Lemmel Syndrome Presenting as Biliary Pancreatitis: A Rare Case of Periampullary Duodenal Diverticulum Diagnosed by MRCP and EUS

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ABSTRACT

Background

Pancreatitis Panniculitis and Polyarthritis (PPP) syndrome is a rare triad of pancreatitis, subcutaneous fat necrosis (panniculitis), and polyarthritis, caused by the extravasation of pancreatic enzymes into the bloodstream. This can result in fat necrosis within various tissues, including bone and joints, leading to painful, swollen joints and skin lesions. Given its rarity, PPP syndrome is often misdiagnosed, and there is no established management protocol. We present a case of PPP syndrome that was initially misdiagnosed and treated as septic arthritis, highlighting the importance of recognizing this uncommon entity.

Case Presentation

A 35-year-old Indian male presented to the emergency department with diffuse joint pain, redness, and swelling for more than 20 days. His medical history was significant for chronic pancreatitis, anemia, pleural effusion, portal vein thrombosis, tuberculosis, and alcohol consumption. He had no recent trauma, travel history, or contact with infectious diseases, including COVID-19. The patient was febrile, tachycardic, and normotensive, with diffuse swelling and tenderness in multiple joints, including the knees, ankles, elbows, and wrists. Laboratory findings showed leukocytosis, elevated inflammatory markers, and elevated amylase levels.

Initial diagnostic imaging, including X-rays, showed no joint abnormalities, and arthrocentesis yielded purulent fluid that tested negative for microorganisms but revealed lipid crystals. This ruled out septic arthritis and raised suspicion for a pancreatic origin. A CT scan of the abdomen confirmed the diagnosis of chronic pancreatitis, with multiple calcifications in the pancreas and portal vein thrombosis. A joint biopsy was performed, and histopathology revealed extensive fat necrosis and the characteristic “ghost cells” of PPP syndrome.

The patient was started on a course of prednisolone, which led to significant clinical improvement, and he was discharged after 29 days of hospitalization. He was placed on a tapering regimen of steroids over six weeks.

Conclusion

PPP syndrome is a rare but important differential diagnosis in patients with recurrent joint effusion, especially in the setting of chronic pancreatitis. Timely recognition and appropriate management can prevent unnecessary surgeries and reduce morbidity. Though there is no established treatment protocol, corticosteroids may offer a promising treatment option for managing extra-pancreatic manifestations. Further research is needed to establish guidelines for the diagnosis and management of PPP syndrome.

Introduction

Lemmel’s syndrome is a pancreaticobiliary disease secondary to periampullary duodenal diverticulum compressing the distal common bile duct. Diverticula from the duodenum are common; the term “peri-ampullary diverticulum” refers to the outpouching of duodenal mucosa that develops within 2 or 3 cm from the ampulla of Vater [1]. The duodenal diverticulum can also cause sphincter of Oddi dysfunction, leading to pancreatitis and cholangitis. Currently, there are very few case reports of this condition, which was first named in 1934 after Dr. Gerhard Lemmel [2]. Here, we report a case of biliary pancreatitis caused by Lemmel syndrome to emphasise its consideration as a rare differential diagnosis [3]. This case report has been reported in line with the SCARE criteria. [4].

CASE PRESENTATION

An 83-year-old Kuwaiti female known case of Parkinson’s disease presented to the emergency department with sudden onset epigastric pain. The pain started after eating soup, non-radiating, associated with facial asymmetry, mouth drop and dysarthria. Her relatives mentioned that dysarthria and mouth drop usually happens to her after any stressful event. There was no other neurological defect, fever, nausea, vomiting, jaundice, or any changes in the bowel or urinary habits. There was no significant family history of malignancy. She had similar pain two years ago, underwent abdominal ultrasound (US) which showed biliary mud and took (Ursodeoxycholic acid) at that time, but she discontinued it on her own. She is also a known case of seasonal asthma, Irritable Bowel Syndrome , and depression with no past surgical history. Currently, she is on Levodopa/carbidopa 100/25 mg, Desvenlafaxine 50mg, Escitalopram 10mg, Olanzapine 10mg, Lidocaine patches 5% patch, Movicol sachet PRN, Prucalopride 2 mg, Boldolaxine 12mg, Rivastigimine 9.5mg/hr patch, Bromazepam 3 mg. She is known to have penicillin allergy.

