

Spontaneous Perforation of the Common Bile Duct in a Toddler: A Diagnostic Dilemma

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ABSTRACT

Spontaneous perforation of common bile duct (SPBD) is very rare but potentially fatal disorder. It is an unusual cause of acute abdomen. It is rarely suspected or correctly diagnosed preoperatively. Clinical presentation is as biliary peritonitis. The aetiology of SPBD is unknown, but proposed theories include congenital mural weakness of the common bile duct, ischaemia, distal biliary obstruction and pancreaticobiliary malunion, infection, and trauma. Bilious abdominal paracentesis, signs of peritonitis and absent free gas in abdominal X-ray will help to confirm the diagnosis. Preoperative recognition is necessary as early surgical intervention gives excellent prognosis. We report an interesting case of SPBD in a 4 years male child who presented to us as an acute abdomen. The literature has been reviewed with emphasis on the diagnostic dilemma of this rare condition. The treatment is surgical and consists of exploration and repair over a T-tube.

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Key words: Spontaneous bile duct perforation; Idiopathic common bile duct perforation; Biliary peritonitis; Biliary ascites

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INTRODUCTION

Spontaneous or idiopathic perforation of the extra hepatic bile duct

and biliary ascites in children is a rare clinical entity. The peak age of occurrence is around 6 months with age ranging from 25 weeks gestation to 7 years^[1]. The first case was described in 1932 by Dijkstra^[2]. Since then about 150 cases have been reported, mostly in infants. SPBD is one of the major causes of jaundice, requiring early corrective surgery. Most of the cases are diagnosed intra-operatively; therefore high level of suspicion is necessary. We report an interesting case of spontaneous bile duct perforation in 4-year-old male child and discuss the diagnostic dilemma in the light of available literature.

CASE REPORT

A 4 year old male child presented to us in the Emergency Department with sudden onset of abdominal pain and distention with vomiting and non-passage of flatus and stool for 5 days. There was no history of hepatobiliary disease or trauma. On examination he was pale and dehydrated, with tachycardia and hypotension. The abdomen was distended and tender. Ascites was present but liver dullness was not masked. Peritoneal tap showed biliary aspirate. No free gas was found in erect abdominal X-ray. Digital rectal examination was found to be acholic stool. His haemogram showed leukocytosis ($18.3 \times 10^9/L$) and neutrophilia (86%). Mild unconjugated jaundice was present and other biochemical tests were normal. Ultrasound of the abdomen revealed massive ascites with normal intra-hepatic biliary radicles. We resuscitated the patient and planned for surgery with diagnostic dilemma.

Operative findings included one liter of bile-stained purulent fluid, thickened gall bladder wall and without any stone and perforation. The stomach and duodenum were unremarkable. A perforation of size 0.5 cm×0.5 cm was present on the anterior surface of supraduodenal part of common bile duct (CBD). (Figure 1) CBD was normal and with no evidence of stone. As per-operative cholangiogram facility was not available, distal CBD patency was checked by passing an 8 Fr infant feeding tube into the duodenum. A cholecystectomy was performed and the CBD rent was repaired over a T-tube. Peritoneal cavity was irrigated with warm normal saline. Abdomen was closed after insertion of drain in sub-hepatic region. T-tube cholangiogram obtained on 12th post-operative day showed free flow of bile into duodenum without stone, obstruction or anomalous pancreaticobiliary junction. Histopathology of margin of perforated CBD showed chronic inflammation. Postoperatively;



Figure 1 Per - operative photograph showing perforation in anterior wall of CBD.

patient made a rapid recovery and was discharged after removing the T-tube on the 16th post-operative day. At follow up of two years, he was doing well.

DISCUSSION

The most frequent cause of spontaneous bile duct perforation is idiopathic. Various theories have been proposed implicating a weakness in the bile duct wall, obstruction distal to the perforation or a combination of both (Table 1).

Table 1 Suggested Causation Of Bile Duct Perforation In Children.

