

Spontaneous perforation of a choledochal cyst, clues for diagnosis

Vijai D. Upadhyaya^a, Basant Kumar^a, Rajat Gupta^a, Moniak S. Sharma^b, Richa Lal^a, Vibhor V. Borkar^b and Rohan Malik^b

Aim Spontaneous perforation is a rare complication of a choledochal cyst that is difficult to diagnose because of its nonspecific clinical presentation. These patients usually present with an insidious course characterized by progressive abdominal distension, vomiting, and pain. Here, we present a series of three cases for clues for diagnosis.

Materials and methods All patients with spontaneous perforation of a choledochal cyst who were presented to our institute from January 2009 to December 2011 were included in the study and symptoms and signs at the time of presentation were analyzed.

Results All patients had clinical jaundice and acholic stools at the time of presentation; pain and vomiting was not a consistent feature. Abdominal paracentesis helped us diagnose these cases.

Introduction

Spontaneous perforation of the choledochal malformations (CM) is a rare complication in children [1–3]; the reported incidence varies from 2 to 18% [2–4]. The diagnosis is often delayed because of its nonspecific presentation; hence, it is very rarely made preoperatively. The presentation of CM perforation may be acute or insidious [3,4], with the latter type being more common [4,5]. Simple peritoneal drainage with a T-tube is an option for the management of such cases in an emergency. Single-stage surgery in the form of excision of the choledochal cyst, cholecystectomy, and Roux-en-Y hepaticojejunostomy is an ideal option but should be attempted only if the patient presents early after perforation and expertise on hepatobiliary surgery is available. In the present series, we focused on the diagnosis and management of a perforated choledochal cyst.

Materials and methods

All patients with spontaneous perforation of a choledochal cyst who were presented to our institute from January 2009 to December 2011 were included in the study. Patients were resuscitated and routine investigation along with ultrasound of the abdomen was performed. Abdominal paracentesis was carried out when investigations and clinical examinations indicated a suspicious biliary peritonitis. After resuscitation and preliminary investigation, patients were explored. After a proper peritoneal lavage, the bile duct was examined for the site of perforation and a T-tube was inserted [after opening the back of the T-tube (size 8 Fr)] through the site of perforation and fixed with a purse-string suture, followed by placement of the abdominal drain in the right subhepatic area. T-tube drainage was replaced with ringer

Conclusion Abdominal paracentesis is very useful in cases of suspected spontaneous perforation of the bile duct. Simple drainage with a T-tube, if possible, is safe, efficacious, has low morbidity, and aids in the establishment of the diagnosis as well as delineation of the anatomy of the concerned region for definitive surgery. *Ann Pediatr Surg* 9:99–102 © 2013 Annals of Pediatric Surgery.

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Departments of ^aPediatric Surgery and ^bPediatric Gastroenterology, Sanjay Gandhi Post Graduate Institute of Medical Sciences (SGPGIMS), Lucknow, India

Correspondence to Vijai D. Upadhyaya, Master of Surgery, MCh, Department of Pediatric Surgery, Sanjay Gandhi Post Graduate Institute of Medical Sciences (SGPGIMS), Lucknow 226014, India
Tel: +91 522 249 05614; e-mail: upadhyayavj@rediffmail.com

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lactate solution initially; once the patients were on full oral feed, it was not replaced. Abdominal drains were removed when the drainage was nil and ultrasound had confirmed that there was no intraperitoneal collection, usually between the 8 and 10th postoperative days. T-tube cholangiogram/magnetic resonance cholangiopancreatography (MRCP) was performed to confirm the diagnosis and to delineate the anatomical details of the pancreaticobiliary region for definitive surgery. This study was approved by the ethical committee.

Results

Three patients were treated during this period, ranging in age between 2 and 10 years. At presentation, all patients had clinical hyperbilirubinemia and abdominal distension without signs of frank peritonitis (no fever, guarding, rigidity, rebound tenderness), although they had mild respiratory discomfort because of abdominal distension and tachycardia because of dehydration (Table 1). Preliminary investigations showed conjugated hyperbilirubinemia with near-normal transaminase, and elevated γ -glutamyl transpeptidase and alkaline phosphatase Table 2. Ultrasound abdomen showed nonvisualization of the bile duct with gross ascites with a contracted gall bladder. On exploration, all patients had gross generalized biliary collection, contracted gall bladder, dilated common bile duct (CBD), and perforation on the dilated CBD. Perforation was seen on the anterior wall of the common hepatic duct proximal to the junction of the cystic duct in one patient, and in the rest of the two patients, it was seen on the posteriolateral aspect of the common hepatic duct. All patients responded well to peritoneal lavage and abdominal drainage with a T-tube in the dilated bile duct; oral feed was started on postoperative day 3 and all patients were on full oral

Table 1 Patient profile and clinical features at the time of presentation

Patients	Age/sex	Abdominal distension	Clinical jaundice	Acholic stools	Dark-colored urine	Pain	Vomiting	Features of peritonitis	Duration between perforation and admission (days)	Follow-up after definitive repair (months)
1	2 year/M	Present	Present	Present	Present	Absent	Present	Absent	5	13
2 ^a	4 year/M	Present	Present	Present	Present	Absent	Present	Absent	5	8
3	8 year/F	Present	Present	Present	Absent	Present	Absent	Absent	8	6

^aOperated at another center by the author.

