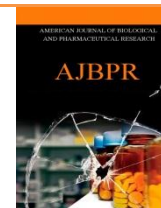




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CLINICAL PRESENTATIONS AND TREATMENT IN ADULT PATIENTS WITH CHOLEDOCHAL CYST –FORTY SIX CASES FROM EASTERN INDIA

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ABSTRACT

A choledochal cyst (CDC) is defined as an isolated or combined congenital dilatation of the intra- and/or extra hepatic biliary tree. It is a rare disease and majority of the patients present either in their infancy or childhood. However, in nearly 20% of the patients with CDC, the diagnosis is delayed till adulthood. Here we analyze the clinical presentation, diagnosis and treatment of congenital bile duct cyst in 46 adults. The clinical data of 46 patients with congenital choledochal cysts admitted from 2005 to 2011 were analyzed in the Department of Surgery, Institute of Post Graduate Medical Education & Research and SSKM Hospital, Kolkata. We also reviewed literature search (Medline) and manual cross-referencing. Female patients outnumbered males. Preoperative imaging is very crucial in evaluating CDC; MRCP is our choice of imaging. Almost all cases, were attempted surgical correction of the defect i.e., complete excision and biliary reconstruction. Complications were duly taken care of and long term follow up is maintained (ongoing). The possibility of the disease should be kept in mind in young adults suffering from symptoms of hepatobiliary disease with dilated bile duct. MRCP is the mainstay of preoperative imaging. . Cyst excision with Roux-en-Y hepaticojejunostomy is recommended as the treatment of choice for patients with type I or type IV cysts. Regular follow up is mandatory.

INTRODUCTION

Choledochal cysts (CDC) are typically a surgical problem of infancy or childhood: the diagnosis is delayed

until adulthood, however, in nearly 23% of patients [1]. A choledochal cyst was first noticed by the German anatomist Abraham Vater in 1723 [2]. Notwithstanding this, the first detailed case in the English literature was that of Halliday Douglas of Edinburgh who, in 1852, described a 17 year-old girl who had jaundice and a tender abdominal mass [3]. Almost a liter of bile was drained percutaneously with some short-lived relief of symptoms.

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Although clinically similar, the presentations and therapeutic strategies for CDC in adults may differ substantially from that of younger patients [4]. It is a rare condition in the West, although it is commoner in Asian females [5]. The relatively high incidence of coexistent hepatobiliary disease such as cystolithiasis, cholelithiasis or pancreatitis increases the difficulty of the surgical management of choledochal cyst [6-8]. Appropriate surgical management of these lesions depends on the anatomic site and the extent of the cystic process. In contrast to pediatric experience, surgery is complicated by associated hepatobiliary pathology (cholangitis, portal hypertension and rarely cholangiocarcinoma) and the added technical difficulties of reoperative biliary surgery [1, 4]. We have analyzed the clinical data of 46 patients to summarize the experiences in diagnosis and treatment of congenital bile duct cyst in adults.

PATIENTS and METHODS

The purpose of the study is to evaluate clinical presentations, types (Todani) [6], diagnostic tools, and surgical management in adult patients (>12 years, as per Institute norm) with CDC at the Department of Surgery, Institute of Post Graduate Medical Education & Research and SSKM Hospital, Kolkata. Review of literature search (Medline) and manual cross-referencing were also done. From January 2005 to December 2011, forty six patients (34 females) were treated and followed up prospectively in our Institute.

All patients were thoroughly evaluated clinically. Proper and relevant investigations were done to establish the diagnosis of CDC and associated hepatobiliary pathology. Hemogram, renal and liver function tests were also performed with imaging studies (USG, ERCP/MRCP, CT accordingly). The aim of surgery is to achieve complete cure. This usually entailed us to excise the CDC totally with bilioenteric anastomosis; wherever this was not possible, the aim is to give maximum palliation to the patients according to her / his age and associated comorbidities. Cyst wall was routinely biopsied for any dysplastic / malignant changes. The follow up of the patients are ongoing.

