Surgical management of non-parasitic hepatic cysts
A single center experience and a review of the literature


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Surgical management of non-parasitic hepatic cysts. A single center experience and a review of the literature.

AIM: We present our experience in the laparoscopic management of the hepatic cysts (SHCs) and the polycystic liver disease (PCLD), and a literature review.

MATERIAL AND METHODS: Between 2005 and 2018, laparoscopic deroofing was performed in 28 consecutive patients. There were 19 cases with SHCs and only 9 cases with PCLD (Gigot's type I). CT scan was performed in all cases to assess the characteristics, dimensions, and exact position of the lesion. Surgery was planned for all patients because of evident and persistent symptomatology.

RESULTS: We have analyzed operative time, surgical procedure, blood loss, hospital stay, complications, and medium follow-up period. All the patients underwent laparoscopic deroofing of the larger cysts and puncturing of the smaller cysts. The total morbidity recorded was 25% (7/28), 3 cases in the group of SHCs (16%) and 4 cases in the PCLD one (44%) and was characterized of 3 cases of ascites through trocar insertion sites after removal of drainage tube and 4 case of pleural effusion.

DISCUSSION: There were no significant group differences in term of length of hospital stay. The follow-up period (a mean of 24 months) confirmed that all the patients remained free of symptoms and relapse of the disease.

Conclusion: The technical feasibility and the good short- and medium-term results made the laparoscopic approach the procedure of choice for the management of symptomatic liver cysts.

KEY WORDS: Hepatic cyst, Liver disease, Minimally invasive surgery

Introduction

Non-parasitic cysts are the most common pathology found in the liver having a prevalence ranging from 2.5 to 5% in the overall population. In the past 20 years, the extensive use of first and second level imaging studies, such as ultrasound (US) and computed tomography (CT) abdomen, has led to an increase up to 18% in the prevalence of these lesions.

Hepatic cysts represent a heterogeneous cluster with regard to pathogenesis, clinical presentation, diagnostic findings and therapeutic management. They are classified as congenital or acquired, based on the pathogenesis. Congenital forms include the simple hepatic cysts (SHCs) and the polycystic liver disease (PCLD). The acquired lesions are represented by hematomas, parasitic cysts, neoplastic and metastatic cysts. These lesions are also classically divided with a different criterion into parasitic hepatic cysts (PHCs) and non-parasitic hepatic cysts (NPHCs), with the latter being the most prevalent worldwide (prevalence of 4-7% in the general population).

Non-parasitic hepatic cysts can be treated surgically when symptomatic, when there is the doubt of a neoplastic origin or when complicated. Various treatment procedures have been proposed, such as a simple US or CT-guided aspiration, injection of...
sclerosing agents, fenestration, enucleation, hepatic resection and transplantation. The recent development of laparoscopic surgery and the short-term benefits associated with it (reduced hospitalization, better cosmetic results and quicker return to normal activities) has produced an increase in resection or fenestration in recent years. We describe the most current management of non-infectious hepatic cysts, thereby discussing differential diagnosis, treatment options and outcomes.

In this study we present our experience in the laparoscopic management of the SCHs and PCLD.

Materials and Methods

Between 2005 and 2018, a total of 28 consecutive patients were referred for evaluation and treatment of simple liver cysts (solitary and multiple cysts) or PCLD to the Department of Medical and surgical Science, University of Foggia. The study group consisted of 11 males and 17 females with a mean age 50 years (range 35-60 years). There were 19 cases with SHCs and only 9 cases with PCLD (Gigot’s type I)³. These patients had not a family history of PCLD and Autosomal Dominant Polycystic Kidney Disease (ADPKD). Demographic data, size of the dominant cyst, cysts sites and ASA score were reported in Table I.

Table I - Demographic data, size of the dominant cyst, cysts sites and ASA score of all patients

<table>
<thead>
<tr>
<th>Data</th>
<th>SHCs (19)</th>
<th>PCLD (9)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (M/F)</td>
<td>7/12</td>
<td>4/5</td>
</tr>
<tr>
<td>Median age (years)</td>
<td>50 (35-60)</td>
<td>46 (43-59)</td>
</tr>
<tr>
<td>Median size, cm (range)</td>
<td>8 (5-17)</td>
<td>11 (10-13)</td>
</tr>
<tr>
<td>Bilateral location, n (%)</td>
<td>5 (26%)</td>
<td>9</td>
</tr>
<tr>
<td>Right hemiliver, n (%)</td>
<td>7 (37%)</td>
<td>/</td>
</tr>
<tr>
<td>Left hemiliver, n (%)</td>
<td>7 (37%)</td>
<td>/</td>
</tr>
<tr>
<td>ASA I, n (%)</td>
<td>16 (84%)</td>
<td>4 (44%)</td>
</tr>
<tr>
<td>ASA II, n (%)</td>
<td>3 (16%)</td>
<td>5 (56%)</td>
</tr>
</tbody>
</table>

