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Endoscopic Ultrasound in the Diagnosis of Low Phospholipid-Associated Cholelithiasis (LPAC) Syndrome: A Case Report

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Abstract | **Case Report**

Low Phospholipid-Associated Cholelithiasis (LPAC) syndrome is an underdiagnosed cause of biliary disorders, typically characterized by recurrent biliary symptoms despite cholecystectomy. Although LPAC classically presents with recurrent biliary pain, acute pancreatitis may represent an unusual revealing manifestation. We report a case of LPAC syndrome diagnosed following an episode of acute pancreatitis. A 45-year-old man with a history of biliary pancreatitis and cholecystectomy presented with epigastric pain and vomiting. Laboratory investigations revealed elevated serum lipase, marked hepatocellular cytolysis, and cholestasis. Contrast-enhanced CT demonstrated Balthazar grade C acute pancreatitis with segmental intrahepatic bile duct dilatation. Transabdominal ultrasound was unremarkable, and magnetic resonance cholangiopancreatography (MRCP) showed peripancreatic inflammation without bile duct dilatation. This imaging discordance prompted further evaluation with endoscopic ultrasound (EUS). EUS, performed after resolution of the acute inflammatory phase, identified multiple punctate hyperechoic intrahepatic foci distributed along the segmental bile ducts, associated with posterior acoustic shadowing and comet-tail artifacts, consistent with intrahepatic cholesterol microlithiasis. Alternative causes of intrahepatic hyperechoic foci were excluded based on imaging characteristics and clinical context. LPAC syndrome was diagnosed, and treatment with ursodeoxycholic acid was initiated. At 6-month follow-up, the patient remained asymptomatic, with normalization of liver biochemical parameters. This case underscores the importance of considering LPAC syndrome in patients with unexplained biliary symptoms or idiopathic pancreatitis after cholecystectomy and highlights the added diagnostic value of EUS when cross-sectional imaging is inconclusive.

Keywords: Case Report, LPAC Syndrome, Pancreatitis.

INTRODUCTION

Low Phospholipid-Associated Cholelithiasis (LPAC) syndrome, also referred to as genetic cholesterol lithiasis, is a distinct form of biliary stone disease. In contrast to common cholelithiasis, LPAC is characterized by the formation of cholesterol stones within both the gallbladder and the intrahepatic bile ducts, typically affecting young adults with normal body mass index and frequently recurring after cholecystectomy [1].

The condition is linked to impaired biliary phospholipid secretion, resulting in defective micelle formation and reduced cholesterol solubilization, ultimately leading to the formation of cholesterol crystals and intrahepatic microlithiasis. Mutations in the ABCB4 gene encoding the MDR3 phospholipid transporter have been identified in 30–50% of patients, reflecting partial genetic penetrance and suggesting additional unidentified genetic contributors.

CASE REPORT

Patient Information

A 45-year-old man with a history of biliary pancreatitis and laparoscopic cholecystectomy performed in 2019, and a family history of cholelithiasis, presented with acute transfixing epigastric pain and vomiting of 48 hours' duration.

Clinical Findings

Physical examination was unremarkable apart from epigastric tenderness. No signs of cholangitis were observed.

Diagnostic Assessment

Laboratory testing showed normal leukocyte count, serum lipase elevated to 12 times the upper limit of normal, elevated transaminases (AST 401 IU/L; ALT 836 IU/L), and mild cholestasis (ALP 1.5× upper limit of normal; total bilirubin 19 mg/L).

Contrast-enhanced CT confirmed Balthazar grade C acute pancreatitis and demonstrated segmental dilatation of the left intrahepatic bile ducts. Transabdominal ultrasound was normal. MRCP showed inflammatory pancreatic changes without evidence of bile duct dilatation or choledocholithiasis.

Given the discrepancy between CT and MRCP findings and the absence of common bile duct stones, further investigation was undertaken after clinical resolution of the acute phase.

