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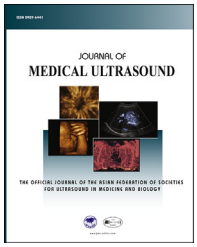

CASE REPORT

Type I Choledochal Cyst Complicated With Acute Hemorrhagic Pancreatitis: A Case Report

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Abstract Choledochal cysts rarely present with acute pancreatitis. We report a patient with type I choledochal cyst(s) who had concomitant acute frank hemorrhagic pancreatitis.

A 14-year-old male noted with a history of recurrent abdominal pain, fever and jaundice. Ultrasonography (US) of abdomen at the Emergency Department depicted distended gall bladder with wall thickening. Apparently dilated intrahepatic ducts (IHDs) and fusiform dilatation of the common bile duct (CBD), and mild dilatation of the pancreatic duct were also noted, suggesting a type I choledochal cyst( ). Computed tomography (CT) demonstrated calcifications in the uncinate process of the pancreas in addition to the similar findings on US. He subsequently underwent choledochal cyst excision with a Roux-en-Y hepaticojejunostomy. After surgical treatment, he has been doing well for 3 years.

Conflict of interest: The authors have no potential conflict of interest.

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Choledochal cyst is one of the most common congenital anomaly of the common bile duct (CBD) [1,2]. The clinical presentations of choledochal cysts vary from patient to patient, and may depend on the patients' age. The symptoms are more subtle and protean in adulthood than in childhood [2,3]. In children, the major manifestations include jaundice, right upper quadrant (RUQ) mass, and intermittent colic abdominal pain [3]. Pancreatitis, although has been described in the literature, is an uncommon complication of choledochal cyst [2—8]. Children may have subtle symptoms or acute pain, the pain can be related to relatively mild pancreatitis; however, acute pain caused by frank hemorrhagic pancreatitis is much rare [4]. We herein report such a case, his disease was diagnosed by using ultrasonography (US) and confirmed by computed tomography (CT) and the subsequent surgery.

Case report

A 14-year-old boy was brought to the emergency department (ED) by his parents, with a chief complaint of intermittent sharp abdominal pain for 2 days. According to the statement of the boy and his parents, he had past experience of frequent abdominal pain and received medication for pain control since 6 years old. Except for his episodes of abdominal pain since childhood, his past history was unremarkable: no diabetes, no hypertension, no hepatitis, no previous history of surgery or allergy. This time, he suffered from more severe sharp abdominal pain than before for two days. Due to increasing intensity of the pain, he was brought to our ED for evaluation. Physical examination revealed tenderness on the right upper quadrant of abdomen with positive Murphy's sign. His abdomen was soft on palpation with normal active bowel sounds. He had no fever, jaundice, nausea or vomiting during the past 2 days. US of abdomen at the ED was done, which depicted distended gall bladder with wall thickening. Apparently dilated intrahepatic ducts (IHDs) and fusiform dilatation of the common bile duct (CBD), and mild dilatation of the pancreatic duct were also noted, suggesting a type I choledochal cyst (Fig. 1). There were evidence of splenomegaly and intraabdominal fluid accumulation in both upper and lower abdomen. Laboratory data reported leukocytosis (WBC: 17,100/ul( ), normal: 4500—10,000/ul), CRP: 0.5 mg/dl (normal: <0.3 mg/dl), amylase: 385 IU/L (normal: 43—116 IU/L), lipase: 643 IU/L (normal: 13—60 IU/

