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Choledochal Cyst: A Retrospective Study Case Series

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Abstract: ***Background and Objectives: Choledochal cyst is an uncommon congenital malformation of biliarysystem, involving cystic dilatation of intrahepatic and/or extra hepaticbile duct. The estimated incidence is one in 1000 live birth in Asian population [1-2] with female to male ratio is 3:1. The objective of this study is to study the presentation, diagnosis, treatment and outcomes of choledochal cyst in our tertiary care center. Materials and Methods: A retrospective review ofpatients, from Jan 2012 to April 2017, who were diagnosed as Choledochal cyst and treated by surgical intervention and their outcomes,in our tertiary care center. Results: We analyzed 16 cases of choledochal cyst. The condition commonly affected the pediatric age group (0 to 10 years)with female preponderance. Most common presenting symptom was pain in abdomen (75%) and jaundice (53%). However the classical triad of abdominal pain, jaundice, and an abdominal mass was not seen in any of the cases. Ultrasonography was diagnostic in all cases. Todani type I cyst was the most common type. Elevenout of 16 cases underwent complete cyst excision with cholecystectomy and Roux-en-Y hepaticojejunostomy without any major complication, however so many cases were not operated due to, not fit for surgery, not willing for surgery etc. Conclusion: Choledochal cyst is a clinical condition occurs, commonly in younger age group that is conveniently diagnosed on ultrasound and has a rewarding outcome if operated.***

Keywords: Choledochal cyst; Case series; Retrospective study; prognosis

1. Introduction

Choledochal cyst is a cystic dilatation of the extra hepatic and/or intrahepatic bile duct, less than 1/100000 patients in worldwide,[1-2] but are more common in Asian population with an incidence of 1:1000 in Asian population.[1-2] It has female preponderance (three to eight times more in women than men) with female: male ratio is 3:1[1-2]. Todani Type I cysts are the most common type and account for approximately 75% of patients. The most accepted theory in pathogenesis of choledochal cyst is Babbitt's theory'[3] is associated with anomalous pancreaticobiliary duct junction (APBDJ) [3-4]. Patients may present at any age with pain abdomen, jaundice and a right upper quadrant mass, however, 60 % of cases are diagnosed before the age of ten years [2]. The main diagnostic tool for detection of a choledochal cyst, in childhood, isultrasonography. Ultrasonography will confirm the presence of an abnormal cyst and magnetic resonance cholangiopancreatography (MRCP) will reveal the detail anatomy. Complete cyst excision with cholecystectomy followed by biliary reconstruction using Roux-en-Y hepaticojejunostomy is the treatment of choice for the extra hepatic component of the disease (type I and type IV Choledochal cyst). In type III, choledochal cyst, ERCP guided sphincterectomy can be done. In type V choledochal cyst (Caroli's disease), liver resection or liver transplantation is indicated. This study was conducted with objective of analyzing the presentation, diagnosis and treatment of choledochal cyst in a tertiary care center.

2. Materials and Methods

This is a retrospective review of the records of the patients who were diagnosed as choledochal cyst and underwent medical and operative intervention at tertiary care center from January 2012 to April 2017. Baseline preoperative investigations which include complete hemogram, urine examination, liver function tests, renal function tests and coagulation profile were recorded. Data regarding the clinical presentation, investigation, surgery, and follow-up were retrieved and analyzed. The research work has been reported in line with the PROCESS criteria. The type of cyst was classified according to the Todani classification.[5] The cases are in follow up every three months in the first year, then every six months for next two years and yearly thereafter.

Additional information

- This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.
- No conflicts of interest.

3. Results

From January 2012 to April 2017, sixteen patients were studied. Eleven patients were underwent surgical intervention however many cases were not operated due to reasons like, not fit for surgery, not willing for surgery etc. Study shows the following results.

Table Showing Age Incidence

Age (years)	*No of Cases*
1 to 10	6
11 to 20	2
21 to 30	4
31 to 40	1
41 to 50	2
51 to 60	1
Total	16

The most common age group is pediatric age group (0-10 yrs.) Commonly Female: male ratio ranging from 3: 1 to 4:1, [1-2]. In this study also most patients are female and one is male patient.In adults [2] commonly in younger age (21-30 yrs.) and rare in old age (single case in this study).

Table Showing Signs and Symptoms

Signs and symptoms	*No of cases*	*Incidence*	*Reference study Incidence [6]*
Abdominal pain	12/16	75.0 %	78 – 90 %
Jaundice	9/16	56.2 %	40 – 50 %
Abdominal mass	0/16	0.00 %	
Cholangitis	2/16	12.5 %	40 – 50 %

Pain in abdomen was the most common presenting symptom (75%) followed by jaundice (56.2 %) and cholangitis (12.5%). None of the case in this series presented with the classical triad of pain abdomen, jaundice and mass per abdomen.

