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Degenerate Intraductal Papillary Mucinous Carcinoma with Fish-Mouth Appearance of the Papilla

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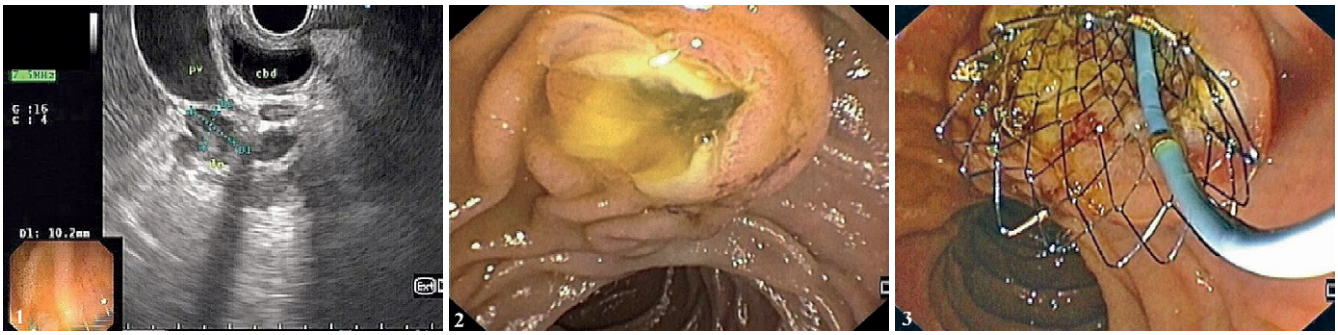

A 68-year old man with chronic alcoholic pancreatitis, presented to the emergency department for abdominal pain and jaundice. Laboratory tests revealed marked hyperbilirubinemia and cholestasis. Abdominal ultrasound showed dilatation of the common bile duct with apparent distal stenosis. Abdominal CT revealed the presence of an 11 mm hypodense cephalopancreatic lesion, with cystic areas, and involvement of the hepatic artery. Adjacent suspicious enlarged lymph nodes were observed. A transendoscopic ultrasonography was then performed confirming the presence of a multiloculated cystic lesion with a solid component, responsible for dilatation of the common bile duct and Wirsung's duct (Fig. 1).

An EUS-guided fine needle aspiration was done for further characterization. For the relief of jaundice it was decided to perform ERCP. With the side-view endoscope a patulous ampulla of Vater with extruding mucus was observed, the so-called, "fish-mouth sign" (Fig. 2), pathognomonic for a main branch intraductal pancreatic mucinous neoplasm (IPMN). Given the invasion of local structures, malignant transformation of the IPMN was assumed, and a short self-expanding metal stent was placed for drainage of the common bile duct (Fig. 3), which allowed neoadjuvant therapy.

The IPMNs arise from the main pancreatic duct and/or branch ducts, and are characterized by intraductal papillary proliferation of mucin-producing epithelial cells exhibiting various degrees of dysplasia [1]. They have a potential for malignant transformation with a well-described adenoma-carcinoma sequence [2]. Evaluation involves CT or MRI of the abdomen, ERCP and EUS [3]. Rarely, on endoscopic evaluation, the ampulla of Vater is described as having the pathognomonic „fish-mouth" appearance, which is the mucinous material being extruded from the pancreatic duct into the duodenum. It is the high viscosity of this mucinous fluid that obstructs the pancreatic duct and may cause pancreatitis.

In our patient, the lesion had progressed to carcinoma with involvement of the adjacent structures.

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Conflicts of interest: None to declare.

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