RESULTS

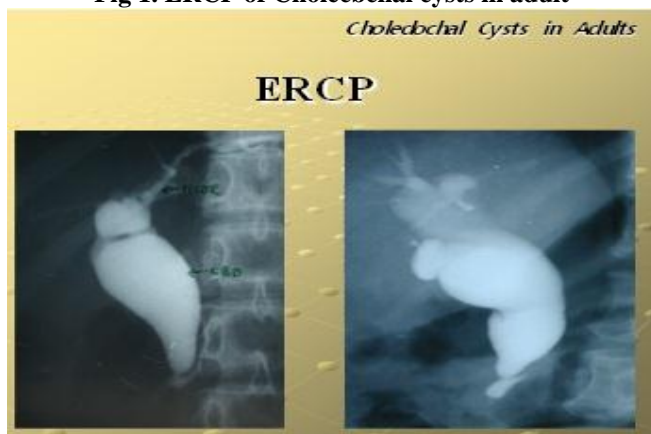
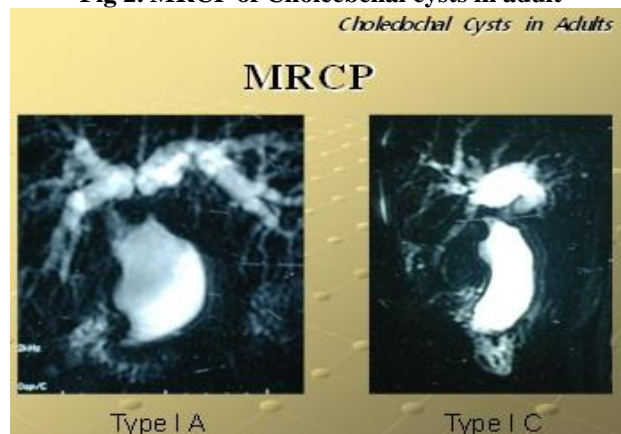
Forty six patients (thirty four females) with wide age range (13-89 years, mean age 32 years) are analyzed in this series over seven years period. The time from the onset of first symptom until the diagnosis is finally made is considerably long; at mean 38 months (2-120 months). Commonest presenting feature is abdominal pain (46=100%), followed by features of cholangitis +/- jaundice (20=45%), and pancreatitis (10=22%); associated conditions like cystolithiasis (12=26%), biliary cirrhosis +/- portal hypertension (4=9%). No patient showed feature of cholangiocarcinoma in our series. Ultrasonic examination was undertaken in 46 cases, ERCP/MRCP was performed in 15/ 25 cases, CT scan in 6 cases. Distribution CDC types in our series shown in Table 1 and Type I is the commonest cyst type.

Surgical procedures: Total cyst excision with bilioenteric anastomosis could be done in 40 case; partial cyst excision in 2 cases due to gross adherence of cyst wall to portal vein. Only cystojejunostomy was performed in a case that had portal hypertension with poor liver function due to cirrhosis. Oldest patient (89 years) in our series, presented with severe cholangitis, in whom cystojejunostomy was performed about 30 years ago. Endoscopic sphincterotomy failed and we could only do a transduodenal sphincteroplasty (*) to relieve jaundice and cholangitis. Cyst excision could not be possible due to dense adhesion of that area. 2 cases of Caroli's disease (Type V) presented with gross cirrhosis, portal hypertension and poor liver function; they were not offered surgery; might be candidates for liver transplantation. All cases were done by open surgical technique and mean operative time is 110 minutes; transfusions were required infrequently. We lost one case (Type I) during postoperative hospital period; her unfortunate demise was due to anastomotic leak (> biliary peritonitis > septicemia > death). Morbidities like wound infections, transient bile leaks were also encountered but were managed successfully with conservative approach. During follow up, all patients were evaluated clinically (esp. features of cholangitis are enquired), with liver function test and imaging as necessary. Two cases of anastomotic strictures (Type IV A) were encountered during the follow up and refashioning of anastomotic strictures was planned.

Table 1. Distribution CDC types in our series

Type (Todani)	Number / % (n=46)	Surgical procedure
I	38 / 83%	Total excision in 35. Partial excision in 2 cases.* Sphincteroplasty in one case
II	0	
III	0	
IVA	6 / 13%	Total Excision in 5; Cystojejunostomy in one case with portal hypertension.
IVB	0	
V	2 / 4%	No operation.



Fig 1. ERCP of Cholebchal cysts in adult

Fig 2. MRCP of Cholebchal cysts in adult

Fig 3. Operative identification of cysts in adult


DISCUSSION

Choledochal cyst is an aneurismal dilatation of the bile duct and is an uncommon finding; accounts for 1% of all benign bile duct condition [9]. It is reported to be more common in Asian populations and in females [5, 10]. Most series show that the most common cyst is the Todani type I [11, 12], which is also found in our series where we have 38 patients with this type of cyst. Similarly, we have a preponderance of female patients in our series (74 % patients).

Between January 2005 and December 2011, we evaluate 46 adult patients (>12 years of age) with CDC in our institution, which is the oldest and most prominent Government teaching hospital of Eastern India and serves a large population; approximately, 30000 new patients attend annually at the Department of Surgery. The small number of patients in our series over a period of seven years indicates the rarity of the disease in adults. Reported incidence of adult patients with CDC from other parts of India also corroborate with our finding: Kashmir (47) [13], Vellore (57) [14], Coimbatore (37) [15, 16], and Varanasi (10) [17]. The wide age range (13-89years), female (74%) and Type I (83%) preponderance, clinical presentations like vague abdominal pain (100%) and cholangitis (45%) - all

have similarity with the published series from various parts of World like, India [13-17], China [7,18], Japan [5], Mexico [19] and also from the Western world [20-22]. The etiology of CDCs is still unclear, although many theories have been put forth. Babbitt's [23] theory of cysts caused by an abnormal pancreaticobiliary duct junction (APBDJ) such that the pancreatic duct and the common bile duct meet outside the ampulla of Vater, thus forming a long common channel, has gained much popularity. This theory postulates that the long common channel allows mixing of the pancreatic and biliary juices, which then activates pancreatic enzymes. These active enzymes cause inflammation and deterioration of the biliary duct wall, leading to dilation [23]. Furthermore, greater pressures in the pancreatic duct can further dilate weak-walled cysts [24].

