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LEMMEL SYNDROME – RARE CAUSE OF CHOLANGITIS : A CASE REPORT

Medical Gastroenterology

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ABSTRACT

Introduction: Lemmel Syndrome is a rare cause of the biliary mechanical compression by the juxtapapillary pseudo-diverticulum, in the absence of gallstones or periampullary tumors. Most periampullary diverticula are asymptomatic. However, complications may occur in about 5%, including diverticulitis, pancreatitis, cholangitis, choledocholithiasis, enterolith, bezoar formation, intestinal obstruction, bleeding and perforation.

Case report: A 60-year-old woman presented with abdominal pain,fever and jaundice of acute onset.
She was hemodynamically stable except for a fever of 101 °F. Initial blood work revealed a WBC 14,500/mm3, ALT 118 IU/L, AST 189 IU/L, ALP 356 IU/L, T bilirubin 7.2 mg/dl. CT and MRI of abdomen revealed periampullary diverticulum compressing distal common bile duct causing proximal dilatation consistent with Lemmel syndrome. Patient was managed with broad spectrum antibiotics and other supportive measures in view of cholangitis. Patient underwent ERCP with CBD stenting. Patient's lab parameters improved steadily and was asymptomatic in few days.

Discussion: Our patient had abdominal pain associated with leukocytosis and cholangitis due to extrinsic compression of the distal CBD. Lemmel syndrome can be transient in nature as a result of a periampullary duodenal diverticulum causing extrinsic compression of the CBD. This can sometimes progress to cholangitis, as seen in our patient. Early diagnosis is necessary to avoid extensive and invasive workup. Management is often supportive, although in some cases with recurrent infection or persistent biliary obstruction, surgical diverticulectomy can be considered.

Conclusion: Lemmel syndrome should be kept as a rare differential when evaluating patients with biliary obstruction.

KEYWORDS

lemmel, diverticulum, cholangitis

INTRODUCTION

Lemmel's syndrome, also known as duodenal diverticulum obstructive jaundice syndrome was first described in 1934 by Lemmel, characterized by obstructive jaundice due to periampullary diverticula, in the absence of cholelithiasis or other detectable obstacle. Very few cases of Lemmel's syndrome have been published and fully investigated (1-3).

Periampullary diverticula (PAD) refer to extraluminal outpouchings of duodenal mucosa that develop within the radius of 2 to 3 cm from the ampulla of Vater (4). PAD are largely asymptomatic but they sometimes can cause both pancreaticobiliary and non-pancreaticobiliary complications. Rarely, obstructive jaundice can develop secondary to PAD in the absence of choledocholithiasis or tumor and is termed Lemmel's syndrome (5).

In the majority of cases, diverticula arise onthe inner or pancreatic border of the duodenum.The possibly of PAD should be kept in mind while interpreting any bile duct imaging.It can create a filling defect in biliary passage; hence, can be mistaken for periampullary tumors or biliary stones.

It can also be misinterpreted as pancreatic pseudocyst when it is large and fluid filled (6).

Herein, we report a case of cholangitis due to Lemmel's syndrome that was successfully managed endoscopically.

CASE ILLUSTRATION

A 67 years old previously healthy female with no significant past medical or surgical history presented to the emergency department with chief complaint of right upper quadrant abdominal pain,fever and jaundice of acute onset.The patient also complained of nausea and decrease in appetite. The patient denied any vomiting, melena, hematochezia, hematemesis or weight loss.No history of malignancy and jaundice in the family.

She was hemodynamically stable except for a temperature of 101 °F. On physical examination, she was icteric and there was tenderness in the right hypochondrium with positive murphys sign. Initial blood work revealed a WBC 14,500/mm3, ALT 118 IU/L, AST 189 IU/L, ALP 356 IU/L, T bilirubin 7.2 mg/dl and non reactive for HbsAg, anti HCV and IgM anti HAV. CECT abdomen revealed diverticulum arising from medial wall of second part of duodenum measuring 2.2x 2.1 cm compressing distal common bile duct causing proximal dilatation of 11 mm ,consistent with Lemmel syndrome. CECT abdomen findings were confirmed by MRCP. Patient was managed with broad spectrum antibiotics and other supportive measures in view of cholangitis. Patient underwent ERCP with CBD stenting. Patient's lab parameters improved steadily and was asymptomatic in few days.

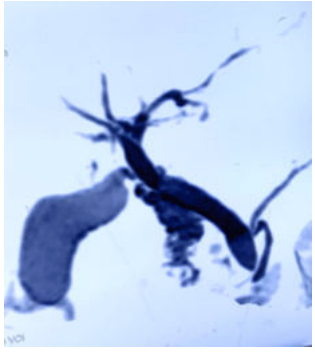

Image 1: MRCP images consistent with Lemmel syndrome

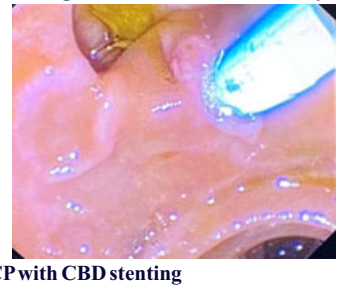

Image 2:ERCP with CBD stenting

DISCUSSION

Lemmel's syndrome is characterized by obstructive jaundice due to PAD, in the absence of cholelithiasis or other detectable obstacle. (1-3) Most patients with Lemmel's syndrome present with jaundice, abdominal pain or acute cholangitis, mimicking periampulary tumors(7,8).

