Spontaneous perforation of the bile duct in a neonate: Drainage or resection?

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Abstract

Spontaneous perforation of the bile duct is a rare disease, and delayed diagnosis without optimal treatment can be fatal. Abdominal drainage with or without repair of perforation seems to be adequate in most case series. We report on a 10-day-old female neonate with spontaneous perforation of the bile duct over the junction of cystic duct and common hepatic duct, who recovered uneventfully with follow-up for 3 years after receiving a single-stage operation of cholecystectomy and biliary reconstruction. Drainage only or resection of the gall bladder or bile duct should depend on the patient’s clinical conditions and intraoperative findings.

Keywords: bile duct; biliary reconstruction; cholecystectomy; neonate; spontaneous perforation

1. Introduction

Spontaneous perforation of the extrahepatic bile duct is rare in infants and neonates. It most often occurs in infants 2–20 weeks of age, and few cases are reported in older infants and children.1–3 The etiology is unknown, but there are many possible causes, such as congenital mural weakness of the common bile duct, relative ischemia of its anterior wall due to posterolateral arterial supply of the bile duct, and anatomic anomalies (such as cyst, diverticulum, and distal biliary obstruction).2,4 Diagnosis is usually confirmed with surgical exploration, and good prognosis depends on early surgical intervention.1,2 We report on a 10-day-old female neonate with spontaneous perforation and nearly total transection of the bile duct at the junction of the cystic duct and common hepatic duct, whose condition was managed by cholecystectomy and biliary reconstruction.

2. Case report

A 10-day-old girl was initially admitted to another hospital for phototherapy because of neonatal hyperbilirubinemia (serum total bilirubin: 15.3 mg/dL) at 2 days old. Bilious vomiting and fever up to 38°C developed 2 days later, and she was transferred to our hospital. Physical examinations revealed fair activity, mild dehydration and abdominal distension. The laboratory data reported white blood cell count as 8200/uL (band form: 5%), C-reactive protein 12.9 mg/dL, and bilirubin (total/direct) 11.5/0.8 mg/dL. Urine culture grew Escherichia coli, and the patient was admitted for medical treatment under suspicion of urinary tract infection with impending sepsis. However, progressive abdominal distension developed 6 days later. Abdominal sonography showed massive ascites, and paracentesis revealed yellow-greenish ascites. An emergency laparotomy found about 100 mL bilious ascites and a perforation, 0.4 cm in diameter, over the anterior wall at the bifurcation of the common hepatic junction, almost transecting the bile duct. Cholecystectomy, biliary reconstruction with Roux-en-Y hepaticojejunostomy, and drainage of the Morison’s pouch by a Penrose drainage tube were performed (Fig. 1). Oral intake was started on...
postoperative Day 4, and the patient was discharged on postoperative Day 10. Three years of postoperative follow-up indicated the patient had recovered well.

3. Discussion

Since Dijkstra reported the first case of spontaneous bile duct perforation in 1932, less than 170 cases have been reported in the literature. Although rare, spontaneous perforation of the bile duct is the third most common cause of surgical jaundice in infants, after biliary atresia and choledochal cyst. Unlike the aforementioned diseases, spontaneous perforation of the bile duct can deteriorate and cause death due to severe sepsis developing in a few days or even hours. Hence, it is very important to make a prompt diagnosis for optimal surgical management.

Clinical features, such as jaundice, abdominal distension, ascites, normal to acholic stool, dark urine, anorexia, vomiting, irritability, mild fever, failure to thrive, peritonitis, and shock, may occur in spontaneous perforation of the bile duct.

If there has been a healthy interval from birth to clinical presentation, the probability of spontaneous perforation of the bile duct is increased. However, clinical features and laboratory evaluations are not pathognomonic, and diagnosis is mostly confirmed by laparotomy.

Surgical methods include simple drainage with or without repair of perforation, cholecystectomy, external biliary diversion, or biliary reconstruction. Drainage often leads to spontaneous closure of a perforated bile duct, but surgical treatment should be individualized. Bile duct stenosis is the most common complication after simple drainage and may progress to biliary cirrhosis and portal hypertension. Intraoperative cholangiography is necessary to confirm the potential anatomical anomaly or distal obstruction. Single-stage cystectomy and Roux-en-Y reconstruction is possible in select patients with perforated choledochal cyst and subsequent bile peritonitis. A staged procedure with peritoneal lavage and T-tube drainage of the biliary tree followed by excision of the cyst and Roux-en-Y hepaticeojunostomy 3 months later may be applied to patients in poor clinical condition.

Perforation of the bile duct or choledochal cyst should be considered in every jaundiced neonate or infant with ascites. In our patient, correct diagnosis was achieved with surgical exploration, and a nearly total transaction of the bile duct was found. Therefore, one-stage operation with cholecystectomy and biliary reconstruction was performed although she was at high risk. Postoperatively, the patient recovered uneventfully with good outcome for 3 years. In conclusion, simple drainage or resection should depend on the patient’s clinical status and intraoperative findings.

References