Radical antegrade modular pancreatosplenectomy for adenocarcinoma of the body of the pancreas in a patient with portal annular pancreas, aberrant hepatic artery, and absence of the celiac trunk
A case report

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Abstract

Rationale: Portal annular pancreas is a rare anatomic variation, where the uncinated process of the pancreas connects with the dorsal pancreas and the pancreas tissue encases the portal vein (PV), superior mesenteric vein (SMV) or splenic vein (SV). Malignancies are quite uncommon in the patients, who have an annular pancreas especially portal annular pancreas. Ectopic common hepatic artery and absence of the celiac trunk (CT) are the other infrequent abnormalities.

Patient concerns: A 74-year-old man suffered from upper abdominal and back pain.

Diagnoses and Interventions: Contrast enhanced computed tomography indicated a low-density mass in the body of the pancreas. Pathological report showed adenocarcinoma of the body of pancreas after radical antegrade modular pancreatosplenectomy (RAMPS).

Outcomes: In the operation, we found the superior vein and portal vein was surrounded by the pancreatic tissue. The left gastric artery and splenic artery originated respectively from abdominal aorta, and celiac trunk was not viewed. In addition, the common hepatic artery was a branch from the superior mesenteric artery.

Lessons: In general, this is a novel clinical case of pancreatic carcinoma happening in the portal annular pancreas which was accompanied with aberrant hepatic artery and absence of the celiac trunk at the same time. Confronted with the pancreatic neoplasms, the possibility of coexistent annular pancreas and arterial variations should be considered.

Abbreviations: CHA = common hepatic artery, CT = celiac trunk, LGA = left gastric artery, PV = portal vein, RAMPS = radical antegrade modular pancreatosplenectomy, SMV = superior mesenteric vein, SV = splenic vein.

Keywords: aberrant celiac trunk, aberrant common hepatic artery, pancreas carcinoma, portal annular pancreas

1. Introduction

Portal annular pancreas is a pancreatic deviance without symptoms, where the uncinated process rings portal vein (PV), superior mesenteric vein (SMV), and/or splenic vein (SV), and connects to the reverse side of the pancreas body.\textsuperscript{[1,2]} Because of the different position of the pancreatic duct and the high incidence of concomitant vascular anomaly, portal annular pancreas is considered to increase the postoperative pancreatic fistula risk.\textsuperscript{[3]} In addition, celiac trunk (CT) usually has 3 branches
including left gastric artery (LGA), common hepatic artery (CHA), and splenic artery (SA). Geoffroy Saint-Hilaire firstly described the case of an absent CT in 1832. Haller described the trifurcation of the CT first, which has been regarded as the normal anatomical appearance. Based on a large series of cases summarized by Matusz et al, the absent CT only occupied 0.19% of the cases (10750 cases from 19 studies). The radical antegrade modular pancreatosplenectomy (RAMPS) procedure is a modification of standard distal pancreatosplenectomy, which can provide high rate of negative tangential margin and survival for this aggressive tumor. Herein, we report a pancreatic body adenocarcinoma case taking place in a patient with the coexistence of portal annular pancreas, aberrant hepatic artery, and absence of the CT.

2. Case report
A 74-year-old man faced with epigastric pain and dorsalgia came to Pancreas Center of The First Affiliated Hospital of Nanjing Medical University. When admitted to hospital, the patient had no vomiting, no jaundice, and anemia in conjunctiva, and no other abdominal symptoms. Contrast-enhanced computed tomography revealed a low-density mass in the body of the pancreas (Fig. 1B) and the pancreas wrapped the PV (Fig. 1A). Laboratory tests revealed raised aspartate aminotransferase, 46.3 U/L (15.0–40.0 U/L), and total bilirubin, 20.9 U/L (5.1–19.0 U/L). Furthermore, carbohydrate antigen 19–9 (CA-19-9) was over 1000 U/mL (<39.0 U/mL), carcinoembryonic antigen was 13.36 ng/mL (<4.7 ng/mL), neuron-specific enolase was elevated to 21.41 ng/mL (<16.3 ng/mL).

