Intraductal papillary mucinous neoplasms (IPMNs) are one of several types of mucinous tumors of the pancreas. They are uncommon ductal epithelial tumors, comprising approximately 10% to 15% of cystic pancreatic neoplasms. IPMNs can be classified into three types – main pancreatic duct IPMN (MD-IPMN), branch duct IPMN (BD-IPMN), and mixed – based on imaging studies (computed tomography or magnetic resonance imaging [MRI] with magnetic resonance cholangiopancreatography [MRCP]) and/or histological examination with endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) [1–5]. MD-IPMN is characterized by segmental or diffuse dilatation of the main pancreatic duct (MPD) of more than 5 mm without other causes of obstruction [2].

A 70-year-old man was transferred to our department on an emergency basis with abdominal pain and fever up to 40°C that had lasted for 1 week. Computed tomography and MRI showed the pancreatic duct dilated to 40 mm (Fig. 1a). In the tail, the dilatation was to 60 mm (Fig. 1b). On endoscopic retrograde cholangiopancreatography (ERCP), mucinous material was seen to bulge out from the dilated ampulla of Vater – an uncommon but essentially pathognomonic sign of IPMN (Fig. 2).

After inspection of the MPD with an extraction balloon, mucinous material and pus were evacuated (Fig. 3). We placed a 9-Fr nasopancreatic drain and withdrew aspirate for microbiological and cytological examination, which showed the presence of polymorphonuclear leukocytes, Gram-negative Pseudomonas aeruginosa, and Gram-positive Enterococcus faecalis. We tried to drain the pancreatic duct, but the mucinous material was too thick, and the procedure was ineffective.

After we had stabilized the patient’s condition, on day 5 we performed a second endoscopic procedure – pancreatic sphincterotomy and peroral pancreatoscopy (POPS) – with a pediatric gastroscopy. Polypoid structures were noted in the distal 30 mm of the MPD (Fig. 4a, b). At the level of the corpus and
tail, the duct was extremely dilated and filled with mucinous material. Biopsy showed IPMN with borderline malignancy. At the end of the procedure, we placed two 10-Fr, 12-cm double-pigtail stents in the MPD (Video 1). After the sepsis had resolved, we referred the patient for surgery.

We have presented a case of MD-IPMN with extreme dilatation of the MPD complicated by pancreatic empyema and sepsis. Drainage procedures and POPS facilitated the diagnosis and successful preparation for surgery. POPS with a pediatric gastroscope allowed a precise preoperative evaluation of the MPD and adequate histological confirmation.

According to published series of cases, the mean frequency of malignancy in MD-IPMN is 61.6%, and the mean frequency of invasive IPMN is 43.1% [3]. No factors consistently predictive of malignancy in MD-IPMN have been identified, including the degree of MPD dilatation, presence of symptoms, and presence of mural nodules [4]. The first cases of IPMN were reported in the 1970s and 1980s. In the 1990s, the term intraductal papillary mucinous neoplasm was coined, and the tumor was established as a distinct entity among pancreatic neoplasms. The Tanaka criteria for the management of IPMN and mucinous cystic neoplasm (MCN) of the pancreas were published in 2012 [5].

Endoscopy_UCTN_Code_TTT_1AO_2AI

Competing interests: None

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DOI http://dx.doi.org/10.1055/s-0034-1393654
Endoscopy 2015; 47: E598–E599
© Georg Thieme Verlag KG
Stuttgart · New York
ISSN 0013-726X

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