Table Showing Types of Todani's Classification

Type of choledochal cyst	*No of cases*	*Reference incidence [6-8]*
Type I	11	50 – 80 %
Type II	0	2 %
Type III	1	1.4 – 4.5 %
Type IV A	3	15 – 30 %
Type IV B	0	
Type V	1	20 %
Total	16	

In this study Todani type I cysts are eleven (68.75%) cases, type III in one (6.25%) case, type IVA are in two (18.75%)cases and type V cyst (Caroli's disease) in one (6.25%) case. In this study three deaths were recorded, related to choledochal cyst. Out of which one was preoperative two were postoperative.

In this study, Serum alkaline phosphatase (ALP) was raised in all cases, raised serum bilirubin was seen in eight (56%) cases. Ultrasonography of abdomen followed by contrast enhanced computed tomography (CECT) abdomen was used to make diagnosis of the cyst. Eleven patients were operated with complete excision of cyst with cholecystectomy followed by biliary reconstruction using Roux-en-Y hepaticojejunostomy. The patient with Choledochocele (type III) has advised for ERCP intervention. The patient with Caroli's disease was advised for liver resection or liver transplantation, but patient had taken DAMA. Two operated patients was presented with recurrent dyspepsia due to duodenal erosions, one is managed conservatively with follow up. The other case was treated with gastrojejunostomy followed by death in early postoperative period. The early postoperative outcome was uneventful in all of operated patients except superficial surgical site infection in four out of eleven operated cases. None of them developed pancreatitis, anastomotic leak or recurrent cholangitis. In late postoperative period, only mild abdominal pain was complained by 2 patients which required no admission and managed on OPD visits. The mean duration of postoperative hospital stay was 15 to 20 days. The histopathology of excised cyst was reported as choledochal cyst in all operated cases. Evidence of anastomotic strictures or malignant transformation was not found in any of the patients in follow-up till now. All the patients are still in follow up as per the protocol.

4. Discussion

Choledochal cyst malformation is characterized by dilatation of the biliary tract in the absence of acute obstruction to the bile flow. Those of these malformations with cystic dilatation constitute the choledochal cyst. Although not proven, the commonly accepted theory [4-5] of their pathogenesis relies on the presence of an anomalous pancreaticobilliary junction (APBJ), which is seen in up to 90% patients with choledochal cyst.

It was first described by Vater and Ezler [9] in 1723.According to Kumar and Rajagopalan (2012) [10] 85% cases are diagnosed before puberty. Similarly in our series also, 43 % of the cases presented below the 15 years of age. Various authors mentioned the female: male ratio ranging from 3: 1 to 4:1.In our series female preponderance is seen with female to male ratio is 15:1. Lipsett et al.[11] reported the classic triad of pain abdomen, jaundice and palpable abdominal mass is seen in less than 20% of the cases. However, in this study none of the patients presented with classical triad of choledochal cystic disease. Lipsett et al. and Jesudason SR [12] mentioned abdominal pain is most common symptom among pediatric and adult patients. According to Singham [6], abdominal pain in pediatric and adult patients has an incidence ranging from 78% to 90%, jaundice and cholangitis being in 40 to 50%. Similarly, in this study abdominal pain was present in about 75% of the cases; jaundice was seen in 56 % and cholangitis in 12.5 % of the cases. According to Kumar and Rajagopalan,[10] 15% of the patients may have features of hepatic changes of cirrhosis, pancreatitis or choledocholithiasis while in the present study none of the patients presented with cirrhosis, pancreatitis or choledocholithiasis. Treem [13] mentioned the incidence of spontaneous rupture of choledochal cyst was 1.8 to 2.8%.

Ultra sonography (USG) is usually the first investigation and is a very sensitive (71 to 97%) in the detection of choledochal cyst. A properly performed high resolution USG is the best screening test [16]. Le et al [14, 15] described 38 cases of choledochal cyst where in all cases USG was the only diagnostic modality. Similarly, in our studyUSG is main diagnostic tool. Though CECT abdomen was done in some cases, findings were similar to USG. So we found USG as sensitive as CECT abdomen for the diagnosis of choledochal cyst. Studies showed CECT abdomen is 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. In patients with type-IVA cysts and Caroli's disease; it is useful to delineate the intrahepatic dilations and the extent of disease such as diffuse hepatic involvement versus localized segmental involvement. Some authors recommend spiral computed tomography to differentiate malignant cyst wall changes from reactive inflammation.Other modalities [16] like Endoscopic Retrograde Cholangiopancreatography (ERCP) or MRCP can precisely visualize the extra hepatic bile duct. ERCP could be used as therapeutic in case of type III with jaundice, though it has its own benefits and complications. MRCP has been advocated as the ideal and non-invasive complete diagnostic modality for choledochal cyst as it ensures accurate visualization of the entire pancreatobiliary system but it is costly so cannot be done in every case.