EUS was performed using a linear echoendoscope with systematic evaluation of the extrahepatic and intrahepatic bile ducts. Hepatic segments II and III were visualized from the gastric position. EUS revealed multiple punctate hyperechoic foci distributed along the intrahepatic bile ducts, associated with posterior acoustic shadowing and comet-tail artifacts (Figures 1 and 2).

Differential diagnoses of intrahepatic hyperechoic foci were carefully considered. Pneumobilia and biliary gas were excluded due to the absence of recent instrumentation and the fixed, non-mobile nature of the lesions without reverberation artifacts typical of intraductal gas. Parasitic infection and post-inflammatory debris were ruled out based on clinical presentation and laboratory findings. Parenchymal calcifications were considered unlikely given the characteristic comet-tail artifact suggestive of cholesterol microlithiasis.

These findings were highly suggestive of intrahepatic cholesterol microlithiasis and, in the appropriate clinical context, were consistent with LPAC syndrome.

Therapeutic Intervention

The diagnosis of LPAC syndrome was established based on recurrent biliary complications after cholecystectomy, positive family history, and characteristic EUS findings. Ursodeoxycholic acid was initiated at a dose of 10 mg/kg/day.

Follow-up and Outcomes

Clinical symptoms resolved within two weeks. Liver enzyme levels progressively normalized over six weeks. At 6-month follow-up, the patient remained asymptomatic, with no recurrence of pancreatitis and stable imaging findings.

DISCUSSION

LPAC syndrome is a rare genetic cholangiopathy characterized by intrahepatic and vesicular cholesterol lithiasis resulting from impaired phospholipid secretion, with an estimated prevalence of approximately 1% among patients with symptomatic cholelithiasis [2].

How this Case Fits the LPAC Phenotype

Although LPAC most commonly affects individuals younger than 40 years, up to 15% of cases are diagnosed after this age threshold [2]. Our patient, aged 45, illustrates that age alone should not preclude consideration of the diagnosis. The recurrence of biliary complications after cholecystectomy and a positive family history further supported the diagnosis.

Acute Pancreatitis as Revealing Manifestation

While recurrent biliary pain is the hallmark presentation, LPAC may be complicated by acute pancreatitis secondary to migration of cholesterol microlithiasis [3, 4]. In this case, pancreatitis was the revealing manifestation, emphasizing the need to consider LPAC in patients with idiopathic pancreatitis following cholecystectomy.

Imaging Discordance and Diagnostic Contribution of EUS

Segmental intrahepatic bile duct dilatation observed on CT was not confirmed on MRCP. This discrepancy may reflect transient inflammatory changes during the acute phase or the limited sensitivity of MRCP for small intrahepatic cholesterol stones. CT is known to have limited sensitivity for cholesterol microlithiasis, and MRCP may appear normal in LPAC [4].

EUS has demonstrated high diagnostic accuracy in detecting microlithiasis missed by CT or MRCP [5, 6]. In our patient, systematic intrahepatic exploration by EUS was decisive in identifying typical comet-tail artifacts along segmental bile ducts. This case therefore supports the integration of EUS into the diagnostic algorithm for post-cholecystectomy patients presenting with unexplained biliary symptoms or idiopathic acute pancreatitis.

Diagnostic Confirmation and Limitations

The diagnosis of LPAC is primarily based on clinical and imaging criteria. Although ABCB4 mutations are identified in 30–50% of cases, genetic testing was not performed in this patient. The diagnosis therefore remains clinically and radiologically supported rather than genetically confirmed.

What This Case Adds

This report highlights three important educational points:

1. LPAC may present as acute pancreatitis several years after cholecystectomy.

2. Imaging discordance between CT and MRCP should prompt further investigation rather than diagnostic dismissal.
3. EUS can provide decisive diagnostic information when cross-sectional imaging is inconclusive.

Treatment

1) Medical Treatment

Treatment is based on long-term ursodeoxycholic acid at 10 mg/kg/day. It increases the pool of hydrophilic bile acids, promoting cholesterol solubilization and dissolution of cholesterol crystals.

