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Intraductal Papillary Mucinous Tumor Simultaneously Involving the Liver and Pancreas
– A Case Report –

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We describe here a 67-year-old man who was diagnosed with a rare case of intraductal papillary mucinous tumors that occurred simultaneously in the liver and pancreas. Abdominal computed tomography showed a tubular and cystic dilatation of the pancreatic duct in the pancreas tail, which suggested an intraductal papillary mucinous tumor (IPMT), and multiple intrahepatic duct stones. The patient underwent a distal pancreatectomy with splenectomy and a lateral segmentectomy of the liver. Microscopic examination showed an intraductal papillary mucinous neoplasms of borderline malignancy in the pancreas and a non-invasive intraductal papillary mucinous tumor with moderate dysplasia of the bile duct. Although several cases of intraductal papillary mucinous neoplasm of the liver (IPNL) without any pancreatic association have been described, the simultaneous presentation of both IPMT of the pancreas and IPNL is very rare. The patient has been doing well for 10 months postoperatively.

Key Words : Pancreas; Bile ducts; Liver; Intraductal papillary mucinous; Neoplasms

Intraductal papillary mucinous tumor of the pancreas (IPMT) has been classified as a separate disease entity, and IMPT is defined as a mucin-producing neoplasm of the main pancreatic duct or the branches thereof, and it shows a cystic dilatation of the pancreatic duct and a papillary proliferation of tumor cells with various degrees of histological dysplasia.[1-3] IPMTs have been reported to constitute approximately 0.4% of all pancreatic neoplasms.[1,2] In contrast to IPMT, intraductal papillary mucinous neoplasm of the liver (IPNL), which shows a histopathologic morphology similar to that of IPMT, has not been classified as a specific disease entity. We treated a rare case of these two homologous lesions that presented concurrently in the liver and pancreas.

CASE REPORT

A 67-year-old man presented to a private clinic with epigastric pain. He had previously been healthy without any medical or surgical disease. He was a heavy smoker with a 40 pack-year smoking history, but he did not have a habit of drinking alcohol. Over the previous 10 days, he had experienced gradually increasing, continuous epigastric pain. Gastrofiberscopy showed multiple gastric ulcers, and he was medically treated for this. As his epigastric pain was not improved by medication, he revisited the hospital. Abdominal computed tomography (CT) showed a 2.2 × 1.9 cm-sized cystic mass in the pancreatic tail with dilatation of the adjacent pancreatic duct, and this all suggested an IPMT. For further evaluation, he was referred to our hospital. There was no palpable mass on the physical examination. The serum lipase concentration was mildly elevated to 55 IU/L (normal, 5.60-51.30 IU/L). The other laboratory findings, including the liver function tests and the serum levels of alpha-fetoprotein, amylase, carcinoembryonic antigen, and carbohydrate-associated antigen 19-9 were all within the normal limits.

A review of his abdominal CT results revealed tubular and cys-

tic dilatation of the pancreatic branch duct with communication to the main pancreatic duct in the pancreas tail, and this all suggested an IPMT (Fig. 1A). In addition, multiple intrahepatic duct stones were identified in segment 3 (S3) of the liver, and these stones were causing focal stricture of the S3 intrahepatic duct (Fig. 1B). The patient underwent a distal pancreatectomy with splenectomy, and a lateral segmentectomy of the liver. The external surface of the resected pancreas was diffusely nodular and hard. The cut surface showed a unilocular cystic lesion (2.5 × 1.0 × 1.0 cm) containing mucinous materials (Fig. 2A). This lesion was communicating with the main pancreatic duct which was diffusely dilated. Multiple scattered yellowish calcifications were noted in the pancreatic parenchyma. The cut surface of the liver showed dilated intrahepatic ducts containing multiple stones but without a definite mass (Fig. 2B).

Microscopically, similar histological findings were noted in the pancreatic ducts and intrahepatic bile ducts, and these findings showed tall columnar cells that contained abundant cytoplasm and intracytoplasmic mucin (Fig. 3A, C). The epithelial cells showed moderate dysplasia and small papillary projections without long slender fibrovascular cores (Fig. 3B, D). No stromal or lymphovascular invasion was detected. The spleen showed no pathology. The patient has shown no evidence of disease for 10 months postoperatively.

