Choledochal cyst of the cystic duct: Report of imaging findings in three cases and review of literature

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Abstract

The choledochal cysts, which refer to the cystic dilatation of the biliary duct, are rare lesions generally seen in children. Choledochal cyst of the cystic duct is an uncommon entity. Often, it is associated with the choledochal cyst of the rest of the biliary tree. Isolated cystic duct choledochal cyst is quite rare. Most of these lesions have been recognized only on surgery. Modern cross-sectional imaging methods have facilitated preoperative recognition and characterization of cystic duct cyst enabling formulation of appropriate management strategy. We report the imaging findings in three cases of choledochal cyst involving the cystic duct. All these cases were correctly diagnosed preoperatively. A review of cases reported in the literature and the role of imaging in guiding the management are also presented.

Keywords: Choledochal cyst; computed tomography; cystic duct cyst; magnetic resonance cholangiopancreatography

Introduction

Choledochal cysts of the biliary tract are well-known entities. However, choledochal cyst of the cystic duct is exceedingly rare. These lesions were often misdiagnosed in the past and were mostly recognized at surgery. Ultrasound is a readily available investigation and the cases of suspected choledochal cyst on ultrasonography (USG) can be further evaluated on magnetic resonance cholangiopancreatography (MRCP). With the advent of modalities like magnetic resonance imaging (MRI) and MRCP, more cases are being recognized preoperatively, which provides a road map to appropriate surgical management.

Case Reports

Case 1
A 26-year-old female presented with pain and jaundice. Laboratory findings included mild elevation of total and direct serum bilirubin and raised levels of alkaline phosphatase. USG done previously had revealed a mass lesion in the gall bladder, with prominent hepatic ducts and the common bile duct (CBD). A tortuous cystic lesion was seen along the course of the cystic duct. The patient was referred for further radiological evaluation. Contrast-enhanced computed tomography of abdomen showed an enhancing mass lesion involving the gall bladder with regional lymphadenopathy. Cystic duct was dilated and tortuous and showed wide communication with CBD [Figure 1A]. A calculus was seen in distal CBD. The patient underwent endoscopic retrograde cholangiopancreatography (ERCP) to remove the obstruction. MRI and MRCP showed grossly dilated and tortuous cystic duct with a wide opening into a prominent common hepatic duct, in addition to gall bladder mass. The CBD was normal in caliber [Figure 1B and C].

The patient underwent laparoscopic radical cholecystectomy for the gall bladder mass. Concurrent excision of the cystic duct and partial excision of the CBD was performed, due to wide opening of the cystic duct into the CBD. The continuity was established by biliary enteric anastomosis. There was no immediate postoperative complication. The patient was discharged on the 7th postoperative day.
Case 2
A 16-year-old male presented with intermittent upper abdominal pain, dark urine, and fever for 1 year. Hyperbilirubinemia was present on biochemical evaluation. Patient had an ultrasound done which indicated the presence of dilatation of the right hepatic duct. A cystic area was reported close to the liver hilum.

Abdominal MRI and MRCP showed fusiform dilation of the cystic duct. The cystic duct opened into CBD through a wide opening. Fusiform dilatation of the right hepatic duct was seen. Left-sided ducts were mildly dilated [Figure 2A-C]. A diagnosis of choledochal cyst involving the cystic duct and intrahepatic ducts was made. The patient underwent open cholecystectomy with cyst excision and Roux-en-y hepaticojejunostomy.

Case 3
A 25-year-old male presented with intermittent right quadrant pain, fever, and jaundice for 5 years. He had been investigated in the past for his symptoms and was carrying multiple biochemical and ultrasound reports documenting the presence of hyperbilirubinemia and choleliathiasis with bilobar fusiform biliary dilatation. He was advised surgery in the past, but had refused to undergo the same. Increasing frequency of the symptoms forced him to seek a definitive therapy.

