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Case Report

Lemmel Syndrome as Rare Cause of Secondary Sclerosing Cholangitis: An Unusual Presentation and Literature Review

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Abstract: Lemmel's syndrome is a rare and misdiagnosed cause of obstructive jaundice, It corresponds to cholestatic disease secondary to compression of the main bile duct by a periampullary duodenal diverticulum with secondary dilation of the extra- and intra-hepatic bile ducts. Approximately 5% of duodenal diverticles can cause symptoms and 1% have complications, with colangitis being the most common. Lemmel syndrome is a type of intermittent obstructive jaundice without the presence of coledocolithiasis. Late diagnosis of this entity is common and may lead to unnecessary further investigations and therapeutic delay. There are no cases reports published associated with secondary sclerosing colangitis.

Keywords: Lemmel syndrome, Juxtapapillary duodenal diverticula, Obstructive jaundice, Sphincter of Oddi dysfunction.

INTRODUCTION

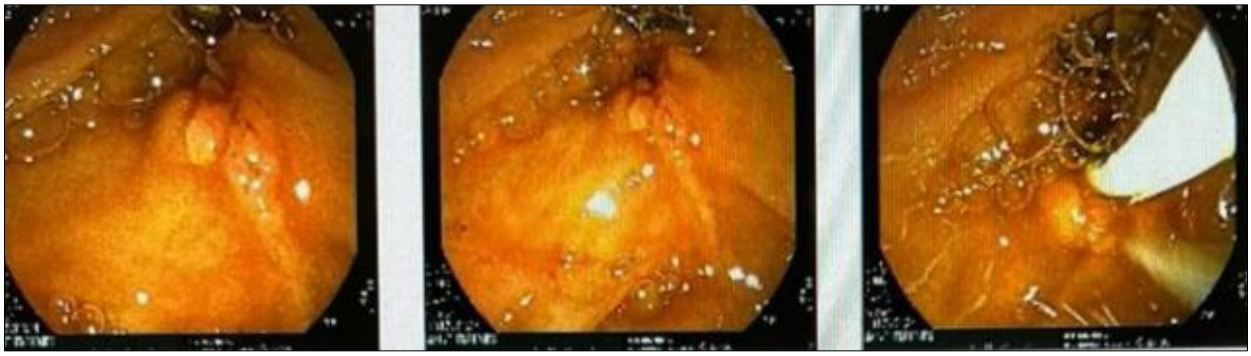
Duodenal diverticula are considered to be infrequent occurrences, with reported incidences approaching up to 20%. These entities gradually gained recognition over an extensive period of time and were initially documented by Chomel, a renowned French pathologist, in the year 1710. The duodenum is the second most prevalent location for diverticula inside the gastrointestinal tract, following the colon. Juxtapapillary or periampullary diverticula (DPA) are typically situated in the second or third segment of the duodenum, namely along the medial wall, provided they are within a distance of less than 2.0 cm from Vater's bulb. Incidental identification is a common occurrence in the recognition of duodenal diverticula. The diagnosis is mostly established with a side-viewing endoscope, specifically employed for the purpose of endoscopic retrograde cholangiopancreatography (ERCP). Lemmel's syndrome is an atypical etiology of obstructive jaundice, characterized by the lack of gallstones and periampullary tumors. This condition develops due to biliary mechanical compression caused by a juxtapapillary diverticulum. The procedure of accurately diagnosing this condition can pose challenges because to its lack of distinct clinical presentation and the potential for its radiological findings to resemble other acute intraabdominal processes.

CASE REPORT

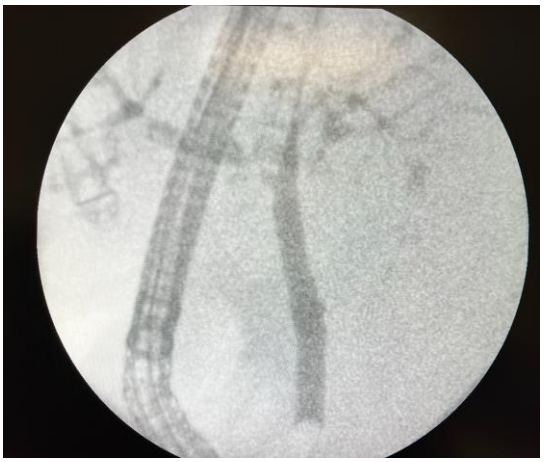
A 64-year-old male patient, without a history of chronic degenerative conditions, presented to the general surgery department with abdominal pain in the right hypochondrium colic type. The pain did not radiate and was accompanied by fever, nausea, and vomiting. The patient was diagnosed with acute cholecystitis and underwent laparoscopic cholecystectomy, which had a