Table II - Clinical findings in all patients

<table>
<thead>
<tr>
<th>Indications</th>
<th>Simple Cysts (19)</th>
<th>PCLD (9)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epigastric pain</td>
<td>16 (87%)</td>
<td>5 (60%)</td>
</tr>
<tr>
<td>Abdominal swelling</td>
<td>15 (80%)</td>
<td>5 (60%)</td>
</tr>
<tr>
<td>Early satiety</td>
<td>7 (40%)</td>
<td>5 (60%)</td>
</tr>
<tr>
<td>Heartburn</td>
<td>2 (13%)</td>
<td>2 (20%)</td>
</tr>
<tr>
<td>Nausea</td>
<td>7 (40%)</td>
<td>4 (40%)</td>
</tr>
<tr>
<td>Vomiting</td>
<td>4 (20%)</td>
<td>4 (40%)</td>
</tr>
<tr>
<td>Supine dyspnea</td>
<td>0</td>
<td>2 (20%)</td>
</tr>
<tr>
<td>Palpable mass</td>
<td>2 (13%)</td>
<td>4 (40%)</td>
</tr>
</tbody>
</table>

Epigastric pain and abdominal swelling, early satiety, heartburn, nausea, vomiting, supine dyspnea and palpable mass were the most common symptoms (Table II). The symptoms were on average 6 months (range, 3-24 months).

Ultrasonography (US), computed tomography (CT) and laboratory tests, were used to make diagnosis ad to rule out parasitic and neoplastic liver cysts. Serum antibodies for Echinococcus were negative such as the tumor markers (α-fetoprotein, carcinoembryonal antigen and carbohydrate antigen 19-9). The initial imaging modality was the US in all cases, followed by CT scan. Simple liver cysts typically presented on US as monolocular, anechoic and sharply demarcated lesions with thin wall and posterior acoustic enhancement. CT scan was performed in all cases to assess the characteristics, dimensions, and exact position of the lesion. On CT scan, SHCs and PCLD appeared as well-demarcated lesion with uniform fluid-density without enhancement after contrast administration.

None of the cysts was complicated, infected or ruptured. Morphological definition and topography of the cystic lesions was also determined by magnetic resonance imaging (MRI). Elective laparoscopic surgery was planned for all patients because of the presence of evident and persistent symptomatology. All the patients underwent laparoscopic deroofing of the larger cysts and puncturing of the smaller cysts. The roof of the cyst is largely excised with monopolar hook and sent for histological examination. Then, the residual cavity was widely in communication with the peritoneum. The evaluation of the cystic fluid, typically clear and serous, is important in order to define the benign nature of the cyst. It is important to examine the cavity of the cyst carefully to preclude both the contingent tissue proliferation and the connection to the biliary ducts. In case the cyst was loculated, all the septa were broken and laid open. In case of deeply cysts their fenestration occurred in a second time and through the superficial ones. In PCLD (Gigot’s type I), this procedure has to be repeated for each cyst and so can become tedious and time consuming, but it is crucial that all cysts are deroofed, because the success of the operation depends on this meticulous treatment ⁵,⁶. Peritoneal drainage was placed in all patients.

Table III - Short-term results

<table>
<thead>
<tr>
<th>Parameter</th>
<th>SHCs (19)</th>
<th>PCLD (9)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hospital stay (days)</td>
<td>5 (range, 3-7)</td>
<td>6 (range, 4-10)</td>
</tr>
<tr>
<td>Mean operative time (min)</td>
<td>55 (range, 40-80)</td>
<td>100 (range, 90-130)</td>
</tr>
<tr>
<td>Ascites n (%)</td>
<td>0</td>
<td>3 (33%)</td>
</tr>
<tr>
<td>Pleural effusion n (%)</td>
<td>3 (16%)</td>
<td>1 (11%)</td>
</tr>
</tbody>
</table>
Results

There was no intraoperative or postoperative mortality in this series. The mean operative time was 55 minutes in simple liver cysts and 100 minutes in PCLD, because of the necessity of treating numerous cysts. There were no conversions. Liquids were started on the same evening as surgery, and a normal diet was started on the first postoperative day (POD). The abdominal drainage was present for 1 days to 4 days. At discharge, the complete resolution of symptomatology was reached in all cases. The histological examinations revealed the typical pattern of the simple liver cysts in any case, without evidence of cystadenoma or carcinoma. Intraoperative complications were not detected in our series. Postoperative morbidity consisted of 3 cases of ascites through trocar insertion sites after removal of drainage and 3 case of pleural effusion. All cases treated conservatively with medical therapy.

