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Acute pancreatitis caused by a duodenal duplication cyst covering the ampulla of Vater

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ABSTRACT

Duodenal duplication cyst is not a common congenital anomaly and the pathophysiology may become very complicated if the cyst is situated at the ampulla of Vater. Here we report a very rare female case of duodenal duplication cyst at the ampulla of Vater, which caused acute pancreatitis due to massive protein plaques in the pancreatic duct. She had a past history of double duodenal atresia and underwent surgery as a neonate. The correct diagnosis could not be determined before the second operation at four years of age and the exact pathophysiology finally became apparent during the operation with a contrast medium study and duodenotomy. We discuss the complicated clinical features and diagnostic and treatment procedures before and during the operation.

1. Introduction

Duodenal duplication is a rare type of intestinal duplication [1], and in some cases duodenal duplication situated at the ampulla of Vater causes severe pancreatitis or cholangitis [2].

We report here the very rare female case who suffered from acute pancreatitis at four years of age, which was caused by a massive protein plug in the pancreatic duct. She had a past history of double duodenal atresia. The acute pancreatitis was caused by the accumulation of protein plug in the dilated pancreatic duct. The mechanism of protein plug formation may be explained as follows: a mixture of bile juice and pancreatic juice accumulated in the duplication cyst that was situated at the ampulla of Vater, and the mixture regurgitated into the pancreatic duct and caused plug formation. We discuss the complicated pathophysiology of this case and review similar cases reported in the literature.

2. Case report

The patient was a four-year-old female. She suffered from an abdominal cyst that had been detected on fetal ultrasound examination and magnetic resonance imaging [3]. After birth, abdominal computed tomography and contrast study revealed double duodenal atresia at the first portion and at the third portion of the duodenum, respectively, and it was suspected that a Y-shaped bile duct drained into the first portion and the second portion of the duodenum. She underwent surgery as a neonate and the duodenal passage was reconstructed by side-to-side anastomosis for the first obstruction and by resection of the atretic membrane for the second obstruction. The intrahepatic and extrahepatic bile ducts were moderately dilated but the presence of an anomalous bile duct tract (i.e., bifid common bile duct) that had been suspected before the operation was not ascertained during the operation. Since then, the patient has been followed at the outpatient clinic with periodic ultrasound examinations.

At one year of age, an ovoid-shaped small defect was detected in the dilated duct at the head of the pancreas by ultrasound examination and it was suspected to be a protein plug in the dilated extrahepatic bile duct (Fig. 1a). At four years old, she complained of sudden onset of abdominal pain and a blood test showed elevated levels of AST (137 IU/l), ALT (46 IU/l), γ-GTP (54 IU/l), and amylase (414 IU/l). Abdominal ultrasound examination showed worsening of dilatation of both the bile duct and pancreatic duct, which suggested obstruction of bile and pancreatic juice flow. Conservative treatment was administered, and the symptoms and signs of pancreatitis subsided. However, after restarting oral intake of foods, she complained of abdominal pain again and her clinical course suggested the need for detailed investigation. Magnetic resonance cholangiopancreatography (MRCP) did not show the presence of pancreaticobiliary mal-junction because the main parts of the dilated biliary tree and pancreatic duct were not depicted perhaps due to protein plug accumulation in them. This accumulation of

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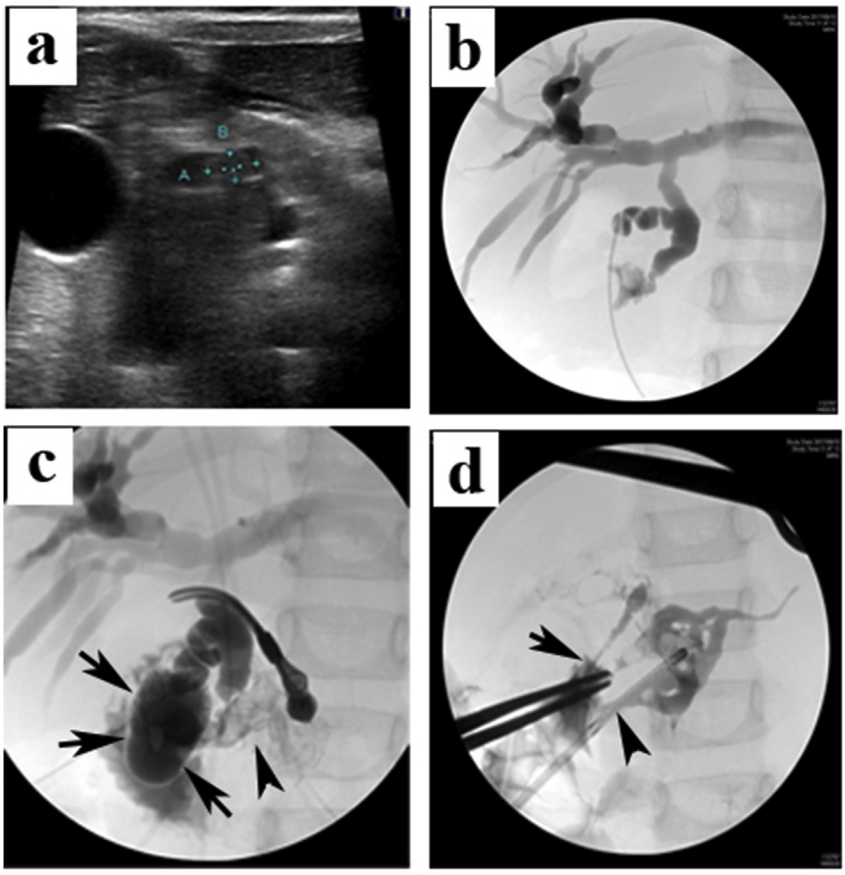

