Case Report

Ruptured choledochal cyst: a rare presentation and unique approach to management

Michael Meschino, Carlos García-Ochoa, Roberto Hernandez-Alejandro

London Health Sciences Centre, Western University, London, ON, Canada

Correspondence to: Roberto Hernandez-Alejandro. Associate Professor of Surgery, Surgical Director of Liver Transplantation, Division of HPB Surgery, Department of Surgery, London Health Sciences Centre, Western University, University Hospital, (C4-211), 339 Windermere Road, P.O. Box 5339, London, ON N6A5A5, Canada. Email: Roberto.HernandezAlejandro@lhsc.on.ca.

Abstract: We present the rare case of a ruptured choledochal cyst (CC) in a young woman presenting with a two-day history of worsening upper abdominal pain. Imaging revealed a contracted gallbladder, dilated common bile duct (CBD), and a large amount of peritoneal fluid. Percutaneous paracentesis was performed, obtaining bilious fluid. Further imaging revealed cystic dilatation of the CBD and the diagnosis of rupture CC type I was made. The patient was initially managed conservatively with percutaneous drains, IV antibiotic therapy, and sphincterotomy through an ERCP. Elective cyst resection and Roux-en-Y hepatojejunostomy was performed 8 weeks later. It is important to differentiate a ruptured CC from other surgical emergencies without exploratory laparotomy. Initial conservative management could be considered, followed by elective resection once inflammation, infection, and other complications have resolved, avoiding the increased risk associated with an emergency operation or two-stage laparotomy.

Keywords: Choledochal cyst (CC); paracentesis; endoscopic retrograde cholangiopancreatography; endoscopic retrograde sphincterotomy; Roux-en-Y hepaticojejunostomy

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Introduction

Biliary cysts (BC) are congenital cystic dilatations of the intra- and/or extra-hepatic biliary tree that are exceedingly rare in North America. Incidence is geographically dependent, being more common in Asia (1/1,000) than in western countries (1/100,000-150,000). Women are more commonly affected than men in a 3-4:1 ratio (1). Traditionally, lesions affecting only the common bile duct (CBD) are referred to specifically as choledochal cysts (CC). In 1969, Babbitt established that the most probable cause of BC relies on an anomalous pancreatobiliary junction (APBJ) (2). According to Yotuyangi, an anomalous proliferation of the biliary epithelial cells before bile duct cannulation during development is one possible explanation (3). The most well-known classification system for BCs was established by Todani (4). There are five types, type I being the most common. Types I, II, III, and IVb are extra-hepatic and type IVa and V are intra-hepatic. BCs have the ability to produce a wide spectrum of complications, including strictures, cholangitis, rupture with biliary peritonitis, and malignancy (5). In this paper, we present the rare case of a ruptured CC managed conservatively at first, followed by elective resection after resolution of biliary peritonitis.

Case report

An 18-year-old female presented to the Emergency Department with a 2-day history of abdominal pain, nausea and vomiting. Vital signs were stable with the exception of mild fever (38.5 ℃). On examination, the patient looked slightly jaundiced and the abdomen was tender on palpation in the right upper quadrant. Past medical history revealed a similar episode 1 year earlier that settled without complication and was never investigated further.

Initial blood work revealed a picture of obstructive jaundice, with an elevated WBC (25.8×10^9 cells/L), total

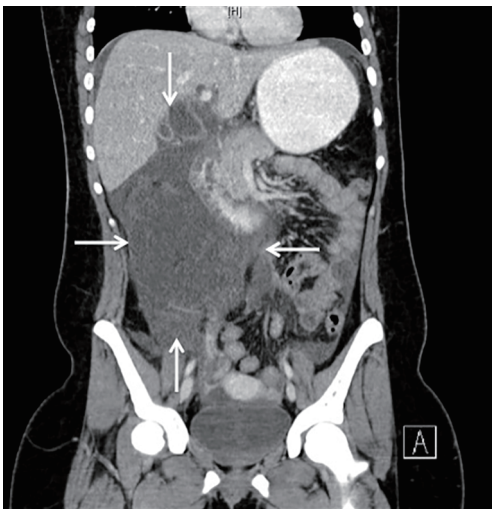

Figure 1 CT at initial presentation in the emergency department, revealing extensive peritoneal fluid collection (white arrows).

