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Lemmel syndrome: A single-center cohort and diagnostic considerations

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ABSTRACT

Objective: Lemmel syndrome is an uncommon cause of obstructive jaundice resulting from extrinsic compression of the common bile duct by a periampullary duodenal diverticulum (PAD) in the absence of choledocholithiasis. Due to its non-specific clinical presentation, diagnosis may be delayed or misinterpreted.

Material and Methods: A retrospective review was conducted on 12 patients diagnosed with Lemmel syndrome at a single tertiary center between 2022 and 2024. Demographic characteristics, clinical presentation, laboratory findings, imaging modalities [ultrasonography (USG), computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP)], endoscopic interventions, and outcomes were analyzed descriptively.

Results: The cohort included 12 patients with a mean age of 70±9 years. All patients presented with clinical or biochemical evidence of obstructive jaundice. USG frequently failed to establish the diagnosis, whereas CT and particularly MRCP demonstrated the anatomical relationship between the PAD and the biliary tree. Endoscopic retrograde cholangiopancreatography served both diagnostic and therapeutic purposes, with biliary stenting performed when indicated. Most patients improved with conservative and/or endoscopic management.

Conclusion: Lemmel syndrome should be considered in elderly patients with unexplained obstructive jaundice. Cross-sectional imaging, particularly MRCP, plays a key role in diagnosis, while endoscopic intervention remains central to management.

Keywords: Lemmel syndrome, periampullary duodenal diverticulum, obstructive jaundice

INTRODUCTION

Lemmel syndrome is an uncommon clinical presentation characterized by extrinsic compression of the common bile duct and obstructive jaundice secondary to a periampullary duodenal diverticulum (PAD) in the absence of choledocholithiasis (1). First described by Lemmel in 1934, this syndrome has increasingly been reported in the literature through case reports and case series, particularly with the widespread use of advanced cross-sectional imaging (2). Its nonspecific clinical presentation and frequent occurrence in elderly patients with multiple comorbidities may lead to diagnostic delay or misinterpretation rather than true epidemiological rarity.

PADs are mucosal outpouchings located around the second part of the duodenum, particularly near the ampulla of Vater (major papilla) (3). Autopsy and endoscopic studies have reported PAD prevalence ranging from 2% to 27%, suggesting the true prevalence may be higher since many cases are asymptomatic (4). Although PADs are relatively common anatomical findings, only a small proportion of patients develop clinically significant complications such as cholangitis, pancreatitis, bleeding, or biliary obstruction, which are reported in approximately 5% of cases (5,6). Lemmel syndrome represents an uncommon complication of this common anatomical entity rather than an intrinsically rare disease. Differential diagnoses for Lemmel syndrome include choledocholithiasis, periampullary tumors, and Mirizzi syndrome (7). PADs can also be classified according to their anatomical relationship with the major papilla (e.g., periampullary or juxtapapillary types), a distinction that may influence endoscopic accessibility and technical success during endoscopic retrograde cholangiopancreatography (ERCP).

For diagnostic evaluation, ultrasonography (USG) is commonly used due to its accessibility and ability to detect bile duct dilatation. However, USG is often insufficient in identifying PADs due to intestinal gas and anatomical variations (8).

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Computed tomography (CT) provides more detailed visualization of the periampullary region, while magnetic resonance cholangiopancreatography MRCP offers superior diagnostic accuracy, especially in suspected cases of Lemmel syndrome. MRCP is crucial for delineating the three-dimensional anatomical relationship between the PAD and the bile/pancreatic ducts. ERCP remains the gold standard, serving both diagnostic and therapeutic purposes, including stone removal and stent placement (9).

In this study, the demographic, clinical, and radiological characteristics, as well as endoscopic findings, of 12 patients diagnosed with Lemmel syndrome in our clinic were retrospectively evaluated. This case series aimed to contribute additional observational data to the existing literature on Lemmel syndrome, with a particular focus on diagnostic evaluation and management in elderly patients. Specifically, it aimed to clarify diagnostic challenges, evaluate the efficacy of imaging and endoscopic modalities, and identify practical therapeutic strategies to improve patient outcomes.

