Case Report

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# Benign Intraductal Papillary-Mucinous Neoplasm of the Pancreas Associated with Spontaneous Pancreaticogastric and Pancreaticoduodenal Fistulas

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**Key Words**
Magnetic resonance imaging · Pancreas · Intraductal papillary-mucinous neoplasm

**Abstract**
Invasive intraductal papillary-mucinous neoplasms (IPMNs) of the pancreas may be associated with pancreaticogastric fistulas as shown by case reports. We report the case of a benign IPMN associated with pancreaticogastric and pancreaticoduodenal fistulas. A 70-year-old woman was admitted with intestinal obstruction. Computed tomography and MRI showed a large dilatation of the main pancreatic duct (>1 cm) with intraductal nodules, and pancreaticogastric and pancreaticoduodenal fistulas. Several features in imaging were present to support a malignant IPMN, so that the patient underwent a pancreaticoduodenectomy. The histopathological examination of the surgical specimen showed a benign IPMN. This case proves that a benign IPMN can cause pancreaticogastric and pancreaticoduodenal fistulas, probably resulting from mechanical factors.



## Introduction

Intraductal papillary-mucinous neoplasms (IPMNs) of the pancreas were first described in 1982 by Ohashi et al. [1] as mucin-producing tumors which developed from the epithelium of the main pancreatic duct (MPD) and its side branches [2]. IPMNs are frequently asymptomatic, but in some cases they may cause epigastric pain. Different types of IPMNs have been distinguished according to their location on the MPD and its branches: lesions of the main duct (18%), branch duct (39%), and combined type (42%) [3].

From the histological and pathological point of view, the lesions are classified according to their invasiveness as benign, noninvasive carcinoma, and invasive carcinoma [3]. IPMNs are generally considered slowly growing tumors with a good outcome after surgical resection, but invasive ones have been associated with a poorer prognosis [4].

Fistulas have been reported between IPMNs and various organs, such as the common bile duct, duodenum and stomach [4, 5]. Several cases of pancreaticogastric fistulas have been described in the literature, but to our knowledge, only 1 case has been reported involving a noninvasive IPMN, which was presumed to be secondary

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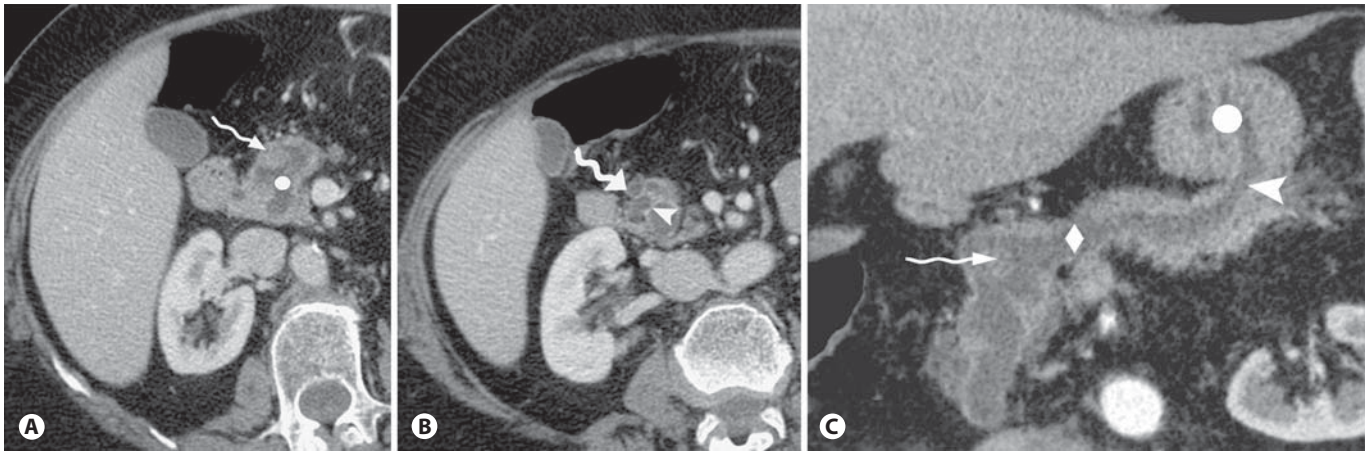


**Fig. 1.** Contrast-enhanced abdominal CT, venous phase. **A** The main pancreatic duct is dilated, measured 18 mm at the level of the pancreatic isthmus (white disc). There is a solid endoluminal nodule of the isthmic pancreas, measuring 16 mm (arrow). This well-defined nodule presents moderate contrast uptake in the venous phase. **B** A 26-mm multilocular cystic lesion (arrow) of the cephalic segment with septa (arrowhead). **C** Curved reformation demonstrates the fistula (arrowhead) between the dilated main pancreatic duct (white diamond) and the body of the stomach (white disc). The large solid nodule of the isthmic pancreas is also visible (arrow).

