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Lemmel's Syndrome: A Rare Cause for Recurrent Cholangitis and Pancreatitis

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Abstract: A 65 year old female presented with the history of obstructive jaundice, recurrent cholangitis and pancreatitis. She was managed conservatively for 2 years in a local hospital. The patient was then referred to our centre. On evaluation, ultrasonography of abdomen showed dilated common bile duct. Contrast enhanced computed tomography of abdomen showed dilated common bile duct and duodenal diverticulum. Magnetic resonance cholangio pancreatography revealed a diverticulum in the second part of duodenum compressing the distal common bile duct. Subsequently we performed a side viewing duodenoscopy, which revealed diverticulum at second part of duodenum and diagnosed as Lemmel's syndrome. Endoscopic biliary sphincterotomy and biliary stenting was performed. Later she improved well.
Keywords: Lemmel's syndrome, peri ampullary diverticulum, obstructive jaundice, recurrent cholangitis, recurrent pancreatitis, endotherapy.

INTRODUCTION:

Way back in 1710 Chomel first described periampullary diverticulum (PAD) [1]. Lemmel syndrome is an unusual cause of obstructive jaundice, recurrent cholangitis and pancreatits due to PAD in the absence of bile duct stones [2]. Duodenal diverticula are extra luminal outpouching of the duodenal mucosa. A duodenal diverticula arising adjacent to ampulla of Vater is termed as periampullary diverticula (PAD). The prevalence varies from 0.16% to 22% [3]. Most cases of PAD are usually asymptomatic. But it could result in obstructive jaundice, cholangitis, pancreatitis, hemorrhage, perforation, fistula and enterolith formation. The reason for obstruction is that PAD compresses the intrapancreatic part of the common bile duct (CBD) which in turn causes the dilatation of both extra- and intrahepatic bile ducts. Here we report a case of Lemmel's syndrome who was diagnosed two years after onset of symptoms.

CASE REPORT:

A 65 year old female from a sub-urban place was referred with a history of recurrent biliary colic for last 2 years. Earlier the patient was hospitalised twice for upper abdominal pain, fever with chills and jaundice and managed conservatively in a local hospital with intravenous fluids, analgesics and parenteral antibiotics. Patient was referred to our centre for further evaluation and management. On examination, patient was febrile, icteric and tenderness in the right upper quadrant of abdomen was present. Other systemic examinations were unremarkable. Laboratory investigation revealed a total count of 13,000 cells/uL with elevated liver function tests [total bilirubin 4.6 mg/dL, direct bilirubin 2.7 mg/ dL, SGOT 110 IU/L, SGPT 160 IU/L, alkaline phosphatase 220 IU/L]. Serum amylase and lipase were 404 and 568 IU/L respectively. Ultrasonography of abdomen (USG) showed dilated CBD. Multi slice contrast enhanced computed tomography (CECT) of abdomen showed dilated CBD and common hepatic duct (CHD) (Figure 1). Magnetic resonance cholangio pancreatography (MRCP) revealed a diverticulum in the second part of duodenum compressing the distal CBD with dilatation of the proximal CBD and no evidence of cholelithiasis or choledocholithiasis (Figure 2). We performed side viewing duodenoscopy, which showed diverticulum at the second part of duodenum. Subsequently, endoscopic biliary sphincterotomy and biliary stenting was performed to relieve the obstruction (Figure 3). Post procedure, fluoroscopic screening showed double pigtail catheter in situ (Figure 4). Biliary symptoms and pancreatitis resolved following the biliary stenting. During follow-up visits, the patient was found to be asymptomatic.

