

Polycystic Liver Disease with Complications: Fenestration by Laparoscopic Approach

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Rezumat

Fenestrarea prin abord laparoscopic în boala polichistică hepatică complicată

Scop: Boala polichistică hepatică izolată este o formă rară de afecțiune ereditară chistică a ficatului, cu transmitere autosomal dominantă. Se definește prin existența a numeroase chisturi de diferite mărimi în parenchimul hepatic, având o evoluție de obicei benignă.

Metodă: Prezentăm cazul unui bolnav de 80 de ani cu boală polichistică hepatică masivă, diagnosticată în urmă cu trei ani, prin ultrasonografie și computer tomografie abdominală. Odată cu apariția complicațiilor date de fenomene de tip compresiv datorită dimensiunilor mari ale chisturilor, bolnavul a prezentat dureri abdominale, grețuri, vărsături și scădere ponderală. Fenestrarea chisturilor prin abord laparoscopic a condus la rezoluția completă a simptomatologiei.

Rezultate: Bolnavul a fost mobilizat în ziua operației, și a fost externat în ziua 9 postoperator după suprimarea tubului de dren.

Concluzii: Boala polichistică hepatică izolată este o afecțiune rară. Tratatamentul chirurgical este indicat numai în cazul

complicațiilor. Abordul laparoscopic este o alternativă terapeutică în caz de nevoie. Bolnavii beneficiază de avantajele chirurgiei miniminvasive.

Cuvinte cheie: chisturi hepatice, abord laparoscopic

Abstract

Aim: Isolated polycystic liver disease is a rare congenital cystic liver disease with autosomal dominant transmission. Its main feature is the presence of a large number of cysts of different sizes in the hepatic parenchyma, which have a benign evolution.

Method: We present the case of an 80 years old male patient with massive polycystic liver disease, diagnosed three years ago by ultrasound examination and abdominal computed tomography scan. The evolution of the disease had been complicated by compressive symptoms, caused by the large dimensions of the cysts. The patient presented with abdominal pain, nausea, vomiting and lost weight. Cyst fenestration through laparoscopic approach resolved the symptoms.

Results: The patient was mobilized on the day of the surgery, and was discharged on the 9th postoperative day, after drainage tube removal.

Conclusions: Isolated polycystic liver disease is rare. Surgical treatment is indicated only if complications occur. The laparoscopic approach is an alternate treatment method, if needed.

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The patients benefit from the advantages of minimally invasive surgery.

Abbreviations: CT-computed tomography, Hct-Hematocrit, Hgb-Hemoglobind, PLT-platelets, IPLD- Isolated polycystic liver disease

Key words: liver cysts, laparoscopic approach

Introduction

Polycystic liver disease is a congenital disorder, defined by the presence of a large number of cysts of different sizes dispersed in the hepatic parenchyma. The cysts originate from the malformation of the embryonic ductal wall lined by functional biliary epithelium (1,2). These cysts grow in size and detach from the intrahepatic bile duct, becoming autonomous structures that lose direct connection to the bile duct. The evolution of the isolated polycystic liver disease is benign; complications can occur exceptionally. The clinical symptoms of polycystic liver disease are most often hardly noticeable or absent, and a complication like intracystic bleeding may trigger the exacerbation of the symptoms (1,3). Liver function of most patients is normal, and positive diagnosis is based mainly on imaging. Treatment options are based on the symptoms and pathology of the cysts.

Case report

We present the case of an 80 years old male patient with massive polycystic liver disease, with compressive symptoms and intracystic bleeding. Three years ago the patient was diagnosed with tumor of the sigmoid colon, as he had rectorrhagia.

Ultrasound examination and CT scan performed before surgery for detection of possible hepatic metastases of the sigmoid colon tumor found multiple hepatic cysts that were asymptomatic.

The surgical treatment of the sigmoid tumor (segmental resection of the sigmoid colon) was performed at another surgery department, and the patient did not receive any oncologic treatment.

Three years after surgery the patient suffered from abdominal pain in the right hypochondrium, nausea, vomiting, and also lost weight. Abdominal ultrasound examination and CT scan confirmed the significant growth of the liver cysts.

Laboratory results were normal – Hematocrit (Htc): 41.9%; WBC: 8700; Platelets (Plt): 231000; Serum glucose: 1,25 g/l; Urea: 5,53 g/l; Creatinine: 0.09 g/l.

Hepatic function was almost normal, with moderate increase of aminotransferase levels: SGPT: 64 U/L; SGOT: 47 U/L.

