nonspecific abdominal pain but there were no other constitutional symptoms. General examination was unremarkable except for mild pallor. The abdomen was grossly distended (Fig. 1) with dilated veins over the abdominal wall. A large mass occupying the whole abdomen was palpable, and extended into the pelvis, more on the left side; however, there was no hepatosplenomegaly or ascites. There was significant flaring of the lower ribs and costal margins on both sides.

The results of routine blood investigations and chest X-ray were normal. Ultrasonographic examination and contrast-enhanced computed tomography of the abdomen and pelvis revealed findings suggestive of a large cystic lesion, and the ovaries were not seen separately (Fig. 2). The α-fetoprotein levels were normal. Under the presumptive diagnosis of a giant ovarian cyst, a laparotomy was performed through an infraumbilical transverse incision. At surgery, a massive cyst measuring $30 \times 25 \times 18$ cm was identified. Further exploration was possible only after decompression of the cyst. Both ovaries were found to be normal and no separate vascular pedicle to the cyst was able to be identified. The cyst was found to have arisen from the undersurface of the right lobe of the liver and the gallbladder was stretched over it (Fig. 3). The cyst was dissected away from the liver using Cavitron ultrasonic surgical aspiration (CUSA) and completely excised. Macroscopically, the cyst was thin-walled and unilocular, and filled with serous fluid. There was no evidence of bile staining in the fluid and no bile leaks were seen from the bed of the cyst after excision. A drainage tube was left in situ. The child had an uneventful postoperative course. The drain was removed on the third postoperative day and she was discharged on the seventh postoperative day.

Histopathological examination revealed findings consistent with a congenital unilocular hepatic cyst with no epithelial lining. No bile duct structures or hepatic parenchymal tissue were seen (Fig. 4).

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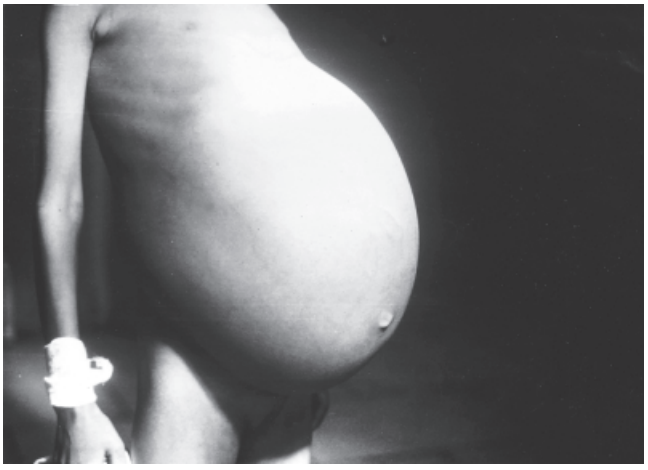

Fig. 1. Clinical photograph of the patient showing the grossly distended abdomen caused by the giant hepatic cyst

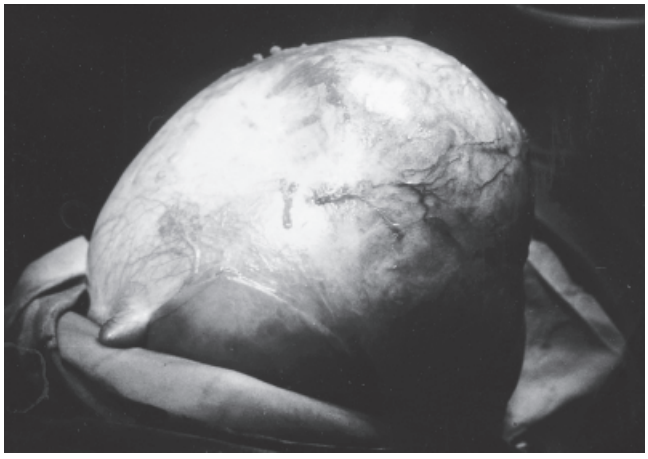

Fig. 3. Intraoperative photograph of the giant hepatic cyst with the right lobe of liver and the gallbladder draped over it

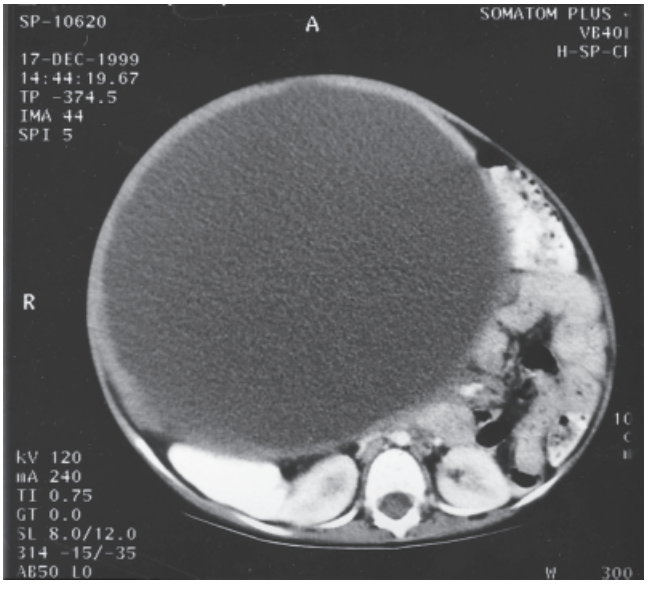

Fig. 2. Computed tomography scan of the patient showing the cyst occupying almost the entire abdomen

