CASE REPORT

Carcinoma of the ampulla of Vater diagnosed after surgical treatment for Lemmel's syndrome

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Abstract Lemmel's syndrome encompasses a range of conditions in which a juxtapapillary duodenal diverticulum exerts mechanical and functional effects on the common bile and pancreatic ducts, leading to jaundice and pancreatitis. In this report, we describe a very rare case of carcinoma of the ampulla of Vater that was detected during postoperative follow-up in a patient who had undergone choledochojejunostomy following a diagnosis of Lemmel's syndrome. We present our clinical and pathological experiences with the diagnosis and treatment of this case as well as a review of the present literature concerning Lemmel's syndrome.

Keywords Juxtapapillary duodenal diverticulum · Lemmel's syndrome · Carcinoma of the ampulla of Vater

Introduction

Juxtapapillary duodenal diverticulum, which is routinely detected in clinical practice via upper gastrointestinal endoscopy and contrast studies, does not exhibit any subjective symptoms in many cases and rarely becomes a clinical problem [1, 2]. However, many pathological conditions comprise Lemmel's syndrome [3] and are characterized by mechanical and functional effects of the juxtapapillary duodenal diverticula on the common bile and pancreatic ducts and the induction of jaundice and

pancreatitis through impaired bile and pancreatic juice excretion.

We experienced a rare case in which a patient was diagnosed with Lemmel's syndrome, underwent choledochojejunostomy (Roux-en Y anastomosis), and was subsequently diagnosed with carcinoma of the ampulla of Vater during postoperative follow-up. This case is reported with a brief literature review.

Case report

A 70-year-old man who had been treated for diabetes mellitus at a neighborhood clinic was referred to our hospital because of hepatic dysfunction in January 2008. After receiving a diagnosis of Lemmel's syndrome, he underwent choledochojejunostomy (Roux-en Y anastomosis) in February 2008. During postoperative follow-up at the neighborhood clinic, he began to exhibit poor glycemic control and weight loss in February 2009 and was referred to our hospital again for a detailed examination in June 2009. Other than his past history of diabetes mellitus, no notable events were observed in either his own life or family history. At the time of the initial surgery, upper gastrointestinal endoscopy (Fig. 1a) revealed a juxtapapillary duodenal diverticulum. A duodenal contrast study (Fig. 1b) revealed similar findings. At the initial admission, endoscopic retrograde cholangiopancreatography was attempted but aborted because the diverticulum prevented identification of the papillary region. Abdominal computed tomography (CT) (Fig. 1c) revealed a stricture of the common bile duct due to the diverticulum and dilatation of the proximal duct but did not indicate intrahepatic bile duct dilatation, stones, or tumors. After the initial admission, the patient's total bilirubin level gradually increased (18.3 mg/dl) and percutaneous transhepatic gallbladder drainage was

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Fig. 1 Imaging findings at the time of the initial surgery **a** Upper gastrointestinal endoscopy reveals a paraduodenal diverticulum (arrow). The duodenal papillary region cannot be observed. **b** Hypotonic duodenography also reveals a paraduodenal diverticulum

(arrow). **c** Abdominal computed tomography revealing stricture of the distal common bile duct due to the paraduodenal diverticulum and proximal dilatation (arrows). There is no evidence of stones, tumors, or other phenomena

Fig. 2 Abdominal computed tomography, Magnetic resonance cholangiopancreatography (MRCP) and Upper gastrointestinal endoscopic findings at readmission (**a**). A dilated pancreatic duct is observed (arrow) and a dilated common bile duct is observed (arrow). There is no apparent evidence of stones, tumors, or other phenomena in the pancreatic head region. **b** As with abdominal computed

tomography, MRCP reveals a dilated common bile duct that had been closed as a blind end in the initial surgery as well as a dilated pancreatic duct (arrows). Although occlusion from the pancreaticobiliary junction to the duodenal papillary region is considered, no apparent abnormalities were observed. **c** A depressed lesion with redness is observed in the duodenal papillary region (arrow)

performed. Based on the above-described course, Lemmel's syndrome was diagnosed and choledochojejunostomy (initial surgery) was performed.

No jaundice was observed upon readmission, although a blood test revealed a mildly elevated transaminase level. The patient's blood glucose and glycosylated hemoglobin levels had increased to 404 mg/dL and 12.7 %, respectively. The following increased levels of tumor markers were also observed: carcinoembryonic antigen, 10.7 ng/mL; carbohydrate antigen 19-9, 374.2 U/mL; sialylated carbonated antigen, 82.0 U/mL; and Duke pancreatic monoclonal antigen type 2 (DUPAN-2), 220 U/mL. Abdominal CT (Fig. 2a) revealed dilated pancreatic and common bile ducts, but no apparent tumor was observed in the pancreatic head region. Similarly, magnetic resonance cholangiopancreatography (Fig. 2b) also revealed a dilated common bile duct that had surgically been closed as a blind

end as well as a dilated pancreatic duct. Although some form of occlusion from the pancreaticobiliary junction to the papillary region was considered, no apparent lesion was observed via imaging. Upper gastrointestinal endoscopy (Fig. 2c) revealed a depressed lesion in the papillary region. A biopsy led to a diagnosis of papillotubular adenocarcinoma. Based on these findings, carcinoma of the ampulla of Vater was diagnosed and pancreaticoduodenectomy (second surgery) was performed. The resected specimen (Fig. 3) revealed a dilated common bile duct, which had been closed as a blind end in the initial surgery, and a hardened, constricted papillary region consequent to the effects of a tumor. A histopathological examination (Fig. 4) revealed malignancy in a 2.5-cm area around the papilla as well as dysplastic diverticular epithelium. The carcinoma of the ampulla of Vater was diagnosed as moderately differentiated adenocarcinoma, pT4 (Du2

