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Lemmel's Syndrome: A Rare Cause of Obstructive Jaundice by Periampullary Diverticulum

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Abstract: ***Obstructive jaundice is a common clinical presentation, most often caused by choledocholithiasis, pancreaticobiliary and periampullary tumors, or other conditions such as strictures or cholangitis. Although* periampullary *duodenal diverticula are frequently asymptomatic, they can sometimes present with obstructive jaundice-in the absence of gallstones or other obstructing lesions like tumors-a condition known as Lemmel's syndrome. Given the rarity of Lemmel's syndrome, a high index of suspicion is necessary to avoid mismanagement, such as unnecessary surgery or misdiagnosis, such as attributing the jaundice to more common causes like gallstones or tumors, and ensure timely appropriate treatment. Treatment depends on patient presentation and may involve conservative management, surgical procedures in the form of excision of the diverticulum, or even endoscopic sphincterotomy or stenting. We report a case of Lemmel's syndrome, successfully diagnosed via magnetic resonance cholangiopancreatography (MRCP) and further confirmed with computed tomography (CT) imaging.***

Keywords: periampullary duodenal diverticulum; lemmel's syndrome; obstructive jaundice; MRCP

1. Introduction

Obstructive jaundice is most often caused by choledocholithiasis and biliary/periampullary tumors. However, it is crucial to be aware of uncommon differential diagnoses to ensure adequate management [1]. Lemmel's syndrome should be considered in the differential diagnosis of obstructive jaundice when choledocholithiasis, pancreaticobiliary/periampullary tumors, and other causes such as primary sclerosing cholangitis or strictures are absent [2]. Periampullary diverticula (PAD) are extraluminal outpouchings of the duodenum that originate within 2-3cm from the ampulla of Vater [3]. While most PADs are asymptomatic, symptomatic PADs are likely underdiagnosed because there are no specific signs or symptoms on imaging [4]. While endoscopic retrograde cholangiopancreatography (ERCP) is the gold standard for diagnosis and allows for therapeutic intervention, computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) are often used for initial evaluation and can suggest the diagnosis. Endoscopic ultrasound can also be a helpful adjunct [5].

2. Case Presentation

An 82-year-old female presented to the hospital with complaints of dark yellow colored urine, white stools and icterus since few days. She denied abdominal pain, itching, fever, vomiting, hematemesis, melena, hematochezia, or weight loss. She had no significant past surgical history and denied any prior episodes of jaundice or abdominal pain. On physical examination, her vitals were stable. Laboratory workup revealed significantly elevated total bilirubin (15.9 mg/dL), direct bilirubin (14.8 mg/dL), ALP (414 U/L), ALT (74.8 U/L) and AST (82.7 U/L,). Viral markers for acute hepatitis were also negative. The CBC showed a normal white blood cell count, and electrolytes were within normal limits. Lab parameters were in favour of obstructive jaundice. Ultrasound of abdomen revealed mild dilatation of intrahepatic biliary ducts, common bile duct and main pancreatic duct. Magnetic resonance pancreaticography (MRCP) was done for further evaluation with an adjunct non-contrast computed tomography (CT) scan.

3. Imaging Findings

MRCP revealed mild dilatation of the common bile duct, intrahepatic biliary radicals and main pancreatic duct [Figure 1]. Coronal T2 weighted and axial T2 weighted [Figure 2] images shows a well-defined T2 hypointense area in the region of pancreatic head which was seen communicating with adjacent duodenal wall representing a duodenal diverticulum. It is seen compressing the intrapancreatic

common bile duct and main pancreatic duct with resultant upstream dilatation. The air filled diverticulum was confirmed on the plain computed tomography scan [Figure 3].

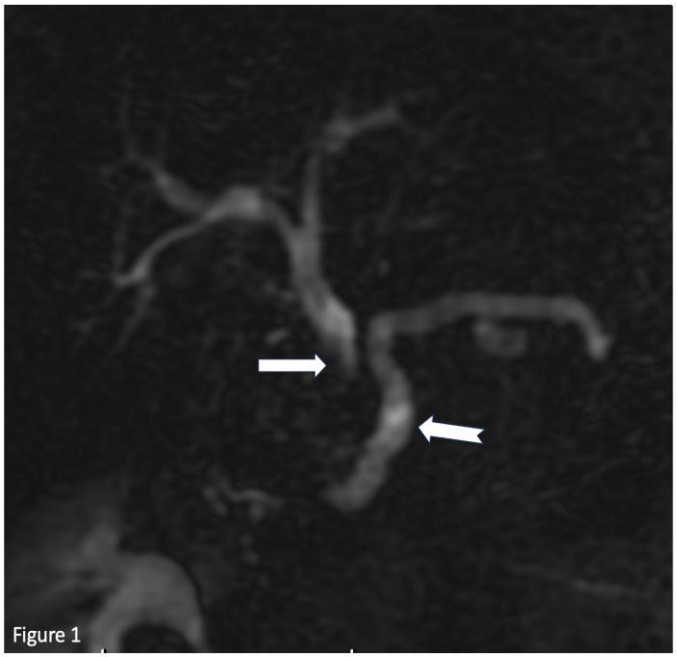

Figure 1: 3D MRCP shows smooth tapering of distal common bile duct with upstream dilatation of common bile duct (white arrow) and main pancreatic duct (notched white arrow).

