An Unusual Cause of Biliary Tract Obstruction: Lemmel Syndrome

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ABSTRACT

BACKGROUND: Lemmel syndrome is a rare and misdiagnosed etiology of obstructive jaundice due to a periampullary duodenal diverticulum causing a mechanical obstruction of the common bile duct. It represents an obstructive jaundice with the absence of choledocholithiasis or pancreaticobiliary tumors. It is an underreported entity due to the absence of specific pathognomonic signs.

CASE PRESENTATION: A 77-year-old-woman admitted for sepsis, due to an ascending cholangitis, underwent a MRCP and a gastroduodenoscopy revealing Lemmel's syndrome. Due to failure of ERCP, the patient underwent surgical derivation.

CONCLUSION: Lemmel syndrome represents an uncommon diagnosis of obstructive jaundice, that shouldn't be neglected if no other organic cause is detected. It is usually asymptomatic, however some patients can develop symptoms and complications such as cholangitis, as is the case of our patient. Imaging allows diagnosis, with MRCP as the modality of choice to confirm diagnosis. Endoscopy is the first line treatment.

KEYWORDS: Lemmel, jaundice, imaging, MRCP, endoscopy

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Introduction

Lemmel syndrome is a rare and misdiagnosed etiology of obstructive jaundice, due to a periampullary duodenal diverticulum causing a mechanical obstruction of the common bile duct. It represents an obstructive jaundice in the absence of choledocholithiasis or pancreaticobiliary tumors.

It is an underreported entity due to the absence of specific pathognomonic signs.

We report the case of a 77-year-old woman, admitted for acute jaundice, to which the diagnosis of a Lemmel syndrome was retained.

Case Presentation

A 77-year-old-woman, with history of chronic smoking and drinking for over 45 years, was admitted for a progressive worsening jaundice with tea-colored urine, clay-colored stool, upper abdominal discomfort, accompanied by fever, chills, and asthenia.

In the emergency room, the patient's fever reached 39°C, associated with tachycardia at a 105 beats per minute.

Her physical examination showed frank jaundice, a non-distended abdomen with mild right upper quadrant tenderness. No hepatomegaly or large palpable gallbladder was palpable.

Laboratory tests revealed an obstructive pattern on liver function tests with alkaline phosphatase = 458 UI/L, γ-glutamyl transferase = 1289 UI/L, total bilirubin = 169 mg/dL; an aspartate aminotransferase = 107 UI/L and an alanine transaminase = 158 UI/L; associated with an increased CRP level of 250 mg/L and a white blood cell count of 16 545/mm^3.

The clinical and laboratory results were highly suggestive of a hepatobiliary sepsis due to ascending cholangitis.

Blood cultures were positive for Escherichia Coli. The patient received antibiotics: ceftriaxone + metronidazole for 7 days, improving recovery of her clinical and biological signs of infection.

Ultrasound of the abdomen revealed a dilated common bile duct with no sign of biliary lithiasis.

A magnetic resonance cholangiopancreatography (MRCP) performed (Figure 1) showed intra- and extra-hepatic biliary dilatation at the level of the ampulla.

Three very large fluid and air filled periampullary duodenal diverticula were also noted measuring: 41 × 25, 24 × 21, and 27 × 20 mm.

A gastroduodenoscopy confirmed the presence of the 3 large peri-duodenal diverticula in D2 (Figure 2).

Imaging and endoscopic findings showed no obstacle other than the 3-giant diverticula, suggesting a Lemmel Syndrome complicated by acute cholangitis.

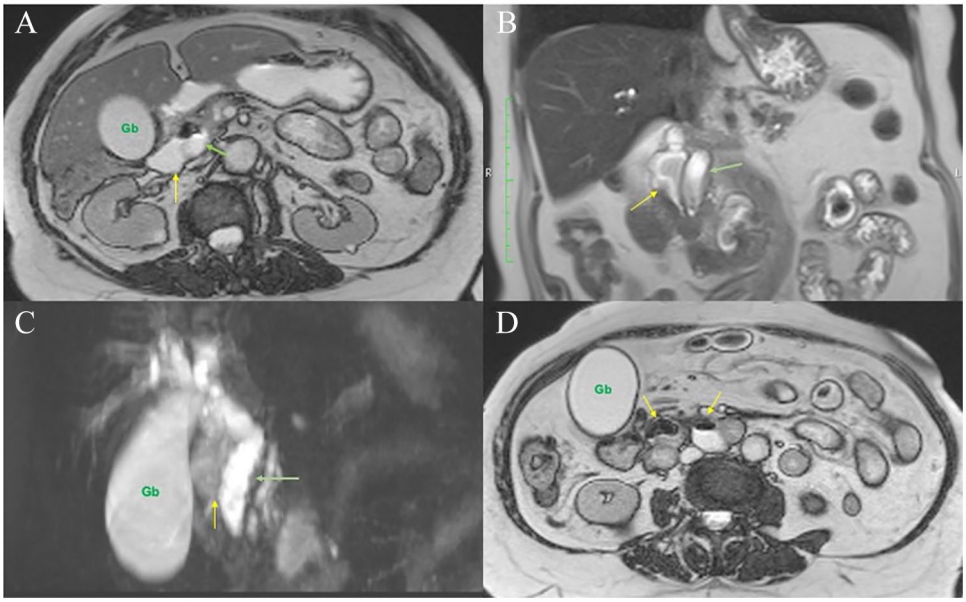

Figure 1. MRCP in axial T2 weighted images (A, D), coronal T2 weighted images (B) and 3D bili sequence (C), showing an important common bile duct dilatation (green arrow; A-C), a gallbladder dilation (Gb; A, C, D) above 3 peri-ampullary duodenal diverticula containing fluid and air (yellow arrow; A-D) causing the obstruction.

