

Diagnosis and management against the complications of human cystic echinococcosis

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Abstract Cystic echinococcosis (CE) (hydatidosis, hydatid disease) is a zoonosis caused by the larval stage of *Echinococcus granulosus*, typically affecting the liver. Hepatic cystic echinococcosis (HCE) is often asymptomatic, and symptoms occur largely when complications develop. Up to one-third of HCE can be shown their complications such as rupture of the cyst, secondary infection, and anaphylactic reaction. Clinically, patients present with pain, obstructive jaundice, cholangitis, anaphylactic reaction, and shock. Early diagnosis and treatment of complications of CE must be very important, since mortality is high when obstruction of the biliary ducts occurs, leading to ascending cholangitis and septicemia, anaphylactic shock, or even life-threatening conditions.

Keywords cystic echinococcosis; complication; diagnosis; treatment

1 Introduction

Echinococcosis is a zoonosis caused by the larval stage of *Taenia Echinococcus*. Human echinococcosis is a significant public health problem despite increased awareness and implementation of preventive measures, especially in endemic regions of China (Xinjiang, Gansu, Tibet, Inner Mongolia, Qinghai, Ningxia and Sichuan). There are two main forms of human echinococcosis, namely, cystic echinococcosis (CE), caused by larval stage of *Echinococcus granulosus* (*E. granulosus*), and alveolar echinococcosis (AE), caused by larval stage of *Echinococcus multilocularis* (*E. multilocularis*). *E. granulosus* occurs worldwide, but *E. multilocularis* is found only in the temperate northern hemisphere. The echinococcal larvae

enter the liver through the portal venous system. The liver acts as a filter, which makes it the most commonly affected organ (50%–75%).

Hepatic cystic echinococcosis (HCE) is often asymptomatic and in many cases is recognized incidentally on imaging studies performed for other reasons. Symptoms occur largely when complications develop [1]. Up to one-third of echinococcosis can be revealed by their complications based on our 7255 hospital case review. One of the most common and serious complications was intrabiliary rupture (IBR) (1%–19.5%) [2–7]. If not diagnosed early and treated in time with the proper procedures, such a rupture may lead to acute obstructive and suppurative cholangitis [8–10].

2 Rupture

Rupture is one of the major complications of HCE. Lewall and McCorkell have classified rupture of the cysts as three types: (1) contained rupture, occurring when only the endocyst was torn and cyst contents were within the pericyst; (2) communicating rupture, defined as the tearing of the endocyst and cyst contents via biliary radicals or bronchioles incorporated into the pericyst; and (3) direct rupture, which describes a tear of both endocyst and pericyst, with cyst contents escaping into the peritoneal or pleural spaces [11]. Generally, the contained rupture may be kept silent clinically, whereas the communicating rupture might cause biliary infection and liver abscess, and the direct rupture can rupture into physiological channels (biliary, bronchial tree), free body cavities (peritoneal, pleural or pericardial) or adjacent organs, which could result in severe clinical consequences.

2.1 Rupture into biliary tree

IBR is a complication of HCE which greatly increases the risk of mortality, due to complications of biliary

obstruction by hydatid debris, secondary pancreatitis, or bacterial cholangitis and septicemia [3–10]. One of the most common and potentially fatal complications of HCE is IBR. Clinically, patients present with pain, obstructive jaundice, cholangitis, or septic shock. Tsitouridis proposed the mechanism of IBR to be based upon three factors: the presence of small bile duct radicles in the pericyst, the condition of the echinococcal cyst wall, and the pressure level of the echinococcal cyst content [12].

Rupture into the biliary tree tends to occur into the right (55%–69.5%) or the left hepatic duct (25%–30%), whereas 10% occurred at the hepatic duct junction, common bile duct, cystic duct, or gallbladder [4,6]. IBR can lead to obstructive, septic or anaphylactic manifestations. Patients commonly present with right upper abdominal pain (82%–100%), obstructive jaundice (57%–100%), fever (70%–90%), acute cholangitis (20%–37%), abdominal mass (22%–39%), and rarely with acute pancreatitis, liver abscess or septicemia, or asymptotically (5%–6%) [3–10]. Between 1999 and 2009, 1465 cases of HCE were treated surgically in our hospital. Of these cases, 62 (4.2%) patients who were diagnosed IBR either pre-, intra- or post-operatively were included in our study. The cysts were localized in the right hepatic lobe in 38 patients (61.3%), in the left lobe in 15 patients (24.2%), and bilaterally in 9 (14.5%) patients. 81 cysts were treated in 62 patients, and 15 (24.2%) patients had multiple cysts. The majority of the cysts were daughter cysts (59.3%, CE₂ according to the World Health Organization-Infomal Working Group on Echinococcosis (WHO/IWGE) classification [13]). Cyst size ranged from 7–21 cm (25.8% between 5 cm and 10 cm, 72.5% greater than 10 cm). The average cyst diameter was 13.7 ± 2.5 cm [14].

