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Single Case

Carcinoma of the Papilla of Vater after Diversion Operation for Pancreaticobiliary Maljunction

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Keywords

Pancreaticobiliary maljunction · Carcinoma · Papilla of Vater

Abstract

Pancreaticobiliary maljunction (PBM) is a congenital malformation that is associated with biliary cancer development. When patients are diagnosed with PBM, a diversion operation is recommended. Although a risk remains for developing residual bile duct carcinoma following diversion, the development of a carcinoma of the ampulla of Vater after a diversion operation for PBM is rare. We present a treated case of carcinoma of the ampulla of Vater after a diversion operation for PBM. A 65-year-old woman presented with abdominal pain. She had undergone extrahepatic bile duct resection and cholecystectomy 2 years 9 months previously for the treatment of type Ic PBM according to the Todani classification. At the current admission for evaluation of the abdominal pain, computed tomography and magnetic resonance imaging showed only dilation of the main pancreatic duct. However, gastrointestinal endoscopy showed a tumor at the papilla of Vater, and biopsy revealed adenocarcinoma of the papilla of Vater. We performed pylorus-preserving pancreaticoduodenectomy, and the pathological diagnosis was moderately differentiated tubular adenocarcinoma of the papilla of Vater with no metastasis to the lymph nodes. The patient remained in good health for 3 years postoperatively. Carcinoma of the papilla of Vater after a diversion operation for PBM is

rare. In this case, a diagnosis could not be made by computed tomography or magnetic resonance imaging; the definitive diagnosis was obtained with gastrointestinal endoscopy. Careful postoperative follow-up with gastrointestinal endoscopy in addition to imaging examination may be needed after a diversion operation for PBM.

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Background

Pancreaticobiliary maljunction (PBM) is a congenital malformation in which the pancreatic and bile ducts are anatomically joined outside the duodenal wall. In addition, the reflux of pancreatic juices into the biliary tract provokes higher rates of biliary tract cancer. So, when patients are diagnosed with PBM, a surgical treatment is recommended before the onset of biliary tract cancer [1–3]. Although a risk remains for developing residual bile duct carcinoma after a diversion operation for PBM, the development of a carcinoma of the ampulla of Vater after diversion is rare. We treated a patient with carcinoma of the ampulla of Vater after a diversion operation for PBM. Only 1 similar case has been reported in the literature [4]. We present our case with a brief review of the literature.

Case Presentation

A 65-year-old woman with abdominal pain presented to a local hospital. She had undergone extrahepatic bile duct resection, cholecystectomy, and choledochojejunostomy at our hospital 2 years 9 months previously for treatment of type Ic PBM according to the Todani classification [5]. At the local hospital, physical examination revealed epigastric pain and computed tomography (CT) showed dilation of the main pancreatic duct (Fig. 1a). The patient improved with 4 days of bed rest at the local hospital. She then presented to our hospital to determine the cause of the abdominal pain and dilation of the main pancreatic duct.

Laboratory data on admission to our hospital were as follows: white blood cell count, 6,200/ μ L; red blood cell count, 404 \times 10⁴/ μ L; hemoglobin, 12.4 g/dL; hematocrit, 38.8%; platelets, 28.6 \times 10⁴/ μ L; total bilirubin, 0.3 mg/dL; aspartate aminotransferase, 22 IU/L; alanine aminotransferase, 20 IU/L; alkaline phosphatase, 317 IU/L; γ -glutamyl transferase, 33 IU/L; amylase, 110 U/L; blood urea nitrogen, 19 mg/dL; creatinine, 0.60 mg/dL; C-reactive protein, 0.04 mg/dL; carcinoembryonic antigen (CEA), 3.9 ng/mL, and carbohydrate antigenic determinant 19-9 (CA19-9), 15 U/mL. The complete blood cell count was within the reference range, and hepatic, biliary, and pancreatic enzyme concentrations were not elevated. The concentrations of tumor markers, including CEA and CA19-9, were normal.