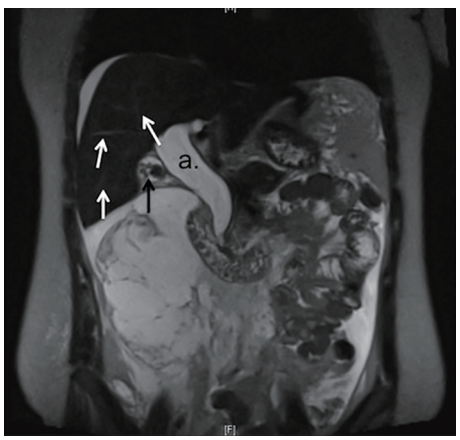

Figure 2 MRCP revealing cystic dilatation of the common bile duct with distal tapering (a), a contracted gallbladder with cholelithiasis (black arrow), and a normal intrahepatic biliary tree (white arrows).

and conjugated bilirubin (72 & 49 µmol/L respectively), and liver enzymes (AST 60 IU/L, ALT 133 IU/L, ALP 146 IU/L, GGT 165 IU/L). Lipase was also elevated (200 IU/L), while serum amylase, lactate, creatinine, and electrolytes fell within normal limits. Beta-hCG was negative.

Ultrasound revealed free fluid in the abdominal cavity, a contracted gallbladder and a dilated CBD. The patient was sent for CT, which showed a large fluid collection extending from the pericholecystic region to the pelvis and collecting in the right paracolic gutter (*Figure 1*). Furthermore, fluid

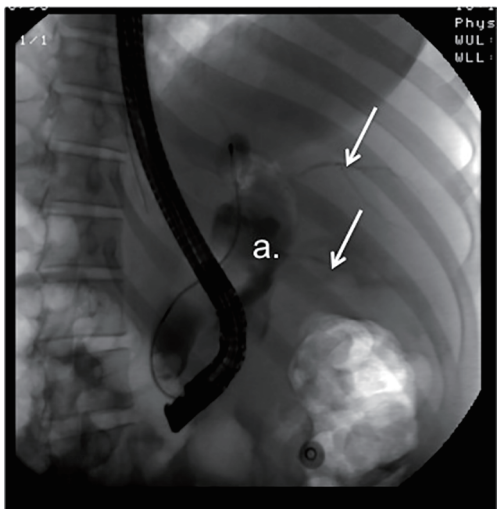

Figure 3 ERCP study showing common bile duct dilatation (a) and normal intrahepatic biliary tree (white arrows).

could be identified tracking retroperitoneally. The CBD dilatation was measured at 3.2 cm in diameter. A MRCP study further characterized the severe CBD dilation with distal tapering and a normal intra-hepatic biliary tree, confirming the diagnosis of CC type I (*Figure 2*).

The patient was admitted and referred to HPB Surgery. A diagnostic paracentesis revealed clear bile. Intravenous fluids and broad-spectrum antibiotics were given, a nasogastric tube was placed and two percutaneous intra-abdominal drains were placed by Interventional Radiology, one sub-hepatic and one inferiorly into the pelvis. An ERCP was performed, including the removal of two small cholesterol stones and a sphincterotomy to relieve pressure in the cyst and promote sealing of the rupture. There was no radiological evidence of bilious leakage from the CBD (*Figure 3*). The patient did well after the procedure and was discharged home after a few days of observation. The plan was to continue conservative management for 6-8 weeks, avoiding laparotomy and definitive surgical management until the perforation healed and the bili-peritoneum had resolved. The percutaneous drains were left in and continued to put out minimal biliary fluid over the next 6 weeks.

