CASE REPORT

A rare choledochal cyst of the cystic duct with an anomalous pancreaticobiliary union in an adult patient

S. Manickam a,*, K. Ramadan b, B.K. Adams c

Departments of aRadiology and Clinical Imaging, bSurgery, Al-Ain Hospital, and cNuclear Medicine, Mafraq Hospital, Abu Dhabi, United Arab Emirates

Introduction

Choledochal cysts of the cystic duct are extremely unusual. Very rarely they occur in conjunction with anomalous pancreaticobiliary union. We present a patient with a cystic duct choledochal cyst with a wide orifice into the common bile duct (CBD) and an anomalous pancreaticobiliary union. These findings were confirmed at surgery.

Case report

A 20-year-old woman presented with a 5-year history of recurrent upper abdominal pain. She had no other symptoms and there was no history of jaundice or fever. Clinical examination was normal. Except for an elevated alkaline phosphatase (136 IU/l; normal range 45-121 IU/l), liver function tests including serum bilirubin and haematological investigations were normal.

Initial ultrasound examination revealed multiple tiny gall bladder calculi with a cystic structure close to the gall bladder neck, which was originally presumed to be Hartmann’s pouch in a tortuous gall bladder. Follow-up ultrasound after 1.5 years (Fig. 1) revealed a fusiform structure postero-lateral to the gall bladder thought to be related to the cystic duct. On this examination, the gall bladder was dilated and a constriction was noted at the junction of the gall bladder neck and cystic duct. This was later proven on histopathological examination after cholecystectomy. There was no dilatation of the intra-hepatic biliary radicles. Tc99m IDA scintigraphy demonstrated a fusiform structure medial to the gall bladder that filled before the latter (Fig. 2a and b). Emptying of these structures occurred in the reverse sequence with gall bladder excretion being delayed (Fig. 2c). The ultrasound findings were confirmed using computed tomography (CT) and also revealed the extent of the cyst. Part of the dilated common bile duct could be seen well in the pancreatic head region (Fig. 3). The exact configuration of the distal CBD and the nature of the pancreatobiliary ductal anomaly could not be appreciated on the axial CT images, which were obtained with a 20 cm field of view (FOV) and slice thickness of 3 mm on a multislice CT machine.

Axial three-dimensional turbo spin-echo (TSE) T2-weighted (T2W) magnetic resonance imaging (MRI) performed on a 1.5 T machine showed the constricted segment of the gall bladder neck well (Fig. 4a). Three-dimensional TSE T2W coronal, thick-slab, breath-hold magnetic resonance cholangiopancreatography (MRCP) images performed with section thickness of 70 and 400 FOV with an acquisition time of 20 s (Fig. 4b) confirmed the cystic duct dilatation and showed a distal narrowing of the CBD, but failed to demonstrate the anomalous pancreaticobiliary duct union (APBDU). In an attempt to delineate the distal CBD an endoscopic retrograde cholangiopancreatography (ERCP) was performed, which demonstrated an ectatic cystic duct opening with a wide orifice into a long narrow anomalous distal CBD. The common hepatic duct was seen dilated proximally unto its division. There was no dilatation of the intra-hepatic biliary radicles. The CBD, which was found narrowed distally, joined the pancreatic duct instead of opening into the ampulla of Vater (Fig. 5). Operative cholangiography confirmed the ERCP findings and showed additionally the dilated cystic duct communication with the distal narrow CBD (Fig. 6).

A choledochal cyst of the cystic duct with a large communicating cyst-CBD orifice and APBDU with a long, distally narrow CBD and associated APBDU were confirmed at surgery. Cholecystectomy was performed and histology did not reveal any gall bladder or cystic duct malignancy.

