Surgical Emergencies in Liver Hydatidosis: Not too Much, but Severe
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Abstract
Liver hydatidosis is a zoosporic caused by *Echinococcus* that has worldwide distribution. There are three types of therapeutic options: surgery, medical treatment and PAIR. But surgery is the treatment that offers better long-term results. Usually surgery for liver hydatidosis is made as a scheduled procedure, but sometimes, severe symptoms provoked by liver cysts must be treated in emergency setting. We have made a review of every complication that could provoke liver hydatidosis: complications related to cysto-biliary communication, intraperitoneal rupture, vascular complications and rupture in surrounding organs. We have made a review of epidemiology, diagnosis and treatment of each complication, focusing when an emergency treatment is needed. We could conclude that only few patients require an emergency treatment due to liver hydatidosis: morbidty and even mortality is high because diagnosis is difficult and sometimes delayed, and we have to face to severe medical situations (anaphylaxis, hypovolemic or septic shock, cholangitis, etc.).

Keywords
Liver; Hydatidosis; Complications; Emergency; Review; Surgery

Abbreviations
HCL: Hydatid cyst of the liver; BDR: Bile Duct Rupture; FIR: Frank Intrabiliary Rupture; ERCP: Endoscopic Retrograde Cholangiopancreatography; HLA: Hydatid Liver Abscess; HAP: Hydatid Acute Pancreatitis CCF: Cyst-cutaneous Fistula

Introduction
Hydatid disease is a parasitic disease caused by the genus *Echinococcus* that is a major health problem in endemic areas [1-13]. The most common clinical presentation is liver hydatid cyst (HCL) [2,3,5,6,9,10,14,15]. The clinical presentation of HCL can be divided into: uncomplicated cases, approximately 80%, and complicated cases, among which we can include: cysto-biliary communication (12% of patients), hepato-thoracic transit, spontaneous or traumatic intraperitoneal rupture (1.6%), vascular complications (0.8%), and perforation of HCL to abdominal organs or skin (0.2%) [2-8,12,16,17].

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How to prevent the occurrence of these complications is very complex because it is difficult to predict the natural story of a hydatid cyst. Currently, the most accepted treatment algorithm is based on the radiological characteristics of the cyst and the presence of symptoms. In this algorithm, active cysts and/or symptomatic cysts should be treated and inactive cysts observed [18]. But some authors support the use of “wait and see” policy in asymptomatic patients with active cysts. The main problem is that does not exist any data that could define which cysts are going to present complications, occasionally inactive cysts could complicate and active cysts will not present clinical problems. We only can define some cysts with high complication risk (large, central, active, with progressive growth), or at increased risk of rupture (peripheral) [19].

Patients with uncomplicated HCL are usually operated electively, but some patients with complicated cysts present clinical situations that require emergency surgery. Our Liver Surgery Unit whenever feasible opts for radical techniques in HCL: total cystectomy or hepatectomy, performing conservative techniques only when radical surgery is not technically feasible. Morbidity, especially biliary-related morbidty, the length of hospital stay and the local recurrence rate is lower in radical techniques [17,20]. In emergency situations, surgeons often do conservative techniques due to local cyst conditions or extreme clinical situation, and these techniques had a higher relapse rate. We will expose in detail the clinical situations that may require emergency surgery.

Complications Caused by a Cysto-Biliary Communication

Communication between HCL and the biliary tree could be clinically silent (5-6% of patients), or could provoke serious clinical scenarios: jaundice, and/or cholangitis, liver abscess, cyst infection, or acute pancreatitis [4,21,22].

Cysto-biliary communication

The rupture of HCL into bile duct (BDR) is the most common complication of HCL. It occurs in 5-25% of patients [16,22-26]. A large series of 2013 patients found a 12% percentage of BDR [8,26]. This variability is due to the absence of an internationally accepted definition of this entity [22,23]. The BDR are classified according to size in minor breaks or single cysto-biliary communication (10-37% of patients with HCL), which is a little communication between the cyst wall and small biliary radicals [21-23], and a bigger BDR call frank intrabiliary rupture (FIR), (3-17% of patients with HCL). In FIR, the content of the cyst could drain into the bile duct and cause obstructive jaundice [4]. When the FIR is greater than 5 mm, some material of the cyst is observed in the bile duct up to 65% of the patients [27]. There is another classification made by Lewall and McCorkell [28] that distinguishes three clinical forms of BDR: contained rupture, direct communication and FIR.

