

Cystic malformation of cystic duct: 10 cases and review of literature

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Abstract

AIM: To evaluate cystic malformations of the cystic duct.

METHODS: Over a 2-year period, we came across 10 cases of cystic malformation of the cystic duct among patients who were investigated in our radiology department with ultrasonography, multidetector computed tomography, or magnetic resonance imaging for abdominal complaints. Radiological diagnosis of cystic malformation of the cystic duct was made on the basis of a dilated, nonvascular cystic structure near the porta hepatis, and visualization of a clear communication with either the gallbladder, normal caliber cystic duct and/or common bile duct (CBD) on at least one imaging modality.

RESULTS: Four of 10 patients had saccular dilatation of the cystic duct. Six patients had fusiform dilatation of the cystic duct, and two of these had only mild fusiform dilatation. Two patients had associated CBD dilatation and one had associated cystic duct calculi and malignancy.

CONCLUSION: Cystic malformations of the cystic duct should be recognized as a distinct type of choledochal cyst and should be added as type VI cyst in Todani's classification.

INTRODUCTION

Congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract is known as choledochal cyst^[1]. Cystic malformations of the main biliary tract are rare, but well known and classified. Cystic malformations of the cystic duct are extremely rare and not well recognized^[2]. Cystic duct malformations are often misdiagnosed or undiagnosed preoperatively^[3-5]. However carefully performed ultrasonography (USG) alone or in combination with multidetector computed tomography (MDCT) or magnetic resonance cholangiopancreatography (MRCP) suggests the correct diagnosis most of the time. Awareness of this type of malformation as a distinct type of choledochal cyst would help in correct preoperative diagnosis and appropriate management.

MATERIALS AND METHODS

Over a 2-year period, we came across 10 cases of cystic malformation of the cystic duct among patients who were investigated in our radiology department with USG, MDCT or magnetic resonance imaging (MRI) for

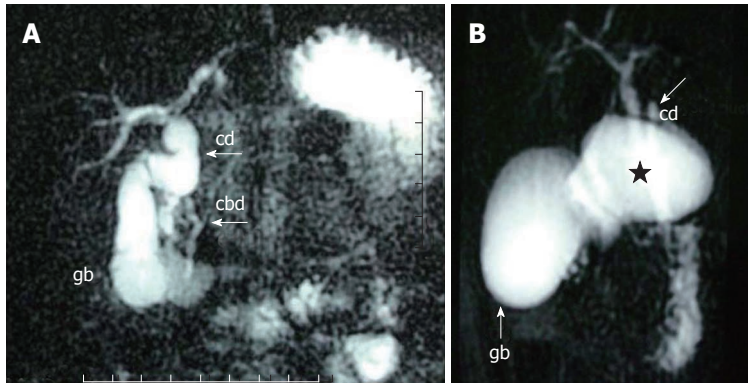


Figure 1 Magnetic resonance cholangiopancreatography. A: Fusiform dilatation of the cystic duct; B: Saccular dilatation of the proximal cystic duct (star). Arrow points to normal caliber distal cystic duct. gb: Gallbladder; cd: Cystic duct; cbd: Common bile duct.