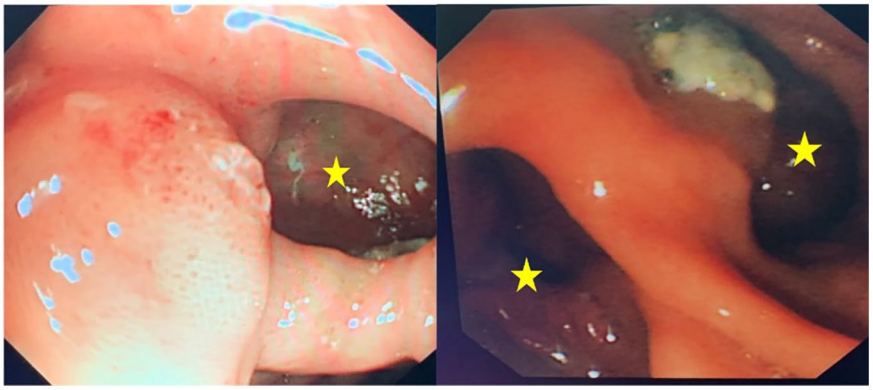

Figure 2. Endoscopic image showing 3 periampullary duodenal diverticula (yellow stars).

Due to technical difficulties during the ERCP procedure, the family requested surgical treatment and the patient had biliary derivation with an end-to-side Roux-en-Y choledocho-jejunostomy technique performed.

She was discharged 2 weeks later with regression of jaundice and good follow-up.

Discussion

Lemmel syndrome was first described in 1934 by Lemmel,[1] as an obstructive jaundice in the absence of gallstones or neoplasm due to a periampullary duodenal diverticulum (PAD).[2] PAD is an extraluminal diverticulum of the duodenal wall located within 2 to 3 cm to the ampulla of Vater.[3]

Its physiopathology isn't certain and specific, but reported mechanisms include 3 mains causes, all due to the proximity of the diverticulum to the ampulla, either causing direct external compression of the common bile duct/ampulla leading to obstruction; or causing a dysfunction of the sphincter of Oddi; or causing direct irritation of the ampulla leading to its chronic inflammation and eventually fibrosis of the papilla.[4-7]

These periampullary duodenal diverticula are often asymptomatic but they may become symptomatic in only 1% to 2% of the cases causing acute abdominal pain and simulating a biliopancreatic colic by an extrinsic obstruction of common bile duct or pancreatic duct.[8]

They can also cause complications in 5% of the cases including[6]:

- *Non-pancreaticobiliary complications* such as: hemorrhage, diverticulitis, perforation, or fistula formation.
- *Pancreaticobiliary complications* such as: acute pancreatitis, cholangitis, bile duct stones, or obstructive jaundice causing the Lemmel Syndrome.

This syndrome is only considered and retained in the absence of choledocholithiasis or any other etiology of obstructive jaundice.

Imaging is the examination of choice to confirm diagnosis. Its modalities include: ultrasonography, CT scan, or MRCP.

- *Ultrasonography*: is usually the first imaging method in the ER showing indirect signs of biliary obstruction: dilation of the biliary ducts.
- MRCP is the examination of choice to confirm diagnosis and evaluate biliary tract anomalies and eliminate differential diagnosis.
- CT scan is usually the first line imaging method to either lead or confirm diagnosis due to its fast acquisition and availability. It can be done with intra veinous contrast enhancement or oral intake of contrast product.

Lemmel syndrome is usually shown as a cavitary lesion of the medial wall of the second or third duodenal portion, either containing gas or fluid, causing obstruction of the distal part of the common bile duct and dilation of the biliary tract above.

Oral contrast intake CT reveals communication of the cavity with the duodenum, confirming diagnosis of the PAD, but also showing its expansion resulting in compression of the common bile duct.

Enhanced contrasted CT and MRCP allow better characterization of the lesion especially when it is filled with fluid, compared to its differential diagnosis: pancreatic abscess, pancreatic pseudo-cyst, pancreatic head cystic neoplasm, or a metastatic lymph node.[4,7,8]

Endoscopic retrograde cholangiopancreatography allows a direct visualization of the diverticulum.[5]

As PAD are usually asymptomatic, treatment is only required when the patient becomes symptomatic, and endoscopic treatment, such as, papillary balloon dilatation or endoscopic sphincterotomy with biliary stent placement represents the treatment of choice, showing a good success rate; however, recurrence rate stays high.

In case of endoscopic treatment failure, surgical approach is preferred and it either includes a trans-duodenal diverticulectomy or an end-to-side Roux-en-Y choledochojejunostomy.[2,3] But these procedures are difficult and have high mortality risk.

Sometimes, a trans-duodenal sphincteroplasty is considered.[5,8]

Conclusion

Lemmel syndrome represents an uncommon diagnosis of obstructive jaundice, that shouldn't be neglected if no other organic cause is detected.

It is usually asymptomatic; however, some patients can develop symptoms and complications such as cholangitis, which was the case for our patient.

Imaging represents the modality of choice to confirm diagnosis especially through MRCP, and endoscopy is the first line treatment.

Informed Consent

Informed consent of the patient was obtained for publication of this case report.

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