A FIR could cause obstruction of the biliary system causing cholangitis and sepsis [8,26]. Other symptoms include fever, right upper quadrant pain, nausea and vomiting, flatulence, weight loss and palpable mass [21,23,24]. It occurs more frequently in cysts located in...
the center of the liver and cysts in advanced stages [21,23]. In 55-60% of cases, FIR is in the right intrahepatic bile duct, in left intrahepatic bile duct (25-30%), and exceptionally at the hilar confluence or gallbladder [21,22].

Diagnosis of FIR can be done pre-, intra- and postoperatively. When the patient has obstructive jaundice, the diagnosis of FIR is relatively simple, and imaging techniques allow us to visualize the FIR. But if there is no jaundice, correct preoperative diagnosis is rarely, and do not exceed 25% [23,24].

Abdominal ultrasound and CT are the best methods to diagnose FIR. The presence of a HCL associated with dilated bile duct and cholangitis is very suggestive of FIR. Ultrasound could differentiate between daughter cysts and stones in the common bile duct [16]. The CT also allows us to evaluate other organs that may be affected by hydatid disease [16]. The cholangio-MRI has a high diagnostic efficiency of FIR. The ERCP (Endoscopic retrograde cholangiopancreatography) has the benefit of being able to perform not only diagnosis, but also therapeutic measures [19,29,30].

Intraoperative signs of FIR are: bile in the cyst, to see an intracystic orifice through which drains bile or bile duct dilated [23,24]. Performing a cholangiography with methylene blue can help you find the communication [23]. The presence of bile in the drainage postoperatively, is an indirect method of recognition of a cystobiliary communication undiagnosed pre or intraoperatively [23]. ERCP with sphincterotomy and biliary stent placement should be done, if output of biliary fistula is high [23].

Surgery is the best treatment of FIR [16,22,25]. Delaying treatment of asymptomatic FIR only could cause a severe cholangitis or liver abscess [16]. Although the data available in the literature are usually retrospective [8], the most accepted surgical strategy in a patient suspecting FIR is to start performing an intraoperative cholangiography. If FIR is confirmed, the procedure performed is: opening bile duct, cleaning of bile duct, choledochoscopy to confirm that the biliary tree is clean and free of cystic content and placement of a T-tube (level III evidence, grade A). The T-tube is easier to perform and less risky than enterobiliary anastomosis, and also allows postoperative monitoring of biliary tree, but is not free of complications [8,22,24,25]. Sphincteroplasty or transduodenal sphincterotomy is currently unused.

A hotly debated topic is how to treat HCL when FIR is present. Total cystectomy with resection of the fistula is difficult to do, especially when cholangitis occurs, so it is only recommended in peripheral cysts. Therefore, the decision about optimal treatment depends on several variables such as: size and location of the cyst, proximity to vessels and features of the cyst wall [8]. The treatment of FIR over 5 mm has been very varied: intracystic suture holes, transistular internal drainage, double drainage, cystojejunostomy, etc. The external drainage methods have a high morbidity and a long hospitalization. Some authors defend performing a choledochoduodenostomy, in elderly patients with obstructive jaundice, biliary dilatation or papillary stenosis [8,24,25,31]. Treating only the cyst or FIR is not enough, but in some patients with significant comorbidities ERCP plus albendazole could be performed for bile duct cleaning [29].

FIR patients require complex interventions. Mortality published on interventions for FIR ranges from 1.8 to 4.5%, with a morbidity ranging 19-43% [23,27]. Most frequent causes of death were sepsis and liver failure. FIR patients should receive postoperative treatment with albendazole for 3 months (Level IV, Grade C) [8].

Conducting a preoperative ERCP with sphincterotomy is emerging as a therapeutic strategy in patients with FIR, but the information is based on retrospective studies [8]. ERCP has a dual diagnostic and therapeutic function [32], defining the FIR and allowing the cleaning of the bile duct, treating cholangitis and biliary obstruction (Figures 1 and 2). This approach let us operate most of the patients with FIR as an elective procedure [8]. ERCP also avoids bile duct exploration during surgery and appears to decrease the rate of postoperative biliary fistula [8,32]. Some authors have recommended routine ERCP in lesions larger than 7.5 cm because seems to be a direct correlation between the size of the cyst with the existence of FIR, but there is no consensus on the therapeutic algorithm [31]. In the postoperative period may be useful for cleaning residual material in the biliary tree or postoperative biliary fistulas [8].