The total morbidity recorded was 25% (7/28), 3 cases in the group of SHC (16%) and 4 cases in the PCLD one (44%). The Table III shows the data compared between the two groups. There were no cases of bile leakage and neither of severe cardiopulmonary complications.

All patients were followed for a mean of 24 months (range, 8 to 48 months). The first follow-up was 30 days after the surgery. The follow-up consisted of the clinical observation, laboratory investigations and US. The long-term outcomes confirmed that all the patients remained free of symptoms and relapse of the disease.

Discussion

The incidence of coincidental cysts has increased up to 18% due to the increasing use of diagnostic imaging. The three important steps, regarding liver cystic lesions, are the following: making the diagnosis of the nature of cysts; determining whether the patient’s symptoms are related to cystic lesions, deciding the right therapy. Usually liver cysts are asymptomatic and only about 5% of patients are symptomatic.

The symptoms may change due to their size, anatomical site, or related complications. The most common symptoms are epigastric pain, nausea, early satiety, vomiting. Complications may include obstructive jaundice, portal hypertension and Budd-Chiari Syndrome. Some rare complications of liver cysts may be: torsion, infection, rupture to the peritoneal cavity, intracystic hemorrhage.

Laboratory findings are generally normal, and sometimes there is only a little alteration of liver function laboratory findings. The most accurate, non-invasive imaging method modality for diagnosis simple cysts is ultrasound where generally cysts appear anechoic spherical or oval shaped, anechoic (fluid filled cavity). Recent advances in CT and MRI technology might result in even higher sensitivity rates. The link between abdominal pain and the presence of cystic lesions should be always put in consideration before any surgical therapy, and other possible causes of pain should always be excluded.

Complicated cysts, echinococcosis and cystic neoplasms (cystadenoma and cystadenocarcinoma), require accurate diagnosis in the early stage in order to prescribe accurate therapy.

Cystadenoma (HC) and cystadenocarcinoma (HCA) are biliary cyst tumors that originate from the biliary epithelium, are generally asymptomatic, and occur more frequently in female patients older than 40 years of age. Abnormal serum and cystic fluid markers, such as CA 19-9 and CEA levels, may be useful for the diagnosis. Diagnostic imaging shows a lesion with internal septations, thickened wall, papillary projections, and calcifications. However, in most cases, differentiation between HC and HCA is not possible.

CEUS enables us to visualize vascular flow within septa or solid components of cysts, which is not present in simple cysts with intracystic haemorrhage. The rupture of blood vessels inside of cystic wall is the mechanism of bleeding in a simple cyst. CEUS can be helpful in differentiating HC and HCA from complicated cysts when USG, CT or MRI is inconclusive. FNA could be useful to exclude complicated cysts. HCA/HC findings are extremely rare (2,2%) with a low percentage of malignancy (0,2%). The presence of multiple hepatic cystic lesions may be linked to PCLD a genetic disease, associated with mutations in the PKD1 or PKD2 gene encoding respectively for polycystin 1 and polycystin 2 that are important for the functioning of the primary cilium. The pathophysiology of PCLD is caused by a malformation of the hepatic ductal plate and cilia of cholangiocytes. As consequence, multiple cysts arise from progressive dilatation of these abnormal ductules that display the same epithelium and structures of functioning cholangiocytes.

PCLD is characterized by a continuous increase in the number of cysts. It is more common in female patients. Pregnancy, multiparity increase the risk for severe hepatic cystic disease, because of the cystic increase in size and number.

There are different clinical classification proposed to grade the severity of PCLD.

Gigot’s classification is used for staging based on CT results:

- type I, less than 10 large cysts (more than 10cm in maximum diameter);
- type II, with multiple and widespread cysts of moderate size, but with remaining large areas of non-cystic liver parenchyma;
- type III, massive, diffuse involvement of liver by small and medium-sized cysts with only a few areas of normal liver tissue.
According to Morino’s classification, PCLD is divided into 2 groups:
- Type 1: limited number of large cysts in the anterior segments of liver;
- Type 2: multiple and widespread small cysts.

Schnellendorfer’s classification is used to differentiate patients in order to propose the best treatment and takes into consideration the severity of symptoms, cysts characteristics, the presence of portal vein or hepatic veins occlusion.

When patients become highly symptomatic or complicated the treatment of hepatic cysts is indicated.

