Spontaneous Perforation of the Common Bile Duct Mimicking Choledochal Cyst

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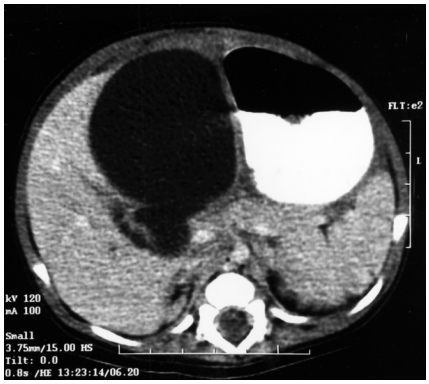
Spontaneous perforation of the extrahepatic bile ducts is very rare, but it is still the second most common surgical cause of jaundice in infants, after biliary atresia [1]. We describe an unusual case, which posed several diagnostic and management difficulties.

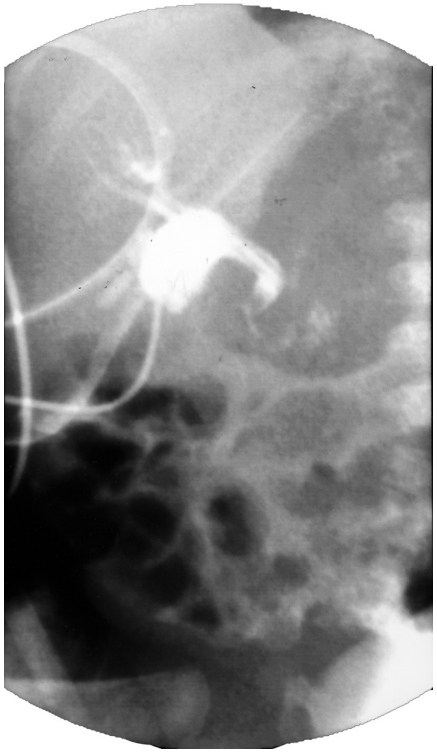
Patient Description

A 1 month old male was admitted because of abdominal distension and pain that had gradually worsened over the preceding 2 weeks. On examination, the upper abdomen was moderately distended and tender, with suspicion of a mass on palpation. The stool was yellow-green in color. The rest of the examination was normal. His white blood count was 19,700, total bilirubin 1.5 mg/dl, direct bilirubin 1.1 mg/dl, gamma-glutamate transferase 320 u/L, alkaline phosphatase 197 u/L, and lactic dehydrogenase 259 u/L. The rest of the liver function tests were within normal limits.

Abdominal ultrasound was followed by computerized tomography [Figure A] which demonstrated a large cyst in the upper mid-abdomen, connected to a mildly dilated common bile duct. Biliary scintigraphy revealed normal hepatic uptake and excretion, but most of the excreted material accumulated in a well-defined structure next to the CBD. The child was diagnosed as having a choledochal cyst and scheduled for surgery. At laparotomy, a 10 cm diameter pseudocyst filled with clear biliary fluid was found in the lesser sac. The CBD was perforated at its junction with the cystic duct. The perforation was oval, 1.1 cm long and 0.4 cm wide. Intraoperative cholangiography demonstrated a normal intrahepatic biliary tree proximally, and good passage distally. There was no evidence of debris or sludge [Figure B]. The abdomen was flushed with saline and a T-tube was inserted into the choledochus via the perforated orifice. The lesser sac was drained.



[A] Preoperative CT showing a huge cyst between the stomach and liver, with connection to the common bile duct.



[B] Intraoperative cholangiography showing the distal intraluminal plug.

Approximately 50–100 ml of bile per day was drained through the T-tube postoperatively. T-tube cholangiography revealed partial obstruction at the papilla of Vater, most probably due to viscous bile sludge. The choledochus was flushed with saline and the papilla was dilated with a balloon catheter via the T-tube. Two days later the baby developed Enterobacter sepsis, which was successfully treated by antibiotics. The T-tube bile drainage decreased gradually, and stopped completely 6 days post-dilatation. The T-tube was closed 10 days later. At the 4 month postoperative follow-up, a T-tube cholangiogram revealed normal anatomy and free passage of contrast material to the duodenum. The T-tube was removed, and the lesser sac drain was removed on the following day.

Three years later, this child underwent adhesiolysis due to small bowel obstruction. The postoperative course was uneventful, and he is now 5 years old and thriving.

Comment

The approximately 70 cases of spontaneous perforation of the common bile duct in infants that have been reported since it was first described by Dijkstrat in 1932 all have a similar mode of presentation

CBD = common bile duct

and site of perforation [2]. The age of presentation is usually between 2 weeks and 2 months of life. These patients usually have uncomplicated birth histories and develop normally until the onset of jaundice, acholic stools or dark urine. Signs of pyrexia or peritonitis are usually absent. Progressive abdominal distension, ascites and failure to thrive may be present and become gradually more prominent, and may be accompanied by vomiting, perhaps from compression of the second part of the duodenum.

Given that the site of perforation is consistently at the junction of the cystic and common bile ducts, it was postulated to be a developmental weakness in the bile duct wall. Necrosis secondary to pancreatic juice reflux caused by pancreaticobiliary mal-union associated with mural immaturity during infancy was also suggested as a possible factor. Viral infection was proposed, while Ando et al. [3] suggested that the perforation of the common bile duct may be related to an abrupt increase in intramural pressure due to obstruction by protein plugs. We consider that the latter might have been the case in our patient, who had partial distal obstruction at the ampulla of Vater. Interestingly, one of the suggested etiologic factors for spontaneous rupture of choledochal cyst is very similar [4], indicating that these two entities might have a common source. Spontaneous perforation of the bile duct has also been associated with a multiple organ disorder, known as Ivemark syndrome, which consists of splenic abnormalities, cardiac pathology, and abnormalities of the gastrointestinal tract [5].

We failed to make the correct diagnosis preoperatively in spite of a proper workup. The leaked bile was well circumscribed in the lesser sac on CT, with continuity to the CBD [Figure B], mimicking choledochal cyst. The diagnosis of choledochal cyst raises the question of performing endoscopic retrograde cholangiopancreatography as part of the investigation. Indeed, ERCP could have made the correct diagnosis, but it is not without morbidity, especially in a 1 month old. Furthermore, the child required surgery in any event and an ERCP would not have changed the overall management.

We also faced a postoperative management problem. At surgery, the distal patency of the CBD was demonstrated by passing an 8 French tube via the perforated orifice into the duodenum. The distal duct was flushed with saline, after which we performed cholangiography. In retrospect, we might have noticed a possible protein plug lodged distally. This missed plug increased the risk of morbidity because it necessitated washout and dilatation of the ampulla of Vater via the T-tube under general anesthesia, and the risk of sepsis.

The perforation may be apparent immediately at laparotomy, or it may be hidden by an inflammatory mass extending from the porta hepatis to the duodenum, as in our case. Careful dissection into the mass will identify the perforated orifice.

We placed a T-tube through the perforation to form a controlled biliary fistula. This technique decompresses the biliary tree and allows healing of the perforation. It gives time for any inspissated bile to clear from the distal bile duct, or as happened here, enables instrumental cleaning. This technique may be performed laparoscopically and perhaps prevent the formation of adhesion and bowel obstruction which would necessitate adhesiolysis.

A word of caution: the surgeon must be absolutely certain that the distal duct is completely clean before closing the abdomen. T-tube insertion is a reliable technique, and it had good long-term results in our case. Finally, a laparoscopic approach may prevent adhesiolysis in the future.

ERCP = endoscopic retrograde cholangiopancreatography

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