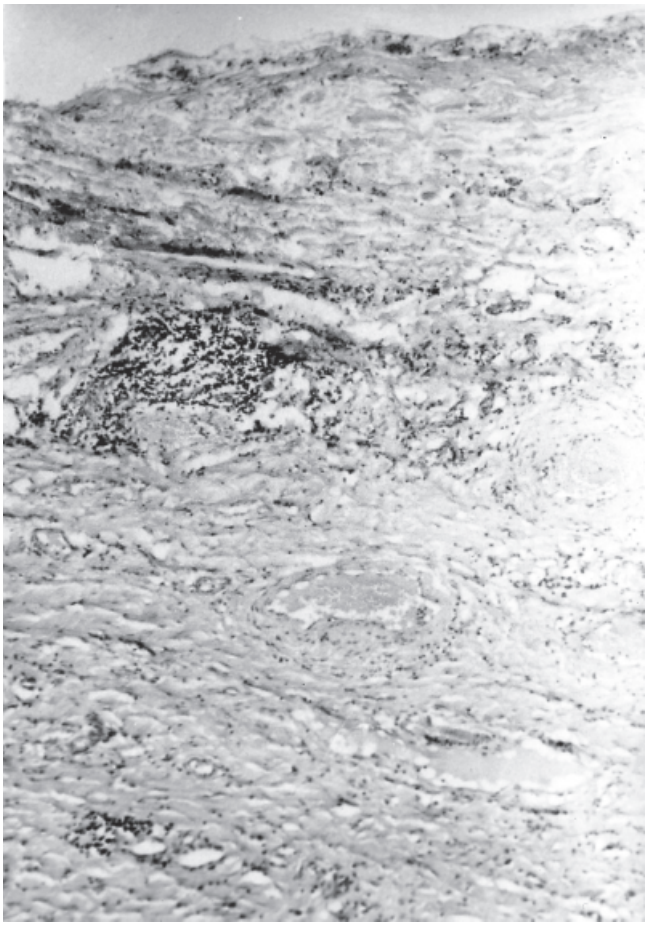

Fig. 4. Section from the cyst wall showing fibrocollagenous tissue with focal lymphocytic infiltration. Lining epithelium was not identified. (H&E, ×10)

Discussion

Congenital solitary nonparasitic cysts of the liver are a well-known clinical entity, with more than 400 cases documented in the world literature.[2] However, the vast majority of cases involved patients in their fifth and sixth decades, with a reported median age of 48 years.[1,3] Conversely, there are few reports of these cysts in children and most pertain to those diagnosed in children younger than 2 years old. Moreover, only four cases of giant nonparasitic liver cysts, greater than 10 cm in diameter, have been reported in children aged 5–10

years old, all of which were documented in the first half of the last century.[4] To our knowledge, there are no similar reports in the recent literature.

The relative rarity and the difficulty in imaging the organ of origin due to the large size of the cyst made establishing a preoperative diagnosis in our patient difficult. Smaller cysts may be diagnosed preoperatively by radiological investigations, but an extremely large cyst can obscure the anatomic site of origin, and a diagnosis may only be made by laparotomy and subsequent histology.

Nonparasitic cysts of the liver are generally benign.[5] While they may be solitary or multiple, large parenchymal cysts are exceedingly rare[6] and can be intrahepatic, partially intrahepatic (as in our patient), or pedunculated.[2] The clinical presentation is usually one of gradual abdominal distension, although a few cases of rapid enlargement have been reported.[2] Although symptoms such as discomfort and pain are occasionally encountered, obstructive jaundice caused by a liver cyst is a very rare complication.[7]

Histologically, these cysts may be classified as cystic mesenchymal hamartoma, solitary nonparasitic cyst, teratoma, or mesenchymoma.[8] The epithelial lining of the internal surface of congenital cysts is usually atrophic and often disappears,[9] as seen in the cyst from our patient.

While small nonparasitic cysts discovered incidentally may not need treatment, large and symptomatic cysts require therapeutic intervention,[10] and various modalities of treatment have been suggested. It is of paramount importance at surgery to aspirate the cyst to determine whether or not it has communication with the intrahepatic bile ducts. If such communication exists, partial excision with internal drainage procedures such as cystojejunostomy can be performed. Other modes of therapy include the injection of minocycline chloride[11] or tetracycline hydrochloride[12] for sclerosis. Enucleation or total excision of the cyst with or without liver resection is the treatment of choice for nonparasitic cysts of the liver,[13,14] although deroofing and partial excision may be indicated if major vascular or biliary ductal structures are present in close proximity to the cyst. The possibility of malignant transformation into adenocarcinoma or squamous cell carcinoma justifies the need for complete excision of the cyst.[15,16]

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