Unusual Presentations of Choledochal Cyst: Case Series and Review of Literature

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Abstract Congenital cystic dilation of the biliary duct is defined as choledochal cyst (CDC). CDC presenting with the classical triad of abdominal pain, lump, and jaundice is seen in only 6 % cases. Cyst excision with hepaticojejunostomy is the standard treatment worldwide. We hereby report five cases of CDC with unusual presentations (gastric outlet obstruction, cyst perforation, giant cystolithiasis, giant cyst, and mixed type) and discuss the challenges faced during the diagnosis and perioperative management of these cases.

Keywords Bile duct · Choledochal cyst · Cystolithiasis · Hepaticojejunostomy

Introduction

Congenital cystic dilation of the biliary duct is defined as choledochal cyst (CDC). Though most of the patients with CDC present in childhood, about 20 % present in adulthood [1]. The classical triad of clinical presentation—abdominal pain, jaundice, and lump—is present in only 6 % of cases [2]. Fifteen to 20 % of patients with CDC present with complications, of which cystolithiasis is the commonest complication. Other uncommon complications range from hepaticolithiasis, intrahepatic abscess, secondary biliary cirrhosis, portal hypertension, and cholangiocarcinoma [3]. Uncommon variants of CDC apart from the classical types as described by Alonso-Lej [4] have been reported in literature. Unusual types and clinical presentation pose difficulties in the

diagnosis and management. Herein, we report five cases of CDC who had unusual features on presentation.

Case Series

Of the 29 cases of CDC managed between January 2009 and December 2013, 5 (17.2 %) cases had unusual presentation.

Case 1

A 12-year-old girl presented with history of pain in the abdomen for 2 years and recurrent bilious vomiting for 7-months duration. Upper gastrointestinal endoscopy showed dilated stomach with external compression at the antropyloric region. Subsequently, contrast-enhanced computed tomography (CECT) was done, which showed type I CDC with gastric outlet obstruction (Fig. 1a). On exploration, dilated stomach with a large CDC (10×7 cm) compressing the antropyloric region was present (Fig. 1b). CDC excision with Roux-en-Y hepaticojejunostomy (RYHJ) was performed. Postoperative course was uneventful and patient is doing well after a follow up of two and half years.

Case 2

A 16-year-old female presented with complaints of abdominal pain for 2 months. Abdominal ultrasonography (USG) revealed a dilated common bile duct (CBD) suggesting CDC. Though surgery was advised on index admission, patient refused the same due to personal reasons. One month later, she presented with jaundice, fever, epigastric lump, and features of local peritonitis. Liver function test showed raised serum bilirubin, 17.25 mg/dl (normal range 0–1.4 mg/dl) and liver enzymes (serum aspartate transaminase, AST), 174 IU/L (normal range 10–50 IU/L); serum alanine transaminase

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Fig. 1 Contrast-enhanced computed tomography (CECT) (a) and intraoperative (b) image of a large choledochal cyst compressing the stomach causing gastric outlet obstruction. Perforated choledochal cyst with

bilioma in the supracolic compartment seen on CECT (c) and intraoperatively (d). Giant cystolithiasis as seen on endoscopic retrograde cholangiography (e) and following cyst excision (f)

(ALT), 152 IU/L (normal range 10–50 IU/L); serum alkaline phosphatase (ALP), 1421 IU/L (normal range 40–129 IU/L). Serum amylase was marginally raised (104 IU/L, normal range less than 96 IU/L). CECT showed type 4a CDC with a loculated collection in the supracolic region with the possibility of post acute pancreatitis fluid collection or contained CDC perforation (Fig. 1c). The collection was contained without evidence of generalized peritonism, hence patient was managed with intravenous antibiotics and USG-guided percutaneous drainage was tried, but it was dry. As patient responded to conservative management, early exploration was not done. Subsequent exploration after 2 weeks showed dense adhesions with a bilioma in the supracolic compartment. Type 4a CDC with cystolithiasis and perforation at the cystic duct-CBD junction was present (Fig. 1d). CDC excision, bilioma drainage, and RYHJ were performed. Patient had an uneventful postoperative course and is doing well after 3 years of follow up.

