Congenital Type 1C Choledochal Cyst: Clinical Presentation and Surgical Treatment

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Abstract Choledochal cysts are uncommon developmental anomalies involving intrahepatic and extrahepatic biliary ducts, and their immediate complete excision is recommended to prevent chronic inflammatory and malignant changes. A diagnosis of a congenital type 1C choledochal cyst was made in an 8-year-old female patient admitted to the emergency unit with complaints of severe upper abdominal pain and vomiting. Under laparotomy, resection of the whole segment of choledochal cyst and gallbladder, assisted Roux-en-Y hepaticojejunostomy was performed. The laparotomy approach involving fine dissection and resection of choledochal cysts with Roux-en-Y hepaticojejunostomy remains superior to the endoscopic approach.

Consent A written informed consent was obtained from the patient for publication of this case report and accompanying images.

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Introduction

Choledochal cysts are rare developmental anomalies involving either extrahepatic or intrahepatic biliary ducts, or both [1–3]. Epidemiologic studies of choledochal cysts show higher frequency in females than in males (3:1 to 4:1), with remarkably high incidence of 1 in 1,000 births among Asian population compared to incidence of 1 in 1,00,000–1,50,000 in Western countries [4, 5]. About two thirds of diagnosed cases of choledochal cysts in Asia occur in Japan [6].

Vater [7] was first to provide an anatomical description of the choledochal cyst. Alonso-Lej et al. [8], based on analysis of 96 reported cases of choledochal cysts, have devised three-category classification and their therapeutic strategies. However, Todani et al. [9] have refined this classification system to include five categories, which become the most acceptable and applicable by most authors.

Etiology of these cysts remains under speculation, as many theories and explanations have been proposed [1]; however, the Babbitt's theory [10] of choledochal cysts formation remains the most popular and accepted. This theory attributes the cyst formation to abnormal pancreaticobiliary duct junction (APBDJ) outside the ampulla of Vater, resulting in a long common channel between the bile duct and the main pancreatic duct allowing the reflux of pancreatic juice into the bile duct. This reflux results in activation of pancreatic enzymes, causing inflammation and weakness of bile duct wall leading to cyst formation.

The present study outlines a case of congenital choledochal cyst involving large segment of bile and common hepatic ducts presented with severe abdominal pain, fever, and mild jaundice. The applied surgical procedure is described in detail

and is compared with other surgical techniques endorsed in the treatment of these congenital choledochal cysts.

Case Report

An 8-year-old female patient with history of repeated attack of upper abdominal pain and bouts of vomiting for the past 2 years was admitted to the emergency unit at Prince Hamza Hospital, Amman, Jordan, during February 2011 with complaints of sever upper abdominal pain radiating posteriorly to interscapular area with attacks of vomiting, yellow discoloration of skin, mild itching, and dark color urine.

On clinical examination she looked sick, cachexic, and febrile with forward-bending posture at the trunk. Mild ting of yellow discoloration of skin and conjunctiva was noticed, and on palpation, sever tenderness in the epigastrium and right hypochondrium was elicited with nonpalpable liver. She was admitted to the pediatrics unit as a case of acute pancreatitis. Laboratory investigations revealed normal serum amylase level, elevated serum levels of direct bilirubin, alkaline phosphatase, ALT, and AST liver enzymes (data not shown).

Magnetic resonance cholangiopancreatography (MRCP) revealed 10×7 cm fusiform cystic swelling of the common bile duct extending into the common hepatic duct with the normal size gallbladder and cystic duct (Figs. 1 and 2). The intrahepatic biliary ducts were slightly dilated, and a diagnosis of type 1C choledochal cyst was concluded. The patient was kept on IV fluid, antibiotics, and analgesic waiting for surgical interference.

Laparotomy with upper right transverse incision was performed and a large fusiform cystic swelling involving the common bile duct and extending into the common hepatic duct up to a level above the opening of cystic duct was

Fig. 1 a and b MRI views of choledochal cyst in a 10-year-old girl prior to surgical excision. The cyst is fusiform-shaped (type 1C) involving the bile and common hepatic ducts. ChC = choledochal cyst; CHD = common hepatic duct; RHD = right hepatic duct; LHD = left hepatic duct; DU = duodenum; CBD = common bile duct

Fig. 2 Cross-sectional MRI view of the choledochal cyst. ChC = choledochal cyst; GB = gallbladder; RK = right kidney; LK = left kidney

observed. The gallbladder, cystic duct, and liver appeared normal. The gallbladder bed was dissected and freed after ligation of cystic vessels to assist dissection and freeing the cyst from portal vein and hepatic artery. The cystic swelling was dissected from porta hepatis down to retroduodenal part of the bile duct. The cystic bile duct was ligated proximal to the junction with the main pancreatic duct, excised and reflected up toward the porta hepatis. The whole segment of the cystic bile duct, common hepatic duct, cystic duct, and gallbladder was excised. The jejunum 40 cm below the ligament of Trietz was transected and the ostium of distal segment was closed using Vicryl 4/0 continuous sutures. The closed-end distal segment of the jejunum was lifted to porta hepatis and sutured with the ostium of the remaining part of the common hepatic duct (hepaticojejunostomy) by end-to-side interrupted full suture using Vicryl 4/0 (Fig. 3).