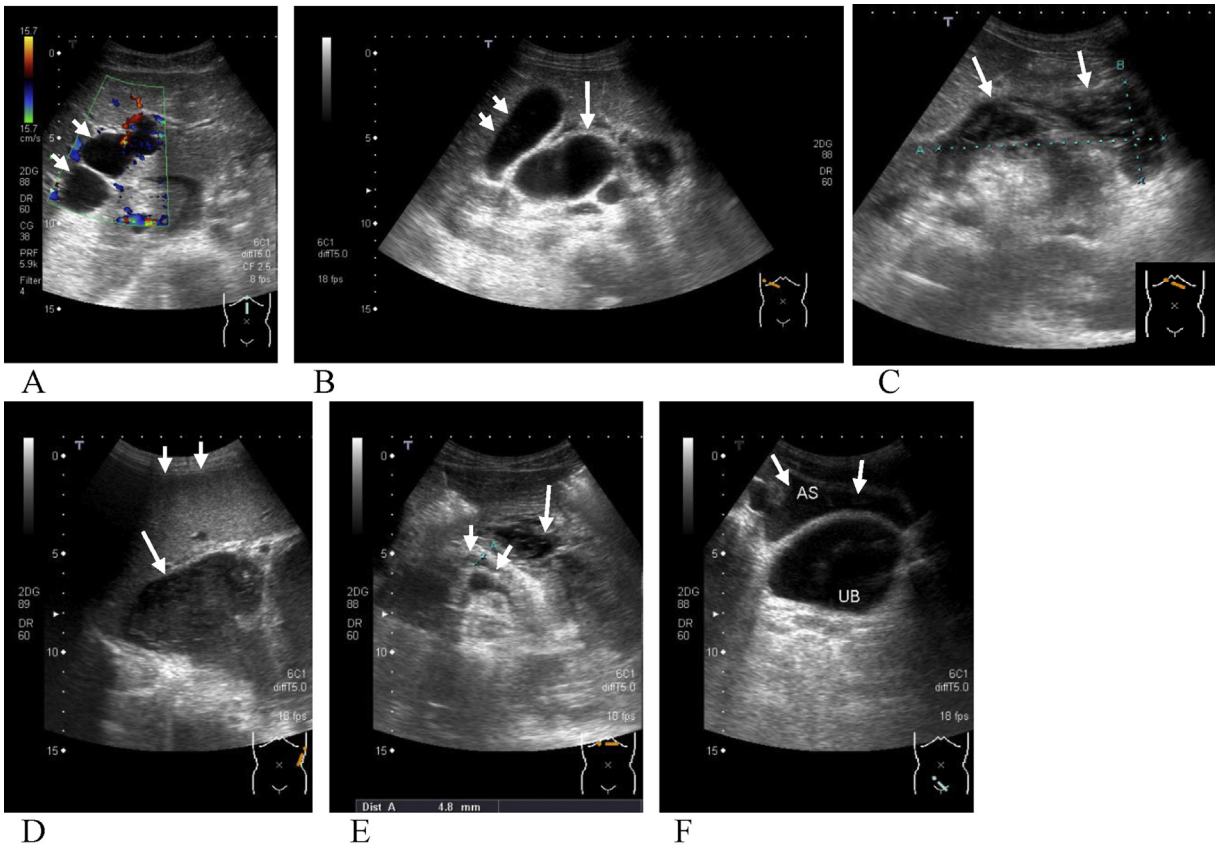

Figure 1 Ultrasonography of the abdomen. (A) Transverse scan of the liver shows dilated IHDs (arrows) in the hilar region (B) Oblique sagittal scan of the right upper abdomen shows mild distention of the gall bladder (small arrows) and apparently dilated CBD (large arrow) (C) Fluid collection in the lesser sac is noted (arrows), surrounding the lateral and inferior margins of the caudate lobe. (D) Similarly, echogenic fluid is evident in the left upper abdomen (large arrow), medial to the spleen (small arrows) (E) The pancreas is essentially in normal size (small arrows), showing mild heterogeneous in echopattern, and associated with mild dilated main pancreatic duct, suggestive of acute (chronic) pancreatitis. Large arrows = lesser sac fluid collection. (F) Pelvic fluid collection (arrows) adjacent to the urinary bladder.

L), Alk-P: 618 IU/L (normal: 104—338 IU/L). Under the impression of choledochal cyst, complicated with acute hemorrhagic pancreatitis, he was admitted to our surgical intensive care unit (SICU). CT was then done, and the diagnosis made by using US was confirmed. The pancreatic fossa was filled with hyperdense structures which were most likely due to mixed necrotic materials and blood clots (Fig. 2). He underwent exploratory laparotomy with total excision of CBD and gall bladder, choledochojejunostomy, and drainage of lesser sac hematoma. The surgical diagnosis was choledochal cyst (type I) caused by congenital anomaly with APBJ1. The abnormally long common channel was 1.7 cm. After surgery, he was sent to SICU for further intensive care. The post-operative course was smooth, he was then moved to surgical ward, stayed for another 2 weeks. After discharge, he had been followed at the out-patient department with US and CT studies for more than 2 years with stable condition, and now has been doing well for 3 years.