satisfactory outcome without any complications. Four weeks after the surgery, the patient returned to the emergency department with complaints of abdominal pain, jaundice, and fever. Liver function tests revealed abnormalities, and acute cholangitis and coledocolithiasis were diagnosed. The condition was successfully resolved through endoscopic retrograde cholangiopancreatography (ERCP), during which a yuxtapapillary diverticulum (known as Lemmel Syndrome) was observed. While hospitalized, the patient experienced another episode of severe cholangitis, prompting the initiation of medical management and a repeat ERCP. The procedure revealed irregularities in the intrahepatic segments of the liver tract, including dilatation and stenosis of the biliary pathway. Our patient presented with features of cholangitis with elevated TLC, ESR, CRP, and deranged LFT and RFT though blood culture was negative. We made the diagnosis confidently with help of similar clinically history, MRCP, CECT of whole abdomen.

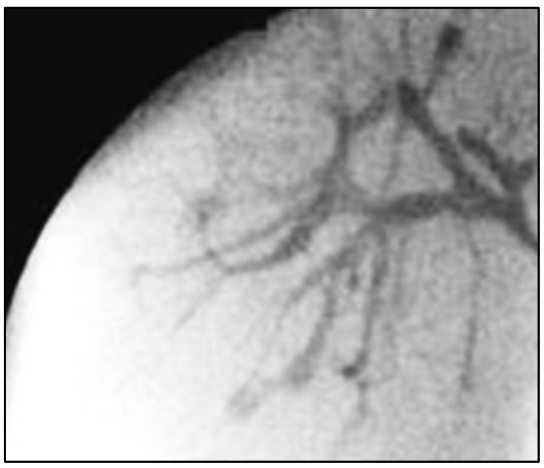
FIGURES



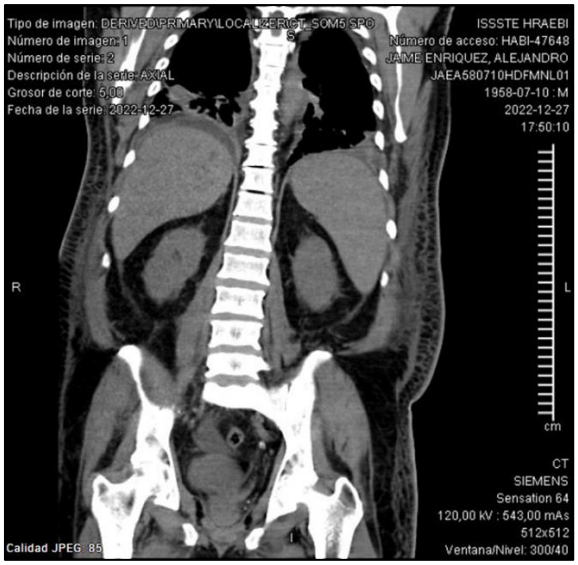
Obstruction of the primary bile duct caused by a periampullary duodenal diverticulum Internal-external biliary drainage catheter



Fluoroscopic image obtained shows a large filling defect



CP-IRM sequences showing intra- and extra-hepatic biliary dilatation



TC Abdomen with chronic macroscopic hepatopathy

DISCUSSION

Diverticula are anatomical formations characterized by the presence of sac-like protrusions in the gastrointestinal tract, which can manifest in any region of the gut wall. The duodenum is the second most prevalent location for diverticula in the digestive tract, with the colon being the most common site. It is then followed by the jejunum, ileum, and stomach. The typical anatomical location of this structure is frequently observed in the second segment, in close proximity to the ampulla of Vater. The prevalence of duodenal diverticula is around 20% among individuals in the overall, non-pathological population. Duodenal diverticula can be categorized into two types: extraluminal and intraluminal. The intraluminal type is typically congenital and occurs as a result of incomplete recanalization of the intestinal lumen. On the other hand, the extraluminal type is the most prevalent and is acquired when weakened mucosa herniates due to protruding large vessels. This type of diverticula can be found either in the intraretropancreatic seat or in the papillary region. This phenomenon is frequently observed in medical practice, however it is characterized by a very low level of auditory disturbance. It is common for patients to exhibit no symptoms. The identification of the duodenal diverticulum typically occurs around the fifth decade of life, either coincidentally during upper endoscopies or due to associated difficulties. In contrast to the sigmoid diverticulum, complications arise in approximately 1-5% of instances involving the duodenal diverticulum.