On examination, she was alert, oriented to time, place, and person. Her vitals: BP 185/95, HR 80, SpO2 98% on RA, T 36.7 °. There was no sign of pallor, jaundice, clubbing, or dehydration., there was epigastric and right hypochondrial tenderness, but Murphy’s sign was negative. Abdomen was not distended and there was no organomegaly, or ascites. Neurological examination consistent with her baseline Parkinson’s disease, showing advanced bradykinesia, rigidity, and tremor in all limbs. There was no restricted eye movement, nystagmus, or pronator drift. Normal sensation in all limbs without signs of lateralization.

Abdominal US showed gallbladder stone measured to be (8.5 mm) in size, with normal wall thickness and no pericholecystic fluid could be detected. Dilated CBD measures 12 mm in maximum caliber with prominent bilobar intrahepatic biliary radicles.

Contrast enhanced abdominopelvic Computed Tomography CT scan revealed evidence of distended gallbladder associated with dilated proximal biliary duct and minimal intrahepatic central biliary ductal dilations. No definable biliary stones. The distal CBD could not be clearly traced. Distended duodenum and proximal jejunum with suggested second duodenal medial aspect diverticular outpouching with no signs of inflammation. Prominent main pancreatic duct, and the pancreas appears slightly bulky regardless of her age, however, it shows homogenous normal parenchymal with no solid or cystic lesions or fluid collections. No peripancreatic inflammatory changes are seen. Further assessment with MRCP was needed.

Further assessment with Magnetic Resonance Cholangiopancreatography (MRCP) which showed evidence of dilated CBD till its distal portion where there is abrupt termination of its lumen by virtue of suspected an ill-defined soft tissue lesion rather than a stone with subsequent mild central intrahepatic biliary radicles dilatation, over distended gallbladder and prominent pancreatic duct giving the appearance of double-duct sign.

Endoscopic Ultrasound (EUS) confirmed the diagnosis of Lemmel syndrome, which showed large bilobed periampullary diverticulum with dilated biliary duct and no obvious mass.
Throughout hospital stay, patient pain was improving. She was treated with aggressive IV fluids and was kept on a fat-free diet and ursodeoxycholic acid. MRCP was arranged to be repeated after three months and follow up in the out-patient clinic.

Investigation	Value	Normal Range
White Blood Cell Count	7.5 × 10⁹/L	4.0–11.0 × 10⁹/L
Haemoglobin	131 g/L	120–160 g/L
Alkaline Phosphatase (ALP)	90 IU/L,	40–130 IU/L
Alanine Aminotransferase (ALT)	<5 U/L,	7–56 U/L
Aspartate Aminotransferase (AST)	28 IU/L	10–40 IU/L
Gamma-Glutamyl Transferase (GGT)	11 IU/L	9–48 IU/L
Total Bilirubin	6 µmol/L	5–21 µmol/L
Direct Bilirubin	1 µmol/L	0–5 µmol/L
Lipase	462 IU/L	0–160 IU/L
Amylase	167 µmol/L	25–125 µmol/L

