CASE REPORT

Spontaneous choledochal cyst perforation in pregnancy with co-existent chronic pancreatitis

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SUMMARY

Choledochal cysts are a congenital disorder of the bile duct, usually presenting in childhood. As they cause weakening of the wall of the bile duct, any distal obstruction, infection or reflux of amylase-rich fluid in the biliary tract can predispose to spontaneous perforation. A 25-year-old pregnant woman at 35 weeks of gestation presented with acute abdominal and fetal distress. On emergency laparotomy, common bile duct perforations were detected. T-tube drainage and caesarean section were performed. A t-tube cholangiogram revealed the diagnosis of a choledochal cyst with an abnormal pancreatobiliary junction and chronic pancreatitis. The patient later underwent cyst excision with Roux en Y hepaticojejunostomy. Spontaneous choledochal cyst perforation in pregnancy can be successfully managed by a two-stage operation.

BACKGROUND

Spontaneous common bile duct (CBD) perforation is an uncommon event. Most cases are due to blockage in the distal bile duct because of a stone, tumour or stricture.^{1 2} Weakening of the bile duct wall as seen in a choledochal cyst patient makes it prone to rupture. Choledochal cyst rupture is most commonly encountered in infancy or early childhood.³ We present a rare case of choledochal cyst rupture during pregnancy with co-existent chronic pancreatitis.

CASE PRESENTATION

A 25-year-old pregnant woman at 34 weeks of gestation presented with pain in the abdomen, vomiting and constipation of 2 days duration. On clinical examination, the patient had epigastric tenderness and fetal heart sounds were present.

INVESTIGATIONS

On ultrasonography of the abdomen, there was multiseptated collection in the right iliac fossa with single live intrauterine gestation. On non-stress test there was evidence of fetal distress.

DIFFERENTIAL DIAGNOSIS

- Preterm labour
- Acute appendicitis with perforation
- Ileal perforation
- Caecal perforation

TREATMENT

In view of the findings, the patient underwent emergency laparotomy with lower segment caesarean section. There was 200 mL of biliopurulent

fluid in the Morison's pouch, lesser sac and right paracolic gutter. The CBD was dilated, oedematous and friable with two perforations each of 5 mm located in the anterior wall of the mid and supra-duodenal part of the CBD (figure 1). The gallbladder, small bowel, large bowel and stomach were normal. Cholecystectomy, placement of T-tube and infant feeding tube via the perforations for external drainage of the bile duct were conducted. Postoperatively, the patient had intra-abdominal collections requiring ultrasound-guided aspirations following which the patient recovered and was discharged with T-tube and infant feeding tube in situ draining 400–500 mL bile per day. On follow-up, bile output from the T-tube gradually reduced. A T-tube cholangiogram performed after 3 weeks revealed a dilated CBD of 2 cm with minimal central intrahepatic biliary radical dilation with contained leak with free passage of contrast into the duodenum (figure 2). There was also opacification of the dilated main pancreatic duct due to a long common channel (figure 2). Based on these cholangiogram findings, diagnosis of a type 1 choledochal cyst with chronic pancreatitis was made. Contrast-enhanced CT of the abdomen showed a small organised collection in the paraduodenal region with a dilated CBD with tubes in situ. Also, the pancreas was atrophic and the main pancreatic duct was dilated with numerous small parenchymal and ductal calcifications. After 6 months of the primary operation, the patient underwent surgery for definitive treatment. Intraoperatively, the posterior wall of the choledochal cyst was densely

Figure 1 Intraoperative picture of choledochal cyst with perforation seen in the anterior wall of the bile duct.

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Figure 2 T-tube cholangiogram showing type 1 choledochal cyst with opacification of dilated and irregular main pancreatic duct via common channel.

adhered to the portal vein, hence Lilly's procedure with Roux en Y hepaticojejunostomy was conducted. Since the patient was not symptomatic for chronic pancreatitis, no pancreatic resection/drainage procedure was performed.

OUTCOME AND FOLLOW-UP

The postoperative course was uneventful and after follow-up of 6 months the patient was asymptomatic. Histopathology confirmed the diagnosis of choledochal cyst and gallbladder showed chronic cholecystitis.

DISCUSSION

Choledochal cysts are usually diagnosed in childhood, but up to 20% of cases are detected after 20 years of age.⁴ Rupture of a choledochal cyst is a rare event with incidence varying from 1.8% to 2.8% in various series.^{5–7} The postulated mechanisms to explain this spontaneous rupture are abnormal pancreatobiliary junction leading to efflux of amylase-rich fluid into the bile duct, obstruction of the common channel by protein plug and raised intra-abdominal pressure.^{4 8 9} Rupture of a choledochal cyst during pregnancy is extremely rare with few cases reported in the literature.^{4 10} Preoperative diagnosis is difficult because of masking of signs and symptoms due to pregnancy. Early diagnosis and treatment is imperative to prevent fetal and maternal mortality. Owing to the rarity of the disease, there are no standard guidelines for management of a perforated choledochal cyst during pregnancy. But based on the reported cases of ruptured choledochal cysts in children and non-pregnant adults, T-tube drainage in an emergency situation followed by definitive surgery of cyst excision and Roux en Y hepaticojejunostomy is the preferred treatment.^{3 4} In one study, the authors performed percutaneous transhepatic cyst drainage as an alternative to T-tube drainage.¹¹ Alternatively, if the patient is in stable condition and the bile duct is not friable, then the cyst excision and Roux en Y hepaticojejunostomy can be performed in the first

operation.¹² In our case, the diagnosis was made intraoperatively and the bile duct was too friable to hold sutures, hence T-tube drainage was performed.

Another important finding in our patient was the presence of chronic calcific pancreatitis. Co-existence of these two conditions is uncommon, with very few reported cases of occurrence in the presence of anomalous pancreatobiliary junction as seen in our case.^{4 13} If the patient is symptomatic or has any complications of chronic pancreatitis, then pancreatic resectional or drainage procedure for choledochal cyst excision may have to be performed. But since in our case the patient was never symptomatic for chronic pancreatitis, only the choledochal cyst was excised. But long-term follow-up is required to look for symptoms and endocrine or exocrine insufficiency.

Learning points

- ▶ In pregnancy, abdominal pain with intra-abdominal collection(s) should raise suspicion of biliary/perforative peritonitis despite the absence of signs of peritonism.
- ▶ In a patient with spontaneous common bile duct perforation, choledochal cyst should be suspected.
- ▶ Choledochal cyst perforation can be successfully managed by a two-staged procedure in pregnancy.

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Competing interests None.

Patient consent Obtained.

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