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Original Article

A case of annular pancreas accompanied with intraductal papillary mucinous neoplasm-case report

Fu-Jian Ji, Xiao-Feng Tian, Liang Wei, Xue-Dong Fang, Hong Cao, Hong-Yong Jin

Department of General Surgery, The China Japan Friendship Hospital of Jilin University, Changchun 130012, China

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Abstract: Objectives: Annular pancreas is a rare congenital anomaly characterized by pancreatic tissues wrapping completely or incompletely around the descending duodenum. In most patients with annular pancreas, onset occurs in early childhood. Adults with annular pancreas are prone to duodenal ulcers and pancreatitis. Intraductal papillary mucinous neoplasm (IPMN) is a type of papillary mucinous secretory epithelial tumor, which originates in the main pancreatic duct and/or branch duct. We report a case of annular pancreas accompanied with intraductal papillary mucinous neoplasm. Methods: A 52-year-old male patient hospitalized due to recurrent upper abdominal pain for one and a half years was enrolled in this study. Results: One case of annular pancreas accompanied with intraductal papillary mucinous neoplasm which manifested as recurrent chronic pancreatitis was found. After pancreaticoduodenectomy, the patient died from uncontrollable gastrointestinal bleeding. Conclusions: To the best of our knowledge, this is the first case in China and the second case worldwide of annular pancreas accompanied with IPMN in English literature.

Keywords: Annular pancreas, intraductal papillary mucinous neoplasm, chronic pancreatitis, pancreaticoduodenectomy, pancreatic fistula

Introduction

Annular pancreas was first reported by Tiedemann in 1818 [1] and was termed in 1862 by Ecker [2], who clarified the pancreatic duct system through case dissection. In 1905, Vidal first performed surgery to treat the annular pancreas which resulted in obstructive symptoms [3]. The rarity of annular pancreas could be evidenced by its prevalence of 3 in 20000 autopsy cases [4] and of 3 in 24519 operative cases [5]. Along with the deeper understanding of annular pancreas and the development of medical imaging techniques, reports on annular pancreas have gradually increased in recent years.

Intraductal papillary mucinous neoplasm (IPMN) was defined by the World Health Organization (WHO) in 2000 as one type of papillary mucin-producing tumors originating in the main pancreatic duct or the major branch pancreatic duct. It is the most common pancreatic cystic lesion and one of the most important precancerous lesions of the pancreas, which is highly associated with pancreatic cancer. It was first reported by a Japanese physician, Takaki, in1982 [6]. IPMN is classified into three groups according to its origin: (1) main-duct, dilation of the main pancreatic duct and tumor located in the main pancreatic duct; (2) branch-duct, dilation of the branch pancreatic duct and tumor not located in the main pancreatic duct; (3) mixed, tumor located in both the main pancreatic duct and the major branch duct [7]. According to its cytology, IPMN can also be subcategorized into four groups: adenoma or low grade dysplasia, borderline tumors or moderate dysplasia, carcinoma in situ and invasive carcinoma [8].

We report here a case of annular pancreas accompanied with IPMN in a 52 year-old man. To the best of our knowledge, this is the first