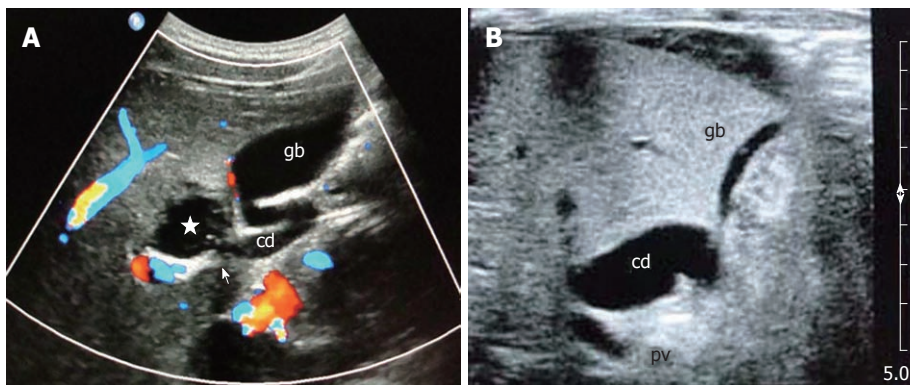


Figure 2 Ultrasonography. A: Focal saccular dilatation of the cystic duct (star), arrow points to communication of the cyst with cystic duct; B: Mild fusiform dilatation of the cystic duct. gb: Gallbladder; cd: Cystic duct.

Table 1 Summary of cases

Patient	Age	Sex	Imaging	Cystic duct dilatation	Associated findings
1	6 d	F	USG, MRCP	Mild fusiform	Gastric perforation
2	4 mo	M	USG, MRCP	Mild fusiform	
3	3 yr	F	USG, MRCP	Fusiform	Fusiform CBD dilatation
4	4 yr	M	USG	Fusiform	
5	4 yr	F	USG, MDCT	Fusiform	Fusiform CBD dilatation
6	7 yr	F	MDCT, MRCP	Fusiform	
7	23 yr	M	USG, MRCP	Saccular	
8	55 yr	F	MDCT	Saccular	Cystic duct calculi and malignancy
9	64 yr	M	MDCT, MRCP	Saccular	
10	75 yr	M	MDCT, MRCP	Saccular	

USG: Ultrasonography; MDCT: Multidetector computed tomography; MRCP: Magnetic resonance cholangiopancreatography; CBD: Common bile duct; M: Male; F: Female.

abdominal complaints. Abdominal pain with or without jaundice was the main presenting symptom. USG was the first imaging modality in six cases. MDCT was the first imaging modality in four cases. A radiological diagnosis of cystic malformation of the cystic duct was

made on the basis of a dilated, nonvascular cystic structure near the porta hepatis and visualization of clear communication with either the gallbladder, normal caliber cystic duct and/or common bile duct (CBD) on at least one imaging modality. Cystic duct diameter greater than that of the CBD in subjective terms and > 5 mm in objective terms was considered abnormal. A diameter of > 5 mm but < 10 mm was considered as mild dilatation.

RESULTS

Patient age ranged from 6 d to 75 years, and there were five male and five female patients (Table 1). Six patients had fusiform cystic duct dilatation. Figure 1A shows fusiform dilatation of the cystic duct in a 7-year-old female child (Patient 6) complaining of recurrent abdominal pain. Note the high insertion of cystic duct and normal caliber CBD. Four patients had saccular cystic duct dilatation. Figure 2A shows focal saccular dilatation of the cystic duct in a 23-year-old male patient (Patient 7) complaining of right hypochondrium pain. Communication of the cyst with the normal proximal cystic duct was clearly seen on USG. Patients 1 and 2 showed a mild fusiform dilatation of the cystic duct. Figure 2B shows mild fusiform dilatation of the cystic duct in a 6-d-old female child (Patient 1) being investigated for neonatal

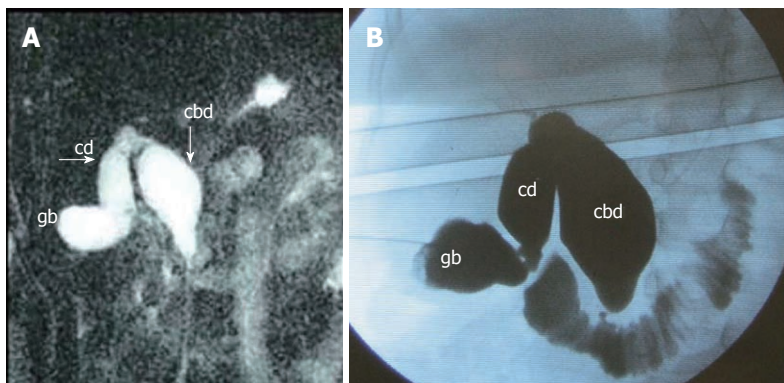


Figure 3 Magnetic resonance cholangiopancreatography (A) and intraoperative cholangiography (B) shows fusiform dilatation of the common bile duct and cystic duct. gb: Gallbladder; cd: Cystic duct; cbd: Common bile duct.

