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Gastrointestinal Imaging *Case Report*

Lemmel syndrome, a rare cause of obstructive jaundice by periampullary duodenal diverticulum: Case report and review of the literature

Massupa Krisem[1], Pornrujee Hirunpat[1], Nuttapat Tungtrongchitr[1]

[1]Ramathibodi Medical School, Chakri Naruebodin Medical Institute, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Samut Prakarn, Thailand.

***Corresponding author:**
Nuttapat Tungtrongchitr, Ramathibodi Medical School, Chakri Naruebodin Medical Institute, Faculty of Medicine Ramathibodi Hospital, Mahidol University, Samut Prakarn, Thailand.

ntungtrongchitr@gmail.com

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ABSTRACT

Lemmel syndrome is a pancreaticoduodenal disease caused by compression of the mid or distal common bile duct by a periampullary diverticulum. This condition should be considered a rare complication of a duodenal diverticulum and an unusual cause of obstructive jaundice. Because of its infrequent occurrence and non-specific clinical presentation, Lemmel syndrome can mimic other conditions. We herein report the clinical and imaging findings (computed tomography, magnetic resonance imaging) of a patient who presented with intermittent abdominal pain and jaundice. Large air-filled outpouching lesions of the duodenum compressed the biliary duct, resulting in upstream biliary ductal dilatation that led to the diagnosis of Lemmel syndrome.

Keywords: Lemmel syndrome, Obstructive jaundice, Periampullary duodenal diverticulum

INTRODUCTION

A periampullary diverticulum (PAD) is characterized by the presence of extraluminal outpouchings of the duodenal wall located within a 2–3-cm radius from the ampulla of vater [Figure 1].[1] PADs are usually asymptomatic, and only 5–10% of affected patients show complications.[2] Lemmel syndrome is an uncommon complication of a PAD that presents with abdominal pain and obstructive jaundice due to extrinsic compression of the common bile duct (CBD) by the PAD without evidence of choledocholithiasis or a tumor.[3,4]

CASE REPORT

An 82-year-old man visited the internal medicine department because of a 2-month history of intermittent abdominal pain. He had also lost 1 kg of body weight during the past 6 months. On abdominal examination, neither a mass nor tenderness was palpated.

A complete blood count showed a white blood cell count of 5.57 × 10^9/L (78% neutrophils), hemoglobin concentration of 13 g/dL, and platelet count of 182 × 10^3/L. Liver function studies showed an aspartate aminotransferase concentration of 45 U/L, alanine aminotransferase concentration of 31 U/L, alkaline phosphatase concentration of 180 U/L, gamma-glutamyl transferase concentration of 176 U/L, total bilirubin concentration of 2.1 mg/dL, and direct

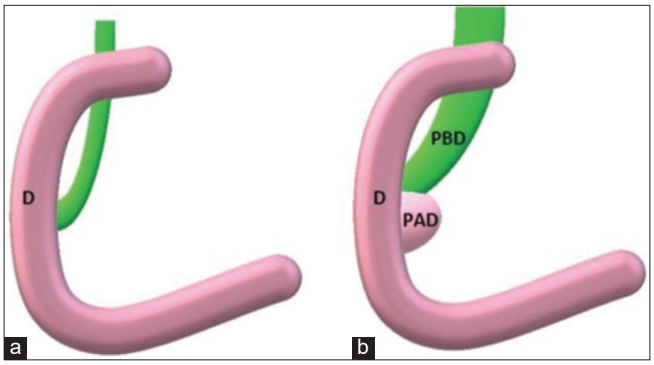

Figure 1: The schematic representation of periampullary diverticulum (PAD). (a) The normal size of PBD which joins to the ampulla of Vater, marking the entry point of bile into the second part of the duodenum. (b) Extrinsic compression of the biliary tract by the PAD causing PBD dilatation. PBD: Principal bile duct, PAD: Periampullary duodenal diverticulum, d: duodenum. The schematic representation was reproduced with permission from "Frauenfelder G, Maraziti A, Ciccone V, Maraziti G, Caleo O, Giurazza F, *et al*. Computed tomography imaging in Lemmel syndrome: A report of two cases. J Clin Imaging Sci 2019;9:23."

bilirubin concentration of 1.0 mg/dL. Hepatitis B surface antigen and anti-hepatitis C virus antibody were negative. The carbohydrate antigen 19-9 and carcinoembryonic antigen concentrations were within normal limits.