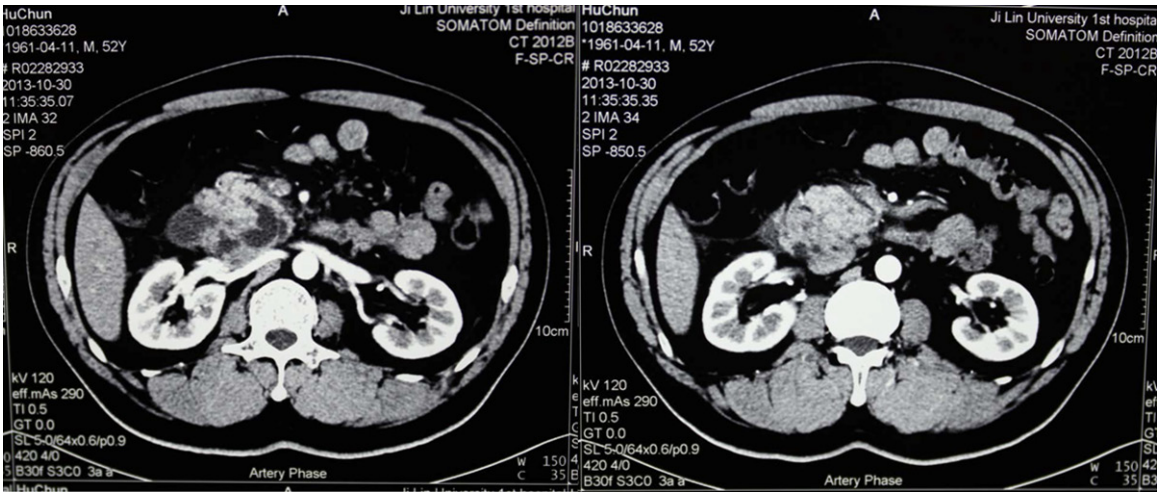

Figure 1. The contrast-enhanced computed tomography (CT) scan revealed the tortuous low density strips in the head of pancreas, which was considered as the dilated branch pancreatic duct.

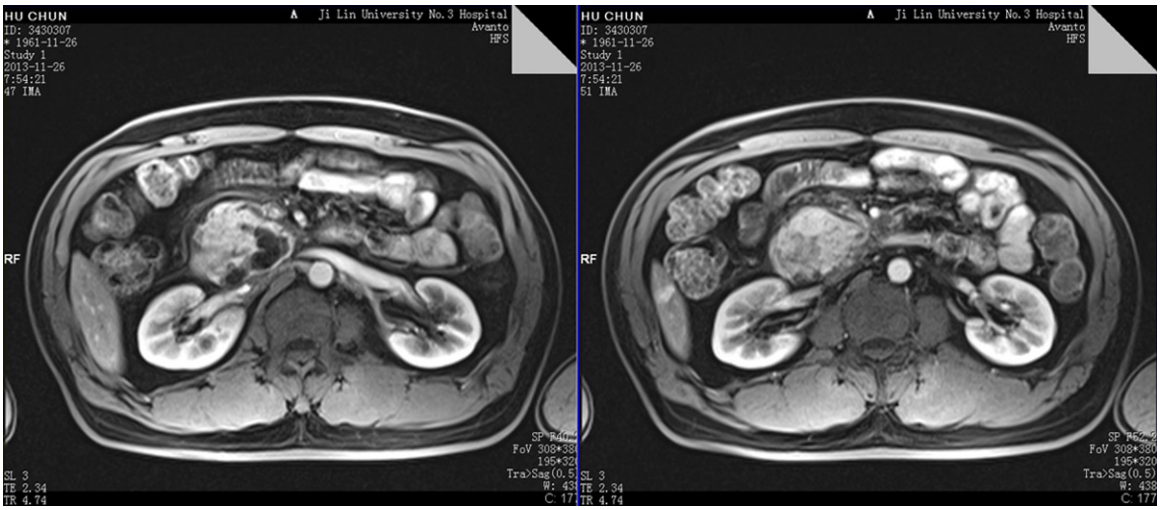

Figure 2. The MRI showed the string-of-beads dilation of branch pancreatic duct connected to the main pancreatic duct in the enlarged head of pancreas.

case in China and the second case worldwide of concurrent association of annular pancreas and IPMN in English literature.

Case report

A 52-year-old male patient was hospitalized with recurrent upper abdominal pain in the past one and a half years. This extremely severe and unendurable pain usually lasted for 1-2 hours, but could be relieved by body flexion. The pain was often followed by diarrhea but not associated with back pain, fever, nausea and vomiting, abdominal distension, yellow discoloration of skin and sclera. The patient denied nocturnal pain but admitted hunger pain when onset occurred. He underwent a total of 6 episodes of abdominal pain which had irregular intervals between one half to three months. He had lost about 10-20 kilograms of body since then. Physical examinations displayed upper abdominal tenderness associated with occasional rebound tenderness and muscle tension. The serum and urine amylase increased significantly. The symptoms could be improved by supportive treatments such as anti-acid therapy and fluid resuscitation. The patient had a vitreous opacity in the left eye which was treated by surgery. He also claimed urethral stones cured

