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Choledochal cyst mimicking as choledocholithiasis: A case report and review of literature

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Abstract

Choledochal cysts are rare congenital anomalies of the biliary system, mostly diagnosed during childhood. In adults, a lower incidence and symptom overlap with more common biliary conditions may hinder the diagnosis. This case study presents a 50-year-old female patient who presented with abdominal pain and multiple gallstones on ultrasonography which also showed a dilation of the common bile duct, presumably left by a stone that had already passed. However, the dilation still existed 3 days later, which raised suspicion of a choledochal cyst. Magnetic resonance cholangiopancreatography was performed, and following surgical consultation, the patient underwent a successful cyst excision by Roux-en-Y hepaticojejunostomy with a smooth recovery leading to hospital discharge. This case highlights the importance of considering choledochal cysts as one possible diagnosis in adults with biliary symptoms and the significance of the more specific imaging modalities such as magnetic resonance cholangiopancreatography.

Keywords

Choledochal cyst, hepaticojejunostomy, case report

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Introduction

Choledochal cysts are rare congenital cystic dilatation of the biliary tract that can present clinically with abdominal pain, jaundice and pancreatitis. Choledochal cysts, if untreated, can lead to complications such as cholelithiasis, cholangitis, and neoplastic transformation.

The precise pathogenesis underlying the development of choledochal cysts remains to be fully elucidated, But, the predominant theory suggests an abnormal pancreaticobiliary junction that allows the reflux of proteolytic pancreatic enzymes into the biliary tree, inciting mural inflammation, erosion, and consequent focal or diffuse dilatation of the extrahepatic biliary system.[1–3] Another commonly held postulation is Babbitt's theory. This states that these cysts are due to an abnormal pancreaticobiliary duct junction (APBDJ). An APBDJ means that the pancreatic duct merges with the bile duct 1–2 cm proximally to the sphincter of oddi.[4] Pancreatic juice reflux and increased pressure into the Common Bile Duct (CBD) can result in dilatation.[3]

The clinical presentation of a choledochal cyst can vary. This patient had a clinical presentation consisting of right hypochondrium pain with vomiting. Vital signs were normal, and the patient did not appear jaundiced. This differs from the classic triad of abdominal pain, abdominal mass, and jaundice which is not seen commonly (<20% cases).[5–7] Furthermore, only 25% of cases in adults show 2/3 symptoms of this triad.[8] Other symptoms may be nonspecific such as nausea/vomiting and complications may be part of the presentation. These include cholelithiasis, cholangitis, pancreatitis, and malignancy.[9,10]

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While right upper quadrant abdominal pain is common, adults may also have signs and symptoms that can mimic other hepatobiliary pathology such as choledocholithiasis or cholangitis, which can present a diagnostic dilemma. This report highlights the importance of considering alternative diagnoses when initial assessment is equivocal for choledocholithiasis. Accurate and prompt diagnosis allows for early surgical management of choledochal cysts to prevent complications such as cholangitis, biliary cirrhosis, portal hypertension, and malignancy.

Case report

A 50-year-old female presented with severe right upper quadrant colic pain associated with biliary vomiting for the past 2–3 days. The patient had a history of intense dieting and reported minimal to no water consumption as a habit for many years.

On arrival, she was afebrile, normotensive (120/80 mmHg) with a Heart rate of 90 beats/min and respiratory rate of 16/min. Her oxygen saturation was 96% on room air. The physical examination was notable for right hypochondrium tenderness. The patient was conscious but not fully oriented to time, place, and person. The rest of the systemic examination was unremarkable, with normal heart sounds and bilateral air entry into the lungs. She had no pallor, icterus, cyanosis, clubbing, edema, or lymphadenopathy.

Laboratory workup showed a normal CBC (Hb—11 gm/dl, TLC-6600/cumm, Platelet Count- 1.71 lakh/cumm), Liver Function Test, and Serum Creatinine (0.8). Serum Amylase (60 U/L) and Lipase (17 U/L) were also within normal limits. As well, alkaline phosphatase was within the normal reference range. Viral markers were nonreactive. Abdominal ultrasound (Figure 1) revealed multiple cholelithiasis with a mildly distended gallbladder and a 12-mm calculus impacted at the neck of the gallbladder as visible in Figure 1. Common bile duct showed mild fusiform dilation measuring 11–12 mm with prominent hepatic ducts. Bilateral nephrolithiasis and a few seedling fibroids in the uterus were also visualized.

Initially, it was thought that the dilated bile duct was due to choledocholithiasis, but since no stones were seen in the common bile duct, it was assumed that the stone had descended further into the biliary tract or the gut. Magnetic Resonance Cholangiopancreatography (MRCP) was performed, which was significant for multiple cholelithiasis, dilated common bile duct (11 mm) and ruled out choledocholithiasis (Figure 2). After 3 days of observation, the dilation of common bile duct persisted. This led to the diagnosis of type 1 choledochal cyst as the cause of dilated common bile duct.