Case 3

An 18-year-old female presented with history of recurrent cholangitis and pain in right hypochondrium for 1 year. Liver function test showed normal serum bilirubin (0.5 mg/dl) but elevated liver enzymes (serum AST, 144 IU/L; serum ALT, 160 IU/L; serum ALP, 1559 IU/L). USG revealed cholelithiasis, dilated CBD (50×40 mm) with a large stone (50×40 mm) in its lumen. Endoscopic retrograde cholangiogram (ERCP) showed grossly dilated CBD with large stone within it, raising the suspicion of CDC (Fig. 1e). Endoscopic stenting was done in view of cholangitis. Subsequently, magnetic

resonance cholangio-pancreatogram (MRCP) was done which showed a type I CDC with large cystolithiasis. Perioperatively, a large CDC with a giant stone within it was found. Hepatoduodenal ligament (HDL) anatomy was distorted due to large stone within the CDC that precluded safe circumferential mobilization of the CDC. Stone was removed by longitudinal choledochotomy that facilitated subsequent complete mobilization of CDC (Fig. 1f). CDC excision with RYHJ was done with an uneventful postoperative course, and patient is healthy after 2 years of follow up.

Case 4

A 13-year-old girl presented with complaints of progressively increasing abdominal lump for 2 months and recurrent cholangitis. A large cystic, nontender lump was present in upper abdomen extending to right iliac fossa on per abdominal examination. USG revealed a hugely dilated CBD with bilateral intrahepatic biliary radicle dilatation (IHBRD). MRCP showed a 14×16×21 cm cystic lesion (CDC) extending up to right iliac fossa, displacing aorta and IVC to the left (Fig. 2a). On exploration, a giant CDC was present occupying the whole upper abdomen and right iliac fossa, displacing stomach, duodenum, and pancreas anteriorly (Fig. 2b). There was no evidence of portal hypertension. As the cyst was difficult to manipulate, hence, the cyst was decompressed by aspirating bile from it. Densely adhered posterior cyst wall was dissected off the portal vein. Because of the long standing pressure by the giant cyst, pancreas became atrophic. Pancreatic duct (PD) was opening into CBD 5 cm proximal to the ampulla of Vater, which resulted in an accidental opening of PD during

Fig. 2 Sagittal magnetic resonance cholangio-pancreatogram (MRCP) (a) and intraoperative image showing the giant choledochal cyst (b). Coronal MRCP (c) and intraoperative image (d) depicting the mixed type choledochal cyst

dissection. Complete excision of CDC was done including the intrapancreatic portion and biliary reconstruction done by RYHJ. Proximal end of pancreas was over sewn and distal end was drained by pancreaticojejunostomy. Postoperative period was uneventful except for grade A pancreatic leak which was managed conservatively. Patient is doing well after a follow up of 16 months.

Case 5

A 32-year-old male presented with history of pain in the right hypochondrium for 10 days and jaundice for 1 week. USG abdomen showed a dilated CBD (11 mm) with mild IHBRD. MRCP was done, which showed type 1 CDC (Fig. 2c). On exploration, fusiform dilatation of CBD was present with a diverticulum arising from its right side. Cystic duct was draining on to this diverticulum, suggestive of mixed type CDC (types I and II; Fig. 2d). Common hepatic duct was normal. Complete excision of CDC with RYHJ was done.

Patient had an uneventful recovery in the postoperative period and is doing well after a follow up of 6 months.

Discussion

Complications are seen in 15–20 % of patients with CDC, among which cystolithiasis, cholangitis, and pancreatitis are commonly reported. Apart from complications, CDC may present with unusual symptoms or types other than the classical types.

Few cases of gastric outlet obstruction in CDC have been reported in literature [3, 5–7]. Can et al. [5] described gastric outlet obstruction associated with a choledochocoele. This presentation is more common in childhood and has been reported in association with duodenal atresia and annular pancreas [6, 7]. Nearly all types of CDC have been reported to be associated with gastric outlet obstruction (Table 1).

