CASE REPORT

Imaging Diagnosis of Type III Choledochal Cyst: A Case Report

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Abstract Choledochocoele is a congenital abnormality of the biliary system, which characterized by a cystic dilatation of intramural segment of the distal common bile duct. Choledochocoele manifests as cystic lesions in the duodenal lumen and resembles duodenal duplication cysts in imaging. We reported a patient with choledochocoele. Magnetic resonance cholangiopancreatography showed a thin-walled sac in the duodenal cavity and a fine tubular structure connecting with the end of the common bile duct. Magnetic resonance cholangiopancreatography is a noninvasive and convenient technique in the diagnosis of choledochocoele.

CHOLEDODCHAL cysts (CCs) are congenital abnormality of the biliary system. It is characterized by cystic dilatation of intrahepatic duct and extrahepatic biliary tree individually or both.¹ According to the modified Todani system, CCs are classified into categories I-V. Type III is a rare form of CCs, also called choledochocoele, which comprises only 4%-6% of all reported cases.² It appears as a cystic dilatation of intramural segment of distal common bile duct protruding into duodenal lumen. Choledochocoele has distinctive

anatomic features and a lower risk of malignancy than other types.³⁻⁴ The clinical presentations and imaging findings of choledochocoele resemble duodenal duplication cysts (DDCs), making their differential diagnosis difficult.⁵ Imaging examinations play an important role in the initial diagnosis, differential diagnosis, surgical planning and long term surveillance of CCs.⁶ The purpose of this study was to demonstrate the importance of magnetic resonance cholangiopancreatography (MRCP) and biliary specific contrast agents in the diagnosis of bile duct cyst.

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CASE DESCRIPTION

A 42-year-old man was admitted to the Peking Union Medical College Hospital on October 29, 2015 for

having been suffering from abdominal pain over one year. There was no significant medical history or family history. Physical examination did not show obvious abnormality. Laboratory data including complete blood count, urinalysis and blood biochemistry were within the normal limits.

In order to confirm the diagnosis, he underwent a series of imaging examinations. Ultrasonography did not show significantly positive signs. After symptomatic treatment, abdominal pain still appeared intermittently. Next, the patient underwent gastroscopy and endoscopic ultrasonography to make a definitive diagnosis. The results revealed a parenteral cystic lesion in the descending section of the duodenum. It was deemed as a duodenal diverticulum (Fig. 1). The upper gastrointestinal barium study displayed a round filling defect in the second portion of the duodenum (Fig. 2). Therefore we excluded the diagnosis of duodenal diverticulum. Abdomen CT revealed a thin-walled cystic mass without nodules causing mild duodenal dilatation and multiple stones in the duodenum (Fig. 3). We considered it as a type III CC or a DDC. In order to confirm the relationship between the cystic lesion and the intestine, MRCP was performed with pineapple juice (PJ). PJ can significantly reduce the liquid in the intestinal cavity itself on T2 signal (Fig. 4). MRCP demonstrated the cystic lesion had no communication with the duodenal lumen. But this modality could not clearly identify its communication with the bile duct. During follow-up dynamic contrast enhancement (DCE)-MRI with specific liver intake contrast agent revealed a slender tube connecting the lower common bile duct with the cystic lesion (Fig. 4). Finally the patient was diagnosed with choledochocoele based on imaging findings.

The operation was carried out. The diagnosis was confirmed by pathological findings. He was discharged on postoperative day 10 with good health condition.