Considering the age of the patient, and associated conditions we performed exploratory laparoscopy under

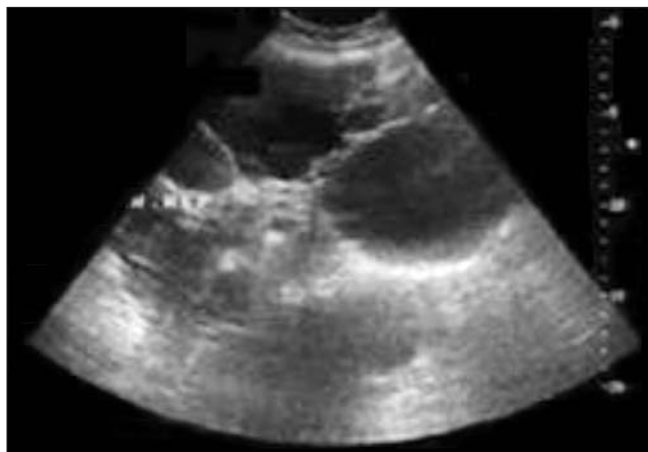


Figure 1. The ultrasound examination in 2007

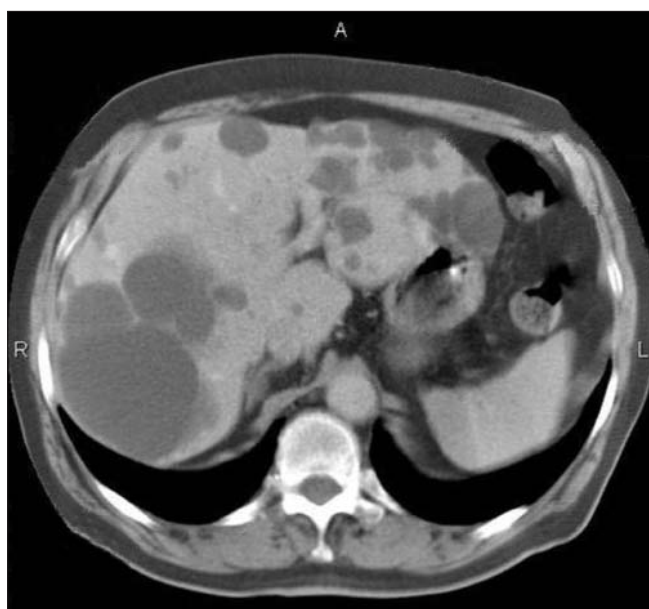


Figure 2. The CT scan examination in 2007

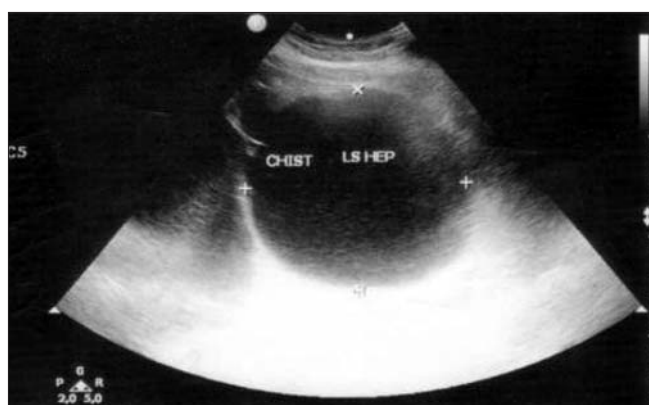


Figure 3. The ultrasound examination in 2010

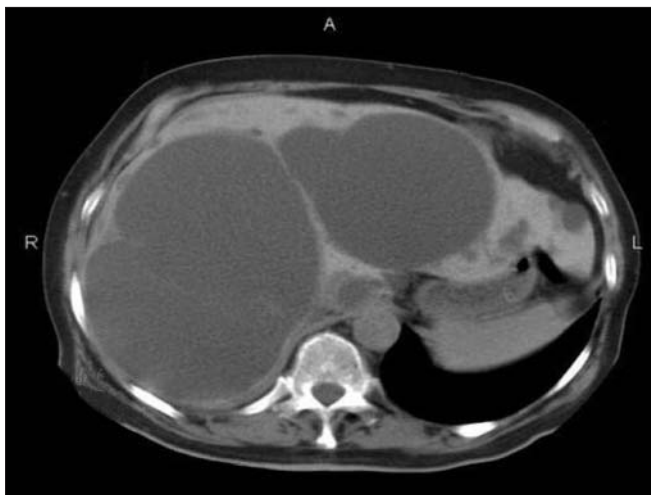


Figure 4. The CT scan examination in 2010

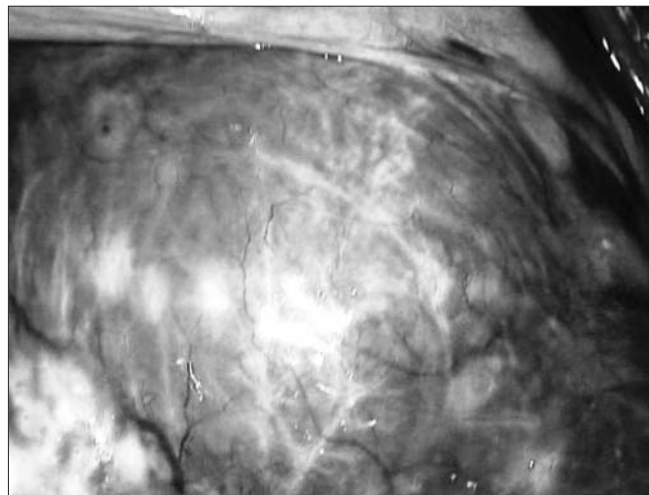


Figure 5. Intraoperative aspect of the cysts

general anesthesia. We found an enlarged liver, with multiple cysts in both lobes, both on the diaphragmatic and the visceral surface of the organ. Some of the cysts were larger than 10 cm. We performed a puncture and flattening of the large cysts leaving a subhepatic drain in place. The content of the cysts was hemorrhagic, and a liver biopsy was sent to histopathology examination.

Surgery: no. 146/29/03/2010 – Flattening of the cysts, liver biopsy, draining.

Histopathology result of the biopsy (cyst wall): on the examined hepatic fragments there are multiple small cysts, lined by simple cubic epithelium. (M. Turcu MD)

The postoperative evolution has been favorable; on the first day approximately 800 ml of ascites-like discharge was eliminated through the subhepatic drain tube, and the volume decreased gradually. On the 8th postoperative day the drain tube was removed. Cyst fenestration using laparoscopic approach led to the resolution of the symptoms..

Results: the patient was mobilized on the day of the surgery, and was discharged on the 9th postoperative day, after drain tube removal.