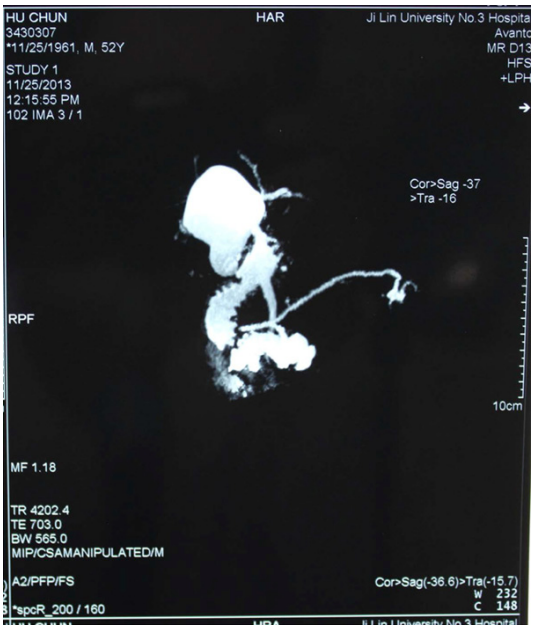

Figure 3. The MRCP displayed a pouch pocket with high signal intensity and irregular edges; It also showed multiple nodular projections in a length of about 50 mm terminated at the end and not connected to the duodenum.

by conservative treatment. His father died of pancreas cancer and his mother died of liver cancer.

No remarkable findings for tumor markers nor complete blood count were found in laboratory investigations. Contrast-enhanced computed tomography (CT) scan revealed the tortuous low density strips in the head of pancreas, which was considered as the dilated branch pancreatic duct (**Figure 1**). The MRI showed the string-of-beads dilation of the branch pancreatic duct connected to the main pancreatic duct in the enlarged head of pancreas. The MRCP displayed a pouch pocket with high signal intensity and irregular edges; The MRCP also showed multiple nodular projections of a length of about 50 mm terminated at the end and not connected to the duodenum (**Figures 2**, **3**). Both CT and MRI demonstrated the existence of pancreatitis and showed that the descending duodenum was encircled in the head of pancreas, which suggested the possibility of the annular pancreas. The ERCP demonstrated the normal main pancreatic duct and the cystic dilation of the branch pancreatic duct with the maximum diameter of about 8.6 mm (**Figure 4**).

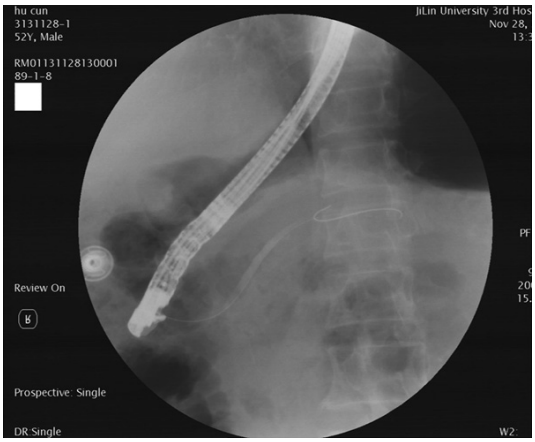

Figure 4. The ERCP demonstrated the normal main pancreatic duct and the cystic dilation of the branch pancreatic duct with the maximum diameter of about 8.6 mm.

In view of these findings described above, a tentative diagnosis of chronic pancreatitis associated with pancreatic head tumor (IPMN possible) and possible annular pancreas was made. During pancreaticoduodenectomy, the annular pancreas and a cystic mass located at the head of pancreas were confirmed (**Figure 5**). A duct to mucosa pancreaticojejunostomy was performed. The operative course was uneventful. Beginning from the seventh day after surgery, about 20 ~ 25 ml of light yellow liquid were collected daily from the right pancreatic intestinal drainage tube. The amylase level in these samples was measured as 6502 IU/L, indicating the pancreatic fistula. Symptomatic treatment was therefore given. On the 13th day after surgery, hemorrhagic anemia due to abdominal bleeding was diagnosed. Appropriate hemostasis treatment was given. On the 17th day after surgery, melena was observed, which led to the diagnosis of gastrointestinal bleeding. Blood transfusion, hemostasis and other symptomatic treatments were given. On the 18th day after surgery, the patient died from the uncontrollable gastrointestinal bleeding.

