
CASE REPORT

Spontaneous Pancreaticoduodenal Fistula is a Rare Complication of Intraductal Papillary Mucinous Neoplasm: a Case Report

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INTRODUCTION

Pancreatic cystic lesions are increasingly identified as cross-sectional imaging becomes more readily available. The prevalence of pancreatic cystic lesions is up to 49.5% in the general population.¹ In patients who undergo abdominal imaging, 13.5% are found to have incidental pancreatic cystic lesions.² Intraductal papillary mucinous neoplasm (IPMN) is among the most common pancreatic cystic lesions; it is characterised by the proliferation of mucin-secreting papillary epithelial cells with consequent dilatation of main or branch pancreatic ducts. Owing to its potential for malignant transformation, IPMNs are of particular interest to radiologists and clinicians alike. Although patients with IPMN are often asymptomatic, they can also present with abdominal pain, jaundice, weight loss and pancreatitis; fistulation to adjacent organs is a very rare complication of IPMN. We report a case of spontaneous pancreaticoduodenal fistula secondary to IPMN.

CASE REPORT

An 81-year-old man with a history of carcinoma of the rectum treated by laparoscopic anterior resection presented with intermittent epigastric pain for 1 week. He denied any symptoms of jaundice, pale stool, tea-stained urine, weight loss or fever. On examination he was afebrile and was not jaundiced; he had epigastric tenderness but no guarding. His blood results showed an elevated white cell count of $13.64 \times 10^9/L$ (normal range $3.7-9.3 \times 10^9/L$) with normal amylase, liver and renal function. He underwent a computed tomography (CT) abdomen and pelvis that demonstrated a diffusely swollen pancreas with homogeneous parenchymal enhancement and a grossly dilated main pancreatic duct, up to 1.3 cm at the pancreatic head (Figure 1). The pancreatic ductal system had a bifid configuration with a dominant duct of Wirsung. There was an abnormal communication between the main pancreatic duct and adjacent D1 segment of the duodenum (Figure 2). No

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Ethics Approval: The patient was treated in accordance with the tenets of the Declaration of Helsinki. The patient provided written informed consent for all treatments and procedures.

Declaration: This case report was presented as a poster at the 8th Joint Scientific Meeting of the Royal College of Radiologists and the Hong Kong College of Radiologists and 27th Annual Scientific Meeting of the Hong Kong College of Radiologists, held on 16-17 November 2019 in Hong Kong.



Figure 1. Computed tomography image of the abdomen showing a diffusely swollen pancreas with peri-pancreatic fat stranding. The main pancreatic duct is dilated along its entire course. The pancreas shows homogeneous parenchymal enhancement.



Figure 2. Computed tomography image of the abdomen showing an abnormal communication (arrow) between the main pancreatic duct at the pancreatic head and D1 segment of the duodenum.

intramural nodule was found within the dilated main pancreatic duct. Overall CT findings were suggestive of pancreaticoduodenal fistula secondary to IPMN. He was treated as a case of mild pancreatitis and discharged home. Subsequent magnetic resonance images of the pancreas showed similar findings to the CT with an abnormal communication between the pancreas and duodenum (Figure 3). There was a small number of dependent hypointense signals within the main pancreatic duct, suggestive of mucin. The pancreatic parenchyma showed diffuse T1 hypointense signals suggestive of chronic pancreatitis; no enhancing components were seen in the arterial or portovenous phases. The patient underwent endoscopic retrograde cholangiopancreatography as an



Figure 3. (a) T1-weighted magnetic resonance image of the pancreas showing diffuse hypointense signal suggestive of chronic pancreatitis. (b) T2-weighted magnetic resonance image of the pancreas showing abnormal communication (arrow) between the pancreas and duodenum.

outpatient that demonstrated an abnormal opening over the medial wall of D1 with mucus plugging and small internal papillary growth. This confirmed the diagnosis of pancreaticoduodenal fistula secondary to IPMN (Figure 4). Biopsy results revealed moderate dysplasia. His carbohydrate antigen 19-9 level was 44.8 U/mL (normal range <35 U/mL). Surgery was offered to the patient at the time but was declined given his advanced age.

On routine follow-up 15 months after his initial presentation, the patient was found to have grossly deranged liver function with an elevated bilirubin 64 $\mu\text{mol/L}$ (normal range 3-21 $\mu\text{mol/L}$), alkaline phosphatase 473 IU/L (normal range 47-168 IU/L) and alanine aminotransferase 254 IU/L (normal range <49 IU/L). CT abdomen demonstrated a new



Figure 4. Photograph taken during endoscopic retrograde cholangiopancreatography showing an abnormal opening at the medial wall of duodenum with communication with the main pancreatic duct. Mucin was noted at the opening with a small papillary growth.

