Agenesis of the dorsal pancreas with chronic suppurative pancreatitis
Case report and literature review
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Abstract
Rationale: Agenesis of the dorsal pancreas (ADP) is a rare congenital anomaly of the pancreas. ADP is associated with some other medical problems such as diabetes mellitus, abdominal pain/bloating, pancreatitis, pancreatic neuroendocrine tumor and so on. In this study, we present a case of ADP with chronic suppurative pancreatitis, summarize the clinical characteristics of the reported cases in China and review the correlative literature.

Patient concerns: A 51-year-old Chinese man, with a history of impaired fasting glucose, presented with jaundice, pruritus and dark urine. Laboratory analysis showed abnormal liver function and elevated carbohydrate antigen 19-9.

Diagnoses: Contrast-enhanced computed tomography demonstrated a mass located at the head of pancreas and complete absence of the body and tail of pancreas. Endoscopic retrograde cholangiopancreatography demonstrated an eccentric malignant stricture about 1.6cm of distal common bile duct.

Interventions: The patient underwent pancreaticoduodenectomy because of the suspicion of pancreatic tumor. The postoperative pathological result was chronic suppurative pancreatitis, with moderate hyperplasia in focal ductal epithelium.

Outcomes: A long-term follow-up shows that the patient is asymptomatic with well-controlled diabetes mellitus and pancreatic exocrine insufficiency.

Lessons: ADP is a quite rare congenital malformation of the pancreas with poorly-understood pathogenesis. The diagnosis of ADP depends on the imaging examination. The therapeutic strategy varies from person to person due to the different accompanying conditions.

Abbreviations: ADP = agenesis of the pancreas, CT = computed tomography, DM = diabetes mellitus, ERCP = endoscopic retrograde cholangiopancreatography.

Keywords: agenesis of the dorsal pancreas, case report, diabetes mellitus, pancreatic anomaly, pancreatitis
1. Introduction

The pancreas develops from dorsal and ventral pancreatic buds arising from the caudal region of the embryonic foregut. The dorsal pancreatic bud contributes to the upper part of the head, body and tail of the pancreas, and the ventral pancreatic bud eventually develops into the uncinate process and inferior part of the head of the pancreas. Agensis of the dorsal pancreas (ADP), also known as congenital short pancreas, is a rare congenital abnormality resulted from the embryological failure of the dorsal pancreatic bud to form the body and tail of the pancreas.[1,2] Up to now, around 100 cases of ADP have been reported worldwide since 191,[3,4] whereas <20 cases have been reported in China as we know so far.[5–19] Here we reported a a case of ADP with chronic supplicative pancreatitis and evaluated the clinical features of patients with ADP in China.

2. Case presentation

A 51-year-old man was admitted to the hospital with a 10-day history of jaundice, pruritus and dark urine. There were no other complaints. The patient underwent cholecystectomy for cholecystolithiasis in another medical unit nine months ago. He complained of impaired fasting glucose for several months yet did not confirm the diagnosis of diabetes mellitus (DM). He had no history of other diseases, and the family history was unremarkable. On admission, the abdomen was soft, not distended, and the patient presented no painful abdomen on palpation, no abdominal guarding and no palpable masses. Physical examination was otherwise within normal limits.

