Management of Complicated Choledochal Cysts

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Key Words

Choledochal cyst · Cholangitis · Hepatolithiasis ·
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Abstract

Objectives: The presentation of choledochal cysts (CDCs) is altered by complications such as acute severe cholangitis, hepatolithiasis, spontaneous perforation, portal hypertension, pancreatitis, malignancy of the biliary tract and previous surgery in the form of internal drainage of the cyst. The management and the outcome of such complicated cysts differ from that of an uncomplicated CDC. This presentation is an analysis of our experience with complicated CDCs. **Patients and Methods:** One hundred and forty-four patients with CDCs were managed at a tertiary level referral hospital in North India, between January 1989 and June 2004. Thirty-three (23%) CDCs were associated with complicating factors: severe cholangitis requiring a biliary drainage procedure prior to definitive operative procedure (n = 11), spontaneous perforation (n = 3), hepatolithiasis (n = 6), acute cholecystitis (n = 1), recurrent acute pancreatitis (n = 1), chronic pancreatitis (n = 1), gastric outlet obstruction (n = 1), portal hypertension (n = 6), biliary tract malignancy (n = 4) and previous internal drainage of the cyst (n = 5). Five patients had more than one complication.

The management outcome of complicated CDCs was compared with that of uncomplicated CDCs. **Results:** Complicated CDCs were significantly more common with type IV-A anatomy. Endoscopic, percutaneous or external surgical biliary drainage procedure was performed in 14 complicated CDCs prior to cyst excision. Desired definitive surgical procedure could be performed in 26/33 (79%) patients with complicated CDCs as compared to 107/111 (96%) patients with uncomplicated CDCs. Four (12%) patients with complicated CDCs had early postoperative complications as compared to 9/111 (8%) patients with uncomplicated CDCs. During a median follow-up of 13 months (2 months to 8 years), 2 patients with complicated CDCs died due to advanced secondary biliary cirrhosis and advanced gall bladder cancer, respectively. Six patients reported complications. Of the 6 patients, 3 required reoperation for a strictured hepaticojejunostomy (n = 2) and hepatolithiasis (n = 1). In contrast, there was no disease-related mortality and only 1 out of 111 patients with uncomplicated cysts had a complication during follow-up. **Conclusion:** Complicated CDCs merit a carefully planned management strategy including percutaneous, endoscopic and surgical procedures. The management may have to be staged. The outcome depends on the nature of complication and the management strategy adopted.

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Fig. 1. Classification of CDCs.

Introduction

Choledochal cysts (CDCs) commonly present with pain abdomen, jaundice and abdominal lump in various combinations [1, 2]. However, the presence of various cyst-related complications may alter the classical presentation and also influence the subsequent management and outcome. Few reports have specifically addressed the problems associated with the management and outcome of complicated CDCs [3, 4]. This study highlights our experience with complicated CDCs. We have also recently reported our experience with 35 cases of type IV-A CDCs treated between January 1989 and December 2002 [5].

Patients and Methods

Data on 144 patients with CDCs, managed between January 1989 and June 2004, at a tertiary level referral hospital in North India, were analyzed retrospectively. There were 76 adults and 68 children in this group. The cyst types were: type I ($n = 86$), type III ($n = 1$), type IV ($n = 56$) and the 'forme fruste' variant ($n = 1$). Figure 1 gives the diagrammatic representation of various cyst types. There were 21 adults (64%) and 12 children (36%, 4 infants) ranging in age from 4 months to 60 years with a female preponderance (19/13; m:f = 0.7:1). Thirty-three of these 144 (23%) cases presented with complications and these were: acute suppurative cholangitis warranting a preoperative biliary drainage ($n = 1$), acute cholecystitis ($n = 1$), spontaneous cyst perforation ($n = 3$), hepatolithiasis ($n = 6$), recurrent acute pancreatitis ($n = 1$), chronic pancreatitis ($n = 1$), gastric outlet obstruction ($n = 1$), portal hypertension

($n = 6$), biliary tract malignancy ($n = 4$) and previous surgery in the form of internal drainage of the cyst ($n = 5$). The duration of symptoms ranged from 3 months to 30 years (median 2 years). The imaging studies used were: abdominal ultrasonography ($n = 33$), contrast-enhanced computed tomographic scan of the abdomen ($n = 9$), endoscopic retrograde cholangiopancreatogram ($n = 8$), magnetic resonance cholangiopancreatogram (MRCP; $n = 4$), and intraoperative cholangiogram ($n = 2$).