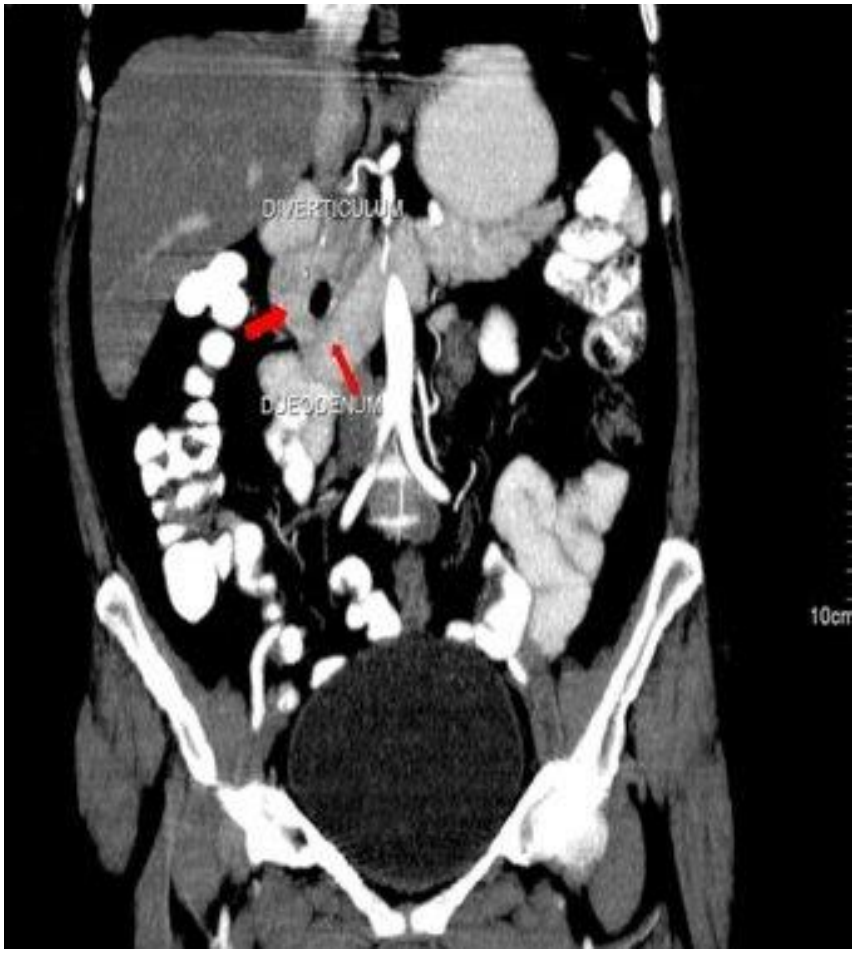

Fig 1: Contrast enhancing computed tomography of abdomen shows the duodenal diverticulum of size 2x3cm which is compressing the distal common bile duct.

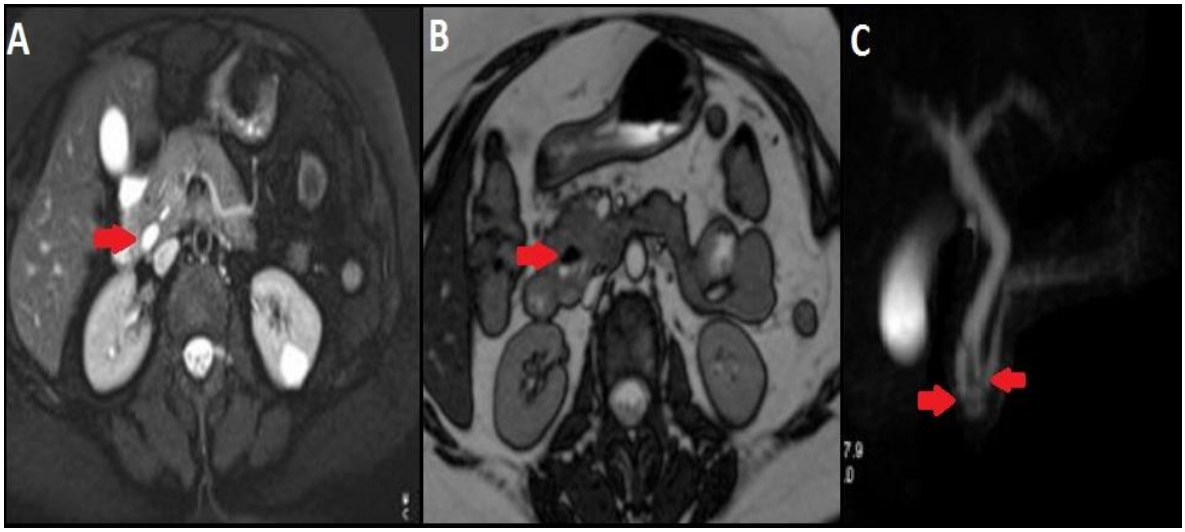

Fig 2: A) Magnetic resonance imaging of abdomen, T2 axial image shows dilatation of CBD B) Out of phase axial image showing the duodenal diverticulum anterior to CBD with air fluid level. C) MRCP shows close relationship of duodenal diverticulum to CBD.