The postoperative pathological diagnosis was as follows: 1) pancreatic intraductal papillary mucinous adenomas without significant infiltration, which was characterized by a moderate dysplasia of some glands (**Figure 6**); 2) annular pancreas; 3) chronic pancreatitis. The immunohistochemistry results were as follows: K-ras (+), E-cadherin (+), Ki-67 (10% +), P53 (< 1% +), bcl-2 (-), CD44 V6 (+).

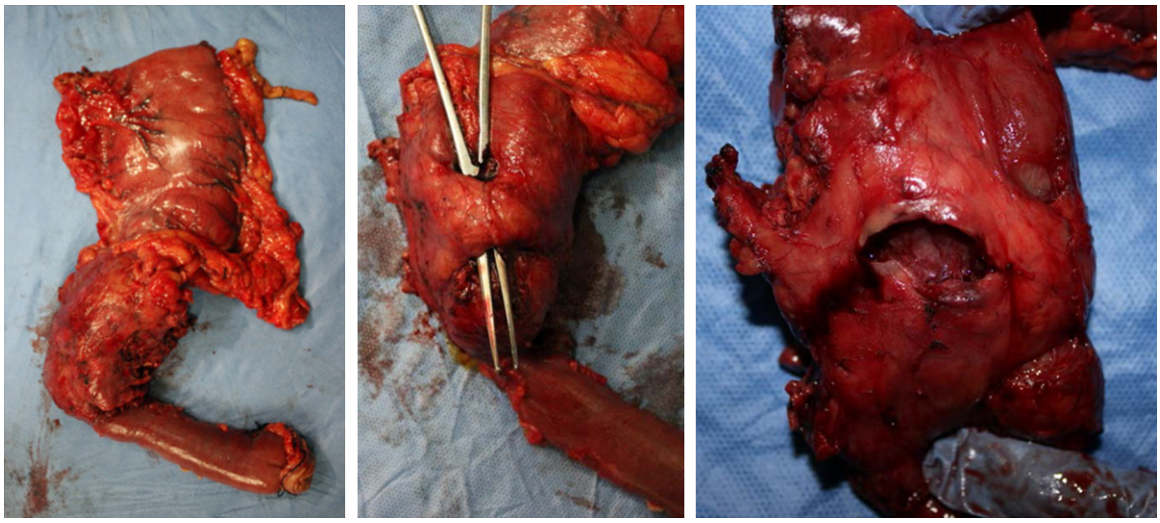

Figure 5. Specimens after surgery. The descending part of the duodenum was completely surrounded by the pancreas. A cystic mass with diameter of about 2.5 cm was seen at the back of the head of the pancreas. The viscous liquid was punctured from the mass. The pancreas was overall hard and swollen with enlarged head of pancreas.

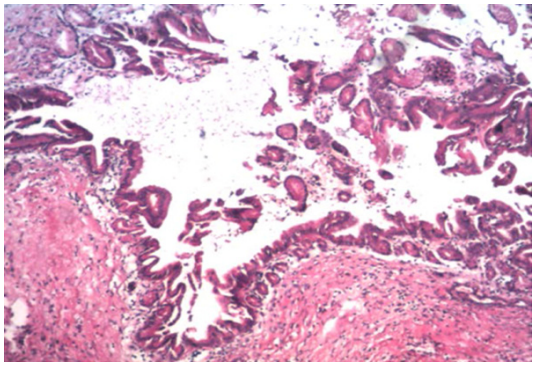

Figure 6. Microscopic findings of the resected specimen showed the dilation of pancreatic duct lined with columnar mucus secreting epithelial cells. Tumor was composed of moderate dysplasia cells arranged in a papillary pattern without infiltration.

Discussions

It has been widely accepted that annular pancreas is due to abnormal rotation of the ventral pancreatic bud, which was first proposed by Lecco [9]. In brief, the adherence of the right ventral pancreatic bud to the duodenum before gut rotation results in a partial or complete ring of pancreatic tissue around the duodenum and subsequent duodenum stenosis. Annular pancreas manifests mainly as duodenum obstruction in children, while more complicated with duodenal obstruction, pancreatitis, or peptic ulcer, in adults. It is seldom associated with obstructive jaundice and malignancy.

