Pathology – Research and Practice 212 (2016) 139–144

Contents lists available at ScienceDirect

Pathology – Research and Practice

journal homepage: www.elsevier.com/locate/prp

Teaching Case

# Intraductal papillary mucinous neoplasm involving pancreaticobiliary maljunction and an aberrant pancreatic duct draining into the stomach: A case report and review of the literature

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ARTICLE INFO

*Article history:*
Received 8 July 2015
Received in revised form 8 October 2015
Accepted 26 October 2015

*Keywords:*
IPMN
Aberrant pancreatic duct system
Ectopic opening
Congenital anomaly

ABSTRACT

Intraductal papillary mucinous neoplasms (IPMN) are characterized by the growth of epithelial components with mucin production in the main pancreatic duct, or a large branch. We report a case of intraductal papillary mucinous carcinoma (IPMC) of the pancreatic head, complicated by pancreaticobiliary maljunction (PBM) and an aberrant pancreatic duct draining into the stomach. A 50-year-old man with malaise and jaundice was found to have a mass in the pancreatic head at a local hospital. He was clinically diagnosed with IPMC with invasion to the stomach and duodenum after extensive endoscopic and radiological evaluation and a pancreaticoduodenectomy was performed. Histologically, most of the lesion exhibited proliferation of atypical glands, with irregular papillary and villoglandular structures lined by mucinous columnar epithelium, which extended intraepithelially along the dilated pancreatic duct wall. Tumor cells spread into the duodenal wall formed mucous nodules. The pancreatic ducts of this lesion uniquely showed two malformations; PBM, and an aberrant pancreatic duct opening to the gastric wall. This case was rare in the sense that IPMC extended into such unique structures. We report a case of IPMC with rare localization. The pathogenesis of this type of tumor in an abnormal pancreatic duct will be discussed.



## Case report

A 50-year-old man was evaluated at a local hospital because of malaise that had started 3 months earlier. A hen's egg-sized tumor was identified in the pancreatic head by an echogram. The patient was referred to our hospital for evaluation and treatment of a pancreatic tumor. His white blood cell count was 6300/μl; C-reactive protein 1.26 mg/dl; total bilirubin 14.7 mg/dl; direct bilirubin 11.9 mg/dl; aspartate aminotransferase 56 IU/l; alanine aminotransferase 45 IU/l; alkaline phosphatase 395 IU/l; γ-glutamyl transpeptidase 42 IU/l. Contrast-enhanced computed tomography scan showed a hypovascular mass in the pancreatic head measuring 3.7 × 3.7 cm. The tumor appeared to spread into the gastric wall and pancreatic duct (Figs. a and b).

Gastrointestinal endoscopy revealed villous or papillary tumors at the ampulla of Vater, minor duodenal papilla, and lesser curvature in the posterior wall of the pylorus (Fig. c–f). Pancreaticoduodenectomy was performed.

Macroscopic examination of the resected specimen showed the mass visible at the pylorus, minor duodenal papilla, and ampulla of Vater. At the cut surface, the specimen revealed a remarkably dilated main pancreatic and common bile duct conjoined 2.0 cm proximal to the duodenal wall, forming a common channel 2.0 cm long. This finding supported the diagnosis of pancreaticobiliary maljunction (PBM). Moreover, an aberrant pancreatic duct measuring 1.5 cm in length and 0.7 cm in diameter was found to connect the main pancreatic duct and the posterior wall of the pylorus. Fig. g is the schematic illustration of the distribution of the tumor in the pancreatic ducts and common bile duct. Histologically, the specimen showed that the papillary or villotubular tumor filled the main and branch pancreatic ducts and common bile ducts (Fig. h). Both the pancreatic and common bile duct were dilated and filled with tumor cells. These two structures formed a common channel (Fig. i). An aberrant pancreatic duct passed through the gastric wall and formed an orifice opening onto the mucosa of the pylorus (Fig. j). The aberrant pancreatic duct showed normal ductal structure surrounded by fibromuscular tissue, and was lined by non-neoplastic

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http://dx.doi.org/10.1016/j.prp.2015.10.010
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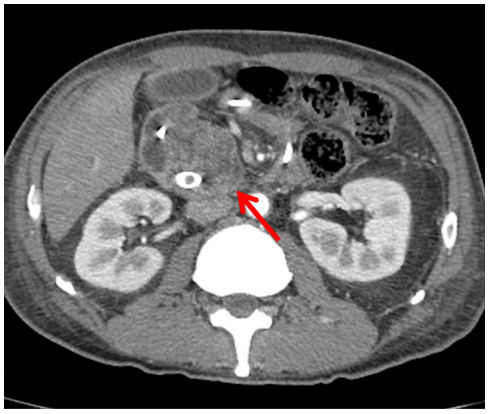

**Fig. a.** Contrast-enhanced *CT scan* showed a *hypovascular mass* in the *pancreatic head measuring* 3.7 × 3.7 cm in size (arrow).

