



JUST BECAUSE IT IS RARE DOESN'T MEAN IT IS IMPLAUSIBLE: LEMMEL'S SYNDROME A CASE SERIES

Gastroenterology

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ABSTRACT

Lemmel's syndrome is a rare pancreaticobiliary complication of duodenal diverticula. It occurs when a duodenal diverticulum causes obstructive jaundice due to a mechanical obstruction of the common bile duct. Other mechanisms like sphincter of Oddi dysfunction can also play a role in pathophysiology. We report three cases of Lemmel's syndrome where liver biochemistry showed obstructive jaundice; but subsequent MRCP showed a massive periampullary diverticulum causing biliary obstruction. Early detection and intervention can prevent needless additional investigations and complications due to obstruction.

KEYWORDS

periampullary diverticulum, obstructive jaundice, biliary obstruction.

INTRODUCTION

Lemmel syndrome is defined as an obstructive jaundice caused by a periampullary duodenal diverticulum compressing the intrapancreatic part of the common bile duct with resultant upstream dilatation of the extra- and intra-hepatic bile ducts [1]. Pseudo-diverticula, or duodenal diverticula, are extraluminal sac-like out-pouchings of the duodenal mucosa that lack a muscularis layer.[2]. Periampullary duodenal diverticula (PAD) are labelled when they form within a 2–3 cm radius of the ampulla of Vater.[3]. PAD are usually asymptomatic, but when they are inflamed at the same time, they may cause pancreaticobiliary complications. [4]. In the case of Lemmel's syndrome, obstructive jaundice may develop as a result of PAD without the presence of choledocholithiasis or a tumour [5]. Here, we present three cases of Lemmel's syndrome.

case 1

A 64-year-old man presented with a 10-day history of progressive jaundice and discomfort in the right upper quadrant of the abdomen, recent onset of fever with chills. There is no history itching, pale stools, weight loss, vomiting, GI bleed. He underwent cholecystectomy for gallstones four years back. On assessment, the patient was awake and alert, with icterus, no pallor, and moderate right upper quadrant tenderness. Lab results revealed an elevated total count on hemogram; Liver function test (LFT) showed direct hyperbilirubinemia (2.4mg/dl) with elevated alkaline phosphatase (ALP) (225IU/L) and GGT of (140IU/L). Serum amylase and lipase were normal; and viral markers (HBsAg, HCV) were negative. Magnetic resonance cholangiopancreatography (MRCP) showed a calculus in the remnant gallbladder, along with a diverticulum in the second part of the duodenum, causing distal CBD compression. During the Endoscopic retrograde cholangiopancreatography (ERCP) PAD was visualized and narrowing in the distal CBD was confirmed (Figure1). A biliary stent was placed during the procedure. Post procedure LFT after 2 days showed down trending of total bilirubin and alkaline phosphatase.

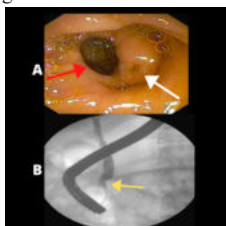


Figure1

A- Endoscopic image of peri ampullary diverticulum (red arrow) in close proximity to the ampulla (white arrow)

B- ERCP image showing the site of obstruction (yellow arrow) caused by the diverticulum.

CASE 2

60-year-old woman with compensated chronic liver disease (CLD) Non-alcoholic steatohepatitis (NASH) related presented with right upper quadrant pain for the past two weeks. She did not have a history of jaundice, abdominal distention, GI bleed, or altered sensorium. Her physical examination was normal. Her LFT showed normal bilirubin, aspartate transaminase (AST), alanine aminotransferase (ALT) but ALP (198 IU/l), Gamma-glutamyl transferase (GGT) (130 IU/L) were elevated. An ultrasound of the abdomen showed chronic liver parenchymal disease with dilated common bile duct (CBD). An MRCP was done to evaluate dilated CBD, which revealed a diverticulum arising from the medial aspect of the second part of duodenum compressing the distal CBD causing upstream dilatation of the biliary system. The PAD and biliary system dilatation were confirmed during the ERCP procedure (Figure 2). Biliary system was stented following which ALP, GGT normalized.

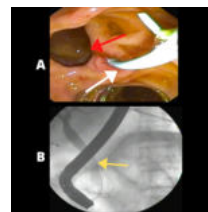


Figure 2

A- Endoscopic image of peri ampullary diverticulum (red arrow) close to the ampulla with a biliary stent in-situ (white arrow)
B- ERCP image showing a dilated biliary system (yellow arrow)

CASE 3

A 40-year-old man presented with right upper quadrant pain, jaundice, and fever for the past two days. On physical examination he was Icteric, and right hypochondrium was tender. His LFT showed total bilirubin (9.8mg/dL), direct bilirubin (6.38 mg/dL), ALP (481 IU/L), GGT (566 U/L). CT scan showed a peri ampullary diverticulum measuring 2cms x 1 cm in the second part of the duodenum compressing the intrapancreatic distal CBD causing upstream dilatation of the biliary system.

ERCP showed two large diverticula in the second part of the duodenum and narrowing of distal CBD (Figure 3). Biliary stenting was done in same sitting. Post procedure his general condition and LFT parameters improved. He was discharged after 3 days with no complications.

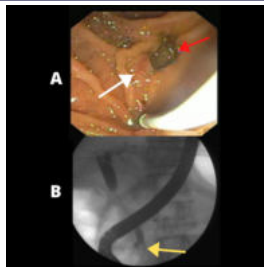


Figure 3

- A- Endoscopic image duodenal diverticula (red arrow) with papilla hidden within it, papilla(white arrow) seen after grasping the mucosal fold below the ampullary ridge of the diverticulum.
 B- ERCP showing a dilated biliary system with distal CBD narrowing (yellow arrow)

DISCUSSION

Diverticula are sac-like protrusions of the whole or a portion of the bowel wall that can occur anywhere in the digestive tract [1]. The colon is the most common location for diverticula, followed by the duodenum [6]. Duodenal diverticula have been found in 2–5% of patients undergoing upper gastrointestinal barium studies and in 7% of patients undergoing ERCP [7]. PAD are duodenal diverticula that occur within 3cm of the ampulla of Vater [8]. Most periampullary diverticuli are asymptomatic and identified by accident, but they can sometimes cause pancreaticobiliary or non-pancreaticobiliary complications.

Haemorrhage, diverticulitis, perforation, and fistula formation are examples of non-pancreaticobiliary complications. Recurrent gallbladder or bile duct stones, obstructive jaundice, acute pancreatitis, or ascending cholangitis are manifestations of pancreaticobiliary complications [9]. Lemmel's syndrome was first identified in 1934 by Lemmel's as obstructive jaundice caused by a periampullary duodenal diverticulum in the absence of gallstones [1]. Computed tomography (CT) scan, MRCP, and barium studies can be used to diagnose this disease. PAD can appear as thin-walled cavitory lesions on the wall of the second portion of the duodenum on CT scan and MRCP. PAD is seen in barium studies as contrast-filled out-pouchings that emerge from the descending duodenal wall [1].

ERCP with sphincterotomy and biliary stent placement is the preferred procedure, which has been linked to a lower risk of morbidity and mortality [10]. Diverticulectomy or biliodigestive anastomosis are two other surgical choices. In cases of obstructive jaundice without choledocholithiasis or tumour, Lemmel's syndrome should be treated as a differential diagnosis. Ignoring the likelihood of this will result in repeated jaundice and possibly cholangitis, raising the patient's risk of mortality and morbidity.

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