Our therapeutic strategy differs from that published in the literature. When a patient with a symptomatic FIR attends the Emergency Room presenting with cholangitis and jaundice, we perform routine medical measures (fluid therapy and antibiotics) and ERCP. If ERCP cannot be performed due to technical problems (no cannulation), or does not solve the clinical situation, we perform...
emergency surgery. But we want to remark that in our center, no emergency surgical procedure due to FIR has been made since preoperative ERCP policy in FIR patients has been adopted (11 cases). In every case, we perform elective surgery a few days after ERCP, and it is done by hepatobiliary surgeons. The preoperative ERCP with bile duct cleaning takes away the need to perform surgery on the bile duct and avoids the morbidity associated to this procedure.

**Hydatid Liver Abscess (HLA)**

HLA infection and subsequent abscess development can occur as a primary abscess, secondary to retrograde infection from the bile duct through a cysto-biliary communication, or after PAIR or D-PAI [16,33]. The HLA is a complication that occurs in 5-15% of patients with HCL [16]. Curiously, the HLA is relatively well tolerated by patients when compared to other liver or intra-abdominal abscesses, and 25% of cases are diagnosed intraoperatively [16]. When symptomatic, the most common symptoms are: right upper quadrant abdominal pain, fatigue, fever and jaundice [16,34]. A small percentage of patients present septic shock, as first sign of HCL, we have observed only in two patients. Patients with HLA had not always leukocytosis [16]. In CT, the presence of a fluid level in HCL and also daughter vesicles are characteristic of HAA, but not always are present. In 50% of patients, there is wall calcification [16].

Surgical treatment is needed when HLA is present [16]. In severe cases of HLA, it is common to perform emergency surgery. Intraoperatively, the surgeon could appreciate a break in HCL that could provoke an acute peritonitis in 10% of patients [30]. When cyst is opened, we could see frequently putrid greenish creamy pus mixed with incomplete or destroyed hydatid membranes without viability [34]. The cultures of intracystic material are positive in 80% of the cases, the most common organisms are *E. coli, Enterobacter* and *Pseudomonas* [16,30]. The classic surgical treatment consisted in performing drainage of the cyst cavity, with or without biliary drainage. But this technique has a poor outcome and often presents serious complications: abscess residual cyst cavity, biliary fistula and chronic suppuration, which usually require reoperation with high morbidity and mortality [34]. Performing radical surgery is associated with decreased morbidity, but not always can be performed due to the characteristics of the cyst or the medical condition of the patient. In these cases, subtotal pericystectomy and placement of drainage tube could be a good technical option [16,34]. The morbidity of surgery ranges from 16 to 37% and mortality is 3% [16]. There is no consensus on the need for postoperative albendazole [16,30]. The patients presenting HLA after PAIR could be treated with percutaneous needle aspiration [33].

In our center, patients with HLA, in addition to general measures of resuscitation and antibiotics, are initially treated with percutaneous drainage of the abscess [35]. If there is clinical improvement, we planned elective surgery, with the advantage of having a more medically recovered patient. If jaundice is present, we also perform a preoperative ERCP, in order to perform elective surgery by hepatobiliary surgeons. If no clinical improvement it is observed after percutaneous drainage or those presenting with acute peritonitis, emergency surgery is performed trying to apply radical techniques or subtotal cystectomy.

**Hydatid Acute Pancreatitis (HAP)**

Hydatid acute pancreatitis (HAP) is a very rare clinical situation [10]. A cystobiliary communication with pass of the cyst content to bile duct is needed to HAP occur [10]. Its etiology is not clear, it may be due to mechanical obstruction of the cyst material that blocks the bile duct and pancreatic duct obstruction, or due to papillary or allergic inflammatory response [10]. The HAP is usually mild and a rapid clinical recovery occurs, probably because the content of the cyst is softer than the stones [10]. There are no reported cases of severe HAP with poor course that have required performing a pancreatic necrosectomy. An ERCP with sphincterotomy and bile duct cleaning usually solves HAP [10]. An elective surgery could be done one week before.