Table 2 Biochemical parameters

Patients	Serum bilirubin (mg/dl)	Conjugated serum bilirubin (mg/dl)	Fluid bilirubin (mg/dl)	ALT/AST (U/l)	GGT (U/l)	Serum amylase (U/l)	Fluid amylase (U/l)
1	5.8 (0.10–1.30)	3.2 (0.0–0.40)	18	38/16 (5–40)	272 (13–86)	38 (16–110)	54
2 ^a	8	4.2	–	26/19	347	46	51
3	5	3.1	14	32/21	286	97	260

Values in parentheses are normal reference values of our lab.

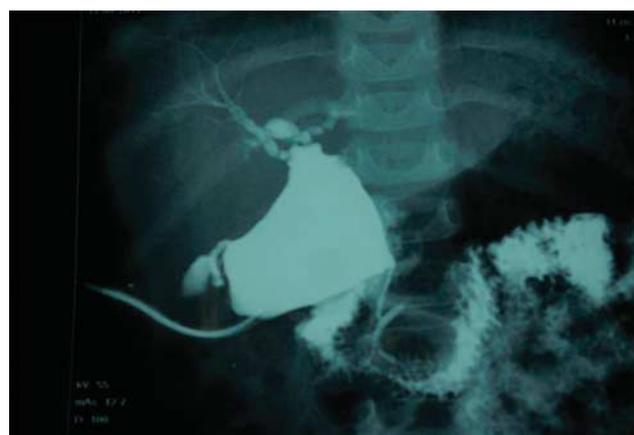
ALT/AST, aspartate aminotransferase/alanine aminotransferase; GGT, γ -glutamyl transpeptidase.

^aOperated at another center by the author.

feed from postoperative day 5. The T-tube drained around 500–700 ml for first 4–5 days, which was stabilized to 200–300 ml/day after 1 week. Cholangiogram showed cystic dilatation of the CBD (Fig. 1) and in one case MRCP was performed to delineate the anatomy (Fig. 2), which showed a type-1 choledochal cyst in all cases. Definitive surgery (Roux-en-Y hepaticojejunostomy after excision of the cyst) was performed after 6 weeks of emergency surgery. All patients are doing well at follow-up and have had no episode of cholangitis or abdominal pain after definitive treatment up to the last follow-up.

Discussion

Spontaneous perforation of the bile duct is a disease entity in which the extrahepatic duct or the intrahepatic duct is perforated spontaneously without traumatic or iatrogenic injury [6]. The exact cause of CM perforation still remains uncertain; several mechanisms have been postulated to explain the spontaneous rupture of CM. The various theories proposed include congenital weakness of the bile duct, pancreaticobiliary anomalies including anomalous union of the pancreaticobiliary such as duct anomalous union of the pancreaticobiliary duct [1,2], pancreatitis, and distal obstruction of the bile duct because of inspissated bile, stones or stenosis, the presence of a diverticulum or abnormal gland of the bile duct wall, viral infection of the bile duct, tuberculosis, necrotizing enterocolitis, and birth trauma. Irritation of immature CM wall from refluxed pancreatic juice [4] was considered as a cause for the perforation explained on the basis of the elevated serum amylase level in these patients, but one of our patients had higher serum amylase level (Table 2). Other authors consider obstruction of the distal CBD [5] along with congenital weakness of the cystic wall as the cause of spontaneous perforation of CM, but in our series, only one patient had a history of pain and none of our cases had sludge or inspissated bile in the distal bile duct on imaging. These findings

Fig. 1

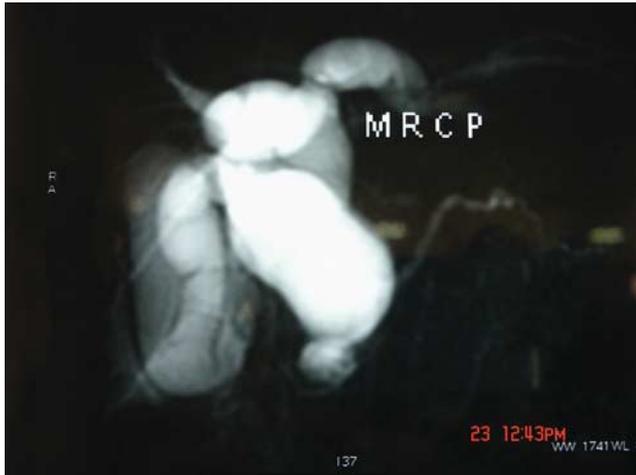
Post operative T tube cholangiogram showing dilated CBD (choledochal cyst).

suggest that reflux of pancreatic juice or distal obstruction is not the only cause for rupture of CM.

