Adult Choledochal Cyst: Intra-operative Surprise. Case Report and Literature Review
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Citation

Abstract
Presentation of a choledochal cyst is rare in adults. The diagnosis of adult choledochal cysts is frequently delayed due to nonspecific clinical symptoms or symptoms obscured by secondary hepatobiliary disease. We report a case of choledochal cyst associated with gallstones in a 15-year-old female which was diagnosed during laparoscopic cholecystectomy.

INTRODUCTION
Choledochal cysts (CCs) are congenital conditions associated with benign cystic dilatation of bile ducts. Although cystic disease of the biliary tree has been described since 1723, much about its etiology, pathophysiology, natural course and optimal treatment remains under debate. CCs are reported mainly in children; however, an increasing number of adult patients have been diagnosed with the disease. Pre-operative diagnosis is essential for proper planning for surgery and to avoid unexpected intra-operative surprise. We report a case of an adult choledochal cyst missed on ultrasonic evaluation of a patient with gallstone symptoms. The diagnosis was established intra-operatively during laparoscopic cholecystectomy by intra-operative cholangiogram and confirmed by post operative MRCP.

CASE REPORT
A 16-year-old female patient presented to the surgical outpatient department at King Fahad Medical City, Riyadh, KSA with a history of recurrent right hypochondrium pain radiating to the right shoulder. She gave no history of jaundice, fever, rigors or previous surgery. She had no history of medical illness. Clinical examination was entirely normal. Laboratory investigations including full blood count, liver function test and urea and electrolytes were within normal limits. The upper abdominal ultrasound was reported as liver normal in size and echogenicity with no focal lesion. The gallbladder showed multiple stones with normal wall thickness and no evidence of acute inflammation. The common bile duct was normal in caliber and there was no dilatation of the intrahepatic bile radicles (figure 1, 2).

Figure 1
Figure 1: Ultrasonography of the gallbladder showing multiple stones with normal wall thickness and no evidence of acute inflammation.
The patient was booked for elective laparoscopic cholecystectomy on the assumptive diagnosis of gall stones. Intraoperatively, the gallbladder was distended, but not inflamed (figure 3). On traction of the gallbladder upwards, a dilated cyst was seen which seemed to be in continuity with the gallbladder just below the level of the cystic duct (figure 4). The cystic duct was dissected (figure 5) and transcystic duct cholangiography was done (figure 6) which confirmed the diagnosis of a choledochal cyst.

**Figure 2**
Figure 2: Ultrasonography of the liver showing a dilated tubular structure posterior to the gallbladder with internal echoes and posterior wall enhancement (findings overlooked in the initial ultrasound report).

**Figure 4**
Figure 4: Huge cyst immediately below and in continuity with the gallbladder and above the duodenum.

**Figure 5**
Figure 5: The cystic duct was dissected and it was obvious that it was in continuity with the cyst.
We proceeded with routine laparoscopic cholecystectomy without dealing with the cyst at this stage. A postoperative MRI confirmed the diagnosis of a choledochal cyst (figures 7 & 8).

Figure 7
Figures 7 & 8: Postoperative MRCP confirming the diagnosis of a choledochal cyst.

Subsequently, the patient had excision of the cyst with hepatojejunostomy without complications. The histopathology report confirmed the diagnosis of a choledochal cyst.

DISCUSSION
Choledochal cysts (CCs) are congenital conditions associated with benign cystic dilatation of bile ducts. They are uncommon in western countries (1) but not as rare in East Asian nations like Japan and China. In North America the incidence of CCs is estimated to be 1/150,000 (2).

Although cystic disease of the biliary tree has been described since 1723, much about its etiology, pathophysiology, natural course and optimal treatment remains under debate (3).

Vaterl is often cited as the first to report a case of CC in 1723, but the first well documented case was reported by Douglas (4) in 1852 when he presented a biliary cyst in a 17-year-old girl.

Alonso-Lej and colleagues (5) proposed the first classification system for CCs in 1959, describing 3 types of bile duct dilation, which has gained wide acceptance. Todani and colleagues (6) expanded this system in 1977 to include the occurrence of intrahepatic and multiple cysts, and this modified classification is now most commonly used by clinicians (7). This classification is based on the cholangiographic morphology, location, and number of intrahepatic and extrahepatic bile duct cysts.