Gastrointestinal endoscopy revealed a tumor of the papilla of Vater, and adenocarcinoma of the papilla of Vater was diagnosed by biopsy (Fig. 1b). Magnetic resonance imaging (MRI) and CT revealed dilation of the main pancreatic duct. However, the tumor at the papilla of Vater was not clear on MRI. There was no obvious accumulation of fluorodeoxyglucose on whole-body fluorodeoxyglucose-positron emission tomography. There was also no distant metastasis. We then performed pylorus-preserving pancreaticoduodenectomy and pancreaticojejunostomy with a diagnosis of carcinoma of the papilla of Vater. Intraoperatively, we performed adhesiolysis of the abdominal wall, liver, and small intestine and found that the tumor at the papilla of Vater was approximately 1 cm in diameter. We removed the pancreatic head and duodenum and performed pancreaticojejunostomy and gastrojejunostomy. The pathological diagnosis was moderately differentiated tubular adenocarcinoma of the

papilla of Vater with no metastasis to the lymph nodes (pT2N0M0 stage IB, International Union against Cancer, UICC). The main part of the tumor was present at the common channel (Fig. 2). The specimen showed p53 expression in the cancer tissue and atypical epithelium of the retained bile duct (Fig. 3). The patient remained in good health with no recurrence for 3 years after the second operation.

Discussion

PBM is frequently associated with biliary cancer [6, 7]. Previous reports have suggested that metaplasia and dysplasia of biliary duct epithelial cells might develop at the site of bile stasis, irritation, and inflammation because pancreatic juice mixed with infected or denatured bile and pancreatic enzymes were activated by the presence of enterokinase [2, 8, 9]. Mutations in the K-ras and p53 genes have also been found in the mucosa of the bile duct in patients with PBM [10–12]. Our patient also showed high expression of p53 in the cancer tissue and atypical epithelium of the bile duct with immunostaining. In a previous report, the rate of developing biliary cancer in patients with PBM with and without choledochal dilation was 21.6 and 42.2%, respectively [2]. Although cholecystectomy, resection of the extrahepatic bile duct, and hepaticojejunostomy have been recommended as standard operations for treating PBM with choledochal dilation, whether the bile duct should be removed when treating PBM without choledochal dilation remains unclear [3]. Even after undergoing a surgical procedure that stops reflux of pancreatic enzymes into the choledochus and prevents the development of biliary cancer, patients with PBM still have a risk of developing residual bile duct carcinoma [9]. The estimated incidence of cancer development after a diversion operation is 0.7%, and the interval between the operation and cancer detection ranges from 1 to 19 years [9]. The development of carcinoma of the papilla of Vater after choledochojejunostomy for PBM is rare; only 1 such report has been published [4]. No reports have discussed the risk factors for carcinoma of the papilla of Vater or the relationship between PBM and carcinoma of the papilla of Vater. In the present case, the main part of the tumor was present in the common channel, which is reportedly the most frequent site of carcinoma of the papilla of Vater [7]. The residual bile duct in the pancreas was atrophic and replaced with atypical epithelium. Therefore, we considered that the PBM caused the carcinogenesis at the common channel and residual bile duct after the diversion operation for PBM. If a tumor at the common channel develops alongside the papilla of Vater, the tumor may manifest as a carcinoma of the papilla of Vater. Careful postoperative examination is important for patients who have undergone a diversion operation for PBM because jaundice is not usually observed in patients who have undergone a diversion operation of the bile duct, although it is one of the most common manifestations of carcinoma of the papilla of Vater [4]. In the present case, the patient had undergone regular follow-up CT examinations. We could not find the tumor by CT or MRI but were able to identify it by gastrointestinal endoscopy. Gastrointestinal endoscopy should be performed regularly as part of the postoperative follow-up of PBM.

Conclusion

We experienced a rare case of carcinoma of the papilla of Vater 2 years 9 months after a diversion operation for the treatment of PBM. In this case, we were able to diagnose the car-

cinoma not by CT or MRI but by gastrointestinal endoscopy. Careful postoperative follow-up with gastrointestinal endoscopy in addition to imaging may be necessary in such patients.

