Case Report

Lemmel's Syndrome: Lesson Based on a Case Report

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Abstract

Lemmel's syndrome is a bile duct disease caused by periampullary duodenal diverticula that develop within 2–3 cm of the Vater papilla. This disease manifests itself as nonobstructive jaundice. In most cases, duodenal diverticula do not cause disease, and only in a small percentage of patients, diverticula cause biliary tract obstruction by extrinsic compression. If the compression is severe, in the long term, it can become complicated with lithiasis and cholangitis. Diagnosis is very difficult, and recurrent biliary symptoms must be directly related to the compression of the duodenal diverticula. Imaging is essential for differential diagnosis and includes conventional contrast radiographs, endoscopic retrograde cholangiopancreatography, computed tomography, and magnetic resonance imaging. The investigations show the dilation of the intra- and extra-hepatic bile ducts in the absence of lithiasis or main pancreatic duct dilatation, compressed by the diverticula, which most frequently originate from the medial wall of the second duodenal tract. The treatment of choice is surgical with removal of the diverticula. Failure to diagnose can cause serious health complications for the patient.

Keywords: Biliary ducts, computed tomography, duodenum diverticula, Lemmel's syndrome, resonance magnetic imaging, ultrasound

Introduction

Lemmel's syndrome (LS) was first described in 1934 by Lemmel[1] as obstructive jaundice caused by a periampullary duodenal diverticulum (PAD) in the absence of biliary lithiasis [Figure 1] that develops at a distance of <2–3 cm from the Vater papilla (VP).[2] The 95% of the PADs are asymptomatic, while 1%–5% cause the syndrome. The most frequent symptoms are right upper quadrant pain, elevated level of transaminases, pancreatic enzymes, and bilirubin. If the bile ducts compression is severe, in the long term, it can be complicated by lithiasis and cholangitis. Recurrent biliary symptoms must have a direct relationship to the compression caused by PAD. Complications related to the diverticulum are: diverticulitis, perforation, bleeding, fistulas, and intestinal obstruction; the pancreatic biliary complications are: obstructive jaundice, lithiasis, acute pancreatitis, and cholangitis. According to the authors, the mechanisms that cause LS include the following:

1. The distal choledochus (common bile duct) or VP can be compressed directly by the PAD which typically contains enterolith or bezoar[3]
2. PAD-induced dysfunction of Oddi sphincter[4]
3. Diverticulitis can cause chronic papillitis and papilla fibrosis.

Instrumental diagnosis includes ultrasound (US), X-ray with contrast medium, endoscopic retrograde cholangiopancreatography (ERCP), computed tomography (CT), and magnetic resonance imaging (MRI). The typical onset of disease is intra- and extra-hepatic bile ducts dilatation in the absence of lithiasis or hepatic ducts dilatation and/or pancreatic duct dilatation. The PAD is most frequently found on the medial wall of the second tract of the duodenum. The symptomatic intermittence is due to the morphological characteristics of the PAD which, in most cases, has a large orifice and allows the food material to be easily evacuated; on the contrary, when the orifice is narrow, the alimentary material accumulates in the PAD, increases the bile ducts compression. The treatment of choice is surgical with the removal of the diverticulum. In elderly patients or subjects at risk, surgery can be avoided by emptying the contents of the diverticulum via ERCP.[5]

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Case Report

An 81-year-old male patient was referred to our imaging department from the medical clinic department of our hospital for intense epigastric pain, elevated transaminases, with a clinical history of cholecystectomy for lithiasis, diverticulosis, gastritis, and frequent episodes of cholangitis for some years. The patient was clinically underweight and had been complaining of low-grade fever for several days. The results of the laboratory tests were as follows: white blood cells: 11,170/μL, C-reactive protein: 2,343 mg/dL, total bilirubin: 2.28 mg/dL, alkaline phosphatase: 379 IU/L, aspartate aminotransferase: 98 IU/L, alanine aminotransferase: 93 IU/L, γ - glutamyl transpeptidase: 581 IU/L. He was subjected to US, MRI, CT, and ERCP. A May Lab "Nine" ultrasound device (Esaote Genova), with convex 3–7.5 Mhz probe, CT device Optima 64 slice (GE Healthcare) and a 1.5 Tesla MRI scanner (Signa, GE), were used. US of liver highlighted: absence of bile ducts dilatation in the right [Figure 2a] and left bile ducts dilatation [Figure 2b], dilatation of common hepatic bile duct, choledochus dilation with a hyperechoic appearance of the biliary walls, and intraluminal stone. The Magnetic Resonance has allowed to highlight: 1. A roundish hypointense mass originating from the second duodenal segment compatible with PAD [Figure 3a]. 2. Dilatation of left intrahepatic bile ducts, common hepatic duct (16mm in diameter) and choledochus (11mm in diameter) and intraluminal stone in the choledochus with a diameter of 10mm [Figure 3b]. 3. Slow enhancement of the choledochus walls after contrast medium administration. 4. Slight dilation of the Wirsung (5 mm in diameter). 5. Vater Papilla hypertrophy. To rule out other abdominal pathologies, the patient underwent CT which confirmed PAD [Figure 3c], left intrahepatic bile ducts dilation and biliary cyst [Figure 3d], bile ducts lithiasis and cholangitis. ERCP confirmed PAD which had a wide (2 cm) collar with fluid/air content, VP hypertrophy, and allowed extraction of choledochus stone. The patient was subsequently treated with anti-inflammatory and antibiotics drugs with regression of pain, fever, and laboratory values normalization; after a week was discharged.