Discussion

Choledochal cysts are dilatations of the biliary tree; their precise pathogenesis remains unclear. Diagnosis in adulthood is uncommon and there is a female predominance.1 The present patient presented with abdominal pain, a common symptom, possibly due to recurrent cholangitis.2 The patient had calculi, known to occur in approximately 8% of individuals with choledochal cysts.3 The constricted gall bladder–cystic duct junction in the present

*Guarantor and correspondent: S. Manickam, P.O. Box: 82464, Al-Ain, United Arab Emirates. Tel.: +971-50-6366754; fax: +971-3-7622407.
E-mail address: kumarmsk@yahoo.com
case was an inflammatory stricture as proven histologically, and could be possibly attributed to recurring attacks of inflammation due to the presence of calculi within the gall bladder. A choledochal cyst of the cystic duct is extremely unusual and this anomaly is not included in Todani’s classification scheme as the disorder most frequently involves the CBD and, less commonly, the hepatic ducts. A small number of reports describe cystic dilatation of the cystic duct or what has been termed a type VI choledochal cyst. However, the association of this condition with an APBDU, which was found in the present patient, has not been reported in the other cases. An APBDU has been observed in up to 40% of patients with choledochal cysts and forms the basis of the aetiological hypothesis that suggests this condition may be due to pancreatic reflux. However, in the present case the cystic duct dilatation could possibly have a similar pathogenesis due to the wide communicat-

Figure 1  Ultrasound study showing a fusiform cystic structure (white arrow) posterolateral to the distended gall bladder (white arrowhead) thought to be a choledochal cyst.

Figure 2  (a) Tc-99m IDA cholescintigraphy demonstrates filling of the CBD and then the fusiform cyst, (black arrow) and early filling of the gall bladder (white arrowhead). (b) Tc-99m IDA cholescintigraphy pre-fatty meal shows filled cyst (black arrow) and gall bladder (white arrowhead). (c) Tc-99m IDA cholescintigraphy post-fatty meal shows emptying of the cyst into the duodenum with continued retention of Tc99m-IDA in the gall bladder (white arrowhead).

Figure 3  Axial CT reveals a cystic structure between the cystic duct (white arrowhead) and the proximally dilated CBD (white arrow), which was found extending up to the superior border of the pancreatic head.
passage of pancreatic secretions into the cystic duct rather than the CBD. Carcinoma of the biliary tract is a well-documented complication of this condition and the present patient was at increased risk; an APBDU on its own has been associated with an increased incidence. This was the rationale for surgical excision of the cyst after cholecystectomy. This procedure reduces, but does not entirely prevent, complications such as cholangitis, lithiasis, pancreatitis and even carcinoma, thus making long-term follow-up essential. The diagnosis is usually made on ultrasound and Tc-99m iminodiacetic acid cholescintigraphy. Both techniques clearly demonstrate the choledochal cyst. The cyst was also visualized on CT and MRCP, although the latter failed to provide clear evidence of the APBDU. This was well delineated on ERCP and operative cholangiogram, which also demonstrated the large cyst–CBD communication, and excluded any associated intra-hepatic biliary disease.

**Figure 4** (a) Axial section MRI shows the constricted portion of gall bladder (white arrow) and the cystic duct dilatation (white arrowhead). (b) Single-slice, thick-slab, breath hold MRCP image shows a constricted junction of the gall bladder and cystic duct (white arrow) where it is folded on itself and the distal insertion of the dilated cystic duct into the CBD (white arrowhead). The distal pancreatic duct was not seen well.

**Figure 5** ERCP demonstrates the constricted gall bladder–cystic duct junction (white arrow) and an ectatic cystic duct and a long narrow anomalous distal CBD joining the pancreatic duct (white arrow head).

**Figure 6** Operative cholangiography in anteroposterior view after excision of the gall bladder at the gall bladder–cystic duct junction. The contrast-opacified dilated cystic duct is seen opening into the distal CBD (white arrow). Distal communication of CBD (white arrowhead) with the pancreatic duct is well shown.
In the present patient, the cyst would appear to have evolved over a period of time as could be appreciated on the serial ultrasound studies. To our knowledge this combination of biliary tract anomalies and evolution of a choledochal cyst over a period of time involving the cystic duct has not been previously reported.

References