Fig. 3 Macroscopic findings of the resected specimen. A dilated common bile duct, which had been closed as a blind end in the initial surgery is observed. The duodenal papillary region is hard and constricted by a tumor (arrow)

Fig. 4 Histopathological findings (hematoxylin–eosin staining, $\times 40$ magnification) malignancy of the duodenal mucosa is observed in a 2.5-cm area around the ampulla of Vater, and the duodenal diverticular epithelium exhibits dysplasia

Panc2), N3, H0, P0, M(–), and Stage IVb [4]. The post-operative course was favorable, and the patient was discharged home 24 days after surgery. However, he developed a recurrence with multiple liver metastases and died approximately 1 year and 6 months after surgery.

Discussion

In 1934, Lemmel collectively described a set of disorders, named “papillen syndrome”, in which juxtapapillary duodenal diverticula compressed the common bile and

pancreatic ducts to obstruct the excretion of bile and pancreatic juice, resulting in jaundice and pancreatitis [3]. At present, this pathological condition is called Lemmel’s syndrome in Japan and is generally not thought to associate with the presence of biliary system stones [5]. This pathological condition is presumed to include not only mechanical compression by the diverticula but also impaired function of the sphincter of Oddi as a result of diverticulitis [6, 7]. Although our case presented with diabetes mellitus and jaundice, no acute pancreatitis or cholangitis was found. Moreover, our search of databases such as PubMed (excluding proceedings) indicated only one previous case of carcinoma of the papilla of Vater presenting with Lemmel’s syndrome [8]. However, to our knowledge, there are no prior reports of any cases in which pancreaticoduodenectomy was performed after surgical treatment for Lemmel’s syndrome similar to what was observed in our case. In one case [8], a scheduled diverticulectomy was canceled and pancreaticoduodenectomy was performed in one stage because papillary carcinoma was diagnosed based on the intraoperative findings. Although Lemmel’s syndrome had been diagnosed preoperatively, there was no apparent imaging finding that showed a diverticulum compressing the common bile duct, as observed in our case. Furthermore, the diverticulum in this previous case was smaller than that in our case. It was assumed that the diverticulum had not affected the morphology of the papillary region or obstructed the view of the region.

In our case, Lemmel’s syndrome was diagnosed by the presence of a juxtapapillary duodenal diverticulum, the absence of stones, and CT images (Fig. 1c) showing stricture of the common bile duct due to the diverticulum and dilatation of the proximal duct. Moreover, because no biliopancreatic system abnormalities were detected for approximately 1 year after surgery, we considered that the diverticulum was very likely responsible. In other words, the initial event of papillary stricture (obstructive jaundice) was assumed to have been caused by the diverticulum, whereas the second event of papillary stricture was caused instead by the papillary carcinoma.

When the patient was diagnosed with Lemmel’s syndrome during the initial hospitalization, the diverticulum prevented sufficient observation of the papillary region. Therefore, it is possible that the papillary carcinoma existed before the diagnosis, although it may have also occurred after diagnosis and treatment. Even if the papillary region had been sufficiently observed, it is also plausible that endoscopy would not have detected abnormalities in this region. Sufficient examination is necessary to reach a diagnosis of Lemmel’s syndrome. Given the previous report of cholangiocarcinoma development during follow-up for Lemmel’s syndrome [9], regular follow-up is

essential even in diagnosed cases. Regarding Lemmel's syndrome and pancreaticobiliary cancer, the number of cases is limited, and there is no clear evidence supporting their association. However, because pancreatitis and cholangitis (choledocholithiasis) are induced in some cases of Lemmel's syndrome, the risk of cancer is concerning when pancreatitis or cholangitis repeatedly occurs [10, 11].

In our patient, the papillary region and diverticulum were observed upon readmission. Accordingly, we cannot rule out the possibility that measures such as reexamination by a different operator or on a different date, procedural innovations [12], the use of different fiberscope models or innovative devices [13], and the clip technique [14] might have allowed observation of this region during the initial hospitalization.

During the process of Lemmel's syndrome diagnosis, as demonstrated by our case, carcinoma of the ampulla of Vater might not be detected at the time of diagnosis and thus remain untreated. A delayed diagnosis is also associated with a risk of reduced treatment opportunities. Therefore, symptoms and findings should not be quickly attributed to juxtapapillary duodenal diverticula (i.e., Lemmel's syndrome). Instead, a detailed examination should be performed while considering the possible presence of tumors in the papillary region.

Conflict of interest There are no financial or other relations that could lead to a conflict of interest.

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