MATERIAL and METHODS

This retrospective descriptive case series was conducted on patients diagnosed with Lemmel syndrome at University of Health Sciences Türkiye, Ankara Etlik City Hospital, between 2022 and 2024, in accordance with the surgical case report 2023 guidelines (10). The study population consisted of individuals who presented with obstructive jaundice and were subsequently found to have extrinsic common bile duct compression secondary to a PAD in the absence of choledocholithiasis or malignancy. A total of 12 patients met these criteria and were included after a thorough review of hospital data storage and patient files.

This study was approved by the Ethics Committee of University of Health Sciences Türkiye, Ankara Etlik City Hospital (approval no: AEŞH-BADEK-2024-1240, date: 18.12.2024), and was conducted in accordance with the Declaration of Helsinki. Written informed consent was obtained from all patients.

Demographic data, including age, sex, and relevant comorbidities, such as hypertension, diabetes mellitus, and cardiac conditions, were obtained from medical records. Additionally, clinical symptoms associated with jaundice, including right upper quadrant pain, nausea, vomiting, and changes in urine and stool color, were documented. Laboratory parameters, including total, direct, and indirect bilirubin levels, as well as liver enzyme levels [alanine aminotransferase (ALT) and aspartate aminotransferase (AST)], were analyzed to assess cholestatic liver injury. Imaging modalities such as USG, CT, and MRCP were meticulously evaluated to confirm bile duct compression due to a PAD.

The therapeutic approach varied depending on clinical presentation and imaging findings. Patients were managed conservatively, with symptomatic treatment including proton pump inhibitors, antiemetics, and intravenous fluid support. In cases where endoscopic intervention was required, ERCP was performed to relieve biliary obstruction, often through the placement of a biliary stent. Procedural details, including the type of intervention performed and any associated complications such as bleeding or perforation, were retrospectively reviewed from patient records. The duration of hospital stay and follow-up period were also recorded.

Patients were included in the study based on the presence of obstructive jaundice supported by elevated serum bilirubin (predominantly direct bilirubin) and liver enzyme abnormalities (ALT and AST), along with imaging or endoscopic confirmation of extrinsic bile duct compression by a PAD. They were excluded if other causes of biliary obstruction, such as Mirizzi syndrome, gallbladder carcinoma, or pancreaticoduodenal tumors, were identified. Additionally, those with incomplete medical records or asymptomatic individuals with incidental PADs were not included in the study.

Imaging modalities were considered "sufficient" when they provided clear visualization of the PAD and its anatomical relationship to the common bile duct, allowing for confident diagnosis of Lemmel syndrome. "Insufficient" imaging lacked adequate resolution or anatomical clarity to establish this relationship.

Statistical Analysis

For statistical analysis, numerical variables were expressed as the mean ± standard deviation or median with minimum and maximum values, while categorical variables were presented as frequencies and percentages. Due to the small sample size, advanced statistical comparisons were not performed, and data analysis was primarily descriptive.

RESULTS

Demographic data and clinical presentation are shown in Table 1. Jaundice was the most common presenting symptom on clinical presentation, observed in all patients (100%), accompanied by scleral icterus and yellowish skin discoloration. Five patients had a history of non-malignant cholangitis or cholecystitis episodes.

Laboratory findings at initial presentation revealed a cholestatic pattern of liver function test abnormalities in all patients (Table 2). The median PAD diameter measured on cross-sectional imaging was 35 mm (range: 20-48 mm). Four patients (33%) had PADs measuring ≥40 mm.

Radiological and endoscopic evaluations played a crucial role in diagnosis. All patients underwent at least one imaging modality, and their diagnostic contributions were categorized as "sufficient" or "insufficient" (Table 3).

