Annular pancreas concurrent with pancreaticobiliary maljunction presented with symptoms until adult age: case report with comparative data on pediatric cases

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Abstract

Background: Annular pancreas (AP) concurrent with pancreaticobiliary maljunction (PBMJ), an unusual coexisted congenital anomaly, often presented symptoms and subjected surgical treatment at the early age of life. We reported the first adult case of concurrent AP with PBMJ presented with symptoms until his twenties, and performed a literature review to analyze the clinicopathological features of such cases comparing with its pediatric counterpart.

Case presentation: The main clinical features of this case were abdominal pain and increased levels of plasma amylase as well as liver function test. A complete type of annular pancreas with duodenal stenosis was found, and dilated common bile duct with high confluence of pancreaticobiliary ducts was also observed. Meanwhile, extremely high levels of bile amylase were detected both in common bile duct and gallbladder. The patient received duodenoojejunostomy (side-to-side anastomosis) as well as choledochojejunostomy (Roux-en-Y anastomosis), and was discharged in a good condition.

Conclusion: AP concurrent with PBMJ usually presents as duodenal obstruction in infancy, while manifests as pancreatitis in adulthood. Careful long-term follow-up is required for children with AP considering its association with PBMJ which would induce various intractable pathologic conditions in the biliary tract and pancreas.

Keywords: Annular pancreas, Pancreaticobiliary maljunction, Pancreatitis

Background

Annular pancreas (AP) is a rare congenital anomaly that is frequently associated with duodenal atresia (DA) or duodenal stenosis (DS) [1]. Pancreaticobiliary maljunction (PBMJ) is another congenital anomaly defined as an anatomical maljunction of the pancreatic duct and the biliary duct outside of the duodenal wall beyond the influence of the sphincter of Oddi, usually forming a markedly long common channel [2]. PBMJ and AP are embryologically closely related entities [3]; however, there have been only a limited number of case reports. To the best of our knowledge, only 11 pediatric cases have been documented in detail, but no adult case has been reported previously in English literature [4-13]. The authors report the first adult case with a literature review to analyze the clinicopathological features of such cases, and emphasize that the unusual coexistence of the two anomalies should be pay much attention in the diagnosis and treatment of recurrent pancreatitis and in the prevention of cancers originating from bile duct and gallbladder.

Case presentation

The patient, a 26-year-old man, presented to our department with abdominal pain. He was born at 40 weeks’ gestational age with a birth weight of 3,010 g. Since his 3 years of age, the patient presented with relatively greater appetite compared with corresponding age cohorts and...
wiggly epigastric mass which would disappear two hours
after meal. He remained free from vomiting and acute
abdomen until the first episode of abdominal pain at the
age of 23 years. Since then, he underwent totally 3 epi-
sodes of acute abdomen, and was diagnosed with acute
pancreatitis.

On presentation, physical examination revealed tender
epigastrium without palpable mass and jaundice. Labora-
tory data showed abnormal liver and pancreatic function
tests with elevated serum levels of aspartate aminotransfer-
ase (85 U/L; normal range, 0–40 U/L), alanine aminotrans-
ferase (72 U/L; normal range, 0–40 U/L), total bilirubin
(32.7 μmol/L; normal range, 0–19.5 μmol/L), amylase (541
U/L; normal range, 25–125 U/L) and lipase (279 U/L;
normal range, <190 U/L). Abdominal sonography revealed
a dilated common bile duct. Computerized tomography
additionally showed the dilation of proximal duodenum
with stenosis at the distal end of descending duodenum.
Magnetic resonance imaging and cholangiopancreato-
graphy revealed fusiform dilatation of the common bile
duct with high confluence of pancreaticobiliary ducts
with a common channel measured 24 mm in length
(Figure 1 A, B). Endoscopy showed an enlarged pylorus and
excessively ectatic duodenal cavity, but endoscopic retro-
grade cholangiopancreatography was not successful due to
duodenal stenosis at the descending part (Figure 2). A ten-
tative diagnosis of acute pancreatitis associated with pan-
creaticobiliary maljunction and annular pancreas was
made.

During laparotomy, a complete type of annular pan-
creas and a fusiform-type dilatation of common bile duct
were confirmed. Air charging demonstrated an enlarged
pylorus and excessively ectatic duodenal bulb with du-
odenal stenosis at the distal end of descending part
(Figure 3). Pancreatic amylase was at extremely high
levels in the bile within the common bile duct and gallblad-
der sampled immediately after laparotomy (5592.8 U/L and
85694.0 U/L, respectively). Transection of the dilated com-
m b o il ed u c ta n dc h o l e d o c h o j e j u n o s t o m yw e r e p e r f o r m e d .

Patient also underwent duodenoejejunostomy for correction
of duodenal stenosis associated with annular pancreas.

The postoperative course was uneventful. The patient
was discharged on the 21th postoperative day on full
oral feeding and without abdominal pain. The patient
has been free from abdominal pain with normal serum
amylase levels in the follow-up period of 12 months
postoperatively.

Discussion
There are three types of pancreatic fusion anomalies: an-
nular pancreas, pancreas divisum and portal annular
pancreas [14]. While portal annular pancreas is the rar-
est and mostly asymptomatic, annular pancreas (AP),
with an incidence ranging from 0.005% to 0.015%, usu-
ally presents as duodenal obstruction in infancy, [15-17].
However, some patients with AP remain asymptomatic.
until into adulthood, when the disorder manifests as pancreatitis as shown in our patient. From the data on pediatric cases, it was indicated that pancreatitis in the setting of AP may be associated with concomitant pancreaticobiliary malformations including pancreas divisum, PBMJ, or duodenal diverticulum [18,19], which might induce to insufficient drainage of pancreatic secretions and bile. In the present case, biliopancreatic reflux associated with PBMJ was probably the cause of the development of recurrent pancreatitis.