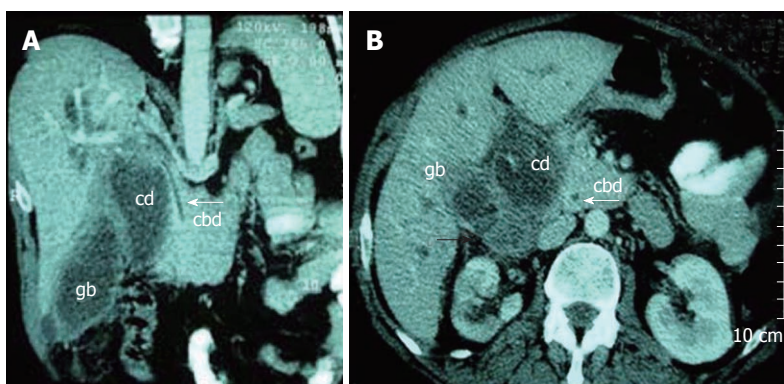


Figure 4 Coronal multiplanar reformatted (A) and axial (B) computed tomography scan shows saccular dilatation of the cystic duct and abnormal soft tissue suggesting malignancy. White arrow points to normal common bile duct. Black arrow points to abnormal soft tissue suggesting malignancy within dilated cystic duct. gb: Gallbladder; cd: Cystic duct; cbd: Common bile duct.

jaundice, vomiting and abdominal distension. Patients 3 and 5 had associated fusiform CBD dilatation. Figure 3 shows fusiform dilatation of CBD and cystic duct in a 3-year-old female child (Patient 3) complaining of abdominal pain. Patient 8 had calculi and malignancy in a dilated cystic duct. Figure 4 shows saccular dilatation of cystic duct in a 55-year-old female patient (Patient 8) complaining of right hypochondrium pain and obstructive jaundice. A normal, distinctly separate CBD was seen (white arrow). Black arrow points to abnormal soft tissue suggesting malignancy.

Findings were confirmed at surgery in Patients 1, 3 and 5-8. Patient 1 had free intraperitoneal air on MRI. Surgery revealed gastric perforation. Intraoperative cholangiography confirmed mild dilatation of the cystic duct as seen on USG and MRI, however, it was not operated upon by the surgeon. Patient 2 had nonspecific complaints and the significance of the radiological finding of mildly dilated cystic duct was not clear, therefore, the patient was advised to undergo follow-up. Patient 4 had abnormal fusiform dilatation of the cystic duct on USG, but refused any further work-up. Patients 9 and 10 had a distinct saccular dilatation of the cystic duct evident on MDCT and MRCP. Figure 1B shows saccular dilatation of proximal cystic duct in a 75-year-old male patient (Patient 10) being investigated for non-related symptoms. Normal caliber distal cystic duct was also seen clearly. This was only an incidental finding on MDCT performed for nonrelated symptoms, and considering the patient's age and absence of any obvious complication, only follow-up was advised.

DISCUSSION

Choledochal cyst is a congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract^[1]. Estimated prevalence of choledochal cysts varies from 1 in 13 000 to 1 in 2 million, and they are rare in the west and far more common in Japan and Asia. Type I choledochal cyst is the most common type (80%-90%)^[6]. There is a female preponderance with a female to male ratio of 3-4:1. Choledochal cyst can present at any age. About two-thirds of patients are clinically symptomatic before the age of 10 years. The classic clinical triad of abdominal pain, jaundice and a palpable right upper quadrant mass is present in 30%-60% of patients presenting in the first decade of life and only in 20% of patients presenting later in life^[1].

