

## Lemmel's Plus Syndrome: An Unusual Cause of Obstructive Jaundice

V.R Rajendran<sup>1</sup>, A M Abdul Latheef<sup>2\*</sup>, Noufal Perumpalath<sup>3</sup>, Ipsit I Menon<sup>4</sup>, Sherinas Rehiman<sup>4</sup>

<sup>1</sup>Professor and Head, <sup>2</sup>Junior Resident, <sup>3</sup>Associate Professor, <sup>4</sup>Radiologist,  
Department of Radiodiagnosis, Government Medical College, Kozhikode, Kerala, India.

### ABSTRACT

Periampullary diverticulum refers to extraluminal outpouching of duodenal mucosa that develops within a radius of 2cm from the ampulla of Vater. Usually asymptomatic; but can be sometimes associated with pancreaticobiliary and non pancreaticobiliary complications. Lemmel's syndrome is a rare entity, refers to obstructive jaundice developing secondary to periampullary diverticulum in the absence of choledocholithiasis and tumour in the head of pancreas. Lemmel's syndrome consists of classic triad obstructive jaundice, cholangitis and pancreatitis. In addition to classic triad our case revealed pneumobilia that has never been reported in literature, here by we coin a new terminology called lemme's plus syndrome that has 4 components: obstructive jaundice, cholangitis, pancreatitis and pneumobilia that is two pathology exists in lemme's plus syndrome element of obstruction and element of perforation.

**Key Words:** Pneumobilia, Lemmel's Plus Syndrome, Ampulla of Vater, Periampullary Diverticulum.

### \*Correspondence to:

**Dr. A M Abdul Latheef,**  
Al Hira, Mannam P.O  
North Paravoor,  
Ernakulam, Kerala, India.

### Article History:

Received: 29-01-2017, Revised: 16-02-2017, Accepted: 05-03-2017

### Access this article online

Website: <a href="http://www.ijmrp.com">www.ijmrp.com</a>	Quick Response code 
DOI: 10.21276/ijmrp.2017.3.2.057	

### INTRODUCTION

62 year old male patient presented with history of yellowish discoloration of eyes and urine for 4 months, recurrent episodes of fever and abdominal pain. Investigations results are as follows; total bilirubin-4.3mg/dl, direct bilirubin-4.1mg/dl, aspartate aminotransferase-112 IU/L, alanine aminotransferase – 262 IU/L, gamma glutamyl transpeptidase – 634 IU/L, alkaline phosphatase -430 IU/L, Total white blood cell count was increased -14400, with a neutrophil predominance of 85%, CA19.9-117.48, Amylase – 790U/L, lipase -1300U/L.

Barium meal follow through revealed multiple diverticuli involving second and third parts of duodenum with barium noted filling the CBD and intrahepatic biliary radicles and the diverticulum in the concave medial aspect of second part of duodenum was noted in relation with the retroduodenal part of CBD. Ultrasonogram revealed dilated CBD and intrahepatic biliary radicle dilatation and pneumobilia. No e/o calculus in gall bladder as well as in the biliary tree. No evidence of mass lesion in head of pancreas. Contrast enhanced CT section of abdomen revealed dilated intrahepatic biliary tree and CBD with CBD measuring 11 mm and pneumobilia. No e/o calculus in CBD and gall bladder. No e/o mass lesion on head of pancreas. Pancreatic duct was normal and measuring 3.4 mm. MRCP demonstrated periampullary diverticulum measuring 2.4 cm diameter and compressing the

terminal CBD causing upstream biliary dilatation and pneumobilia. The CBD distal to diverticulum appears collapsed. Upper gastrointestinal endoscopy confirmed the periampullary diverticulum with ampullary opening displaced due to diverticuli.

### DISCUSSION

Periampullary diverticulum refers to extraluminal outpouching of duodenal mucosa that develops within a radius of 2cm from the ampulla of Vater.<sup>1</sup> Lemmel's syndrome is a rare entity, refers to obstructive jaundice developing secondary to periampullary diverticulum in the absence of choledocholithiasis and tumour in the head of pancreas.<sup>2</sup>

Duodenum is second most common site for diverticulum after colon.<sup>3</sup> Most common site for diverticulum in duodenum is concave medial aspect of second part of duodenum. Incidence of duodenal diverticulum varies based on study; it is 1-6 % in upper GI contrast studies and 12-27% in endoscopy studies. Incidence increases with age. 90% are solitary. 70% occurs in second part of duodenum.<sup>4,5</sup>

Juxta papillary duodenal diverticula are classified into 3 types: I – Most common type. Major papillae located within the diverticulum. II – Papillae located within the margin of diverticulum. III – Papillae near the diverticulum.