A follow-up CT performed 6 weeks later showed resolution of the bili-peritoneum and decompression of the CC (*Figure 4*), establishing the success of initial conservative management. The patient was scheduled for elective resection and reconstruction at 8 weeks after initial presentation. The abdomen was accessed through a right subcostal incision and the anatomy was revealed through

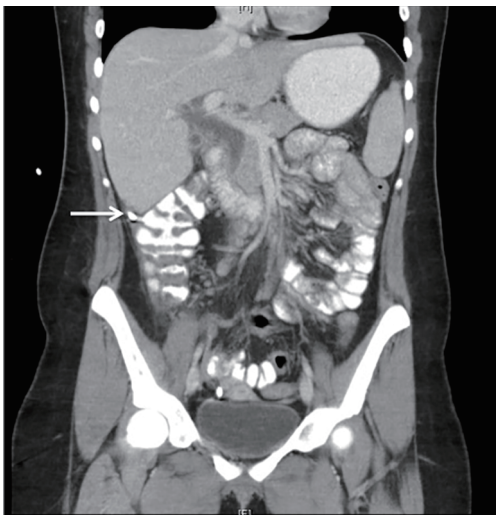

Figure 4 CT after 6 weeks of conservative management showing percutaneous drain placement (white arrow), resolution of the biliperitoneum and decompression of the choledochal cyst.

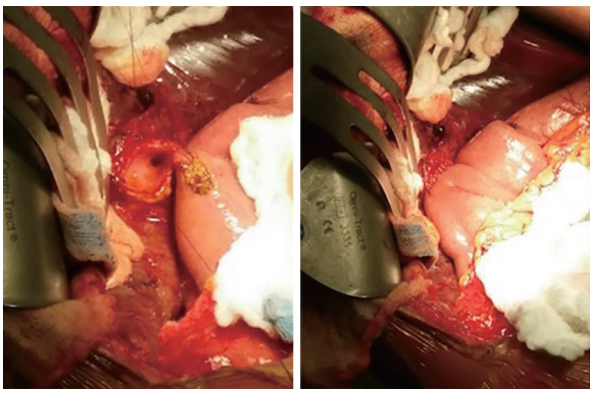

Figure 5 Reconstruction (intrahepatic Roux-en-Y hepaticojejunostomy).

minimal adhesions. Exploratory laparotomy revealed small collections of bile around the percutaneous drains. The cyst was identified and no other areas of abnormality were visualized. The likely site of rupture was identified within the CBD as an area of fibrosis and thinning of the wall. Mobilization of the duodenum was performed through a Kocher maneuver. The portal vascular structures were identified to avoid injury. Excision of the cyst as close to the pancreatic junction as possible, together with cholecystectomy, was performed with minimal blood loss. Intrahepatic hepaticojejunostomy was performed through a Roux-en-Y jejunal limb (*Figure 5*). The patient did well after surgery with no evidence of leak and was discharged home on post-operative day 5.

Pathology reported an acute on chronic cholecystitis with cholelithiasis. Inflammatory atypia of surface epithelium of the CC was noted with no evidence of malignancy. The patient was seen in clinic 4 weeks after surgery with no complications. She remains asymptomatic with normal liver function 1 year later and continues to be followed annually.

Discussion

Biliary peritonitis secondary to rupture of a CC is a rare presentation described in less than two percent of cases (6-8). Patients usually present as children (only about 20-30% present as adults) with the classic triad of palpable abdominal masses, pain in right upper quadrant and jaundice (9,10). BCs may present with different types of complications, including cystolithiasis, cholangitis, pancreatitis, and malignancy; the later has a 20-fold increased risk in BCs compared with the general population and increases with age (11).

The underlying cause for BCs is unknown. The most accepted theory involves an APBJ, which is found in 50-80% of BCs (2). In APBJ, the distal portion of the CBD and the pancreatic duct are formed 1-2 cm proximal to the ampulla of Vater and sphincter of Oddi, creating a large common channel where biliary and pancreatic secretions can pool. Due to greater secreting pressures in the pancreas, this mixture of secretions can reflux into the biliary tree. The biliary juices activate the pancreatic enzymes, creating an inflammatory environment which weakens the mucosa and dilates the walls of the CBD, thus making it more prone to cystic degeneration, dysplasia and eventually malignancy (12). In this case, however, imaging revealed a normal APBJ.

