ORIGINAL RESEARCH PAPER

Radio-Diagnosis

A RARE CASE OF ISOLATED RIGHT POSTERIOR BILE DUCT CHOLEDOCHAL CYST IN AN ADULT PATIENT

KEY WORDS: Choledochal Cysts, Bile Duct Cysts, Biliary Anatomy

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ABSTRACT

Choledochal cyst are rare congenital intrahepatic, extrahepatic or both biliary tree dilatations. It is seen in significant frequency in childhood, with a higher prevalence in females. Their presentation in adults is rare and repeatedly associated with complications. Choledochal cyst arising from a single segmental intrahepatic bile duct is even rarer. Herein, we present a rare case of a 56-year-old male patient with a choledochal cyst arising from the right posterior bile duct in the background of chronic cholecystitis. Magnetic Resonance Cholangiopancreaticography (MRCP) showed a multilobulated cystic lesion in the liver which was carefully assessed for its origin and reported to be arising from the right posterior bile duct. The surgery was done where an intraoperative cholangiogram was performed which confirmed the imaging findings. Isolated intrahepatic biliary cysts should also be considered in the differential diagnosis when intrahepatic cystic lesions are encountered. Detailed radiological evaluation and multidisciplinary discussion become crucial in such cases.

INTRODUCTION

Choledochal cysts are dilated portions of the biliary tract. There are five types with a few recognised subtypes according to the Modified Todani classification: Type I involves dilatation of the common bile duct; Type II is a diverticulum of the common bile duct; Type III (choledochocele) involves the intraduodenal portion of the common bile duct; Type IVa consists of multiple intrahepatic and extrahepatic cysts, while Type IVb involves multiple extrahepatic cysts only; and Type V (Caroli disease) consists of multiple intrahepatic cysts. The incidence in western population is 1 in 1,00,000 -1,50,000 live births and Asian populations is 1 in 1000 to 13000 [1]. There is predominance in the female population, which is more common in the first decade. Adults more likely present with biliary or infectious problems and are more likely to progress to choledocholithiasis and malignant transformation [2].

Intrahepatic choledochal cysts arising from a single segmental bile duct are extremely rare. Diagnosis in an adult is even rarer. We herein report a case of a choledochal cyst arising from a segmental branch of the right posterior bile duct in an adult which was surgically excised followed by closure of the communication. Given its communication with a single intrahepatic segmental duct and absence of diffuse biliary involvement, this case most closely resembles a focal or segmental form of Type V (Caroli disease), though the focal and isolated nature distinguishes it from classic Caroli disease, which typically involves multiple intrahepatic ducts diffusely.

Case Report

A 56-year-old male presented with complaints of intermittent abdominal pain with fever and jaundice. No previous imaging studies were available for review, and detailed history did not elicit any prior episodes of cholangitis. Ultrasonography done elsewhere showed an irregular shaped cystic lesion at the porta with few internal echoes and was reported as a hepatic cyst along with mild diffuse gall bladder wall edema with no calculus. The patient then presented to our hospital for further investigation and management. An MRCP was done on an outpatient basis which showed a multilobulated cystic lesion measuring 2.4 x 4.4 x 3.1 cm in the right lobe of the liver in the region of segment IVB/right posterior sector with dependent sludge and multiple calculi within [Figure 1]. Further careful assessment of the biliary anatomy revealed that the cystic lesion was communicating with the right posterior bile duct [Figure 2(a-f)]. MRI and MRCP were crucial in this evaluation, as they provide superior soft tissue contrast and direct multiplanar visualisation of biliary anatomy compared to CT, enabling precise delineation of the communication between the cystic lesion and the biliary tree. No other pancreatico-biliary anomaly was detected. The gall bladder appeared normal with no calculi within [Figure 3] and showed no obvious features of acute inflammation. No obvious communication of the gall bladder with the cystic lesion was seen to suggest contained perforation. Rest of the visualised organs did not show any significant abnormality.

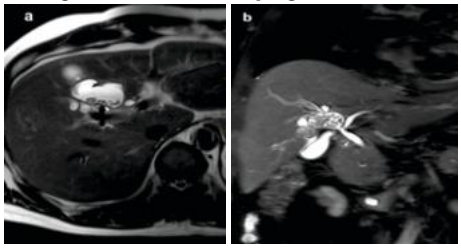

Figure 1: T2 Weighted Magnetic Resonance Images Showing a Lobulated Cystic Lesion in Segment IVB of Liver with Multiple Calculi Within.

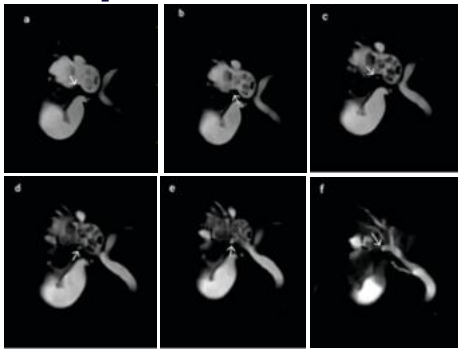

Figure 2(a-f): MRCP Images Showing Communication of the Cystic Lesion with the Right Posterior Bile Duct (Short White Arrows).

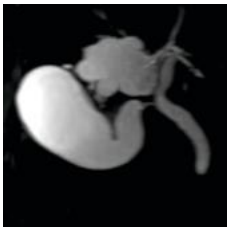

Figure 3: MRCP Image Showing Gallbladder Separate from the Lesion.

