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Case Report

Lemmel Syndrome: A Rare Cause of Right Hypochondrial Pain Secondary to Duodenal Diverticulum

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Abstract

Although, it is a reflex to link right hypochondrial pain and obstructive jaundice to choledocholithiasis, there are many rare causes out of which Lemmel syndrome is one of them. It occurs when a duodenal diverticulum causes mechanical obstruction of the common bile duct. Imaging modalities play a vital role in the diagnosis as it can be easily misdiagnosed. We report the case of a 55-year-old male presenting with right hypochondrial pain and loss of appetite. On physical examination, there was tenderness in the right upper quadrant. Initial blood investigations revealed an elevated total leukocyte count with a neutrophilic predominance and an elevated liver function test. A contrast enhanced CT was ordered that revealed two duodenal diverticula causing mass effect on the distal CBD and the pancreatic duct. Subsequently, the patient was managed conservatively that led to a significant decline in the signs and symptoms.

Keywords: Lemmel Syndrome; Obstructive Jaundice; Duodenal Diverticulum; Right Hypochondrial Pain

Introduction

There exists a plethora of causes when it comes to right hypochondrial pain along with obstructive jaundice out of which choledocholithiasis is the most common one [1]. A rare cause of obstructive jaundice with right upper quadrant abdominal pain in the absence of other pathology is Lemmel syndrome [2]. It was first described by Lemmel in 1934. Lemmel syndrome is characterized by obstructive jaundice secondary to diverticula arising from the periampullary duodenum in the absence of choledocholithiasis or neoplasm [3]. The term periampullary diverticulum is used when the diverticulum is located within 2-3 cm of the ampulla of Vater [4]. They are mostly located in the second part of the duodenum. After taking informed consent from the patient, we report a case of Lemmel syndrome that was successfully managed with conservative measures.

Clinical Case

A 55-year-old male presented to our hospital with complaints of right upper quadrant abdominal pain, nausea, and loss of appetite for a duration of 1 month. He is a known case of diabetes mellitus with a past history of mesh hernioplasty 3 years back. He also gave history of significant weight loss along with complaints of fatigue and weakness. The pain was insidious on onset, colicky in nature, non-radiating and intermittent with no aggravating or relieving factors. There is no change in bowel and bladder habits as well. The patient denied any fever, chills, melena, hematemesis, hematochezia and altered mental status. His general physical examination revealed a positive Murphy sign but was negative for icterus. His routine laboratory tests like complete blood count, inflammatory markers, liver function tests, renal function tests,

serum electrolytes and urine routine examination were ordered. The results demonstrated an elevated white blood count with a neutrophilic predominance, direct hyperbilirubinemia with a total bilirubin of 3 mg/dL and a rise in ALP to 240 U/L.

An ultrasound of the abdomen revealed a distended gall bladder with normal walls and no evidence of gall stones. The liver was found to be normal in size and echo pattern. A contrast enhanced computed tomography of the abdomen was done that revealed two duodenal diverticula causing moderate mass effect on the distal CBD and pancreatic duct resulting in dilatation of the common bile duct to 10.1 mm (Figure 1). The pancreatic duct diameter at the site of obstruction is 3 mm (Figure 2).

Figure 1: A large duodenal diverticulum in the pancreatico-duodenal groove of dimensions 73 mm X 34 mm. Maximum calibre of the common bile duct is 10.1 mm.

Figure 2: Both the duodenal diverticula cause mass effect on the pancreatic duct (marked with a white arrow) that leads to a diameter of 3 mm.

A small 17.4 x 13.2 mm duodenal diverticulum was noted, most likely arising from the anterior and superior wall of the second part of the duodenum located anterior to the duodenal ampulla (Figure 3). Another large diverticulum measuring 73.6 x 34.9 mm was noted in the region of pancreatico-duodenal groove arising from the 3rd part of duodenum with multiple food particles within (Figure 4). The large duodenal diverticulum was also displacing the uncinate process and the head of pancreas superiorly. Both the duodenal diverticula have mild wall thickening and mucosal enhancement. There is no evidence of gall stones or focal strictures in the biliary tree.

Figure 3: A small duodenal diverticulum measuring 17 x 13 mm from the superior and anterior wall of the 2nd part of duodenum.

Figure 4: The large periampullary duodenal diverticulum (marked in white arrow) with features of mucosal enhancement and wall thickening in the coronal section of the CT scan of the abdomen and pelvis.

In addition, the stomach and the proximal duodenum were normal with no air-fluid levels (Figure 5) and hence, no proximal obstruction. The other abdominal organs were normal. The liver was normal in size with uniform density. No focal lesion was seen, and no abnormal enhancement was noted. An incidental finding of a cortical cyst was noted on the upper pole of the right kidney.

Figure 5: No air fluid levels seen in the stomach and the duodenum. Thicker arrow indicates stomach and thinner arrow indicates duodenum.

We decided to keep the patient on conservative management and decided that no surgical intervention was warranted. The patient was subsequently made nil per oral (NPO), and an NG tube (Ryle's tube) was placed to ensure bowel rest. He was then started on a 7-day course of intravenous antibiotics like metronidazole and ciprofloxacin.

