

MANAGEMENT OF ADULT CHOLEDOCHAL CYSTS: A 10 – YEAR EXPERIENCE

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Abstract: The aim of this paper is to show the different diagnostic procedures and treatment, in patients diagnosed with choledochal cysts. Choledochal cysts are congenital anomalies of the bile ducts and include cystic dilatation of the extrahepatic and intrahepatic biliary ducts or both. The study shows six patients, diagnosed as having type Ic choledochal cysts, according to the Todani classification. Diagnosis was established by clinical and radiographic findings including: ultrasound (US), magnetic resonance cholangiopancreatography (MRCP) and intraoperative cholangiography (IOC). All patients underwent surgical treatment with a total cyst excision, cholecystectomy and Roux-en-Y hepaticojejunostomy, followed by a good and uneventful recovery. Total cyst excision is recommended for reducing cyst related complications and risk of cholangiocarcinoma. Also, a long term review, is necessary due to recurrent cholangitis or pancreatitis, that may occur in patients with these cystic lesions. The epidemiology, diagnosis, surgical treatment, and risk of cancer in choledochal cysts were reviewed according to relevant published literature.

Keywords: choledochal cyst, ultrasound, magnetic resonance cholangiopancreatography, intraoperative cholangiography, hepaticojejunostomy

INTRODUCTION

Choledochal cysts are congenital anomalies of the bile ducts and are defined as abnormal, disproportionate, and cystic dilatation of the bile duct. The incidence of bile duct cysts ranges from 1 in 13.000 to 1 in 300.000 live births, with more than 60% of them being diagnosed in the first 1 to10 years of life. ^[1 - 4] Although the clinical picture can be vague and nonspecific, the diagnosis is facilitated by modern imaging techniques.

MATERIALS AND METHODS

In a retrospective review of all adults presenting with choledochal cysts to our department, between 2006 and 2015, medical and operative records for these patients were reviewed. Detailed analyses were made of the clinical presentation, radiological and biochemical findings, anatomical anomalies, management, complications and outcomes.

Investigation of the patients was dictated by their presentation, but generally involved ultrasound (US), magnetic resonance cholangiopancreatography (MRCP) and intraoperative cholangiography (IOC). All patients underwent surgical treatment with a total cyst excision, cholecystectomy and Roux-en-Y hepaticojejunostomy followed by a good and uneventful recovery.

RESULTS

Six patients (4 women and 2 men) were identified, aged between 25 and 57 years old. According to the modified Todani classification all our patients had type Ic cysts (see Table 1).

All patients presented with symptoms of recurrent abdominal pain, weight loss, nausea, and occasional vomiting. The time duration of symptoms before referral ranged between 1 month and 10 years. These symptoms

evolved episodically, with periods of calm, alternating with those of emphasized symptoms (usually after eating difficult to digest foods or after intellectual/physical intense effort) and were relatively unresponsive to drug therapy.

On general physical examination there were no significant changes, except the poor nutritional status caused by weight loss.

Abdominal ultrasound revealed for four of our six patients a well defined collection, with homogenous content within the projection of the pancreatic head (see Figure 1).

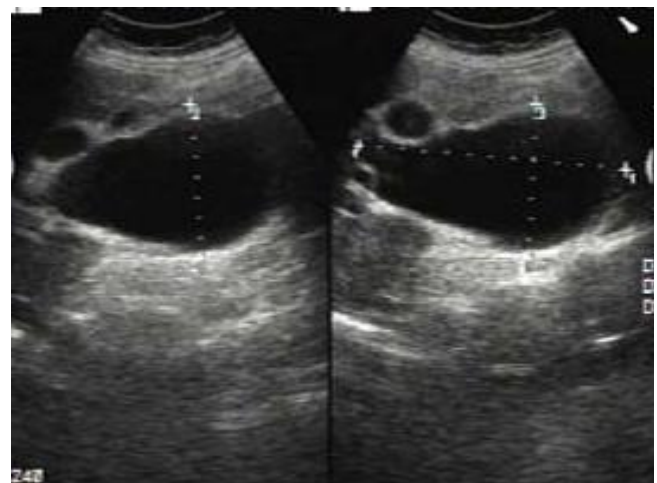


Figure 1: Ultrasound examination showing choledochal cyst.