GI surgery was consulted, and the patient was advised for cholecystectomy and choledochal cyst excision. Choledochal cyst excision by Roux-En-Y hepaticojejunostomy was performed. The patient tolerated the procedure well. Gallbladder and choledochal cyst specimens were sent for pathological examination. Gallbladder measuring 8 cm × 5 cm × 4 cm, had a congested external wall and wall thickness of 0.4 cm. The cut surface revealed ulcerated mucosa and multiple calculi. Microscopic examination of the choledochal cyst, a

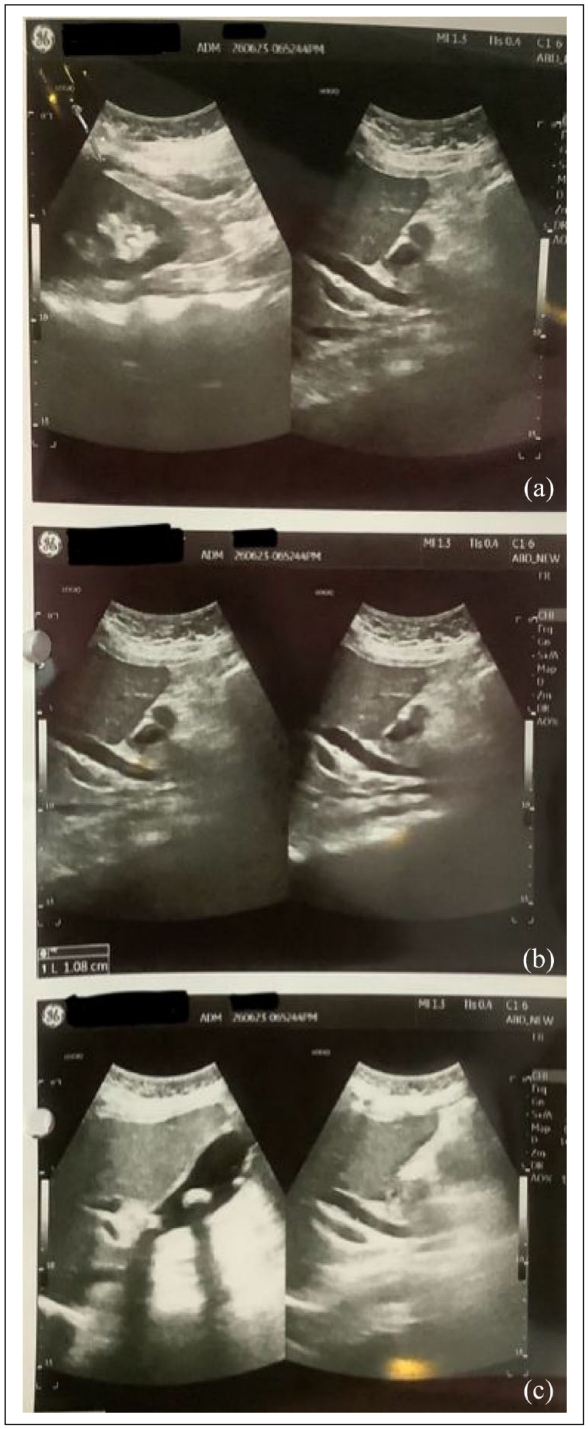

Figure 1. Ultrasonography (USG) scan (a and b) results showing mildly dilated CBD with prominent hepatic ducts. There is no calculus in the visualized part of CBD. USG scan (c) depicting multiple calculi in gall bladder. CBD not visualized.

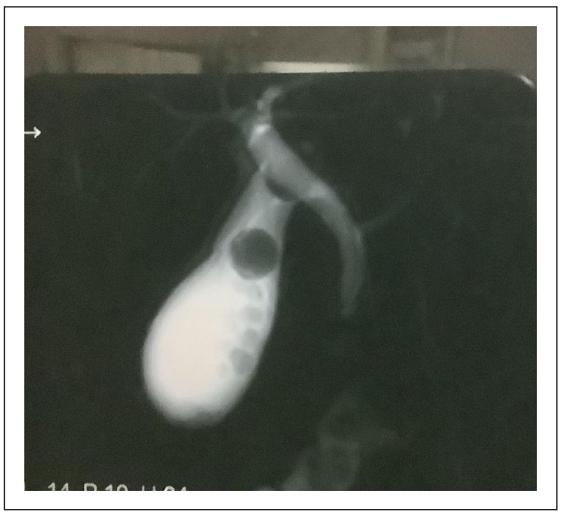

Figure 2. MRCP scan depicting multiple gallstones, dilated common bile duct (11 mm).

grey-white tissue piece measuring 2.5 cm × 0.3 cm, showed wall inflammation.

The patient tolerated the procedure well. She was kept Nil-By-Mouth (NBM) initially and was moved to a liquid diet followed by a semisolid diet as she improved. The patient was able to pass stool and the drain was removed. Follow-up abdominal ultrasound performed after 7 days showed post-surgical mild pneumobilia and post-cholecystectomy status.