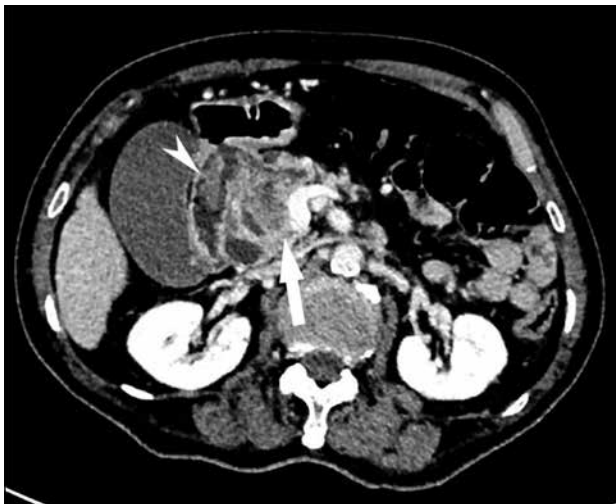


Figure 5. Computed tomography image of the abdomen 15 months after initial presentation showing a new irregular enhancing mass suggestive of carcinoma at the head and uncinate process of the pancreas with connection to the main pancreatic duct (arrow) and extension to the duodenum.

heterogeneous and irregular lesion with enhancing solid components at the pancreatic head and uncinate process (Figure 5) suggestive of carcinoma. The lesion was connected to the main pancreatic duct with suspicious extension to the duodenum via the fistula. The common bile duct was now grossly dilated to 1.9 cm with associated bilateral intrahepatic duct dilatation. Most recent carbohydrate antigen 19-9

was markedly elevated at 90.0 U/mL. The patient underwent internal external percutaneous transhepatic biliary drainage with subsequent biliary stent insertion and his liver function gradually improved. The lesion was deemed inoperable, and the patient subsequently received palliative radiotherapy.

DISCUSSION

IPMN is one of the most common pancreatic cystic lesions, classified according to its location: main duct IPMN, branch duct IPMN, or mixed type IPMN. They can also be classified according to varying degrees of dysplasia, ranging from benign adenoma to invasive carcinomas. Main duct IPMN is defined as diffuse or segmental dilatation of the main pancreatic duct >5 mm in the absence of other causes of obstruction. Main duct IPMN is known to have a higher risk of malignant transformation of up to 60% whereas the risk of malignancy of branch duct IPMN is 11%.^{3,4} Magnetic resonance cholangiopancreatography is currently considered the investigation of choice to evaluate pancreatic cystic lesions as it allows better visualisation of ductal communication, nodules and septae.⁵

Most IPMNs do not progress to carcinoma, but radiological risk stratification remains an important part of clinical assessment. Patients with lesions with high-risk features can be offered early surgical intervention or appropriate follow-up examination. The international consensus Fukuoka guidelines describe several features that suggest a high risk for malignancy, including a main pancreatic duct diameter ≥ 10 mm, enhancing mural nodule >5 mm and biliary obstruction. Patients with these features who are surgically fit should undergo surgical resection. Patients with worrisome features including cyst size ≥ 3 cm, enhancing mural nodule <5 mm, or main duct diameter 5 to 9 mm should undergo endoscopic ultrasound. Surgery should be considered in the presence of any of the following on endoscopic ultrasound: mural nodules ≥ 5 mm, main duct features suspicious of involvement, or cytology suspicious of or positive for malignancy. If no such feature is present, patients should be followed up with either CT, magnetic resonance imaging or endoscopic ultrasound, depending on the size of the lesion.⁵

Although many patients with IPMN remain asymptomatic, some present with symptoms such as abdominal pain or weight loss, or complications such as pancreatitis. Fistulation to the adjacent organs is a very rare complication of IPMN with only a few reports in

the literature. In a retrospective study of 423 patients with IPMN, 26 fistulas to adjacent organs were found in eight patients (1.9%).⁶ The majority of fistulas were found in the duodenum, followed by the stomach, common bile duct and the colon. The type of IPMN most commonly involved with fistulation was main duct IPMN, followed by mixed type IPMN and branch duct IPMN. Although the imaging findings of all patients with IPMNs and fistulations were suggestive of malignancy, histology from 27% of fistulas following surgery did not demonstrate atypia. It was previously thought that fistulations could only be found in malignant IPMNs, but studies have now shown that they can also be found in benign lesions.⁶⁻⁸ Hypotheses on the pathogenesis of fistula formation include excessive mucin production resulting in an increase in mechanical pressure on the pancreatic ducts or direct invasion by the primary tumour.^{9,10}

CONCLUSION

Fistulation to adjacent organs is a rare complication of IPMN and can occur in both malignant and benign lesions. The duodenum is the most commonly involved organ. Radiologists and clinicians should be aware of this potential complication since pancreatic cystic lesions including IPMN are increasingly detected due to wide availability of cross-sectional imaging. We present a rare case of spontaneous pancreaticoduodenal fistula secondary to IPMN that later progressed to carcinoma.

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