Magnetic resonance cholangiopancreatography was performed in 3 cases and confirmed the existence of cystic formations, with fluid and fine sediment, overlapping the common bile duct topography, just below the union of the

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right and left hepatic ducts, imprinting the adjacent tract segment and the cephalic pancreas (see Figure 2).

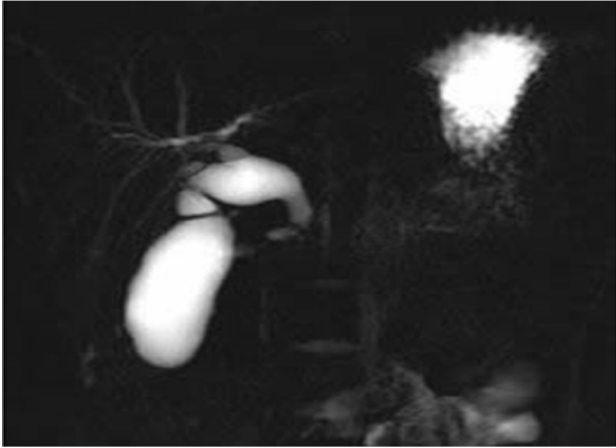


Figure 2: Magnetic resonance cholangiopancreatography showing choledochal cyst.

All patients underwent elective definitive surgery in our unit, comprising cyst excision, cholecystectomy and Roux-en-Y hepaticojejunostomy.



Figure 3: Gallbladder – resection piece.

For one patient the intraoperative findings were particular: after releasing the underside of the liver by adherentolysis, a monstrous sized bile duct was found (10-12/6-7 cm in diameter, double a normal intestinal loop). The walls of the bile duct were much thickened and stiff, and the morphological parietal changes gradually diminished in proximal direction, toward the hepatic hilum, where the walls became slimmer, suppler. These macroscopic findings were later confirmed by the microscopic and electro – optical examinations. The liver had steatotic aspect and the gallbladder was pear-shaped, normally sized, with thickened walls, fleshy, with the cyst discharging into the cystic bag (see Figure 3 - 5). Anterograde cholecystectomy was performed, followed by a puncture cholangiography. The images obtained showed bile duct dilatation, the transition of contrast into the duodenum, but - despite the fact that there were injected 40 ml of Odiston - intrahepatic bile ducts were not opacified (see Figure 6).

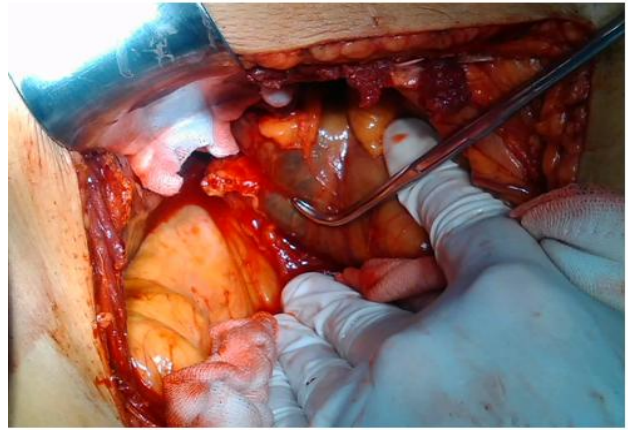


Figure 4: Choledochal cyst – intraoperative aspect, after cholecystectomy

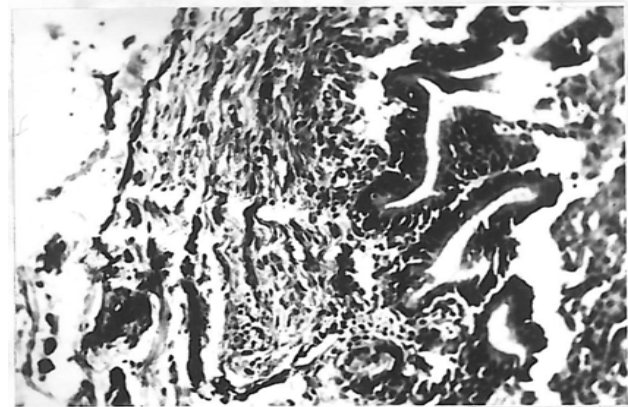


Figure 5: Microscopic aspect of the intrahepatic bile duct (Van Gieson method): you can see the normal structure of the mucosa, inflammatory infiltration of the lumen and the edematous thickening of the duct wall.

