CASE REPORT

Experience of surgical management of rare cases of choledochal cyst in last five years—tertiary care reflection

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Abstract

Choledochal Cyst (CC), also known as the biliary cyst, is one of the rare inherited anomalies of intrahepatic and/or extrahepatic biliary system characterised by varying degrees of cystic dilatation of the biliary tract without acute obstruction. The prevalence ranges from 1 in 13,000 people to 1 in 2 million people with preponderance in Asia[1], particularly in Japan. Moreover, the presentation also varies in children and adults, and is usually vaguer and non-specific in adults. The prevalence is even lower in males, with female to male ratio being 3:1-4:1[2]. We present here three cases of adult choledochal cysts excised in our surgical unit in the last five years. We discuss the aetiopathogenesis, presentation, diagnosis, surgical treatment, and complications of choledochal cysts based on the available literature. It is crucial to establish a multidisciplinary group of professionals, that comprises paediatric surgeons, pathologists, paediatric gastroenterologists, physiotherapists, nutritionists, oncologists, and radiologists, to get acceptable outcomes in diagnosing and treating children with choledochal cysts.

Keywords: Choledochal cyst, Roux enY Anastomosis, Cholangiocarcinoma, RHC mass.

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Introduction

Choledochal Cyst (CC), also known as biliary cyst, is one of the rare inherited anomalies of intrahepatic and/or extrahepatic biliary system characterised by varying degrees of cystic dilatation of the biliary tract without acute obstruction. The prevalence ranges from 1 in 13,000 people to 1 in 2 million people with preponderance in Asia, particularly in Japan. The condition was initially explained by Vater and Ezler in 1723, and very few cases have been reported since then. More than 70 percent of the cases present during the first decade of life and only rarely during adulthood. Moreover, presentation also varies in children and adults; it is usually vaguer and non-specific in adults. The prevalence is even lower in males, with female to male ratio being 3:1-4:1. We present three cases of choledochal cysts excised in our North Surgical Unit at Mayo Hospital during the last five years. One is of a 14-year-old boy and the other of a 22-year-old girl, both of whom did not have any feature except pain and tenderness in the epigastrium and the right hypochondrium. The third case was of a 13-year-old girl. All the cases were published after the consent of their guardians. The aetiopathogenesis, presentation, diagnosis, surgical treatment, and complications of choledochal cysts based on the available literature are also discussed.

Case 1

A 14-year-old boy was admitted to the surgical unit due to complaint of pain in the epigastrium and right hypochondrium since the last three days. The pain was sudden in onset, of severe intensity, radiating to the arms, and was associated with nausea, vomiting, and fever. There was no history of jaundice but the patient reported having similar pain one year back which was relieved with

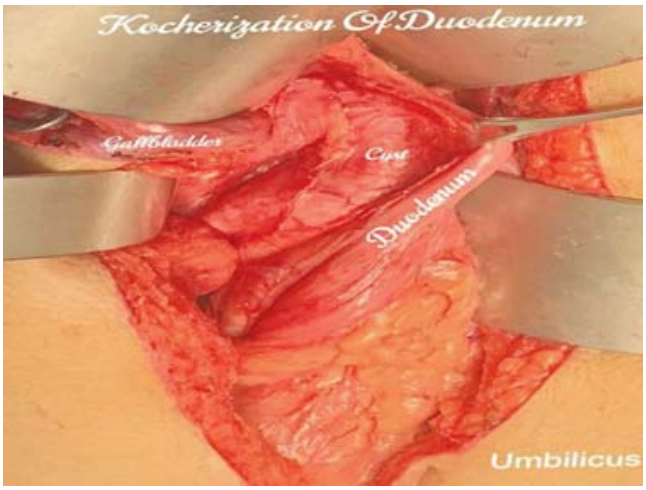

Figure-1: Kocherisation of the Duodenum.

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Figure-2: Hepaticojejunostomy

medication. On physical examination, there was tenderness in the epigastrium and the right hypochondrium but no palpable mass was noted. Subsequent ultrasound and MRCP reports revealed a tapering dilatation of the proximal as well as mid part of the common bile duct which measured 64x46x46mm; hence, a diagnosis of Choledocal cyst type I was established. Accordingly, the cyst was excised and Hepatico-jejunostomy with end-to-side anastomosis was done (figs # 1,2). He had unremarkable post-operative recovery with good clinical outcome and was discharged on the 13th day after surgery.