Initial abdominal ultrasound revealed CBD and central intrahepatic bile duct dilatation without a demonstrable cause of obstruction. Therefore, the patient was referred for a contrast-enhanced abdominal computed tomography (CT) scan, which also revealed diffuse dilatation of the intrahepatic bile ducts and CBD with abrupt tapering of the distal CBD [Figure 2a and b]. In addition, a large air-filled PAD showing narrow communication with the duodenal lumen was located at the distal CBD [Figure 2c].

Magnetic resonance imaging (MRI) of the upper abdomen and magnetic resonance cholangiopancreatography (MRCP) was performed to better characterize the biliary tract. These imaging examinations also demonstrated a large 8.3-cm air-filled PAD that was mechanically compressing the distal CBD and distal main pancreatic duct, resulting in upstream dilatation of the CBD (1.6 cm in diameter), proximal intrahepatic duct, and pancreatic duct (0.5 cm in diameter), giving the double-duct sign. Coronal T2-weighted MRI of the abdomen was demonstrated in [Figure 3a and b]. There was no evidence of choledocholithiasis or a tumor. Therefore, the presumptive diagnosis was a PAD compressing the bile duct with upstream biliary ductal dilatation and cholestasis, compatible with Lemmel syndrome.

Endoscopic retrograde cholangiopancreatography (ERCP) was performed to confirm the diagnosis. Cholangiography showed dilatation of the CBD with no filling defect [Figure 3c]. The patient was advised and counseled on this condition. At the time of this writing, he had undergone 6 months of follow-up, during which time he had remained asymptomatic with normal laboratory values.

DISCUSSION

The duodenum is the second most common site in the gastrointestinal tract for diverticulum formation after the colon, with an incidence rate of about 22% of the population.[2] The most common type of duodenal diverticulum is a PAD. Patients with PADs are usually asymptomatic. Complications can occur in about 5% of patients and include hemorrhage, perforation, fistula formation, diverticulitis, cholangitis, pancreatitis, gallstone formation, and rarely obstructive jaundice.[3,5,6] Lemmel syndrome is a type of obstructive jaundice caused by extrinsic compression of the biliary tract by the PAD without stones or tumors.

Various pathogeneses of Lemmel syndrome have been proposed, including the following. One possible pathogenesis involves chronic fibrosis of the papilla (papillitis chronica fibrosa) caused by direct mechanical irritation due to chronic inflammation of the papilla or chronic periampullary diverticulitis.[7,8] Second, a PAD may cause dysfunction of the sphincter of Oddi.[8,9] Third, the CBD or ampulla may be mechanically compressed by the PAD, resulting in obstructive jaundice as occurred in our patient.[10]

The reported characteristics of PADs vary in size depending on individual cases. They can range in size from very small, around 2–3 mm in diameter, to very large, up to 5 cm.[1,11] While duodenal diverticula are common, the occurrence of a large PAD (>4 cm) is rare. In addition, having huge diverticulum compress both of the CBD and pancreatic ducts, resulting in jaundice representing Lemmel syndrome and causing double duct sign like in our case, is also uncommon.

To the best of our knowledge, only a few reports have described a duodenal diverticulum associated with the double-duct sign that mimicked a malignancy.[12] The presence of double duct sign is typically associated with carcinoma of the pancreatic head and an ampullary tumor, which were differential diagnoses in our case.[13] However, in our patient, CT and MRI showed the double duct caused by the compressive effect of the large PAD. Furthermore, MRCP and ERCP demonstrated biliary duct dilatation without a filling defect. Pancreatic head and ampullary malignancies were considered unlikely because there was no evidence of a focal pancreatic or ampullary mass and no abnormal enhancement or restricted diffusion areas that are typically seen in malignancies. In addition, the patient had an uneventful clinical course throughout 6 months of follow-up with normal tumor marker concentrations.

