A Predisposing Factor for Spontaneous Choledochal Cyst Perforation: Esophageal Dilatation Procedures

Koledok Kistinin Perforasyonu için Bir Predispozan Faktör: Özofagus Dilatasyon İşlemleri

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Abstract

Cyst rupture is one of the rarest complications of choledochal cysts (CC). We report an 8-year-old boy with CC rupture leading to bile peritonitis following repeated esophageal dilations for corrosive stricture, and discuss how the esophageal dilatation procedures might constitute a predisposing factor for CC perforation.

Keywords: Choledochal cyst, Perforation, Esophageal dilatation

Introduction

Cyst rupture with bile peritonitis is one of the rarest complications of (CC) [1]. Although the etiology of this complication remains unknown in many cases, precipitating factors such as blunt abdominal trauma, pregnancy, and anatomical abnormality have been postulated [2]. We report a case of CC rupture with bile peritonitis following repeated esophageal dilatations, and discuss how the esophageal dilatation procedures might constitute a predisposing factor for CC perforation.

Case Report

An 8-year-old boy presented to the pediatric surgery department of our hospital with a three-day history of nausea, vomiting, and abdominal pain. A medical history revealed that he had undergone 11 esophageal dilatation procedures in the last year (five times with balloon dilators following six times with push dilators) for esophageal strictures related to ingestion of a caustic substance. Balloon and push dilators had been used under midazolam sedation and general anesthesia, respectively.
The last dilatation procedure had been performed four months prior to presentation. At that time, no history of previous abdominal pain, fever, jaundice, or trauma was noted. On physical examination, the patient did not have a fever. However, he had rebound tenderness in the right lower quadrant and abdominal guarding without rigidity, but no icteric sclera.

Laboratory investigations revealed a hemoglobin level of 13.3 g/dL, a white blood cell count of 17,000/mL, an amylase level of 163 U/L, and an alkaline phosphatase level of 367 U/dL with normal levels of serum transaminases. Plain abdominal radiography findings were normal. Abdominal ultrasonography (USG) revealed minimal ascites in all the quadrants of the abdomen. The patient was subjected to laparotomy through a right paramedian incision because the provisional diagnosis was acute perforated appendicitis. Approximately 200 mL of turbid and bilious peritoneal fluid was aspirated. However, the biliary tree was not explored. Intraoperatively, the appendix had an edematous appearance, but it was not perforated. Histopathologic examination of the appendix was normal. Postoperatively, the complaints and symptoms of the patient gradually intensified.

The patient developed fever and his ascites increased markedly. The patient was then referred to our pediatrics department for further evaluation. Physical examination revealed a markedly distended and diffusely tender abdomen with fluctuation, and 3 cm of hepatomegaly. His scleras were icteric. Laboratory investigation results, including complete blood count, blood glucose, electrolytes, serum glutamic-oxaloacetic transaminase, serum glutamic-pyruvic transaminase, renal function tests, and urine analysis, were within normal limits. Serum total bilirubin, direct bilirubin and amylase levels were 5.2 mg/dL, 2.1 mg/dL, and 308 U/L, respectively. Ascites fluid obtained via paracentesis was turbid and bilious. A biochemical analysis showed a total bilirubin level of 38.7 mg/dL, direct bilirubin level of 14.3 mg/dL, and amylase level of 2004 U/L in the ascites. A hepatobiliary iminodiacetic acid scan revealed an enlarged common bile duct and free extravasation of bile into the peritoneal cavity (Figure 1a and 1b).

An abdominal computed tomography (CT) scan showed a normal-sized gallbladder, distension of the common bile duct, and ascites (Figure 2). The patient was diagnosed with a rupture of the extrahepatic biliary tract and underwent emergent laparotomy. Surgical exploration showed a large amount of intraperitoneal bile and biliary peritonitis. The ductus choledochus was dilated fusiformly. A type 1 choledochal cyst with a perforation on the posterior wall of the cyst was identified. Cholecystectomy, total cyst excision, and Roux-en-Y hepaticojejunostomy were performed. The postoperative recovery was uneventful, and the patient was discharged on the 18th day of hospitalization. He was still well 6 months later.

**Discussion**

Choledochal cysts are generally considered congenital because they occur in fetuses and neonates. In contrast to adults, the classic presentation of a CC with the triad of jaundice, abdominal pain, and a right hypochondrial mass is rare in children, and is seen in only 15% to 25% of cases at the initial visit [2]. Our patient had never had such a triad.

