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Case Report

A Resected Case of Mucinous Adenocarcinoma of the Duodenum, Mimicking Intraductal Papillary Neoplasm of the Bile Duct

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Duodenum mucinous carcinoma is very rare, and the prognosis of the patient is very bad, especially when the tumor is invasive to other organs. In this case, duodenum carcinoma was invasive to common bile duct and transverse colon. Mucinous fluid, which was secreted from a duodenum tumor, was found in the dilatated bile duct. The intraductal papillary neoplasm of the bile duct was considered a differential diagnosis. We performed aggressive resection and had a good prognosis. A 74-year-old woman received a diagnosis of cholangitis and was treated with antibiotic drugs. Endoscopic retrograde cholangiopancreatography revealed a defect in the lower common bile duct with the mucoid fluid. We suspected intraductal papillary neoplasm of the bile duct, but no malignant cells were detected. One year later, gastrointestinal fiberscopy revealed a villous tumor in the postbulbar portion of the duodenum; adenocarcinoma was detected in biopsy specimens. Computed tomography revealed dilatation of the duodenum with an enhanced tumor, and dilatation of both the common and intrahepatic bile ducts. Magnetic resonance cholangiopancreatography revealed that the duodenum was connected with the common bile duct and ascending colon. We resected the segmental duodenum, extrahepatic bile duct, left lobe of liver, a partial of the transverse colon, and associated lymph nodes. Although the advanced duodenal carcinoma had poor prognosis, the patient was alive, without recurrence, 5 years after the operation.

Key words: Duodenal carcinoma – Mucinous carcinoma – Intraductal papillary neoplasm of the bile duct – Duodenal segmentectomy

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Duodenum carcinoma is rare and reported to be found in only 0.3% to 0.4% of all gastrointestinal tract cancers. Mucinous carcinoma of the duodenum is very rare, and there have been only a few case reports. In this case, duodenum carcinoma was invasive to common bile duct and transverse colon. In addition, the imaging of the tumor was similar to intraductal papillary neoplasm of the bile duct (IPNB). There was no case report describing a patient like this one. Overall survival at 2 years with advanced duodenal carcinoma was reported to be 11.7%.[1] We report a successfully resected case of primary mucinous carcinoma of the duodenum invasive to common bile duct and transverse colon.

Case Presentation

A 74-year-old woman was admitted to our hospital for further investigation and treatment of repeated cholangitis; she had no other past medical history. When the patient was hospitalized for cholangitis 1 year previously, computed tomography (CT) did not reveal any tumors or any other cause for the cholangitis. Endoscopic retrograde cholangiopancreatography (ERCP) revealed a defect in the lower common bile duct with accumulation of mucoid fluid, and the papilla of Vater was expanded because of the presence of mucus (Fig. 1). The mucus was removed by using a balloon catheter, which was inserted into the upper bile duct. No malignant cells were detected in brush cytology samples from the bile duct. IPNB was suspected because of the mucus in the bile duct, but there was no evidence of the intraductal tumor; the patient was treated with antibiotic drugs. However, she experienced repeated cholangitis and was readmitted to our hospital 1 year later because of a high fever. The laboratory data upon readmission were as follows: white blood cell count, 13,200 cells/μL; hemoglobin level, 9.0 g/dL; hematocrit, 27.1%; platelet count, 243,000 cells/μL; C-reactive protein, 18.3 mg/dL; total bilirubin level, 1.3 mg/dL; aspartate aminotransferase level, 22 IU/L; alanine aminotransferase level, 20 IU/L; lactate dehydrogenase level, 193 IU/L; alkaline phosphatase level, 784 IU/L; and γ-glutamyltransferase level, 205 IU/L. The levels of some tumor markers were elevated, which are as follows: carcinoembryonic antigen, 1.2 ng/mL; carbohydrate antigen 19-9, 45,272 U/mL; DUPAN-2, 601 U/mL; and SPAN-1, 8481 U/mL. CT revealed an enhanced tumor in the duodenum, and dilatation of the common bile duct and intrahepatic bile duct at the left lobe of the liver (Fig. 2). The common bile duct wall was thickened and enhanced and hilar bile duct was narrow by the tumor. Magnetic resonance cholangiopancreatography revealed dilatation of the bile duct, and that the duodenum was connected with the hilar bile duct and ascending colon (Fig. 3). ERCP revealed stenosis in the hilar portion of the bile duct and mucoid fluid in the lower common bile duct. No malignant cells were detected in brush cytology samples taken from bile duct at this time, similar to what had been observed 1 year previously. Gastrointestinal fiberscopy revealed a villous tumor in the postbulbar portion of the duodenum (Fig. 4a). The villous adenoma was detected as an adenocarcinoma in the biopsy specimens. Colonofiberscopy revealed a papillary mass in the hepatic flexure; adenocarcinoma was not detected in biopsy specimens. ^{18}F-fluorodeoxyglucose-positron emission tomography/CT (^{18}F-FDG-PET/CT) revealed accumulation of fluid in the left lobe of the liver, where the maximum standardized uptake value (SUVmax) was 6.06, and accumulation in the common bile duct and duodenal tumor, where the SUVmax was 4.81 (Fig. 4b). It was considered invasion to left hepatic duct by the dilatation of left hepatic bile duct.