Abdominal MRI and MRCP revealed choledolithiasis and gross fusiform dilatation of right, left, and common hepatic ducts. Calculus was seen in the dilated right hepatic duct. The cystic duct was dilated, tortuous, and opened into normal caliber CBD with a narrow neck [Figure 3A-C]. A diagnosis of type IV choledochal cyst with cystic duct involvement and choledolithiasis was made. Open cholecystectomy with complete excision of the dilated extrahepatic biliary ducts was done and continuity was established with roux-en-y hepaticojejunostomy for right and left ducts separately. The hepatolithiasis were retrieved during surgery.

Discussion
Choledochal cyst refers to isolated or combined dilatation of extrahepatic and intrahepatic biliary system of congenital origin.[1] The prevalence varies widely, ranging from 1:30,000-50,000 in the western population[2-4] to 1:1000 in Asia.[4] Females are more commonly affected. Majority of the cases present in childhood. Alonso-Lej and colleagues proposed the first classification for choledochal cyst in 1959,[5] which was expanded by Todani in 1977.[6] Bode and Aust reported the first case of cystic dilatation of cystic duct in 1983.[7] Since then, the description of this entity has largely been in the form of isolated case reports and case series. Several terms including choledochal cyst of cystic duct,[8] cystic duct cyst,[9] type VI choledochal cyst,[10,11] choledochocele of cystic duct,[2] and cystic malformation of
cystic duct\textsuperscript{[12,13]} have been used. We carried out a search on the Medline database and Google search using the above terms and the term choledochal cyst. A total of 18 studies with 31 cases were found.\textsuperscript{[1,2,7-22]} Table 1 contains a summary of these cases, including our series.

The most widely accepted theory for occurrence of choledochal cyst postulates that an anomalous pancreatic biliary ductal junction (APBDJ) with long common channel is the primary abnormality.\textsuperscript{[7,16,23]} APBDJ has been reported in 33-90.2\% of choledochal cysts.\textsuperscript{[23,24]} It has been suggested that abnormal APBDJ results in reflux of pancreatic enzymes in the CBD. Due to loss of sphincteric action of the duodenum, the pancreatic enzymes cause weakening of the walls of the biliary channels ultimately leading to dilatation. This has mostly been seen with type I cyst and supported by the level of pancreatic enzymes in biliary secretions. However, this does not completely explain the pathogenesis of the cysts where the CBD is normal. Another popular hypothesis includes a ganglionicosis with proximal biliary dilatation.\textsuperscript{[29]} According to De et al.,\textsuperscript{[14]} cystic duct cyst may result from a combination of APBDJ, acute angulation, and wide orifice of cystic duct hepatic duct junction. However, APBDJ has been observed only in three cases.\textsuperscript{[11,14,21]} Also, wide opening of the cystic duct into hepatic duct was not found in all cases. APBDJ was not seen in our cases. Hence, in our view, other factors may be responsible for cystic duct dilatation. Congenital stricture\textsuperscript{[26]} and aganglionic segment with dilatation of proximal segment\textsuperscript{[6,15]} have also been reported as the possible etiologies. The evaluation of other factors which might be responsible requires further research.

The clinical presentation of patients with cystic duct cyst was generally the same as seen with other types of the choledochal cyst, and included abdominal pain, fever, and jaundice. Most of the patients also had associated complicating biliary tract disease ranging from cholangitis, cholecystitis, cholelithiasis, gall bladder polyp, cystolithiasis, etc.\textsuperscript{[1,3,11,12,13,17-22]} Gall bladder carcinoma was seen in three cases\textsuperscript{[9,12]} including one of our cases.

Association of cystic duct cysts with other congenital biliary abnormality is well documented,\textsuperscript{[8,11,12,14,16,17,19,21]} which makes the case for inclusion of this entity within Todani’s classification. However, due to wide variation in the appearance, exact classification has been a matter of debate. Serrena et al.\textsuperscript{[11]} proposed the inclusion of cystic duct cyst as type VI in Todani’s classification. Loke et al.\textsuperscript{[9]} suggested that these lesions may be variant of type II 2 choledochal cysts. Recently, Mishra et al.\textsuperscript{[21]} have proposed a new modification, wherein cystic duct cysts are included in type II and mixed variants are grouped as type VI.

