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An analysis of Choledochal Cysts in Adults

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Abstract

Background

Choledochal Cyst (CCD) is not an isolated entity but a constellation of pathological abnormalities in the pancreatohepatobiliary system. Alonso-Lej et al (1959) proposed an anatomical classification. Todani et al (1977) described the choledochal cysts into five types based upon the anatomy of the cyst and distribution within the hepatobiliary tree. Multiple theories have been proposed to explain the origin of bile duct cysts .

Methods

It is a a retrospective review of 8 patients who underwent surgery in our tertiary care hospital during the period January 2016 to 2018 , Department of Surgical Gastroenterology , Govt. KMC Hospital, Chennai

Results

Eight cases of choledochal cyst were managed ; 6 female (75%) and 2 males(25%) .Age ranged from 28 years to 64 years. According to modified Todani's classification there were 7 cases of Todani type I (87.5%)and one case of Type IV A.(12.5%). Surgical intervention included complete excision of the cyst with Roux-en-Y Hepaticojejunostomy in 6(75%) patients and Lilly's procedure in one patient (12.5%)and non-operative management in one patient. There were no evidence of Malignancy reported in any of these patients.

Conclusion

Choledochal cyst is rare in adults. Studies suggest that nearly 80% of individuals with bile duct cysts are complicated by any one of these potential problems like cystolithiasis, pancreatitis, cholangiocarcinoma, intrahepatic abscess and cirrhosis with portal hypertension. The surgical strategy aims for single stage complete excision of the cyst with hepaticojejunostomy.

Keywords: Choledochal Cysts. ,Hepaticojejunostomy., APBBDJ

Introduction

Choledochal cyst (CCD),though rare in adults it is reported more commonly in Asian countries particularly from Japan than from Western countries. [1,2]. In Japan the incidence is about 1 in 13,000 (1-3)whereas in England(4,5) the incidence is 1 in 2 million patients.. This affects mainly females and the ratio in female to male is 3:1 across the globe. [3,4]. The accepted etiopathogenic event in choledochal cyst formation is presence of anomalous biliopancreatic duct junction (APBDJ) allowing pancreatic juice to reflux into biliary tree [2-5].

Bile stasis and stone formation is associated with choledochal cyst and malignancy transformation is at high risk in whole biliary tree (10,14).The cyst anatomy and the Todani classification of the disease can be accurately imaged by preoperative Magnetic Resonance Cholangio Pancreatography(MRCP).The treatment of choice for Type I Choledochal cyst and the extra hepatic component of type IV choledochal cyst is complete cyst excision with cholecystectomy and Hepatico-Jejunostomy by Roux-en-Y technique

Materials and Methods

It is a a retrospective review of 8 patients who underwent surgery in our tertiary care hospital during the period 2015-2018.Data regarding the clinical presentation, investigation, operation, and follow-up were analysed. The type of cyst was classified according to Todani classification.

Results

Eight cases of choledochal cyst were found; 6 female (75%) and 2 males(25%) .Age ranged from 28 years to 64 years. According to modified Todani" s classification there were 7 cases of Todani type I (87.5%)and one case of Type IV A.(12.5%). The predominant symptoms were recurrent abdominal pain in 7 patients and and one patient had jaundice. . Investigations included ultrasound abdomen and CT Scan abdomen in all patients ERCP in 2 patients, and MRCP in 5. Surgical intervention included complete excision of the cyst with hepaticojejunostomy in 6 (75%) patients and Lilly's procedure in one patient (12.5%)and non-operative management in one patient. No evidence of malignancy was reported in any of these patients.

Table 1: Demographic details, management, and outcome of patients.

Serial no.	Age& Sex	Symptom	Types	Investigations	Procedures	Post-operative	Hospital stay days
1	28 F	Pain abdomen	Type I f	USG ,CECT Scan ,MRCP	Cholecystectomy, Cyst excision, Hepaticojejunostomy	smooth	10
2	45 F	Jaundice	Type I f	USG ,CECT Scan, ERCP	Cholecystectomy, Cyst excision, Hepaticojejunostomy		8
3	38M	Pain abdomen	Type I f	USG,CECT Scan, MRCP	Cholecystectomy, Cyst excision, Hepaticojejunostomy		8
4	60F	Jaundice	Type I f	USG ,CECT Scan, MRCP	Cholecystectomy, Cyst excision, Hepaticojejunostomy		13
5	53F	Pain abdomen	Type IV A	USG ,CECT Scan, MRCP	Non-operative mgmt		6
6	7OF	Pain abdomen	Type I f	USG ,CECT Scan, ERCP	**Lilly' procedure**		12
7	37F	Pain abdomen	Type I f	USG ,CECT Scan, MRCP	Cholecystectomy, Cyst excision, Hepaticojejunostomy		7
8	51M	Pain abdomen	Type I f	USG ,CECT Scan	Cholecystectomy, Cyst excision, Hepaticojejunostomy		8

