Enteric duplication cyst of the pancreas with duplicated pancreatic duct

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Abstract (159 words)

Enteric duplication cyst is one of the rarest forms of cystic lesion of the pancreas. We report a unique case of an enteric duplication cyst of the pancreas, which was communicating with a duplicated pancreatic duct. A 7-year-old girl with severe acute abdominal pain was found to have a large cyst which was smoothly communicating with the dilated pancreatic duct in the pancreatic tail. Analysis of cyst fluid showed elevated levels of amylase, CEA and CA19-9 and no epithelial cells. Intraoperative cyst-pancreatography revealed the pancreatic duct was duplicated in the tail: one duct was communicating with the cyst, and the other was dilated within the pancreatic tail. The patient underwent spleen- preserving distal pancreatectomy and complete cyst excision without complication. As preoperative diagnosis of duplication cyst of the pancreas is difficult, this condition should be considered during differential diagnosis of atypical cystic lesions of the pancreas. Complete excision is desirable for the management of duplication cyst of the pancreas.

Key Words: Duplication cyst of the pancreas, pancreatic duct, cyst fluid analysis

The majority of pancreatic cysts in children are pseudocysts caused by trauma, acute pancreatitis or infection. True pancreatic cysts, both with or without systemic diseases such as von Hippel-Lindau disease or polycystic kidney disease, are rare. Among these, one of the rarest forms of pancreatic cyst is enteric duplication cyst of the pancreas[1]. The most common sites for enteric duplication cysts are the jejunum and ileum, followed by the mediastinum, colon and stomach[2]. The pancreas is one of the rarest sites for dupliction[3]. Herein, we report a large enteric duplication cyst of the pancreas that communicated with one end of the duplicated pancreatic duct in the pancreatic tail.

1. Case Report

A 7 year-old girl was admitted to a regional general hospital with increasingly severe abdominal pain after milder symptoms for a week. She was transferred to our hospital with a diagnosis of ‘pancreatic cyst’, which was found by a CT scan. She had no history of recurrent abdominal pain, abdominal trauma, or pancreatitis. Laboratory data showed elevated levels of serum amylase (2369 IU/L), lipase (3450 U/L), and C-reactive protein (4.54 mg/dL). The abdominal CT scan (Fig.1A, 1B) showed that a huge cystic lesion was located in the left side of her abdominal cavity, the cyst was communicating with the dilated pancreatic duct in the pancreatic tail, and a small amount of cyst fluid had leaked into the abdominal cavity. The cyst wall was smooth and thin, with some calcification, and without any solid component in the cyst.

Her abdominal pain was cured with conservative therapy in a few days. Magnetic resonance cholangiopancreatography (MRCP) revealed a dilated pancreatic duct in the pancreatic tail with large cystic lesion. Endoscopic retrograde cholangiopancreatography (ERCP, Fig.1C) showed a displaced, but non dilated pancreatic duct in the pancreatic head and an apparently dilated distal pancreatic duct, which was communicating with the cyst. No abnormalities such as pancreatic divisum, heterotopic pancreas or duodenal duplication were found in the head and uncinate process of the pancreas and duodenum by preoperative imaging studies. About 150 ml of serous, clear cyst fluid was obtained percutaneously and cyst fluid analysis revealed elevated level of amylase (199000 IU/L), lipase (450000U/L), CA 19-9 (1556 U/mL), CEA (69.6 ng/mL) and CA125 (2090 U/mL). Cytological analysis of the cyst fluid showed no epithelial cells. The cyst was enlarged to the same size observed before aspiration within a few days.

The patient was operated upon with the diagnosis of ‘pancreatic cyst’. A large, smooth cyst, which was communicating with the pancreatic tail, was found in her left retroperitoneum (Fig. 2A). The cyst was separated easily from the descending colon and the retroperitoneum without any tight adhesion. The pancreatic tail was drained by an anomalous large vein running into the splenic vein, without any small connecting vessels between the pancreatic tail and splenic vein (Fig. 2A). The gross appearance of the cut end of the cyst resembled that of an intestine (Fig. 2B). After removing the extra-pancreatic portion of the cyst,

cyst-pancreatography from the cut end of the cyst (Fig.3) revealed that the pancreatic duct was duplicated in the pancreatic tail, and that the other end of the duct was also dilated within the pancreatic tail. As we concerned about the possibility of a cyst recurrence and malignant disease, we decided to perform distal pancreatectomy without splenectomy to remove both the cyst and all dilated pancreatic ducts. Histological examination showed that the cyst lumen was covered with stratified squamous and ciliated columnar epithelium with a longitudinal and circular muscle layer (Fig. 4). Based on these findings, this lesion was diagnosed as an enteric duplication cyst of the pancreas. The patient's postoperative recovery was uneventful, and she experienced no abdominal pain after the operation.

