Dysontogenetic liver cysts and their surgical management

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Dysontogenetische Leberzysten und ihre chirurgische Therapie


Schlüsselwörter: Lebererkrankung, dysontogenetische Zysten, polyzystische Lebererkrankung, Chirurgie.

Summary. Background: Liver cysts occur with a prevalence of 4–7% in the population. We present our experience in the management of these cysts.

Methods: Between 1994 and 2003, 42 patients (33 women, 9 men) were referred for surgery of dysontogenetic liver cysts. There were 19 patients with polycystic liver disease (PLD), 18 with simple cysts, 4 with Caroli disease, and one with a malignant cyst. All patients were symptomatic.

Results: Laparoscopic management was undertaken in 21 patients with simple cysts (n = 13) and PLD Gigot type 1 (n = 8). In the other patients, open procedure was chosen due to the extent of the disease. Caroli disease was monolobar, and segment resection 2/3 and left hemihepatectomy were performed. The patient with malignancy underwent right hemihepatectomy.

Conclusions: Treatment of choice for patients with symptomatic simple cysts and PLD Gigot type 1 is laparoscopic deroofing and fenestration. In advanced-stage PLD, we recommend combined hepatic resection and cyst fenestration techniques, often as a bridging to transplantation. In monolobar and symptomatic Caroli disease, surgical resection represents the best treatment.

Key words: liver disease, dysontogenetic cysts, polycystic liver disease, surgery.

Introduction

Dysontogenetic liver cysts are reported with a prevalence of 4–7% in the population [1]. These cysts are usually small, but also large cysts can remain asymptomatic. Congenital cysts can be solitary, multiple, or multiple and diffuse in the liver parenchyma (polycystic liver disease, PLD).

Caroli disease is an uncommon congenital disorder characterized by segmental dilatation of the bile ducts, causing chronic cholestasis and hepatolithiasis. It is considered to be an autosomal recessive inherited condition.

Liver cysts are usually found incidentally on radiologic imaging like ultrasonography or computed tomography (CT).

Hepatic cysts are normally asymptomatic. Symptomatic patients tend to have large cysts. Pain may be related to pressure on related structures and lead to nausea, vomiting, obstructive jaundice, or early satiety [2].

Clinical presentation can be complicated by hemorrhage, rupture, torsion, or infection [3–5]. Treatment is limited to symptomatic patients and patients with malignancy. Various possibilities for treatment are proposed, including nonsurgical procedures such as percutaneous aspiration, with or without alcohol injection [5–7], and
surgical procedures, including open or laparoscopic fenestration and deroofing, and liver resection [2–21].

The first technique of intraperitoneal fenestration and deroofing was described by Lin et al. in 1968 [18]. Nowadays this technique is considered by many to be the treatment of choice for simple cysts and PLD, performed as a single procedure or in combination with liver resection.

We present our experience in the management of patients with dysontogenetic liver cysts.

Material and methods

Between 1994 and 2003, 42 patients (33 women, 9 men) were referred to our center for the management of cystic liver disease. Their median age was 55 years (range, 30–79 years). There were 19 patients with PLD, 18 with simple giant cysts of the liver, one with a malignant cyst, and four with Caroli disease.

All patients underwent preoperative ultrasonography and CT.

The median size of solitary simple cysts was 15.5 cm (range, 10–20 cm); the median diameter in patients with PLD was 11 cm (range, 5–22 cm).

All patients were symptomatic; presenting complaints were abdominal pain or symptoms related to compression of adjacent organs including nausea, vomiting, or early satiety.

Four patients with simple cysts showed a mild elevation of the liver enzymes glutamic oxalacetate transaminase, glutamic pyruvic transaminase, and gamma glutamyl transpeptidase; these liver enzymes and total bilirubin level were elevated in five patients with PLD. Creatinine was elevated in two patients with PLD and additional polycystic kidney disease (PKD). In all patients, PLD was associated with PKD.

Four patients underwent surgery for Caroli disease. Their median age was 47 years (range, 40–54). The disease was monolobar in all patients. The presenting symptoms were abdominal pain and recurrent episodes of choledithiasis. All patients had evidence of cystic dilatation of the intrahepatic bile ducts with calculi, both seen on ERCP.

Finally, a 60-year-old woman underwent surgery for treatment of a neoplastic cyst. She presented with abdominal pain and distention. The cyst was solitary with a diameter of 20 cm and with cyst wall nodules and solid components suspected of being malignant, as assessed by CT. Tumor marker CA 19-9 was not elevated.

Results

Laparoscopic management was undertaken in 21 patients with solitary giant cysts (n = 13) and PLD Gigot type 1 (n = 8). In the other patients, open procedure was chosen due to the extent of the disease. In five patients with simple cysts, open procedure was conducted. In two cases, this was due to adhesions resulting from preceding laparotomies; in three other patients, due to suspicion of neoplastic disease.

Laparoscopic procedure was successfully completed in 20 patients. In one patient with PLD, bleeding from a superficial hepatic vein necessitated conversion to open procedure. The bleeding was due to coagulation injury. Two postoperative complications occurred: one patient developed pneumothorax following placement of a cava catheter and was treated by thorax drainage; another patient had biliary leakage, which was treated conservatively. In two patients, laparoscopic cholecystectomy was done simultaneously. The mean operative time was 100 min (range, 70–120 min). The mean duration of hospitalization was 10 days (range, 8–14 days). There was no postoperative mortality. The median follow-up time was 27 months (range, 7–52 months). There were no symptomatic recurrences.

Open procedure was performed in 11 patients with PLD (Fig. 1): one left hemihepatectomy, deroofing in 2 patients, segment resection 2/3 plus deroofing in 6 patients, segment resection 5/6 plus deroofing in 2 patients.

There were four incidents of complications: a case of biliary leakage was managed conservatively; two patients developed pneumothorax following insertion of a cava catheter, and in one patient the abdominal drain tore off and had to be removed by re-laparotomy on the fourth postoperative day.

The mean operative time was 145 min (range, 120–180 min). The mean duration of hospitalization was 15 days (range, 12–42 days). There was no postoperative mortality. The median follow-up time was 60 months (range, 24–98 months). One symptomatic recurrence occurred 84 months following the first operation. The female patient had massive hepatomegaly with compression of the inferior vena cava. Upon surgery it was not possible to decompress the inferior vena cava, because the cysts had massively encroached upon it. Liver transplantation was attempted two weeks later, but the patient developed a hepatopulmonary syndrome with right heart failure and died during transplantation.

Histological examination of all specimens, simple cysts and PLD, revealed cuboidal and flat monolayer epithelia without dysplasia in the walls of the cysts.

All patients with PLD underwent CT scan one year after surgery.

Fig. 1. PLD specimen after left hepatic resection via open approach