In our patient, medical therapy alone was sufficient, with rapid clinical and biochemical improvement.

Statins may be added in case of hypercholesterolemia. Estrogen-progestin therapy should be discontinued when applicable.

2) Interventional and Surgical Management

Endoscopic or radiological drainage is indicated in cases of cholangitis or liver abscess. Stone extraction may be performed using Fogarty balloon or Dormia basket. Lithotripsy may be indicated for large stones [7].

Liver resection may be considered in localized disease with recurrent cholangitis despite adequate medical therapy. Liver transplantation remains exceptional and is reserved for end-stage liver disease [7].

Limitations

This report has several limitations inherent to single-case studies. First, the findings are based on a single patient and therefore cannot be generalized to the broader population of patients with suspected LPAC syndrome. Second, genetic testing for ABCB4 mutations was not performed, and the diagnosis remained supported by clinical and imaging criteria rather than molecular confirmation. Finally, endoscopic ultrasound is an operator-dependent technique, and diagnostic reproducibility may vary according to expertise and equipment availability. Despite these limitations, the concordance of clinical history, imaging findings, and therapeutic response strongly supports the diagnosis of LPAC syndrome in this case.

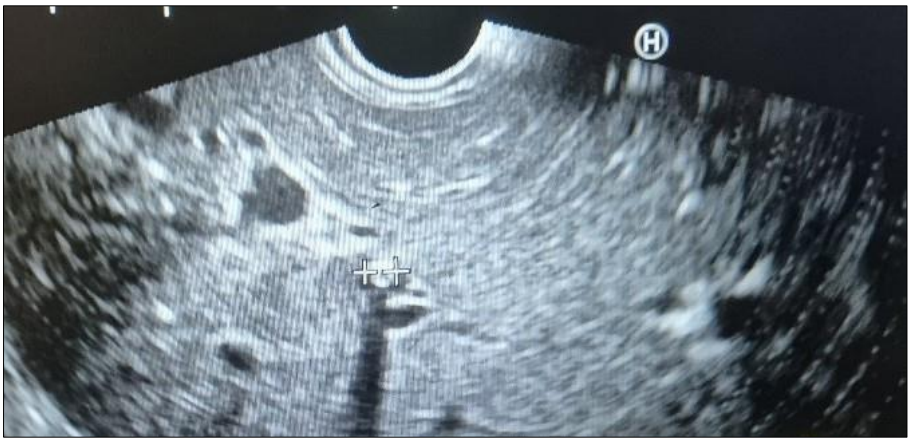

Figure 1: EUS image of hyperechoic intrahepatic formation with posterior shadow cone

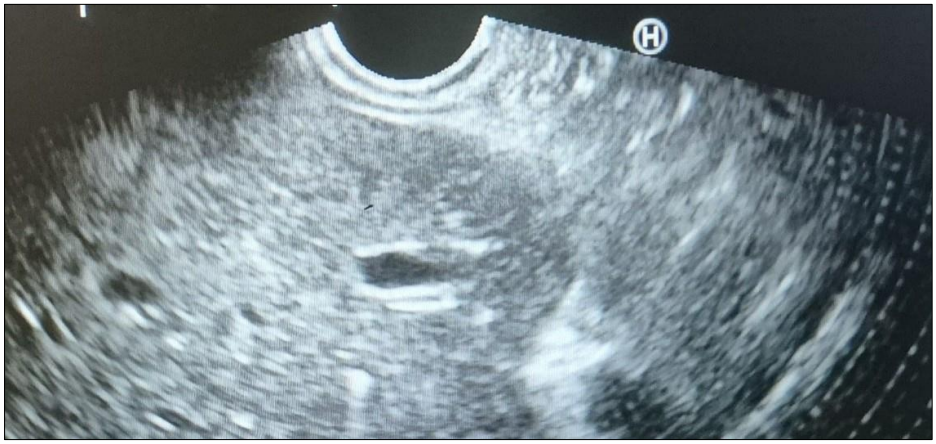

Figure 2: EUS image of hyperechoic intrahepatic formation producing a comet-tail artifact testifying of their cholesterolic character

CONCLUSION

This case illustrates the diagnostic challenges of LPAC syndrome in a post-cholecystectomy patient presenting with acute pancreatitis. Imaging discordance between CT and MRCP should not exclude biliary microlithiasis. Endoscopic ultrasound provided critical diagnostic information when standard imaging was inconclusive.