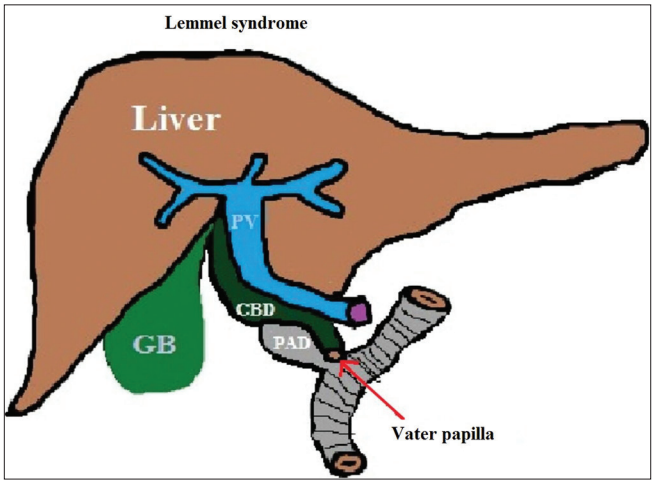

Figure 1: Summary diagram of the anatomical structures involved in patients with LS. In LS, the PAD compresses the CBD. PV: Portal vein, GB: Gallbladder, CBD: Common bile duct, PAD: paravaterian diverticulum, LS: Lemmel syndrome

Discussion

In LS, diagnostics play an essential role in patient management and PAD can also be studied using Conventional Radiology with oral administration of contrast medium that allows it to be localized and show its morphology. CT and MRI are second-level examinations that in addition to PAD can highlight complications of the disease such as cholangitis, biliary tract lithiasis, Wirsung dilation, and pancreatitis. In CT and MRI, PAD generally appears as a cavity originating from the second section of the medial duodenal wall, near the papilla, bounded by thin walls, and may contain air, liquids, enteroliths, or bezoars; in some cases, the papilla may be inside the diverticulum. However, this aspect of PAD is not specific and can sometimes be confused with other pathologies such as pancreatic neoplasms, secondary lymph nodes, pancreatic pseudocysts, and abscesses; therefore, the study must always be deepened with ERCP, which is the most suitable procedure because it allows the direct visualization of PAD, its content and the relationship it contracts with the VP. ERCP also allows to intervene in eventual biliary tract lithiasis, to place a stent in the biliary tract to favor a rapid inflammation regression and allows the emptying of the PAD. PAD, while causing obstructive jaundice in the absence of gallstones, favors the biliary tract lithiasis; in fact, according to some authors, the excessive proliferation of bacteria that produce beta-glucuronidase causes the deconjugation of the glucuronides of bilirubin that precipitate as calcic bilirubin stones; therefore, it is possible in the long term the formation of biliary tract stones in patients with LS.[6] US is the first level examination for the study of the biliary tract, but due to the artifacts generated by intestinal meteorism, the duodenum

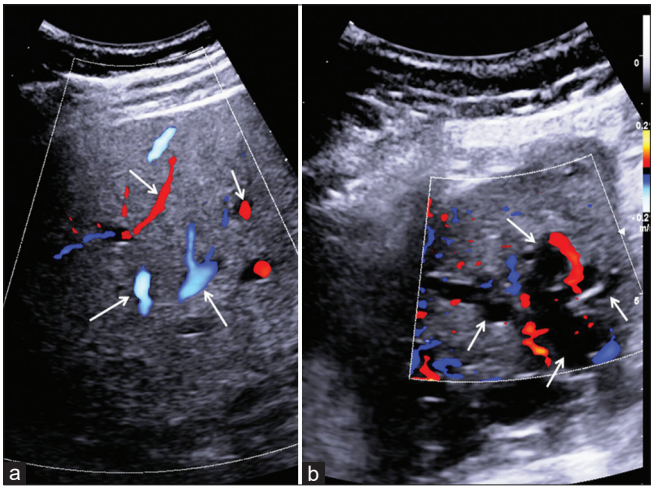

Figure 2: Ultrasound of the liver. (a): Ultrasound scans show no dilation of the right bile ducts; Hepatic veins (arrows) and (b) left bile ducts dilatation (arrows)