Primary or true duodenal diverticula represent mucosal out-pouchings devoid of muscle layer. Duodenal diverticula are classified based on their location. Among the different types of duodenal diverticula, periampullary duodenal diverticula are the most common. The majority of periampullary diverticula are asymptomatic; however, biliopancreatic complications such as recurrentbiliary calculi, obstructive jaundice, cholangitis, acute or chronicpancreatitis can

result from mechanical compression by a large,distended due to poorly emptying diverticulum or due to motilitydysfunction of the sphincter of Oddi, reflux of intestinal contentinto the ducts and bacterial overgrowth (9). Complications relatedto inflammation such as diverticulitis, hemorrhage, perforation orfistula formation may also occur (5).

Diverticula of the gastrointestinal tract are outpouchings of all or part of the intestinal wall whichcan occur anywhere throughout the gastrointestinaltract. The duodenum is second most common site of diverticula in the gastrointestinal tract after colon, followed by jejunum, ileum and stomach. Duodenal diverticula was first reported byPierre Jean Baptiste and first welldocumented report was made by Morgagni in 1762 andit was regarded an anatomic curiosity until 1913 whenradiological demonstration was done by JT Case, whodisplayed roentgenograms of 4 cases (10,11)

It is difficult to ascertain the true prevalence of duodenal diverticula; they are seen in 1-6% of upper gastrointestinal radiologic exam, 12-27% of endoscopic studies and in 15-22% of autopsies (8,12).Duodenal diverticula are rare below age 40 years and has a tendency to increase with age (6,12).

They can be classified as congenital or acquired and intraluminal or extraluminal. They typically occur in the periampullary region, along the medial aspect of the second and third part of the duodenum (13,14). Incidence of Lemmel's syndrome seems higher in patients with intradiverticular papilla than in patients with a juxtapapillary diverticulum, possibly because of their larger size and closer relation to ampulla (8).

Pathologic mechanisms through which Lemmel's syndrome is thought to occur include the following. First, diverticulitis or direct mechanical irritation of PAD cause chronic inflammation of the ampulla and lead to chronic fibrosis of papilla (papillitischronica fibrosa). Second PAD may cause dysfunction of the sphincter of oddi. Third food debris flowing into the diverticulum cause distal CBD or ampulla directly compressed mechanically by PAD (1-3,15,16) Diagnosing Lemmel's syndrome could be challenging, but being aware of this condition is important to avoid mismanagement and it begins with identification of PAD. PAD are best demonstrated using a side-viewing endoscope during ERCP. On CT scan or MRCP, PAD appear as thin-walled cavitary lesions situated on the medial wall of the duodenum second part that typically contain gas. However, PAD are sometimes filled with fluid and frequently be misinterpreted with pancreatic pseudocyst, pancreatic abscess, cystic neoplasm in the pancreas head or even metastatic lymph node (17-19). Therefore, high index ofsuspicion is necessary to establish right diagnosis insuch cases.

The therapeutic options for Lemmel syndrome are surgical resection, endoscopic intervention and conservative treatment (3). The most simple treatment of Lemmel's syndrome is endoscopic sphincterotomy to release CBD obstruction (8). There is now consensus that elective surgical treatment of asymptomatic or minimally symptomatic diverticulum is not justified. Surgical procedures for diverticula in the second part of duodenum are often considered difficult since it requires mobilizationof the duodenum which is retroperitoneal. Surgical or endoscopic interventions should only be reserved for symptomatic

diverticulum. Diverticulectomy for abdominal discomfort and indefinite pain is dangerous and unrewarding; it carries a high morbidity and mortality. Only 50% of patients treated with diverticulectomy were relieved of their symptoms (6).

CONCLUSION

In summary, Lemmel syndrome is a rare cause of obstructivejaundice and should be included in the differential diagnosis of biliary obstruction when a periampullary duodenal diverticulumis demonstrated. It is important to have a high index of suspicion to establish an accurate diagnosis, as Lemmel syndrome can mimic several benign and malignant abnormalities in the periampullary region.

REFERENCES:

[1]. Kang, H. S., Hyun, J. J., Kim, S. Y., Jung, S. W., Koo, J. S., Yim, H. J., & Lee, S. W. (2014). Lemmel's syndrome, an unusual cause of abdominal pain and jaundice by impacted intradiverticularenterolith: case report. *Journal of Korean medical science*, *29*(6), 874–878. https://doi.org/10.3346/jkms.2014.29.6.874

[2]. Rouet, J., Gaujoux, S., Ronot, M., Palazzo, M., Cauchy, F., Vilgrain, V., Belghiti, J., O'Toole, D., &Sauvanet, A. (2012). Lemmel's syndrome as a rare cause of obstructive jaundice. *Clinics and research in hepatology and gastroenterology*, *36*(6), 628–631.https://doi.org/10.1016/j.clinre.2012.05.002.