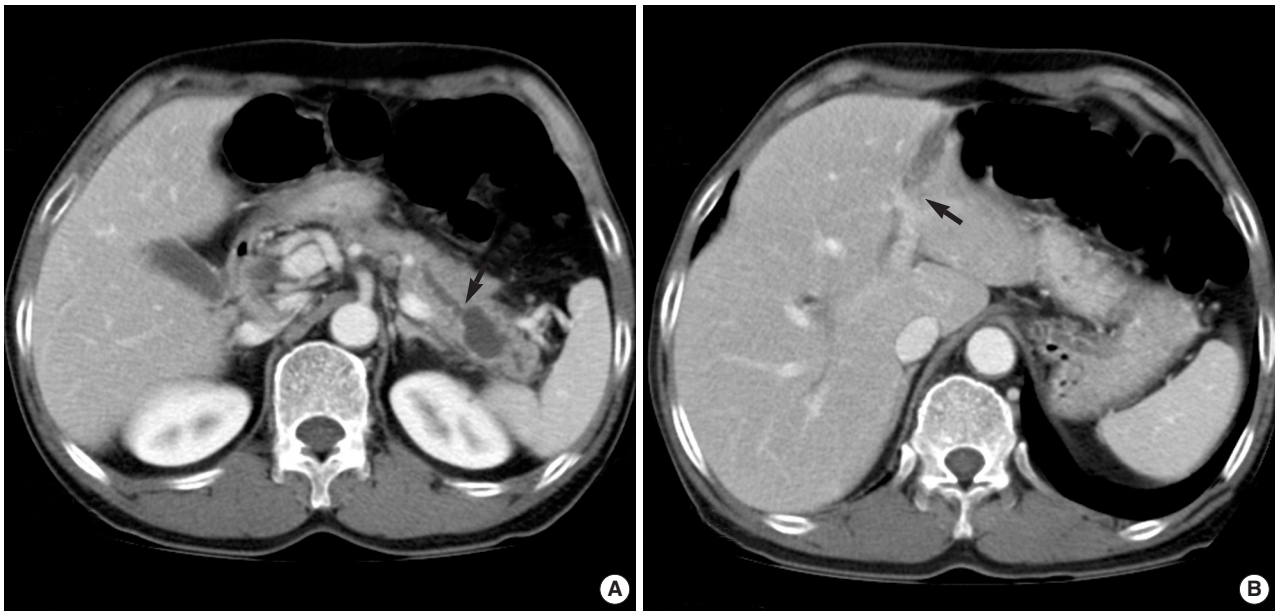

Fig. 1. Abdominal computed tomography (CT) reveals a cystic mass of communicating with dilatation of the main pancreatic duct in the pancreatic tail (A) and multiple intrahepatic duct stones in segment 3 of the liver with dilatation of the intrahepatic duct (B).

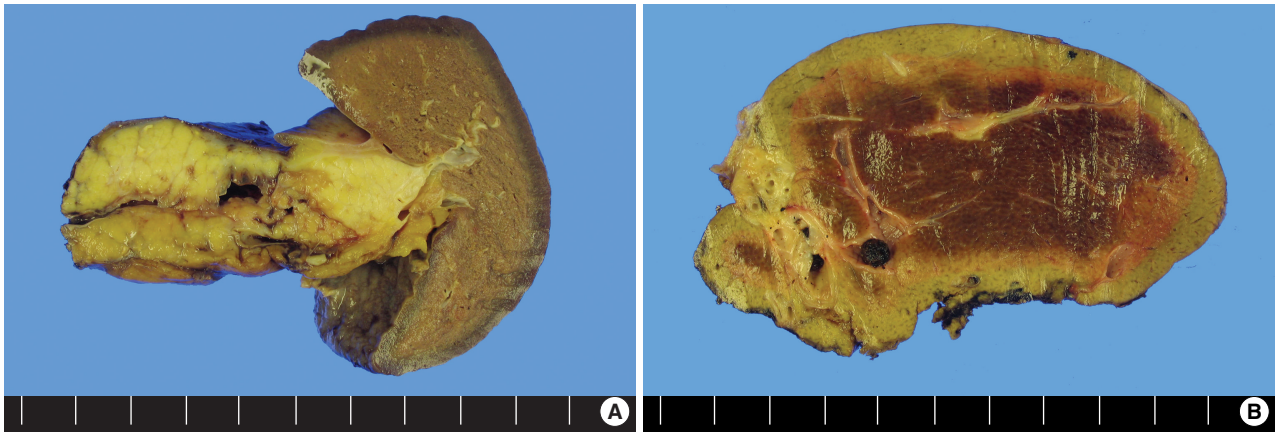

Fig. 2. Distal pancreatectomy with splenectomy specimen shows a unilocular cystic mass-like lesion with the dilated main pancreatic ducts (A). Left lateral segmentectomy of the liver shows dilated intrahepatic ducts with multiple stones (B).