Statistical analysis was done using the Fisher's exact test. Statistical significance was defined as $p < 0.05$.

Results

The cyst types in 33 patients with complicated CDC were: type I ($n = 11$), type III ($n = 1$) and type IV ($n = 21$). Complications found to be associated with the different anatomical cyst types are listed in table 1.

Acute suppurative cholangitis ($n = 11$), unresponsive to intravenous antibiotics, required preoperative biliary drainage in all patients in the form of a percutaneous transhepatic biliary drainage ($n = 10$) and a surgical drainage with a T-tube choledochocystostomy ($n = 1$). One patient died of cholangitis and septicemia leading to multisystem organ failure despite a percutaneous biliary drainage. Cyst excision was performed successfully 6–12 weeks later in 8 of the 10 remaining patients. Two patients did not report for definitive surgery after a biliary drainage procedure.

Table 1. Complicated CDCs and cyst anatomy

Nature of complication	Patients	Cyst type
Acute suppurative cholangitis	11	I (n = 3) IV-A (n = 8)
Acute cholecystitis	1	IV-A (n = 1)
Spontaneous perforation	3 ^a	I (n = 1) IV-A (n = 2)
Hepaticolithiasis	6 ^a	IV-A (n = 6)
Recurrent acute pancreatitis	1	III (n = 1)
Chronic calcific pancreatitis	1	IV-A (n = 1)
Gastric outlet obstruction	1	IV-A (n = 1)
Portal hypertension	6 ^a	I (n = 2) IV-A (n = 4)
Biliary tract malignancy	4	I (n = 4)
Previous internal drainage	5 ^a	I (n = 1) IV-A (n = 4)

^a More than 1 complication was present in 5 patients.

Spontaneous cyst perforation presented as bile peritonitis (n = 2) and localized subhepatic biloma (n = 1). The initial management was a percutaneous drainage for the biloma (n = 1) and a T-tube choledochocystostomy and peritoneal lavage for bile peritonitis (n = 2). Cyst excision (n = 3) was performed 6–12 weeks later.

All 6 patients with hepaticolithiasis had type IV-A anatomy with a history of recurrent cholangitis. One patient required preoperative biliary drainage for the same. Cyst excision was performed in all 6 patients. One of these 6 patients subsequently required a left lateral segmentectomy because of recurrent cholangitis consequent to intrahepatic cystic dilatations and calculi largely limited to the left lateral segment.

Recurrent episodes of acute pancreatitis presented in 1 patient with choledochocoele. Endoscopic deroofting had to be abandoned in this patient because of bleeding, and the patient was then successfully managed surgically with transduodenal sphincteroplasty.

One patient had associated symptomatic chronic pancreatitis with a drainable pancreatic duct on MRCP. Hence, a lateral pancreaticojejunostomy along with cyst excision and roux-en-y hepaticojejunostomy was performed.

Another patient presented with gastric outlet obstruction due to extrinsic compression by the cyst which resolved following cyst excision.

Portal hypertension (n = 6) manifested as hematemesis (n = 2), refractory ascites (n = 3), and with ascites, jaundice and cholangitis (n = 1). The cause of portal hyperten-

sion was secondary biliary cirrhosis (n = 5) and extra hepatic portal vein thrombosis (EHPVO) probably secondary to recurrent cholangitis (n = 1). Shunt surgery for portal decompression was attempted in the patient with EHPVO but was unsuccessful. Cyst excision was performed successfully in 3 out of 6 patients. In 2 patients, an internal drainage of cyst was resorted to because of excessive bleeding from the collaterals in the hepatoduodenal ligament. One patient did not report for definitive surgery after a percutaneous biliary drainage for recurrent severe cholangitis.