This case received a diagnosis of duodenal cancer and direct invasion to the hilar bile duct, with mucosal fluid accumulation in the left lobe of the liver, on the basis of the findings with clinical imaging and the analysis of biopsy specimens. We performed segmental resection of the duodenum, resection of the extrahepatic bile duct, left hepatic lobectomy, partial resection of the transverse colon, dissection of associated lymph nodes, and choledochojejunostomy; the operating time was 981 minutes, and blood loss was 2953 mL. The histologic diagnosis was papillary adenocarcinoma of the duodenum with mucinous production and invasion to the bile duct and transverse colon, with no evidence of metastasis to the lymph nodes and liver (Fig. 5). Inflammatory granulation tissue and abscess formation with prominent neutrophil infiltration was observed in the liver, but no tumor was observed.

After the operation, we drained and washed the intra-abdominal abscesses. The patient was discharged on the 29th day after surgery, and adjuvant chemotherapy with tegafur-uracil was administered for 1 year. Five years after the operation, she is alive without any recurrence.

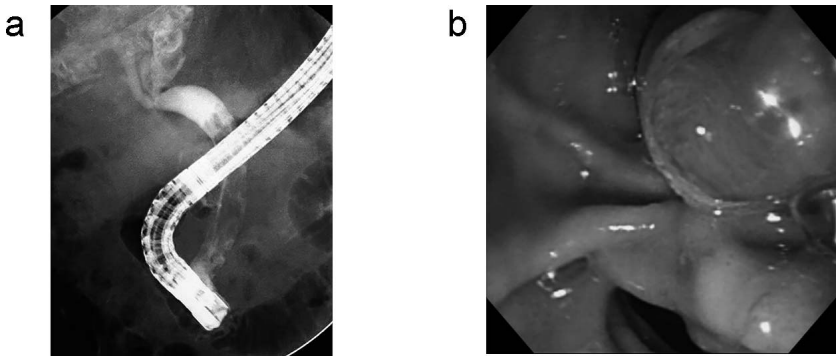

Fig. 1 ERCP findings. (a) ERCP revealed that the common bile duct was dilatated and the lower bile duct filled with mucoid fluid. The mucus was removed by using a balloon catheter, which was inserted into the upper bile duct. (b) ERCP revealed an expanded papilla of Vater due to the presence of mucoid fluid.