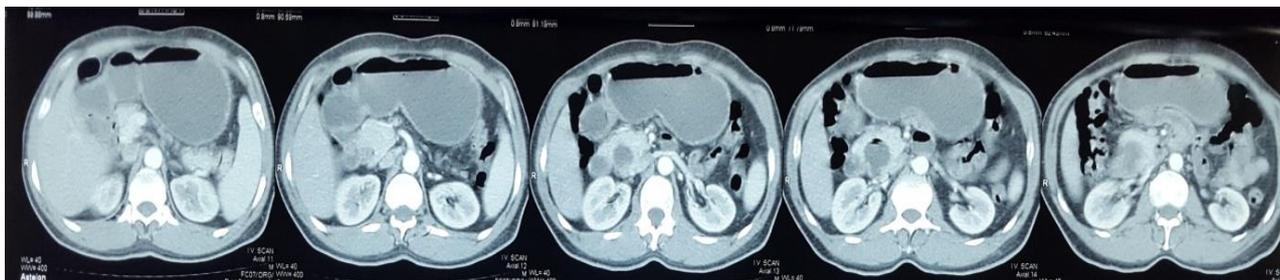


Figure 1: Axial post contrast section of upper abdomen showing pneumobilia and air densities in the gall bladder.

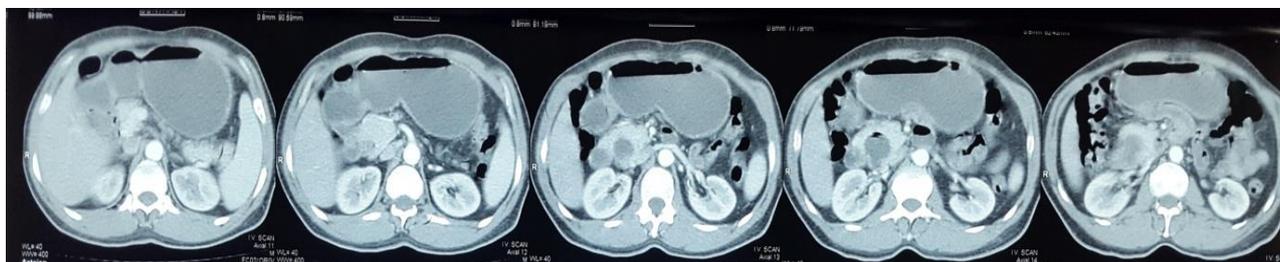


Figure 2: Axial post contrast section of upper abdomen showing peripapillary diverticulum with air fluid level, no evidence of mass lesion head of pancreas.

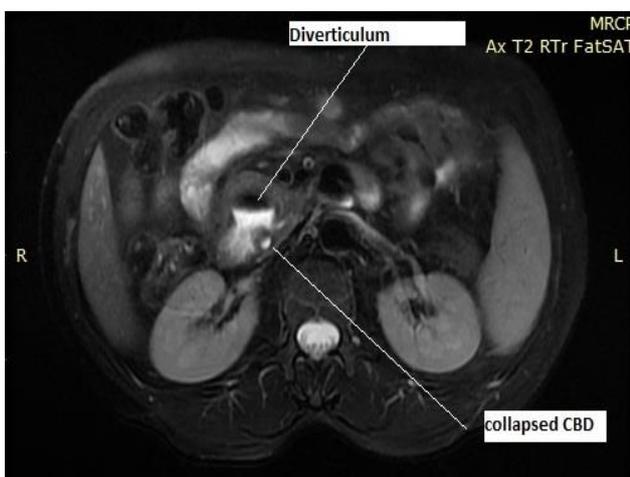


Figure 3: Axial T2 weighted imaging showing the peripapillary diverticulum in the concave medial aspect of D2 with collapsed CBD distal to diverticulum seen in the inferior aspect.

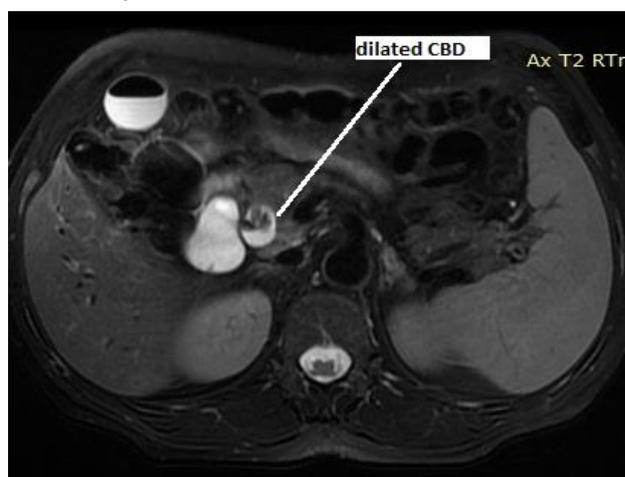


Figure 4: Axial T2 weighted images showing the dilated supra-pancreatic CBD above the level of diverticulum.



