Management of rupture of choledochal cyst

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Abstract One of the rare complications of choledochal cysts is rupture. In majority of the cases, the cause of rupture is unknown. Reconstructive surgery is the treatment of choice. We describe three patients with choledochal cyst rupture, who were admitted with acute abdomen. Diagnosis of biliary ascites with peritonitis was made in all the three patients. At surgery, two patients underwent T-tube placement, and definitive repair was done electively. One patient underwent definitive repair of ruptured choledochal cyst, but died due to septicemia. External bile drainage would be safer in emergency condition.

Keywords Biliary peritonitis · Choledochal cyst · Cyst rupture

Introduction

Choledochal cysts are uncommon congenital malformation described typically in childhood. They are definitively treated by excision of the extrahepatic bile duct and anastomosis of the hepatic duct with either jejunum or duodenum [1]. The incidence in the population is 1:5,000 [2], and is three times higher in females [3]. The prevalence is higher in Asian countries, especially in Japan [3]. The most common choledochal cyst is type I, with diffuse or segmental fusiform dilatation of the common bile duct. This type accounts for 50% to 85% of cases [4]. The classic symptomatic triad of cholestatic jaundice, abdominal pain, and a palpable abdominal mass occurs in only approximately 10% to 25% of the cases [5]. Although spontaneous rupture of a choledochal cyst is a rare complication, and can sometimes be the initial manifestation of a choledochal cyst, it should be considered in the presence of bile-like fluid in peritoneum, and differentiated from spontaneous perforation of the bile tract. In spontaneous rupture of choledochal cyst, cystectomy, cholecystectomy, and Roux-en-Y hepaticojejunostomy is the treatment of choice [6]. We report three cases of ruptured choledochal cysts; in 2 of the 3 cases, we had done T-tube external drainage as an emergency procedure as both the patients were seriously ill. Both these patients responded to the procedure and survived.

Case Reports

Case 1

A 13-month-old girl visited a local hospital because of abdominal distension, pain, and vomiting. Ultrasonography (USG) showed cystic dilatation of the common bile duct and mild ascites. For further treatment, she was referred to our institute with a diagnosis of acute intestinal obstruction. On admission, her vital parameters were: temperature 36.6°C, blood pressure 98/62 mmHg, pulse rate 140/min, and respiratory rate 34/min. She had distended abdomen and jaundice but did not have an abdominal lump. Laboratory tests demonstrated elevated total serum bilirubin and liver enzyme levels. She continued to be irritable and her vital signs had deteriorated. An emergency surgery was planned with a preoperative diagnosis of biliary peritonitis, made on the basis of previous USG showing cystic dilatation of common bile.
duct with ascites, and presence of jaundice. Laparotomy revealed ruptured choledochal cyst with loss of almost entire anterior wall of the cyst (Fig. 1). Peritoneal lavage was done and T-tube was placed at the perforation site. After placing the T-tube on posterior wall of the ruptured cyst, limbs of the T-tube were introduced into the common bile duct ends which were proximal and distal to the perforated cyst and purse string sutures were applied. Contrast study through the T-tube confirmed the free flow of bile to the duodenum without leakage and pancreatobiliary malunion (Fig. 2). Definitive surgery, that is, hepaticojejunostomy with Roux-en-Y anastomosis was done 12 weeks later. The patient was discharged on the 18th postoperative day without any remarkable event, and is doing well at 8 months of follow up.

Case 2

A 36-month-old girl had been suffering from abdominal pain for 1 week. After being referred to a local hospital, she was found to have choledochal cyst by computed tomography and was transferred to our hospital. On admission, she was lethargic and her temperature was 37.2°C, blood pressure 102/56 mmHg, pulse rate 136/min, and respiratory rate 28/min. She had distended abdomen but did not have jaundice or palpable abdominal mass. Laboratory tests revealed marked anemia and slight hepatic dysfunction. Treatment with broad spectrum antibiotics started, but fever, and abdominal pain with marked distension continued. At emergency abdominal surgery, bile drainage through T-tube placement into the cyst was done. About 1 L of bile was aspirated at the operation. The general condition of the patient improved postoperatively. Definitive surgery was carried out 8 weeks later, and the patient was discharged on the 12th postoperative day. She has been visiting our OPD for the last 1 year and thriving well without any complication.

Case 3

A 18-month-old boy presented to our emergency department with the complaints of abdominal distension, constipation, and vomiting. The patient was referred to us from a primary health care center with a provisional diagnosis of acute intestinal obstruction. On examination, the patient was found to have abdominal distension, but did not have jaundice or palpable abdominal mass. His laboratory parameters were normal. An x-ray of the abdomen showed few air fluid levels with ground glass appearance. The USG of the abdomen revealed distended bowel loops with ascites. An ascitic tap was found to have clear bile. An emergency laparotomy was done which showed a ruptured choledochal cyst. After aspirating 1,200 mL of bile, definitive repair was done, but died on the 4th postoperative day due to ongoing septicemia.

Discussion

Choledochal cysts present with classical triad of jaundice, abdominal pain, and a palpable mass; however, the course of the disease can be complicated [3]. Spontaneous rupture of choledochal cyst is considered rare, and has been reported in 1.8% to 2.8% of patients in various series [5]. Bile peritonitis following spontaneous rupture may lack peritoneal signs and could be difficult to diagnose but can be made by
hepatobiliary scintigram demonstrating an extrahepatic bile leakage [1]. The diagnosis of rupture should be considered in the presence of bile-like peritoneal fluid at the time of emergency laparotomy [1]. In our patients, we had done abdominal paracentesis to check the nature of ascitic fluid; a provisional diagnosis of biliary tract pathology was made since the ascitic fluid contained bile.

In the past, most reported cases have been managed with external drainage of the cyst followed by a second procedure to excise the cyst and reconstruct the biliary tract. Presently, most researchers, especially from the western world, recommend primary reconstructive surgery as treatment of choice [6, 7]. Reconstructive surgery may be tolerable even in emergency condition if a patient’s preoperative condition is stable [7]. When patients present late and have septicemia, they may not tolerate major definitive surgery. In case 3, the patient treated with primary reconstructive surgery died because of septicemia, which suggests that an emergent biliary drainage might have been safer. The condition of patients improves after external biliary drainage, and the patient may be discharged few days after operation. In our case, we discharged both the patients on the 5th postoperative day in satisfactory general condition with T-tube in situ. The patients underwent definitive surgery at a later date.

To conclude, external bile drainage would be safer in emergency condition, especially when patients present late with sepsis and comorbidities. Definitive surgery should also be regarded as a procedure with some risk of postoperative complications.

References