In three reviews of 1433, 188 and 11 patients with CC, the incidence of spontaneous rupture of a CC leading to biliary peritonitis has been reported to occur at rates of 1.8%, 2.1%, and 18%, respectively [1]. The etiology of this compli-
cation remains obscure in many cases. The initial presentation with perforation and small size of the cyst make it difficult to determine the cause of rupture of CC. However, precipitating factors, such as blunt abdominal trauma, pregnancy and labor, and an additional anatomical abnormality have been postulated to lead to spontaneous rupture of CCs. Mechanisms such as distal obstruction of the common bile duct with protein plugs, congenital weakness and anoxic necrosis of the cyst wall, and irritation of the mucosa of the bile duct by reflux of pancreatic enzymes into the biliary tree have been proposed [2,3].

Our patient had undergone 11 procedures of esophageal dilatation (5 with balloon dilators following 6 with push dilators) in the last year. Balloon catheter dilatation is reported to be a safe and effective procedure for the treatment of esophageal strictures in the pediatric population [4]. This procedure can be performed under conscious sedation [5] or under general anesthesia [4]. All of the balloon catheter dilations in our patient were performed under conscious sedation with midazolam. This drug decreases events such as retching, the gag reflex and salivation during the dilatation procedure, but it cannot completely prevent them [6]. The gag reflex is mainly related to the increased intra-abdominal pressure (IIAP), which may have caused reflux of pancreatic fluid into the biliary tract in our patient.

In addition, an in vitro study investigating the acute effects of IIAP mimicked by a laparoscopic intervention showed that pneumoperitoneum-induced IIAP causes structural alterations which may be related to ischemia-reperfusion injury in rat terminal ileum [7]. Another study has reported that the alteration in the intraabdominal pressure may affect oxygenation of the rabbit colon [8]. In the light of these explanations, IIAP during balloon dilatations in our case may be said to have caused perfusion abnormalities in the cystic wall, which is especially vulnerable to ischemia. This is because type 1 CCs, such as the one observed in the present case, is thought to form as a result of a suboptimal blood supply [3].

Subsequent esophageal dilatation procedures in our patient were performed under general anesthesia, which cannot lead to IIAP. Findings from animal studies suggest that general anesthetics can affect abdominal hemodynamics and tissue oxygenation. Desflurane is shown to impair hepatic and small intestinal O2 reserve capacity [9]. The mean surface PO2 of the liver and small intestine is reported to decrease by 20% during isoflurane and enflurane anesthesia [10]. Furthermore, mesenteric artery blood flow and mesenteric oxygen delivery are decreased by xenon administration. Halothane has been shown to cause a marked decrease in mesenteric blood flow and mean hepatic surface PO2, but does not cause impairment of the oxygenation of the gut mucosa [10].

These adverse effects do not mean that general anesthetics cause severe hypoxia in normal tissues, but it suggests that they might lead to failure in the oxygenation of tissues such as type 1 CC, which have suboptimal blood supplies [3]. Most perforations reported in the literature, as in the present case, involve the anterior or posterior wall of the choledochal cyst, suggesting such perforations are an ischemia-related event. CC perforation, in our case, occurred four months after the last esophageal dilatation. Whether esophageal dilatations are solely responsible for perforation is questionable, but they are more likely to be a predisposing factor. In our patient, repeated esophageal procedures might have weakened the cystic wall secondary to ischemia and failed oxygenation, thus facilitating cyst rupture.

Although it has been reported that a high-resolution abdominal USG is the best screening test for the diagnosis of pediatric CC, we did not perform this test. USG and CT are both helpful in showing cystic masses in relation to the pancreatic head and porta hepatitis. However, CT is considered to be more accurate in demonstrating the status of the distal part of the common bile duct, which may be obscured by bowel gas on USG [10]. The bowel preparation for USG could not be performed in this case because our patient was in an emergency state. For this reason, abdominal USG would not have allowed for identification of the CC in our case.

In the first operation, although bilious peritoneal fluid had been aspirated, the biliary tree was not evaluated. In addition to the bile-like fluid from the paracentesis, histopathologically-normal appendix suggested bile duct pathology and prompted us to evaluate the biliary tree carefully. Therefore, in the presence of bilious peritoneal fluid, examination of the biliary tract should be mandatory to avoid the use of additional diagnostic approaches and secondary operations, as occurred in the present case.

In conclusion, esophageal dilatation procedures may be a predisposing factor for CC perforation. Any fluid suggestive of bile in the peritoneal cavity should alert the physicians to the possibility of perforation of the bile duct and rupture of a CC. Bilirubin levels in ascites fluid should be measured to assess this possibility, and the biliary tree should also be evaluated.
Conflict interest statement The authors declare that they have no conflict of interest to the publication of this article.

References