

## CASE REPORT

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# Management of the Mucin Filled Bile Duct. A Complication of Intraductal Papillary Mucinous Tumor of the Pancreas

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### ABSTRACT

**Context** Biliary obstruction secondary to intrabiliary mucin is a relatively rare presentation of malignant intraductal pancreatic mucinous tumor. We report a case of unresectable intraductal pancreatic mucinous tumor associated with obstructive jaundice secondary to intrabiliary mucin. This case and a review of the literature, highlight the difficulty in obtaining sustained palliation from jaundice using endoscopically placed biliary stents or percutaneously placed biliary catheters due to rapid occlusion with thick mucin secreted by the tumor. Furthermore, this case differs from that commonly seen in the setting of pancreatic adenocarcinoma, where endoscopic or percutaneous biliary drainage is usually successful at long-term palliation from jaundice.

**Case report** We report a case of obstructive jaundice secondary to invasive intraductal pancreatic mucinous tumor associated with dilated bile ducts containing copious amounts of mucin. The diagnosis of intraductal pancreatic mucinous tumor was established based on diagnostic findings on computed tomography scan and endoscopic retrograde cholangiopancreatography. The tumor was unresectable due to vascular invasion. Attempts at endoscopic biliary drainage proved unhelpful with the patient experiencing rapid occlusion of the biliary

stents with thick mucinous material leading to recurrent cholangitis. The patient eventually underwent a choledochojejunostomy leading to complete and sustained resolution of the cholestasis.

**Conclusion** If intraductal pancreatic mucinous tumor in association with intrabiliary mucinous obstruction is deemed unresectable, surgical biliary bypass seems to be superior to endoscopic biliary drainage and should be performed on initial presentation. This is due to rapid occlusion of biliary stents with thick mucin leading to frequent stent changes and recurrent cholestasis.

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### INTRODUCTION

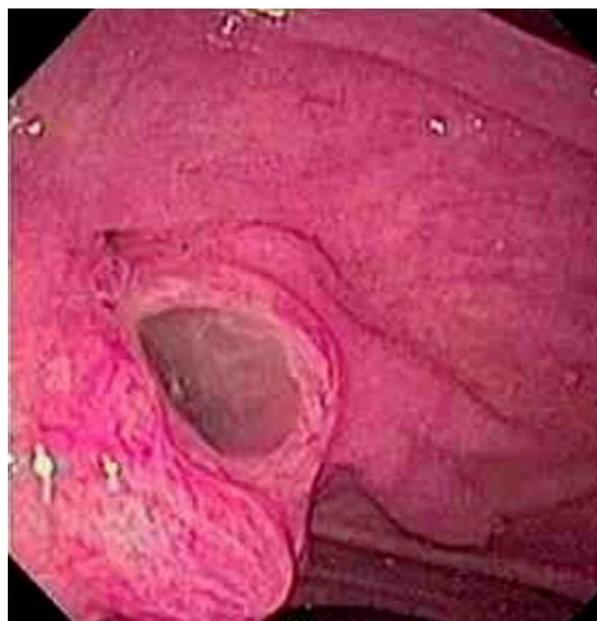
Since the first reported case in 1982 by Ohhashi *et al.* [1], intraductal pancreatic mucinous tumor (IPMT) has become a distinct clinico-pathologic entity, which is increasingly being recognized throughout the world. It is more common in males and usually presents in the seventh decade [2]. It can present with chronic or recurrent pancreatitis, jaundice, anorexia, weight loss, back pain, steatorrhea or diabetes [2, 3, 4]. It is characterized pathologically by a dysplastic pancreatic ductal epithelium, which typically secretes mucin and is associated with dilation of the main pancreatic duct and/or its side branches [5]. The dysplastic epithelium in

these tumors can become malignant in approximately a third of patients. These tumors can invade nearby structures such as the duodenum, stomach and bile duct [6]. Biliary obstruction secondary to IPMT is either from direct invasion into the bile duct wall leading to a stricture formation or a fistula forming between the mucin-containing tumor and the lumen of the bile duct leading to mucin build up in the lumen of the distal bile duct. Several other diseases can lead to the accumulation of mucin within the biliary tree. These should be distinguished from IPMT [7, 8, 9]. These include biliary intraductal papillary mucinous tumor, mucin producing biliary papillomatosis and, hepatobiliary cystic tumors.

