Multiple stone formation in a remnant choledochal cyst

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The recommended surgical method for treating congenital biliary dilation is excision of the entire extrahepatic bile duct, with a hepaticoenterostomy to stop reflux and stasis of pancreatic secretions in the gallbladder and dilated bile duct. Late complications of surgical treatment include anastomotic strictures, cholangitis, and stone formation in the intrahepatic bile ducts. This report describes a very rare late complication, in which protein stones formed in a remnant choledochal cyst in the pancreas 23 years after resection of a congenital choledochal cyst. Our case highlights the necessity for complete cyst resection as close as possible to the portion buried in the pancreatic bed at the time of primary choledochal cyst resection. (Korean J Med 77:227-231, 2009)

Key Words: Choledochal cyst; Remnant; Late complication

INTRODUCTION

A choledochal cyst is a congenital disorder characterized by a dilated bile duct. Babbitt et al. proposed in 1969 that choledochal cyst might be closely associated with anomalous pancreaticobiliary ductal junction (APBDJ) which would allow pancreatic juice to reflux freely into the common bile duct, producing cyst formation and recurrent bouts of cholangitis. Such a chemical stimuli and inflammation is presumed to be cause of high association between congenital biliary dilation and hepatobiliary malignancies. Therefore, for the treatment of congenital biliary dilation, the excision of the entire extrahepatic bile duct with a hepaticoenterostomy is the recommended surgical method to stop reflux and stasis of pancreatic secretions in the gallbladder and the dilated bile duct. However, late complications during long-term follow-up after surgical treatment have been reported, such as anastomotic strictures, cholangitis, and stone formation in the intrahepatic bile ducts. In addition, various post-operative complications related to the pancreas, including pancreatitis, pancreatic stones, and cancer have been reported, even after primary cyst excision.

We report a very rare late complicated case of multiple stone formation within the intrapancreatic portion of a remnant bile duct, which occurred 23 years after primary excision of an ex-
Figure 1. (A) Preoperative T2-weighted magnetic resonance imaging shows a 4×3 cm remnant choledochal cyst containing multiple stones and air bubbles. A choledochojejunostomy loop is seen below the liver. (B) Magnetic resonance cholangiopancreatography (MRCP) shows cystic pooling with filling defects. (C) Endoscopic retrograde cholangiopancreatography (ERCP) shows that the pancreatic duct and remnant choledochal cyst share a common opening. A dilated remnant choledochal cyst is observed. (D) The moment the catheter was inserted, a large amount of white fluid was released from the ampulla of Vater.

CASE REPORT

A 55-year-old woman was admitted to the Catholic University Kangnam St. Mary Hospital because of epigastric pain for 4 days. Twenty-three years previously, she had undergone excision of a choledochal cyst with reconstruction by Roux-en-Y hepaticojejunostomy as treatment for Todani’s type Ia congenital biliary dilation, which was confined to the extrahepatic bile duct. She had been in her usual state of good health with no symptoms until 23 years after surgery, when she developed upper abdominal pain.

Physical examination on admission revealed moderate tenderness in the epigastrium without other abnormalities. With respect to laboratory data, amylase and lipase were elevated to 153.2 U/L (normal, 40–130 U/L) and 434.4 U/L (normal, 7–50 U/L), respectively. Other laboratory data were within normal limits, as follows: white blood cell count, 5,520/mm³; hemoglobin, 13.1 g/dL; hematocrit, 37.9%; platelets, 243×10⁹/L; total protein, 6.2 g/dL; albumin, 3.5 g/dL; AST, 22 U/L; ALT, 10 U/L; alkaline phosphatase, 60 U/L; lactate dehydrogenase, 310 U/L;
total bilirubin, 0.8 mg/dL; cholesterol, 186 mg/dL; triglycerides, 86 mg/dL; and glucose, 107 mg/dL.

Abdominal computed tomography (CT) revealed a cystic lesion in the pancreas head measuring 4×3 cm in size. Air-fluid levels were seen, and multiple small air density lesions were observed within the fluid. Magnetic resonance imaging (MRI) demonstrated a 4×3 cm cystic lesion containing sludge and multiple stones in the pancreatic head. Following gadolinium infusion, thin cyst wall enhancement was seen in this cystic lesion, suggesting an inflammatory condition of the cyst (Fig. 1A). On magnetic resonance cholangiopancreatography (MRCP), the pancreatic duct was unremarkable and the remnant cyst arose from a common channel to the distal portion of the pancreatic duct (Fig. 1B). On endoscopic retrograde pancreatography (ERCP), a definite communication between the remnant choledochal cyst and the main pancreatic duct was not seen; but they crossed with an obtuse angle in the distal portion of the main pancreatic duct and formed a long common channel (Fig. 1C). The main pancreatic duct was not dilated and did not have any stones or plugs, although the expanded choledochal cyst compressed the pancreatic duct in the head portion. A disturbance in excretion of the pancreatic enzymes may cause pancreatitis. When the catheter was inserted into the remnant choledochal cyst, large amounts of whitish fluid and protein plugs were released (Fig. 1D). For temporary decompression, the endoscopic retrograde biliary drainage catheter was inserted into the remnant cyst.