Table 1: Laboratory Investigations on Admission

DISCUSSION

Lemmel syndrome was first described in 1934 by Lemmel, as an obstructive jaundice in the absence of choledocholithiasis or neoplasm due to a periampullary duodenal diverticulum (PAD) [5]. PAD typically occurs along the medial aspect of the second or third part of the duodenum [6]. Although the majority of patients with PAD are usually asymptomatic, complications can occur in about 5% of cases. These complications include bleeding, perforation, diverticulitis, pancreatitis, choledocholithiasis, cholangitis, jaundice, enterolith formation, intestinal obstruction, etc. [7][8]. There are various etiologies regarding the pathogenesis leading to the development of Lemmel syndrome: First, direct mechanical irritation of periampullary diverticula may cause chronic inflammation of the ampulla, which leads to fibrosis of the papilla. Second, periampullary diverticula may cause dysfunction of the sphincter of Oddi. Third, as in our case, the distal common bile duct (CBD) or ampulla can be compressed mechanically by periampullary diverticula, which showed in our patient MRCP the appearance of double-duct sign [9]. Based on recent case reports, only few described a double-duct sign that mimicked a malignancy [10]. Presence of double-duct sign is typically associated with carcinoma of the pancreatic head and an ampullary tumor, which were differential diagnoses in our case [11] [12]. Imaging is the examination of choice to confirm diagnosis. Its modalities include US, CT or MRCP. Ultrasonography is usually the first imaging method in the ER showing indirect signs of biliary obstruction: dilation of the biliary ducts. MRCP is the examination of choice to confirm diagnosis and evaluate biliary tract anomalies and eliminate other differential diagnoses. CT is usually the first line imaging method to either lead or confirm diagnosis due to its fast acquisition and availability. It can be done with intravenous contrast enhancement or oral - contrast [13]. In clinically indicated cases, Endoscopic Cholangiopancreatography (ERCP) is another alternative that can be diagnostic and therapeutic [14]. In patients with mild symptoms, diet modification and diverticular lavage may offer complete resolution of symptoms. In more severe cases, endoscopic biliary cannulation and sphincterotomy may provide adequate relief and circumvent the need for invasive surgical intervention [15].

In comparison, other documented cases have shown variability in treatment options, with ERCP and sphincterotomy as first-line interventions, but surgical resection may be necessary in cases where ERCP is not feasible or fails [16]. The current case, managed conservatively after diagnosis by imaging, highlights the effectiveness of non-invasive approaches. However, in cases of recurrent cholangitis or severe biliary obstruction, surgical options, including diverticulectomy or hepaticojejunostomy, have proven effective [17].

Lemmel syndrome, though rare, should always be considered in patients presenting with obstructive jaundice in the absence of gallstones or neoplasia [17][18] . The presentation of this case further demonstrates that early recognition and appropriate diagnostic modalities are critical for avoiding unnecessary invasive procedures.

CONCLUSION

This case adds to the limited but growing body of literature on Lemmel syndrome. It underscores the importance of considering periampullary duodenal diverticula in the differential diagnosis of patients presenting with obstructive jaundice without choledocholithiasis or malignancy [17][18]. As the diagnostic difficulty of Lemmel syndrome remains a challenge, clinicians should utilize advanced imaging techniques such as MRCP and ERCP, which can offer both diagnostic and therapeutic benefits [12].

Given that the majority of periampullary diverticula are asymptomatic, clinicians should be vigilant for symptomatic cases that can lead to serious complications such as cholangitis, pancreatitis, or biliary obstruction. Treatment decisions should be based on the severity of symptoms, and while endoscopic interventions like ERCP are often sufficient, surgical options should be considered when endoscopic management fails [16]. Further studies are necessary to further explore long-term outcomes and recurrence rates in patients managed with non-surgical versus surgical approaches [16].

Although Lemmel syndrome is a rare cause of biliary pancreatitis, it should be considered in the absence of choledocholithiasis or neoplasm. Presence of PAD and double-duct sign should raise the suspicion of Lemmel syndrome. Imaging is the modality of choice to confirm diagnosis. As for treatment, it mainly depends on the patient's symptoms and complications. These may include conservative, endoscopic, and surgical intervention. Failure to recognize this condition may lead to mistreatment, which could lead to serious health complications.

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