We report a case of obstructive jaundice secondary to invasive, unresectable IPMT associated with dilated bile ducts containing copious amounts of mucin. Our case highlights the difficulty in providing sustained biliary drainage by endoscopic methods due to rapid occlusion from the thick mucin produced by the tumor. This type of cholestasis differs from that commonly seen in the setting of pancreatic adenocarcinoma, where endoscopic or percutaneous biliary drainage is usually successful at relieving jaundice for sustained periods of time.

## CASE REPORT

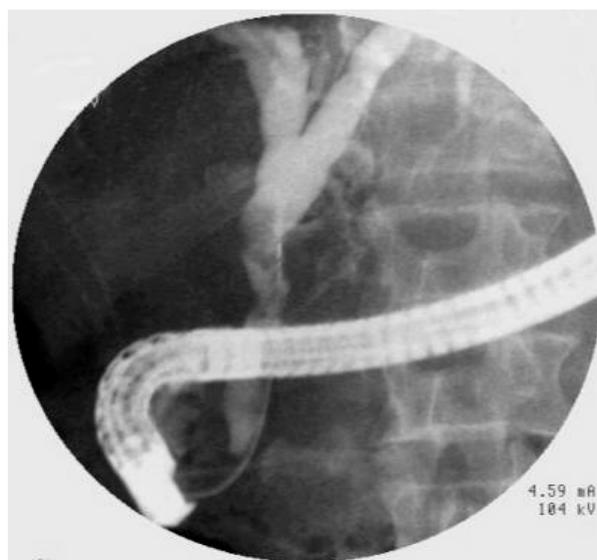
A 74-year-old male presented with cholangitis. He had lost 30 lb in weight over the preceding one year. CT of the abdomen revealed an 8.6x3.7 cm heterogeneous mass in the head of the pancreas compressing the portal and superior mesenteric veins. Endoscopic retrograde cholangiopancreatography (ERCP) showed a massively dilated pancreatic duct with multiple filling defects consistent with mucus plugs (Figure 1). The common bile duct (CBD) was dilated and filled with mucinous material (Figure 2). A biliary sphincterotomy was performed and the biliary mucinous debris was removed. A 10 French plastic biliary stent was placed leading to improvement in his jaundice. A diagnosis of IPMT was made based on endoscopic and



**Figure 1.** Appearance of ampulla on ERCP: The pancreatic orifice was patulous and engorged with clear mucinous secretions.

imaging studies. Due to invasion to the portal and superior mesenteric veins, as seen on CT images, the tumor was deemed to be unresectable.

He presented one month later with cholangitis. On ERCP the CBD was dilated to 20 mm and was filled with thick mucinous



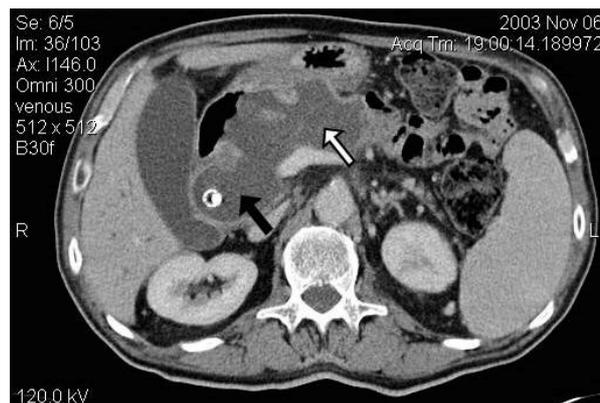
**Figure 2.** Endoscopic cholangiogram reveals a dilated biliary tree with distal and mid common bile duct containing extensive filling defects which on extraction was found to consist of copious amounts of mucinous secretions.