BC classification started in 1959 with Alonso *et al*., in which they described only three extra-hepatic types of cysts. Later in 1977, Todani *et al*. included intrahepatic cysts and by 2003 the classification was refined once more to include APBJs. Presently, the most commonly used classification is Todani *et al*. modified version which consists of five types and subtypes of cysts (*Table 1*) (13,15).

Risk factors predisposing to rupture are not well described in the literature. In this case, ERCP revealed multiple stones in the distal CBD as well as fibrosis and inflammatory atypia. It is possible that increased pressure and inflammation of the cyst as a result of choledocholithiasis predisposed this patient to spontaneous rupture.

Imaging is necessary to make the correct diagnosis of

Table 1 Biliary cyst classification (13,14)

Types	Characters
I	
Ia	Cystic dilatation of the hepatic and the CBD. The cystic duct arises from the BC
Ib	Focal cystic dilatation from the CBD, most commonly in the distal portion
Ic	Fusiform dilatation of the extra-hepatic bile duct that runs from the APBJ and may extend as far as the intrahepatic ducts
II	Saccular diverticula that are connected to the CBD through a narrow stalk
III	Also known as choledochoceles. Intra-duodenal cystic dilatations of the distal portion of the CBD. They may be lined by either duodenal or biliary epithelium
IV	
IVa	Consists of multiple intra- and extra-hepatic dilatations
IVb	Multiple extra-hepatic dilatations only
V	Also known as Caroli disease. One or more intrahepatic cysts or dilatations

CBD, common bile duct.

ruptured CC. This is especially true in the setting of acute rupture and biliary peritonitis where an accurate diagnosis may call for conservative management and avoidance of emergency surgery. Ultrasound is often the first imaging tool used because of its lack of invasiveness, availability and low cost, but CT is more sensitive at visualizing the intrahepatic ducts (14,16).

Both ERCP and MRCP are highly sensitive at confirming the diagnosis (17), although MRCP is preferred due to the invasiveness and risk of complications (pancreatitis, hemorrhage, perforation) associated with ERCP (18). However, ERCP potentiates the ability to conservatively manage a ruptured cyst through sphincterotomy or papillotomy, as was done in this case.

Over the years there has been a wide variety of approaches to management of ruptured CC. From 1960 to the 1980s, the treatment of choice was laparotomy and external biliary drainage with excision and biliary reconstruction occurring months to years later (19,20). However, in the 1990s many believed that the best management involved performing a single surgery at the moment of the BC rupture (21).

In recent years, the literature on the topic has changed again. Franga *et al*. in 2013 and Fragulidis *et al*. in 2007 , stated that the best-suited approach to a BC rupture was to perform a laparotomy-guided T-tube drainage followed by an interval cystectomy and hepaticoenterostomy (18,22). In 2008, Kang *et al*. suggested that a percutaneous trans-hepatic cyst drainage preceding definite surgery was the best option for BC rupture with peritonitis (23). In most cases, cystenterostomy (internal drainage) of the BC alone has been abandoned due to its high risk of malignancy, which has been reported in as many as 50% of cases (24).

A single stage surgery may be a suitable treatment for patients who are stable and free of other immediate complications. However, in a patient presenting with acute rupture and peritonitis, the risk of complications during definitive surgical management may be too high. In the acute setting, the placement of a T-tube and delayed excision with biliary reconstruction may be appropriate for patients that are not in optimal condition to tolerate surgery. However, due to the fact that this option consists of two surgeries, additional comorbidities may exist.

Conclusions

This case highlights the benefit of an accurate diagnosis and initial conservative management of BC rupture. Drainage of the peritoneal fluid and ERCP-guided sphincterotomy allowed decompression and temporary seal of the rupture. The benefit of a single elective surgery after resolution of peritonitis may be significant to reduce complications and improve long-term outcomes involving biliary function.

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Informed Consent: Informed consent was obtained from the patient for publication of this case report and any accompanying images.

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