Laboratory assessment revealed: leukocytes $6.0 \times 10^9$ cell/L (reference values from $4.0 \sim 10.0 \times 10^9$ cells/L), total bilirubin $139.5 \mu$mol/L (2.1–17.3), direct bilirubin $55.1 \mu$L/L (0–5.8), alanine aminotransferase $1822 \text{U/L}$ (0–40), alkaline phosphatase $442 \text{U/L}$ (40–135), γ-glutamyl transpeptidase $378.1 \text{U/L}$ (5–40), and both serum amylase and lipase were within the normal range, carbohydrate antigen 19–9 $103.1 \text{U/mL}$ (0–30.0), carbohydrate antigen $24.2 \sim 143 \text{U/mL}$ (0–25.0), carbohydrate antigen 50 $160 \text{U/mL}$ (0–20.0). An abdominal computed tomography (CT) scan showed a mass located at the pancreas head with the dilation of intrahepatic and extrapapillary bile duct, and complete absence of the body and tail of pancreas (Fig. 1C–G). Major duodenal papilla was normal by electronic duodenoscopy (Fig. 1A). Endoscopic retrograde cholangiopancreatography (ERCP) demonstrated an eccentric malignant stricture about 1.6 cm of distal common bile duct. Figure 1B showed remarkably dilated common bile duct, intrahepatic bile duct and the location of the bile duct obstruction as well. Moreover, only the pancreatic duct with normal diameter located at the head of the pancreas was demonstrated. Combined with the analysis of the imaging examination results, abnormal liver function and elevated tumor marker levels, carcinoma of head of pancreas suspected. With thorough preoperative discussion and communication with the patient’s family, the patient underwent a laparotomy operation, during which a mass located at the pancreas head, the absence of the body and tail of pancreas was confirmed (Fig. 1F). Intraoperative frozen section examination was conducted twice. The first result showed no obvious atypia in the inspected pancreas tissue, yet the subsequent result was chronic supplicative pancreatitis, with moderate hyperplasia in focal ductal epithelium. The surgery was continued after adequate communication with the patient’s family. The gallbladder, common bile duct, duodenum, and the pancreas head were firstly resected as the routine pylorus-preserving Whipple procedure. Then cholecystjejunostomy and gastrojejunostomy were performed routinely. In the present case, this procedure was almost equivalent to total pancreatectomy. Thus, there was no pancreaticojejunostomy or the other pancreatic drainage routes. The postoperative pathological result was chronic supplicative pancreatitis, with moderate hyperplasia in focal ductal epithelium as well. Figure 2 shows the representative hematoxylineosin staining of pancreatic tissue images. Generally, the blood glucose of the patient was under control during the perioperative period. Fasting blood glucose values (venous blood) after operation were shown as follows: 1st day 25.34 mmol/L, 3rd day 9.73 mmol/L, 5th day 8.96 mmol/L, and 14th day 8.94 mmol/L. Moreover, the patient’s postoperative glucose values of peripheral blood were monitored every 4 hours, fluctuating from approximately 13.8 to 24.8
and the tail of the pancreas, and gives rise to the accessory pancreatic duct, which is also called duct of Santorini.\[2,20\] Any failure in the development of the dorsal bud therefore leads to an absence of a functional pancreatic body, tail, and duct of Santorini. This anomaly could be partial or complete. In partial ADP, the minor papilla, duct of Santorini, or the pancreatic body are present. In complete ADP, the neck, the body, and the tail of the pancreas, duct of Santorini, and minor papilla are all absent.\[21\]

The exact genetic pathogenesis of ADP is still poorly understood. *Hepatocyte Nuclear Factor 1-Beta* and *GATA Binding Protein 6* genes were proven to be correlated with the embryonic development of the pancreas.\[22,23\] Besides, experiments in mice showed that *homeobox protein HB9* gene and *retinaldehyde dehydrogenase 2* gene mutation or deficiency resulted in ADP.\[24,25\] Consistent results, however, were not observed in humans.

According to the study of Schnedl et al, only 53 cases of ADP have been reported from 1911 to 2008.\[3\] Most ADP patients were asymptomatic, yet commonly associated with DM and other additional medical problems such as pancreatitis, abdominal pain, polysplenia syndrome, and so on. A recent study further summarized the demographics of 53 cases of ADP in the published studies of the Medline and ISI Web of Science Databases from 2008 to 2015.\[4\] Except for the common associated diseases like DM and pancreatitis, it described 9 cases of pancreatic adenocarcinoma, who underwent total pancreatectomy.