2. Discussion

Duplication cysts of the pancreas present with various symptoms such as abdominal pain, nausea and vomiting, and a palpable mass[1]. Pancreatitis and the mass effect are considered as common etiologies of abdominal pain by these lesions [1, 3]. In addition to these mechanisms, fluid leakage or rupture of the cyst can cause severe abdominal pain, as in our case.

Differential diagnosis of pancreatic cysts includes pseudocyst, various true cysts and cystic neoplasm of the pancreas. Several studies indicate that cyst fluid analysis helps to differentiate between these lesions. Van der Waaij *et al.* reported that an elevated level of

CEA (>800 ng/mL) suggests mucinous cystic neoplasms, low CEA (<5 ng/mL) or CA 19-9 (<37 U/mL) suggests serous cystadenoma or pseudocyst, and that an elevated level of amylase can be observed in both cystic neoplasms and pseudocysts[4]. According to their criteria, relatively high values of CEA and CA19-9, and the highly elevated amylase level in our case, are not indicative of either cystic neoplasm or pseudocyst. There are two reports of cyst fluid analysis of duplication cysts involving the pancreas[5, 6]. Levels of CEA and amylase were increased in both of these reports, as in our case. Thus, elevated CEA and amylase in the cyst fluid would represent characteristics of a duplication cyst of the pancreas. The viscosity of the cyst fluid is significant information for differential diagnosis of cystic lesion of the pancreas. Although preoperative cyst fluid was not viscous in our case, it is common to find that cyst fluid is mucinous or viscous in the duplication cyst[3, 5, 7, 8], which is secreted by mucosa on the cyst lumen. As tumor markers such as CEA, CA 19-9 and CA125 and the vicosity of the cyst fluid are indicative of epithelial lesions including mucinous cystic neoplasms, intraductal papillary tumors and enteric duplication cysts, these features of the cyst fluid in duplication cyst of the pancreas, comprising elevated levels of amylase and CEA, and the viscosity, suggested the possible misdiagnosis of the duplication cyst as a cystic neoplasm.

There are several case reports of enteric duplication cysts communicating with the pancreatic duct[7, 9-12]. Most of these cases are located on the head of the pancreas, and

possess a gastric type mucosa in the cyst. One explanation for the connection between the duplication cyst and pancreatic duct is due to the result of inflammation and ulceration within the cyst that may perforate into a duct of the adjacent pancreas creating an internal fistula[13]. In our case, however, the cyst was smoothly communicating with the dilated pancreatic duct. This morphological continuity strongly suggests that pancreas, or pancreatic duct, is the origin of the duplication cyst in our case. Our case had other abnormalities associated with the pancreas; duplicated distal pancreatic ducts in the pancreatic tail, and an anomalous single large vein in the pancreatic tail without small drainage veins running into the splenic vein. As the head and uncinate process of the pancreas was normal in this case, developmental malformation in the dorsal pancreas would explain the abnormality of our case including duplication cyst.

Although an enteric duplication cyst of the pancreas is quite a rare lesion, this condition should be considered during differential diagnosis of cystic lesion of the pancreas. Radiological examination involving a CT scan, MRCP and ERCP coupled with cytological and chemical cyst fluid analysis provides useful information for preoperative diagnosis. A consideration only of elevated CEA and viscosity of the cyst fluid might result in a misdiagnosis of these lesions as a cystic neoplasm of the pancreas. A definitive diagnosis is usually made on the basis of histological examination after surgical resection. A complete resection, if possible, would therefore be an appropriate option for the treatment of enteric

duplication cyst of the pancreas because of the potential complication and difficulty associated with preoperative differentiation from cystic neoplasm. In case the preoperative workup confirms a duplication cyst without any diagnostic ambiguity, laparoscopic surgery could be considered in the management.

Figure legends

Fig. 1. Preoperative imaging studies:

(A) Abdominal CT; Transverse section: A large cyst (C) existed at the tail of the pancreas. A dilated pancreatic duct (black arrow) was detected in the pancreatic tail.

(B) Abdominal CT; Coronal section: The cyst was smoothly communicating with the dilated pancreatic duct (black arrow). The cyst wall was thin and smooth, and a solid component was not found. A small amount of cyst fluid had leaked from the cyst (white arrow).

(C) **Endoscopic retrograde cholangiopancreatography (ERCP)**: ERCP showed a displaced, but not dilated pancreatic duct in the pancreatic head and an apparently dilated distal pancreatic duct, which was communicating with the large cystic lesion indicated by dotted line (cyst).

Fig. 2. Intraoperative photograph.

A: The outer surface of the cyst (C) was smooth, and easily separated from the surrounding retroperitoneal tissue. The cyst originated from the pancreatic tail (P). The pancreatic tail was drained by a large anomalous vein (white arrow) and normal small drainage veins running into the splenic vein were not found.

