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The Role of Endoscopy for the Diagnosis of Intraductal Papillary Mucinous Tumor of the Pancreas

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Intraductal papillary mucinous tumor is a recently recognized rare pancreatic tumor. Because the tumor includes a broad spectrum of malignancy grade, preoperative assessment of disease extent and differentiation between benign and malignant forms are essential. Among a variety of diagnostic modalities, endoscopy plays important roles to assess intraductal papillary mucinous tumor. ERCP is a traditional diagnostic tool and enables pancreatic juice sampling for cytology and guided forceps biopsy. EUS is an accurate modality for the diagnosis of intraductal papillary mucinous tumor. Certain endosonographic features are highly indicative of a malignancy. The addition of fine needle aspiration capability further enhances diagnostic accuracy. IDUS can be used for a more detailed evaluation of intraductal papillary mucinous tumor in assessing the intraductal spread of the tumor and its pancreatic parenchymal invasion. Pancreatoscopy makes possible a definite diagnosis of intraductal papillary mucinous tumor allowing biopsy under direct vision and provides valuable information in assessing the extent of the lesion. Newly developed video pancreatoscope makes it possible to make a more detailed morphologic assessment of the tumor and its grade of malignancy. With a combination of these endoscopic modalities, intraductal papillary mucinous tumor can now be effectively imaged and diagnosed preoperatively.
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Intraductal papillary mucinous tumor (IPMT) has been recently recognized as a unique pancreatic tumor with an indolent biologic behavior and favorable prognosis.[1] The tumor spreads along the pancreatic duct, replacing the normal epithelium,[2] and includes a broad spectrum of histopathologic disorders, such as hyperplasia, adenoma, and adenocarcinoma.[3] It is documented that the development of IPMT includes the so-called adenoma–carcinoma sequence, where the ultimate form of malignant progression is invasive carcinoma.[3,4]

IPMT often presents with acute pancreatitis of mild to moderate severity. It has been reported that approximately one-fourth of patients with IPMT experience symptoms including epigastric discomfort and/or pain and backache.[5,6] Diabetes, steatorrhea, and jaundice are the possible symptoms according to the progression of the disease. However, two-thirds of patients with IPMT do not display any symptoms. In cases like these, IPMTs may be discovered during routine workup for other diseases or at autopsy.

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IPMTs are usually classified into three types: main duct type (MDT), branch duct type (BDT), and combined type, according to the site and extent of involvement.[7,8] MDT-IPMT is characterized by a diffusely or partially dilated main pancreatic duct filled with excessive mucin. The inner surface of the dilated duct frequently contains mural nodules. BDT-IPMT affects one or more branches of the pancreatic duct, which consequently show cystic dilation. The dilated branch duct may contain solitary or multiple tumors and/or viscid mucin. Any combination of the above two types of IPMT is designated as combined-type IPMT. In combined-type IPMT, in addition to the presence of BDT-IPMT, the main pancreatic duct contains papillary growth of the tumor that produces excessive mucin.

Distinction of IPMT from chronic pancreatitis is of clinical significance, especially in patients with MDT-IPMT. IPMT is typically easily distinguishable from other tumors, such as serous cystadenomas, solid-pseudopapillary tumors, acinar cell cystadenocarcinomas, and cystic endocrine tumors. However, the distinction between IPMT and the mucinous cystic neoplasm of the pancreas (MCN) remains to clarified. The definition and classification proposed by the World Health Organization[9] and the Armed Forces Institute of Pathology[10] have provided considerable guidance.

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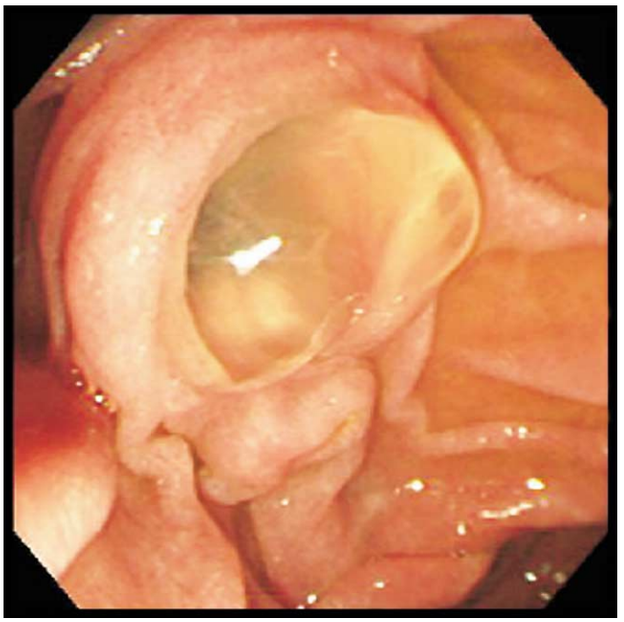

Figure 1 A patulous ampullary orifice with mucus secretion in a patient with a MDT-IPMT. (Color version of figure is available online.)