Four patients, all presenting with jaundice, were detected to have an associated biliary tract malignancy at presentation. These were: locally advanced gall bladder cancer (infiltrating into the liver and hepatoduodenal ligament; n = 2); lower end cholangiocarcinoma (n = 1), and an ampullary carcinoid tumor (n = 1). Cholecystectomy and internal drainage (n = 2) were performed for gall bladder carcinoma; cyst excision was not done due to the advanced stage of malignancy. The other 2 patients successfully underwent a pylorus-preserving pancreaticoduodenal resection (PPPD).

Five patients presented after a previous internal drainage of the CDC (choledochocystoduodenostomy in 4, and choledochocystogastrostomy in 1 patient) performed elsewhere. All 5 patients, presented with recurrent pain, jaundice and cholangitis. Two patients required a preoperative biliary drainage for control of cholangitis refractory to antibiotics. Cyst excision was performed successfully in all 5 patients.

The overall management approach is summarized in table 2. The desired surgical procedure could be performed in 26 patients (79%). These were: cyst excision (n = 23), PPPD (n = 2) and transduodenal sphincteroplasty (n = 1) for choledochocoele.

One patient died of acute myocardial infarction in the postoperative period. Nonfatal complications were observed in 4 (12%) patients. These were: hepaticojunostomy leak (n = 3) and a low output controlled external pancreatic fistula (n = 1). The bile leak responded to percutaneous biliary diversion in all 3 cases, while the controlled external pancreatic fistula closed spontaneously.

Follow-up information was available for 24 (77%) patients (range 2 months to 8 years, mean 22 ± 25 months, and median 13 months) in the complicated cyst group. While on follow-up, 2 patients died of advanced gall bladder cancer and liver failure, respectively. Of the remaining 22 patients, 4 patients who had had total cyst excision developed recurrent cholangitis on account of anastomot-

Table 2. Management of complicated CDCs

Complication	First stage surgical/nonsurgical intervention	Definitive surgical procedures
Acute severe cholangitis (n = 11)	Endoscopic/percutaneous transhepatic biliary drainage (n = 10) Surgical external drainage (n = 1)	Cyst excision (n = 8) ^a
Hepaticolithiasis (n = 6)	Percutaneous transhepatic biliary drainage (n = 1) ^b	Cyst excision (n = 6)
Spontaneous cyst perforation (n = 3)	External biliary drainage of the cyst (n = 2) Percutaneous drainage of biloma (n = 1)	Cyst excision (n = 3)
Acute pancreatitis (n = 1)	Endoscopic papillotomy (failed)	Transduodenal sphincteroplasty (n = 1)
Chronic calcific pancreatitis (n = 1)	nil	Cyst excision + lateral pancreaticojejunostomy (n = 1)
Gastric outlet obstruction	nil	Cyst excision (n = 1)
Portal hypertension (n = 6)	Splenectomy + devascularization (n = 1) Percutaneous cholecystostomy (n = 1) Percutaneous transhepatic biliary drainage (n = 1) ^b	Cyst excision (n = 3) Internal drainage of cyst (n = 2) (1 patient lost to follow-up)
Biliary tract malignancy (n = 4)	nil	Cholecystectomy + internal cyst drainage (n = 2) Pancreaticoduodenal resection (n = 2)
Previous internal drainage (n = 5)	Percutaneous transhepatic biliary drainage/endoscopic drainage (n = 2) ^b	Cyst excision (n = 5)

^a One mortality despite percutaneous drainage; 2 cases lost to follow up after a percutaneous drainage.

^b Also included in the acute severe cholangitis group.

ic stricture (n = 2) and hepaticolithiasis (n = 2). Three of them underwent reoperation: revision of hepaticojejunostomy (n = 2) and left lateral segmentectomy (n = 1). The fourth patient with cholangitis responded to antibiotics. The patient with EHPVO also developed recurrence of cholangitis after internal drainage of the cyst. This was managed by endoscopic biliary stenting. One patient, who underwent a PPPDR along with cyst excision for an associated ampullary carcinoid tumor developed stenosis of the pancreaticojejunostomy anastomosis

3 years after the resection. A pancreaticogastrostomy was performed at revision surgery.

