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Case Report

Intraductal papillary mucinous neoplasm mimicking a duodenal tumor*

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ABSTRACT

Intraductal papillary mucinous neoplasms of the pancreas are characterized by ductal dilatation, mucus secretion, and intraductal papillary growth. Intraductal papillary mucinous neoplasms can cause fistulation and extend to adjacent organs. However, they rarely present as large mass in the small bowel that causes bowel obstruction. Herein, we describe a case of intraductal papillary mucinous neoplasm that protruded into the duodenal lumen through the ampulla of Vater, presented as a large duodenal tumor, and developed duodenal obstruction. A 78-year-old woman was admitted to the emergency department with a 2-day history of vomiting and epigastric pain and 2 kg weight-loss in 2 months. Esophagogastroduodenoscopy showed a duodenal mass; however, the endoscope could not pass further, and visualization of the entire tumor was impossible. Computed tomography and magnetic resonance imaging revealed a heterogeneous enhancing mass measuring 8 cm in the second portion of the duodenum, which continued further as an intraductal mass of the pancreas. The main pancreatic duct was dilated, and the parenchyma was atrophied. The biopsy showed a well-differentiated adenocarcinoma and an intact overlying duodenal mucosa. Surgical option of treatment was offered to the patient; however, she refused it due to her advanced age and personal religious beliefs. Thus, we conclude that the experience and knowledge gathered through this patient regarding intraductal papillary mucinous neoplasms could provide further understanding of this disease and evolve subsequent patient care.

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Introduction

Intraductal papillary mucinous neoplasms (IPMNs) of the pancreas account for up to 25% of surgically resected neoplasms and up to 50% of incidentally detected pancreatic cystic lesions [1]. They can undergo malignant transformation and in-

vade the adjacent organs. However, IPMNs of the pancreas rarely form a large duodenal mass that extends through the major duct opening of the duodenum. Here, we present a case of pancreatic IPMN that protruded into the duodenal lumen, which presented as a large duodenal tumor and developed duodenal obstruction.

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Case report

A 78-year-old woman presented to the emergency department of our hospital with a 2-day history of vomiting and epigastric pain. She also reported a loss in weight by of 2 kg within the past 2 months. Moreover, 1 month prior, she experienced epigastric burning and nausea, which restricted her ability to eat anything except watery rice porridge. She also complained of occasional black stools. The patient appeared acutely ill; however, the abdomen was generally soft on physical examination. Laboratory examinations revealed decreased levels of hemoglobin, 8.4 g/dL [reference range; 12-16 g/dL]; hematocrit, 28.0% [reference range; 36%-48%], and elevated lipase level, 85 U/L [reference range; 0-67 U/L]. Esophagogastroduodenoscopy (EGD) demonstrated a large mass in the second portion of the duodenum; however, the endoscope could not pass through due to the obstruction caused by the mass occupying the duodenal lumen. Computed tomography (CT) revealed gastric distension and marked dilatation of the pancreatic main duct (up to 2.5 cm) from the head and tail of the gland associated with parenchymal atrophy. Multiple intraductal nodules were observed, which were mildly enhanced (Figs. 1A and B). Oblique coronal reconstruction images showed that the intraductal mass of the pancreas protruded through the major papilla and formed a large polypoid mass inside the duodenal lumen in its second portion (Fig. 2). Magnetic resonance imaging (MRI) was performed after removing the gastric contents using a Levin-tube (nasogastric tube), and there was no gastric distension observed in the CT. Further, MRI and magnetic resonance cholangiopancreatography (MRCP) demonstrated the presence of multiple frond-like, hypointense papillary projections within a dilated main pancreatic duct and in the second portion of the duodenum (Figs. 3A and B). The structure was diagnosed as an IPMN of the pancreas, which extended through the ampulla of Vater and formed a duodenal mass. Biopsy revealed a well-differentiated adenocarcinoma and an intact overlying duodenal mucosa (Figs. 4A and B). Surgical treatment was offered to the patient; however, she refused it due to her advanced age and personal religious beliefs. Therefore, we opted for close monitoring of the patient instead.

Discussion

IPMNs are pancreatic exocrine tumors composed of intraductal papillary growth of mucin-containing neoplastic cells in the pancreatic duct. IPMNs account for less than 10% of all pancreatic neoplasms [2], and their incidence has increased with the development of diagnostic techniques. Notably, 50% of the incidentally detected pancreatic cysts are IPMNs [1]. They usually occur in the sixth and seventh decades of life, and affects males more than females.

The clinical presentation of IPMN includes epigastric discomfort or pain, nausea, vomiting, weight loss, jaundice, backache, and diabetes. Radiological diagnostic tools for IPMN include ultrasonography (US), CT, MRI, and MRCP. CT or MRI are performed for further evaluation and differential diagnosis when pancreatic duct dilatation or cystic lesions are detected on ultrasonography. IPMN are classified into 3 types according to their site and extent of involvement: main duct type (MD-IPMN), branch duct type (BD-IPMN), and mixed or combined type. CT shows one or more cystic dilatations in the pancreas (BD-IPMN) or diffuse or segmental dilatation of the main pancreatic duct (MD-IPMN). This main duct type IPMN (MD-IPMN) is characterized by dilatation of the main pancreatic duct of >5 mm without any other cause of obstruction. This increases the sensitivity for radiological diagnosis of MD-IPMN; however, decreases the specificity [2]. According to guidelines, main pancreatic duct (MPD) dilatation of 5-9 mm is considered as a "worrisome feature" and MPD dilatation of \geq 10 mm is one of the "high risk stigmata" [3]. In addition, CT and MRI findings of high risk malignant lesions include a mass of >3 cm, presenting enhanced solid component, and metastasis of the lymph nodes [2]. In our case, the MPD dilatation of 2.5 cm, multiple enhanced mural nodules, and mass measuring of 8 cm indicate that the lesion is malignant. Recent reports have shown that MRCP is more sensitive than endoscopic retrograde cholangiopancreatography (ERCP) [2,4]. Furthermore, MRCP is not operator-dependent and is non-invasive. Furthermore, MRCP shows a high signal intensity of the whole dilated ductal system and cystic mass complex, even when the pancreatic duct is obstructed by mucin or mural nodules [5]. In