material. The mucinous material was cleared with balloon extraction and a 10x8 mm uncovered Zilver metal stent (Wilson-Cook Medical Inc., Winston-Salem, NC, USA) was placed. Antibiotics were also administered. Although the patient initially improved, the cholangitis recurred two weeks later. Repeat cholangiogram showed that the lumen of the metal stent was occluded with thick mucus plugs. The mucoid material was again balloon extracted and a 10 French 12 cm Cotton-Leung stent (Wilson-Cook Medical Inc., Winston-Salem, NC, USA) was placed through the metal stent.

Despite these efforts at endoscopic biliary drainage, his jaundice worsened with further weight loss. A repeat triphasic CT scan of the abdomen 10 weeks after the initial scan showed that the tumor had increased in size to 4.8x12 cm (Figure 3). There was evidence of biliary dilation and tumor invasion of the mesenteric vessels. The patient then underwent a choledochojejunostomy, cholecystectomy and gastrojejunostomy. He had no complications following surgery and was discharged from hospital on post-operative day 10. One month following the biliary bypass his jaundice has resolved and he has had no further episodes of cholangitis.

## DISCUSSION

Biliary obstruction can develop during the course of IPMT. Two factors contribute to the pathogenesis of biliary obstruction in the setting of IPMT. The first and most commonly seen presentation is direct extension of the pancreatic cystic tumor into the bile duct wall, as in the more common adenocarcinoma of the pancreas causing a biliary stricture. In this situation, biliary drainage is usually achievable endoscopically and/or radiologically. The second and less commonly seen presentation is when the mucin secreting tumor fistulizes into the bile duct and mucin accumulates in the biliary tree causing occlusion of bile flow. Our experience and evidence from the literature suggest that the latter presentation responds



**Figure 3.** CT scan depicts dilated mucin filled common biliary duct containing plastic stent (black arrow). Adjacent to the common biliary duct a greatly dilated and mucin filled pancreatic duct can be seen as well (white arrow).

poorly to endoscopic and radiologic drainage and usually requires surgical drainage [3, 10]. Kurihara *et al.* from Japan reported two cases of malignant IPMT associated with biliopancreatic fistula [10]. In their study, both patients presented with symptoms of biliary obstruction. One of the patients failed multiple attempts at endoscopic biliary drainage due to stent occlusion from thick mucinous secretions. Both subjects underwent surgical resection of the tumor. They also reviewed the literature and found 21 additional cases of biliopancreatic fistula associated with IPMT. Eighteen cases were from Japan, two cases from Europe and one case from the United States. With the exception of four cases, all patients underwent surgical resection. The patients undergoing surgery were all found to have malignant IPMT. One of the patients spared resection died of intractable cholangitis despite percutaneous transhepatic biliary drainage. Data on the outcome of the remaining three patients treated palliatively were not available.

Interestingly, in around half of the patients reported by Kurihara *et al.* [10], the pathogenesis was thought to be due to the mucous lakes associated with cystic pancreatic tumor bursting into the bile duct rather than direct tumor invasion, as evident by the lack of stromal invasion at the site of the pancreaticobiliary fistula in resected

specimens. In our patient, block resection and complete pathological examination of the tumor was not possible due to vascular invasion. The metal stent removed during surgery was covered with dysplastic cells suggesting direct invasion of the pancreatic tumor into the bile duct.

Our patient presented with biliary obstruction due to biliary mucin in the setting of malignant IPMT. The degree of invasion and vascular involvement seen on imaging studies precluded surgical resection and the biliary obstruction was initially treated with endoscopic biliary drainage. This failed to relieve the jaundice due to rapid occlusion of the stent with thick mucinous material. The uncovered biliary metal stent also got rapidly occluded with mucinous material leading to cholangitis. The patient eventually underwent surgical biliary bypass and achieved successful long-term relief of cholestasis.

