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Case report

Lemmel's syndrome: Presentation of an uncommon cholangitis cause and a risk factor for failed endoscopic retrograde cholangiopancreatography. Case report

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ABSTRACT

Introduction: Lemmel's syndrome is a rare pathology that presents jaundice or dilation of common biliary tract, secondary to duodenal diverticula. Due to infrequency, a high suspicion has to be considered to avoid omitting this diagnosis, mainly in patients who present direct hyperbilirubinemia without choledocholithiasis.

Presentation of case: A 76-year-old female admitted to the emergency department with cholangitis, secondary to an ampullary diverticulum (papilla was located inside the diverticulum). As a consequence, the endoscopic retrograde cholangiopancreatography failed and a biliodigestive derivation (choledochal duodenum anastomosis) was performed.

Discussion and conclusions: This case is an example of an unusual clinical and anatomical presentation of duodenal diverticulum. This unusual presentation is an example of the importance of not overlooking a diagnosis, which can lead to more severe complications such as cholangitis. It is also important to consider that cases like this are risk factors for failed endoscopic management and surgical possibilities procedures should also be considered in the management.

1. Introduction

Lemmel's syndrome is a rare pathology that presents jaundice and/or dilation of the common biliary tract, secondary to duodenal diverticula [1,2]. Due to its infrequency, a high suspicion is required, to avoid omitting this diagnosis, mainly in direct hyperbilirubinemia patients without choledocholithiasis [3]. The described syndrome is based mainly on the clinical presentation of the patient. Although the initial management is endoscopic, there are particular cases in which it fails, and it's necessary to resort to surgery [4].

We present a case of a 76-year-old female admitted to the emergency department with cholangitis, secondary to an ampullary diverticulum (papilla inside diverticulum). As a consequence, the endoscopic retrograde cholangiopancreatography failed and a biliodigestive derivation (choledochal duodenum anastomosis) was performed.

Here, we present a literature review that reflects the rare presentation of this pathology and the established endoscopic or surgical management.

This work has been reported in line with the SCARE criteria [5].

2. Case description

A 76-year-old female patient, without clinical, familiar or surgery history, admitted to the emergency department with moderate abdominal pain in the right hypochondrium, with generalized jaundice, fever, choluria and absence of weight loss precedents.

Complementary tests revealed an increased direct bilirubin, elevated liver enzymes and negative tumoral markers, which points out a cholestatic pattern, that suggest an obstructive pathology. The patient was admitted with cholangitis diagnose for clinical treatment and assessment. Furthermore, the ultrasound showed a dilated intra and extrahepatic bile duct. In addition, tomography and nuclear magnetic

Abbreviations: ERCP, endoscopic retrograde cholangiopancreatography; MRCP, magnetic resonance cholangiopancreatography; CT, computed tomography.

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resonance cholangiography (MRCP) presented dilatation of the intrahepatic bile duct up to 7.6 mm, without intrahepatic lithiasis, 14 mm hepatocholedocus without inner lithiasis (Fig. 1A). At its distal end, a thinning of benign appearance is observed due to extrinsic compression related to a middle side diverticulum of the second duodenal portion which measures 29 × 24 mm (Fig. 1B).

The patient underwent an endoscopic retrograde cholangiopancreatography (ERCP), in which the papilla cannulation failed, due to its location inside of the duodenal diverticulum. Therefore, surgery was decided, to solve the benign secondary cholangitis obstruction caused by duodenal diverticulum (Lemmel's syndrome). Biliary digestive derivation was performed, a choledocho-duodenal anastomosis, by a general surgeon (Figs. 2 and 3). The post-surgical evolution was satisfactory, the patient remained hospitalized for 5 days until the antibiotic scheme was completed. No complications were reported. She stayed under control for 2 years and she did not present episodes of biliary obstruction nor cholangitis.

3. Discussion

Lemmel's Syndrome is an uncommon cause of jaundice or common biliary tract dilation. The main causes are choledocholithiasis or pancreatic-biliary and periampullary tumors [2]. Because of the infrequency of the pathology, a high awareness is required to avoid overlooking the diagnosis in patients with direct hyperbilirubinemia without choledocholithiasis and should be supported by complementary studies, as it was performed with our patient [3].