Ultrasonography (US) and computed tomography (CT) have been routinely used to confirm the diagnosis of HCE. In endemic areas, the diagnosis of HCE has become easier, especially after the introduction of US and CT. In our study, sensitivities of US and CT were each 100%. However, the diagnosis of IBR is very difficult. The only direct sign of IBR is a visible cyst wall defect or communication between the cyst and the biliary tree [15]. US and CT demonstrated the place of rupture in 25%–77% of cases, overall [3,4,7,15]. We calculated sensitivity of US and CT for IBR to be 27.4% and 35.3%, respectively. The highly suggestive direct sign of rupture is identified in approximately 75% of cases. Secondary signs of IBR, such as a dilated biliary radical with intraluminal hydatid debris, are present in approximately 75% of cases [15].

Endoscopic retrograde cholangiopancreatography (ERCP) can be considered as an effective IBR diagnostic tool and had a sensitivity of 86%–100% [16,17]. In our study, eight patients with clinically suspected or confirmed cystobiliary communication were managed by ERCP. In all cases, the common bile duct contained germinative

membrane pieces and vesicles and the exact place of the rupture was demonstrated. Magnetic resonance cholangiography (MRCP) now offers a noninvasive diagnostic alternative without the risks associated with ERCP. Modern MRCP depicts accurately the details of a hepatic echinococcal cyst, as well as the communication with the biliary tree. A small cystobiliary communication cannot always be excluded by ERCP, and MRCP should be actively sought as an adjunct diagnostic method prior to surgery [18].

The main management priorities for patients with IBR are surgical removal of the cyst and all cystic elements, treatment of the residual cavity, clearance of cystic material from the biliary tree, and drainage of the biliary tree. Although opinions on management of the residual cyst cavity may differ, we prefer to perform partial cystectomy in all our patients; we have observed improved healing, likely due to decreasing the size of the cyst cavity and minimizing the risk of secondary infection.

There is a general consensus that common bile duct exploration with meticulous clearance of cystic material from the biliary tree is mandatory in every patient with IBR. Decompressing the pressure of the biliary system is the most effective way to reduce the likelihood of mortality from secondary complications. Various techniques have been recommended for biliary tract drainage, including choledochoduodenostomy [19], T-tube drainage [10], and transduodenal sphincteroplasty. A recent report suggested that routine biliary decompression using a T-tube (which is our standard) in patients with cyst-biliary complications was associated with lower incidence of postoperative bile leaks [5]. Placement of a T-tube enables simple monitoring of the biliary drainage during the postoperative period and prevents unnecessary bilio-enteric reconstruction and its undesirable long-term consequences. Also, it is a much easier and faster technique in the high-risk patient and is free of the perioperative morbidity of a bilio-enteric anastomosis or sphincteroplasty. However, Roux-en-Y choledochojejunostomy may be necessary for large cysts in which major ducts are disrupted.

Postoperative biliary fistulae are the most common complication in IBR of HCE. We generally elect to suture-ligate biliary-cystic communications. Suture ligation is recommended by several authors due to ease of performance [3,5,20]. In our series, only three patients developed postoperative biliary fistulae despite suture ligation, likely due to the fact that the source of bile leakage could be seen in only 37.7% of cases (11.7%–17.1% in the literature [3,21]). US [22] and telescopic visualization [23] have been suggested as aids to visualization, but these methods are insufficient to demonstrate whole ruptures. Intraoperative methylene blue injection has been described as very useful for fistula localization in the most difficult cases [24]. We performed intraoperative methylene blue instillation in 15 patients and were able to demonstrate the exact

location of rupture in 14 of them. We routinely advocate this technique to patients with HCE for the evaluation of the cystic cavity, because it may decrease the rate of complications caused by these communications.