All patients initially received medical management, including proton pump inhibitors, antiemetics, and intravenous fluid support. ERCP was considered for those with significant biliary obstruction, particularly those with elevated bilirubin levels. Among the five patients who underwent stenting, three demonstrated significant clinical and biochemical improvement, whereas two elderly patients with multiple comorbidities showed only partial resolution of the symptoms. The mean duration of hospital stay was 12±6 days (range: 2-56), and the average follow-up duration was 8 months.

Table 1. Demographic and clinical characteristics of the study population (n=12)

Variable	Value
Age, mean ± SD (range), years	70±9 (43-87)
Sex, n (%)	
Female	7 (58.3%)
Male	5 (41.7%)
Comorbidities, n (%)	
Hypertension	9 (75%)
Diabetes mellitus	6 (50%)
Cardiovascular disease	7 (58.3%)
Chronic obstructive pulmonary disease	2 (16.7%)
Myeloma	1 (8.3%)
Bullous pemphigoid	1 (8.3%)
Presenting symptoms, n (%)	
Jaundice	12 (100%)
Right upper quadrant pain	6 (50%)
Dyspepsia	10 (83.3%)
History of cholangitis/cholecystitis	5 (41.7%)

SD: Standard deviation.

Table 2. Laboratory findings at presentation

Parameter	Median [min-max], n=12
Total bilirubin (mg/dL)	3.05 [0.31-9.6]
Direct bilirubin (mg/dL)	1.7 [0.12-5.5]
ALT (U/L)	166.5 [11-419]
AST (U/L)	161 [13-471]

ALT: Alanine aminotransferase, AST: aspartate aminotransferase.

One patient with a known history of advanced gastric malignancy and multiple systemic comorbidities died during the follow-up period. Mortality was attributed to progression of the underlying malignancy and sepsis and was not considered to be directly related to Lemmel syndrome or to any diagnostic or therapeutic endoscopic intervention performed during hospitalization. The remaining patients were discharged after resolution of the obstructive jaundice, with no major complications noted during outpatient follow-up.

DISCUSSION

This case series adds further observational data to the expanding literature on Lemmel syndrome, particularly in an elderly and comorbid patient population. The findings corroborate previously reported diagnostic challenges, imaging characteristics, and outcomes of endoscopic management in routine clinical practice. Lemmel syndrome may be overlooked in elderly patients with nonspecific presentations and multiple comorbidities; careful integration of clinical findings with appropriate cross-sectional imaging and endoscopic evaluation remains essential for accurate diagnosis and effective management.

PADs are generally asymptomatic and are often detected incidentally during endoscopic or radiological imaging. However, their anatomical proximity to the common bile duct or pancreatic duct may lead to severe clinical conditions such as obstructive jaundice, cholangitis, or pancreatitis (11). In the current series, elevations in liver enzymes in addition to hyperbilirubinemia can be adequately explained by biliary obstruction and, in some cases, associated cholangitis, which are well-recognized biochemical manifestations of extrahepatic biliary compression. These non-specific symptoms, especially in patients with multiple comorbidities, may mimic malignancy or choledocholithiasis, leading to misdiagnosis and inappropriate initial work-up. Previous studies have emphasized that Lemmel syndrome can present with atypical symptoms and is often misdiagnosed as malignancy, choledocholithiasis, or Mirizzi syndrome (12). Similarly, in the present series, several patients presented with dyspeptic complaints and non-specific

Table 3. Diagnostic performance of the imaging and endoscopic modalities (n=2)

Modalities	Patients evaluated, n	Sufficient, n (%)	Insufficient, n (%)	Not performed, n (%)
USG	11	3 (27.3)	7 (63.6)	1 (8.3%)
CT	10	5 (50)	5 (50)	2 (16.7%)
MRCP	9	6 (66.7)	3 (33.3)	3 (25%)
EGD	12	8 (66.7)	4 (33.3)	0 (0%)
ERCP	7	5 (71.4)	2 (28.6)	5 (41.7%)
EUS	4	4 (100)	0 (0%)	8 (66.7%)

USG: Ultrasonography, CT: Computed tomography, MRCP: Magnetic resonance cholangiopancreatography, EGD: Gastroduodenoscopy, ERCP: Endoscopic retrograde cholangiopancreatography, EUS: Endoscopic ultrasound.

abdominal pain in addition to jaundice, which may further complicate USG diagnostic evaluation.