Diverticula are protrusions with sac shape of all or segments of the intestinal wall that can occur anywhere in the gastrointestinal tract. They can occur at any part of the gastrointestinal tract. The most frequent place where diverticula are located is in the colon followed by duodenum. Duodenal diverticula have been reported in 2–5% of patients undergoing upper gastrointestinal barium studies and in 7% of patients undergoing endoscopic retrograde cholangiopancreatography (ERCP) [6].

Diverticula are classified in extraluminal and intraluminal, the majority are extraluminal. The extraluminal diverticula are an acquired hernia due to a defect in the intestinal wall as a consequence of vessel entrance. Therefore, its incidence is higher with age [9]. The intraluminal duodenal diverticulum is a rare congenic anomaly as a result of the incomplete cannulation of the intestinal lumen [2]. Duodenal diverticula are frequently located 3 cm from the ampulla of Vater, these diverticula are denominated periampullary diverticulum, peripapillary or perivaterian [8,9]. Diverticula which contain papilla

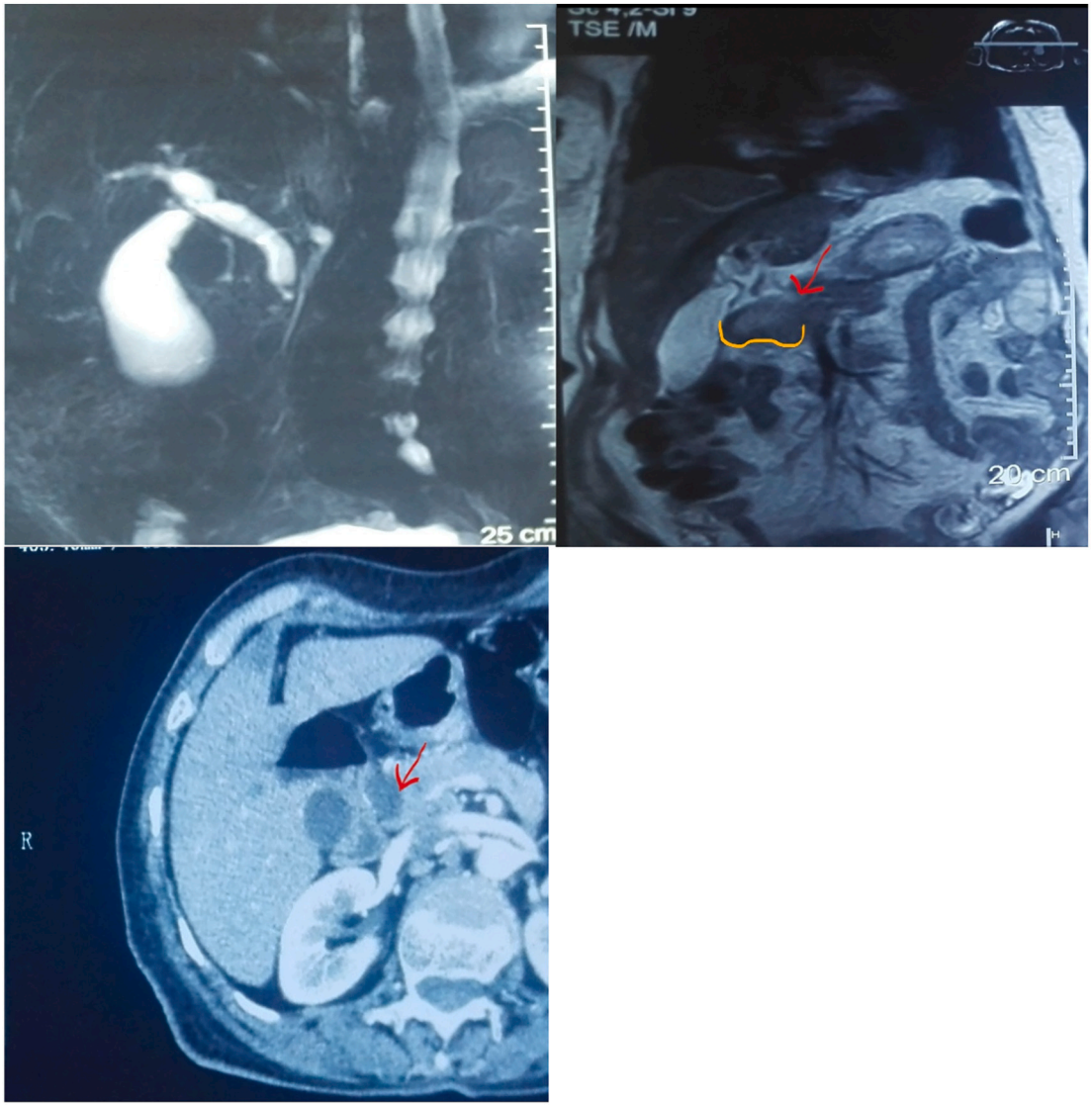

Fig. 1. A: MRCP: Choledocus dilation without calculous. B: MRCP: Diverticulum at the middle duodenal wall C. Computed axial tomography scan (CT): duodenal section.