When patients present with the symptoms described, the first step toward making the correct diagnosis is imaging. USG is the first choice. Sensitivity of USG in making the diagnosis is 71%-97% [25]. Furthermore, given that USG is noninvasive and inexpensive, it is the modality of choice for follow-up surveillance. Endoscopic Ultrasound (EUS) has been proven useful as it does not have any of these limitations



and allows good visualization of the intrapancreatic portion of the common bile duct [26].

Computed tomography (CT) scans are useful in showing continuity of the cyst with the biliary tree, its relation to surrounding structures and the presence of associated malignancy. It is superior to USG in imaging the intrahepatic bile ducts, distal bile duct and pancreatic head [27]. Although ERCP has been reported to be the most sensitive imaging modality for CDCs, this sensitivity does fall in certain situations. Recurrent inflammation and scarring may make cannulation of the ampulla difficult or impossible and may cause partial or complete obstruction at any point of the biliary tree, with no resultant biliary imaging. Finally, the sensitivity of ERCP and the quality of images is operator-dependent [28]. Given the concerns regarding cholangiography, MRCP is now considered to be the gold standard [24, 28-32]. We also agree with the literature and USG followed by MRCP is our choice of investigation and also we prefer USG for follow up surveillance.

Surgical treatment, in general, is demonstrated by two procedures: internal drainage without removal of the cyst (Cystoentero-anastomosis) or resection of the cyst with biliary-enteric anastomosis. Cystoentero-anastomosis is associated with biliary stasis, secondary biliary calculus, recurrent cholangitis, hepatic abscess and biliary cirrhosis [33-36]. It is more frequent for adult patients to have history of internal drainage or placement of prosthesis for biliary drainage than pediatric patients [37]. We had one case of an 89 years old man where cystojejunostomy procedures were attempted 30 years ago and he presented to us with cholangitis and jaundice. Cystojejunostomy was performed in one patient that had portal hypertension with poor liver function due to cirrhosis.

For this reason, surgical treatment of choice is cyst resection [6, 35-40] with variations according to the type. Type I and type IV resection of the cyst with bilioenteric anastomosis; type II: resection and if necessary an anastomosis; type III: sphincteroplasty or sphincterotomy. We have experience of operating only Type I & IVA CDC. There was no Type II or III CDC in our series. All extrahepatic cysts should be excised. Postoperative evolution will be satisfactory if biliary flow is adequate with symptom resolution in >80% of cases [6]. This was the case in >90% of our patients.

In patients with intrahepatic cysts the treatment is controversial. Some authors suggest the possibility of hepatectomy or hepatectomy, [46, 47] but a wide hepatojejunal anastomosis is probably useful with close postoperative monitoring [6, 33, 41]. In our series, we have 6 cases of type IVA cysts. We could excise 5 cases of extrahepatic cysts and in one case we did cystojejunostomy as mentioned earlier. We recommend close monitoring, especially in patients with intrahepatic involvement [41].

We have all operations done by open method though laparoscopic and robotic cyst excision followed by bilioenteric anastomosis is accepted method of treatment [10, 15, 16, 46-49]. Palanivelu *et al* from Coimbatore, India reported 35 laparoscopic cyst excision [15,16]. This recent publication reports the largest series of laparoscopic treatment of bile duct cysts in adults. It establishes the feasibility and favorable outcomes for this approach in adults.

Postoperative morbidity is mainly due to surgical wound infection, cholangitis and leakage of the anastomosis, which in most cases are managed conservatively [44] and it is same in our series. We have one death in the early postoperative period. In the follow up period, to patients developed anastomotic strictures (both type IVA CDC) and reoperation was planned. So, the post operative anastomotic stricture is high after type IVA cyst excision, seen in our series.

CONCLUSION

Over the years, very few diseases have caught on to the fancy of the surgeons the world over. The diseases are rare but fine and sterling. These are usually difficult to diagnose and treat, primarily because of their rarity, so that a surgeon has a very limited exposure in his lifetime. Proper diagnosis and treatment of such disease yield an unparalleled amount of pleasure and satisfaction brims over to cheer the otherwise dull and monotonous routine of every surgeon's daily life. Choledochal cyst is one such disease entity.

Choledochal cyst is a congenital disease mainly diagnosed in pediatric patients. When found in adults, management is surgical in the majority of cases. The initial imaging of the biliary tree should be a simple ultrasound, and in most patients this will lead to the diagnosis of CDCs. To delineate biliary anatomy for operative planning, ERCP is commonly used for this; the risk of cholangitis warrants the use of MRCP instead whenever possible. CT and MRI are useful modalities to diagnose and determine the extent of intra-hepatic disease, such as type-IV A and type-V cysts. Treatment should include resection of the cyst with bilioenteric anastomosis. Internal drain towards a loop of small bowel is not recommended because of the possibility of complications and probability of chronic damage to the liver or the development of a malignancy in the cyst remnant. In our experience, surgical management with resection has been possible with low morbidity and mortality.

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CONFLICT OF INTEREST:

The authors declare that they have no conflict of interest.



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