Figure 2: Coronal T2 weighted (A) and axial T2 weighted (B) images shows a T2 hypointense area representing a duodenal diverticulum (star) which is seen compressing the intrapancreatic common bile duct (chevron) and main pancreatic duct (arrow) with resultant upstream dilatation.

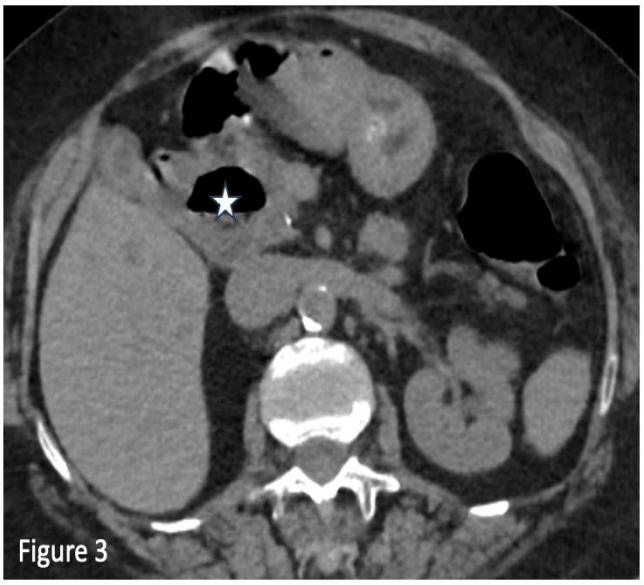

Figure 3: Computed tomography plain study shows air filled outpouching arising from duodenum confirming duodenal diverticulum (star).

The patient underwent endoscopic retrograde cholangiopancreaticography (ERCP) allowing a sphincterotomy with placement of a biliary stent. Post operative laboratory workup showed significant improvement in the parameters.

4. Discussion

Obstructive jaundice caused by periampullary duodenal diverticulum (PAD) in absence of choledocholithiasis or tumor is known as Lemmel syndrome [2]. Duodenal diverticula occur in 5 to 10% of the general population, with incidence increasing with age , typically being discovered around 80 years old [1]. Diverticula often arise from the second or third part of the duodenum [6]. Juxta-ampullary diverticula, those located within 2-3 cm of the ampulla of Vater, represent 75% of duodenal diverticula and are most commonly implicated in Lemmel's syndrome [6,7]. The precise pathophysiology remains unclear, but several mechanisms have been proposed. These include: 1) Chronic fibrosis of the papilla, possibly secondary to periampullary diverticulitis and chronic inflammation of the ampulla; 2) Functional obstruction due to the location of the diverticulum affecting the sphincter of Oddi; and 3) External compression of the common bile duct or ampulla of Vater by the diverticulum itself [7]. The clinical symptoms of Lemmel's syndrome, are nonspecific, meaning they can mimic other biliary or abdominal conditions. The main manifestations include right hypochondrial pain, recurrent attacks of acute abdominal pain (which may be colicky in nature), and jaundice [4]. Patients may also experience nausea, vomiting, or weight loss. Diagnosis of Lemmel's syndrome relies on imaging. Barium studies can identify the diverticulum but aren't primary. CT and MRCP visualize the diverticulum and biliary tree; unenhanced scans may show a thin-walled outpouching with gas or fluid, potentially mimicking a lymph node or pancreatic pseudocyst, especially if large and near the pancreas. Oral or/and intravenous contrast improves accuracy [4]. CT helps in differentiation of intraluminal air and calculus, both of which are visualized as filling defects on MRCP. ERCP, using a side-viewing endoscope for optimal ampulla visualization, is the gold standard, allowing both diagnosis and therapeutic interventions like sphincterotomy or stenting [8]. The majority of these periampullary diverticula are asymptomatic and only occasionally cause complications such as diverticulitis, perforation,and fistula formation. Diverticula may also complicate pancreatobiliary tree leading to obstructive jaundice, choledocholithiasis, cholelithiasis, cholangitis and acute pancreatitis [9]. Excision of the periampullary diverticulum is indicated when biliary obstruction is present. This is a complex surgical procedure, due to the anatomical location and proximity to vital structures, and is associated with increased mortality and morbidity [8]. In high-risk cases, such as those with significant patient comorbidities or where surgical expertise is a concern, less invasive endoscopic alternatives like sphincterotomy or stenting are suitable options [8].

5. Conclusion

Lemmel's syndrome is a less commonly recognized cause of biliary tract obstruction. It is diagnosed when a periampullary diverticulum is seen compressing the extrahepatic bile duct in a case of obstructive jaundice and all other common causative pathologies are ruled out. Magnetic resonance cholangiopancreatography plays a vital role in evaluation of the biliary tree and eliminate other differential diagnoses. Computed tomography helps in confirming the diagnosis of diverticulum due to presence of intraluminal air or positive oral contrast within the diverticulum.

Conflict of Interest: None declared

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