Operative details

Pre-operative Drainage procedures were carried out in two patients with ERCP and biliary stenting who had earlier recurrent symptoms of cholangitis. Surgical strategy included complete choledochal cyst excision, cholecystectomy and Hepatico-jejunostomy in all patients except in one where Lily' procedure was carried out because of extensive inflammation and inflamed posterior wall which was adherent to portal vein .The operating time varied from 150 minutes to 240 minutes. The duration of hospital stay ranged from 7days to 13 days.

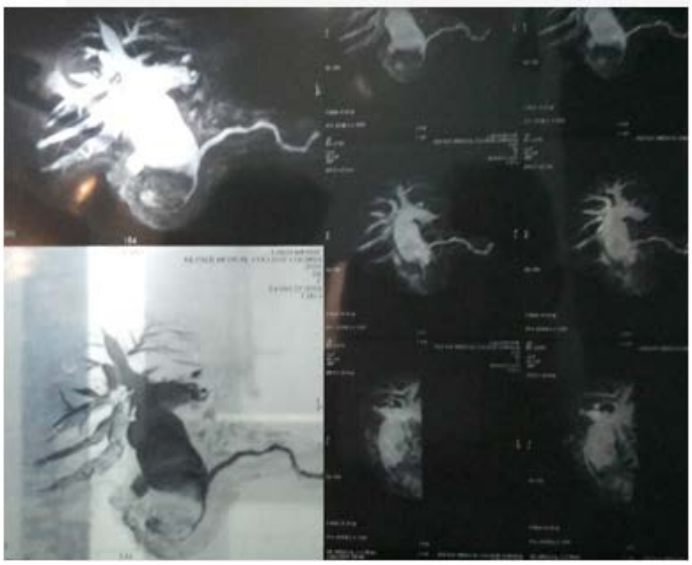

Figure 1: (a) MRCP showing type I- Massive Fusiform Choledochal cyst with large stones, choledochal cyst.

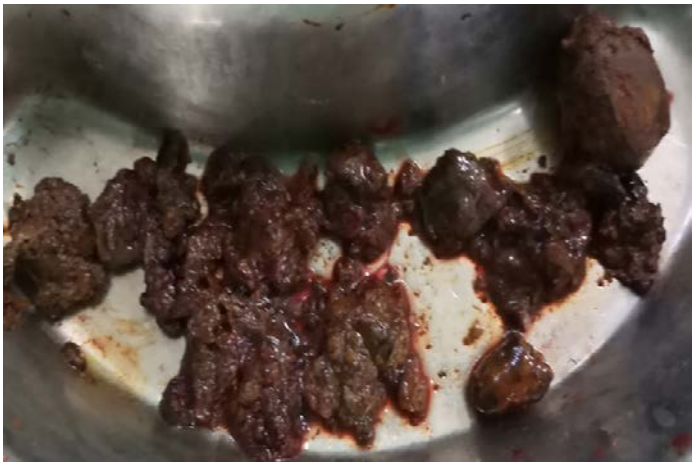

Figure 2: After the Resected specimen, plenty of large stone alongwith numerous gravel seen.

