



# Obstructive cholangitis by mucus from an intraductal papillary mucinous neoplasm with pancreatobiliary fistula treated by endoscopic septotomy and direct peroral cholangioscopy: a case report

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## Abstract

Intraductal papillary mucinous neoplasms (IPMNs) occasionally form a fistula to adjacent organs, resulting in obstructive jaundice and cholangitis due to mucus obstruction. Although some procedures such as endoscopic nasobiliary drainage are attempted, they often do not work adequately because of high mucus viscosity. Herein, we report the case of an 87-year-old man with obstructive cholangitis treated by endoscopic septotomy and mucus suction with direct peroral cholangioscopy using conventional endoscopy. The patient incidentally showed a branched-type IPMN in the pancreatic head on computed tomography (CT) approximately 10-years ago. Although the patient's tumor had grown slowly and he occasionally developed cholangitis, he did not want surgery. He was admitted to our hospital because of cholangitis by mucus obstruction with a PB fistula. Endoscopic retrograde cholangiopancreatography (ERCP) and septotomy were performed. Septotomy made the duodenal papilla a large orifice, thereby facilitating spontaneous drainage of mucus. In addition, conventional endoscopy with a large working channel enabled direct access into the orifice and smooth mucus suction, thereby alleviating his cholangitis. In conclusion, septotomy and direct peroral cholangioscopy using conventional endoscopy could be useful to control biliary tract infection and obstructive jaundice due to mucus obstruction from an IPMNs with PB fistula.

**Keywords** Pancreatobiliary fistula · Intraductal papillary mucinous neoplasm · Endoscopic septotomy · Direct peroral cholangioscopy

## Introduction

Intraductal papillary mucinous neoplasms (IPMNs) are defined as intraductal papillary growth tumors with mucin secretion. In general, these tumors grow slowly. However, surgery should be considered for patients with “high-risk stigma” (obstructive jaundice with a pancreatic head cystic lesion, an enhanced mural nodule  $\geq 5$  mm, main pancreatic duct (MPD) size  $> 10$  mm) or “worrisome features” (a cyst of  $\geq 3$  cm, an enhancing mural nodule  $< 5$  mm, thickened,

enhanced walls, MPD 5–9 mm) because these tumors can become malignant and increase the risk of conventional pancreatic ductal adenocarcinoma (PDAC) [1]. Occasionally, IPMNs form a fistula to adjacent organs, consequently resulting in obstructive jaundice and cholangitis due to mucus obstruction when the pancreatic duct communicates with the common bile duct (CBD), thereby causing difficulty in the treatment of these patients.

Herein, we report the case of cholangitis by mucus obstruction from an IPMN accompanied by a pancreatobiliary (PB) fistula successfully treated by endoscopic septotomy and mucus suction with direct peroral cholangioscopy using conventional endoscopy.

The patient provided informed consent for this case report.

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## Case report

An 87-year-old man was admitted to our hospital with a 3-day history of fever and abdominal pain. He smoked 15 cigarettes a day from 20 to 50 years of age and consumed 180 mL of sake per day for 67 years. He underwent distal gastrectomy and Billroth-I reconstruction at the age of 62 because of gastric cancer. Incidentally, a branched-type IPMN was observed at the pancreatic head on computed tomography (CT) about 10-years ago. Although the patient's tumor had grown slowly and become a mixed-type (defined as MPD > 5 mm and cysts > 5 mm that communicate with the MPD) tumor and he occasionally developed cholangitis, he did not agree to surgery. About a year ago, he developed obstructive cholangitis and CT demonstrated a PB fistula between the dilated pancreatic duct and common bile duct (CBD; Fig. 1). Then he underwent endoscopic sphincterotomy (EST) and cholangitis improved.

