CASE REPORT

Magnetic resonance cholangiopancreatography diagnosis of choledochal cyst involving the cystic duct: report of three cases

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ABSTRACT. Here we describe three cases of choledochal cyst involving the cystic duct or isolated dilatation of the cystic duct. All cases were visualised on MR cholangiopancreatography (MRCP) imaging. We report findings of ultrasonography, CT and MRCP. These cases are extremely rare; nine cases have been reported in the English-language literature. This anomaly was not included in Todani's classifications of choledochal cysts, a system that is accepted worldwide. We think that this variant anomaly will be included in this classification system in the near future.

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Case report

Case 1

A previously healthy 59-year-old woman presented with a month-long history of intermittent back pain. There was no liver function test abnormality. Mild increased α -fetoprotein (AFP, 18.24 ng mL⁻¹; normal range ~10 ng mL⁻¹) was noted. Initial work-up included a transabdominal ultrasonography, which showed a hypoechoic mass replacing the fundal portion of the gallbladder (Figure 1a). On abdominal CT, a soft-tissue mass was noted in the fundus of the gallbladder with suspicion of hepatic invasion (Figure 1b); multiple enlarged lymph nodes were identified in the peripancreatic, portocaval, aortocaval, and left paraaortic regions (Figure 1b), suggesting the possibility of malignancy.

Focal fusiform dilatation of the cystic duct without a definite cause of obstruction was identified and was about 22 mm in diameter and 34 mm in length (Figure 1c).

The extrahepatic bile duct and common bile duct were of normal diameter.

MR cholangiopancreatography (MRCP) was performed for further evaluation. On thick-slab coronal oblique MRCP imaging, fusiform dilatation of the cystic duct with normal diameters for the intrahepatic and extrahepatic bile ducts was seen (Figure 1d).

There was no evidence of bile duct stone or stenosis in the transitional portion of the cystic duct. Also, there was no evidence of definite anomalous union of the pancreaticobiliary duct (AUPBD). This patient was diagnosed with advanced gallbladder cancer with lymph

nodes metastasis combined with choledochal cyst involving the cystic duct only.

Case 2

A 48-year-old woman was transferred from a local clinic because of bile duct dilatation. The patient complained of intermittent dyspepsia and mild abdominal pain for 6 weeks. But there was no definite tender point or Murphy's sign on physical examination. On CT scan performed at an outside institution, mild wall thickening of the gallbladder was identified with luminal contraction and suspicion of gallbladder stones. Demonstrable dilatation of the common bile duct was present with normal appearance of the intrahepatic bile duct and pancreatic duct. The laboratory findings showed an increased level of total bilirubin (1.2 mg dL⁻¹; normal range, 0.2–1.0 mg dL⁻¹); other laboratory results including serum tumour markers were within normal limits. Abdominal MRI (Figure 2a) and MRCP (Figure 2b) were performed, showing fusiform dilatation of the common bile duct and cystic duct of about 7 cm in length and 3 cm in diameter; the ducts were joined by a wide opening, but no visible cause of obstruction was identified. There was no evidence of definite AUPBD. After 1 week of clinical observation, the total bilirubin level returned to normal. The patient was conservatively treated for 5 years because she refused surgery; nevertheless there was the attendant risk of malignancy. The evidence of concomitant malignancy was not demonstrated during the follow-up period.

Case 3

A 15-year-old girl presented with a 4 day history of postprandial epigastric and right upper quadrant abdominal pain, fever and chills. On physical examination, the

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Figure 1. 59-year-old female with a choledochal cyst involving the cystic duct and advanced gallbladder cancer (a) Transabdominal ultrasound image shows a hypoechoic solid mass replacing the fundal portion of the gallbladder (asterisk). (b) Axial contrast-enhanced CT during delayed phase shows a focal hypodense mass in the gallbladder (arrows) and multiple enlarged lymph nodes in the aortocaval and left paraaortic portion (arrowheads). (c) More superior level of axial contrast-enhanced CT shows marked dilatation of cystic duct (arrows) and multiple hypodense lymph nodes in the peripancreatic portion. (d) Thick-slab coronal oblique MR cholangiopancreatography shows fusiform dilatation of the cystic duct (arrow) with normal diameter of the intrahepatic and extrahepatic bile ducts.

patient appeared acutely ill with abdominal tenderness and a positive Murphy's sign. Clinically the patient was suspected of having acute cholecystitis.