In patients with uncomplicated cysts (n = 111), there were 2 early postoperative deaths. The causes were intraoperative portal vein injury (n = 1) with subsequent coagulopathy because of massive blood transfusion and biliary anastomotic leak leading to sepsis (n = 1). Nonfatal complications occurred in 9 (8%) patients. These were: bilioenteric anastomotic leak (n = 5, biliary diversion in 1), external pancreatic fistula (n = 2, closed spontane-

Table 3. Comparison of complicated with uncomplicated CDCs

Parameter	Complicated cyst (n = 33)	Uncomplicated cyst (n = 111)	p value
Type IV-A anatomy	21 (64)	35 (31)	0.001*
Cyst excision	26 (79)	107 (96)	0.003*
Internal drainage procedures	5 (22)	4 (4)	0.02*
Early postoperative complications	4 (12)	11 (10)	0.35
Delayed complications (including mortality)	8 (33)	1 (0.9)	0.0001*

Figures in parentheses indicate percentages. * p value significant.

ously) and intra-abdominal bleed (n = 2, 1 required re-exploration).

Follow-up was available for 76 (70%) patients with uncomplicated CDCs (range 1 month to 11 years, mean 17 ± 24 months, median 12 months). There were no disease-related deaths during follow-up. One patient developed anastomotic stricture extending into the right ductal system for which a right hepatectomy was performed.

The patients with complicated CDCs were compared with those with uncomplicated cysts (table 3). Complicated CDCs were frequently of type IV-A anatomy; at surgery, complete excision was less likely, and these patients had internal drainage procedures more often. At follow-up, disease-related mortality and long-term complications were more common in the complicated CDCs group.

Discussion

Complicated CDCs were more frequently of type IV-A anatomy in this series, and this has also been observed in the literature [3, 6, 7]. Type IV-A cysts by virtue of their intrahepatic cystic component and intrahepatic bile stasis, are more prone to develop complications [8].

Complications related to CDCs have been broadly divided into infective and noninfective complications [3]. Amongst the infective complications, fulminant cholangitis is common, especially in adults [9, 10]. Failure to respond to intravenous antibiotics should prompt an early percutaneous or endoscopic biliary drainage; external surgical drainage of the cyst may occasionally be required when a percutaneous or an endoscopic procedure fails to provide adequate biliary decompression, as was experienced in one of our patients. Excision of the CDC in presence of acute suppurative cholangitis has been reported to be associated with high morbidity and mortality. Staged

management in the form of an initial biliary decompression followed by cyst excision 6–12 weeks after the control of cholangitis, has been found to give a satisfactory outcome [3, 11–14].

Spontaneous perforation of CDC is rare, reported in 1–2% of cases [15, 16]. The etiopathogenesis of these perforations has been attributed to a sudden rise in intracystic pressure as a result of the blockage of the common channel by a protein plug, compounded by weakening of the cyst wall by recurrent chemical cholangitis secondary to long-standing reflux of pancreatic juice into the biliary system [17, 18]. These patients present with bile peritonitis and the diagnosis is confirmed by bile aspiration on abdominal paracentesis together with an ultrasonographic documentation of a CDC. The recommended management is emergency laparotomy, peritoneal lavage and external biliary drainage preferably by a T-tube inserted into the CDC [17]. The perforation may sometimes present as a localized bile collection (biloma), which may be managed by percutaneous drainage. Complete cyst excision with a bilioenteric reconstruction performed 6–12 weeks after the drainage procedure gives a satisfactory outcome as was observed in our patients.

Hepaticolithiasis is more often associated with intrahepatic cystic dilatations with or without downstream congenital ductal stenosis [7]. An intraoperative cyst endoscopy is a valuable aid in detecting and washing off the intrahepatic ducts of calculi and debris [7].

For residual intrahepatic calculi, a 'jejunal access limb' provides a useful access for subsequent choledochoscopic clearance of the intrahepatic biliary tree. Percutaneous transhepatic cholangioscopy and lithotripsy is another useful approach to the management of intrahepatic calculi [7]. Hepatic resection may be indicated for calculi and cysts limited to one part of the liver causing recurrent cholangitis and segmental fibrosis. One of our patients did require a left lateral segmentectomy for the same purpose.