The patient underwent RAMPS. During the operation, the pancreatic parenchyma was encircling the SMV and PV. The uncinate process was connected with the body of the pancreas (Fig. 2A–C). Distant metastasis was not observed during surgical exploration. In addition, the CHA and CT was not located over the body of the pancreas. The LGA and SA originated from aorta abdominis directly. The CHA was a branch from the superior mesenteric artery. In other words, this is a patient without CT (Fig. 3A and B). The operation was smooth (Fig. 4A and B), and the pathological report of the sample showed pancreatic ductal adenocarcinoma with regional lymph node metastasis (TNM staging: T2N1M0, stage IIB) (Fig. 5). Pancreatic fistula, bleeding, delayed gastric emptying, and other complications were not observed during the period after operation. The patient discharged from the hospital for 11 days after surgery.

3. Discussion
Portal annular pancreas surrounding PV and/or SMV is known as a congenital variation, and has been taken into account as a risk factor for postoperative complications of pancreatic surgery. Pancreas is developed from the ventral and dorsal buds in embryogenesis. As we known, the body and tail of pancreas are formed by the ventral bud. The uncinate process of pancreas is developed from the dorsal bud. The head portion of pancreas is originated from the 2 buds. During early embryogenesis, the ventral duct will take a posterior rotation. Usually, the dorsal duct and ventral duct will fuse on the PV’s right. Rarely, they will connect on the other side, thus encasing the PV.

Based on the morphology of the pancreatic duct and the relationship with portal confluence, Joseph suggested the following 3 types: type I, the ventral bud is fused with the body of the pancreas and the pancreatic duct courses behind the PV; type II, the main pancreatic duct divides into 2 ducts, and the ducts course to the PV anteriorly and posteriorly; and type III, the PV is encased by the uncinate process with a normal pancreatic duct, which course to the PV anteriorly. All the types can be divide into a, b, and c, based on the location of the SV. In our case, the portal pancreas belongs to type III B.

Contrast-enhanced computed tomography may be a good method to diagnose this deviance before surgical intervention. Multidimensional images can describe the complete variation easily. Magnetic resonance imaging is another useful method.
Figure 2. (A) Intraoperative photo showing portal annular pancreas. Uncinate process of pancreas (thin arrow), body of pancreas (thick arrow). (B) The annular pancreas encased the PV and (SMV). Margin of neck of pancreas (thin arrow). (C) An illustration. PV = portal vein, SMV = superior mesenteric vein.

Figure 3. (A) Intraoperative picture showing the LGA and SA originate respectively from the celiac artery. The CHA arise from the SMA. (B) An illustration. CHA = common hepatic artery, IMV = inferior mesenteric artery, LGA = left gastric artery, PV = portal vein, SA = splenic artery, SMA = superior mesenteric artery.

Figure 4. (A) This picture revealing the stump of the SA (thin arrow) and stump of the SV (thick arrow) after resection. (B) An illustration. CHA = common hepatic artery, IMV = inferior mesenteric artery, PV = portal vein, SA = splenic artery.
which can offer positive information about the ductal variation which often coexist in portal annular pancreas patients. Considering of this deviance preoperatively is significant for surgeons to take appropriate vascular and pancreatic anastomosis, which may decrease the risk of pancreatic fistula. For the variation of the CT in this case, according to the statements made by Matusz et al. the occurrence rate of this anomaly were concluded by the earlier case reports. The typical complete CT took the most part of the cases (about 90.70%) and the bifurcate aberrant CT occurred in 6.09% of 10,750 cases from 19 studies; however, the incidence of the absence of the CT was very rare (only 0.19%). The first classification system which described the absent CT as a morphological type was that of Morita. Based on the embryological studies of Tandler, Morita depicted 5 types for the CT, including the CT, absent CT, gastrosplenic trunk, hepatogastric trunk, and hepatosplenic trunk. According to the classification of Morita, the present anomalous case belongs to type III. Identify that the arterial deviations are significant for abdominal surgery procedure in this area. Arterial variations can also provide important information during procedures such as chemoembolization of pancreatic and hepatocellular carcinoma. What more, RAMPS procedure was a new method to cut out the tumor in the body and tail of the pancreas, which was proposed in 2003. The novel procedure achieved high survival rate in Mitchem et al and Strasberg et al’s research. However, RAMPS procedure was applied in annular pancreas in this case, the effect remains to be observed. In conclusion, for all we know, the present report is the first clinical case of pancreatic carcinoma happening in the portal annular pancreas which was accompanied with the aberrant hepatic artery and the absence of the CT at the same time. Confronted with the pancreatic neoplasms, the possibility of coexistent annular pancreas and arterial variations should be considered. The video materials during the operation was list in the supplementary data; http://links.lww.com/MD/C5; http://links.lww.com/MD/C6 and http://links.lww.com/MD/C7.

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