2.2 Intraperitoneal rupture

Intraperitoneal rupture is defined as the release of brood capsules, protoscoleces, and even daughter cysts from the ruptured HCE into the peritoneal cavity. In acute symptomatic rupture, peritoneal irritation, acute abdominal symptoms, and allergic reactions occur. The incidence is about 1% to 4% [25,26]. In silent rupture, the patient presents with disseminated abdominal hydatidosis. Multiple cysts develop throughout the peritoneal cavity causing intestinal obstruction, gross abdominal distention, ascites, and cachexia several years after the rupture. This phenomenon has been known as ‘secondary echinococcosis.’

In endemic areas the sudden onset of bile-colored ascites in an otherwise healthy person, regardless of the fact that no hepatic cyst is visible, raises suspicion of intraperitoneal rupture. US and CT were the main diagnostic methods with 85% and 100% sensitivity, respectively, in identifying hydatid cyst rupture in our study. Emergency surgery remains the only effective therapy for a ruptured cyst. Surgical procedure consists of removing the cyst and possible daughter cysts spilled into the abdominal cavity in combination with peritoneal washing [25].

2.3 Adjacent organs rupture

The cyst ruptures into adjacent organs, e.g. the stomach, duodenum or small intestine. Those patients affected developed hydatidemia (hydatid cyst and membranes in the vomitus) and hydatidenteria (passage of hydatid membranes in the stools) with diminishing of clinical symptoms.

2.4 Pleural cavity rupture

HCE located in the dome of liver close to the diaphragm tend to generate intrathoracic complications. Infection and continued expansion of the cyst causes pressure, erosion, and adhesion to the diaphragm. Expansion of the cyst stretches and ruptures the diaphragm and the cyst eventually becomes contiguous with the pleural space, and then, with increasing intracystic pressure, the cyst ruptures into the pleural cavity.

3 Secondary infection

Hydatid cysts localized in the liver cause compression of the biliary system in up to 90% of the cases, which leads to

lesions resulting in biliary communication in 80% of the cases. The ensuing development of biliary-cystic communications allows small amounts of hydatid fluid to enter the biliary tree [20]. Biliary-cystic communication is a prerequisite for bacterial contamination. The incidence of secondary infection of HCE in the literature ranges from 11.0% to 27.1%. In our current reviews, it is 12.8% in a total of 1465 CE cases.

The most frequent symptoms of bacterial infected HCE include abdominal pain, fever, chills, jaundice, nausea, vomiting, and anemia. Signs of right pleural effusion, such as dyspnea and pleural pain, develop if the abscess extends through the diaphragm. Diagnosis of a secondary infection of HCE is based on a case history, clinical findings, imaging techniques, immunodiagnostic tests, blood cell count, and blood cultures. Without timely administration of antibiotics and drainage procedures, infected HCE remains uniformly fatal. The most common causes of death include sepsis, multiorgan failure, and hepatic failure. Percutaneous drainage has become the standard of care and should be the first intervention considered for small cysts. For cysts greater than 5 cm, ruptured cysts, and multiloculated cysts, surgical drainage is generally recommended over percutaneous intervention.

Occasionally, the entire cyst content undergoes aseptic necrosis, the parasite dies, and the cyst is filled with amorphous, yellow, debris that must be distinguished from the pus of secondary infection. Essentially, this is the gross appearance of a multivesicular cyst and not a suppurated cyst.

4 Anaphylactic reaction

CE antigens or toxic albumin is the most important components, which may cause allergic shock after cyst rupture spontaneously or secondarily. The degree of anaphylactic reaction depends on where the cyst ruptures. The anaphylactic mild reaction only represented skin erythema, itch, measles, and nausea in those patients. Lewall and McCorkell reported that 4 of 24 patients (16.7%) with ruptured hydatid cysts had allergic symptoms [11]. In our current reviews, anaphylaxis was only 0.3% within a total of 1465 CE cases. However, fatal anaphylaxis after cyst rupture has been described. The symptoms of anaphylactic shock are varied, but may often include such things as a constriction of the airway, light-headedness and fainting, swelling of the neck and face, itching, and low blood pressure. The diagnosis of anaphylaxis is clinical and does not rely on laboratory testing. In endemic areas, when typical symptoms are noted in association with acute abdominal symptoms, diagnosis is virtually certain. When unclear, ancillary testing may help establish the diagnosis. Immediate emergency treatment is required for this type of shock.