The association of acute or chronic pancreatitis with CDC has been explained by an underlying anomalous pancreaticobiliary junction which permits free intermixing of pancreatic and biliary juices and protein plugs in the common channel. The incidence of acute pancreatitis with CDCs has been variously quoted to range from 0 to 70% [19–21]. The true incidence of pancreatitis with CDC is difficult to decipher from the literature because serum amylase or lipase levels are not routinely done for pain abdomen in patients with CDC, and the definition of pancreatitis in the setting of CDC is variable. Todani et al. [22] described patients with CDC who had recurrent bouts of abdominal pain and mild elevation in serum amylase but no evidence of pancreatitis at surgery. This ‘psuedopancreatitis’ and hyperamylasemia was associated with high biliary amylase levels and attributed to pancreaticobiliary reflux. While most episodes of pancreatitis are mild and can be managed conservatively, recurrences can only be eliminated by cyst excision and bilio-pancreatic disconnection [19–23].

In patients with associated symptomatic chronic pancreatitis, treatment options include: cyst excision combined with a lateral pancreaticojejunostomy if the pancreatic duct is adequately dilated or pancreaticoduodenectomy for predominantly pancreatic head pathology [24–28].

Choledochoceles, per se, are rare but are more often known to present as pancreatitis; the incidence in the literature varies from 19 to 45%. The treatment of choice in the current era is endoscopic deroofting of the choledochocoele together with an adequate sphincterotomy to permit free drainage of the pancreaticobiliary tree. Surgery is indicated for large choledochoceles, suspicion of a mass lesion within the cyst or failure of endoscopic therapy as was the indication in our patient [29].

Portal hypertension in CDCs may be secondary to biliary cirrhosis, compression of the portal vein by the CDC or portal vein thrombosis [30–32]. Excision of the CDC in the presence of portal hypertension is a major and hazardous undertaking due to the presence of extensive collaterals in the hepatoduodenal ligament and may be associated with increased postoperative morbidity and mortality [33]. Cyst excision may have to be abandoned in favor of internal drainage in these patients [3, 6, 34]. Decompression of the portal venous system in the form of a portosystemic shunt may be required before the cyst can be approached for excision, especially in patients who have bled from the varices [35]. In the current series, cyst excision could be achieved in only 3 of 6 patients, while internal drainage had to be performed in 2 patients. Pa-

tients with advanced biliary cirrhosis and portal hypertension would eventually merit liver transplantation.

A malignancy of the biliary tract is associated with CDC in 2.5–26% of cases, and carries a dismal prognosis on account of delayed diagnosis, advanced stage at presentation and multicentricity of the lesion [36–41]. This translates into fewer radical resections, and even after radical resection, most patients succumb within 2 years of the diagnosis [42, 43].

It is estimated that 13–69% of patients who have had an internal drainage procedure would need reoperation for abdominal pain, jaundice, recurrent cholangitis, calculi, pancreatitis and portal hypertension because of strictured cystenterostomy and stasis within the retained cyst [6, 34, 44, 45]. It is also estimated that the risk of malignancy in the retained cysts may be as high as 50% [15]. Reoperation and excision of the cyst in these patients may not be an easy task on account of extensive fibrosis and/or portal hypertension. Excision of the cyst was, however, performed successfully in all 5 patients with previous internal drainage in our series. It has been recommended that cyst excision should be performed in all patients who have had a prior internal drainage, even if asymptomatic [46–48].

Management strategy for complicated CDCs needs meticulous planning and multimodality approach. The therapeutic strategy needs to be tailored to an individual case. Percutaneous and endoscopic biliary drainage procedures are useful adjuncts to the surgical management, which itself may have to be a multistaged process. The outcome is satisfactory in patients with appropriately managed infective complications. The presence of portal hypertension or malignancy may preclude a technically satisfactory cyst excision in many instances, and the ultimate prognosis is largely governed by the progression of liver pathology or malignancy.

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