Weakness in wall of bile duct	Distal obstruction or stenosis
Congenital weakness ^[10]	Abnormal or inspissated bile ^[10]
Choledochal cyst ^[11]	Congenital stenosis of the ampulla of Vater ^[10]
Ischemia of bile duct ^[11]	Common bile duct stenosis ^[10]
Pancreatic reflux ^[4,11]	Congenital anomalous pancreaticobiliary junction ^[1,5,6,10,11]
Pancreatitis ^[11]	Stones ^[1,10]
Birth trauma ^[10]	Protein Plugs ^[1,10]
Torsion of gall bladder	Biliary sludge ^[10]
Presence of Diverticulum ^[10]	Biliary stones ^[10]
Abnormal glands of the bile duct wall ^[10]	Slow bile transit and biliary stasis ^[1]
Tuberculosis of CBD ^[12]	
Necrotising enterocolitis ^[13]	
Viral infection of the bile duct ^[10]	

Although bile duct perforation and choledochal cyst are considered as different entities, there is growing evidence that both may be interrelated problems with a common pathogenesis. 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^[3].

The condition presents a diagnostic dilemma because of its rarity, and absence of characteristic diagnostic finding, therefore a high index of suspicion is required. Previous history of biliary tract disease is usually absent. The presentation of SPBD may be acute or insidious, with the latter type being more common (80%) and characterized by progressive jaundice, painless abdominal distension, fluid hernia and acholic stool. The acute form is less common (20%) and presents with fever, vomiting and signs of fulminant peritonitis, toxemia, shock, with or without icterus, which raise the suspicion of rupture of a hollow viscus^[4].

Laboratory evaluations are not pathognomonic. Conjugated bilirubin and alkaline phosphatase levels may be elevated. An ultrasound will show free or loculated intraperitoneal fluid with normal intra and extra hepatic ducts^[4,5]. Paracentesis carried out under ultrasonography guidance may reveal bile-stained fluid. Hepatobiliary scintigraphy can show that the intraperitoneal fluid

originated from the biliary tract. It is highly sensitive and specific for SPBD^[6]. Endoscopic retrograde cholangiopancreatography [ERCP] is useful diagnostic and therapeutic procedure^[7]. Drip infusion cholangiography (DIC-CT) using meglumine iotroate is useful in pancreaticobiliary maljunction, biliary stricture and perforation. The 3D DIC- CT clearly demonstrated reflux of the contrast material into the main pancreatic duct as well as a long common channel, dilated biliary tree, and duodenal lumen. It is a valuable method in the evaluation of patients with suspected APBJ^[8]. Finally, peritonitis with absence of pneumo-peritoneum, bilious peritoneal tap and acholic stool are considered pathognomonic for SPBD.

Management is controversial and it depends on the condition of the child and availability of intra - operative T-tube cholangiogram. If it shows normal passage of dye into the duodenum, simple peritoneal drainage will be sufficient as the leak is expected to close spontaneously. If there is any evidence of distal CBD obstruction or if one is not able to obtain an intra - operative cholangiogram, then the condition is best managed by a T-tube drainage and peritoneal lavage^[9]. We did not have intra - operative cholangiogram facility and so we repaired the CBD over T-tube. It was presumed that perforation is due to biliary sludge or inspissated bile, which clears by itself. Emergency ERCP may be an alternative therapy of SPBD in infants and children. It helps not only in the diagnosis but it also provides endoscopic stenting and decompression. Drawback of this procedure is the availability of proper equipment and endoscopic expertise^[7].

Exploration of the porta-hepatis may be hazardous at emergency and therefore simple peritoneal drainage with T-tube drainage is recommended even if there is a distal obstruction. This entails less morbidity and has a good chance of curing the condition or at least stabilizing the patient for second look definitive surgery^[6]. Pancreatico-biliary malunion requires biliary intestinal anastomosis to prevent biliary cirrhosis, portal hypertension, recurrent pancreatitis, and ultimately biliary carcinoma^[4,5,6]. However this can be done at a second laparotomy when inflammation has settled. Repair of the perforation is unnecessary and may be hazardous because there is also the risk of postoperative stricture. With recent advances in laparoscopic surgery, diagnosis and percutaneous drainage is the alternative^[1].

CONCLUSION

SPBD is a rare and potentially fatal disorder. The condition presents a diagnostic dilemma. It is emphasized that SPBD should always be considered in young children who present with ascites, pale stools and the absence of impressive jaundice and peritonitis without pneumoperitoneum. The symptoms warrant abdominal paracentesis to assist in the diagnosis. Therefore a high degree of suspicion is required for its pre-operative diagnosis. Prognosis remains excellent with prompt diagnosis and treatment. Minimal surgery in the form of T-tube and peritoneal drainage is mainstay of treatment.

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