Due to the language reasons, the aforementioned systematic reviews did not include all the ADP cases in China. In the present study, we performed a literature search using the terms “agenesis of the dorsal pancreas,” “dorsal pancreatic agenesis,” “congenital short pancreas” from the Medline and Chinese databases-CNKI, Wanfang, Baidu Scholar. Overall, 15 articles and 18 cases of ADP (including the present case) were included in this literature review. The clinical features of 17 cases reported in Chinese region were summarized in Table 1.\[5–19\] These cases aged from 15 to 67 and comprised 5 males and 13 females, among whom one case was confirmed by autopsy.\[6\] The first case of ADP was also incidentally found by autopsy in 1911.\[26\] In Chinese cases, most cases (11/18) had DM or impaired glucose tolerance, which was similar to the previous reviews.\[1,4\] Most of the islet cells locate at the body and tail of the pancreas, thus ADP contributes to the development of DM.\[27\] Abdominal pain is another common presenting symptom with this anomaly, yet it may be resulted from other diseases like pancreatitis or pancreatic tumor in some patients with ADP. In our review, abdominal pain was detected in 7 of 18 cases of ADP. Of note, gastrointestinal malrotation was described in 3 of 18 Chinese ADP cases. It is regarded that the dorsal and ventral pancreatic buds fuse during rotation of the gut tube at the seventh week of gestation,\[1,4\] which may cause the linkage between the pancreatic anomaly and gastrointestinal malrotation. The pathological result showed that our ADP case was associated with chronic supplicative pancreatitis, with moderate hyperplasia in focal ductal epithelium of the pancreas, which was actually considered to be the precancerous lesion of the pancreas. Previous studies have confirmed that ADP patients may combine with chronic pancreatitis and peripancreatic tumors including pancreatic adenocarcinoma, neuroendocrine tumor, solid pseudopapillary tumor of the pancreas, and cholangiocarcinoma.\[18,26–31\] Cholangiocarcinoma and solid pseudopapillary

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**Figure 2.** Representative HE staining of pancreatic tissue images from the reported case of ADP (×20 objective).
Table 1
Characteristics of reported cases of dorsal pancreas agenesis in China.

| Case | Author/year (reference) | Age, y/sex | Symptoms | Family history | Diagnostic method | Type | Associated diseases |
|------|-------------------------|------------|----------|----------------|-------------------|------|---------------------|
| 1    | Wang et al, 1990[5]     | 54/M       | Abdominal pain | NA             | Ultrasound, ERCP, CT, Surgery | Complete | DM, Dilated biliary trees |
| 2    | Lao and Mo, 2002[6]     | 15/F       | NA       | NA             | Autopsy           | Complete | NA                  |
| 3    | Ma et al, 2003[7]       | 35/F       | Epigastric pain, fever, jaundice | NA             | Ultrasound, CT | Complete | Cholelithiasis |
| 4    | Wu et al, 2007[8]       | 43/F       | Jaundice, low fever | NA             | ERCP, MR, surgery | Complete | D, Intestinal malrotation, Distal cholangiocarcinoma |
| 5    | Lin et al, 2009[9]      | 23/F       | Unremarkable | Unremarkable | CT, MR            | Complete | DM                  |
| 6    | Wei and Sun, 2009[10]   | 23/F       | Unremarkable | NA             | CT, Surgery       | Partial | Solid-pseudopapillary tumor of the pancreas |
| 7    | Lin and Chen, 2013[11]  | 35/M       | Unremarkable | Unremarkable | CT                | Partial | DM                  |
| 8    | Zhou et al, 2014[12]    | 56/M       | Unremarkable | Unremarkable | CT                | Complete | DM                  |
| 9    | Li et al, 2015[13]      | 23/F       | Unconsciousness | Unremarkable | CT                | Complete | Polysplenia, Gastric malrotation |
| 10   | Wu, 2015[14]            | 32/F       | Unremarkable | NA             | CT                | Partial | Acute pancreatitis, Polysplenia |
| 11   | Zhang and He, 2015[15]  | 52/F       | Abdominal pain | NA             | CT                | Complete | Gastric volvulus, Intestinal malrotation |
| 12   | Li et al, 2015[16]      | 23/F       | Hyperglycemia | Unremarkable | MR               | Complete | DM                  |
| 13   | Yang et al, 2016[17]    | 30/M       | Epigastric pain | Unremarkable | CT, MR            | Complete | Diabetic ketoacidosis |
| 14   | Mei et al, 2020[18]     | 65/F       | Abdominal pain, nausea, bloating, acid regurgitation | Unremarkable | CT               | Complete | DM                  |
| 15   | Zhong et al, 2020[19]   | 67/F       | Lower back pain | NA             | CT, MR, endoscopic ultrasound | Complete | Impaired glucose tolerance |
| 16   | Present case            | 51/M       | Jaundice, pruritus, dark urine | NA             | ERCP, CT, surgery | Complete | DM, Chronic supplicative pancreatitis, Moderate hyperplasia in focal ductal epithelium of the pancreas |