His blood pressure on admission was 117/95 mmHg, pulse rate 68 beats/min, and body temperature 38.0 °C. His palpebral conjunctiva was icteric, and the abdomen was soft and flat with mild tenderness. Laboratory analysis revealed the following: white blood cell count, 9500/mm<sup>3</sup>; C-reactive protein, 8.69 mg/dL; total bilirubin, 4.08 mg/dL; aspartate aminotransferase, 117 U/L; alanine aminotransferase,

72 U/L; alkaline phosphatase, 2288 IU/L; and gamma-glutamyltransferase, 361 U/L. Nonenhanced CT still revealed a dilated CBD accompanied by a PB fistula similar to the CT 1 year ago. Therefore, he was started on the antibiotic, ceftriaxone (2 g/day) on the first day of admission after the diagnosis of acute cholangitis.

However, his condition did not improve despite the administration of antibiotic, and he underwent endoscopic retrograde cholangiopancreatography (ERCP) on the sixth hospital day. His duodenal papilla was markedly dilated, showing a “pig-nose appearance,” and was obstructed by greenish gelatinous mucus (Fig. 2a). We attempted mucus suction and removal through a duodenoscopy and a balloon catheter, respectively, but this attempt was unsuccessful. Further, we tried to insert a conventional endoscopy into the papilla, but failed as well. He underwent a second ERCP on the 14th hospital day because of prolonged jaundice, although he had become afebrile after the first ERCP. Because mucus suction was unsuccessful through duodenoscopy, the septum between the pancreatic duct and CBD was cut with a bow-knife catheter to make the duodenal papilla a single, large orifice (Fig. 2b). Mucus suction became easier as forward-viewing conventional endoscopy with a large working channel allowed access into the duodenal papilla following septotomy. Then, a large amount of discharge along with bile was drained (Fig. 2c), and the cholangitis was alleviated. No tumors were observed in the CBD. He underwent a third ERCP on the 21st hospital day because he was febrile again. The ERCP revealed obstruction of the duodenal papilla with copious mucus. The mucus in the CBD was removed by suction through conventional endoscope, and no pus was observed. Subsequently, mucus in the dilated main pancreatic duct was removed and white pus was discharged (Fig. 3a), suggesting pancreatic ductitis. We could observe the inside of the dilated pancreatic duct, and a specimen from the villous tumor on the wall of the pancreatic duct was obtained (Fig. 3b); pathology demonstrated an adenoma.

The patient's condition improved immediately after the third ERCP and the patient left the hospital on the 27th hospital day. The post-treatment course has been uneventful for more than a year.

## Discussion

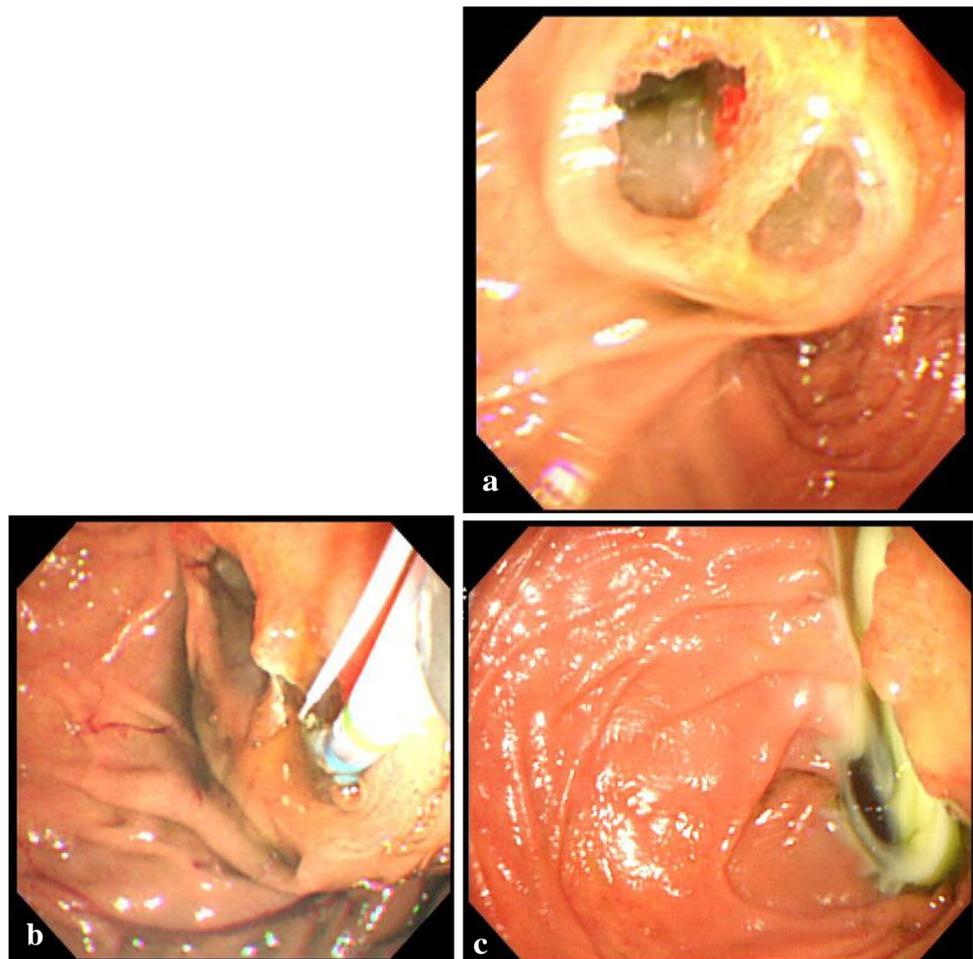
In this case of cholangitis due to mucus obstruction from an IPMN, accompanied by a PB fistula in an elderly man, the CT had revealed the IPMN 10 years ago, but he had not agreed to surgery.