The disorder known as Lemmel's syndrome was initially documented by Lemmel in 1934. It is characterized as a cholestatic disease that arises as a result of the obstruction of the primary bile duct caused by a periampullary duodenal diverticulum. Various etiologies exist with respect to the pathophysiology underlying the emergence of Lemmel syndrome. The potential explanation of this phenomenon might be attributed to mechanical irritation of the pancreatico-biliary ampulla, resulting in the development of chronic inflammation and subsequent fibrosis of the papilla. Sphincter dysfunction can be caused by peripheral artery disease (PAD) in certain instances. Lemmel's syndrome may arise as a result of mechanical compression exerted on the distal common bile duct or ampulla by pancreatico-duodenal (PAD) structures. The clinical presentation characterized by recurring episodes of ascending cholangitis, along with corresponding imaging abnormalities, indicates the most probable diagnosis. In the case of Lemmel syndrome, it is common for patients to exhibit symptoms such as jaundice, pain in the abdomen, or acute cholangitis, which may manifest repeatedly. The individual in question exhibited clinical indicators that are indicative of cholangitis, such as an increased level of C-reactive protein and abnormal results from liver function tests.The characteristic computed tomography (CT) presentation of a duodenal diverticulum is characterized as a slender-walled, circular accumulation of gas and oral contrast substance located adjacent to the inner edge of the junction between the second and third segments of the duodenum. Duodenal diverticula can exhibit regions of high signal strength, which can be attributed to the presence of fluid, as well as regions of low signal intensity, which can be attributed to the presence of gas, as observed on T2-weighted magnetic resonance imaging.

The diagnosis of Lemmel syndrome can be achieved through the direct observation during endoscopic retrograde cholangiopancreatography, utilizing a side-viewing endoscope. However, the utilization of CT and MRI, due to their ability to generate images in several planes, can provide a noninvasive means of diagnosing periampullary disorders while also facilitating the elimination of alternative conditions. The available treatment modalities encompass surgical resection, endoscopic intervention, extracorporeal shock

wave lithotripsy, and conservative care, as illustrated in the presented vignette. The surgical excision in this anatomical area is a special challenge because to the frequent necessity of manipulating the retroperitoneal duodenum. The successful treatment of Lemmel syndrome, which arises from chronic fibrous papillitis or malfunction of the sphincter of Oddi, involves the implementation of endoscopic sphincterotomy to alleviate the obstruction in the biliary system. In conclusion, Lemmel syndrome is an infrequent etiology of obstructive jaundice and warrants consideration in the differential diagnosis of biliary blockage when there is evidence of a periampullary duodenal diverticulum. Maintaining a heightened level of suspicion is crucial in order to achieve a precise diagnosis, as Lemmel syndrome has the potential to imitate several noncancerous and cancerous anomalies in the periampullary area.

CONCLUSION

Lemmel syndrome is a medical disorder that exhibits a range of characteristics that lead to changes in both the microscopic and macroscopic structure. Consequently, this sequence of events ultimately gives rise to the occurrence of cholecystitis and coledocolithiasis, so causing a protracted obstruction within the biliary system. The obstruction leads to a condition known as biliary stasis and colangitis, which subsequently facilitates the development of pigment stones and inflammatory stenosis. These pathogenic mechanisms contribute to the worsening of cholestasis, ultimately resulting in the progression of sclerosing cholangitis. Sclerosing cholangitis is a medical condition characterized by a relatively low occurrence rate and a tendency to often be inadequately identified. The condition poses a diagnostic and therapeutic dilemma as a result of its unpredictable prognosis, marked by elevated death rates and fast advancement towards hepatic cirrhosis. In the end, liver transplantation emerges as the ultimate therapeutic recourse for this specific medical disease.

CONFLICTS OF INTERESTS

The authors declare no conflicts of interest. The patient has signed the informed consent form and consented to the use and publication of confidential information and images for scientific and non-profit purposes. The submitted paper has been read and endorsed by all authors.

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