Journal of Population Therapeutics & Clinical Pharmacology

RESEARCH ARTICLE
DOI: 10.53555/vj2jvw78

A HIDDEN ANOMALY: DELAYED PRESENTATION OF A GIANT CHOLEDOCHAL CYST IN ADULTHOOD

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Abstract:
Choledochal cyst, a rare congenital anomaly involving bile duct typically present as cyst. There are 5 subtypes of choledochal cyst are classified by Todani, in which type IV is more commonly seen and type I and IV have much greater risk of malignancy then any other types which are commonly benign in nature. Not many cases of choledochal cyst are reported in literature. He we are reporting a case of thirty-five-Year-old lady presented with a large upper abdominal mass but without jaundice and on evaluation it was found out be a giant choledochal cyst of typeIVa and patient underwent dilated choledochal cyst excision with Roux-en-Y-hepaticojejunostomy and cholecystectomy at the junction of right and left hepatic duct. Keywords: Bile duct cyst, Choledochal cyst, giant choledochal cyst.

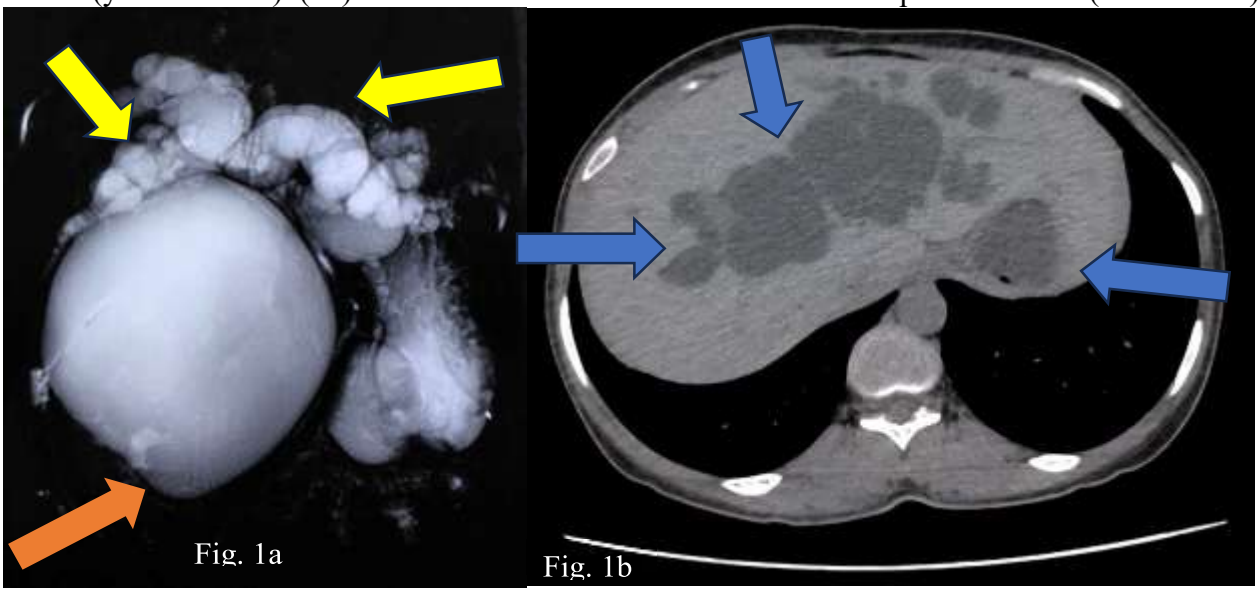
Introduction:
Choledochal cysts are rare congenital bile duct anomalies that primarily affect young women and children, particularly those of Asian descent, with a higher prevalence in Japan and Taiwan. The first classification system for these cysts was introduced by Alonso-Lej et al. in 1959, dividing them into three types (I-III). In 1977, Todani and colleagues revised this system by adding two additional types (IV and V) and further subdividing Type I into three distinct subcategories [1,2]. A choledochal cyst exceeding 10 cm in size is classified as a giant cyst. These cases are exceptionally rare, with only a limited number documented in medical literature. Due to their uncommon occurrence, initial diagnosis can be challenging, and their large size presents additional difficulties in surgical management, as complete excision is the preferred treatment approach [2]. Approximately 80% of choledochal cysts are diagnosed in infants and young children within the first decade of life. In Western countries, choledochal cysts are rare, with an incidence of 1 in 100,000 to 1 in 150,000. However, they are more commonly observed in Asian populations, where the prevalence Is significantly higher i.e. as high as 1:1000[3]. Additionally, these cysts are found approximately four times more frequently in females than in males [4]. Type I is the most common form of choledochal cysts, representing 80% of cases and primarily affecting infants and young children [5]. Although choledochal cysts are more commonly diagnosed in children, around 25% of cases manifest later in adulthood [6]. Anomalous pancreaticobiliary duct union (APBDU), in which the common bile duct and pancreatic duct merge outside the duodenum, is present in 30% to 70% of cases and may play a role in the development of choledochal cysts [7,8]. The common channel theory is the most widely