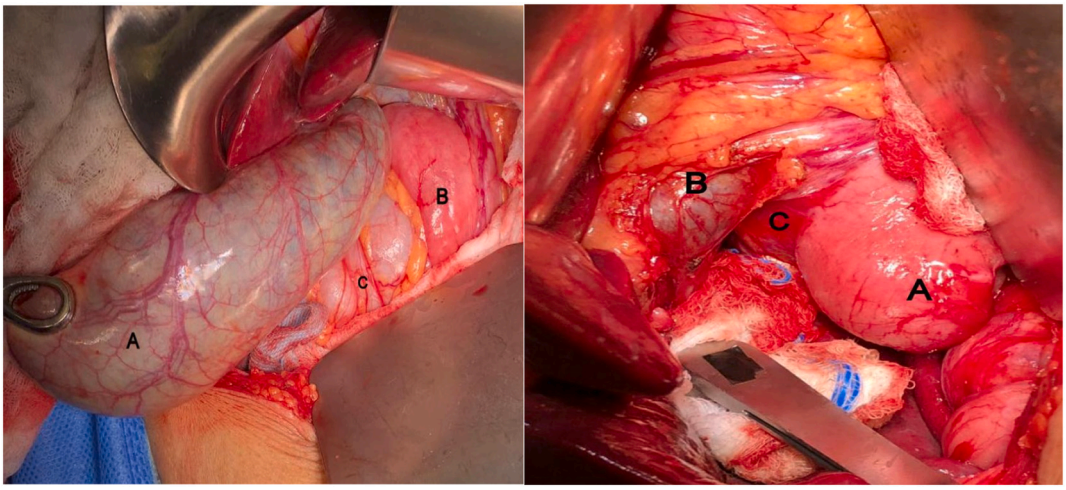

Fig. 2. A: Surgery beginning, structure identification and cholecystectomy, Gallbladder (A), Duodenum (B) and Colon (C).
B: Duodenum identification (A) Biliary tract (B) and diverticulum (C).

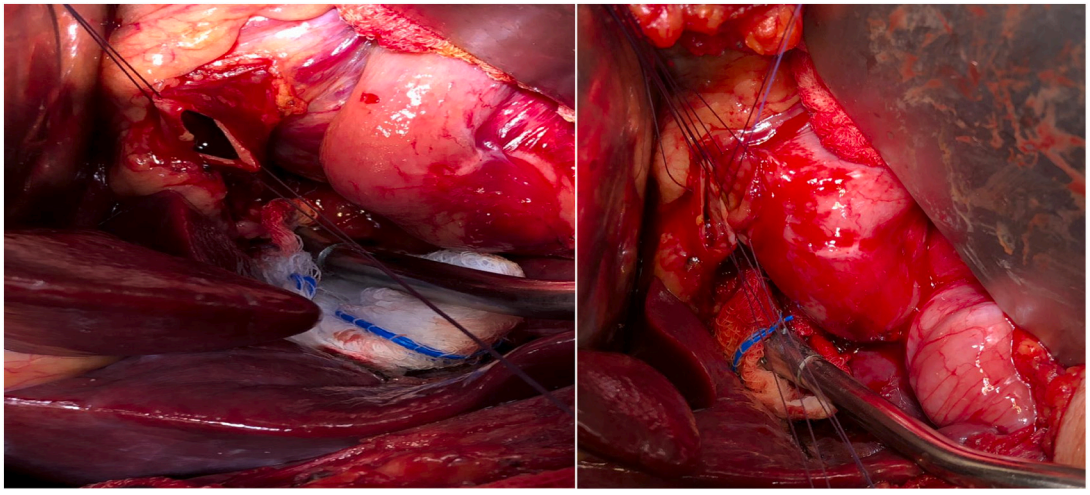

Fig. 3. A: Biliary tract preparation for anastomosis; anchoring spots are placed.
B: Finished Choledocho duodenum Anastomosis.