After the antibiotic course was over, there was complete resolution of symptoms and follow up was done with a CT scan after 21 days that revealed a significant reduction in inflammation and the patient was tolerating oral feeds well. Follow up laboratory values like the total leukocyte count and inflammatory markers also came back to normal level. His discharge was uneventful.

Discussion

A diverticulum is a mucosal outpouching and when arising from the intestinal wall, it is known as a gastrointestinal diverticulum [5]. It Is mainly devoid of the muscle layer [6]. The diverticulum compresses the distal common bile duct causing cholestatic jaundice [7]. It is mainly classified based on their location into three different types based on the position of the major papilla in relation to the diverticulum [4]. Most of them are asymptomatic but can cause pancreaticobiliary complications if inflamed [8]. A mucosal outpouching that arises 2-3 cm from the ampulla of Vater is termed as a peri-ampullary diverticulum [4]. The pancreatico-biliary disease characterized by common bile duct obstruction secondary to a periampullary diverticulum is the main pathological finding in Lemmel syndrome [11]. Clinically, the patient complains of right upper quadrant pain and the laboratory findings include elevated leukocyte count and liver function tests [10].

The periampullary diverticulum does not cause any symptoms in only 10% cases. Some complications pertaining to the diverticulum secondary to inflammation are diverticulitis, perforation, and hemorrhage [9,12-14]. The diverticulum is a continuation of the duodenal lumen and acts as a nesting pouch as in our case. This nesting pouch contains food particles acting as a reservoir for bacterial growth thereby, leading to diverticulitis [15].

Lemmel syndrome also can lead to complications involving the pancreas and the biliary tree like acute pancreatitis, cholangitis and recurrent gall bladder or bile duct stones [16]. There are multiple etiologies that lead to the development of Lemmel syndrome. Firstly, the periampullary diverticulum can lead to papillary fibrosis due to constant mechanical irritation. Secondly, sphincter of Oddi dysfunction can occur due to the periampullary diverticulum. Lastly, there could be mechanical obstruction of the distal common bile duct due to the duodenal diverticulum as seen in our case [17].

Imaging is extremely critical to diagnose Lemmel syndrome. Once an abdominal ultrasound is negative for stones in the biliary tree, Lemmel syndrome should be the amongst the differential diagnoses. The next best step should be to do a contrast enhanced abdominal computed tomography must be done. The oral contrast with a multiplanar visualization shows the contrast material entering the diverticulum. Specifically, the venous phase should be given a lot of importance.

Normally, a 55-year-old male should have a common bile duct with a normal calibre of 5.5 mm. However, in our case, the

maximum calibre of CBD proximal to obstruction was 10.1 mm while the calibre of CBD at the level of obstruction was 4.07 mm. The calibre of the pancreatic duct is 3 mm (Figure 2) with minimal prominence of the central intrahepatic biliary radicles. In addition to the two duodenal diverticula, multiple small diverticula were also noted along the splenic flexure and proximal descending colon that suggests that the patient is prone to develop diverticula.

Based on the literature review, an ERCP with a side viewing endoscope is gold standard for diagnosing Lemmel syndrome. In addition, an endoscopic ultrasound has also proven to show similar results [17]. ERCP also has the added advantage of allowing an endoscopic sphincterotomy, a bile duct stent implantation or endoscopic extraction of the entrapped material [18]. Using imaging studies like CT and MRCP, the periampullary diverticulum can be visualized as a thin walled cavitary lesion on the medial wall of the second part of the duodenum [19].

Based on the clinical presentation, pathological findings and pathophysiology, Lemmel syndrome can be treated with different methods. Endoscopic approach has had a high success rate based on the literature review [20]. When the pathology found in Lemmel syndrome is due to chronic fibrosis of the papilla or sphincter of Oddi dysfunction, then the best treatment modality would be ERCP with sphincterotomy [21]. In cases of endoscopic failure, surgical interventions like simple diverticulectomy, diverticular inversion or transduodenal sphincteroplasty have been described. All these interventions are associated with a high level of morbidity and mortality. It is crucial to individualize the treatment plan according to the characteristics of the patient, location of the diverticulum and experience of the medical team.

Several cases of Lemmel syndrome have been reported. Desai K et al reported the case of a 25-year-old female presenting with unintentional weight loss and fatigue was found to have a large periampullary diverticulum that was then treated conservatively [5]. In our case, the patient recovered within the next 3 weeks with conservative management. Due to the resolution of symptoms and normalization of laboratory values, surgical intervention was not considered.

Conclusion

In a patient presenting with right hypochondriacal pain, icterus, direct hyperbilirubinemia with no evidence of gall stones, it is important to include Lemmel syndrome as part of the differential diagnosis especially when the patient has diverticula arising from other parts of the bowel as well. Considering Lemmel Syndrome in a setting of such a clinical picture would reduce the delay in diagnosis of the patient and definitive management of the patient.

Conflict of Interest

There is no financial interest or conflict of interest that exists.

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