Fig. 1. (a) An abdominal ultrasound image taken at 1 year old. At the pancreatic head region, a dilated tract was ascertained and a small, slightly high echogenic mass was detected. We noticed it as a protein plug that had formed in the common bile duct at that time. (b) Intraoperative cholangiography at 4 years of age showed a slightly dilated biliary tree, but Y-shaped double common bile ducts were not shown. The pancreatic duct was not depicted, which suggested that the patient did not suffer from mal-junction of the pancreaticobiliary tree. (c) After clamping the hepatic duct, the lower biliary tree was selectively examined by a contrast medium study. In this study, a cystic lesion was first seen in the duodenum (arrows), and then a dilated pancreatic duct (Wirsung duct) was seen (arrowhead). In the pancreatic duct, many defects were ascertained, and they were later identified as many protein plugs. (d) After unroofing the duodenal duplication cyst and washing out protein plugs in the dilated pancreatic duct, a contrast medium study was again performed via the common bile duct and pancreatic duct. The main pancreatic duct and common bile duct joined at the ampulla of Vater (arrow), and the Santorini duct was also revealed (arrowhead). The distal pancreatic duct was not dilated.

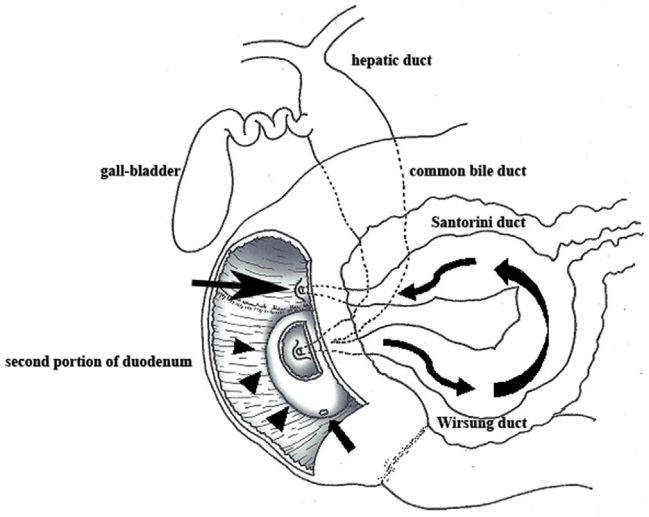

Fig. 2. Schematic view of the architecture of the pancreatic duct and biliary tree, and the duodenal duplication cyst. The duplication cyst (arrowheads) completely covered the ampulla of Vater, and a pin-hole (small arrow) existed at the caudal part of the cyst which worked as a drainage route. The pancreatic duct at the head of the pancreas was dilated, although the pancreatic duct at the pancreatic body and tail was not dilated. The Santorini duct (large arrow) was also dilated. The etiology of plug formation in these dilated pancreatic ducts is as follows: (1) Both bile juice and pancreatic juice accumulated in the duplication cyst. (2) Then, the mixture of bile juice and pancreatic juice regurgitated into the main pancreatic duct because of intra-cystic high pressure due to the small outlet. (3) Many protein plugs gradually formed in the main pancreatic duct and the protein plugs extended into the Santorini duct over four year (curved arrows).

protein plug was considered to be the cause of pancreatitis and cholangitis but the exact etiology of the protein plug formation was not apparent despite several imaging examinations. We finally decided to perform a contrast medium study to reveal the etiology of the obstruction in the pancreatic duct and/or biliary tree under laparotomy. During the surgery, we first performed the contrast medium study via the gallbladder. The common bile duct was a single tract and was mildly dilated, and a protein plug was not detected in the extrahepatic bile duct (Fig. 1b). The pancreatic duct was not depicted in this study and mal-junction of the pancreaticobiliary tree was ruled out (Fig. 1b). We noticed that when the hepatic duct was clamped, a cystic lesion was depicted in the duodenum. The lower biliary tree was selectively examined by contrast medium (Fig. 1c), and the cystic lesion was suspected to be a duodenal duplication cyst. In this study, after detecting the cyst, a prominently dilated pancreatic duct gradually became apparent, and the dilation seemed to be caused by regurgitation of fluid from the duplication cyst (Fig. 1c). In order to ascertain the cyst in the duodenum, duodenotomy was performed and the cystic lesion was ascertained at the second portion of the duodenum. A pin-hole existed at the caudal side of the cyst and nearly all of the protruding cyst wall was resected to open its space. After unroofing the cyst, the ampulla of Vater was recognized at the true duodenal wall and some protein plugs were scattered around it. On intraoperative pathology consultation, microscopic examination of frozen sections of the resected wall of the cyst revealed that the cyst wall was composed of duodenal wall. Therefore, the cyst was defined as a duodenal duplication cyst and not a choledochocele. We then recognized another orifice at the cranial side of the duplication cyst, which was the orifice of the Santorini duct. From this orifice, many protein plaques came out into the duodenal lumen and we washed the pancreatic duct to remove these protein plugs. After removing almost all plugs, a contrast medium study was attempted to reveal the entire pancreatic duct (Fig. 1d). The pancreatic duct at the