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Case Report

Choledochal Cyst Todani Classification Type IC and Choledocholithiasis Presenting as Recurrent Pancreatitis in a 7-Year-Old Girl – A Case Report

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Abstract:

Background: Choledochal cyst is an uncommon but important cause of biliary pathology in the pediatric population which can lead to choledocholithiasis and recurrent pancreatitis. High clinical suspicion, comprehensive imaging, and multidisciplinary evaluation are essential for accurate diagnosis and treatment. Definitive management is achieved through surgical intervention.

***Case*:** A 7-year-old girl experienced recurrent postprandial right upper quadrant and epigastric pain for three years, with fluctuating pancreatic and hepatic enzymes. Initial ultrasound and magnetic resonance cholangiopancreatography (MRCP) revealed ductal dilatation with biliary sludge, while multislice computed tomography (MSCT) suggested autoimmune pancreatitis, leading to temporary steroid response. Symptoms persisted, and contrast-enhanced magnetic resonance imaging (MRI) demonstrated fusiform common bile duct dilatation with debris, confirming Todani type IC choledochal cyst and choledocholithiasis. She underwent laparoscopic cyst and gallbladder excision with Roux-en-Y hepaticojejunostomy. Intraoperative and histological findings verified sludge, gallstones, and chronic cholecystitis.

***Discussion*:** Choledochal cysts should be considered in pediatric recurrent pancreatitis, particularly with biliary obstruction. Advanced imaging and differential diagnosis are crucial. Surgical excision with biliary reconstruction is the treatment of choice.

Conclusion: Early recognition and timely surgical intervention are essential to prevent long-term complications.

Keywords: autoimmune pancreatitis, biliary obstruction, hepaticojejunostomy, laparoscopic surgery, magnetic resonance cholangiopancreatography

Introduction

Choledochal cysts (CCs) are uncommon congenital malformations of the biliary system, defined by segmental or diffuse dilatation of the intra- and/or extrahepatic bile ducts.[1] The Todani classification categorizes CCs, with type I being the most common (80–90% of cases).[2-4] Choledocholithiasis, the presence of stones within the common bile duct, often arises secondary to congenital biliary anomalies and may manifest as biliary colic, obstructive jaundice, or recurrent acute pancreatitis.[5, 6] Untreated CCs may lead to acute recurrent pancreatitis (ARP) through episodic obstruction at the pancreaticobiliary junction.[6] Recurrent pancreatitis in children poses substantial diagnostic complexity, necessitating evaluation for structural, genetic, metabolic, and immune-mediated etiologies.[7] Autoimmune pancreatitis (AIP) can mimic biliary causes, especially when imaging and antibody tests overlap.[8, 9]

We report a 7-year-old girl with a Todani type IC choledochal cyst complicated by choledocholithiasis, manifesting as recurrent pancreatitis and definitively treated via laparoscopic excision with Roux-en-Y hepaticojejunostomy. This case is unique because of the prolonged diagnostic course, autoimmune pancreatitis–like features, and delayed identification of a structural biliary cause in a resource-limited setting.

Case

A 7-year-old girl initially presented with acute postprandial pain in the right upper quadrant and epigastrium, associated with nausea, reduced appetite, and a low-grade fever, but without jaundice. Laboratory tests revealed marked pancreatic and hepatic enzyme elevations: amylase 233 U/L (reference 25–101 U/L), lipase 162.6 U/L (reference ~60 U/L), AST 536.2 U/L (reference <34 U/L), and ALT 410.4 U/L (reference <55 U/L). In this first episode, abdominal ultrasonography demonstrated acute pancreatitis accompanied by dilation of the pancreatic duct, proximal common bile duct, cystic duct, and intrahepatic biliary branches. Her symptoms resolved within a few days following conservative therapy. Three months later, she re-presented with similar abdominal pain and vomiting. Laboratory testing again showed elevation of hepatic and pancreatic markers: AST 295.6 U/L, ALT 460.9 U/L, Amylase 246.8 U/L, Lipase 162.4 U/L, and markedly elevated GGT 305.0 U/L (normal <33 U/L). Hepatitis B serology was non-reactive. She was diagnosed with acute recurrent pancreatitis (ARP) and improved with supportive care.