IPMNs typically progress slowly but surgery is recommended for patients with “high-risk stigma” or “worrisome features” because these tumors may become malignant or

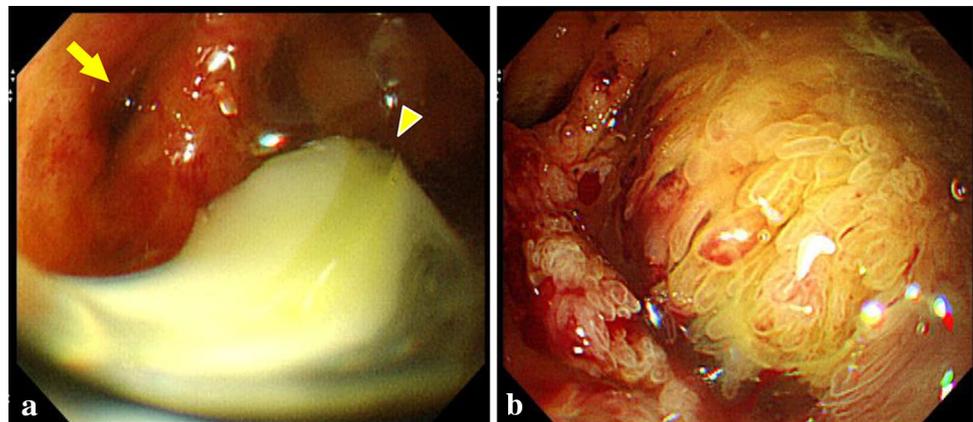


**Fig. 1** Enhanced computed tomography demonstrated a pancreaticobiliary fistula between the dilated pancreatic duct and common bile duct (yellow arrow)

**Fig. 2** Endoscopic retrograde cholangiopancreatography (ERCP) revealed a dilated duodenal papilla, with a pig-nose appearance (**a**). The septum was cut by a bow-knife catheter during the second ERCP (**b**). The pus was drained from the common bile duct (**c**)



**Fig. 3** The third endoscopic retrograde holangiopancreatography revealed no pus in the common bile duct (yellow arrow), but showed white pus in the dilated pancreatic duct (yellow arrow head) (**a**). The villous tumor existed on the wall of the pancreatic duct (**b**)



accompany a PDAC [1]. IPMNs typically develop in the elderly, and a follow-up observation is often performed for surgical stress. However, complications such as pancreatitis with mucus obstruction or fistula formation to adjacent organs occasionally develop during the follow-up.