Although genetic confirmation was not obtained, the convergence of clinical history and characteristic EUS findings strongly supported the diagnosis of LPAC syndrome. Early recognition and treatment with ursodeoxycholic acid may result in favorable outcomes. Further studies are warranted to better define the role of EUS within the diagnostic strategy of suspected LPAC syndrome.

Author Contributions

- Sara Hdiye (corresponding author) contributed to patient management, and manuscript drafting.
- Firdaouss Ait Iken and Chaimae Hdiye participated in data collection, imaging interpretation (CT, MRCP, EUS), and literature review.
- Ahlame Benhamdane and Tarik Addajou contributed to patient follow-up and biochemical data analysis.
- Hassan Seddik participated in critical revision of the manuscript for important intellectual content and provided clinical supervision.

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Informed Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. The patient was informed that all personal identifiers would be anonymized and that the information would be used for educational and scientific purposes.

Patient Perspective

I experienced recurrent biliary pain and acute pancreatitis even after cholecystectomy, which was stressful due to multiple tests and uncertainty. I felt relieved when LPAC syndrome was diagnosed by endoscopic ultrasound. Treatment with ursodeoxycholic acid was easy and well-tolerated, and my symptoms resolved within six months. I am grateful to the medical team for their thorough care and clear communication.

Competing Interests: The authors declare no competing interests.

BIBLIOGRAPHY

1. Rosmorduc O, Poupon R. Low phospholipid associated cholelithiasis: association with mutation in the MDR3/ABCB4 gene. *Orphanet J Rare Dis*. 2007;2:29. doi:10.1186/1750-1172-2-29.
2. Dong C, Condat B, Picon-Coste M, et al. Low-phospholipid-associated cholelithiasis syndrome: Prevalence, clinical features, and comorbidities. *JHEP Rep*. 2021;3(2):100201. doi:10.1016/j.jhepr.2020.100201
3. El Moutaoukil N, Zouaoui M, Hnach Y, Azouaoui M, Aqodad N. Considerations for Low Phospholipid-Associated Cholelithiasis (LPAC) Syndrome: Report of Three Cases. *Cureus*. 15(10):e48082. doi:10.7759/cureus.48082
4. Gille N, Karila-Cohen P, Goujon G, et al. Low phospholipid-associated cholelithiasis syndrome: A rare cause of acute pancreatitis that should not be neglected. *World J Hepatol*. 2020;12(6):312-322. doi:10.4254/wjh.v12.i6.312
5. Kondo S, Isayama H, Akahane M, et al. Detection of common bile duct stones: comparison between endoscopic ultrasonography, magnetic resonance cholangiography, and helical-computed-tomographic cholangiography. *Eur J Radiol*. 2005;54(2):271-275. doi:10.1016/j.ejrad.2004.07.007
6. Smith I, Ramesh J, Kyanam Kabir Baig KR, Mönkemüller K, Wilcox CM. Emerging Role of Endoscopic Ultrasound in the Diagnostic Evaluation of Idiopathic Pancreatitis. *Am J Med Sci*. 2015;350(3):229-234. doi:10.1097/MAJ.0000000000000541
7. Goubault P, Brunel T, Rode A, Bancel B, Mohkam K, Mabrut JY. Low-Phospholipid Associated Cholelithiasis (LPAC) syndrome: A synthetic review. *J Visc Surg*. 2019;156(4):319-328. doi: 10.1016/j.jviscsurg.2019.02.006.