The patient was then admitted to the hospital for an exploratory laparotomy with cholecystectomy and SOS liver resection/hepaticojejunostomy. A post-contrast MRCP imaging was also done before the surgery to rule out any associated malignant alteration and did not reveal any abnormal enhancing soft tissue. The patient underwent an open cholecystectomy with excision of the biliary cyst. Perioperatively, the gall bladder was dilated containing sludge but no stones. Bile was aspirated and sent for culture which did not grow any organisms. Intraoperative cholangiogram was performed to delineate the biliary anatomy which confirmed the communication of the exophytic biliary cyst with the right posterior bile duct. The biliary cyst was then excised and was found to contain bile with stones and sludge.

Histopathology reports revealed that the biliary cyst had a fibrous wall with acute on chronic inflammatory infiltrates with no signs of dysplasia or malignancy. The excised specimen of the gallbladder showed chronic lympho-plasmacytic infiltrate throughout the wall with no specific signs of malignancy. The stones retrieved from the biliary cyst were sent for analysis and were found to be pigment stones, consistent with a biliary origin.

Post-operatively, ultrasound of the abdomen was performed and showed a small localised collection in the operative bed. The post-operative period was otherwise uneventful. The patient was discharged with a diagnosis of choledochal cyst with chronic acalculous cholecystitis.

DISCUSSION

Choledochal cysts are rare congenital dilatations of the biliary tract. The exact etiology remains uncertain; however, various theories have been proposed. The most accepted one is the proposal by Babbitt in 1969 [3], which states that choledochal cysts are caused by an anomalous junction of the pancreatic and the choledochal duct outside the duodenal wall, proximal to Oddi's sphincter during the embryonic period; with a formation of a common channel that results in reflux of the pancreatic secretions into the bile duct, and activation of the pancreatic enzymes and deconjugation of bile acids [4,5,6]. The combination of activation of the pancreatic enzymes and deconjugated bile acids in the biliary drainage system generates increased intraductal pressure and inflammation, leading to dilatation with cyst formation [5,7].

There are five types with a few recognised subtypes according to the Modified Todani classification. The various subtypes have been classified on the basis of anomalous unions (Komi's classification) and their anatomical locations (Todani's classification) [8]. Type I is the most common type, accounting for approximately 80–90% of cases. A rare Type VI choledochal cyst also exists, where there is isolated dilatation originating from the cystic duct [9]. The present case, involving a single segmental intrahepatic bile duct cyst communicating with the right posterior duct, represents a very unusual variant. Truly isolated segmental intrahepatic choledochal cysts communicating with a single sectoral duct in adults have been reported only in isolated case reports in the literature [7,8], underscoring their extreme rarity.

Choledochal cysts typically presents in childhood but about 25 % can also present in adulthood [10]. Symptoms vary however they can often be asymptomatic. Most common presentation in children include abdominal pain or discomfort, abdominal mass and jaundice. In adults, the clinical manifestations are usually non-specific and it is commonly associated with an increased rate of hepatobiliary pathology such as cholangitis, stone formation, cyst rupture, secondary biliary cirrhosis, obstructive jaundice and malignancy (cholangiocarcinoma) [11].

The most important imaging modalities helping in establishing the diagnosis are Multislice Computer tomography (MSCT), Magnetic resonance imaging (MRI), or Magnetic resonance cholangiopancreaticography (MRCP). MRI and MRCP are considered superior to CT for biliary evaluation, as they offer detailed non-invasive visualisation of the biliary ductal anatomy, cyst communication, and soft tissue characteristics without radiation exposure. However, often the preliminary diagnosis is made on ultrasonography.

With regards to treatment and management of choledochal cysts, it includes total cyst excision, cholecystectomy and Roux-en-Y hepaticojejunostomy depending on the variant [12].

An important differential diagnosis to consider in cases presenting with intrahepatic cystic lesions containing calculi is hepatolithiasis (primary intrahepatic stones). Hepatolithiasis can occur without cholelithiasis and may present as an intrahepatic cystic dilatation with stones, mimicking a choledochal cyst. In the present case, several features favour the diagnosis of a congenital choledochal cyst over hepatolithiasis: the cystic nature of the lesion with a distinct wall, the absence of a history of recurrent cholangitis, negative bile culture, and histopathology showing a fibrous-walled cyst without features typical of chronic stone disease such as biliary epithelial hyperplasia or extensive fibrosis. However, the MRCP appearances and the presence of acute on chronic inflammatory infiltrates on histopathology do acknowledge an overlap with hepatolithiasis, which cannot be entirely excluded. The stones were sent for biochemical analysis to aid in this distinction. This distinction is clinically relevant, as hepatolithiasis may not require the same surgical approach and may be associated with a higher recurrence risk. Choledochal cysts can be misinterpreted as other cystic lesions of the liver, and hepatolithiasis must be specifically included in the differential diagnosis when intrahepatic cystic lesions with calculi are encountered.

CONCLUSION

Choledochal cyst is a congenital disease, and the present case is unusual as the patient was asymptomatic until 56 years of age. Isolated exophytic cystic dilatations of the intrahepatic biliary ducts are extremely rare in adults, with only isolated case reports in the literature [7,8]. Preoperative diagnosis can be challenging and requires detailed radiological evaluation, particularly MRI and MRCP, and multidisciplinary discussion. The differential diagnosis of such lesions should always include hepatolithiasis, which may have overlapping imaging features. Surgeons and radiologists alike should recognise the possibility of atypical variations in choledochal cysts that do not fit within standard classifications. Stone analysis and thorough histopathological examination are essential post-operatively to aid in the definitive diagnosis.

Conflict of Interest

Authors declare no conflict of interest for this article.

Acknowledgment

None.

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