Statement of Ethics

Approval from the Ethics Committee was not required for this case report. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Disclosure Statement

The authors declare that they received no financial support in association with this case report. The authors also declare that they have no competing interests.

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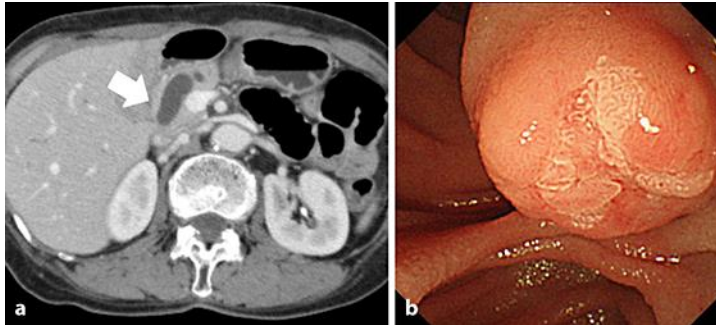


Fig. 1. **a** Contrast-enhanced CT showing dilation of the main pancreatic duct on the peripheral side of the papilla of Vater. The main pancreatic duct is indicated by the arrow. **b** Gastrointestinal endoscopy showing the tumor at the papilla of Vater. The pathological diagnosis according to biopsy examination was adenocarcinoma of the papilla of Vater.

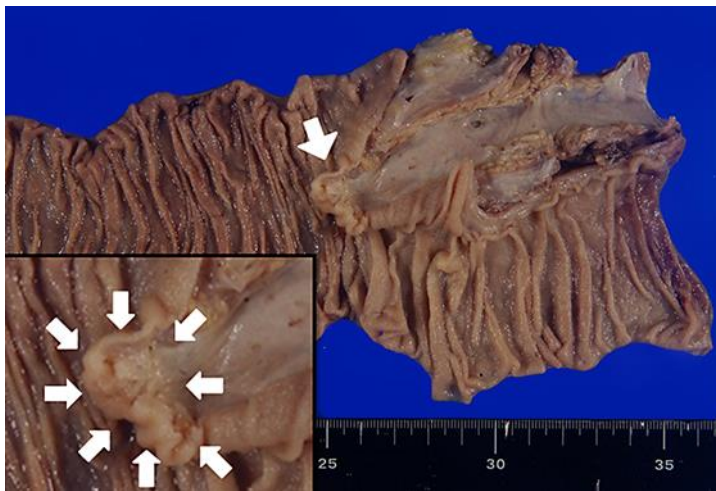


Fig. 2. Pathological findings showing a 15-mm tumor at the papilla of Vater. The tumor is indicated by arrows.

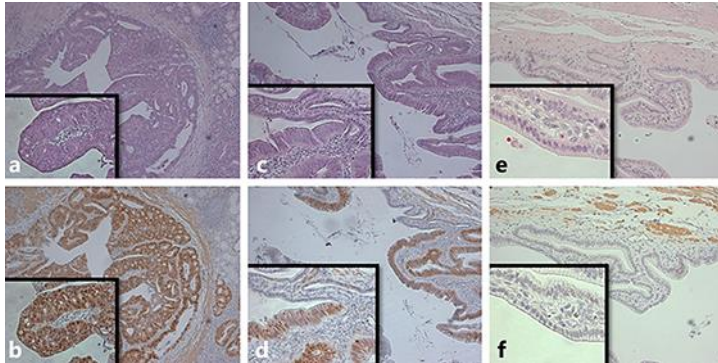


Fig. 3. **a** Examination of the resected specimen revealed a diagnosis of moderately differentiated tubular adenocarcinoma of the papilla of Vater. **b** The cancerous tissue showed p53 expression with immunostaining. **c** The common bile duct, which was retained in the pancreas, was atrophic and replaced by atypical epithelium 1 cm from the papilla of Vater and at the blind end 3 cm from the papilla of Vater. **d** The atypical epithelium of the retained bile duct showed p53 expression with immunostaining. **e** The common bile duct with normal epithelium. **f** The normal epithelium of the bile duct did not show p53 expression with immunostaining. Magnification for all microscopy images, $\times 100$, $\times 200$.