Choledochal cysts were originally classified into three types by Alonso-LEJ *et al*^[7] in 1959. Other variations were subsequently recognized and the above classification was modified by Todani *et al*^[8], which is presently the most accepted classification. The classification of Todani *et al*^[8] describes five types of choledochal cysts, but does not describe choledochal cysts of the cystic duct.

Cystic malformation of the cystic duct is a less known entity. On extensive review of the literature, we could find only 16 such cases and most of them were limited to sporadic case reports^[2-5,9-17]. In most of the reported cases a precise diagnosis of choledochal cyst of the cystic duct was made only intraoperatively^[11,6]. Only Yoon

et al^[17] diagnosed all three cases preoperatively with CT or MRI. Over a span of 2 years, we encountered 10 cases of congenital dilatation of the cystic duct diagnosed preoperatively by imaging. Age range of our patients corresponded to other types of choledochal cysts. No sex predilection was found in our patients.

Presenting features and complications of choledochal cysts of the cystic duct are similar to other types of choledochal cysts. They might be asymptomatic and incidentally detected, or present with varying degrees of abdominal pain, jaundice, cholangitis, calculus disease and malignancy.

Although preoperative diagnosis of cystic malformation of the cystic duct can be challenging, in most cases a correct preoperative diagnosis can be suggested on careful radiological imaging. Endoscopic retrograde cholangiopancreatography can delineate the anatomy definitively, however, it is invasive in nature. Tc99m iminodiacetic acid cholescintigraphy can be used to determine the biliary origin of the cyst^[5,15]. Intraoperative cholangiography can be a useful adjunct at surgery^[9,10]. Helical CT cholangiography appears to be an ideal modality for accurately diagnosing such abnormalities, being noninvasive and highly informative^[12]. MRCP or MDCT can also suggest the diagnosis most of the time. USG would be an initial imaging modality in many cases and it can accurately depict the cystic nature of the abnormality, diagnose any associated complications, and differentiate from a vascular etiology with the help of Doppler ultrasound. We found USG to be extremely helpful in correctly localizing the abnormality to the cystic duct, by tracing its connection to the gallbladder or normal portion of the cystic duct on one end, and tracing its connection to the CBD at the other end. However, USG is operator dependent and may fail to depict the biliary origin of the cyst. Therefore, a second line of investigation would be required in most cases for better delineation of anatomy, enhancing the preoperative diagnostic confidence and providing a road map to the surgeon. In our experience MRCP or MDCT along with carefully focused USG can suggest correct preoperative diagnosis most of the time. Any dilated nonvascular cystic structure near the porta hepatis should be evaluated for its connection with the biliary tract and its relationship with the gallbladder, and the cystic duct and CBD should be clearly delineated.

The normal diameter of the cystic duct varies from 1 to 5 mm^[18]. Two distinct patterns of cystic duct dilatation were found in our patients: fusiform dilatation ($n = 6$) and saccular dilatation ($n = 4$). Saccular type choledochal cyst of the cystic duct may be confused with type II choledochal cyst (diverticulum of CBD)^[11]. Saccular type choledochal cyst of the cystic duct can be distinguished from type II choledochal cyst only if a normal narrow caliber cystic duct can be identified between the choledochal cyst and CBD. If the choledochal cyst of the cystic duct has a wide opening into the CBD, it cannot be distinguished from type II choledochal cyst

either radiologically or pathologically.

The exact etiology of choledochal cysts is not clear. Two theories are most prevalent. Babbitt has proposed the theory of anomalous pancreaticobiliary junction (APBJ) with a long common channel, resulting in reflux of pancreatic juice into the biliary tract, with resultant inflammation, scarring and biliary dilatation^[1]. This can explain type I, III and IV cysts but does not explain type II and V cysts where the CBD is normal. Some believe that these cysts are congenital due to distal aganglionosis and proximal dilatation^[19]. The initial case reports of choledochal cysts of the cystic duct do not discuss APBJ. Weiler *et al*^[13] and Noun *et al*^[14] describe an association of choledochal cysts of the cystic duct with APBJ, although no conclusive etiological role has been established. The exact etiology remains debatable, although some authors favor congenital etiology over APBJ as far as choledochal cysts of the cystic duct are concerned^[10].