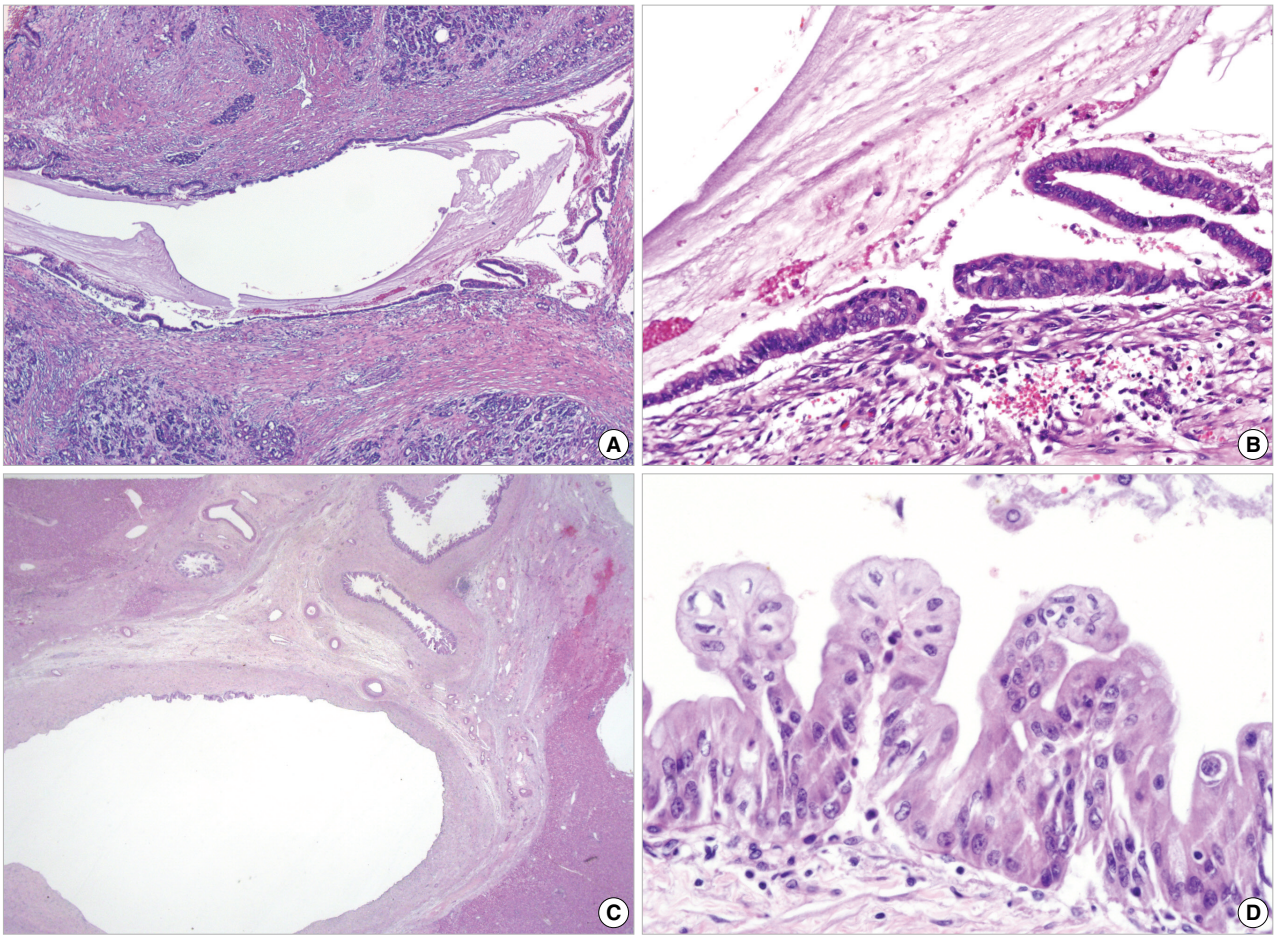

Fig. 3. The dilated pancreas ducts contain mucinous materials (A) and the tumor cells show moderate dysplasia (B). The tumor cells lining dilated intrahepatic bile ducts show cellular atypism and small papillary proliferation (C, D).

DISCUSSION

IPNLs are, also called intraductal papillary mucinous neoplasms of the bile ducts (IPMN-Bs), mucin-producing bile duct tumors (MPBT), biliary papillomatosis, or biliary papillary tumors,[4] and IPNLs have been identified in various clinical conditions, including hepatolithiasis, recurrent pyogenic cholangitis and abscess. The incidence of IPNL in association with hepatolithiasis was reported to be as high as 30%.[5,6] However, IPMT has not been definitively associated with any clinical condition except smoking.

Although IPMT is frequently associated with malignancy of other organs, including the stomach, colon, esophagus and lung, the simultaneous occurrence of IPMT and IPNL is very rare,[3,7] and only three such cases have currently been reported.[8-10] Two of those cases were described to show focal adenomatous change with mild cellular atypism.[8,9] One case by Yamaguchi *et al.*[10] was the simultaneous occurrence of histologically malignant IPMT and IPNL. Hepatolithiasis has not been mentioned in the previous reports. The hepatic lesions in the previous cases consisted of cystically dilated intrahepatic bile ducts that showed mass-like lesion, which was similar to IPMT.[8-10] Whereas the present case showed mildly dilated intrahepatic ducts without a definite mass.

Molecular tests, including assays for *K-ras* oncogene mutations, have shown that IPMT follows a genetic course similar to that of the colorectal adenoma-carcinoma sequence in the progression to carcinoma.[3,11] IPMT and IPNL have recently been shown to share several molecular and embryologic aspects. For example, *K-ras* mutations have been detected in 29% of the IPNLs as well as in the hyperplastic lesions of the pancreas including IPMT.[12,13] Although we could not rule out the possibility that the two neoplasms occurred coincidentally in our case, the similar histology of the pancreatic and hepatic lesions suggests that both tumors

may have a similar pathogenesis. We can speculate that both lesions were premalignant and so they might have accumulated fewer genetic abnormalities than carcinomas. Further studies on the carcinogenesis of IPMT and IPNL may clarify this hypothesis.

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