Different therapeutic options have been proposed for symptomatic patients: sclerotherapy with percutaneous aspiration of the cyst, reserved for those patients who are not eligible for surgery and general anesthesia; laparoscopic or open surgical fenestration also known as deroofing, consisting of an excision of the cystic roof to provide communication between the cyst and the peritoneal cavity; hepatic resection and liver transplantation.

The surgical management in patients with PCLD is:

- Open technique to conclude the operation in safety.
- Robotic procedures, consideration must be given to the patient’s condition.
- Laparoscopic deroofing in PCLD type I; resection or liver transplantation (OLT) in PCLD type II/III medical management may be vary. The somatostatin analogue, which have inhibitory effect on cholangiocyte proliferation, reduces the volume of polycystic livers but with modest clinical effect.

Only in a selected group of patients with symptomatic PCLD, somatostatin analogues are indicated.

The correct approach for the therapy of cystic liver disease remains a clinical challenge. All possible therapeutic possibilities have to be evaluated and depend strictly on the patient’s condition.

In our series, we have treated with laparoscopic deroofing patients affected both by SHCs and PCLD type I.

In fact, we think that in Gigot’s type I, symptoms are not connected to the size of the entire liver but to the size of the largest cysts and that the main target of laparoscopic deroofing is to obtain complete decompression of cysts, leading to disappearance of the patient’s symptoms.

Our study focused on a selected group of patients who underwent laparoscopic fenestration whose symptoms were specifically related to cysts. For this reason, this therapeutic approach provided immediate complete relief of symptoms for both SHCs and PCLD.

The procedure was completed with success by laparoscopic approach for all 28 cases. The location of cysts didn’t represent a contraindication to the laparoscopic approach. In our series, we recorded neither mortality nor intraoperative complications, the postoperative morbidity has been 25%, with no case of recurrence and without the necessity of reoperation, because of the complete regression of the cystic disease and the symptomatology. The long-term outcomes confirmed that all the patients remained free of symptoms and of radiologic refilling of cysts.

Conclusions

The right approach for the therapy of cystic liver disease remains a clinical challenge. All possible therapeutic possibilities have to be evaluated and depend on the patient’s condition. The management in patients with PCLD is more complex.

With laparoscopic surgery, it is possible to reach important goals: symptom relief equivalent to that with open fenestration with the benefits of laparoscopic surgery by reducing the hospital stay. The technical feasibility and safety as well as the good short- and medium-term results made the laparoscopic approach the best choice for the management of patients with symptomatic liver cysts.
**Riassunto**
La presenza di cisti epatiche è una condizione patologica abbastanza diffusa, con una prevalenza tra il 2,5 e il 5% tra la popolazione globale. Il riscontro diagnostico è notevolmente aumentato negli ultimi anni, indipendentemente dalla presenza di sintomatologia per l’uso diffuso degli esami ecografici, oltre naturalmente alla TC. Possono essere di natura congenita o acquisite. Tra le congenite riconosciamo le cisti epatiche semplici, e la malattia polyclistica epatica. Tra le acquisite invece le lesioni cistiche possono essere di diversa natura, neoplastica, metastatica parasitaria o angiomatosa. Le indicazioni al trattamento, per le cisti non parasitari esiste quando queste sono sintomatiche, complicate o vi sia il sospetto di una malattia cancerosa. Diversi trattamenti sono possibili, ma l’asportazione e la fenestrazione con tecniche chirurgiche mini-invasive sono sempre più realizzate. In questo studio presentiamo la nostra esperienza nel trattamento laparoscopico, delle cisti epatiche semplici e nella malattia epatica policistica. Dal 2005 al 2018, 28 pazienti sono stati arruolati per il trattamento di cisti epatiche. Tutti i pazienti sono stati sottoposti a trattamento laparoscopico in regime di elezione. Si è proceduto ad un deroothing delle cisti più grandi mentre le più piccole sono state dette per aspirare. Nei casi di malattia policistica epatica (tipo I secondo Gigot) la procedura è stata ripetuta per ciascuna delle cisti. Non ci sono stati casi di conversione a chirurgia ad addome aperto, e i pazienti sono rimasti in follow-up in periodo da 12 a 48 mesi. In nessun caso vi è stata una ricomparsa della sintomatologia. Il corretto approccio al trattamento delle cisti epatiche rimane una sfida. Il trattamento alla malattia epatica policistica è ancora più complesso. Le tecniche mini-invasive e la laparoscopia offrono la possibilità di un trattamento sicuro efficace e in grado di raggiungere obiettivi terapeutici importanti.

**References**