Patra *et al* has pointed out that the preoperative diagnosis of annular pancreas is often difficult and the diagnosis mainly relies on medical imaging [10]. The upper gastrointestinal series can display the dilation of the stomach and the upper duodenum, as well as the stenosis of the descending duodenum. However, it cannot ensure that it is the pancreatic tissues that surround the duodenum. CT and MRI can show the pancreatic tissues completely or incompletely encircling the narrow segment of duodenum. ERCP is specific in the diagnosis of annular pancreas, since it can illustrate the circling pancreatic duct surrounding the narrow duodenum segment. However ERCP is invasive and thus it will probably be replaced by MRCP. Endoscopic ultrasound is helpful in the diagnosis of annular pancreas.

Treatment is not necessary for annular pancreas without symptoms. Surgical bypass of the duodenum is indicated in severe stenosis. For annular pancreas associated with chronic pancreatitis, pancreatic duct stones, or malignancies which could not be excluded, pancreaticoduodenectomy should be considered.

There are no specific symptoms or signs related to IPMN in the early stage. It often manifests as recurrent acute or chronic pancreatitis due to the intermittent obstruction of the pancreatic duct resultant from the mucinous secretion of tumor cells. The characterized imaging findings

of IPMN include the dilation of the pancreatic duct, the cystic mass of the pancreas, the swelling of duodenal papilla and the mucinous secretion, all of which can be identified by ERCP specifically. The CT scan and the endoscopic ultrasound are also helpful for the diagnosis of IPMN. The Sendai Guidelines published in 2006 provide a solid foundation to begin risk stratification for IPMN and have been widely recognized by physicians as guidance for treatment of IPMN [11]. According to the Sendai Guidelines, the surgery resection for all main duct IPMN and mixed IPMN in reasonable surgical candidates is highly recommended as the malignant risk increases in these two types of IPMN; Resection is also recommended for branch duct IPMN that is symptomatic, > 3 cm, have mural nodules or demonstrate cyst-aspirate cytology. The prognosis of IPMN depends on the tumor characteristics.

The concurrent association of annular pancreas and IPMN was previously reported by Jang et al, with diarrhea as the major manifestation [12]. The case we described here, different from the previously reported case, presented with pain followed by diarrhea. We summarized our experiences from this case as following:

1): Reasons for delayed diagnosis. The diagnosis of annular pancreas accompanied by IPMN was not made for as long as one and half years until surgery. The possible reasons include our insufficient knowledge of the annular pancreas because of its low incidence, the lack of symptoms of duodenum obstructions in this case. The delayed diagnosis is also partially caused by the limitations of the imaging techniques we employed such as the limited length of gastric endoscopy. In addition, the duodenal papilla is superior to the annular pancreas in this case, which prevented our further observation on annular pancreas besides the IPMN.

2): The pancreaticoduodenectomy is the best treatment option for annular pancreas accompanied by IPMN manifested as recurrent chronic pancreatitis. This operation can cure the recurrent chronic pancreatitis by resection of the involved pancreas, can prevent the duodenum obstruction caused by the stenosis of the descending duodenum, can prevent the gastrojejunostomy anastomotic ulcer by gastrectomy, and can avoid the possible malignancy by resection of the IPMN in the head of pancreas. However, this operation is highly risky, especially for the patient with recurrent chronic pancreatitis, and thus should be operated by the senior and experienced physician.

3): The pancreatic fistula is a common complication of pancreaticoduodenectomy and highly risky. To prevent and treat the possible pancreatic fistula, the appropriate pancreaticojejunostomy and the postoperative treatments should be carefully chosen.

4): The abdominal bleeding in this case was probably due to the corrosion by pancreatic liquid. The gastrointestinal bleeding might be caused by stress ulcer.

There are no essential associations between annular pancreas and IPMN, although they share the same manifestation as pancreatitis. It is highly possible that the chronic pancreatitis is secondary to the annular pancreas and/or IPMN, especially for those without reasonable predisposing factors such as heavy meal and alcohol ingestion. Treatment and operative protocols have to be individualized.

Disclosure of conflict of interest

None.

Address correspondence to: Hongyong Jin, Department of General Surgery, The China Japan Union Hospital of Jilin University, No. 829 Xin'min Road, Chaoyang District, Changchun 130012, China. Tel: +86-0431-84997620; Fax: +86-0431-84997620; E-mail: hongyongjin537@yeah.net

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