Discussion

Choledochal cysts are a very uncommon cystic dilatation of the biliary tract. They occur mostly in infants and children. In fact, about 80% of choledochal cysts are diagnosed in patients before the age of 10 years old.[11,12] Not only this but the incidence is greater in east Asian communities (1/13,000 in Japan vs 1/100,000 in western nations).[13,14] It is important to note that this presents much more frequently in females (4:1).[11]

The most widely used system to classify choledochal cysts was speculated by Todani and colleagues in 1977.[2] These classifications are from I to V, as shown in Figure 1 below, with type I being the most prevalent (80%–90%).[2,7,8,15,16] Type I cysts appear as an anechoic cystic lesion which communicates with the biliary tract. For this case, it was determined that this patient had a type I cyst as imaging, as discussed below, showed a fusiform extrahepatic dilatation of the CBD (11 mm).

Abdominal ultrasound is an appropriate initial imaging test to delineate a choledochal cyst and related pathology. Its sensitivity is about 71%–97%.[17] Endoscopic retrograde cholangiopancreatography (ERCP) is excellent for the diagnosis of choledochal cysts and other related conditions of the biliary tree. However, the ERCP is very invasive and has its associated complications.[13] On the other hand, Magnetic Resonance Cholangiopancreatography (MRCP) is mentioned as being as good as the ERCP and has the added benefit of not having complications and risks associated with ERCP.[18] A review from 2009 even stated that it can well delineate features of the biliary and pancreatic ducts. However, an ERCP may not achieve this as well as the MRCP in the context of a choledochal cyst as there might be tight structures in the distal part of the cyst.[13] CT scans are also said to be accurate for diagnosing cysts.[5] However, reports have mentioned that they have been missed and later detected on MRCP.[19] As mentioned, this patient had an initial U/S of the abdomen that showed a distended gallbladder and gallstones. The U/S was able to detect a dilatation of the CBD. As the Liver Function Test (LFT) was normal, an MRCP was later done and showed an 11 mm choledochal cyst (Figure 2).

As mentioned, choledochal cysts are benign but can have aggressive complications. This includes but is not limited to pancreatitis, cholangitis, biliary cirrhosis, and malignancy. The sequelae of pathogenesis may be because of its association with an anomalous pancreaticobiliary junction.[18] These cysts allow bile to reflux which can activate pancreatic enzymes leading to pancreatitis.[3] This could be suggested by a study where 57% of patients with choledochal cysts had anomalous pancreaticobiliary union on ERCP. Of this subset of patients, all developed pancreatitis.[20] Also, cholangitis is a common complication of this condition and can also occur after surgical management.[18] Biliary cirrhosis is another important complication to mention. Biopsies taken during surgery showed this to be present in 40%–50% of cases.[21,22] Fortunately, this patient did not have these complications which were justified by the normal vital signs, lipase/amylase, CBC, LFTs, and renal function. As well there was no jaundice which was reassuring.

For a type 1 cyst, both hepaticoduodenostomy and Roux-en-Y hepaticojejunostomy (RYHJ) have been reported in the literature. However, RYHJ is preferred over the former procedure.[1] A hepaticoduodenostomy has much more postoperative reflux and gastritis as shown in a meta-analysis.[23] As well, this procedure is associated with higher rates of gastric/biliary cancer as compared to RYHJ.[24,25] Thus, a RYHJ was preferred and was the surgery of choice in this case after the cyst was resected. As well, due to the coexisting cholelithiasis, the patient also underwent a cholecystectomy.

Postoperative complications are seen more in adults than in children.[6,26–28] Later complications (>30 days) include cancer, cholangitis, anastomotic stricture, and cirrhosis. These complications can occur in up to 40% of adu lts.[6,15,24,27,29–31] The 5-year survival rate is over 90% for patients who underwent a resection and thus the prognosis is good.[32–34] The risk for developing cholangiocarcinoma is 10% in patients with choledochal cysts and this risk does not reduce to population baseline levels post-surgery.[9,10]

Conclusion

This case report highlights the uncommon occurrence of choledochal cysts in adults and the significance of considering choledochal cysts as a differential diagnosis in adults presenting with biliary symptoms. Advanced imaging techniques, such as MRCP, played an important role for accurate and timely diagnosis. Our case also contributes to the existing literature by reiterating the importance of the Todani classification system in categorizing choledochal cysts and guiding appropriate therapeutic approaches. Early identification and surgical intervention, as demonstrated in our case, play a pivotal role in preventing potential complications associated with choledochal cysts.

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Author contributions

MPS was responsible for writing the introduction. AMF and HRB were responsible for writing the discussion. MBI wrote the case report. MWA was responsible for writing the abstract and conclusion. RDM overviewed the article, made edits, literature search, and was responsible for supervision. All authors approved the final article and agreed to be equally accountable for this work.

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Ethics approval

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Informed consent

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