Discussion

Polycystic liver disease is more frequently associated with polycystic kidney disease, but may as well occur as an isolated and distinct genetic disorder. (1,3) Isolated polycystic liver disease (IPLD) has an autosomal dominant transmission by a genetic mutation different from that of the patients with polycystic kidney disease (1). Recently two genes responsible for IPLD have been discovered: the PRKCSH gene, located on the 19p chromosome, and the SPC 63 gene, located on the 6q chromosome (1). These genes encode glycoproteins synthesized in the endoplasmic reticulum, and their mutation results in faulty processing of a key regulatory substance of biliary cell growth (1). Finally, all these mutations lead to formation of biliary cysts in the liver. IPLD is much less frequent than poly-

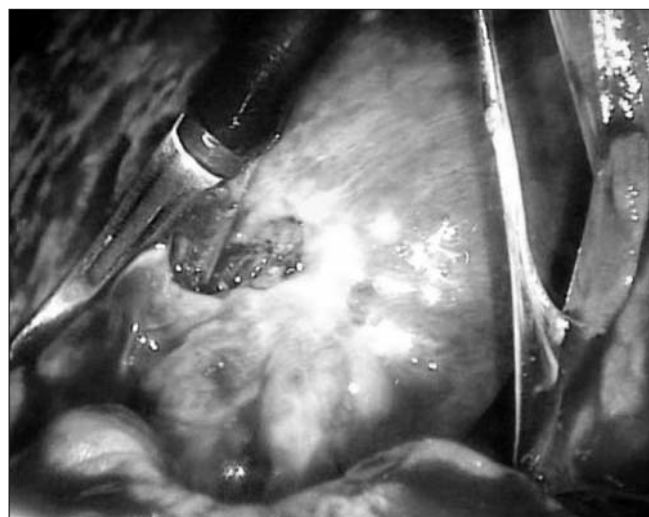


Figure 6. Cyst fenestration

cystic kidney disease, with prevalence less than 0.01 (1,3,4). In most cases polycystic liver disease is asymptomatic, but may display progress in time, and in rare cases may impact hepatic function or it may become symptomatic through the apparition of complications. Complications are very rare: intracystic bleeding, infection, posttraumatic rupture (3,5).

Symptomatic patients have massive hepatomegaly. Depending on the size and distribution of the cysts, critical vascular or ductal structures may be compressed. Portal vein compression may lead to portal hypertension, as well as associated conditions like splenomegaly and ascites (1).