**Intraperitoneal rupture of HCL**

Intraperitoneal rupture is rare and occurs in 1, 6-8% of patients with HCL [5,7,8,15,36]. Factors of HCL rupture are: superficial cysts, large cysts (>10 cm), and viable cysts [15,36]. Etiology of the rupture can be spontaneous or traumatic, but a high-energy trauma is not required for it to occur [11,12,15]. When the cyst ruptures, a widespread of the parasite throughout the abdominal cavity could occur [7]. Although there are some cases of asymptomatic disseminated hydatidosis (Figure 3) [8], the most common clinical symptoms are severe abdominal pain, with peritonitis caused by peritoneal irritation due to cyst fluid and membranes [15,36]. Nausea, vomiting, hemoperitoneum and jaundice also could occur [36]. Some patients (1-7.5%) have several degrees of anaphylactic shock presenting with fever, urticaria and pruritis [2,5,7,9,14].

Lewall and McCorkell [28] classified Intraparenchymal ruptures into 3 types: type I, contained rupture with intact pericystic, type II, break into the biliary ducts and type III, complete rupture with cyst contents in the abdominal cavity [15,25]. Ultrasound is useful but the best method to diagnose a ruptured HCL is CT [5,8,9,14,15,36].

Intraperitoneal rupture requires immediate medical treatment against allergic reaction, and emergency surgery to drain the fluid and membranes and cleaning of the abdominal cavity with a scolicid solution [15,36]. Primary HCL should be also treated [5,6,15]. There is no consensus on which is the most effective scolicid: hypertonic saline, povidone and hydrogen peroxide [14]. If during the procedure the patient has hemodynamic problems, it could be washed out and quickly closed planning a second delayed surgical procedure [8]. Mortality is between 6-12% and morbidity is between 10 and 35% [8,14,15,36]. Postoperatively, we have to treat these patients with albendazole for at least 3 months [5,9,14]. Recurrence after rupture is frequent (7 to 29%), so it is recommended to perform a strict follow-up.
up to these patients. When a multiple site recurrence exists, it is a very demanding surgical procedure [36].

Vascular Complications

There are very few vascular complications due to HCL, but could be extremely severe:

Inferior vena cava

A HCL can cause compression of the inferior vena cava causing thrombosis and cavernomatous transformation, or exceptionally creating a fistula between the HCL and cava vein [1,3,37,38].

Portal vein

The portal vein may be affected by extrinsic compression of a HCL causing severe degrees of portal thrombosis, even portal cavernomatosis, or could create a real fistula with daughter vesicles in the bloodstream [2].

When HCL cause compression, elective surgery of the cyst could solve it. In the exceptional cases of fistula between HCL and vessels, we have to face a situation of extreme emergency, with anaphylactic and hemodynamic shock requiring emergency surgery with high morbidity and mortality.

Breaking to Other Organs

As mentioned before, the bile duct is where most often breaks a HCL. It may break in other organs, but it is extremely rare. The reason of low frequency is because the resistance of the intestinal wall is higher than that offered by the bile duct [6]. HCL Fistulas are described in stomach, duodenum, pancreas, colon and skin [6,39-42]. A CT scan is the method of choice in these patients [6].

Colon

Patients had a lower gastrointestinal bleeding that can be self-limiting or cataclysmic [6]. In certain cases, we observe hydatidenteria or hydatidorrhoea that is a very specific data [6]. Emergency surgery involves resection of the affected segment of colon, and to treat HCL that caused the fistula [6].

Stomach and duodenum

Is exceptional, we should perform excision of the cyst and gastric or duodenal resection.

Skin

The existence of a cyst-cutaneous fistula (CCF) is an extremely rare complication of hydatid disease, and only 7 cases have been published until 2006 [39-44] (Figure 4). The patients had the primary cyst in the liver in 5 cases [40,43]. All were primary cysts without previous surgery. The CCF occurs because the cyst grows and invades the abdominal wall, causing an inflammatory process that causes necrosis and fistulization, initially in subcutaneous tissue and subsequently, in some isolated cases in the skin causing a CCF [39]. Treatment consists of excision of the skin, subcutaneous tissue and affected wall, and treating of the HCL. Sometimes, the parietal abdominal defect requires complex reconstructions.

Conclusion

Usually surgery for liver hydatidosis is a scheduled procedure, but sometimes HCL must be treated in emergency setting. Several complications provoked by HCL could need emergency surgery: complications related to cysto-biliary communication, intraperitoneal rupture, vascular complications and rupture in surrounding organs. We could conclude that only few patients require an emergency treatment due to liver hydatidosis, ERCP and percutaneous drainage could let us perform scheduled instead of emergency surgeries. Morbidity and even mortality is high because diagnosis is difficult and sometimes delayed, and we have to face to hazardous intraoperative situations.

References


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Volume 2 • Issue 1 • 1000111 • Page 5 of 5

Top