Discussion

Choledochal cyst is generally believed to be a congenital biliary disorder. It is characterized by benign cystic dilatations of the extrahepatic and/or intrahepatic biliary tree. The etiology of choledochal cyst can be multifactorial, including congenital anomaly of the biliary system and anomalous pancreatobiliary junction (APBJ) [1,2,9—14]. The so-called anomalous pancreas to biliary junction is defined as a long common channel with a length greater than 1.5 cm [1,2,9].

The vast majority of patients with choledochal cysts have APBJ [1,2,9]. When there is abnormal junction between the pancreatic and biliary ducts, the pancreatic secretions and enzymes reflux into CBD, and result in inflammation and weakening of CBD, which is subsequently dilated [9]. Some other experts proposed that choledochal cyst may be part of congenital anomaly of biliary system or multiple congenital anomaly [1,9,11].

Choledochal cyst is more prevalent in Asian than in Western countries, and more than 33% of all reported cases are from Japan. There is a female predilection, with a female-to-male ratio of 3:1—4:1. The majority (67—80%) of patients have been diagnosed before 10 years of age [1,2,9]. They are classified into 5 types according to Todani [15], i.e., 1. fusiform dilatation of the CBD; 2. diverticulum of the CBD; 3. Choledococele; 4. multiple segmental dilations of the biliary tree including CBD; and 5. Caroli disease.

The classical presentation of choledochal cysts is a triad of abdominal pain, jaundice and a palpable lump seen in