Despite its widespread use as an initial diagnostic modality, demonstrated limited sensitivity in detecting PAD-related extrinsic compression in most cases. This finding aligns with prior literature, where even large PADs may be overlooked due to interference from bowel gas and anatomical overlap. Conversely, CT can visualize mass-like lesions or diverticular extensions in the periampullary region, yet it may present difficulties in differential diagnosis (Figure 1) (8). The current findings indicated that MRCP played a crucial role in establishing the diagnosis (Figure 2). By providing a three-dimensional assessment of the relationship between PADs and the biliary and pancreatic ducts, MRCP may be essential in differentiating Lemmel syndrome from choledocholithiasis and malignancies. These findings support the consideration of early MRCP evaluation, particularly in elderly patients with obstructive jaundice of unclear etiology. Given its non-invasive nature and superior anatomical resolution, MRCP should be considered early in the diagnostic algorithm for elderly patients with unexplained biliary obstruction.

Endoscopic interventions, including gastroduodenoscopy (EGD) and ERCP, represent the cornerstone of both diagnostic and therapeutic approaches in Lemmel syndrome (12). In the present series, gastroduodenoscopy was performed in all patients to identify the position of the major papilla within or adjacent to the PAD (Figure 3). In some cases, assessment was technically challenging due to large size of the PAD and altered periampullary anatomy. ERCP facilitated both anatomical clarification and therapeutic biliary stenting, effectively resolving obstructive jaundice (13). However, technical difficulty was encountered in two cases with massive PADs, where complete ERCP procedures could not be performed.

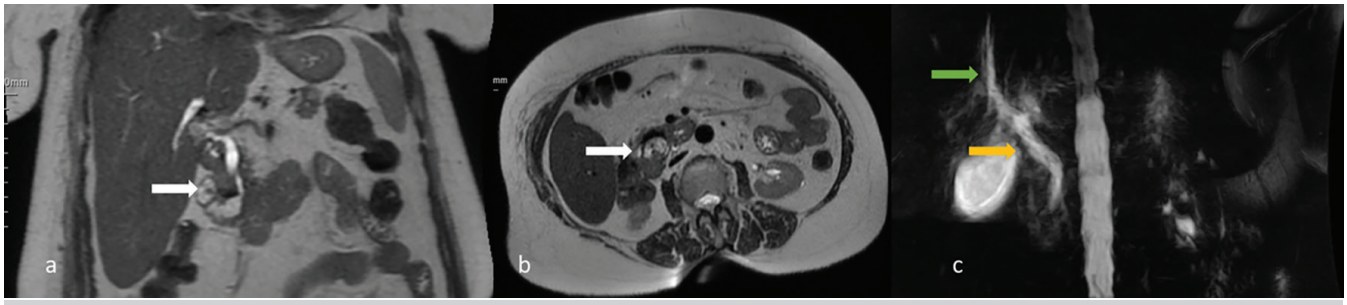

Figure 1. Upper abdominal magnetic resonance imaging: coronal and axial T2 sections showed a periampullary duodenal diverticulum (PAD) (white arrows) (**a, b**). Magnetic resonance cholangiopancreatography: Biliary sequence showing a PAD causing dilatation of the common bile duct (yellow arrow) and the proximal part of intrahepatic bile duct dilatation (green arrow) (**c**).