We studied the age and sex distribution of the disease. Of the 28 cases for which information regarding age was available, 17 were adults\textsuperscript{[1,2,11-15,17-19,22]} and 11 were children.\textsuperscript{[7,12,17-19,24]} Data regarding sex were available for 25 patients. Seventeen patients were females\textsuperscript{[1,2,7,9,11,13,19-21]} and eight were males.\textsuperscript{[17,19,22]} The clinical presentation of these patients was same as in those with choledochal cysts.

A plethora of imaging tools have been used for evaluation in these cases, and include USG, nuclear imaging, multidetector computed tomography (MDCT), ERCP, and MRI with MRCP. The liver-specific contrast agents show biliary excretion and are used to acquire T1-weighted MR cholangiogram which has excellent spatial resolution. Hepatobiliary iodo di acetic acid (HIDA) scan is used to study the biliary anatomy and diagnose obstruction of the biliary tree. In most of the early reports, the correct diagnosis was made only at surgery.\textsuperscript{[12,18,19]} This is most likely due to lack of awareness of the entity and limitations of earlier imaging tools. With modern imaging, these cases are being correctly recognized preoperatively.

USG is generally the first modality employed for investigation of biliary symptoms. A cyst of variable size is seen in relation to portahepatis or CBD. Communication of the cyst with both gall bladder/cystic duct and CBD is seldom seen\textsuperscript{[19]} owing to the cyst being larger and additional abnormalities like dilated extrahepatic biliary tree and shadowing from calculi. Moreover, ultrasound is operator dependent. In the series of Maheshwari et al.,\textsuperscript{[19]} definitive sonographic diagnosis was made in only one out of six cases where USG was used.
Table 1: Summary of reported cases of cystic duct cysts