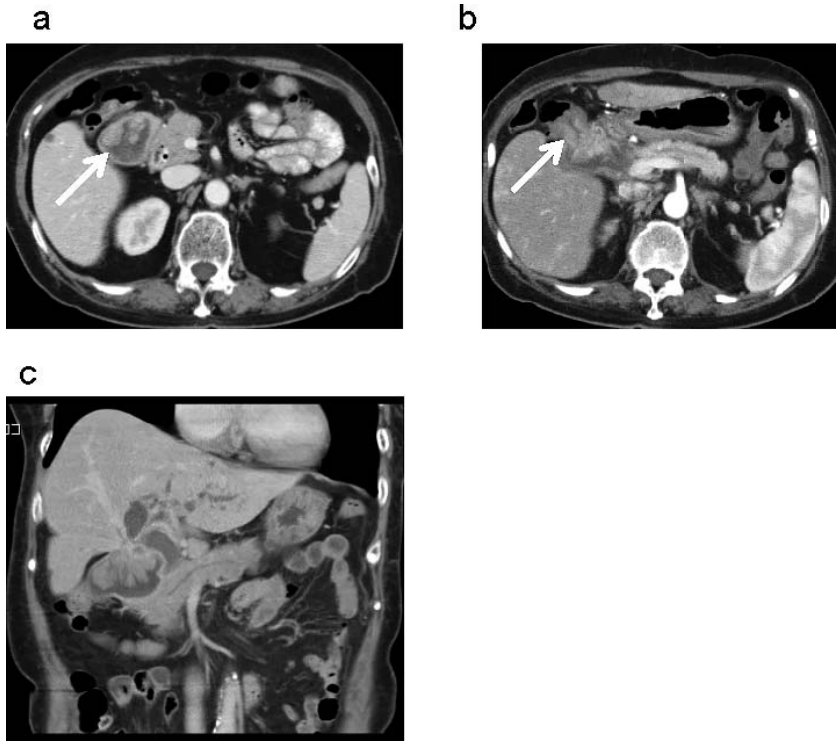

Fig. 2 Abdominal enhanced CT findings. CT images were taken on the day of admission. Axial view (a and b) and coronal view (c). (a) Abdominal CT imaging revealed an enhanced tumor in the extended duodenum. The arrow on the figure indicates the duodenal carcinoma. (b) Invasion of the transverse colon was suspected. The arrow on the figure indicates the tumor invasion to the transverse colon. (c) Coronal view of the CT revealed invasion to the hilar bile duct and dilatation of the left hepatic bile duct and common bile duct.

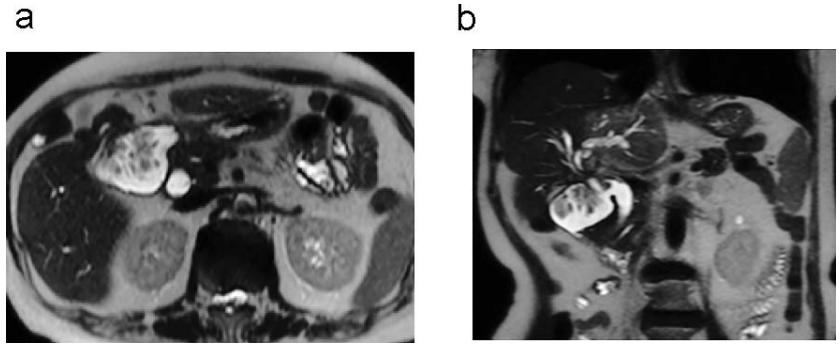

Fig. 3 Magnetic resonance imaging (MRI) findings. MRI revealed the connection of the duodenum and hilar bile duct, and duodenal carcinoma was detected in a T2-weighted image. Axial view (a) and coronal view (b).