The Japan Pancreas Society performed a multi-institutional, retrospective study of 1379 patients with IPMT. Clinicopathological features and postoperative long-term outcomes were investigated. IPMT was most frequently found in men and in the head of the pancreas. Prognostic indicators of malignant IPMT included advanced age, presence of symptoms, abundant mucus secretion, presence of large nodules and/or large cysts, marked dilation of the main pancreatic duct, and main duct- or combined-type IPMT. The 5-year survival rate of IPMT patients after resection was 98% to 100% in adenoma to noninvasive carcinoma cases, 89% in minimally invasive carcinoma cases, and 58% in invasive carcinoma cases.[11]

IPMT may now be effectively imaged and diagnosed with a variety of modalities, including transabdominal ultrasound (US), helical computed tomography (CT), magnetic resonance imaging (MRI), or magnetic resonance cholangiopancreatography (MRCP). However, endoscopic diagnostic tools, such as endoscopic retrograde cholangiopancreatography (ERCP) and endoultrasonography (EUS), are also indispensable for making a therapeutic strategy for IPMT. More recently, intraductal ultrasonography (IDUS) and pancreatoscopy have been introduced to evaluate IPMT and have played an important role in assessing the grade of malignancy and the extent of the lesion preoperatively. In this chapter, the role of endoscopic diagnostic modalities for IPMT is discussed, especially focusing on techniques and usefulness of a more recent modality of pancreatoscopy.

ERCP

The classic description of IPMT, the presence of mucin extruding from a patulous ampulla, is most commonly seen in the presence of an advanced intraductal neoplasm[1,12,13] (Fig. 1). It is this endoscopic finding that initially brought this disease to our attention. This unique widening of the orifice is sometimes observed at the minor ampulla and/or the major ampulla. A growing awareness of this indication of IPMT has resulted in an increasing number of patients diagnosed during routine upper gastrointestinal endoscopy in Japan.[14] ERCP typically reveals any dilation of the main pancreatic duct or branches with filling defects due to the presence of either mural nodules or mucin. Full visualization of the ductal system in either MDT- or BDT-IPMT by ERCP is frequently difficult due to the presence of vicid mucin. MRCP is the preferred method of use to visualize the entire outline of the ductal system in such cases (Fig. 2A and B). Communication between BDT-IPMT and the main pancreatic duct is usually evident. ERCP may fail to detect branch ductal ectasia in up to 12% of patients because of abundant intraductal or intracystic mucin.[15,16] It can be useful to aspirate the mucin and to use a balloon catheter to occlude the main duct during injection of contrast medium to preclude reflux into the duodenum.[17,18]

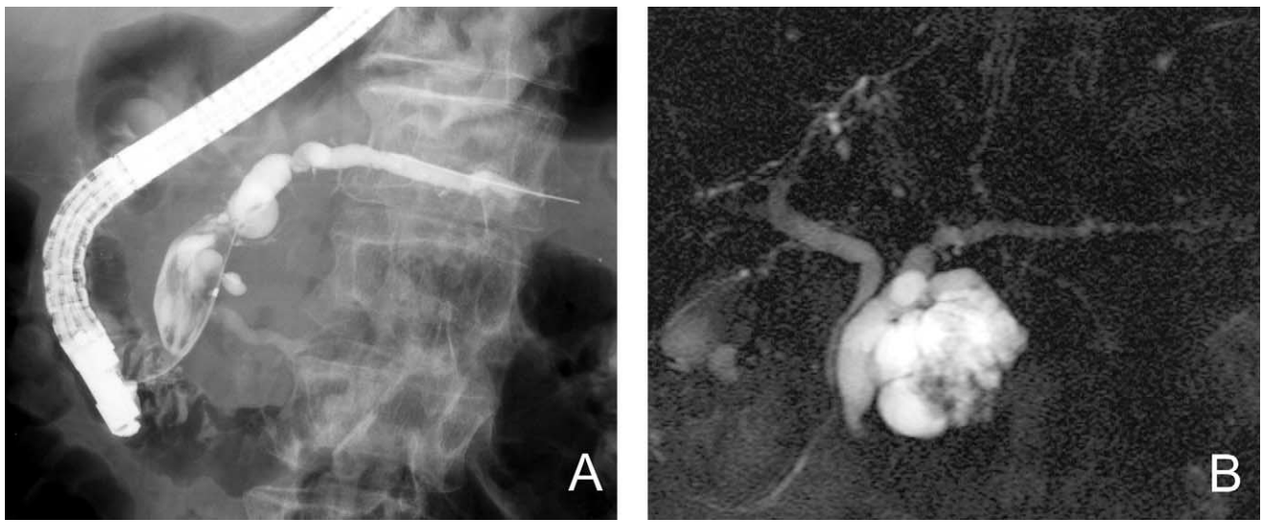

Figure 2 A case of BDT-IPMT. (A) ERCP showing a dilation of the main pancreatic duct with a filling defect due to mucin at the pancreas head. Dilated branch ducts are not fully opacified. (B) MRCP (the same case). A marked dilation of branch ducts are visualized.