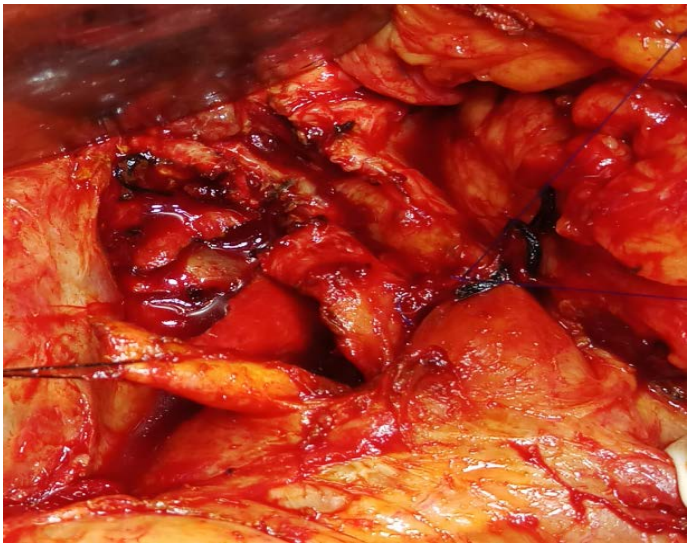

Figure 3: Intraoperative view of large choledochal cyst dissected out

All patients were symptom-free, except for one patient who developed 2 episodes of recurrent cholangitis during their follow-up and were managed successfully with antibiotics. No anastomotic strictures or malignancy was noted in any of these patients during follow-up.

Discussion

Pathogenesis of Choledochal cyst ; The anomalous pancreaticobiliary junction leads to reflux of pancreatic juice into bile duct, leading to ineffective bile flow which in turn results in increased intraductal pressure, chronic inflammation, and its associated carcinogenic effect [14,17,19,].The existence of long comman channel varies from 68% to 94% in adult series [2-7].

The initial investigation of choice is **ultrasoun**d abdomen in evaluating the choledochal cyst due to easy availability and ease of performing it ,which may give a clue to the next line of approach to this uncommon disease.(3,10,11).The difficulties with this investigations are technical quality, to differentiate the cyst from other causes of gallbladder distension and may underestimate this rare diagnosis**.** To know more about extrahepatic and intrahepatic extension of biliary system dilatation, Computed tomography scan will be helpful.(3,10,11)

MRCP is the most important investigation(**Figure-1**) in delineating the cyst anatomy including its site, size, shape, and can detect the anomalous pancreatico biliary ductal junction (APBDJ)and it has the advantage of being noninvasive in nature(21). It facilitates the reliable diagnosis of Caroli's disease based on finding of cystic intrahepatic cavities communicating with intrahepatic biliary tree (22).By taking brush and biopsy cytology during ERCP/PTC , the diagnosis of cholangio-carcinoma can be established in 40-60% of patients. (23).

The **complications** are stone formation within the cyst and intrahepatic ducts (**Figure-2**),recurrent cholangitis, pancreatitis, spontaneous cyst rupture(10,11,22) portal hypertension (7,8)secondary to coexisting congenital hepatic fibrosis, acute pancreatitis(30-70%) due to abnormal pancreaticobiliary junction with protein plug impaction(10,11).The risk of malignant transformation is the major concern which is well documented in the literature(1-15) and the whole biliary tree is at risk.

The main motto of **surgical intervention** is complete excision of the cyst wall wherever possiple in order to avoid long term complications as mentioned above. By doing internal drainage procedures (cystoduodenostomy or cystojejunostomy) these complications can be exacerbated(4-6). Cholecystectomy is done due to high risk of gallbladder malignancy being reported.(13,18).The ideal treatment is complete excision of choledochal cyst(**Figure-3**) and Hepatico jejunostomy with normal available hepatic ducts. The exact level of anastomosis is a balance between the need for complete cyst resection and the need to achieve widely patent anastomosis [11,13,14].

In few patients due to recurrent cholangitis and marked adhesion of surrounding tissues, it may be difficult to remove the posterior wall of cyst from the portal vein and in such patients a rim of posterior cyst wall is left behind(**Lilly's procedure**) (24)

Conclusion

Choledochal cysts should be suspected in patients with uniform dilatation common bile duct from intrapancreatic portion to hilum of liver or beyond that into the liver where there is no attributable etiological factors. Once suspected I t needs to be evaluated immediately for early diagnosis and confirmation, because of its potential life threatening complications including malignancy. The imaging modality of choice remains MRCP which will guide us in delineating the anatomical details, anomalous junction of pancreato biliary system and lithiasis within the bile ducts. Complete excision of the choledochal cyst with Roux-en-Y Hepaticojejunostomy is the ideal treatment wherever possible without compromising the anastomotic level. Hepatic resection is indicated in few patients with localised intrahepatic type IV A. Diffuse intrahepatic caroli'disease should be offered for liver transplantation. Nowadays laparoscopic cyst excision with hepaticojejunostomy is being practised in high volume centres with highly experienced laparoscopic surgeons.

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Conflict of interests

The authors declare that there is no conflict of interests

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