accepted explanation for the etiology of choledochal cysts. It suggests that an abnormally long common channel, along with an ineffective Sphincter of Oddi at the junction of the common bile duct and pancreatic duct, contributes to their development [9,10]. While typical symptoms include abdominal pain, abdominal mass and jaundice our case features an adult 35year old female with abdominal pain, abdominal mass but without jaundice.

Case History:

Thirty-Five-year-old female presented with right upper abdomen pain and a noticeable lump in the same region for past 1year. The pain was insidious in onset, severe, intermittent, and colicky in nature, associated with body aches and weakness. It was exacerbated by food intake, squatting, and weightlifting but relieved by analgesics. The lump first noticed a year ago was progressive but not associated with jaundice, fever or diurnal variations. Abdominal examination revealed a 12 cmX10 cm oval lump in the right hypochondriac region which was mildly tender, firm in consistency and moved with respiration. Laboratory investigations showed elevated Alkaline phosphatase-489U/L, SGPT-76U/L, SGOT-61.20U/L levels and direct bilirubin levels-0.53mg/dl. MRCP(Figure 1) revealed a cystic dilation of common bile duct, of 9.1X 11.3X13.5 cm (APXTRXCC) with no obvious opening appreciated with extension into the intrahepatic bile ducts consistent with a giant choledochal cyst type IVa and an APBDU. She was planned for open cholecystectomy and excision of the choledochal cyst with Roux-en-Y hepaticojejunostomy under general anaesthesia was done, Liver transplant wasn't done as there was no signs of Liver Failure. Intraoperatively (Figure 2), the cyst was densely adherent to surrounding structures like duodenum and pancreas. It was a large cyst 10X12cm in size, cyst wall was incised and greenish bilious fluid was drained out following which cyst was excised in toto. Postoperatively (Figure 3) period was uneventful, the drain was removed on the third day, and the patient was discharged on the seventh postoperative day. Histopathological examination (Figure 3) of the excised cystic tissue revealed dense fibrous stroma, smooth muscle fibres and chronic inflammatory infiltrates comprising lymphocytes and plasma cells consistent with a choledochal cyst. Sections of the gallbladder (GB) showed chronic cholecystitis.

Figure 1: (1a) MRCP shows a choledochal cyst(Orange arrow) with dilated intrahepatic ducts(yellow arrow). (1b) HRCT transverse section confirms intrahepatic dilations (blue arrows).



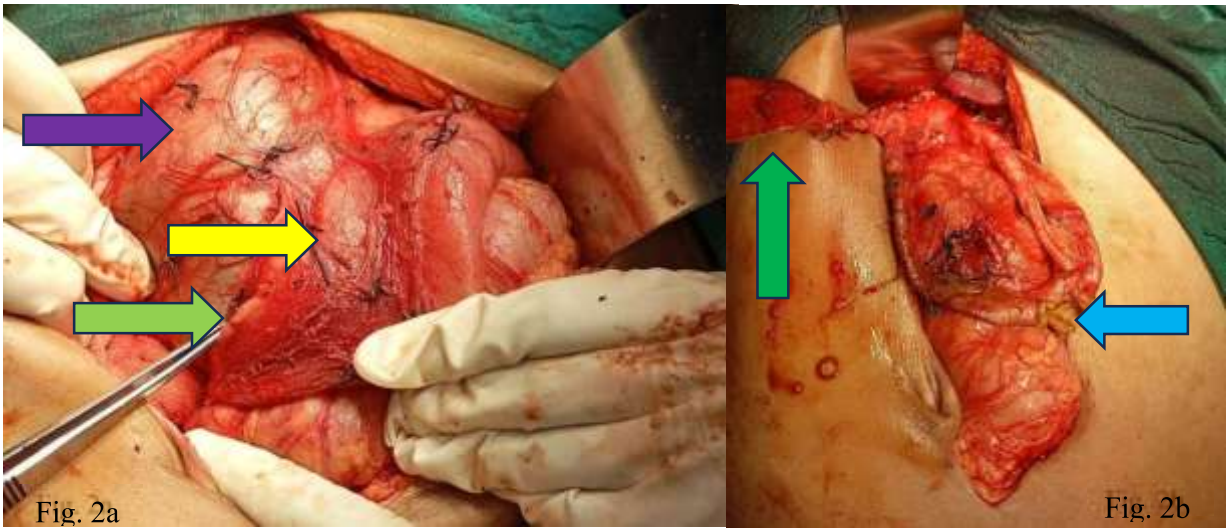


Figure 2: (2a) Operative image reveals a giant choledochal cyst (purple arrow) behind the duodenum (yellow arrow) and small tissue of pancreas at forcep tip(green arrow). (2b) Shows the cyst's cut end (Blue arrow) at the pancreas's superior border and gall bladder superiorly(dark green arrow).