The number and size of the cysts increase with age. The hereditary polycystic liver usually contains more than 20 cysts, which generally replace over 50% of the liver parenchyma. This represents an important differentiation criterion from multicystic (non-hereditary) liver (1,5). The size and distribution of the cysts are variable. There are large cysts, located primarily at the surface of the liver, accessible to

surgical fenestration, and also multiple small cysts distributed in the entire parenchyma, like posterior cysts that are less accessible to fenestration (1). Imaging studies describe two types of cysts in patients with polycystic liver disease: intrahepatic cysts and peribiliary cysts (1). Intrahepatic cysts are less frequent, located largely peripherally and have large dimensions (10 mm to 80 mm). Peribiliary cysts are usually smaller than 10 mm in diameter (1,6).

Positive diagnosis is based mainly on imaging studies: ultrasound examination, magnetic resonance imaging and CT scan.

The ultrasound appearance of polycystic liver is relatively typical, displaying multiple transonic, round or oval shaped images, with variable dimensions (from 1 cm to 10 cm). The transonic aspect of the cysts disappears in case of intracystic bleeding or in case of superinfection of the cyst (3). In these cases the content of some cysts will appear hypoechogenic. The liver parenchyma between the transonic hepatic cysts appears normal. CT and MRI are more sensitive methods (detecting cysts under 1 cm diameter), and are used especially in the diagnosis of complications. Biochemistry parameters are rarely useful, as typically the liver parenchyma is typically preserved. GGT may increase moderately, while the hepatic function is normal.

The evolution of isolated polycystic liver disease is benign, and complications are exceptional.

Polycystic liver disease does not require therapy. Treatment is indicated in case of complications.

The cysts can be evacuated and a sclerotic substance can be injected into their cavity using ultrasound or CT guided percutaneous puncture of the cyst. This is also called delaying therapy (1,7). This therapy is particularly useful in cases with rapid expansion of dominant cysts, infected and hemorrhagic cysts.

For more severe, symptomatic cases the therapeutic options include surgical procedures (open or laparoscopic) like cyst fenestration, fenestration combined with partial hepatic resections or hepatic transplant (4,8,9).

Cyst fenestration is a common treatment of massive polycystic liver disease.

The approach may be through laparotomy or laparoscopy (5,6,10,11). In the majority of cases this leads to resolution of the symptoms. Advantages of laparoscopy include: low postoperative morbidity, shortened hospital stay (5,6,7,11). Cyst fenestration may represent a temporary relief of symptomatic disease, but these cysts will relapse in 22%-75% of the patients (1).

Partial liver resection has high perioperative morbidity and it is indicated in cases resistant to decompression (1,2).

Generally, fenestration is the only and most acceptable treatment for polycystic liver disease patients, where the size of the liver may decrease after the collapse of the cysts (3,10). Although reported morbidity and mortality rates for open and laparoscopic fenestration are similar, symptomatic recurrence is slightly higher in the case of laparoscopic technique (1). Massive perioperative fluid loss during removal of large cysts may lead to severe dehydration (6). Complications of cyst

fenestration include ascites in all patients (6,9). Generally, this diminishes after the third postoperative day, being eliminated through the drain tube, which is maintained indwelling 5-26 days after surgery, in case diuretics are used (6). The persistence of ascites following fenestration is due to the fact that the peritoneum is unable to absorb more than 900 ml of liquid per day (6).

Cyst fenestration combined with partial hepatic resection (2) is a viable option for patients with uneven distribution of cysts, but with certain areas of normal hepatic parenchyma. Due to the distorted anatomy of these patients, segmental liver resection is a technical challenge (2,9). The resection – fenestration combination is appropriate for patients with heterogeneous cyst patterns; during this combined procedure the segments with large numbers of diffusely distributed small cysts are resected in combination with fenestration of larger cysts in other areas of the liver. This will enable the reduction of the size of the liver (2).

Liver transplant is performed in the case of patients whose symptoms cannot be improved by other interventions (8,9). Liver transplant is the last line of treatment for patients with refractory symptoms. Usually, liver transplant results in excellent improvement of symptoms in these patients, but nevertheless it is a high risk procedure, associated with mortality rates between 10% and 33% (8,9).

The compromise between less invasive procedures, providing immediate, but short term relief, and a variety of invasive therapies offering better long term results, which are however technically more difficult due to the significantly distorted anatomy of these patients must be considered in each case individually. High risk of complications associated with aggressive surgical therapy must be considered when establishing the appropriate treatment for a patient (1).

Conclusions

Polycystic liver disease is a rare disease. Surgery is indicated only in cases with massive liver cysts with compression, or in case of intracystic bleeding or infection. Cysts size and distribution patterns determine the therapeutic options available for individual patients. The laparoscopic approach is an alternate treatment method, if needed. The patients benefit from the advantages of minimally invasive surgery.

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