The mechanisms of connection to the adjacent organs include (1) direct invasion of the IPMN into the adjacent

organs, (2) perforation into the adjacent organs caused by elevated pressure in the pancreatic duct due to unregulated mucin production, and (3) mucus migration across the mucinous lake in the stroma [2]. Kobayashi et al. reported that penetration was observed in the duodenum (67%), stomach (44%), CBD (33%), colon (6%) and small intestine (6%) (overlapping results) [3]. Obstructive jaundice

and cholangitis occur due to obstruction by the mucus that passes through the fistula to the CBD.

Yamaguchi et al. reported that the mean survival rate of the resected IPMNs with PB fistula and the unresected IPMNs with PB fistula was 47.9 and 10.4 months, respectively. The causes of death due to unresected IPMNs were mainly cancer, liver failure, or cholangitis. Therefore, appropriate treatment of these causes may improve the prognosis of the unresected IPMNs with PB fistula [4]. Although alternative procedures such as following are attempted if resection of the IPMNs with PB fistula is difficult, the efficacy of these procedures is unsatisfactory. The self-expandable metallic stent cannot cover the fistula completely and is often occluded or displaced [5]. Intermittent drainage through endoscopic nasobiliary drainage [6] or percutaneous transhepatic biliary drainage [7] is useful to improve obstructive jaundice, but it is not feasible for long-term use because of its narrow lumen. Notably, palliative techniques such as choledochojejunostomy may be an alternative, if possible [8].

In our case, despite post-EST papilla, mucus suction and removal from the CBD through a duodenoscopy and a balloon catheter was unsuccessful, respectively. Therefore, the septum between the pancreatic duct and CBD of the dilated duodenal papilla, which had a “pig-nose appearance,” [9] was cut by a bow-knife catheter through duodenoscopy to facilitate spontaneous mucus drainage. We presumed that septotomy facilitate spontaneous discharge of mucus for following two reasons. First, the large orifice enables easy mucus discharge. Second, mucus is expected to move into the duodenum easier rather than the bile duct by septotomy that reach the fistula and make common channel large. Furthermore, following septotomy, a conventional endoscopy allowed direct access into the dilated duodenal papilla and facilitated mucus suction because of a large working channel and forward-viewing specification.

Sung et al. have reported that direct peroral cholangioscopy is useful for diagnosing IPMNs with PB fistulas [10]. Recently, direct peroral cholangiography using the mother–baby scope system, ultra-slim endoscopy, and Spy-Glass™ (Boston Scientific, Natick, USA) has been widely used since the image quality and operability have improved [11]. However, because these endoscopes have a small working channel, procedures through the endoscope are restricted and suctioning highly viscous mucus is often difficult. The conventional endoscope (GIFH260, Olympus, Tokyo, Japan) we used, has a 2.8-mm diameter working channel, which enabled smooth mucus suction compared to an ultra-slim endoscope (e.g., GIFPQ260, Olympus, Tokyo, Japan) which has a 2.0-mm diameter working channel. Brauer et al. and Jung et al. reported that usefulness of argon plasma coagulation and endoscopic mucosal resection to the tumor of the bile duct under direct peroral cholangioscopy using a

conventional endoscopy, respectively [12, 13]. Accordingly, if a conventional endoscope can be inserted into the duodenal papilla, the treatment options will increase.

Miao et al. also reported the safety of septotomy for cannulation to the bile duct in a patient in whom insertion of a catheter into the bile duct was difficult [14]. Septotomy makes the duodenal papilla a large orifice, thereby facilitating not only spontaneous drainage of mucus but also smooth insertion of a conventional endoscope. When obstructive cholangitis repeats hereafter, these techniques may help to suppress mucus production from IPMNs. In conclusion, septotomy and mucus suction with direct peroral cholangioscopy using conventional endoscopy with a large suction channel could be useful to control biliary tract infection and obstructive jaundice due to mucus obstruction from an IPMNs accompanied by PB fistula.

## Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

**Human rights** All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

**Institutional review board statement** This study was approved by Totori Prefectural Central Hospital Clinical Ethics Review Board.

**Informed consent** Informed consent was obtained from all patients for being included in the study.

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