Case 2

A 22-year-old female patient was admitted in our surgical unit due to exacerbated epigastric pain over the last three months. History dated back to one year before her admission with the onset of pain. The pain was gradual in onset, radiating to the back, had been increasing progressively, and was associated with multiple episodes of vomiting with no signs and symptoms of jaundice. On physical examination, the patient was conscious with good vital signs and tenderness over the right hypochondrium and the epigastric region. A diagnosis of Choledochal Cyst Type I was established based on MRCP. Exploratory laparotomy showed a cystic dilatation of the common bile duct extending from 1cm below the hepatic confluence to just above the head of the pancreas and measured 8x10cm. Subsequently, it was excised along with the gallbladder, and Roux-enY Hepaticojejunostomy with enteroenterostomy of the jejunum was performed. She had an uneventful post-operative recovery and was discharged on the 12th day after surgery.

Case 3

A 13-year-old girl was admitted to our surgical unit with pain in the abdomen for the last four months. She had a few episodes of vomiting also but no fever. The pain was felt in the epigastric region, and was non-radiating and generalised. She never had jaundice in the past. On examination, her vital signs were stable and she was comfortable in the bed; she had swelling in the abdomen which was around 10x8cm in the epigastric region, non-tender and moving with respiration. It was diagnosed as Choledochal cyst type 1 on MRCP. The cyst was excised, along with Roux–en-Y hepaticojejunostomy reconstruction. It was around 10x15cm cyst extending from the confluence of hepatic duct up to the common bile duct. The patient had a smooth recuperation and was discharged on the eighth postoperative day.

Literature Review and Discussion

Cystic dilation of the biliary system can affect any region, starting from the liver to the duodenum. In 1959, Alonso-LEJ et al divided it into three types depending on the place of development[3]. Later in 1977, Todani et al added two more categories of CCs to this categorisation, resulting in the current five-category categorisation[4]. This categorisation is made on the basis of cholangiographic

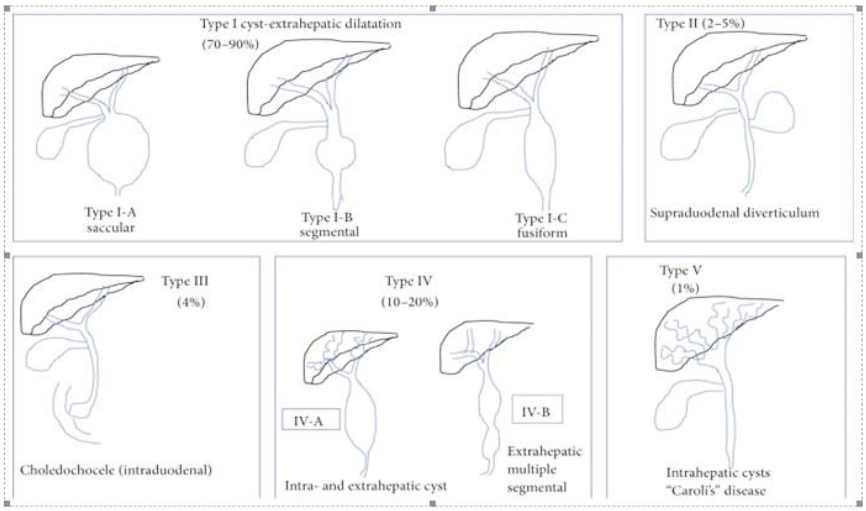

Figure-3: Todani's classification of Choledochal Cyst

anatomy, site and several intrahepatic and extrahepatic biliary tree cysts.It can involve the extrahepatic bile duct (types I, II, and III), the intrahepatic bile duct (type V), or both (type IVa), with type I being the most common (80-90 %) (Fig # 3).

Some specialists, however, criticise the categorisation for being focussed on the simplistic anatomy of the biliary tract and not focussing more on aetiology. The genesis of a choledochal cyst is still unclear, which has resulted in the creation of several ideas[5]. The most widely accepted explanation is Babbitt's, which states that an abnormal pancreaticobiliary junction (APBJ) causes cyst formation [6]. APBJ occurs due to the merging of biliary and pancreatic ducts 1 to 2cm proximal to the Oddi sphincter. The formation of a long passage (>15mm) makes way for the backflow and blending of pancreatic and biliary secretions, hence triggering pancreatic enzymes. This increase in intraductal pressure contributes to biliary tree dilatation, inflammation, epithelial injury, chromosomal instability, and cancer[6].

According to certain research, high amounts of amylase and higher amounts of phospholipase A2 and trypsinogen in CCs bile strengthen this notion[7]. Some specialists, however, disagree with Babbitt's notion, claiming the presence of APBJ in only 50% to 80% of the cases and also reflux was not present in antenatally diagnosed CCs[6]. Another idea, on the other hand, proposes an incomplete occlusion of the bile duct, resulting in greater proximal bile duct pressure and ultimate dilatation, first of the extrahepatic segment and then of intrahepatic component[8].