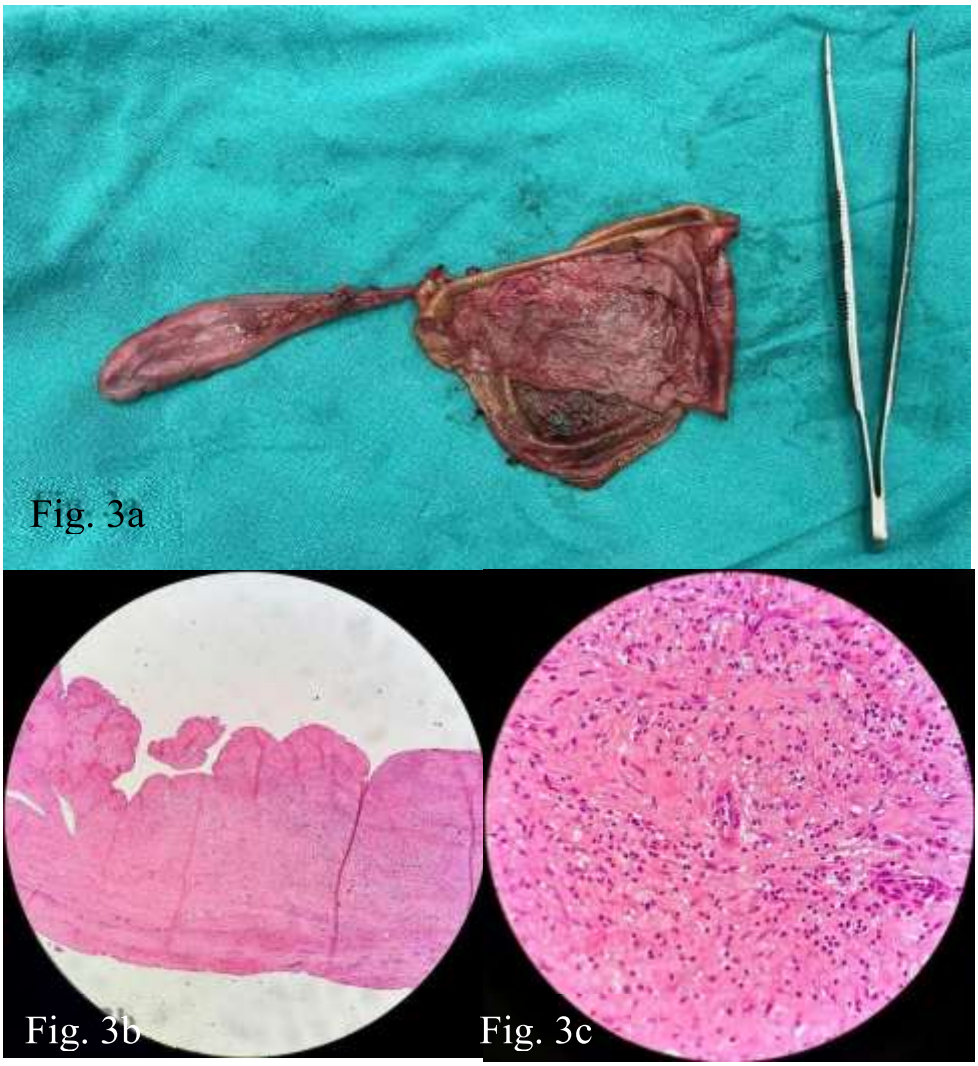


Figure 3: (3a) Postoperative image of the resected cyst and gallbladder. (3b) Histopathology (10x) shows inflamed cystic walls. (3c) Microscopy (40x) reveals smooth muscle fibers, fibroblasts, and infiltration.