Laboratory evaluation showed mild elevation of the total bilirubin level (1.9 mg dl^{-1} ; normal range $0.2\text{--}1.0 \text{ mg dl}^{-1}$) and direct bilirubin level (1.0 mg dl^{-1} ; normal range $0.0\text{--}0.5 \text{ mg dl}^{-1}$), and elevated levels of aspartate aminotransferase (70 IU l^{-1} ; normal range $0\text{--}40 \text{ IU l}^{-1}$), alanine aminotransferase (66 IU l^{-1} ; normal range $0\text{--}45 \text{ IU l}^{-1}$), γ -glutamyl transpeptidase (222 IU l^{-1} ; normal range $5\text{--}37 \text{ IU l}^{-1}$) and lactate dehydrogenase (435 IU l^{-1} ; normal range $232\text{--}410 \text{ IU l}^{-1}$) were identified.

On ultrasonography performed at an outside hospital, an abnormally dilated common bile duct was noted. The abdominal CT showed fusiform dilatation of the extrahepatic bile duct, 4 cm in diameter, and fusiform dilatation of the cystic duct, 2.5 cm diameter and 6.5 cm length, without a definite cause of obstruction (Figure 3a,b).

Multiple small gallbladder polyps were also identified (Figure 3a). On MRCP, fusiform dilatation of the extrahepatic bile duct and combined fusiform dilatation of the cystic duct with wide opening were demonstrated (Figure 3c). There was no evidence of definite AUPBD. Cyst excision and Roux-en-Y hepaticojejunostomy was performed.

Grossly, a cystic lesion was observed along the distal portion of cystic duct (Figure 3d). Histological analysis demonstrated a partial lining of columnar cells supported by fibrous tissue with mild chronic inflammation, compatible with choledochal cyst.

Discussion

Choledochal cysts are traditionally associated with cystic dilations of the extrahepatic duct. In Western populations, the incidence is 1 per 30 000–50 000 [1]. Asian

Figure 2. 48-year-old female with a choledochal cyst involving the cystic duct. (a) T_2 weighted axial image shows marked dilatation of the choledochal cyst in the retropancreatic portion (arrow). (b) Thick-slab coronal oblique MR cholangiopancreatography shows fusiform redundant dilatation of the common bile duct involving the cystic duct via a wide communication (asterisk) and abrupt normalisation of the distal common bile duct (arrows).

populations have a higher incidence, estimated at 1 in 1000 in Japan [2]. This disease is now reported with increasing frequency, probably because of improved diagnostic tools, especially MRCP.

This disease entity is associated with an AUPBD, the reported incidence of this association is 33–90.2% [3, 4]. The classic triad of jaundice, pain and a right hypochondrial mass is present in approximately 6–38% of cases [2, 5]. This classic presentation appears less frequently nowadays because of earlier diagnosis and treatment. In children, incidental biliary ascites was reported after spontaneous or traumatic perforation of a choledochal cyst [6]. Biliary carcinoma occurs in 2.5–39.4% of patients with biliary cysts [7, 8]: the degenerated mucosa of a choledochal cyst increases the risk for carcinomatous development due to chronic irritation, ulceration and subsequent regeneration of cyst epithelium [9].

The widely accepted classification system for choledochal cysts, which was devised by Todani et al [10] (Figure 4), is based on cholangiographic morphology, location and number of intrahepatic and extrahepatic bile duct cyst.

Type I cysts are confined to the extrahepatic bile duct and can be further subdivided into IA (entire cystic involvement of extrahepatic bile duct), IB (segmental cystic involvement of extrahepatic bile duct) and IC (diffuse or cylindrical dilatation of extrahepatic bile duct). Type II cysts are true diverticula of the extrahepatic bile duct. Type III cysts are also referred to as choledochoceles and are confined to the extrahepatic bile duct within the duodenal wall. Type IV cysts have multiple features, which can include both an extrahepatic and an intrahepatic component. Type IV cysts can be further subdivided into type IVA, involving both the extra- and intrahepatic bile duct, as well as type IVB, involving multiple segmental dilatations of the extrahepatic bile duct. Finally, type V (Calori's disease) cysts are confined to the intrahepatic bile duct. Calori's disease is thought to be an inherited condition and is clinically

associated with intrahepatic gallstone formation and a high incidence of renal disease.