DISCUSSION

Type III cyst is a rare type of CCs, characterized by a cystic dilatation of the bile duct terminal bulging into duodenal lumen.⁶⁻⁷ It is similar to DDCs, which also present as cystic structures and typically protrude into the second or third portion of the duodenum.⁸ DDCs are extremely rare^{6, 9} and account for 5% of all gastrointestinal duplications.¹⁰ DDCs are usually located in the mesenteric side and share their circumference with the intestine. They have a muscle coat and may or may not communicate with the gut lumen. Both CCs and DDCs can communicate with the bile duct. Except for connecting with the bile duct, DDCs can communicate with the pancreaticobiliary duct, especially with the pancreatic duct. They have much in common about clinical manifestations and imaging findings.¹¹

It is difficult to distinguish them using preoperative diagnostic procedures. The key to differentiating them is to determine the relationship between the lesions and the lumen as well as bile duct. Cholangiography and upper gastrointestinal barium studies have been proposed to differentiate them. Choledochocoele could be filled with contrast during cholangiography but not be filled during barium studies, whereas DDCs could not be filled when scanned with both studies.¹² It may be sufficient to identify larger lesions with the two methods, but for smaller lesions the sensitivity and specificity of the two techniques are low. In addition, cholangiography is invasive. Recently the commonly used imaging techniques are ultrasonography,

Figure 1. Endoscopy (A) revealed a parenteral cystic lesion (star) in the descending duodenum. Endoscopic ultrasonography (B) showed a cystic lesion in the duodenal lumen with many stones (star).

Figure 2. The upper gastrointestinal barium study displayed a round filling defect (arrow) in the second portion of the duodenum.

CT, MRCP and endoscopic retrograde cholangiopancreatography (ERCP) to verify CCs. Ultrasonography is economic, convenient and widely used, but it's extremely susceptible to interference of bowel gas. Multi-detector computed tomography (MDCT) is very helpful in surgical planning, especially in the accurate delineation of extent of the dilated bile ducts. MDCT can also identify cyst wall thickening and intracystic masses that develop secondary to malignancy. But it's difficult to display the relationship between cystic lesions and the bile ducts, duodenal lumen. MRCP and DCE-MRI well settle these problems with PJ and

Figure 3. Axial (A) and coronal (B) CT scan images of the abdomen revealing a cystic mass with multiple stones in the duodenum lumen (arrow). Coronal CT scan (B) showing a thin-walled cystic lesion (arrow) without nodules accompanied with mild duodenal dilatation.

Figure 4. Magnetic resonance cholangiopancreatography (MRCP) (A-C) and dynamic contrast enhancement (DCE)-MRI (D-F) images.

A. Pineapple juice can reduce the pre-existing gastrointestinal tract liquid (long arrow) in T2 signal and distinguish bowel water from liquid in the cystic lesion (short arrow) on T2WI.

B. MRCP showing multiple filling defects in the cystic cavity (arrow).

C. 2D MRCP.

D-F. DCE-MRI revealing the process of contrast medium excreted into the cystic lesion during discharge phase (arrows) and clearly displaying a slender tube (arrow, F) connecting the lower common bile duct with the cystic lesion.

hepatocyte-specific contrast agent. PJ has shorter T1 and T2 than other juices. After oral PJ, fluid pre-existing in the lumen is suppressed, but liquid in the cystic cavity is still bright on T2WI.¹³ It can clearly show the relationship of the cystic lesion with the lumen. The liver specific contrast agent excreted by the bile duct can clearly show the relationship between the cystic lesion and bile ducts. Despite ERCP remains the most widely used diagnostic tool for identification of choledochocoele, with a reported diagnostic sensitivity of 97%,⁵ it is invasive and the patients are difficult to tolerate. Nevertheless MRI is safe, non-invasive and no ionizing radiation. Furthermore, MRI does not carry the risks of ERCP including cholangitis, duodenal perforation, hemorrhage and pancreatitis. MRCP has distinct advantages to assess cyst lesions, size, site and shape of bile duct dilatation.¹⁴ The image quality of MRCP is more susceptible to motion artifacts than ERCP, which can be compensated by breath-hold sequences and respiratory triggered scanning.¹⁵ Sacher *et al*¹⁴ identified that MRCP has a 96%-100% detection rate for CCs, which making it the best choice for preoperative evaluation. MRI can entirely replace ERCP in assessing the relationship between the cystic lesion and bile duct, making guidance for surgical planning. This may benefit from PJ and liver specific contrast agent. It may be achieved in all patients with choledochocoele who cannot be verified by ERCP. After a series of tests, the patient was diagnosed as type III choledochal cyst by MRCP, and MRI with administration of the specific contrast agent of biliary tract confirmed the diagnosis. MRCP is noninvasive and convenient in the diagnosis of choledochocoele.