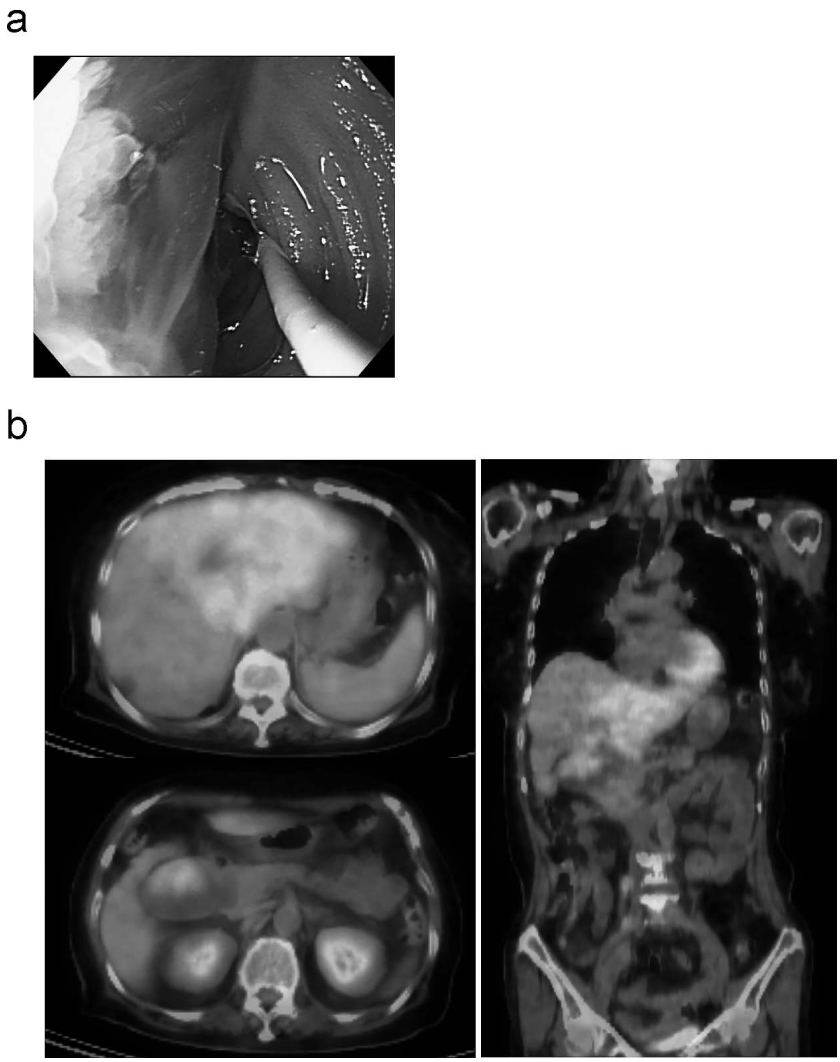

Fig. 4 (a) Gastrointestinal fiberscope (GIF) revealed villous tumor with mucin fluid in the postbulbar portion of the duodenum and an adenocarcinoma was detected in biopsy specimens. (b) ^{18}F-FDG-PET/CT revealed accumulation in the duodenal tumor (SUVmax, 4.81) and in the left lobe of the liver (SUVmax, 6.06).

Discussion

Duodenal carcinoma has been reported to be found in 0.3% to 0.4% of all gastrointestinal tract cancers.[2] The mean age of patients with duodenal cancer is 68 years, and the male to female ratio is 1.9:1 to 2.75:1.[3–5] Most (56.5–74.0%) duodenal cancers are located in the second portion of the duodenum, and only 4% are located in the first portion.[6] Duodenal mucinous adenocarcinoma is very rare and only reported in a few case reports.[7–9]

The common symptoms of duodenal carcinoma are abdominal pain, gastrointestinal tract bleeding, body weight loss, nausea, vomiting, and jaundice.[3] Because the medical examination reveals little information about such cases, duodenal carcinoma is often only diagnosed at an advanced stage. Surgical resection is the only radical curative treatment for duodenal cancer. Some reports have suggested that the prognostic factors are disease stage, cytology, total bilirubin, and positive resection margins.[3,5,6] The overall survival at 5 years in resectable cases was reported to be 40% to 71%, but in nonresectable cases it was 0%.[5,6,10] The overall survival at 2 years for patients with advanced duodenal carcinoma, especially with serosal invasion, was reported to be only 11.7%.[1] On the contrary, it was reported that lymph node–positive duodenal carcinoma is curable,[11] and the 5-year survival of a patient with pN0 was not significantly different from that of a patient with pN1.[10] Resection of tumors with positively expanding margins is useful for duodenal cancer.[12]