The first priority is to protect the airway and maintain adequate blood pressure, and may use antihistamines, such as diphenhydramine, and corticosteroids, such as prednisone, to further reduce symptoms (after lifesaving measures and epinephrine are given), and the emergency operation is also important.

5 Postoperative complications of hepatic cystic echinococcosis

Surgery still remains the cornerstone of the treatment for hepatic hydatid cysts. The desired goals of successful surgical management include the extinction of parasites, obliteration of the remaining cyst cavity, and prevention of postoperative complications, while minimizing the mortality rate of the treatment itself. Numerous surgical techniques (radical, non-radical, and laparoscopic) for the treatment of HCE have been described. However, the surgical procedure would be associated with numerous complications, including local recurrence, cavity-related complications (infection of residual cavity, biliary fistulas), secondary biliary cirrhosis and peritoneal dissemination. The rate of postoperative complications ranged between 8% and 80% [27,28]. Early local recurrence and cavity-related complications are the main problems affecting the success of the surgical management of hydatid liver disease.

5.1 Local recurrence

Recurrence is defined as the appearance of new active cysts after therapy of intrahepatic or extrahepatic disease. The failure to achieve permanent control of the primary treated cyst is considered to be the cause of the local recurrence. Local recurrence rate after surgery is between 1.1% and 9.6% in different series [27,28]. In our study, the rate is 2.5%. Here were some reasons for recurrence of hydatid disease: (1) The fluid of the cyst spilling into the peritoneal cavity, caused by rupture of cyst or incorrect manipulation with no sufficient protection of cyst-around viscera in the first operation. In puncture progress, if there are too many protoscolices in the cyst fluid (400 000/mL), spillage of cyst fluid may cause recurrence of hydatid cyst with a high incidence. (2) In the first operation, protoscolices comes into blood through laceration of liver parenchyma around the incision of the ectocyst wall, which may be contaminated by cyst fluid, or through laceration of liver parenchyma caused by overexerting in the process of scrubbing the membrane of ectocyst wall. (3) Puncture failure, which cannot reaspirate daughter cysts or matrix. There may still be protoscolices in the residual cavity, which may cause the growth of the new cysts. (4) Improper protoscolices agents and applied procedures. And (5) secondary infection.

Diagnosis and treatment for local recurrence are similar

to those for the primary disease. Radical interventions are also suggested in patients with recurrence that previously underwent surgery conservatively. However, these radical operations are technically more difficult, and reoperations have higher mortality rates. Due to the decreased rates of recurrence after radical operations, pericystectomy could be performed with lower morbidity and mortality rates in selected patients with solitary, small, and peripherally located cysts. Conservative operation with inadequate incision, good exposition, and spreading of the disease is also a proper choice, especially in a difficult case. Routine albendazole therapy should begin seven days before intervention and continue for three months thereafter.

5.2 Postoperative biliary leakage

Postoperative biliary leakage is the most common HCE complication. Theoretically, hepatic hydatid cysts communicate with the biliary system in up to 80% of the cases. 55% of the biliary–cystic communication may develop biliary leakage after operation. In order to reduce the complication rate, the defect must be directly sutured during operation and its application has been recommended by several hydatid centers, but communication between cyst cavity and biliary duct is difficult to be seen, although residual cyst cavity is fully explored during operation. In present studies, 11.7%–17.1% of the communications were determined during the intraoperative insistent examination of the cavities, and this led to the high incidence of postoperative biliary leakage [3,21]. For this reason, when a biliary communication is suspected during operation, the retrograde methylene blue infusion technique described in the present study can safely be performed. Surprisingly, in some cases, biliary leakage or fistulas may develop postoperatively without any objective sign, and most of the leakage closed spontaneously [14,20]. The thought that biliary leakage is not an important morbidity may be the cause of this under-reporting. Nevertheless, biliary leakage, whether transient or persistent, can be the source of secondary cavity infections. If not properly drained, it may result in abscess formation in the residual cavity or leakage to the peritoneum and eventually leading to bile peritonitis. If drained effectively, an external biliary fistula may develop, representing the most common complication in most of the leakage closed.

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