In summary, malignant IPMT can involve the bile duct and produce biliary obstruction through the accumulation of thick mucinous material within the bile duct. Endoscopic biliary stenting with an uncovered metal stent or plastic stent is usually unsuccessful due to rapid occlusion of the stents with thick mucinous material. A covered metal stent could theoretically provide better occlusion of the fistula in the distal bile duct but there is no data in the literature to support this. If the fistula is not covered by the stent, it may also be susceptible to clogging as well. Although not used in our case, percutaneous transhepatic catheter drainage would encounter the same risk of clogging with the mucinous material and lead to frequent catheter changes. This was illustrated by the case in Kurihara's review [10] that died of cholangitis despite percutaneous transhepatic catheter drainage. Thus, surgical biliary bypass offers the best chance for successful biliary drainage in patients that present with a mucin filled bile duct in the setting of unresectable IPMT. Continued attempts at endoscopic management of these patients can be associated with risk of recurrent cholangitis, persistent cholestasis and adverse outcomes and should be avoided.

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**Keywords** Cholangitis; Cholangiography; Neoplasms, Cystic, Mucinous, and Serous; Palliative Care

**Abbreviations** CBD: common bile duct; IPMT: intraductal pancreatic mucinous tumor

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### References

1. Ohashi K, Murakami Y, Maruyama M. Four cases of mucin producing cancer of the pancreas on specific findings of the papilla of Vater. *Prog Dig Endosc* 1982; 20:348.
2. Balzano G, Zerbi A, Di Carlo V. Intraductal papillary mucinous tumors of the pancreas: incidence, clinical findings and natural history. *JOP. J Pancreas (Online)* 2005; 6(1 Suppl.):108-11. [PMID 15650294]
3. Tanaka M. Intraductal papillary mucinous neoplasm of the pancreas: diagnosis and treatment. *Pancreas* 2004; 28:282-8. [PMID 15084972]
4. Suzuki Y, Atomi Y, Sugiyama M, Isaji S, Inui K, Kimura W, et al. Cystic neoplasm of the pancreas. A Japanese multiinstitutional study of intraductal papillary mucinous tumor and mucinous cystic tumor. *Pancreas* 2004; 28:241-6. [PMID 15084964]
5. Loftus EV Jr, Olivares-Pakzad BA, Batts KP, Adkins MC, Stephens DH, Sarr MG, DiMagno EP. Intraductal papillary-mucinous tumors of the pancreas: clinicopathologic features, outcome, and nomenclature. Members of the Pancreas Clinic, and Pancreatic Surgeons of Mayo Clinic. *Gastroenterology* 1996; 110:1909-18. [PMID 8964418]
6. Jung IS, Shim CS, Cheon YK, Bhandari S, Cha SW, Moon JH, et al. Invasive intraductal papillary mucinous tumor of the pancreas with simultaneous invasion of the stomach and duodenum. *Endoscopy* 2004; 36:186-9. [PMID 14765321]

7. Ishada M, Seki K, Honda K, Kimura T, Katayama K, Hirose K, et al. Intraductal mucinous tumors occurring simultaneously in the liver and pancreas. *J Gastroenterology* 2002; 37:1073-78. [PMID 12522542]

8. Somgyi L, Dimashkieh H, Weber FL Jr, Buell J. Biliary intraductal papillary mucinous tumor: diagnosis and localization by endoscopic retrograde cholangioscopy. *Gastrointest Endosc* 2003; 57:620-2. [PMID 12665789]

9. Lee SS, Kim MH, Lee SK, Jang SJ, Song MH, Kim KP et al. Clinicopathologic review of 58 patients with biliary papillomatosis. *Cancer* 2004; 100:783-93. [PMID 14770435]

10. Kurihara K, Nagai H, Kasahara K, Kanazawa K, Kanai N. Biliopancreatic fistula associated with intraductal papillary-mucinous pancreatic cancer: institutional experience and review of the literature. *Hepatogastroenterology* 2000; 47:1164-7. [PMID 11020905]

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