| Author and year of publication | Number of cases | Imaging modalities used | Cystic duct abnormalities | Associated biliary tract abnormalities | Diagnosis preoperative/intraoperative | Management |
|-------------------------------|----------------|-------------------------|---------------------------|-----------------------------------------|----------------------------------------|------------|
| Bode et al., 1983[2]          | One            | USG, Tc99 m PIPIDA scan, PTC | Fusiform dilatation with narrow neck | Obstructed CBD, cholangitis             | Intraoperative                          | Cholecystectomy, cyst excision, cholecdochoduodenostomy |
| Champetier et al., 1987[3]    | Two            | USG, IVC, ERCP           | Not known                 |                                         |                                        |                                                        |
| Serena Serradel et al., 1991[4] | One            | OCG, USG                 | Cystic dilatation of cystic duct with redundant cystic duct | Cystolithiasis                          | Intraoperative                          | Cholecystectomy, cystic duct excision |
| Loke et al., 1999[5]          | One            | USG, CT, ERCP          | Dilated cystic duct with wide opening into the CBD | Cystolithiasis                          | Intraoperative                          | Cholecystectomy, cyst excision with RYHJ               |
| Bresciani et al., 1999[6]     | One            | USG                     | Cyst of cystic duct with anomalous duct joining the cyst to right hepatic duct | Chronic calculouscholecytis            | Intraoperative                          | Video laparoscopic en bloc resection of cyst and gall bladder with ligature with a clip of the cystic duct and anomalous duct |
| Baj et al., 2002[7]           | One            | USG, oral CT cholangiography | Fusiform dilatation, wide opening | None                              | Preoperative                            | Patient refused surgery                             |
| Weiler et al., 2003[8]        | One            | USG, ERCP            | Not known                 | APBDJ                                  | Preoperative                            | Excision of cyst, CBD with cholecystectomy and RYHJ  |
| Manickam et al., 2004[9]      | One            | USG, HIDA scan, CT, MRC | Not known                 | APBDJ                                  | Not known                               | Excision of cyst with cholecystectomy                |
| Yoon et al., 2009[10]         | Three          |                         |                          |                                        |                                        |                                                        |
| Chan et al., 2009[11]         | One            | CT, MRCP              | Fusiform dilatation with wide opening in the CBD | CBD dilatation, gall bladder polyps     | Preoperative                            | Cyst excision, RYHJ[1]                               |
| Conway et al., 2009[12]       | One            | CT, ERCP, HIDA         | Fusiform dilatation with narrow opening in CBD | Cholelithiasis, chronic cholecystitis  | Intraoperative                          | Excision of cystic duct cyst                         |
| Ghatak et al., 2010[13]       | One            | Not known              | Sacculardilatation        | Fusiform dilatation of CBD             | Not known                               | Excision of cyst, CBD, RYHJ[1]                       |
| Khanuja et al., 2010[14]      | One            | USG, MRCP             | Cystic dilatation with wide opening into the common bile duct | Dilatation of common hepatic duct and CBD | Preoperative                            | Excision of cyst, gall bladder, and common hepatic duct with hepaticojejunostomy  |
| De et al., 2011[15]           | One            | USG, CT, MRC          | Cystic duct cyst with wide opening into CBD and normal distal CBD | Cholecystitis                          | Intraoperative                          | Excision of cyst, gall bladder, and distal CBD, hepatocenterostomy |
| Maheshwari et al., 2012[16]   | Ten             | USG, CT, MRCP         | Fusiform dilatation in six, sacculardilatation in four | Fusiform CBD dilatation: Two cases Cystic duct calculi and malignancy: One case | Preoperative                            | Surgical management of cyst: Five cases, details of surgery not known Surgery for other indications, no intervention for cystic duct cyst; One case Expectant management: Three cases Refused follow-up: One case Excision of cystic duct and part of CBD with RYHJ |
| Shah et al., 2013[17]         | One            | USG, MRCP             | Cystic dilatation with wide opening | Cholecystitis                          | Preoperative                            | Excision of cystic duct and part of CBD with RYHJ    |
| Mishra et al., 2013[18]       | Two            | USG, MRCP             | Case 1: Fusiform dilatation with wide opening | Case 1: CBD, diverticulum, choledochocele, and cholelithiasis | Preoperative                            | Case 1: Excision of CDC with RYHJ[1], derooing of the choledochocele Case 2: CDC excision with RYHJ[1] |
| Kesici et al., 2013[19]       | One            | USG, MRCP             | Fusiform dilatation       | Cholelithiasis                          | Preoperative                            | Elective excision of gall bladder and cystic duct cyst offered to the patient |

Contd...

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Abdominal MRI with MRCP is preferred for further evaluation of sonographic abnormalities due to lack of radiation and high contrast resolution which results in excellent depiction of biliary anatomy and relationships. Origin of cyst from the cystic duct can be easily ascertained and the associated biliary abnormalities are well seen. Normal cystic duct diameter has an upper limit of 5 mm. The cystic duct dilatation may be fusiform or saccular; the fusiform variety is more common. The width of opening of cyst into the CBD is important in cases of isolated cyst due to management implications and must be specifically mentioned in the report. According to Maheshwari et al., cystic duct cyst with a wide opening cannot be distinguished from type II choledochal cyst. However, we feel that with the use of MRI and MRCP, such problem seldom arises.

Management of cystic duct cyst is influenced by multiple factors. These include association of other anomalies, type of opening of the cyst into the CBD, and surgical expertise available. According to Bode et al., the management should aim at prompt diagnosis, examination of the entire biliary tree, complete cyst excision, establish continuity of the biliary tree and intestines, and cholecystectomy. Some authors advocate laparoscopic resection in cases where the isolated cyst has narrow opening into the CBD. For cases with a wide opening, associated dilatation of other parts of biliary tree and complete resection of the dilated segment with re-establishment of biliary intestinal communication is the preferred treatment. In most cases, an open surgery was performed. Bresciani et al. performed video laparoscopic excision. Laparoscopic method was used in our first case as well.

**Conclusion**

In conclusion, we have reported three different variants of cystic duct cyst. Ultrasound and abdominal MRI with MRCP are helpful for preoperative diagnosis and mapping other biliary abnormalities in most cases, which is crucial for appropriate surgical management.

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