Table 1 Cases of choledochal cyst presenting with gastric outlet obstruction

	Number of cases	Age (years)/sex	Type	Associated anomaly	Management
Lal et al. [3]	1	NR	Type IVa	None	Cyst excision with hepaticojejunostomy
Can et al. [5]	1	21/M	Type III	None	Cyst excision with reimplantation of intramural pedicle
Komuro et al. [6]	2	1. Neonate/male 2. Neonate/male	1. Type II 2. Type IVa	1. Annular pancreas 2. Multiple anomalies	1. Cyst excision, choledochojunostomy, and side-to-side duodeno-duodenostomy 2. Cyst-enterostomy
Iwai et al. [7]	1	4/F	Type I	Duodenal atresia in neonatal period	Cyst excision and hepaticojejunostomy
Current case	1	12/F	Type I	None	Cyst excision with hepaticojejunostomy

NR not reported

Complete excision of the CDC with RYHJ is the recommended treatment.

Spontaneous perforation is a rare complication of CDC with a reported incidence of about 2 %. Exact etiology is unknown, but is supposed to be the association of CDC with anomalous pancreatic duct–biliary duct junction (APBDJ) and reflux of pancreatic juice into the CBD resulting in weakness and rupture of the cyst wall [8]. However, some workers suggest abnormal rise of ductal pressure due to the blockage of common channel as the cause of spontaneous perforation [9]. Mural immaturity of the cyst wall may be the cause of higher incidence of perforation in infancy compared to older children [10]. The rarity in adults may be due to cyst wall thickening from repeated attacks of cholangitis. The most common site for perforation is the cystic duct–CBD junction. Patients may present with contained bilioma or free peritoneal perforation. Percutaneous drainage should be done in cases with bilioma, whereas a free perforation mandates peritoneal lavage with T-tube drainage of CBD. Although definitive excision of the cyst, in both circumstances, is performed after 6–12 weeks, there are few reports of single-stage treatment [11]. In our case, the bilioma was contained with thick wall and devoid of significant inflammation in HDL. Hence, we opted for a single-stage definitive treatment.

Cystolithiasis is the most common complication of CDC in adults with incidence up to 70 %. Stones in CDC are usually soft and pigmented, but the viscid bile may cause a large stone cast in the cyst. Large stone casts have been reported in type II CDC and in association with portal hypertension [12, 13]. Large stone cast may pose difficulties intraoperatively due to its size, pericystic adhesions from recurrent cholangitis, or associated portal hypertension. Extreme care is warranted during posterior dissection of the cyst to avoid portal vein injury.

Few reports of giant CDC are available in the literature which was managed by Roux-en-Y-choledochocyst-jejunostomy or RYHJ [14–16]. Preoperative detection of APBDJ may be difficult in a giant CDC. It may be encountered intraoperatively, as in our case, and may require pancreaticoduodenectomy. Aspiration of the large cyst may aid in its intraoperative manipulation and the dense pericystic adhesions cautions the posterior dissection especially from the portal vein and retropancreatic area. Any injury to pancreatic duct should be managed meticulously, either by pancreaticoduodenectomy or drainage of the distal pancreatic stump.

Todani et al. [17] modified the Alonso-Lej classification of CDC into five types. Several authors have reported unusual types of CDC apart from these five types, the most common being isolated cystic dilatation of cystic duct (type VI CDC) [18]. Another variant described in both adults and pediatric age groups are mixed types I and II, where there is fusiform dilatation of CBD with a lateral diverticulum [19–23]. The

diverticulum arises from the right side of the dilated CBD and there is a definable cystic duct between gall bladder and diverticulum. Preoperative diagnosis is easily missed due to overlap of cyst and the rarity of entity. Only four cases of mixed type have been reported in pediatric patients and five cases in adults. Though the inclusion of this variant in the Todani's classification is debated, the surgical treatment is not different from the classical types.

Conclusion

The current series highlight various unusual presentations of CDC, which may pose diagnostic dilemma and significant impact on its surgical management. One should be aware of these variations both in the preoperative and intraoperative period, or else inadvertent injury may occur with significant postoperative complications.

Conflict of Interest The authors declare no potential conflicts of interest.

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