Figure 5: Barium meal follow through revealed shows multiple diverticuli involving second and third part of duodenum with barium noted filling the CBD and intrahepatic biliary radicles and the diverticulum in the concave medial aspect of second part of duodenum was noted in relation with the retroduodenal part of CBD.

Majority of peripapillary diverticulum are asymptomatic, however biliopancreatic complications can occur such as recurrent biliary calculi, obstructive jaundice, cholangitis, pancreatitis.

Pathologic mechanism through which Lemmel's syndrome is known to occur include:

1<sup>st</sup> – Diverticulitis or direct mechanical irritation of peripapillary diverticulum causing chronic inflammation of ampulla and leading to chronic fibrosis papillae.<sup>6</sup>

2<sup>nd</sup> – Dysfunction of sphincter of oddi.<sup>7</sup>

3<sup>rd</sup> – Distal CBD/ampulla directly compressed mechanically by peripapillary diverticulum that is filled with enterolith or bezoar.<sup>8,9</sup>

In our case the cause of obstruction by the diverticulum is direct mechanical compression as MRCP is revealing dilation of CBD proximal to diverticulum whereas CBD distal to diverticulum appears collapsed.

In addition to classic triad of Lemmel's syndrome – cholangitis, pancreatitis and obstructive jaundice; imaging in our case revealed significant pneumobilia possible cause could be recurrent diverticulitis causing thinning of CBD and perforation of diverticulum into CBD.

## CONCLUSION

Although periampullary diverticulum appears to be rare cause for obstructive jaundice, it should be considered in the differentials in case of absence of choledocholithiasis and lesion in head of pancreas. It can also be mistaken for cystic neoplasm of pancreas, pancreatic pseudocyst. In a patient with obstructive jaundice One can consider a diagnosis of Lemmel's plus syndrome if there is element of both obstruction and perforation in the biliary tree secondary to a periampullary diverticulum.

## REFERENCES

1. Lobo DN, Balfour TW, Iftikhar SY, Rowlands BJ. Periampullary diverticula and pancreaticobiliary disease. *Br J Surg*. 1999 May; 86(5):588-97.
2. Lemmel G. Die Klinische Bedeutung der Duodenal Divertikel. *Arch Verdauungskrht* 1934; 46: 59-70.
3. Chomel JB. Report of a case of duodenal diverticulum containing gallstones. *Histoire Acad R Sci Paris* 1710: 48-50.
4. Zoepf T, Zoepf DS, Arnold JC, Benz C, Riemann JF. The relationship between juxtapapillary duodenal diverticula and disorders of the biliopancreatic system: analysis of 350 patients. *Gastrointest Endosc* 2001;54:56-61.
5. Wu SD, Su Y, Fan Y, Zhang ZH, Wang HL, Kong J, Tian Y. Relationship between intraduodenal peri-ampullary diverticulum and biliary disease in 178 patients undergoing ERCP. *Hepatobiliary Pancreat Dis Int* 2007;6:299-302.
6. Manabe T, Yu GS. Duodenal diverticulum causing intermittent-persistent cholestasis: associated with papillitis chronica fibrosa. *N Y State J Med* 1977; 77: 2132-6.
7. Tomita R, Tanjoh K. Endoscopic manometry of the sphincter of Oddi in patients with Lemmel's syndrome. *Surg Today* 1998; 28: 258-61.
8. Rouet J, Gaujoux S, Ronot M, Palazzo M, Cauchy F, Vilgrain V, Belghiti J, O'Toole D, Sauvanet A. Lemmel's syndrome as a rare cause of obstructive jaundice. *Clin Res Hepatol Gastroenterol* 2012; 36: 628-31.
9. Nishida K, Kato M, Higashijima M, Takagi K, Akashi R. A case of Lemmel's syndrome caused by a large diverticular enterolith at the peripapillary portion of the duodenum. *Nihon Ronen Igakkai Zasshi* 1995; 32: 825-9.

**Source of Support:** Nil.

**Conflict of Interest:** None Declared.

**Copyright:** © the author(s) and publisher. IJMRP is an official publication of Ibn Sina Academy of Medieval Medicine & Sciences, registered in 2001 under Indian Trusts Act, 1882.

This is an open access article distributed under the terms of the Creative Commons Attribution Non-commercial License, which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited.

**Cite this article as:** V.R Rajendran, A M Abdul Latheef, Noufal Perumpalath, Ipsit I Menon, Sherinas Rehiman. Lemmel's Plus Syndrome: An Unusual Cause of Obstructive Jaundice. *Int J Med Res Prof*. 2017; 3(2):273-75. DOI:10.21276/ijmrp.2017.3.2.057