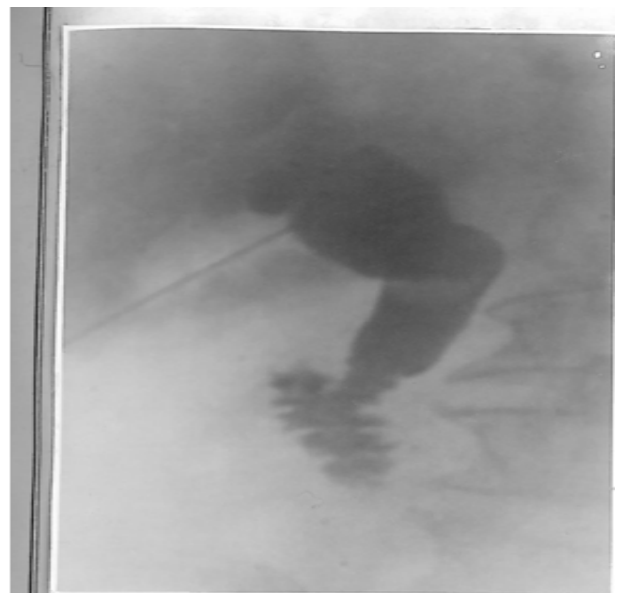


Figure 6: Choledochal cyst – intraoperative cholangiography.

We decided to completely remove the choledochal cyst. The choledoch was circularly isolated and then immediately sectioned above the duodenum. The distal end was closed with transfixing wire. The dilated bile duct was dissected by the proximal end of the other elements of the hepatic pedicle; the dissection becoming difficult in the middle segment of the common bile duct, with risk of infringement of the portal vein, common bile duct was sectioned 1 cm below the junction of the two hepatic channels and there were discharged approximately 80-100 ml of turbid bile, with muddy, brown sediment. Proximal stump was spotted with wires attached, starting the dissection in reverse, up to 2-3 cm from the place where it was initially interrupted. Further dissection being risky, through a longitudinal incision, the channel was transformed into a gutter, after which the choledochal wall was maximally removed, in plain sight, leaving in place a previous blade, whose mucosa was removed by skinning, scraping.

Postoperative recovery of all patients was uneventful and patients were discharged after 10 to 14 days, with a follow-up period of 1 to 8 years.

DISCUSSION

The first description of a choledochal cyst was made by Vater in 1723 and it wasn't until 1959, when Alonso-Lej et al. published the first systematic description of choledochal cysts, based on the clinical and anatomic findings in 96 cases. They classified choledochal cysts into three types and described the therapeutic strategies for each type. This classification was further modified by Todani and colleagues, who included five major types and several subtypes as seen in Table 1.^[4 - 9] The most frequent type encountered is the first one (80% of patients), also confirmed by our series in which all cases had a type Ic anomaly.

Table 1: Classification of choledochal cysts
(as proposed by Alonso-Lej and modified by Todani)

Type I	Dilatation of extrahepatic biliary ducts. Type I cysts may be further classified as cystic (Ia), focal (Ib) or fusiform (Ic).
Type II	Diverticulum of the extrahepatic biliary ducts.
Type III	Cystic dilatation of the intramural portion of the common bile duct, known as <i>choledochoceles</i> .
Type IVa	Lesions that involve both intra- and extrahepatic portions of the bile ducts.
Type IVb	Multiple cysts limited to the extrahepatic bile ducts.
Type V	Synonymous with <i>Caroli disease</i> , the lesions are abnormalities of the intrahepatic bile ducts resulting in multiple segmental intrahepatic cystic biliary dilatations.

Believed to be congenital in origin, the exact etiology of choledochal cysts remains unknown. Multiple etiologic theories have been proposed for the origin of this pathology. The embryological theory proposed by Yotsyanagi in 1936, suggested that the appearance of this type of lesion is determined by an unknown event during weeks 8 to 12 of intrauterine life. Although it was scientifically proven to some extent, the theory could only be valid for the dismorphic type and not for all encountered situations.