DM = diabetes mellitus, F = female, M = male, NA = not available, CT = computed tomography, ERCP = endoscopic retrograde cholangiopancreatography, MR = magnetic resonance.

tumor of the pancreas were described to be combined with Chinese ADP patients in our review as well.[8,10]

Generally, the diagnosis of ADP depends on combination of imaging examinations, including CT and ERCP or magnetic resonance cholangiopancreatography by demonstrating the absence of the pancreatic body, the tail and the missing duct of Santorini as well. ERCP is an invasive procedure and operator-dependent for successful identification of opacify of the main and accessory pancreatic duct. By contrast, magnetic resonance cholangiopancreatography has its advantage by clearly presenting the pancreatic duct morphology noninvasively. Ultrasound has limitations because of nonvisualization of the body and tail of the pancreas due to the interference from bowel gas or technical failure.[33,34] Although the application of endoscopic ultrasound in the diagnosis of ADP has been proved to be effective and reliable.[19] In the present case, CT scanning and ERCP was firstly performed preoperatively, and surgery finally confirmed the diagnosis of ADP by revealing the absence of the body and tail of the pancreas.

The differential diagnosis of ADP includes pancreatic fat replacement and distal pancreatectomy. The former occurs because of the atrophy of the distal pancreatic parenchyma, and MR is helpful to make the differential diagnosis by demonstrating the different fat signal of the head of the pancreas.[18] In the cases associated with the absence of splenic vein and relevant operation history, distal pancreatectomy should be considered.[15] Treatment is not necessary for asymptomatic ADP patients. The main goal of the therapy for ADP is to relieve the associated symptoms. A low-fat diet and diabetes control are recommended for the patients of ADP with DM.[36] If the ADP patients have pancreatitis, pancreatic enzymes could be administrated to reduce the pancreatic secretion and promote pain relief.[37] When pancreatic tumors or other malignant medical problems are suspected in ADP patients, surgical therapy like pancreatoduodenectomy, even total pancreatectomy should be taken into consideration. Moreover, pancreatin supplement could be useful in ADP patients complicated with the symptoms of exocrine pancreatic insufficiency.[18]

The present ADP case presented with chronic pancreatitis, with moderate hyperplasia in focal ductal epithelium of the pancreas, which was not reported before. The treatment in this patient was equivalent to total pancreatectomy.

To summarize, ADP is a quite rare congenital malformation of the pancreas associated with abdominal pain, DM, or other diseases. Reported cases of ADP are rare. Therefore, in-depth investigations should be performed, for appropriate management.

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Author contributions

LX and XB designed the research, summarized the present case and wrote the manuscript and contributed equally to this work; NM, PJ, FY and YZ reviewed the literatures and conducted the data search; NM conducted the follow up of the present case and assisted in editing the manuscript and the figure. All authors read and approved the manuscript.

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