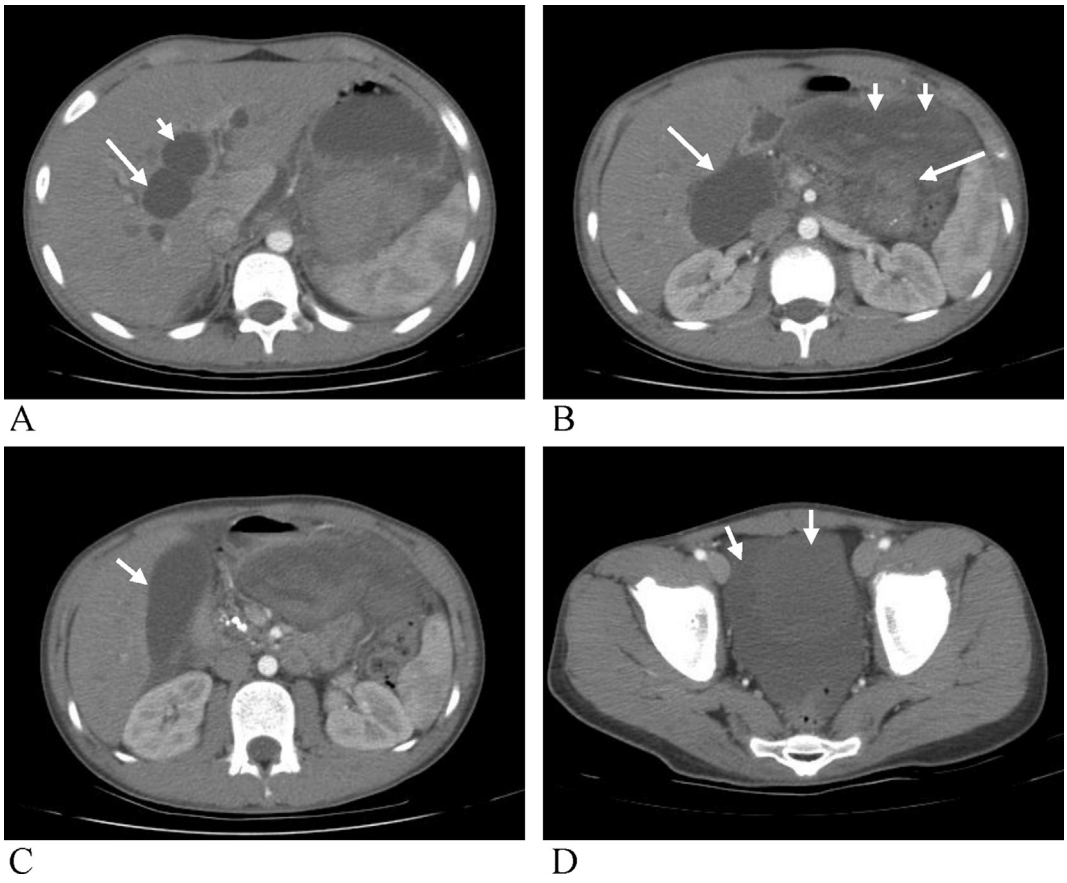

Figure 2 CT scan of the upper abdomen and pelvis also shows similar findings: (A) Dilatation of the IHDs (arrows) (B) Dilatation of the CBD (large short arrow) and relatively hyperdense fluid collection in the left upper abdomen (small arrows). Structures with soft tissue density in the pancreatic bed (large long arrow) are most likely due to mixed necrotic material and blood clots. (C) Mild distension of the gall bladder (arrow) (D) Pelvis fluid collection (arrows).

13—63% of patients [16]. Presentations of choledochal cysts vary from patient to patient, and may depend on the patients' age. The symptoms are more subtle and protean in adulthood than in childhood. In children, the major manifestations include jaundice, right upper quadrant (RUQ) mass, and intermittent colicky abdominal pain. While in adults, the triad of jaundice, abdominal pain and RUQ mass is noted in only 10—20%. The most common symptom in adult is abdominal pain. Some patients have only subtle symptoms. Morbidities of choledochal cyst include: biliary stone (80%), cholangitis, liver abscess, liver cirrhosis, portal hypertension, spontaneous rupture of bile duct, and pancreatitis [2—5]. Rarely, patients with choledochal cyst may develop cholangiocarcinoma [16], however, the reported rate of this malignancy in patients with choledochal cysts has been as high as 9—28%, about 25—40 times of incidence compared to normal population [16,17].

Lal et al. reported a large series of complicated choledochal cysts in 144 patients over 15 years but with a low occurrence of pancreatitis (1.4%). In their study, one patient with a choledochocele had repeated episodes of acute pancreatitis which were managed successfully with a transduodenal sphincteroplasty, and another patient had chronic pancreatitis [18]. Swisher et al. reviewed 32 adult patients who were treated for choledochal cysts. Thirty documented episodes of pancreatitis in 18 patients (56.3%) were noted; this was seen in all types of choledochal cysts and was not related to the age, gender or race of the patient. All eight patients with an abnormal pancreaticobiliary junction developed pancreatitis as compared to only two of the six patients with normal pancreatic duct anatomy ($P < 0.006$). In their series, only one patient with a type I choledochal cyst had chronic pancreatitis [19]. An anomalous anatomical arrangement of the pancreaticobiliary ductal junction (long common channel more than 15 mm) allows reciprocal reflux of bile and pancreatic juices into the biliary tree, which can cause inflammation, ectasia and dilatation [20,21]. Amylase can be found in the bile and may explain the increased incidence of cholangiocarcinoma and gall bladder carcinoma [22]. Similarly, the entry of bile into the pancreatic duct might lead to recurrent acute pancreatitis [23]. In search of the literature Gouda et al. identified only 11 published reports of chronic pancreatitis in a patient population treated for choledochal cysts of which six are confirmed cases of chronic calcific pancreatitis. Chronic pancreatitis is an uncommon association in patients with choledochal cysts [6]. Our young patient presenting acute pain caused by frank hemorrhagic pancreatitis is much rare.

In summary, the cross-sectional imaging modalities such as US, CT, MRI in conjunction with MRCP, are good tools for diagnosing choledochal cyst. MRCP may be competitive to ERCP in the diagnosis of biliary system disease. In this case, we used US, CT to make a correct diagnosis of choledochal cyst. The interpreters of hepatobiliary imaging studies should bear in mind the following normograms of the biliary anatomy; i.e., the normal CBD diameter (<0.7 cm), the normal IHD diameter (<0.3 cm at hilar region), and normal length of common channel (<1.5 cm), and the types of choledochal cysts so that a diagnosis can be made correctly when abnormal CBD feature is encountered. Acute pancreatitis is a rare occurrence in patients with type I choledochal cyst(s) and only six cases have been reported in the literature. Our patient with choledochal cyst(s) associated with pancreatitis was treated surgically with uneventful clinical course.

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