During the second year of illness, she developed her most severe biochemical flare, with Amylase 832 U/L and Lipase 1771 U/L (approximately 7–15× pediatric upper limits of normal), while bilirubin, alkaline phosphatase, and complete blood count were within pediatric reference ranges. Intermittent hair thinning was also reported. Because no structural abnormality had been detected previously despite multiple recurrences, AIP was considered. ANA IFA was positive at 1:100, fine-speckled; C3

was mildly elevated at 132.2 mg/dL (pediatric normal 90–180 mg/dL). A multislice computed tomography (MSCT) scan demonstrated features compatible with AIP. Given limited availability of autoimmune diagnostic assays in Bali, she was treated empirically with intravenous methylprednisolone 2 mg/kg/day, later transitioned to oral taper, along with weekly Methotrexate 10 mg/BSA. Symptoms initially improved, but after approximately three months she developed dyspepsia. Esophagogastroduodenoscopy revealed erosive fundal gastritis, prompting discontinuation of immunosuppressive therapy.

Shortly after cessation, abdominal pain recurred. Laboratory values again showed elevated pancreatic and hepatic markers: Amylase 433 U/L, Lipase 413 U/L, AST 229 U/L, and ALT 311 U/L. Because of an incomplete steroid response and continued episodes, further structural evaluation was pursued. High-resolution abdominal magnetic resonance imaging (MRI) was obtained and findings were consistent with a Todani type I-C choledochal cyst with upstream biliary stasis due to distal obstruction **(Figure 1A–B)**. These MRI findings established a clear anatomic cause, a choledochal cyst and choledocolithiasis, for her recurrent pancreaticobiliary obstruction.

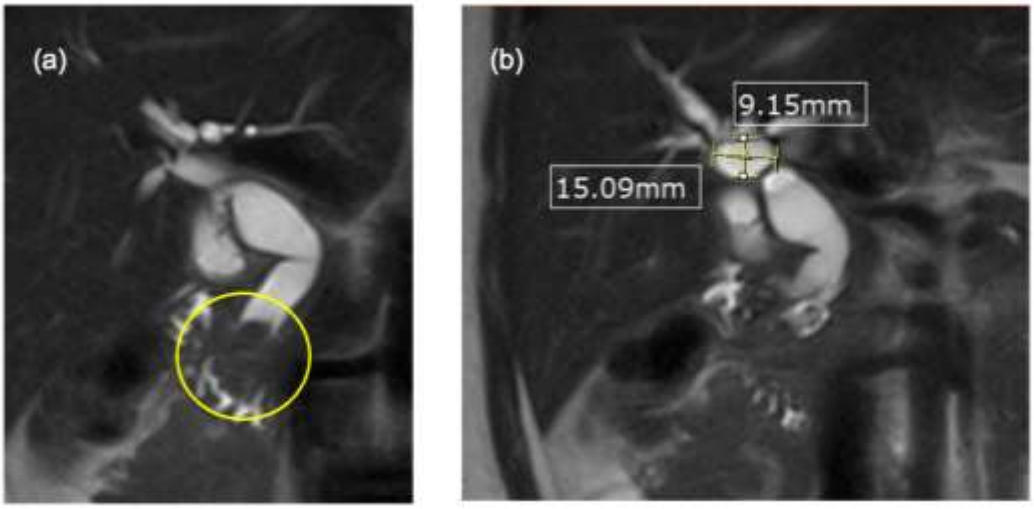

Figure 1. Contrast-enhanced abdominal MRI

a) Suspected choledocholithiasis on the distal common bile duct with dilated left and right intrahepatic duct, common hepaticus communis and the proximal common bile duct.