Until now, there were totally 12 cases of concurrent AP with PBMJ, of which the 11 pediatric cases were described previously, Table 1. Data from these reports showed that most of these patients with AP as well as PBMJ have subjected both duodenoduodenostomy and flow-diversion surgery simultaneously or metachronously. However, prophylactic flow-diversion surgery remains controversial. The recent case reported by Komuro had just received duodenoduodenostomy for AP while prophylactic flow-diversion surgery had not been performed for concomitant PBMJ with bile duct dilatation. According to our experiences, the extremely high levels of pancreatic enzymes in common bile duct or gallbladder should be an important indication for flow-diversion surgery.

PBMJ can be divided into PBMJ with biliary dilatation and PBMJ without biliary dilatation [20]. Out of the 12 reviewed cases, 10 cases (83.3%) were found with dilated common bile duct. All seven AP associated DA cases showed fusiform dilatation of the common bile duct at 2–12 years of age. However, no choledochal cyst or marked dilation was discovered soon after birth when they underwent initial surgery for the duodenal obstruction. Thus, it should be suggested that although it is often called “congenital” bile duct dilatation, the bile ducts probably undergo a gradual development of dilatation caused by PBMJ after birth.

The diagnosis of PBMJ in patients with AP is often delayed and even missed, not only because of the gradual

### Table 1 Reported cases of annular pancreas accompanied with pancreaticobiliary maljunction

| Case | Author     | Year | Sex | Age at duodenal surgery | Duodenal Lesion associated with AP | Age at biliary surgery | Bile duct lesion associated with PBMJ |
|------|------------|------|-----|-------------------------|-----------------------------------|-----------------------|--------------------------------------|
| 1    | Komura [5] | 1991 | F   | 2 years                 | DS                                | 2 years               | Not dilated                          |
| 2    | Okada [4]  | 1993 | M   | 2 days                  | DA                                | 12 years              | If, IIf                              |
| 3    | Okada [4]  | 1993 | F   | 7 days                  | DA                                | 3 years               | If, IIf                              |
| 4    | Nakamura [6]| 1993 | F   | 3 days                  | DS                                | 5 years               | If                                   |
| 5    | Komuro [7] | 2000 | M   | 11 months               | DS                                | 13 months             | Intrapancreatic cyst                  |
| 6    | Sugimoto [8]| 2002 | F   | 0 day                   | DA                                | 2 years               | If, IIf                              |
| 7    | Oowari [13]| 2003 | F   | 1 day                   | DA                                | 8 years               | If                                   |
| 8    | Shih [14]  | 2005 | F   | 3 days                  | DA                                | 7 years               | If, IIf                              |
| 9    | Iwai [9]   | 2009 | F   | 1 day                   | DA                                | 4 years               | If, IIf                              |
| 10   | Okuyama [12]| 2010| M   | neonatal                | DA                                | 3 years               | If                                   |
| 11   | Komuro [11]| 2012 | M   | 3 years                 | DS                                | NOT                  | Not dilated                          |
| 12   | Present case| 2013 | M   | 26 years                | DS                                | 26 years              | If                                   |

M = male; F = Female; DA = duodenal atresia; DS duodenal stenosis, If fusiform dilatation of the common bile duct, IIf fusiform dilatation of the intrahepatic bile duct; NOT = not operated; PBMJ = pancreaticobiliary maljunction.
and variable development of bile duct dilatation, but also because of lacking accurate diagnostic maneuver for PBMJ [21]. Thus, although combination of AP and PBMJ is a rare condition, careful follow-up is required for the patients with AP, taking into account that PBMJ might present clinical symptoms after several years following definitive surgery for AP. Delayed or missed diagnosis of concurrent PBMJ might sometimes cause serious disease or even death. PBMJ is commonly associated with carcinoma of the bile duct and gallbladder [22,23]. Meanwhile, bile may also reflux into the pancreatic duct via PBMJ in some conditions, such as with cholangitis or bile stasis in the biliary tract. Refluxed bile may activate pancreatic enzymes, and may thus cause recurrent acute pancreatitis and subsequent chronic pancreatitis, which is related to pancreatic carcinoma [24]. Thus, it is very important for AP patients to receive MRCP and/or ERCP to confirm the coexistence of PBMJ, and long-term careful follow-up make much of senses to the prevention and treatment for the recurrent pancreatitis of such patients.

Conclusions
AP concurrent with PBMJ usually presents as duodenal obstruction in infancy, while manifests as pancreatitis in adulthood. Although prophylactic flow-diversion surgery remains controversial, careful long-term follow-up is required for patients with AP associated DA or DS considering the association of concurrent PBMJ with various intractable pathologic conditions in the biliary tract and pancreas.

Consent
Written informed consent was obtained from the patient for publication of this Case report and any accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Abbreviations
AP: Annular pancreas; PBMJ: Concurrent with pancreaticobiliary maljunction; DA: Duodenal atresia; DS: Duodenal stenosis.

Competing interests
The authors declare that they have no competing interests.

Authors’ contributions
LC contributed to the design of the study and direction of its implementation. FZT conceived and designed the experiments and supervision of the field activities. LC and T.J.Z. carried out the prepared the Materials of patient and prepared the literature review as well as the Discussion sections of the text. JDR and ZLL conducted the data analysis. All authors read and approved the final version of the manuscript.

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