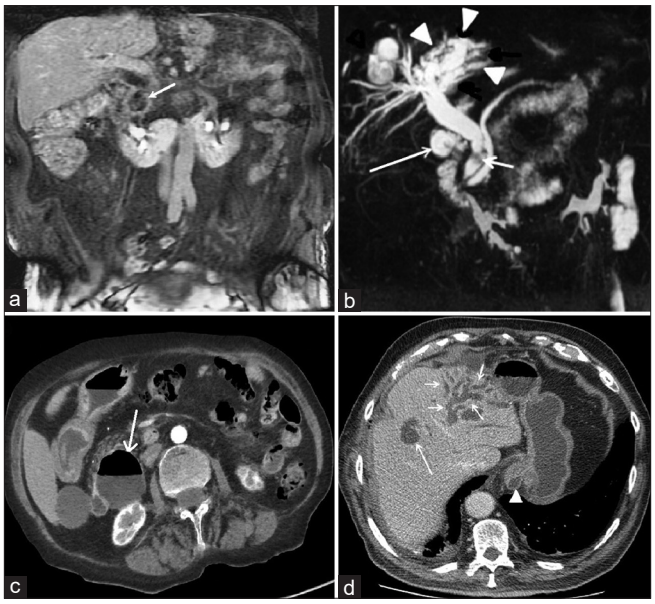

Figure 3: CT and MRI findings of LS. (a) MRI in coronal view shows PAD (arrow). (b) MRI-cholangiography shows left hepatic bile ducts dilatation (arrows head). PAD (long arrow). choledochus lithiasis (short arrow). (c) The CT in axial view shows a rounded image with an air/fluid level corresponding to the PAD (arrow). (d) CT in axial view shows left bile ducts dilation (short arrows). Biliary cyst (long arrow). CT: Computed tomography, MRI: Magnetic resonance imaging, LS: Lemmel's syndrome, PAD: Paravaterian diverticulum

cannot be studied ultrasonographically; however, this method can provide useful elements for the LS suspicion, in fact, the intrahepatic biliary tract dilatation and the absence of stones, associated with the chronicity of symptoms and laboratory alterations, should lead to suspicion of PAD; however, many patients are still discharged after antibiotic and anti-inflammatory medical therapy without a definitive diagnosis. A similar case occurs for the diagnosis of superior mesenteric artery syndrome for which ultrasound, while not allowing the diagnosis of duodenal compression, in the presence of typical symptoms (vomiting, nausea, and weight loss) and indirect signs (reduction of aortomesenteric angle), reinforces the suspicion of LS and provides indications for further investigation with second level investigations.[7] After having ascertained the PAD, to demonstrate compression of the biliary tract, it is necessary that the diverticulum is full of intestinal contents, and this is not always possible, especially when the neck of the diverticulum is wide and promotes emptying. In our case, the suspicion of LS was induced by two factors:

1. The MR, in addition to highlighting the PAD, showed a significant prolapse of the VP which is almost always caused by extrinsic compression
2. After a careful analysis of the clinical history, we discovered that the patient had already undergone several hospitalizations for the same symptomatology, during which he had been subjected to instrumental investigations (US, CT, and MR) that had not shown lithiasis of the biliary tract, so the stone we discovered could not be the cause of the chronic painful episodes due to cholangitis but probably a long-term complication.

The most suitable second-level examination, in our opinion, is the CT because it allows to highlight the dilatation and inflammation of the biliary tract, locate the PAD and to exclude other causes of compression (vateromas, papillitis, neoplasms, and lymph nodes hepatic hilar). The management of LS depends on symptoms and complications. In paucisymptomatic patients, conservative pharmacological treatment may be sufficient, while in patients with chronic obstructive jaundice, recurrent pancreatitis, hemorrhages, perforation, intestinal obstructions, the treatment must be surgical with the removal of the diverticulum.[8] When surgery is not possible, laparoscopic diverticulectomy may be a viable alternative.[9] In patients who are too old or in poor physical condition, emptying the diverticulum with ERCP can cause immediate decompression of the biliary tract, and combined with extracorporeal shock wave lithotripsy[10] also allows the extraction of intrahepatic stones.

Conclusions

LS is a rare disease and should be suspected in patients with recurrent episodes of obstructive jaundice in the absence of biliary tract lithiasis or in patients with recurrent episodes of acute pancreatitis in absence of biliary tract lithiasis. US is the first-level examination and can induce diagnostic suspicion but requires diagnostic integration with second-level methods to highlight the PAD, any complications and exclude other causes of compression. Failure to diagnose LS can expose patients to serious health complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent form. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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