b) Fusiform dilatation of the common bile duct (CBD) with suspected intraductal sludge or calculi, consistent with a Todani Type IC choledochal cyst and associated choledocholithiasis

Patient was definitively treated via laparoscopic excision with Roux-en-Y hepaticojejunostomy by the pediatric surgery division. Intraoperative imaging **(Figure 2A–D)** provides a stepwise depiction of the surgical field. Reconstruction was performed with a Roux-en-Y hepaticojejunostomy and a side-to-side jejunojejunostomy, with tension-free anastomoses. A 14 Fr subhepatic drain was placed. Postoperatively, she remained hemodynamically stable in the PICU, without bile leakage or infectious complications. Enteral feeding resumed on postoperative day four. Histopathology confirmed a choledochal cyst without dysplasia or malignancy **(Figure 3)**. She was discharged on postoperative day seven and has remained asymptomatic with normal laboratory results on follow-up.

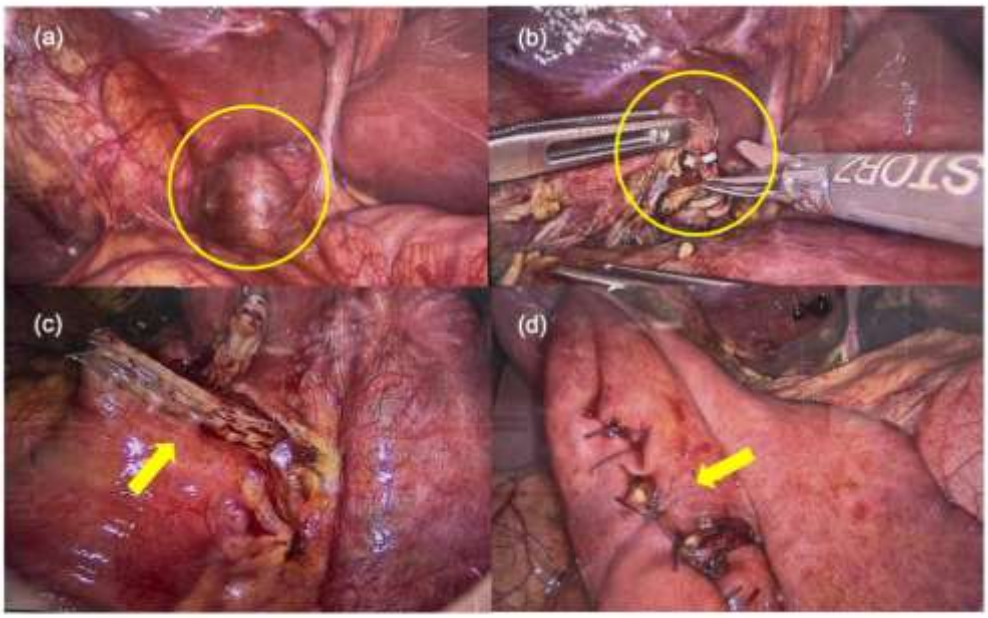

Figure 2. Laparascopic Roux-en-Y hepaticojejunostomy
a) Fusiform choledochal cyst
b) Ligated choledochal cyst
c) Hepatico-jejunostomy
d) Jejunum-jejunostomy

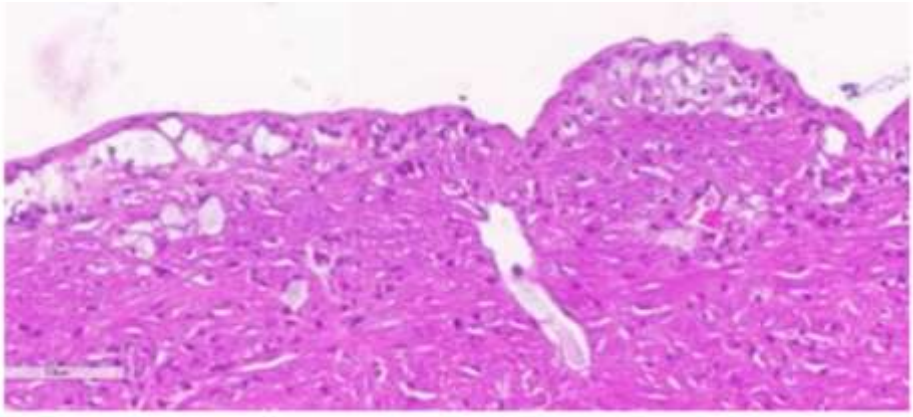

Figure 3. Histomorphology result showed columnar epithelium with no sign of malignancy