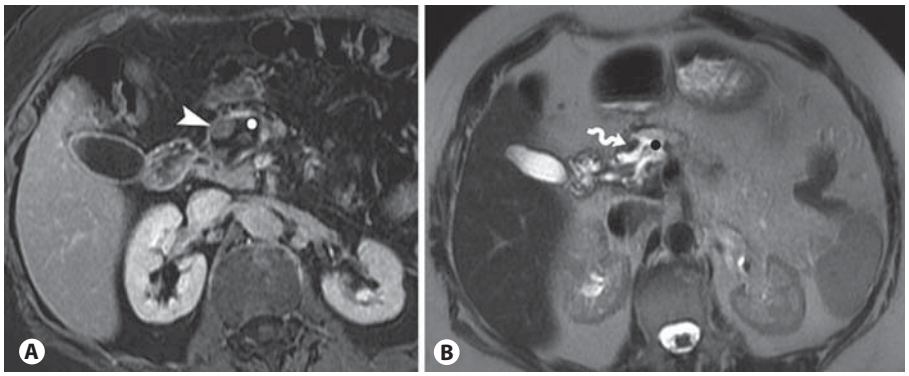


**Fig. 2.** Contrast-enhanced T1-weighted MRI, delayed phase (**A**) demonstrates the dilatation of the main pancreatic duct (white disc on **A** and black disc on **B**), as well as the presence of the solid nodule in the MPD (arrowhead) with moderate contrast uptake. This nodule appears hypointense (arrow) on the T2-weighted sequence (**B**).

to an endoscopic retrograde cholangiopancreatography (ERCP) [4].

We report a case of benign IPMN associated with spontaneous pancreaticogastric and pancreaticoduodenal fistulas.

## Clinical Case

A 70-year-old female patient was admitted due to intestinal obstruction that spontaneously resolved within the next 24 h. Her medical history included hypertension and an incarcerated inguinal hernia, for which she had been operated 10 years earlier.

On physical examination, the abdomen was distended without peritoneal signs. Laboratory results showed an increase in CRP at 10 mg/l and increased WBC (16,000 WBC/ml). The hepatic and pancreatic enzymes were normal.

On the abdominal CT, the jejunum was dilated without any anatomical obstacle. The MPD was dilated (18 mm in diameter at the level of the pancreatic isthmus), with 3 endoluminal nodules (2 in the head and 1 in the isthmus), measuring 9–16 mm in diameter each. These well-delineated nodules presented moderate contrast uptake after injection of contrast media. A 26-mm multilocular cystic lesion was found in the head of the pancreas. A fistula between the caudal part of the distal MPD and the gastric fundus was suspected (fig. 1). The pancreatic parenchyma appeared fibrotic and moderately atrophic. No metastases, enlarged lymph nodes or vascular abnormalities were described.

To further investigate the cystic lesion and its relationship with the MPD, an abdominal MRI was then performed which confirmed the global dilatation of the MPD, the presence of nodules within it and the 26-mm multilocular cystic lesion located in

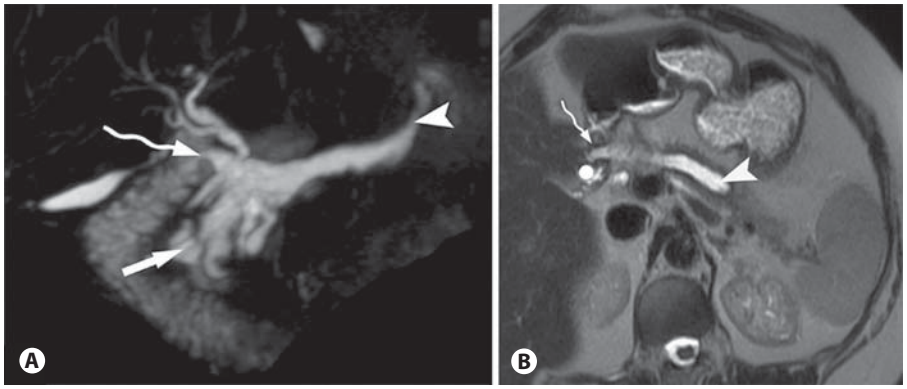

**Fig. 3. A** 3D sequence demonstrates the communication between the multilocular cystic lesion (straight arrow) of the pancreatic head and the MPD, the pancreaticoduodenal fistula (curved arrow) and the pancreaticogastric fistula (arrowhead). **B** Axial T2-weighted sequence shows the dilatation of the MPD (arrowhead) and the pancreaticoduodenal fistula (arrow). White disc: duodenum.