The most widely accepted theory is that cystic dilatation of the bile ducts is related to an anomalous pancreaticobiliary ductal union.^[9] This anomalous union, between the pancreatic duct and the common bile duct, located far from the duodenum results in a long common channel that allows the reflux of pancreatic secretion into the biliary tract, consequently causing enzymatic destruction of the mucosa, which favours inflammation, ectasia and ultimately dilatation. The theory is strongly supported by the presence of amylase in bile but it cannot account for the minority of cases with normal ductal entry into the ampulla of Vater.

Another plausible etiological theory for choledochal cysts is represented by obstruction of the common bile duct, a theory that was suggested by experimental animal studies. In neonatal animal models, ligation of the common bile duct causes a dilatation, morphologically resembling a type I choledochal cysts, whereas in adults, generalized dilatation of the whole biliary system can be seen.^[10-12]

This paper does not intend commenting on these theories, which are widely reported in valuable works, that analyze factual material appreciably in number and variety. Note however that all theories have a strong hypothetical character, thus not allowing a general consent on the means used in prevention of these defects. On the other hand, their interpretation by all authors as inherited distortions could be in the near future, the premise for solving them through genetic engineering.

In terms of the clinical presentation, the classical triad of jaundice, right upper quadrant mass and abdominal pain is present in only a minority of patients (0-17%). It is more commonly seen in children than in adults, and 85% of the children have at least two features of the triad at presentation, compared with only 25% of adults. Other presenting features of choledochal cysts are cholangitis, pancreatitis and biliary peritonitis from cyst rupture.^[13]

Ultrasound (US) is the first-line imaging investigation that can guide diagnosis, even from early intrauterine life. US allow the evaluation of the entire intrahepatic and extrahepatic biliary system and gallbladder, and can also demonstrate the associated complications such as cystolithiasis, cholangitis, and malignancy. Other important imagistic investigations are computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP) and endoscopic retrograde cholangiography (ERCP), the last two being the diagnostic methods of choice for biliary ductal pathology.^[14] Intraoperative cholangiography (IOC) states the lesion type and offers important guidance in the surgical approach.^[15,16]

A very important step in the management of choledochal cysts is identifying any associated hepatobiliary pathology. The most common accompanying condition in adults with choledochal cysts is cystolithiasis, followed by pancreatitis, cholangitis, portal hypertension and malignancy.^[17,18]

The surgical management of choledochal cysts is based on the cyst type and associated hepatobiliary pathology. In general, all bile duct cysts should be excised and bile flow is re-established by mucosa-to-mucosa biliary enteric anastomosis. The treatment of choice for type I bile duct cysts in adults is total ablation of the cyst and cholecystectomy followed by Roux-en-Y hepaticojejunostomy. The advantages of this procedure are as follows: low risk of malignancy by cyst excision; reduced incidence of anastomotic strictures; and it ensures the transit of bile through an intestinal segment that has peristaltic movement towards the digestive tract, without the possibility of reflux into the bile ducts.

Treatment of type II cysts depends on size of neck of the cyst at junction with the common bile duct, as in these cases the neck may be closed primarily or with T-tube decompression of the common bile duct.

Until recently, the treatment of choice for choledochoceles was transduodenal cyst excision with or without sphincterotomy but lately, endoscopic sphincterotomy and cyst unroofing has become the treatment of choice.^[13]

The extrahepatic component of type IVa and IVb cysts is approached as a type I choledochal cyst. The extent of the resection in type IVa cysts is controversial. Some authors advocate management by excision of the extrahepatic component, only with hepaticoenterostomy but when the intrahepatic disease is localized it would seem reasonable to perform the relevant partial hepatectomy. The same partial hepatectomy is practiced for Caroli disease.^[13]

The laparoscopic approach should be also considered for all types of choledochal cysts.^[19, 20]

CONCLUSIONS

Although considered a rare condition, choledochal cysts must be mentioned in the context of a rudimentary bile syndrome, occurring in children, young adults and rarely in old age.

Clinical manifestations are nonspecific in adults.

Among imagistic examinations, abdominal ultrasound has indicative value, but usually other methods are required (MRCP, ERCP, CT). Intra-operative cholangiography allows correct evaluation of the biliary tree and uncertain anatomical situation.

Surgery, the only effective management, must pursue, as much as possible, the removal of the lesion due to the risk of complications, malignancy being the most feared.

The monitoring of operated patients is required throughout their whole life, given the possibility of late complications.

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