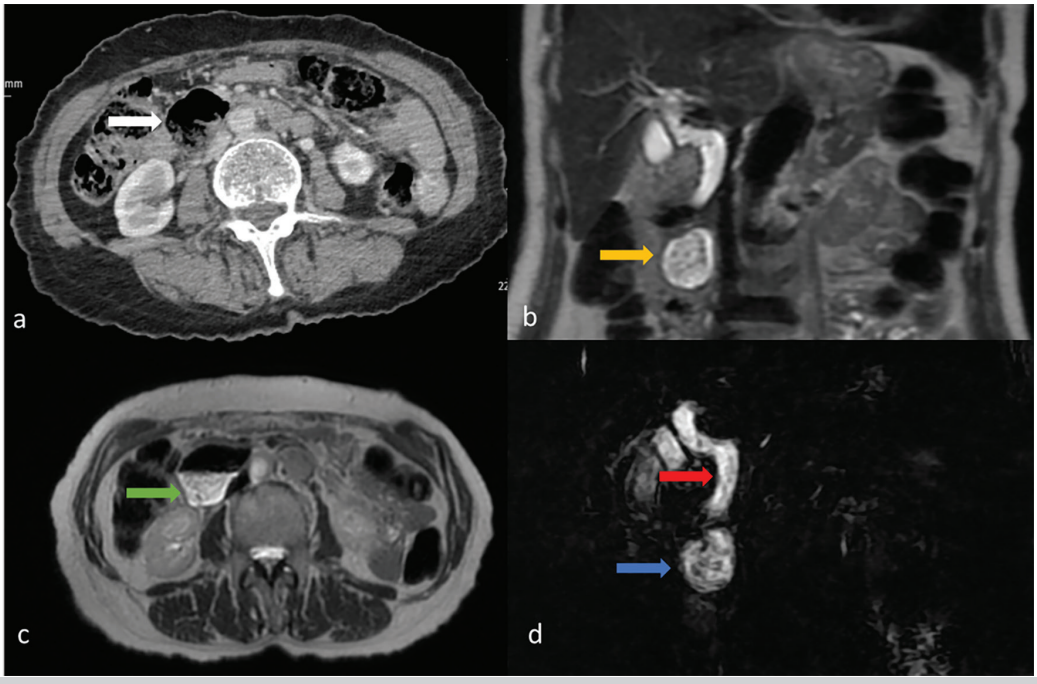

Figure 2. Abdominal computed tomography scan demonstrating a periampullary duodenal diverticulum (PAD) (white arrow) with resultant upstream dilatation of the common bile duct (**a**). Magnetic resonance cholangiopancreatography: Coronal (**b**) and axial (**c**) T2 sections showing a PAD of the same patient (yellow and green arrows). 3D biliary sequence showing a PAD (blue arrow) causing dilatation of the common bile duct (red arrow) (**d**).