In 1958, Caroli (8) described in detail the distressing course of the condition.

No strong unifying etiologic theory exists for choledochal
cysts. The pathogenesis is probably multifactorial (7). Although many theories have been put forth, Babbitt’s (9) theory of cysts caused by an abnormal pancreaticobiliary duct junction such that the pancreatic duct and the common bile duct meet outside the ampulla of Vater, thus forming a long common channel, has gained much popularity. Empting of pancreatic duct into the common bile duct allows pancreatic secretions to reflux into the common bile duct, where the pancreatic proenzymes become activated, damaging and weakening the bile duct wall. Defects in epithelialization and recanalization of the developing bile ducts and congenital weakness of the ductal wall also have been implicated. The result is formation of a choledochal cyst.

CCs are reported mainly in children; however, an increasing number of adult patients have been diagnosed with the disease. About 20-30% of CCs are diagnosed in adults (10).

Most of CCs present in the first year of life; adult presentation is rare and usually at this stage is associated with complications. The classic triad of intermittent abdominal pain, jaundice, and a right upper quadrant abdominal mass is found only in 20% of patients.

The diagnosis of adult choledochal cysts is frequently delayed due to nonspecific clinical symptoms or symptoms obscured by secondary hepatobiliary disease.

Pre-operative diagnosis is essential for properly planned surgery and to avoid unexpected intra-operative surprise. There are only few cases of CCs that were diagnosed intra-operatively. Sharp et al. reported a case of choledochal cyst found during a cholecystectomy using a transcystic intra-operative cholangiogram (11).

Gibbs et al. reported a case which was almost similar to our case in which a Hispanic female presented with classic findings consistent with biliary colic. A preoperative ultrasound revealed multiple gallstones and mild dilatation of the common bile duct. At the time of laparoscopy, she was found to have a dilated common bile duct, cystic duct, and gallbladder. Further dissection was discontinued, a cholecystectomy made, and a percutaneous transabdominal cholangiogram through the gallbladder was performed, which revealed a type I choledochal cyst (12).

A choledochocele may be easily overlooked by the conventional diagnostic methods, such as upper gastrointestinal series, intravenous cholangiogram, abdominal ultrasound, and computed tomography (13). Multidetector computed tomography with reformatted imaging, magnetic resonance cholangiopancreatography, and endoscopic retrograde cholangiography represent the important techniques providing the anatomical resolution and detail required to properly diagnose and classify choledochal cysts and their associated abnormal features of the biliary tree, as well as their pancreaticobile duct union (14).

Magnetic resonance cholangiopancreatography (MRCP) is now considered to be the gold standard (15, 16, and 17). MR cholangiography can be used to confirm the diagnosis of choledochal cysts and define the extent of involvement preoperatively (18). MRCP offers diagnostic information that is equivalent or superior to that of ERCP for the evaluation of type I choledochal cysts in adults and because this modality is non-invasive, it should be the preferred imaging technique for examination of adult patients with choledochal cysts (19).

Matos et al. compared MR cholangiopancreatography and endoscopic retrograde cholangiopancreatography in eight patients with choledochal cysts. They concluded that MRCP provides information equivalent to that provided with ERCP, without potential complications, for the preoperative assessment of choledochal cysts. Dynamic secretin-stimulated MRCP studies might help better understand the pathophysiologic characteristics of this entity (20).

Complications of choledochal cysts in adults include cholecystitis, recurrent cholangitis, biliary stricture, choledocholithiasis, recurrent acute pancreatitis, and malignant transformation into cholangiocarcinoma (2).

In the past, choledochal cysts were often treated using drainage procedures; however, the optimal treatment used today is likely to involve the complete excision of the extrahepatic duct, cholecystectomy, and Roux-en-Y hepaticojejunostomy.

**SUMMARY**

CCs are reported mainly in children; however, an increasing number of adult patients have been diagnosed with the disease. Adult presentation is rare and the diagnosis is frequently delayed due to nonspecific clinical symptoms or symptoms obscured by secondary hepatobiliary disease. Pre-operative diagnosis is essential for proper planning for surgery and to avoid unexpected intra-operative surprise.
MRCP provides a valuable non-invasive diagnostic modality for establishing the diagnosis in suspected cases.

References
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