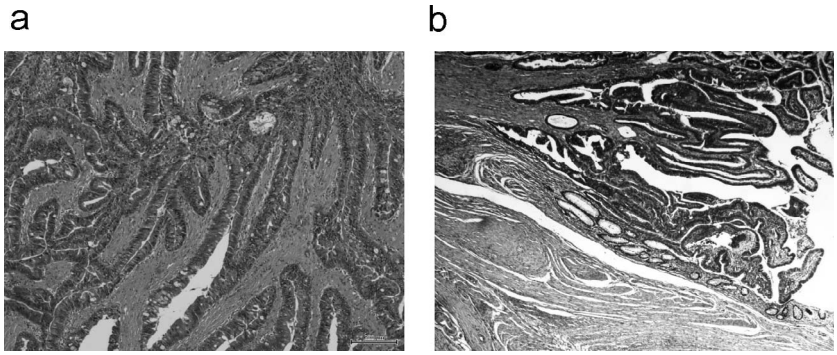

Fig. 5 Pathologic findings. (a) Microscopic findings. The pathologic diagnosis is papillary adenocarcinoma of the duodenum with mucus production. (b) This figure revealed that duodenal carcinoma invaded the transverse colon.

IPNB has recently been proposed as a new disease concept—that is, a papillary or villous proliferating biliary epithelium producing mucus in the bile duct—and it is considered a tumor of borderline malignant potential. A characteristic feature of images of IPNB is a dilatated bile duct containing mucoid fluid. The images in this case were similar to those of IPNB. When the presence of mucoid fluid was observed at the time of the first ERCP, it was thought that there was already penetration into the bile duct from the duodenum; however, since then, signs that contrast medium in the bile duct was discharged to the duodenum have not been observed. The patient was treated with antibiotic drugs, and her condition improved. However, this delayed the operation and made it difficult to make a diagnosis of duodenal carcinoma at an early date.

Prominent unusual accumulation in the left lobe of the liver on the preoperative ^{18}F-FDG-PET/CT images was considered to have been due to inflammation occurring as a secondary effect of the stasis of mucus in the bile duct because of the tumor invasion to the left hepatic bile duct. Left hepatectomy was required close to the invasion; we should get further information with intraoperative ultrasound examination to define the neoplastic extension. The invasion to left hepatic bile duct was not found by the pathology report. It was thought that the dilatation of left bile duct was caused by the narrowing and mucus.

Duodenal segmentectomy associated with intestinal derotation was reported as a safe procedure for the treatment of primary adenocarcinoma of the third and fourth portions of the duodenum.[13,14] When the complete oncologic (R0) resection is possible, duodenal segmentectomy is regarded as the effective procedure.[14] The prognosis of patients with duodenal carcinoma who were treated with duodenal segmentectomy or pancreatoduodenectomy was not significantly different.[15] Segmental resection may be appropriate for a case of duodenal carcinoma without invasion of the pancreas. In this case, duodenal tumor was located at the opposite side of the pancreas from the first portion of duodenum and invasion to hilar bile duct. It was considered that there was no invasion to the pancreas and the lower part of bile duct. Duodenal segmentectomy was performed to preserve the pancreas, and complete oncologic (R0) resection was confirmed by the pathology report. Because 5 years have passed without tumor recurrence, we believe that curative resection has been performed.

It was reported that the use of adjuvant chemoradiation therapy does not influence survival.[16] However, our patient was treated with adjuvant chemotherapy with tegafur-uracil for 1 year. She has been disease-free for 5 years after the operation.

Conclusion

We report a successfully resected case of primary mucinous carcinoma of the duodenum. Mucinous fluid from the duodenum tumor was found in the common bile duct, and IPNB was considered the differential diagnosis. Even if there is invasion to other organs from a duodenal cancer, we think that a prognostic improvement is obtained by aggressive excision.

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