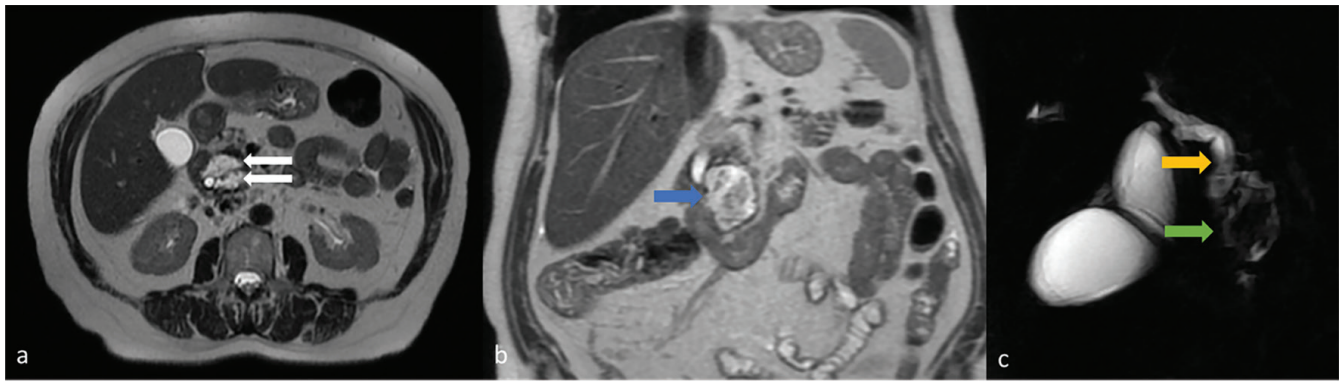

Figure 3. Magnetic resonance cholangiopancreatography: Axial T2 sections showing two periampullary duodenal diverticulum (PAD) adjacent to each other (white arrows) (**a**) and coronal T2 sections showing a PAD (blue arrow) (**b**). 3D biliary sequence showing a PAD (green arrow) causing dilatation of the common bile duct (yellow arrow) (**c**).

Endoscopic ultrasound (EUS) was utilized in four patients and contributed to the diagnostic evaluation.

As reported in previous literature, EUS offers detailed visualization of extraluminal structures and may serve as a valuable adjunctive modality when ERCP is technically challenging (12). Nevertheless, EUS remains operator-dependent, and its diagnostic performance may vary according to institutional expertise.

The presence of advanced age and multiple systemic comorbidities in most patients in our study added complexity to the management of Lemmel syndrome. Coexisting conditions, such as cardiovascular, endocrine, or respiratory diseases, not only contributed to diagnostic delays but also complicated treatment planning. In patients with significant comorbidities, conservative or endoscopic management was prioritized over radical surgical interventions. However, in cases where conservative treatment was inadequate, surgical options were considered. In our series, one patient who exhibited persistent symptoms and recurrent clinical episodes despite medical treatment underwent gastrojejunostomy with Braun anastomosis, which was successfully performed. Postoperative follow-up revealed no additional complications. However, our experience suggests that conservative or endoscopic management may be appropriate as an initial approach in clinically stable patients.

Study Limitations

This study had several limitations, including its single-center, retrospective design and small sample size, which precluded comparative statistical analysis. Although the PAD size measurements were available, the limited sample size also restricted meaningful assessment of potential correlations between the PAD diameter and biochemical parameters. Additionally, some diagnostic procedures (e.g., USG, ERCP) are operator-dependent, introducing variability in diagnostic accuracy. Despite these constraints, the series contributes additional descriptive data within the context of a relatively uncommon clinical presentation.

In conclusion, clinicians should maintain a high index of suspicion for Lemmel syndrome in elderly patients presenting with unexplained obstructive jaundice. A structured diagnostic pathway—initiating with MRCP and supplemented by endoscopy—may reduce delays and optimize outcomes. Given the rarity of the condition and the lack of standardized protocols, larger multicenter studies are warranted to validate these observations and refine treatment strategies.

CONCLUSION

Lemmel syndrome should be considered in elderly patients presenting with obstructive jaundice, as its clinical presentation can often mimic other causes of biliary obstruction. Diagnosis and management rely primarily on advanced imaging and endoscopic techniques, particularly MRCP and ERCP, with EUS serving as a complementary tool when needed. Given the complexity of the condition, a multidisciplinary approach involving gastroenterologists, surgeons, and radiologists is essential to ensure optimal patient outcomes and to tailor individualized treatment strategies.

Ethics

Ethics Committee Approval: This study was approved by the Ethics Committee of University of Health Sciences Türkiye, Ankara Etlik City Hospital (approval no: AEŞH-BADEK-2024-1240, date: 18.12.2024), and was conducted in accordance with the Declaration of Helsinki.

Informed Consent: Written informed consent was obtained from all patients.

Footnotes

Author Contributions

Surgical and Medical Practices - E.E., S.D., M.S.S., Ş.B.; Concept - E.E.; Design - E.E., S.D.; Data Collection or Processing - E.E., S.D., Ş.E.; Analysis or Interpretation - E.E., S.D., M.S.S.; Literature Search - E.E., S.D.; Writing - E.E., S.D.

Conflict of Interest: No conflict of interest was declared by the authors.

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