

## Case Report

# Large Congenital Solitary Non-Parasitic Cysts of the Liver: Laparoscopic Removal

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Kuwait Medical Journal 2001, 33 (2): 166-168

### INTRODUCTION

Congenital solitary non-parasitic cysts of the liver (CSNCL) are clinically rare. Seen mostly in adults, they are usually small and asymptomatic. When they become large in size, they may present with abdominal pain or an abdominal mass. This paper describes the incidental finding of a large cystic abdominal mass in an asymptomatic child, which was proven at laparoscopy and later by histopathology to be a CSNCL.

### CASE REPORT

An eight-year-old Saudi male child was referred for an abdominal ultrasound examination (US) as part of a routine investigation for fever of a few days duration. Physical examination did not reveal any abnormal findings and no palpable mass was felt in the abdomen or pelvis. Laboratory tests including complete blood count, erythrocyte sedimentation rate (ESR), liver function tests and urinalysis were all within normal limits. His abdominal US showed a large unilocular purely cystic mass in the right hypochondrium extending from the inferior surface of the liver superiorly, down to the level of the umbilicus. It measured 4 x 7 x 18 cm in size and was apparently separable from all abdominal organs including the liver, gallbladder, pancreas, kidneys and spleen. Contrast-enhanced computed tomography (CECT) of the abdomen demonstrated a homogeneous non-enhancing, water dense mass with no discernible wall and no mural nodules, lying closely adjacent to the wall of the gallbladder, and displacing the bowel loops towards the left side (Fig. 1,2,3).

Differential diagnoses of mesenteric cyst and enteric duplication cyst were considered. A pre-operative Elisa test for hydatid disease was done and proved negative. At laparoscopy, a large transparent cyst was seen protruding from the inferior surface of the right lobe of the liver and was

attached to the gallbladder wall. Aspiration of clear yellowish cyst fluid followed by deroofing of the cyst wall was performed. Histopathologic examination of the cyst wall showed findings typical of a congenital solitary non-parasitic cyst of the liver. The post-operative period was uneventful and the patient discharged in good health on the third post-operative day.

### DISCUSSION

Cystic disease of the liver has been classified according to Lambruschi and Rudolf<sup>[1]</sup> into parasitic and non-parasitic cysts. Parasitic cysts are usually the result of echinococcus infection, while the non-parasitic cysts may be either acquired or congenital. The acquired cysts are more common and can occur in association with neoplastic disease or as a sequel of traumatic injury. The congenital cysts, however, are much less frequently seen and are thought to arise from obstruction of congenitally derived aberrant bile ducts that undergo cystic dilatation<sup>[2]</sup>. They may be multiple, seen in association with the familial polycystic kidney disease, or solitary, which is not associated with renal, pancreatic or splenic cysts.

The congenital solitary hepatic cysts are clinically very rare, with fewer than 300 patients being documented in the literature<sup>[3]</sup>. They are mostly seen in the fourth and fifth decades of life and are rare in childhood. Only 17 CSNCLs presenting in patients less than two years of age are found in the literature<sup>[2,4]</sup>. A survey from the Mayo Clinic on necropsy specimens revealed an incidence of 0.16%<sup>[5]</sup>. However, systematic surveys with advanced non-invasive abdominal imaging techniques have now shown that asymptomatic hepatic cysts are found more commonly in the general population with a prevalence rate of up to 4.7% and a sharp rise in incidence with age<sup>[6]</sup>. The reported female to male ratio ranges from 9 to 4:1<sup>[2,7]</sup>.

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CSNCL may be intrahepatic or pedunculated and are unilocular in 80% of the patients<sup>[4]</sup>. They vary in size, usually ranging between 1 and 15 cm in diameter, with the right lobe being involved twice as often as the left<sup>[2,5]</sup>. The small cysts are incidental findings in two-thirds of the cases, but the large uncomplicated cysts may cause an abdominal mass and a feeling of abdominal discomfort, fullness, or pain. Complications may occur, presenting with the picture of acute abdomen including infection, intra-cystic hemorrhage, rupture of the cyst with consequent peritonitis, and torsion of a pedunculated cyst<sup>[5,6]</sup>. Jaundice is another uncommon complication occurring in about 5% of the patients, usually due to extrinsic pressure of the cyst on the hepatic biliary tree<sup>[8,9]</sup>. Although these cysts are benign, they may undergo subsequent malignant transformation especially when the cyst is lined with squamous epithelium. This occurs exceedingly rarely with only five cases of squamous cell carcinoma arising in a CSNCL reported in the literature<sup>[3]</sup>.



Fig. 1a

**Fig. 1a:** CECT scan of the abdomen showing a large cystic mass protruding from the inferior surface of the liver, closely adjacent to the gallbladder wall.



Fig. 1b

**Fig. 1b:** CECT scan of the abdomen at a lower level. The cyst does not show any discernible enhancing capsule.



**Fig. 2:** Non-enhanced CT scan of the abdomen reveals the same mass extending down in front of the right kidney, indenting and displacing the ascending colon medially.



**Fig. 3:** Post-CECT plain film of the abdomen shows medial displacement of the gastrograffin-filled ascending colon and hepatic flexure by the cyst.

CSNCL are not difficult to diagnose if they are within the substance of the liver. However, when they arise at the periphery or are pedunculated, other differential diagnoses are to be considered. These include choledochal cysts, which are due to congenital dilatation of the common bile duct (CBD) presenting during infancy with cholestatic jaundice. An US in these cases reveals markedly dilated CBD, with or without dilatation of intra-hepatic biliary radicles, thus differentiating it from CSNCL<sup>[2]</sup>.

Other types of non-hepatic developmental cysts that can occur in infancy are duplication cysts of the intestine. These are commonly located in the terminal ileum and ileo-cecal valve, but can extend up to the inferior surface of the liver, mimicking a liver cyst. They are lined by mucosa and muscular layers, however, giving them a characteristic sonographic morphology of an anechoic mass surrounded by echogenic mucosa and a hypoechoic halo, which represents the muscular layer. Debris may rarely be seen inside the cyst due to mucus secretions or hemorrhage. Cysts of lymphatic origin, such as lymphangiomas, are also seen during infancy presenting with painless abdominal masses. They are usually multilocular, have only an endothelial lining and may contain chyle or blood. On CECT scan, septation and attenuation values of fat and water may be demonstrated. Mesenteric, ovarian and exophytic renal cysts are other differential diagnoses not to be mistaken for CSNCL<sup>[2,6]</sup>.

The treatment of choice for CSNCL is now laparoscopic cyst excision or deroofting. This was first introduced in 1991 and is being more widely used recently due to its major advantages over the open surgical techniques. These advantages include reduced morbidity and patient hospitalization. This technique is usually curative with very few recurrences and complications being reported in the literature. Open deroofting, however, is reserved for cysts inaccessible by the laparoscope<sup>[6,10,11]</sup>. It is not known, however, whether or not laparoscopic deroofting and not removing the cyst in its entirety exposes patients to the risk of developing squamous cell carcinoma. In most of the reports available<sup>[6,10,12,13]</sup> the follow-up periods were, generally, very short - in the range of few months to three years- and, usually only the presence or absence of symptoms and the recurrence of the cysts were investigated. Long term follow-up studies are necessary to evaluate for

the possibility of late malignant transformation.

To our knowledge, this is the first case reported in Kuwait of a child with a large CSNCL incidentally discovered and successfully removed by the laparoscopic approach.

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