Discussion

Unlike most reported cases, this patient demonstrated overlapping clinical and radiologic features suggestive of autoimmune pancreatitis, leading to temporary immunosuppressive treatment before the definitive biliary etiology was identified.

The Role of Choledocal Cysts in Choledocholithiasis and Recurrent Pancreatitis

Choledocholithiasis is uncommon in children compared to adults.[5, 6] When clinical signs are present, they frequently resemble other gastrointestinal conditions. Diagnostic confirmation of gallstones and choledocholithiasis primarily depends on imaging, with transabdominal ultrasonography serving as the initial modality of choice. Nevertheless, ultrasound has limited sensitivity for detecting stones within the common bile duct, especially when ductal dilatation is absent. In such cases, advanced imaging techniques such as magnetic resonance cholangiopancreatography (MRCP) or contrast-enhanced computed tomography (CT) are often necessary to better define the biliary anatomy.[7]

The presence of gallstones or biliary sludge in the common bile duct may intermittently obstruct the pancreaticobiliary junction, leading to retrograde bile flow, premature activation of pancreatic enzymes, and subsequent pancreatic inflammation. In the pediatric population, recurrent inflammation without timely diagnosis and intervention may result in progressive pancreatic damage and chronic sequelae.[10, 11] Markers of biliary pancreatitis in children include significantly elevated liver transaminases (particularly ALT >150 U/L), hyperbilirubinemia, elevated gamma-glutamyl transferase (GGT), and pancreatic enzymes such as serum amylase and lipase.[5, 6] Poffenberger et al. also emphasized the importance of clinical findings such as right upper quadrant (RUQ) tenderness, biliary colic, and postprandial exacerbation of symptoms.[5]

Choledochal cysts (CCs) represent congenital malformations of the biliary tract, defined by cystic dilatation of the bile ducts. According to the Todani classification, type IC is marked by fusiform enlargement of the extrahepatic bile duct and is often linked to an anomalous pancreaticobiliary junction (APBJ). This anatomical variation allows pancreatic secretions to reflux into the biliary system, leading to chronic inflammation and injury of the biliary epithelium.[1, 2, 10, 11] Definitive diagnosis often requires advanced imaging, with MRCP and CT being especially useful for mapping the biliary anatomy and identifying complications.[10, 11]

Comprehensive assessment for choledochal cysts is crucial in pediatric patients presenting with choledocholithiasis, as these cysts represent a frequent underlying etiology in children. Failure to identify and treat an associated choledochal cyst can leave the disease unaddressed, predisposing to recurrent gallstone formation and

chronic pancreatitis. In the present case, a Type IC choledochal cyst (CC) was diagnosed based on imaging that demonstrated fusiform dilatation, suspected choledocholithiasis, and a history of recurrent pancreatitis. Surgical management involved cholecystectomy combined with laparoscopic Roux-en-Y hepaticojejunostomy to correct the biliary anomaly. Histopathological analysis revealed chronic inflammatory changes, consistent with long-standing biliary tract pathology.[1, 4]

