Choledochal cyst of the proximal cystic duct: a taxonomical and therapeutic conundrum

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ABSTRACT

Isolated choledochal cysts involving the cystic duct are rare. We present a case of a choledochal cyst involving only the proximal cystic duct, and discuss the taxonomic and therapeutic challenges. There is a need for a clearly defined classification system for these cysts as they may be categorised as either type II or type VI cysts. The optimal treatment remains debatable, with some authors recommending a bilioenteric reconstruction owing to the wide cystic duct–bile duct junction. However, we suggest that a cholecystectomy should be performed with examination of the specimen and frozen section in case of any abnormality rather than upfront bile duct excision. In addition, given the rarity of this condition and the paucity of long-term data, we recommend meticulous follow-up for development of any malignancy.

KEYWORDS

Type VI choledochal cyst – Cystic duct cyst – Isolated cystic duct dilation – Cholecystectomy

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Choledochal cysts (CDCs) involving the cystic duct are a rare entity and dilations restricted to the cystic duct are rarer still. The limited literature on isolated cystic duct CDCs makes decisions on the management of these patients difficult. We present our experience of a patient with an isolated proximal cystic duct CDC, and then review the existing literature and debate the management options available.

Case history

A 29-year-old woman presented to our department having experienced a single episode of abdominal pain 4 weeks earlier. Ultrasonography revealed a type I CDC. Further evaluation with magnetic resonance cholangiopancreatography (MRCP) showed isolated dilation of the proximal cystic duct with a wide opening into the common bile duct (CBD), suggestive of a CDC of the cystic duct (Fig 1). The CBD appeared normal and there was no associated anomalous pancreatobiliary duct junction (APBDJ). This resulted in the dilemma of whether to offer the patient a simple cholecystectomy alone or to excise the CBD. Although a cholecystectomy would leave a large rent in the CBD and the standard of care in most CDCs is CBD excision, it was felt that excision would constitute overtreatment.

The patient was eventually listed for a cholecystectomy and intraoperative assessment of the CBD. The intraoperative findings replicated the findings on imaging. Only the proximal cystic duct was dilated, with a normal cystic duct beyond it draining the gallbladder. The dilated cystic duct had a wide opening into the CBD, with a diameter of 1.5cm

at its junction with the CBD. The CBD was 5mm in diameter. A simple cholecystectomy was performed. The cystic duct was divided flush with the CBD and the rent repaired primarily (Fig 2). The patient recovered well and was discharged on the second postoperative day. The histology was suggestive of CDC without any evidence of dysplasia.

Discussion

A total of 52 cases with cystic duct CDCs have been reported to date. However, most of these reports deal only with the radiological appearance of these cysts, with surgical management being described in only 24 patients. Among these, over a third of the cases ($n=9$, 38%) were associated with other anomalies of the biliary system, the most common being concomitant involvement of the CBD in the form of a type I or type IV cyst.¹

Only 15 cases of surgical management of isolated cystic duct CDC have been described (Table 1).^{2–16} A notable aspect is the relatively older age of patients with these cysts (median: 29.5 years, range: 4 months – 75 years, $n=15$) compared with patients with other types of CDC. Association with an APBDJ was seen in only two patients (13%) with isolated cystic duct CDCs. This incidence is much lower than that reported with other types of CDC (35–90.2%).^{17,18} There appears to be an interesting association between cystic duct CDCs and systemic diseases such as systemic lupus erythematosus, thyroiditis, goitre and diverticulosis.^{6,19}

Diagnosis and treatment of a cystic duct CDC associated with dilation of the intra and/or extrahepatic CBD is

Figure 1 Magnetic resonance cholangiopancreatography showing the gallbladder with a dilated cystic duct and a wide opening into the common bile duct. Note the normal diameter of the common bile duct proximally and distally.

straightforward and dictated by the associated lesion. In contrast, the management of isolated cystic duct cysts is controversial. These can be divided broadly into two groups: cases with a narrow (normal) cystic duct–CBD junction and those with a wide junction (such as in our case). The main challenge in diagnosing the cystic duct cyst preoperatively is in establishing the origin of the cystic lesion and delineating the CBD separately. Preoperative identification is difficult and many of these cases have been misreported (as CDC I, II or IV) on radiological workup. The accuracy of preoperative diagnosis is unsatisfactory in the reported literature, with less than half ($n=7$, 47%) of the patients having been diagnosed correctly prior to surgery.

The case of an isolated cyst of the proximal cystic duct is interesting not just because of its rarity but also because of the taxonomic and therapeutic possibilities that exist. At first, it may appear to be a simple case of what is now referred to as a type VI CDC.⁴ However, can one truly exclude the possibility of a type II CDC involving the mid-CBD at the site of insertion of cystic duct? Moreover, should one not classify isolated cystic duct CDCs separately from those associated with dilations of the CBD?

Detractors may contend that this is just academic curiosity and would not have clinical consequences. Nevertheless, the fact that the taxonomy has implications for uniformity in reporting, audit and data interpretation (especially in the current scenario of virtually non-existent literature) reflects the importance of an accurate,

Figure 2 Intraoperative photograph of cystic duct choledochal cyst (arrow) with normal common bile duct proximally and distally (yellow slings). The right hepatic artery (blue sling) was separated from the lesion. Inset shows operative specimen.

universally accepted classification. Furthermore, one cannot be sure that isolated cystic duct CDCs and those with CBD dilation represent the same entity and so classifying them separately would be prudent.