The ostium of the proximal cut segment of the jejunum was sutured end-to-side with the distal segment of the jejunum (Roux-en-Y procedure) using Vicryl 4/0 interrupted single-layer seromuscular sutures (Fig. 3). The abdominal incision was closed without drain, and the excised specimen was sent for histopathology. The histopathologic examination of the excised tissue confirmed the diagnosis of a benign choledochal cyst, involving common bile and hepatic ducts with normal gallbladder tissue (Fig. 4).

Postoperatively, the patient recovered unremarkably; the pain and jaundice disappeared and he was discharged on the fifth postoperative day.

Fig. 3 Schematic drawing of the applied Roux-en-Y hepatoenterostomy surgical procedure

Fig. 4 Light microscopic view of the excised choledochal cyst **a** showing fibromuscular tissue partially lined by simple epithelial mucosa with no malignant changes. The gallbladder tissue **b** appeared normal (a and b $\times 10$)

Discussion

Choledochal cysts are congenital anomalies of the biliary tract being more common among Asian population [4–6]. Developmentally, biliary ducts develop from a hepatic diverticulum in the ventral wall of caudal end of primitive foregut early in the fourth week of intrauterine [11, 12]. This small diverticulum will develop into extrahepatic and intrahepatic biliary ducts, gallbladder, and ventral pancreas. Recanalization of the lumen of common duct starts at the end of the fifth week and moves slowly distally. By the sixth week, the common duct and the ventral pancreatic bud rotate 180° clockwise around the duodenum. Early in the seventh week, the bile and pancreatic ducts end in closed cavities of the duodenum. The muscle of the sphincter of Oddi develops from a concentric ring of mesenchyme surrounding the preampullary portion of bile and pancreatic ducts.

Etiology of choledochal cysts remains speculative; however, the pathogenesis is most probably multifactorial. In majority of patients with choledochal cysts, a developmental abnormality at the junction between the common bile duct and the main pancreatic duct can be demonstrated. This occurs when the pancreatic duct empties into the common bile duct more than 1 cm proximal to the ampulla of Vater [10].

Miyano and Yamataka [6] have documented such abnormal junctions in 90–100 % of patients with choledochal cysts. They attributed the formation of choledochal cysts to the reflux of activated pancreatic proenzymes into the common bile duct which cause damage and weakness of the bile duct wall. Defects in the epithelialization and recanalization of developing bile ducts, and congenital weakness of the duct wall due to distal aganglionosis have also been implicated [13, 14].

Choledochal cysts have been classified according to the system published by Todani et al. [9] into five major classes:

1. Type I cysts are the most common and represent about 80–90 %. They consist of saccular or fusiform dilatations

of the common bile duct, which involve either a segment of the duct or the entire duct. This type is further subdivided into the following:

- Type IA is saccular in shape and involves either the entire common bile duct or the majority of it.
 - Type IB is saccular and involves a limited segment of the bile duct.
 - Type IC is more fusiform in shape and involves most or all of the bile and common hepatic ducts.
2. Type II choledochal cysts appear as an isolated diverticulum protruding from the wall of the common bile duct with or without involvement of the common hepatic duct.
 3. Type III choledochal cysts arise from the intraduodenal portion of the common bile duct and are alternately termed “choledochocele”.
 4. Type IV cysts are multiple saccular dilatations of either intrahepatic or extrahepatic bile duct, or both. They are subdivided into the following:
 - Type IVA cysts consist of multiple dilatations of the intrahepatic and extrahepatic bile ducts.
 - Type IVB cysts consist of choledochal cysts which are multiple dilatations involving only the extrahepatic bile ducts.
 5. Type V or Caroli disease consists of multiple dilatations limited to intrahepatic bile ducts.

In our present case, the excised cyst represented the category of type 1C choledochal cyst. Shah et al. [15] in a retrospective study of 32 children and 47 adults, with choledochal cysts in regard to their presentation, management, histopathology, and outcomes, found that a history of biliary surgery, pancreatitis, cholangitis, early postoperative complications, and late postoperative complications occurred more frequently in adults than in children; the classic triad of abdominal pain, jaundice, and palpable right upper abdominal

mass were more frequent in children than in adults; fibrosis of the cyst wall was peculiar to children; and the signs of inflammation, hyperplasia, and long-term complications were more frequent in adults. From these findings, they concluded that choledochal cysts of children and those of adults should be considered as separate entities.

Several surgical procedures have been employed in the treatment of choledochal cysts [16–19]. These procedures vary between laparotomy and laparoscopic approaches; however, laparoscopic complete cyst resection, assisted Roux-en-Y reconstruction, and hepaticojejunostomy remain the procedure of choice by majority of surgeons.

In conclusion, we aimed to resect extrahepatic biliary ducts as much as possible preserving the last segment of the bile duct joining the main pancreatic duct. The procedure applied reduces the risk of subsequent malignant changes in the remaining stump. Others have advocated a procedure leaving a longer stump of common hepatic duct with mucosal resection as an alternative measure of protection against cancerous changes. Our personal experience and view suggest that a longer de-mucosal stump is more liable to malignant changes, and stump stricture is quite likely to occur.

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Conflict of Interest All authors have no conflict of interest

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