(intradiverticular papilla) are also known as ampullary diverticula and according to the previously described characteristics, our patient's case corresponds to an extraluminal ampullary diverticulum as shown in the MRCP. Diverticula which do not contain papilla are also named juxta-papillary or juxtampullary [7].

Currently, the Li-Tanaka classification has been proposed. It is based on the number of periampullary diverticula and their anatomic relation to the major papilla. Type I describes the case when papilla has an intradiverticular location and it is not located next to the edge of the diverticulum. Type II describes the case when papilla is located at the edge of the diverticulum or less than 1 cm outside of the edge (IIa inside the edge and IIb less than 1 cm outside the edge). Type III describes when its location is more than 1 cm outside edge and type IV describes when two or more diverticula are present (IVa if papilla is located in the edge of one of the diverticula and IVb if the papilla is located more than 1 cm from both diverticula) [8].

Only 5% of the patients present symptoms. Therefore, the syndrome is usually revealed as an incidental finding, during the study of other disease or when performing a upper endoscopy [1]. Among symptomatic cases, like our patient, most of Lemmel's syndrome patients show jaundice, abdominal pain or acute cholangitis, these symptoms could manifest intermittently [9]. Regarding laboratory tests, most of the cases present leukocytosis, an increase of inflammatory markers such as globular sedimentation speed and reactive c protein, an increment of direct and total bilirubin, hepatic enzymes, alkaline phosphatase and gamma-glutamyl transferase [10].

Imaging is extremely important to identify and diagnose Lemmel's syndrome accurately [11]. Ultrasound could reveal biliary tract dilation. However, its ability to examine intestinal pathology is limited, therefore it cannot identify duodenal diverticula, which could be assessed by CT, MRCP or ERCP. This last one method could be therapeutical [2]. In our patient's case, two image studies were performed to establish the diagnosis.

In CT and MRCP, periampullary diverticula could appear as cavitary lesions of the thin walls of the duodenal second portion mid walls as it has been described in our case [11]. Regarding the ERCP, literature reports that a lateral vision endoscope during the procedure is considered as the gold standard [4]; and that endoscopic sphincterotomy is the treatment of choice in case a biliary or pancreatic complication appears, [1] as it is associated with a low risk of morbidity and mortality [6]. Noting that if the ampulla is intradiverticular, then, its cannulation can be difficult [1]; as it happened in our case in which the papilla cannulation failed, due to its location inside of the duodenal diverticulum. Then, surgical treatment was decided.

Duodenal diverticula management is based on clinical presentation. In these cases, asymptomatic patients are not generally treated unless complications occur. While in oligosymptomatic patients, conservative treatment consists of nasogastric decompression and A wide spectrum antibiotic treatment in case of perforation [2,12]. Although, endoscopic intervention, ball dilation and extracorporeal shock wave lithotripsy have been described [6].

Surgery is reserved for cases in which endoscopic and conservative treatment fail. Although, literature considers diverticulectomy as the standard surgical procedure [4,10], currently, there is no consensus about the surgical technique to be performed [1]. Surgical resection is particularly difficult in this region as it often implies retroperitoneal duodenum mobilization [9]. Diverticulectomy is always associated with a high morbimortality due to a risk of biliary pancreatic tract lesion. Consequently, it is imperative to locate the ampulla of Vater through duodenotomy, with an anterograde form through the cystic tract or by choledochotomy [1]. Some authors mention that associating biliary digestive derivation lowers the risk of lesion [6]. Even so, an enterogastric derivation, especially if there is a local inflammation or diverticular perforation risk [1]. In our case, it was decided to perform a derivation, after the ERCP management failed, to warrant proper biliary flux and to solve the initial cholangitis complication.

4. Conclusion

In patients with cholangitis, in absence of choledocholithiasis or neoplastic pathology, Lemmel's syndrome has to be considered to avoid overlooking the diagnosis.

In the cases in which endoscopic treatment through ERCP fails, considerated the procedure of choice, surgery is indicated to solve the obstructive process.

The presence of periampullary diverticula is a cause of difficult biliary tract cannulation during ERCP.

Ethical approval

We have written consent of the patient to publish this article.

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Credit authorship contribution statement

Ruth A Rojas – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Guarantor

The corresponding author is the guarantor of submission.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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Declaration of competing interest

Nothing to declare.

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