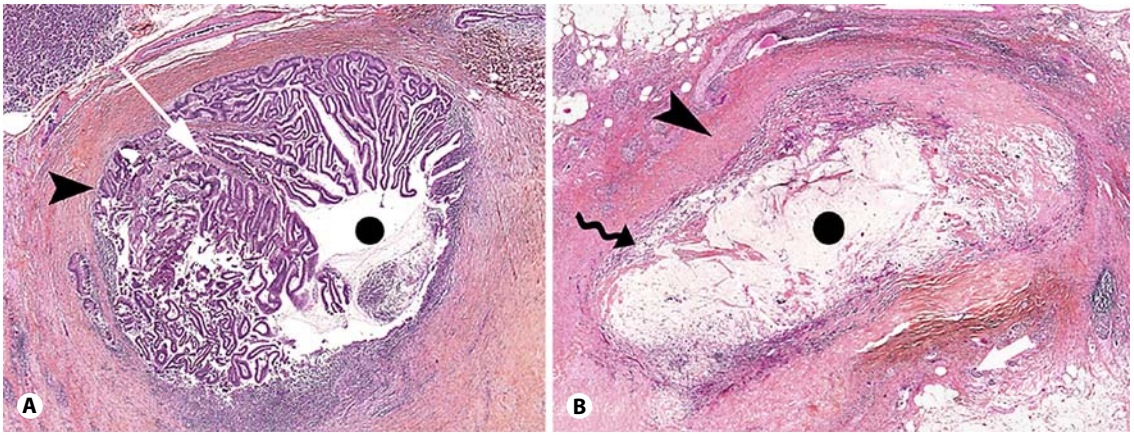

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**Fig. 4. A** Gross papilla formation (white arrow) mixed with mucin, occupying the dilated MPD lumen (black disc). The pancreatic parenchyma is not invaded (arrowhead). **B** In some places, peripheral mucin lakes (black disc) without tumor cells are found. Mucin lakes do not communicate with the MPD, and are surrounded by many inflammatory cells (black arrow), fibrous changes (arrowhead) and pancreatic islets (white arrow).

the head of the pancreas. Moreover, MRI showed solid nodules in the cyst that were characterized by sharp and regular limits, a hypointense signal on T2-weighted sequences, a slightly hyperintense signal on T1-weighted sequences, and moderate contrast uptake. On the heavily T2-weighted sequences, a 6-mm pancreaticogastric fistula was confirmed and MRI showed an additional pancreaticoduodenal fistula. MRI demonstrated clearly the presence of a communication between the MPD and the cystic lesion in the pancreatic head on the 3D sequences (fig. 2, 3). There was no mass effect on the biliary tree.

The imaging features suggested the diagnosis of a malignant IPMN. An extended cephalic pancreaticoduodenectomy was performed.

Histological examination of the surgical specimen showed a diffuse dilatation of the MPD with an intraductal papillary pattern and mucin production. The dilatation of the MPD and its branches formed a multicystic lesion, all cysts communicating between each other and with the MPD. There was no papillary pattern in the first 2 cm of the pancreaticoduodenal fistula nor in the caudal segment of the pancreas. The dilated ducts and the cyst were lined by columnar mucin-containing epithelial cells forming papillary structures and showing low-grade dysplasia. The papillary pattern was similar to that of villous adenomas of the gut indicating an 'intestinal-type' lesion (fig. 4). In some places, ductal ruptures were found with peripheral mucin lakes, but without tumoral cells.

Although the lesion did not invade the surrounding pancreatic parenchyma, it appeared diffusely pale and firm, with increased inflammatory cells and fibrotic changes, reflecting lesions of chronic pancreatitis. The duodenum was not invaded.

The final diagnosis was an 'intestinal-type' benign IPMN, with pancreaticogastric and pancreaticoduodenal fistulas, initially revealed by a self-limited intestinal obstruction. Three years after surgery, the patient was asymptomatic with no sign of recurrence.

## Discussion

To our knowledge, this is the first case of a proven benign IPMN associated with 2 fistulas between the MPD and surrounding digestive organs, these fistulas being diagnosed by MRI.

Although, CT showed a cystic dilatation of the MPD, nodules and a cystic lesion in the head of the pancreas, MRI allowed a more precise diagnosis of the lesions showing: an additional fistula, nodules and communication between the cystic lesion and the MPD. Considering that the cystic lesion was located on a branch duct but also involved the MPD, as shown by the MRI, we conclude that it is a combined type IPMN.

The second original feature in our case is the association of a benign IPMN with pancreaticodigestive fistulas. Nevertheless, in the initial reading of the MRI and CT, we overestimated the malignant signs: dilatation of the MPD to >10 mm, the presence of nodules in its lumen and fistulas. Consequently, we based the indication for a pancreaticoduodenectomy on these findings.

