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An Unusual Cause of Abdominal Pain

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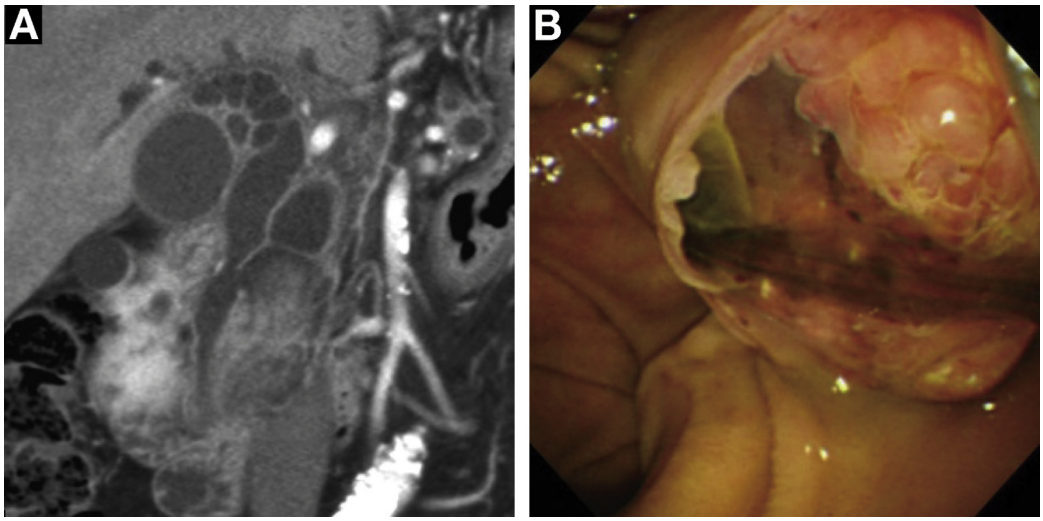

Question: An 88-year-old woman was admitted to our hospital complaining of upper abdominal and back pain. She had a history of hypertension. On admission, she was febrile with a temperature of 38.4°C, blood pressure was 122/54 mmHg, and her pulse rate was 84 beats per minute. Her abdomen was soft, nondistended, and nontender. Admission blood work revealed a white blood cell count of 7,800/mm^3. Her liver panel showed a total bilirubin of 1.1 mg/dL (normal, 0.1-1.1); alanine aminotransferase, 62 IU/L (normal, 3-49); aspartate aminotransferase, 47 IU/L, (normal, 9-37); alkaline phosphatase, 438 IU/L, (normal, 104-338); and γ-glutamyl transpeptidase, 168 IU/L (normal, 6-71). Tumor markers were normal: carcinoembryonic antigen, 2.4 ng/mL (normal, <5.0) and carbohydrate antigen 19-9, 17 U/mL (normal, <37). To evaluate further the abnormal liver enzymes, abdominal ultrasound followed by a contrast-enhanced CT of the abdomen was performed (Figure *A*). Side-viewing endoscope showed duodenal papilla (Figure *B*).

What are the findings of the abdominal CT and side-viewing endoscope and what is the diagnosis?

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Conflicts of interest
The authors disclose no conflicts.

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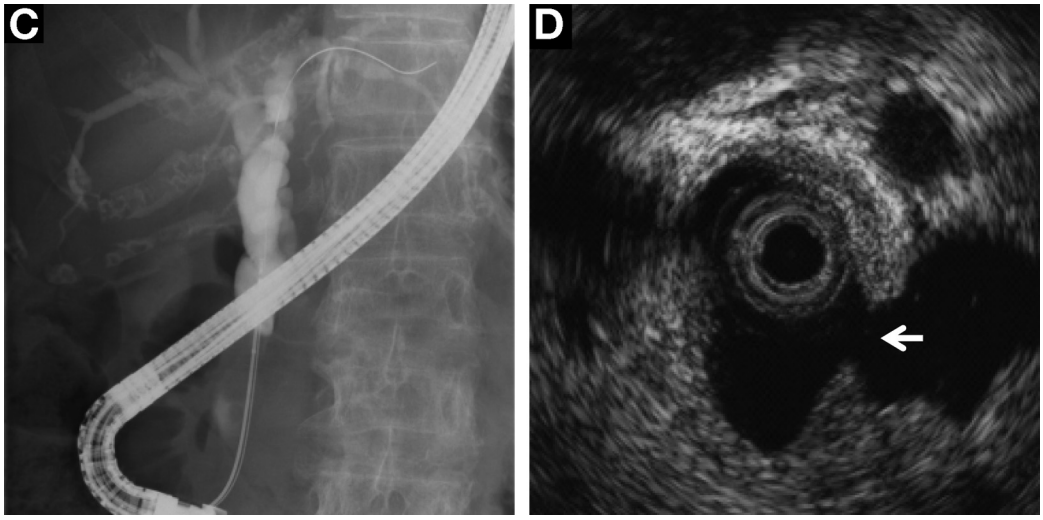
Answer to Image 1: Pancreatobiliary Fistulas Associated With Intraductal Papillary Mucinous Neoplasm



CT showed a tumor with papillary growth in the pancreatic head associated with a dilated main pancreatic duct, and a suspected fistula connecting to the common bile duct (CBD; Figure *A*). Duodenal papilla images revealed dilated pancreatic and bile ducts with mucin extrusion (we call this the 'pig-nose appearance'; Figure B). Endoscopic retrograde cholangiopancreatography revealed a dilated biliary tree with an extensive filling defect in the distal CBD (Figure *C*). Intraductal ultrasonography confirmed a distinct pancreatobiliary (PB) fistula (Figure *D, arrow*).

The patient underwent a pylorus-preserving pancreatoduodenectomy, resulting in a final diagnosis of pancreatic ductal adenocarcinoma derived from intraductal papillary mucinous neoplasm (IPMN). She has been doing well for 18 months after surgery. Fistula formation between the pancreas and adjacent organs (duodenum, CBD, stomach) has been reported in $\leq$6.6% of IPMN cases.[1,2] In addition to the duodenal papilla of IPMN described as a "fish-mouth" appearance,[3] the "pig-nose" appearance of the duodenal papilla, in which both the bile and pancreatic ducts are dilated, may be a unique signature of PB fistulas associated with IPMN.

References

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