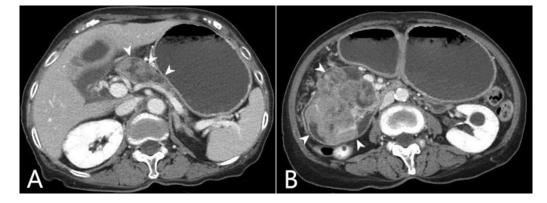


Fig. 1 – Contrast-enhanced axial computed tomography scans. (A) Mild-enhancing papillary nodules were observed in the dilated main pancreatic duct. Severe atrophic changes were observed in the pancreatic parenchyma. (B) A heterogeneous enhancing lobulated contoured mass of 8 cm was seen in the second portion of the duodenum.

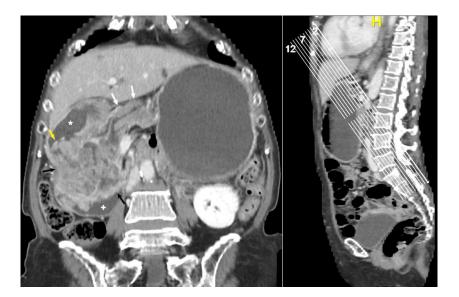


Fig. 2 – Contrast-enhanced oblique coronal reconstruction scans. The intraductal mass of the pancreas (white arrows) protruded through the major papilla and formed a large polypoid mass inside the duodenal lumen (star: first portion, cross: second portion of the duodenum) in its second portion (black arrows). Some overlying duodenal mucosae is intact (yellow arrow).

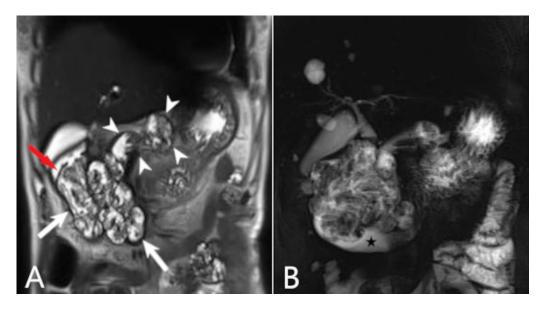


Fig. 3 – Coronal T2 magnetic resonance imaging and MRCP. (A,B) Multiple frond-like, hypointense papillary projections within a dilated main pancreatic duct (arrowheads) and duodenal second portion (white arrows) are observed. Some overlying duodenal mucosa was intact (red arrow). MRCP showed findings similar to those of T2 coronal MRI scans (star: lumen of the duodenum).

such cases, ERCP is often inadequate in visualization of the entire pancreatic ductal system.

IPMNs compose a large spectrum ranging from adenomas to invasive cancer with various degrees of severity [5]. IPMNs have malignant potential; currently, most authors agree that their evolution towards the carcinoma stage is slow and is the lag time between IPMN adenoma and IPMN with invasive cancer is estimated to be approximately 3-6.4 years [4,6].

IPMNs can extend, cause fistulation, and rupture adjacent organs. The pattern of rupture is classified into 2 types; tumor

invades surrounding tissue of fistula (60% of cases, invasive type) and the inner pressure of the pancreatic duct increases as the tumor grows or extends by itself (40% of cases, automatic type) [5]. Our patient experienced the latter type.

Surgical resection is the treatment of choice for IPMN, mostly for resection of MD-IPMN, mixed variants, and symptomatic BD-IPMN [2–4].

Malignant IPMN have a more favorable prognosis than ductal adenocarcinomas and are associated with a lower incidence (22%) of lymph node metastasis [4].

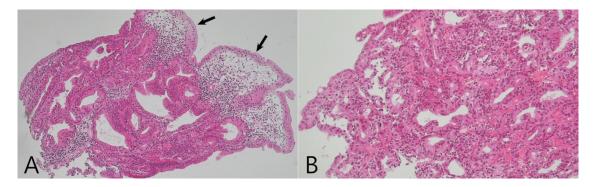


Fig. 4 – Histopathological findings. (A) The biopsy specimen showed a tumor with normal overlying duodenal mucosa (hematoxylin and eosin stain, \times 40, arrows). (B) The tumor was composed of columnar cells with rich cytoplasm and a back-to-back arrangement and were consistent with well-differentiated adenocarcinoma (hematoxylin and eosin stain, \times 100).

Conclusion

Thus, experience and knowledge regarding the variable findings of pancreatic IPMNs might aid in understanding the pathophysiology of this disease and improving patient care.

Patient consent

Consent for publication has been obtained.

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