Subsequently, Todani et al [11] revised their classification of Type I choledochal cysts based on AUPBD. Type IA is a cystic dilatation of the extrahepatic bile duct.

Type IB is a segmental dilatation with no presence of AUPBD. Type IC refers to a fusiform, diffuse or cylindrical dilatation of the extrahepatic bile duct with AUPBD. Moreover, the dilatation often extends continuously into the intrahepatic duct. Lastly, type IVA shows primary ductal stricture, especially around the hepatic hilum and umbilicus.

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. It has become clear that these operations have an unacceptable rate of long-term complications, most notably malignant transformation [12, 13].

The traditional classification of choledochal cysts by Todani et al [10] comprises five different types of biliary cyst, but this classification system does not cover all kinds of bile duct cysts; cystic dilatation of the cystic duct is not included. Serradel et al [2] described one case of isolated dilatation of the cystic duct as "type VI choledochal cyst", an addition to the Todani classification. Our first case of a cyst localised exclusively to the cystic duct appears to be similar to that of Serradel et al. Our first case showed isolated fusiform dilatation of the cystic duct combined with gallbladder cancer; this cyst did not involve the extrahepatic duct or evidence of AUPBD.

Our second patient presented with combined fusiform dilatation of extrahepatic and cystic ducts with a wide communication between them; this patient also had multiple gallbladder polyps. In our third patient, separate fusiform dilatation of the extrahepatic bile duct and cystic duct were identified. All three cases were diagnosed preoperatively with CT or MRCP. There were no evidence of AUPBD in all three cases.

Figure 3. 15-year-old girl with a choledochal cyst involving the cystic duct and multiple gallbladder polyps. (a) Axial contrast-enhanced CT during the portal venous phase shows cystic dilatation of the cystic duct (asterisk) and a dilated extrahepatic duct (arrow) and multiple small gallbladder polyps (arrowheads). (b) Sagittal multiplanar reconstruction CT image shows fusiform dilatation of the extrahepatic bile duct and a fusiform dilated cystic duct (asterisk). (c) Thick-slab coronal oblique MR cholangiopancreatography shows a well-demonstrated choledochal cyst involving extrahepatic bile duct and combined cystic duct dilatation (arrow). (d) Photograph of an opened, resected specimen showing a fusiform-dilated cystic duct (asterisk) and extrahepatic bile duct (arrow) (G, gallbladder). On microscopic examination (not shown), the wall of the extrahepatic bile duct was partly lined by columnar cells and supported by fibrous tissue with mild chronic inflammation, and compatible with choledochal cyst.

Our cases each represent different types of choledochal cysts. These are extremely rare, with only nine cases reported in the English literature [2, 14–20]. The published reports predominately highlight clinical and surgical findings, not diagnostic findings such as MRCP. Of the reported nine cases, isolated dilatation of the cystic duct was found in four cases and combined dilatation of the cystic duct and common bile duct was found in five cases. There were no cases diagnosed with radiological imaging before surgery; most cases were diagnosed as Todani II choledochal cyst, duplicated gallbladder or Mirizzi syndrome. Only one case had AUPBD that showed combined dilatation of the cystic duct and common bile duct [17]. There were no reports of AUPBD in cases of isolated dilatation of the cystic duct. Some authors have suggested a different theory for the occurrence of choledochal cyst

in isolated cystic ducts, including congenital cystic duct stricture and proximal ductal dilatation [21], or an associated functional abnormality and absence of ganglion cells in choledochal and cystic duct dilatation [22].

It is very important to recognise and correctly diagnose this rare biliary anomaly as choledochal cysts preoperatively to avoid performing laparoscopic cholecystectomy. Treatment of choledochal cysts is via cholecystectomy, with excision of the cyst and transection of the cystic duct, because of the possibility of malignancy. If there is a wide opening between the cystic duct and common duct, Roux-en-Y choledochal jejunostomy and total cyst excision are required [14].

In conclusion, we have reported three rare cases of congenital cystic dilatation of the cystic duct, one case represented an isolated finding and two cases occurred in

Figure 4. Choledochal cyst was described according to Todani's classification system.

combination with a classic choledochal cyst. Radiological evaluation, especially with MRCP, is critical for the correct pre-operative diagnosis and treatment plan. If similar cases are reported in the future, we propose that these variant anomalies be included in the classification of choledochal cysts.

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