Diagnostic Challenges and Possibility of Autoimmune Pancreatitis

Autoimmune pancreatitis (AIP) is a rare cause of chronic pancreatitis that can be difficult to diagnose in children due to its nonspecific symptoms and overlap with other pancreatic diseases. The initial consideration of autoimmune pancreatitis arose because the patient exhibited recurrent episodes of pancreatitis without a demonstrable structural etiology on early imaging, an indication in which AIP should be evaluated according to the International Consensus Diagnostic Criteria (ICDC), which synthesize parenchymal and ductal imaging, serology, other-organ involvement, histopathology, and response to steroids as the core diagnostic domains.[12] The MSCT demonstrated diffuse gland enlargement with a classic "sausage-shaped" pancreatic morphology, fulfilling an ICDC Level 1 parenchymal imaging criterion, thereby strengthening the preliminary suspicion of AIP.[13] Due to substantial diagnostic constraints in Bali, where IgG4 quantification, pancreas-specific autoantibodies, and OOI-related assays are not routinely available, the immunologic assessment was restricted to ANA immunofluorescence and ANA profile testing, representing an incomplete but pragmatic approach to fulfilling the serologic domain. This limitation significantly reduces diagnostic confidence and increases over-reliance on partial criteria and steroid trials.[12-14] Methotrexate was instituted as a steroid-sparing immunomodulator following an initial intravenous methylprednisolone course to permit outpatient continuation of immunosuppression and to limit cumulative glucocorticoid exposure.[14, 15] Methotrexate use in this patient (oral, weekly dosing as per institutional practice) therefore represented a pragmatic approach to maintain disease control in the outpatient setting. However, Methotrexate use in this case should not be interpreted as standard pediatric practice but rather as a context-driven decision in a resource-limited setting.

Nonetheless, AIP was ultimately excluded when the patient's symptoms recurred during and after steroid tapering, and her pancreatitis biomarkers did not exhibit the expected biochemical resolution, a pattern discordant with the rapid and consistent steroid-responsive trajectory documented in both adult and pediatric AIP cohorts.[16-18] These considerations prompted renewed etiologic investigation. MRI evaluation was performed in consultation with a pediatric radiology subspecialist subsequently revealed choledocholithiasis and a Todani type IC choledochal cyst, providing a

definitive structural cause for the patient's recurrent pancreatitis and replacing the earlier working diagnosis of AIP.

Timely Diagnosis and Surgical Intervention

The mainstay of treatment for choledochal cysts, particularly when complicated by choledocholithiasis and pancreatitis, is complete cyst excision and biliary-enteric reconstruction. Roux-en-Y hepaticojejunostomy is the preferred surgical technique due to its durability and reduced risk of complications such as bile reflux and cholangitis compared to hepaticoduodenostomy.[19, 20] The laparoscopic approach, although needed advance skill and experience, offers advantages such as reduced surgical trauma, faster recovery, and improved cosmesis and has become standard in specialized pediatric centers.[21]

In this patient, laparoscopic Roux-en-Y hepaticojejunostomy with complete cyst and gallbladder excision was performed. Consistent with pediatric recommendations, postoperative evaluation following choledochal cyst excision requires longitudinal monitoring to detect late complications such as anastomotic stricture, recurrent ductal dilatation, cholangitis, or hepatic fibrosis.[22] Current pediatric series and guidelines advocate structured follow-up comprising clinical evaluation and liver function tests at 1, 3, and 6 months postoperatively, supplemented by abdominal ultrasonography or MRCP during the first postoperative year and annually thereafter, or more frequently if symptoms or biochemical abnormalities emerge.[23, 24] In this patient, an early outpatient review on postoperative day 14 demonstrated complete clinical resolution and normalized laboratory parameters.

Conclusion

This case emphasizes the diagnostic challenges of pediatric biliary pancreatitis in children, particularly when autoimmune markers and imaging results yield borderline findings. Structural biliary disease should remain a diagnostic priority in pediatric recurrent pancreatitis, even in the presence of autoimmune-like features. Given the often nonspecific clinical manifestation, a multidisciplinary and comprehensive evaluation is essential through early collaboration among pediatric gastroenterology, radiology, surgery, and other relevant specialties to facilitate timely diagnosis and optimize clinical outcomes.

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This case report has received consent for publication from the subject and parents.

Conflict of Interest

This case report has no conflict of interest

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