Aside from taxonomical issues, the optimal treatment poses an equally difficult challenge. The operative management of an isolated cystic duct cyst with a narrow cystic duct–CBD junction is fairly straightforward and cholecystectomy is the universally accepted treatment. Conversely, surgical treatment of isolated cystic duct cyst with a wide cystic duct–CBD junction (as in our case) is contentious. The options include either a simple cholecystectomy and excision of the cyst or concomitant excision of the CBD and bilioenteric anastomosis. One may argue in favour of CBD excision with bilioenteric reconstruction given the premalignant nature of CDCs and the fact that the whole of the biliary epithelium is at risk of neoplasia. In addition, a wide cystic duct–CBD junction would make closure of the CBD (which would be necessary if it was not being excised) technically difficult, and fraught with the possibility of postoperative leaks and future strictures.

The counterargument is that if the disease is limited to the cystic duct, removing the CBD could be unnecessary. Since the CBD is not dilated, the bilioenteric reconstruction would be similarly at risk of complications such as leak and stricture. Moreover, the delayed age at presentation (median: 29.5 years), frequent association with systemic illness and the fact that only around 13% of cases have been associated with an APBDJ (Table 1) raises the possibility of alternative pathologies contributing to the origin of an isolated cystic duct cyst. This is bound to influence what is perceived as complete treatment for the cyst. Presence of an

Table 1 Summary of literature on surgical management of isolated type VI CDCs

Author	Age / sex	Pain	Jaundice	Preoperative diagnosis	Final diagnosis	Cystic duct–CBD junction	Associated anomaly	Details of surgery
Bode, 1983 ²	7 F	-	+	CDC II	Intraoperative	Narrow	Biliary obstruction	Cholecystectomy, excision of cyst, bile duct exploration, duodenotomy and choledochoduodenostomy
Champetier, 1987 ³	49 M	+	-	Chronic cholecystitis	ERCP	Narrow	Gallstones	Cholecystectomy
Serena Serradel, 1991 ⁴	? F	+	+	CDC II	Intraoperative	Narrow		Cholecystectomy
Bresciani, 1998 ⁵	30 M	+	-	Chronic cholecystitis	Intraoperative cholangiography	Narrow	RHD joining the cyst	Laparoscopic cholecystectomy + division of RHD
Loke, 1999 ⁶	29 F	+	+	CDC VI	ERCP	Wide	SLE	Cholecystectomy + CBD excision + RYHJ
Weiler, 2003 ⁷	40 F	+	-	CDC VI	ERCP	Wide	APBDJ	Cholecystectomy + CBD excision + RYHJ
Manickam, 2004 ⁸	20 F	+	-	CDC VI	MRCP	Wide	APBDJ	Cholecystectomy
Iype, 2005 ⁹	33 F	+	-	CDC VI	MRCP	Narrow		Cholecystectomy
Chan, 2009 ¹⁰	41 M	-	-	CDC II	Intraoperative	Narrow	Gallstones	Laparoscopic cholecystectomy
Conway, 2009 ¹¹	52 F	+	-	CDC	Intraoperative	Narrow		Cholecystectomy
De, 2011 ¹²	18 F	+	+	CDC IV	Intraoperative	Wide		Cholecystectomy + CBD excision + RYHJ
Shah, 2013 ¹³	10 F	+	-	CDC VI	MRCP	Wide		Cholecystectomy + CBD excision + RYHJ
Tzedakis, 2013 ¹⁴	32 M	+	+	CDC VI	MRCP	Narrow		Laparoscopic cholecystectomy
Kaselas, 2016 ¹⁵	10 F	+	-	CDC I	Intraoperative	Wide		Cholecystectomy
Youn, 2016 ¹⁶	0.3 M	+	-	CDC VI	MRCP	Narrow		Cholecystectomy

APBDJ = anomalous pancreatobiliary duct junction; CBD = common bile duct; CDC = choledochal cyst; ERCP = endoscopic retrograde cholangiopancreatography; MRCP = magnetic resonance cholangiopancreatography; RHD = right hepatic duct; RYHJ = Roux-en-Y hepaticojejunostomy; SLE = systemic lupus erythematosus

APBDJ mandates excision of the CBD to disconnect it from the refluxing pancreatic juice. Besides, at least part of the risk of neoplasia results from the APBDJ and its absence may confer some protection.

Even though the literature is sparse and the conclusions that can be drawn from such scarce data are limited, it is notable that no case of malignancy has been reported arising in patients with isolated cystic duct cysts. Surgical management has been described in only six cases of an isolated cystic duct cyst with a wide cystic duct–CBD junction.^{6–8,12,13,15} Of these, four patients underwent excision of the CBD with bilioenteric anastomosis and two underwent cholecystectomy with primary CBD closure (Table 1).

In the absence of substantial literature on the long-term outcomes of management, operative options should be discussed with the patient preoperatively based on the evidence available, and possible short and long-term complications considered. A pragmatic approach would be to plan for a cholecystectomy with intraoperative assessment and frozen section of the cyst. Any involvement of the CBD, presence of abnormal mucosa in the cyst or intraoperative frozen section showing atypical cells would mandate CBD excision and bilioenteric reconstruction. In all other cases, a cholecystectomy with closure of the CBD (either primarily or over a stent) would suffice.

We believe that the category of type VI CDC should be reserved for isolated cystic duct dilation (whether proximal or in its entirety) and cysts with associated CBD dilation should be grouped according to the classification of the CBD cyst (which decides the management). These patients should be followed up for life or until there is concrete evidence regarding the risk of malignancy in this subgroup of patients.

Conclusions

Isolated CDC of the proximal cystic duct is an extremely rare entity, and presents a taxonomical and therapeutic dilemma. On the basis of the literature available, we suggest a simple cholecystectomy is performed in the absence of any

high risk features. Patients should be kept on follow-up surveillance for a malignancy arising in the future.

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