Spontaneous perforation of CM can present as acute abdomen secondary to biliary peritonitis [4,7] or as delayed biliary pseudocyst formation [8]. All patients in our series had insidious bile duct perforation, characterized by progressive jaundice and painless abdominal distension with no features of frank peritonitis. Clinical jaundice is not a frequent occurrence in these types of patients and is seen in around 38% cases [3,4] but all patients in our series had clinical jaundice at the time of presentation. None of our patients presented with features of frank peritonitis or pseudocyst formation as described by various authors.

Biochemical investigations indicated cholestatic jaundice with near-normal liver function tests, which differentiate spontaneous biliary peritonitis from biliary atresia and

Fig. 2



MRCP showing choledochal cyst.

neonatal hepatitis [9]. Ultrasound abdomen showed free fluid in the peritoneal cavity with a contracted gall bladder and nonvisualization of the CBD with normal intrahepatic biliary radicles; none of our patients had localized fluid collection around the porta or had a pseudocyst in contrast to previously published reports [9,10].

Computed tomography scan of the abdomen shows the same finding as ultrasound, but few authors have considered it as an important diagnostic tool for such cases [11]. MRCP represents a noninvasive method of evaluating the biliary tree and, owing to its excellent anatomical and contrast resolutions, MRCP can delineate bile duct perforation, but this was not done in our patients because of logistic constraints. Hepatobiliary scintigraphy with ^{99m}Tc iminodiacetic acid derivatives may be diagnostic in cases of bile duct perforation [10,12]. Abdominal paracentesis is useful in the diagnosis of bile duct perforation when ascitic bilirubin levels are several folds higher than the serum bilirubin level, although it may increase the risk of infection [13]. The presence of bile in the peritoneal cavity associated with obstructive jaundice in the absence of liver derangement is considered pathognomonic for bile duct perforation [14]. In the present series, for all patients who were presented as an emergency and on the basis of history (history of recent passage of pale stool), clinical evaluation (presence of jaundice, gross ascites), preliminary biochemical examination (liver function test showing conjugated hyperbilirubinemia, markedly elevated alkaline phosphatase and γ -glutamyl transpeptidase, with marginally elevated transaminases), and ultrasonography finding (contracted gall bladder, nonvisualization of the CBD with normal intrahepatic biliary radicles, and gross ascites with low-level internal echoes), bile duct perforation was suspected. In our experience, diagnostic paracentesis for suspected bile duct perforation is safe and the risk of fulminant infection is minimal as suggested by different authors [12,13]. Thus, proper assessment of history, clinical examination, and preliminary investigation will provide a clue for the diagnosis of this entity, which can be further

supported bedside by abdominal paracentesis. In developing countries such as India, because of limited resources and financial constraints, diagnostic paracentesis is one of the very useful tests for the diagnosis of this entity.

Surgical management of the biliary-tree perforation remains controversial, but the goals of such surgical management are to halt continuing abdominal contamination with infected bile by means of peritoneal drainage [14], that is, surgical closure of the perforation or T-tube drainage [10], and to treat the associated biliary pathologic features later or in the same sitting [7,10,11,15] by open or laparoscopic surgery [16]. The operation should be tailored according to the patient's general condition, severity of peritonitis, and exploratory findings. In the present series, we opted for two-stage surgery in all cases (except one, operated outside) because patients presented after 5 days of suspected perforation, had poor general and nutritional status, and intraoperatively, the wall of the cyst was extremely friable and inflamed. We placed a T-tube from the site of the perforation in all cases, as recommended by other authors [16–18]. Depending on the size of the perforation and the cyst, the T-tube was tailored and fixed with a purse-string suture and the abdomen was drained with a subhepatic drain. Single-stage definitive surgery as indicated by a few authors should only be attempted if the patient presents early enough after perforation, the general condition of the patient is good, and expertise on surgery is available.

Conclusion

Abdominal paracentesis is very useful in the diagnosis of suspected spontaneous perforation of the bile duct. Simple drainage with a T-tube, if possible, results in less morbidity and stabilizes the patient and helps in establishing the diagnosis. In developing countries such as India, because of limited resources and delayed presentation, laparotomy with a peritoneal lavage and abdominal drain, with or without T-tube drainage, is life saving.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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