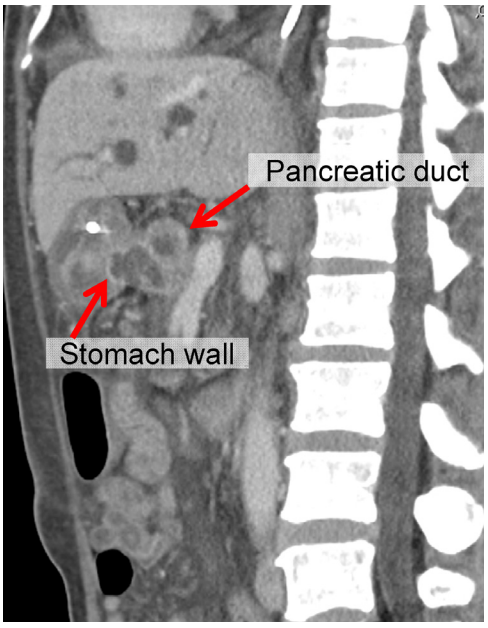


**Fig. b.** The tumor appeared to spread into the gastric wall and pancreatic duct (arrow).

epithelium in the lumen (Fig. k). The tumor mainly showed intraductal growth, and was lined by atypical columnar cells with mucin production. The tumor was mainly composed of well-differentiated adenocarcinoma (tub1), with cellular and structural atypia, while an adenomatous component was also observed in part (Fig. h, inset). Well-differentiated adenocarcinoma invaded in the muscular layer of the duodenum forming mucous lakes (Fig. l).

Immunohistochemically, the tumor cells were positive for AE1+3, CK7, CK20, EMA, CAM5.2, CK18, CK19, MUC5ac, and MUC2, partly positive for MUC6, weakly positive for CDX2, and negative for MUC1. Immunohistochemical phenotype of the tumor cells of the aberrant pancreatic duct were MUC1−/MUC2+/MUC5AC+/MUC6−, which were different from the epithelial cells of the gastric mucosa (Fig. m). Accordingly, we diagnosed this case as intraductal papillary mucinous carcinoma (IPMC), invasive, derived from intraductal papillary mucinous neoplasm (IPMN). Immunohistochemically, phenotype of the mucin was intestinal and gastric-foveolar type.

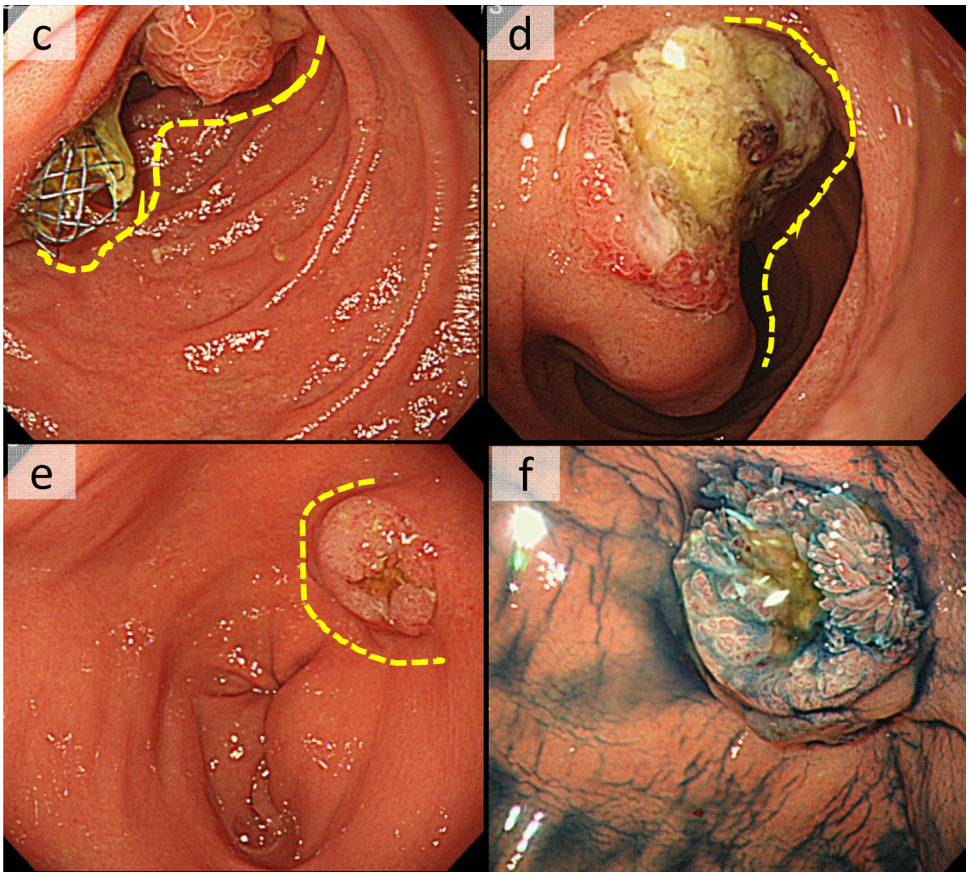


**Fig. c–f.** Gastrointestinal endoscopy revealed villous or papillary tumors at the papilla of Vater (c), minor duodenal papilla (d) and lesser curvature in the posterior wall of pylorus (e and f).