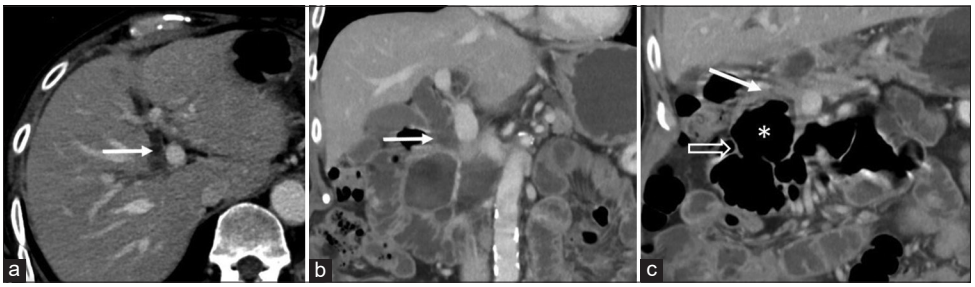

Figure 2: An 82-year-old man with Lemmel syndrome who presented with intermittent abdominal pain and jaundice. (a and b) Axial and coronal contrast-enhanced computed tomography (CT) scan of the abdomen shows dilation of the intrahepatic duct (arrow in a) and common bile duct (CBD) (arrow in b) without demonstrable cholelithiasis. (c) CT image with coronal reconstruction shows a large air-filled periampullary duodenal diverticulum (asterisk) showing narrow communication with the duodenal lumen (open arrow) was located adjacent to the distal CBD (arrow in c).

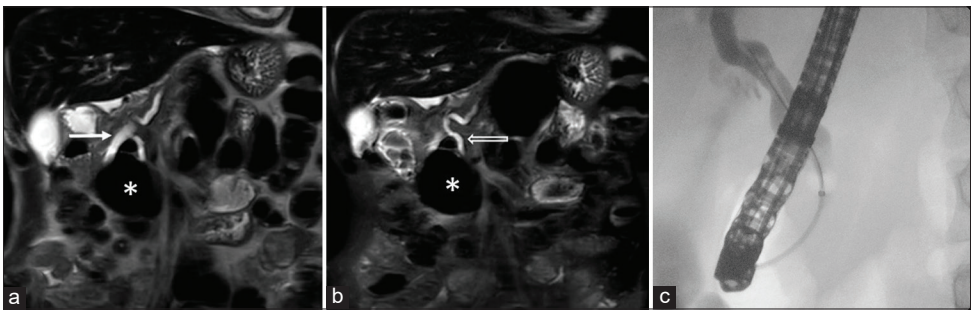

Figure 3: An 82-year-old man with Lemmel syndrome who presented with intermittent abdominal pain and jaundice. (a and b) Coronal T2-weighted magnetic resonance imaging of the abdomen shows an 8.3-cm periampullary diverticulum (asterisk) mechanically compressing the biliary duct with distal common bile duct (CBD) (arrow in a) and pancreatic ductal dilatation (open arrow in b). Occluded cholangiogram showed dilated CBD without filling defect (c).

A few reports have described content within the diverticular. A diverticulum usually contains air, an air-fluid level, food content, stone, or debris. Its appearance can sometimes mimic a pancreatic pseudocyst, pancreatic abscess, cystic pancreatic tumor, metastatic lymph node, or choledochal cyst.[3] Our patient had only an air-filled PAD with no sign of inflammation. Furthermore, there were no signs of remote carcinoma or metastatic disease elsewhere, making these conditions unlikely in this case. The importance of internal contents of a PAD remains a topic of debate in the medical literature. The presence of internal contents such as stones, debris, or bile secretion in the PAD can potentially obstruct the opening of the pancreatic or biliary ducts, leading to complications such as choledocholithiasis, pancreatitis, or cholangitis. However, the extent to which the internal content of the diverticulum plays a role in overall risk of complications is not clear and may depend on a variety of factors. Hence, the future research is needed to determine its significance in the diagnosis, management, and treatment of this condition.

Imaging is important to identify and diagnose Lemmel syndrome. Ultrasound may help to evaluate biliary ductal dilatation, but it cannot be used to examine bowel pathology. CT and MRCP are the imaging modalities of choice to diagnose and rule out stones or other periampullary diseases. ERCP is the gold standard for diagnosis and can be performed along with endoscopic intervention.[14]

CONCLUSION

Lemmel syndrome is a rare cause of obstructive jaundice that should be considered in the differential diagnosis of biliary obstruction. CT, MRCP, and ERCP are essential tools for the diagnosis and exclusion of other pancreaticobiliary diseases. Recognition of this condition should be kept in mind when imaging studies show both a PAD and double-duct sign.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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