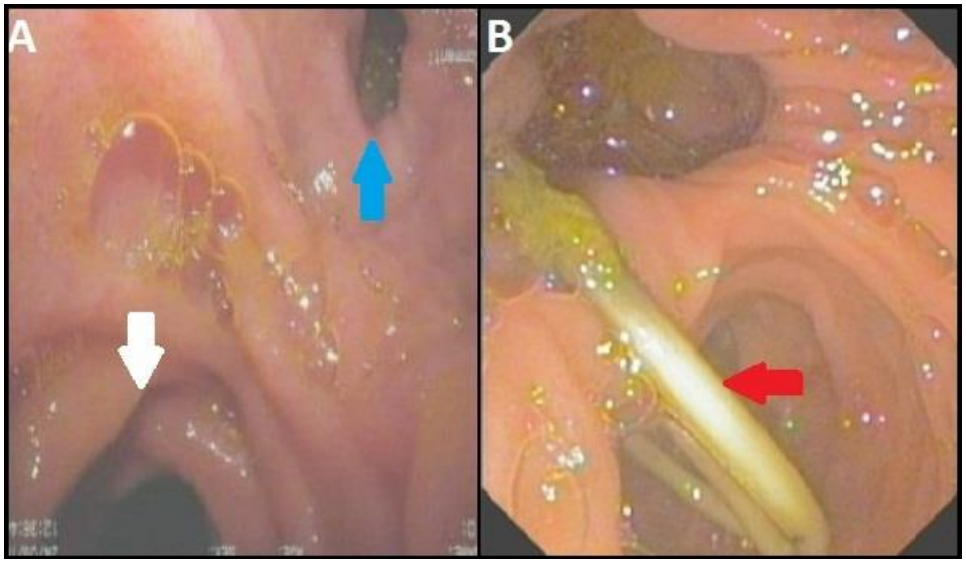

Fig 3: A) Side viewing duodenoscopy shows the diverticulum at second part of duodenum (blue arrow) and white arrow indicates the lumen of the duodenum. B) 7FrX8cm biliary double pigtail catheter seen in lumen with bile flow (red arrow).

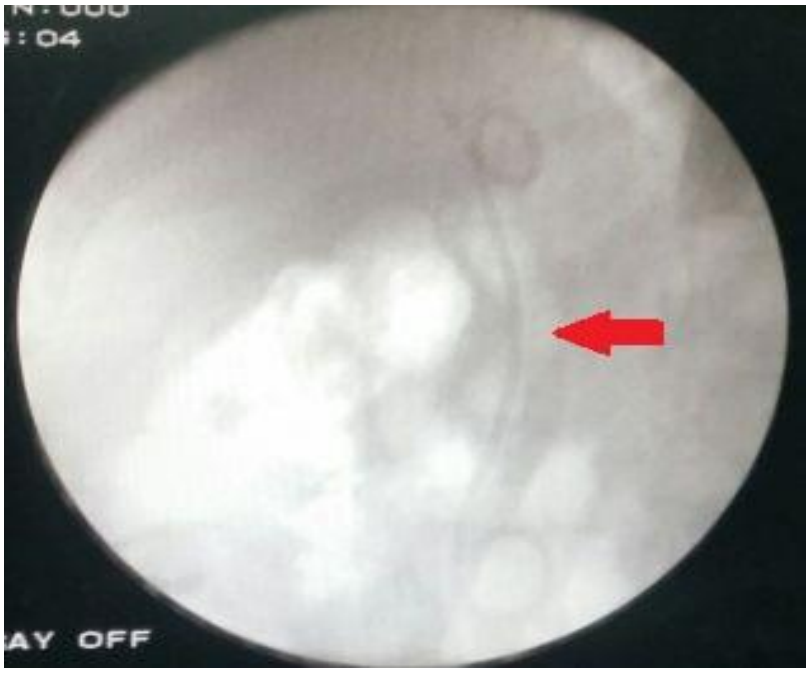

Fig 4: Fluoroscopic image shows the double pigtail catheter in situ (arrow).

DISCUSSION:

Pathogenesis of Lemmel's syndrome includes the following a). PAD which may be filled with bezoar or enterolith that can cause direct compression of distal CBD or ampulla [4, 5] b). Sequele of diverticulitis result in chronic fibrosis of papilla (papillitis chronic fibrosa) [6] c). PAD can also cause sphincter of Oddi dysfunction [7]. All these changes result in obstructive jaundice, recurrent cholangitis and pancreatitis in the abscence of bile duct stones. The important life threatening complications associated with this syndrome are bleeding or perforation of the diverticulum. MRCP plays an important role as a non-invasive tool to diagnose Lemmel's syndrome [8]. The side viewing duodenoscopy is a minimally invasive procedure used for both identifying PAD and performing therapeutic procedures like sphincterotomy and biliary stenting [9]. On the other hand, the available various surgical options for Lemmel's syndrome are CBD exploration, bilio enteric anastomosis and diverticulectomy [10]. Rarely major surgery like Whipple's procedure may be required in case of perforation of PAD [11]. Hence management of this syndrome is mainly depends on the clinical presentation. Our case presented with obstructive jaundice, recurrent cholangitis and pancreatitis. There was no evidence of bezoar or enterolith. She responded well to endoscopic biliary sphincterotomy and biliary

stenting. Two years after onset of symptoms only, the diagnosis of Lemmel's syndrome was established by doing imaging and side viewing duodenoscopy in our patient. So we emphasis Lemmel's syndrome should be kept as a differential diagnosis in any patient who is undergoing initial diagnostic work up for hepato biliary pancreatic symptoms.

CONCLUSION:

In patients with hepato biliary pancreatic symptoms, high index of suspicion is needed to diagnose Lemmel's syndrome and appropriate management should be instituted to prevent fatal complications.

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AUTHOR CONTRIBUTIONS:

Durairaj Segamalai wrote the manuscript and is the article guarantor. All the other authors revised the manuscript.

FINANCIAL DISCLOSURE: None to report.

ETHICS

Informed consent was obtained for this case report.

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