It has also been proposed that type II (real CBD diverticulum) and type III (choledochocele) CCs are caused by biliary duplication cysts and biliary or duodenal duplication cysts, respectively[9]. An obstruction at the ampulla of Vater is thought to result in a distal intramural common bile duct localised dilatation[10]. Caroli disease, commonly known as type V CC, is an autosomal recessive condition thought to be caused by ductal plate embryological distortion[11]. Caroli disease is linked to biliary atresia because the pathophysiology of ductal plate deformity is similar, but it is also linked to autosomal recessive polycystic kidney disease (PKD) and, less commonly, autosomal dominant PKD[12].

CCs can be silent for many years until being detected until imaging examinations for an unrelated condition are performed. Clinical signs differ between children and adults[13]. Children were more likely than adults to have the traditional triad of jaundice, discomfort in the right hypochondrium, and a palpable lump (85 percent versus 25 percent, respectively[2]. The majority of the youngsters present with only two of these three symptoms. Abdominal discomfort is the most prevalent clinical manifestation in adults[14]. Sludge and stone development owing to bile stagnation, repeated cholangitis, and pancreatitis owing to pancreatic secretions and enteric material backflow into the biliary network are some of the consequences of CCs.

There are no laboratory tests that can diagnose Choledocal cysts.

Ultrasonography and MRCP are two imaging techniques used to diagnose CCs[15]. Abdominal ultrasonography can detect choledochal cysts throughout pregnancy and is frequently the first imaging modality used after birth. It has a high sensitivity (71 to 97 percent) for detecting them, but a significant weakness is its failure to distinguish them from other anomalies and its failure to identify the aetiology in around one-third of cases[16]. When compared to ultrasonography, CT cholangiography is more efficient in determining cystic dilatations as well as the number of dilatations, particularly in type IV and type V. It has a sensitivity of 90% for detecting CCs and is useful for surgical approaches, especially when MRCP cannot be done. With a sensitivity of 90 percent to 100 percent, MRCP is the gold standard for identifying CCs[17]. The key advantage of this methodology and extensive usage is its precision, non-invasive nature, lack of radiation usage, and lack of consequences which include bleeding, perforation, pancreatitis, or cholangitis seen in ERCP. It is also capable of detecting other ductal disorders. However, it is unable to detect minor ductal anomalies and tiny CCs. Although ERCP has the best predictive value for CCs and is curative, it is only utilised in selected instances of CC due to complications, ionizing radiation exposure, and its intrusive nature.

Another technique that can be utilised in new-borns to differentiate between choledochal cysts and biliary atresia, as well as to diagnose spontaneous cyst rupture, is the technetium-99 HIDA scan[18]. The management strategy for CCs is determined by the type of cyst according to the classification. The mainstay of therapy is surgical excision and bile flow restoration.

The goal of surgery is to prevent long-term CC consequences, such as pancreatitis, cholangitis, liver cirrhosis, and malignant change[19]. Type I and IV CCs are cured by total cyst removal and bile flow restoration by a Roux-en-Y hepaticojejunostomy (HJ) or hepaticoduodenostomy (HD)[20]. Previously, cholecystectomy with internal or exterior drainage was also advised. The probability of type II and type III cysts to

develop cancer is low. Type II is treated by simple cyst removal or diverticulectomy, as well as the closure of the neck of diverticulum[10]. Type III choledochal cysts (choledochocele) can be managed by sphincterotomy only in selected cases, complemented with a sampling of cyst epithelium to eliminate dysplasia as well as to determine the type of epithelium coating the cyst, because the biliary mucosal lining is linked to greater chances of having malignant disease. If the cyst becomes too big, it can be marsupialised during ERCP or by marsupialisation using a transduodenal route[21].

If the Caroli disease (Type V CCs) is isolated or unilocular, segmental hepatectomy is the best choice; if the illness is widespread or bilocular, an orthotopic liver transplant is required[22]. Furthermore, individuals with type V CCD who have congenital hepatic fibrosis are predisposed to portal hypertension and oesophageal varices. The biggest issue with CCs is the possibility of developing malignancy. Tumours are more prevalent in type I and IV CCs and less prevalent in types II, III, and V CCs[23]. That is why cyst removal is critical for preventing malignant transformation.

Conclusion

It is crucial to establish a multidisciplinary group of professionals, that comprises paediatric surgeons, pathologists, paediatric gastroenterologists, physiotherapists, nutritionists, oncologists, and radiologists, to achieve greatest outcomes in diagnosing and treating children with choledochal cysts.

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Conflict of Interest: The person who signed the letter as head of department is also one of the authors of the article.

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