The patient underwent laparotomy under general anesthesia for excision of the remnant choledochal cyst. The pancreatic head was exposed by blunt and sharp dissection of adhesions due to the previous surgical procedures. Intraoperative ultrasonography identified strong echoes with acoustic shadows in the pancreatic head. The cyst was carefully dissected from the pancreatic parenchyma. Infusion of contrast medium from the cyst revealed a communication between the cyst and the main pancreatic duct. The remnant cyst was then resected near the main pancreatic duct. The excised cyst contained numerous stones (several millimeters in diameter), gravel, and sludge. The stone had a milk-white color, which is typically found in proteinaceous stones.

Histologic examination of the cyst revealed a fibrous, thickened bile duct wall with mild chronic inflammation, congestion, and hemorrhage, accompanied by slightly hyperplastic glands. No dysplastic changes were noted in the remnant ductal wall. By post-operative day 8, amylase and lipase had normalized. The follow-up CT scan at the same time revealed a marked fluid collection in the peri-pancreatic and hepatic areas. Percutaneous tube drainage was performed. The patient had received conservative care for 2 months in the outpatient department. She had an uneventful course and was free of symptoms.

DISCUSSION

We have reported a rare case of multiple stone formation within the intrapancreatic portion of a remnant bile duct, which occurred 23 years after primary excision of an extrahepatic choledochal cyst. Why the remnant choledochal cyst was complicated by marked dilatation and stone formation after very long time from primary surgery is unknown, but we proposed possible consecutive insults as the basis. The first insult was the incomplete resection of the cyst during the primary operation. As the pancreatic duct lies close to the intrapancreatic cyst wall, there is always a risk of injury to the pancreatic duct when removing the bottom of the cyst buried in the pancreatic parenchyma during surgery. Her abdominal CT scan data performed at another hospital for a urologic problem 2 years earlier showed an abnormal pancreas finding with a 2 cm cyst in the pancreatic head. This suggested a remnant choledochal cyst without inflammation or stones. However, this is insufficient alone to explain why an uncomplicated remnant cyst became markedly dilated and contained multiple stones. We think the second insult may have been protein plugs or pancreatic ductal calculi which can play a role in persistent choledochal dilatation. Finally, the etiology of pancreatic protein plug formation remains uncertain. Some investigators have suggested an increased viscosity of pancreatic secretions due to elevated protein concentrations, leading to protein plug formation and a temporary blockage of the pancreatic duct by a protein plug, a pancreatic calculus, or dysfunction of the sphincter of Oddi. The obstruction of the pancreatic output may cause dilatation of the choledochal remnant.

After all, a pitfall of choledochal cyst excision is the remain-
ing intrapancreatic portion of the dilated bile duct. Some authors recommend partial cyst excision leaving behind the intrapancreatic portion of the cyst to avoid pancreatic injury.\(^\text{16-18}\) Others have proposed complete excision of the cyst, including the portion buried in the pancreas.\(^\text{19}\) They suggest that the narrow segment connecting the cyst to the pancreatic duct must be exposed by dissection of the outer plane of the cyst wall.\(^\text{19}\) However, it is controversial how much of the cyst buried in the pancreas should be resected. Recently, Miyano and colleagues\(^\text{20}\) suggested intraoperative endoscopy may be useful for deciding the level of adequate excision of the choledochal cyst in the pancreas. In addition, we think that confirmation of a complete cyst excision buried in the pancreas using intraoperative cholangiography is easier and a more convenient method, if available. In the present patient, in whom adhesions and scarring were formed by the previous surgery, intraoperative ultrasonography was useful for detecting the cystic lesion in the pancreas and intraoperative cystography was beneficial for preventing injury to the main pancreatic duct. Alternatively, preoperative endoscopic placement of a nasopancreatic tube in the main pancreatic duct and intraoperative pancreateography and/or a leak test with use of a dye solution during manipulation of the cystic lesion may be helpful.

In this report, we have described a very rare late complication, in which protein stones formed in a remnant choledochal cyst in the pancreas 23 years after resection of a congenital choledochal cyst. Our case highlights the necessity of complete cyst resection close to the pancreatic duct. It should be emphasized again that the cyst should be excised as far as possible at the time of primary choledochal cyst resection, and potential injury to the main pancreatic duct should be avoided. In a case in which it is impossible to remove the intrapancreatic choledochal cyst at primary surgery, careful long-term surveillance is encouraged with regular pancreatic ultrasonography or magnetic resonance cholangiopancreatography.

**REFERENCES**


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