Discussion:

The incidence of bile duct cysts has increased in recent years, with more than half of the reported cases originating from Japan. A study from Finland estimates that the incidence has risen from 1 in 128,000 to 1 in 38,000 over the past four decades. Although this condition was previously more common in children, recent studies indicate that it now occurs at similar rates in both adults and children [11]. Despite multiple proposed theories, the exact etiology of choledochal cysts remains uncertain. The most widely accepted explanation is the theory introduced by Babbitt in 1969 [12]. Also, studies by Miyano et al have shown approximately 90-100% anomalous junction in choledochal cyst patients in 1977 [14]. In our case also anomalous pancreatico-biliary junction has noted in MRCP.

Regarding clinical presentation there was no consensus exists in the online literature of any commonest manifestation [15]. The clinical manifestations in recent research shows that children (ages 1–18 years) most commonly presents with symptoms like nausea, vomiting, and abdominal pain. Whereas in adults are nausea, vomiting, jaundice abdominal pain and palpable mass [16,17]. Our patient also showed one or two of the above symptoms. Early diagnosis and treatment of choledochal cysts are essential for ensuring optimal outcomes.

However, delayed diagnosis may occur due to factors such as limited healthcare resources low parental health literacy, and minimally symptomatic cases [12]. In most cases ultrasound is considered as first radio imaging modality. To investigate the condition, specific imaging techniques such as Ultrasonography, Computed Tomography, have proven effective in detecting bile duct dilations and gallstones. However, MRCP is currently considered the gold standard and the primary choice for diagnostic evaluation [11]. Our patient underwent MRCP where diagnosis of giant choledochal cyst of type IVa was confirmed. Though, there was a delay in diagnosis but the major contributary factor here was patient negligence lack of education and resources. Giant choledochal cyst have been reported in literature. When comparing the dimensions of choledochal cysts reported in previous studies with our case, it is evident that there is a wide variability in cyst size presentation. Harikrishnan et al. reported a massive extrahepatic bile duct dilatation measuring 23 × 15 cm, indicating significant cystic enlargement up to the intrapancreatic portion. Similarly, Atwine et al. documented an even larger cystic mass measuring 30 × 25 × 20 cm, containing more than 2 liters of bilious fluid, emphasizing the potential for extreme cystic expansion. In contrast, Budipramana et al. described a relatively smaller, fusiform cyst measuring 14 × 14 × 1 cm. Compared to these cases, our patient presented with a cystic dilation of the common bile duct measuring 9.1 × 11.3 × 13.5 cm (AP × TR × CC) on MRCP, which, while substantial, remains within the lower to mid-range of sizes described in the literature. This comparison underscores the considerable heterogeneity in choledochal cyst dimensions, which may influence both clinical presentation and surgical management strategies[17,18,21].

The Optimal treatment approach of choledochal cysts involves initial management of complications, followed by definitive surgical excision and reconstruction. The standard treatment for bile duct cysts is surgical, with specific approaches based on classification type. Type I, II, and IV cysts typically require resection due to the risk of neoplastic transformation. Type I and V cysts are completely resected, along with cholecystectomy, followed by biliary reconstruction using Roux-en-Y hepaticojejunostomy. For type IVa cysts, partial hepatectomy is also necessary [15]. Currently, the most widely accepted surgical approach for type IVa is complete excision of the cyst with biliary-enteric drainage restored through either hepatic-duodenostomy or Roux-en-Y hepaticojejunostomy done [20]. A laparoscopic approach is currently being investigated as a minimally invasive surgical option for managing choledochal cysts in children [19]. The Malignant potential of choledochal cyst increase with age particularly in type-I and IV cyst [21]. Our patient was also a high-risk case but the Histopathological Examination showed no signs of malignancy. Therefore, the histopathological evaluation as well as thorough pre-operative evaluation is a must.

In conclusion, Giant Choledochal cyst beyond childhood a very rare diagnosis adding the diagnostic and surgical challenge during its treatment course. There are very few giant choledochal cyst is reported in literature and varied presentation of symptoms add to its scarcity. Commonly used

treatment protocol is cyst excision and Roux-en-Y hepaticojejunostomy due to the risk of malignancy in future. Advancements in healthcare and greater access to high-precision medical imaging have facilitated early diagnosis allowing timely therapeutic interventions and reducing the risk of complications. So, keeping in mind its incidence; non- specific clinical features and diagnostic rarity we as clinicians should make use of the available diagnostic tools to our utmost advantage and confirm our diagnosis.

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List of Abbreviations:

Abbreviation	Definition
SGPT	Serum glutamic pyruvic transaminase
APBDU	Anomalous pancreaticobiliary duct union
SGOT	serum glutamic-oxaloacetic transaminase
MRCP	Magnetic Resonance Cholangiopancreatography
HRCT	High-Resolution Computed Tomography
Fig	Figure