[3]. Takakura, K., Koido, S., Kajihara, M., Fukue, R., Takami, S., Ito, Z., Kobayashi, H., Matsumoto, Y., Tsukinaga, S., Odahara, S., Yukawa, T., Uchiyama, K., Misawa, T., Ohkusa, T., &Tajiri, H. (2014). Repetitive acute cholangitis and pancreatitis due to Lemmel syndrome: A case report.

[4]. Lobo, D. N., Balfour, T. W., Iftikhar, S. Y., & Rowlands, B.J.(1999). Periampullary diverticula and pancreaticobiliarydisease. *The British journal of surgery*, *86*(5), 588–597. https://doi.org/10.1046/j.1365-2168.1999.01121.x

[5]. Gerhard Lemmel. Die klinischeBedeutung der Duodenaldivertikel. *Digestion.* Vol. 56(1-2):59-70. DOI: 10.1159/000196978

[6]. Rizwan, M. M., Singh, H., Chandar, V., Zulfiqar, M.,& Singh, V. (2011). Duodenal diverticulum and associated pancreatitis: case report with brief review of literature. *World journal of gastrointestinal endoscopy*, *3*(3), 62–63. https://doi.org/10.4253/ wjge.v3.i3.62

[7]. Yoneyama, F., Miyata, K., Ohta, H., Takeuchi, E., Yamada, T., & Kobayashi, Y. (2004). Excision of a juxtapapillary duodenal diverticulum causing biliary obstruction: report of three cases. *Journal of hepato-biliary-pancreatic surgery*, *11*(1), 69–72. https://doi.org/10.1007/s00534-003-0854-7

[8]. Chiang, T. H., Lee, Y. C., Chiu, H. M., Huang, S. P., Lin, J. T., & Wang, H. P. (2006). Endoscopic therapeutics for patients with cholangitis caused by the juxtapapillary duodenal diverticulum. *Hepato-gastroenterology*, *53*(70), 501–505.

[9]. Desai, K., Wermers, J. D., &Beteselassie, N. (2017). Lemmel Syndrome Secondary to Duodenal Diverticulitis: A Case Report. *Cureus*, *9*(3), e1066. https://doi.org/ 10.7759/cureus.1066

[10] .Teven, C. M., Grossman, E., Roggin, K. K., & Matthews, J. B. (2012). Surgical management of pancreaticobiliary disease associated with juxtapapillary duodenal diverticula: case series and review of the literature. *Journal of gastrointestinal surgery : official journal of the Society for Surgery of the Alimentary Tract*, *16*(7), 1436–1441. https://doi.org/10.1007/s11605-012-1856-z

[11]. Mahajan Sanjay, K., Rajesh, K., ChandelUpender, K., & Jatinder, M. (2004). Duodenal diverticulum: review of literature. *Indian journal of surgery*, *66*(3), 140-145.

[12]. Lobo, D. N., Balfour, T. W., &Iftikhar, S. Y. (1998). Periampullary diverticula: consequences of failed ERCP. *Annals of the Royal College of Surgeons of England*, *80*(5), 326–331.

[13]. Kua, J. E., Seah, A., & So, J. B. (2005). Periampullary diverticulum: a case of bleeding from a periampullary diverticulum. *Annals of the Academy of Medicine, Singapore*, *34*(10), 636–638.

[14]. Harford, W. V. (1998). Diverticula of the hypopharynx and esophagus, the stomach and small bowel. *Sleisenger and Fordtran's Gastrointestinal and Liver Diseases. 6th (Ed). Philadelphia: WB Saunders Co*, *1*, 313-6.

[15]. Chen, Q., Li, Z., Li, S., Ding, X., Liu, Z., Wu, C., Gong, J., &Zuo, G. (2010). Diagnosis and treatment of juxta-ampullary duodenal diverticulum. *Clinical and investigative medicine. Medecinecliniqueet experimentale*, *33*(5), E298–E303. https://doi.org/10.25011/cim.v33i5.14355

[16]. Macari, M., Lazarus, D., Israel, G., &Megibow, A. (2003). Duodenal diverticula mimicking cystic neoplasms of the pancreas: CT and MR imaging findings in seven patients. *AJR. American journal of roentgenology*, *180*(1), 195–199. https:// doi.org/10.2214/ajr.180.1.1800195.

[17]. Sherlock, S., & Dooley, J.S. (2001). Imaging of the Biliary Tract: Interventional Radiology and Endoscopy.

[18]. Mathis, K. L., & Farley, D. R. (2007). Operative management of symptomatic duodenal diverticula. *American journal of surgery*, *193*(3), 305–309. https://doi.org/10.1016/j.amjsurg.2006.09.024

[19]. Martins, P. N., Benckert, C., Vetzke-Schlieker, W., Pratschke, J., Tullius, S. G., &Neuhaus, P. (2007). Intraduodenal diverticulum associated with a double common bile duct causing recurrent pancreatitis and cholangitis: report of a case. *Surgery today*, *37*(4), 320–324. https://doi.org/10.1007/s00595-006-3401-8