Manickam *et al*^[5] have reported a case of choledochal cyst of the cystic duct, with apparent evolution of the choledochal cyst over a period of 1.5 years. However, it is not clearly evident from their article, and it appears to us that the choledochal cyst was mistaken for Hartmann's pouch on initial USG. However, the concept of evolution is interesting and needs to be considered. Two of our 10 patients were infants (aged 6 d and 4 mo) and they showed a distinct abnormal dilatation of the cystic duct, but it was not a sizeable cyst and measured > 5 mm but < 10 mm in diameter. Four patients who were in the pediatric age group (3-7 years) had a fusiform cystic duct dilatation of > 10 mm. All four adult patients (23-75 years) had saccular dilatation of the cystic duct. A choledochal cyst may evolve over a period of time due to repeated infection and inflammation. Mild choledochal cysts diagnosed in infancy or childhood, and not treated surgically for whatever reasons, need to be followed up closely to look for apparent evolution and complications.

Treatment for type I, II and IV choledochal cysts is complete excision and subsequent biliary-enteric anastomosis for drainage^[1]. Some treat type II cysts by simple excision^[9]. There are no uniform guidelines for the treatment of choledochal cysts of the cystic duct. Some treat isolated choledochal cysts of the cystic duct with a narrow opening in the CBD with simple cholecystectomy and cyst excision either by open surgery or laparoscopically^[2,5,9,10] and some prefer complete excision with biliary drainage^[13,14]. We recommend complete excision with biliary drainage, in view of the increased risk of developing bile duct and gallbladder cancer in the presence of cystic biliary duct anomalies and APBJ.

In conclusion, cystic malformations of the cystic duct share features common to other types of choledochal cysts in Todani's classification. Although rare, they need to be recognized. We recommend modification of Todani's classification and inclusion of cystic malformations of the cystic duct as type VI choledochal cysts. It is not clear whether the milder forms of abnor-

mally dilated cystic duct seen in our infantile patients are an arrested form of the symptomatic choledochal cyst or have the potential to grow into symptomatic cysts with age. Such cases require long-term follow-up to determine the exact significance, future course of events and appropriate management.

COMMENTS

Background

Choledochal cysts of the cystic duct are considered to be very rare. They are not described in the widely accepted Todani's classification of choledochal cysts. There is a lack of awareness of this entity, resulting in missing the diagnosis altogether or confusing it with other types of choledochal cysts, and inappropriate treatment.

Research frontiers

There is a lack of proper guidelines for diagnosis and management of choledochal cysts of the cystic duct. In this study, the authors highlight preoperative diagnosis and appropriate management of choledochal cysts of the cystic duct.

Innovations and breakthroughs

To date, only a few sporadic case reports have been published in the literature, of which, most cases were diagnosed correctly only during surgery. In the present study, the authors presented a series of cases correctly diagnosed preoperatively. Furthermore, the condition may not be as rare as previously thought, and the authors suggest that choledochal cysts of the cystic duct should be included as a distinct entity in Todani's classification of choledochal cysts.

Applications

Awareness of choledochal cysts of the cystic duct as a distinct type of choledochal cyst would lead to its consideration in the preoperative differential diagnosis, and correct preoperative diagnosis will help with appropriate treatment.

Terminology

Choledochal cyst is congenital dilatation of the biliary tract. Todani's classification describes five types of choledochal cysts depending upon the part of the biliary tract involved, but does not describe choledochal cysts of the cystic duct.

Peer review

The article is well written. The information is interesting, new and important.

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