B: The cut end of the extra-pancreatic portion of the cyst (white arrow) resembled the cut end of an intestine. (P: pancreatic tail)

Fig. 3. Intraoperative cyst-pancreatography.

After resecting the extra-pancreatic portion of the cyst, cyst-pancreatography was performed from the cut end. Dotted lines indicate the resected part of the cyst. The pancreatic duct was duplicated in the pancreatic tail: one was dilated and communicating with the cyst, and the other duct was dilated within the pancreatic tail (black arrow).

Fig. 4. Histological findings of the cyst.

The cyst lumen was covered with stratified squamous and ciliated columnar epithelium with a longitudinal and circular muscle layer (Hematoxylin and eosin, x 40).

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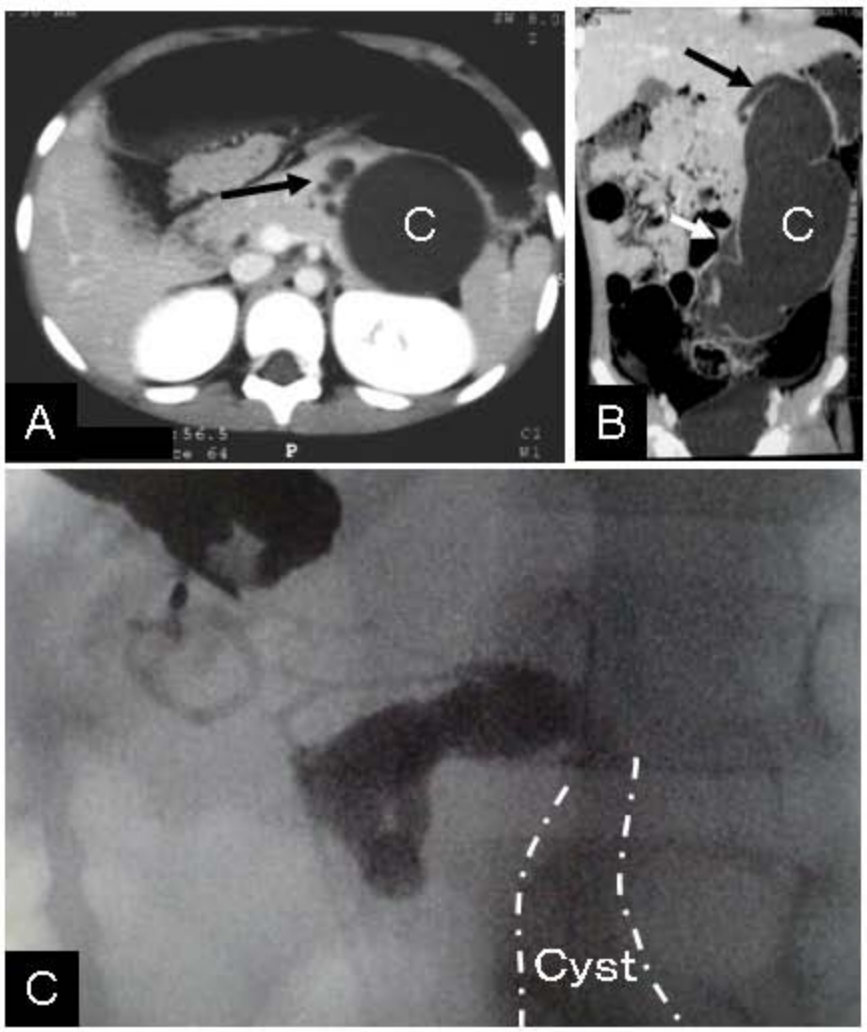
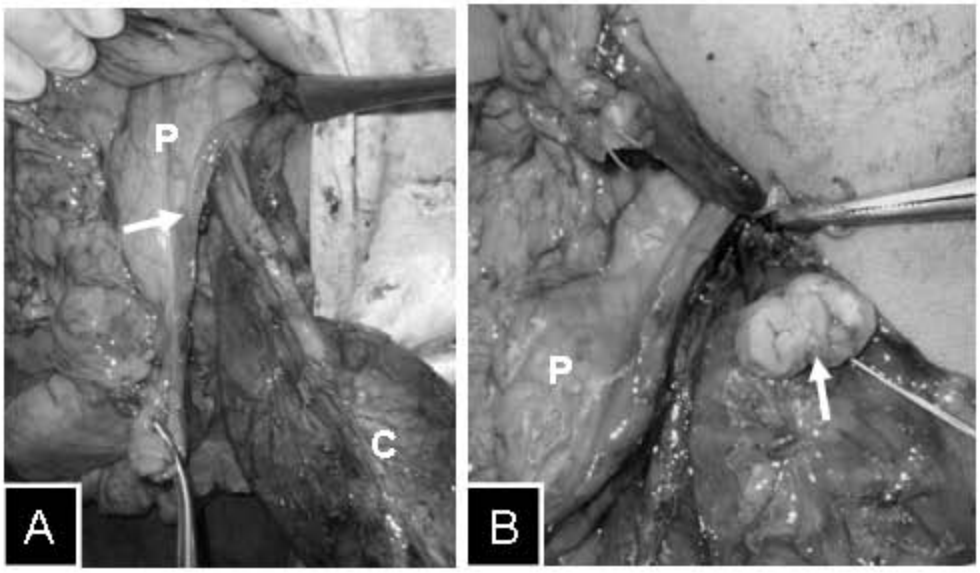
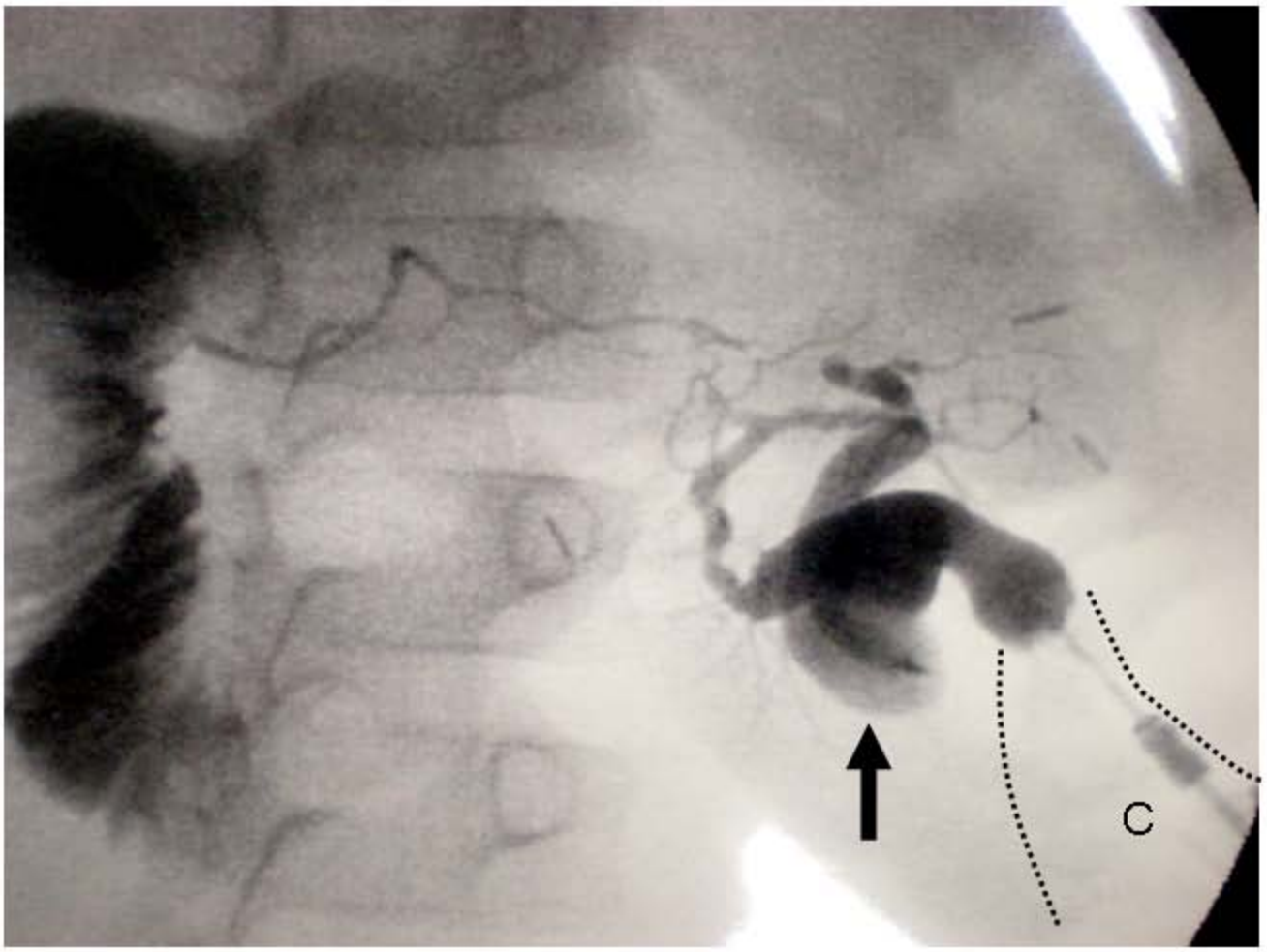
Fujishiro Fig.1



Fujishiro Fig.2



Fujishiro Fig.3



Fujishiro Fig.4