Conflicts of interest statement

The authors have no conflicts of interest to disclose.

REFERENCES

- de Vries JS, de Vries S, Aronson DC, et al. Choledochal cysts: age of presentation, symptoms, and late complications related to Todani's classification. *J Pediatr Surg* 2002; 37(11):1568-73. doi: 10.1053/jpsu.2002.36186.
- Jablonska B. Biliary cysts: etiology, diagnosis and management. *World J Gastroenterol* 2012; 18(35):4801-10. doi: 10.3748/wjg.v18.i35.4801.
- Bhavsar MS, Vora HB, Giriappa VH. Choledochal cysts: a review of literature. *Saudi J Gastroenterol* 2012; 18(4):230-6. doi: 10.4103/1319-3767.98425.
- Todani T, Watanabe Y, Narusue M, et al. Congenital bile duct cysts: classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. *Am J Surg* 1977; 134(2):263-9. doi: 10.1016/0002-9610(77)90359-2.
- Law R, Topazian M. Diagnosis and treatment of choledochocoeles. *Clin Gastroenterol Hepatol* 2014; 12(2):196-203. doi: 10.1016/j.cgh.2013.04.037.
- Koffie RM, Lee S, Perez-Atayde A, et al. Periapillary duodenal duplication cyst masquerading as a choledochocoele. *Pediatr Surg Int* 2012; 28(10):1035-9. doi: 10.1007/s00383-012-3116-4.
- Katabathina VS, Kapalczynski W, Dasyam AK, et al. Adult choledochal cysts: current update on classification, pathogenesis and cross-sectional imaging findings. *Abdom Imaging* 2015; 40(6):1971-81. doi: 10.1007/s00261-014-0344-1.
- Kim YJ, Kim YK, Jeong YJ, et al. Ileal duplication cyst: Y-configuration on *in vivo* sonography. *J Pediatr Surg* 2009; 44(7):1462-4. doi: 10.1016/j.jpedsurg.2009.03.034.
- Macpherson RI. Gastrointestinal tract duplications: clinical, pathologic, etiologic, and radiologic considerations. *Radiographics* 1993; 13(5):1063-80. doi: 10.1148/radiographics.13.5.8210590.
- Antaki F, Tringali A, Deprez P, et al. A case series of symptomatic intraluminal duodenal duplication cysts: presentation, endoscopic therapy, and long-term outcome (with video). *Gastrointest Endosc* 2008; 67(1):163-8. doi: 10.1016/j.gie.2007.08.006.
- Atkinson HD, Fischer CP, de Jong CH, et al. Choledochal cysts in adults and their complications. *Int Hepatol Pancreatol Biliary Assoc* 2003; 5(2):105-10. doi: 10.1080/13651820310001144.
- Mourra N, Chafai N, Bessoud B, et al. Colorectal duplication in adults: report of seven cases and review of the literature. *J Clin Pathol* 2010; 63(12):1080-3. doi: 10.1136/jcp.2010.083238.
- Arthurs OJ, Graves MJ, Edwards AD, et al. Interactive neonatal gastrointestinal magnetic resonance imaging using fruit juice as an oral contrast media. *BMC Med Imaging* 2014; 22(14):33. doi: 10.1186/1471-2342-14-33.
- Sacher VY, Davis JS, Sleeman D, et al. Role of mag-

netic resonance cholangiopancreatography in diagnosing choledochal cysts: case series and review. *World J Radiol* 2013; 5(8):304-12. doi: 10.4329/wjr.v5.i8.304.

15. Makin E, Davenport M. Understanding choledochal malformation. *Arch Dis Child* 2012; 97(1):69-72. doi: 10.1136/adc.2010.195974.