The radiological criteria reported to differentiate benign and malignant IPMNs are not standardized in the literature. Solid nodules inside the pancreatic ducts (PPV 91%), dilatation of the MPD (>6–10 mm; Sensitivity 87%), the presence of intracystic septa >2 mm in thickness, tumor size >30 mm for the branch-duct type, protrusion of the MPD into the ampulla of Vater, bile duct dilatation, intraductal calcifications and enlarged peripancreatic lymph nodes [2, 6] are all considered indicators of malignancy. By contrast Gupta et al. [3] consider only the two following criteria for malignancy: the main-duct type and the presence of a large communication between a branch duct-type lesion and the MPD. The presence of solid nodules is not considered a criterion for malignancy by these authors, as they found that only 50% of the lesions associated with nodules were malignant in their experience, while 25% were associated with noninvasive carcinoma and 25% with adenomas.

The presence of pancreaticogastric and pancreaticoduodenal fistulas was another feature suggesting a malignant lesion. Fistulas between the IPMN and the adjacent organs [4, 5, 7, 8] are usually associated with an invasive carcinoma and involve the duodenum (64%), the common bile duct (56%) or the stomach (17%) [5]. There are 5 reported cases of IPMN associated with a pancreaticogastric fistula in the literature. Four of these were malignant tumors and the 5th was a noninvasive carcinoma occurring after ERCP; in this case, it was assumed that the determining factor for the appearance of a fistula was the increase in pressure in the pancreatic duct during ERCP [4]. The physiopathological mechanism for the formation of pancreaticogastric and pancreaticoduodenal fistulas is not entirely known. For invasive carcinomas, the supposed mechanism may be a contiguous invasion [7]. By contrast, in our case, no sign of invasion was observed. Mucin lakes were present at the contact of branch ducts and may have increased the intraductal pressure, resulting in ductal dilatation and wall disruption, finally leading to the fistula [4, 5]. The chronic inflammatory pattern of the pancreatic tissue may also have contributed to the formation of fistulas [4]. This pattern has been described by Yamada et al. [9] as a rim-like enhancement.

## Conclusion

The present case report suggests that the accumulation of mucin inside the pancreatic ducts in the context of an IPMN may be responsible for the formation of pancreaticodigestive fistulas, without any sign of malignancy of the tumor itself. Cross-sectional noninvasive imaging, such as CT and MRI, are the most effective tools to investigate IPMNs complicated by fistulas, while more invasive techniques such as ERCP are not required. However, it still remains difficult to precisely define the radiological criteria of malignancy for IPMNs. Indeed, despite the presence of solid nodules, thick septa and dilatation of the MPD, the histological analysis of the surgical specimen revealed a benign tumor in our patient. Surgical decisions should thus be made cautiously in such patients on the basis of these imaging techniques and take into account the age, comorbidities and surgical risk.

## References


1 Ohashi K, Murakami Y, Maryuama M, et al: Four cases of mucous secreting pancreatic cancer. Prog Dig Endosc 1982;20:348–351.

2 Sahani DV, Kadavigere R, Blake M, Fernandez-Del Castillo C, Lauwers GY, Hahn PF: Intraductal papillary mucinous neoplasm of pancreas: multi-detector row CT with 2D curved reformations – correlation with MRCP. Radiology 2006;238:560–569.

3 Gupta R, Mortele KJ, Tatli S, et al: Pancreatic intraductal papillary mucinous neoplasms: role of CT in predicting pathologic subtypes. AJR Am J Roentgenol 2008;191:1458–1464.

4 Koizumi M, Sata N, Yoshizawa K, et al: Post-ERCP pancreatogastric fistula associated with an intraductal papillary-mucinous neoplasm of the pancreas – a case report and literature review. World J Surg Oncol 2005;3:70.

5 Okada K, Furuuchi T, Tamada T, et al: Pancreatobiliary fistula associated with an intraductal papillary-mucinous pancreatic neoplasm manifesting as obstructive jaundice: report of a case. Surg Today 2008;38:371–376.

6 Ogawa H, Itoh S, Ikeda M, Suzuki K, Naganawa S: Intraductal papillary mucinous neoplasm of the pancreas: assessment of the likelihood of invasiveness with multisection CT. Radiology 2008;248:876–886.

7 Jung IS, Shim CS, Cheon YK, et al: Invasive intraductal papillary mucinous tumor of the pancreas with simultaneous invasion of the stomach and duodenum. Endoscopy 2004;36:186–189.

8 Lee SE, Jang JY, Yang SH, Kim SW: Intraductal papillary mucinous carcinoma with atypical manifestations: report of two cases. World J Gastroenterol 2007;13:1622–1625.

9 Yamada Y, Mori H, Matsumoto S: Intraductal papillary mucinous neoplasms of the pancreas: correlation of helical CT and dynamic MR imaging features with pathologic findings. Abdom Imaging 2008;33:474–481.