Initial classification by Alonso-Lej *et al*. in 1959 described 3 types of choledochal cysts, type I–III.[8,11] Later Todani *et al*. in 1977 modified it by adding type IV and V [7]. Modified Todani *et al*. classification is most commonly used by surgeons.

Lipsett [8,11] and Cho et al. mentioned complete excision of extrahepatic component of choledochal cyst combined with cholecystectomy, followed by Roux-en-Y biliary reconstruction as the treatment of choice for type I and IV choledochal cyst. And depending on cyst type (Todani type IV, V), further intervention may be necessary. Cholecystectomy is carried out due to the high risk of associated gall bladder malignancy [17].Likewise among all patients we operated complete cyst excision with cholecystectomy followed by biliary reconstruction using Roux-en-Y hepaticojejunostomy was performed. We recommend the same as the main stay of treatment for choledochal cyst Type I and Type IV. Type II cysts should be excised entirely and in the presence of an APBDJ, biliary enteric diversion by Roux-en-Y hepaticojejunostomy is appropriate. Type III cysts which may not involve APBDJ can be treated with endoscopic drainage, transduodenal excision or sphincteroplasty. However, in case of extensive intrahepatic dilation with complications, such as stones, cholangitis, or biliary cirrhosis in type IV A, other options, such as hepatic resection in case of unilobar disease and liver transplantation in bilobar disease can be considered as per literature [18,19].Surgical treatment of Caroli's disease type V ranges from resection if the disease is unilobar to liver transplantation when diffuse disease is detected [32].

Early complications of cyst excision and hepaticoenterostomy include anastomotic leak, pancreatic leak with injury to the pancreatic duct, bowel obstruction due to intussusception, and bowel kinking due to manipulation or adhesions. Late complications include peptic ulcer disease, cholangitis, biliary calculi, pancreatitis, liver failure and cancer.[27,30,31,34]. However in this study two patients were reported with late complication as peptic ulcer disease due to duodenal erosions. Fibrosis and inflammation of cyst tissue at the time of surgery, such that the anastomosed margins are friable, result in poor healing, leakage and anastomotic stricture. Because fibrosis and inflammation increase with age, surgical complications become more common with older age at surgery, and surgery should be done as early as possible [28,29]. Cholangitis and calculi usually occur as a result of anastomotic stricture leading to bile stasis [28]. The reported incidence rate of anastomotic stricture following Roux-en-Y hepaticojejunostomyis 4.1%.[19, 20,21]However, a wide anastomosis between the hepatic hilum and intestine may prevent anastomotic stricture.Tao et al. [33] suggested minimum diameter of stoma to be three cm and observed 92% success rate with biliary reconstruction. Choledochal cyst associated cholangiocarcinoma, has incidence about 10-30% [17, 20,21]. It was not found in any of our patients. This may be due to the fact that the risk of associated cholangiocarcinoma increases with age. However, it is likely that with increased number of patients, the rate of cholangiocarcinoma will meet what is found in the larger population. Similarly, due to the short follow up in this study, patients recently diagnosed and treated for choledochal cyst, it is not possible to evaluate the long term outcomes.

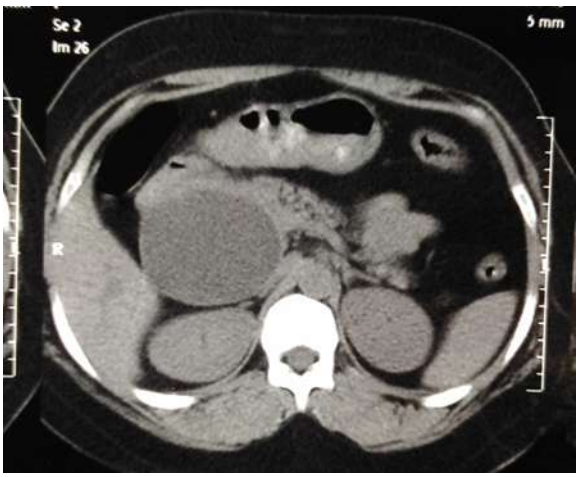

Figure 1

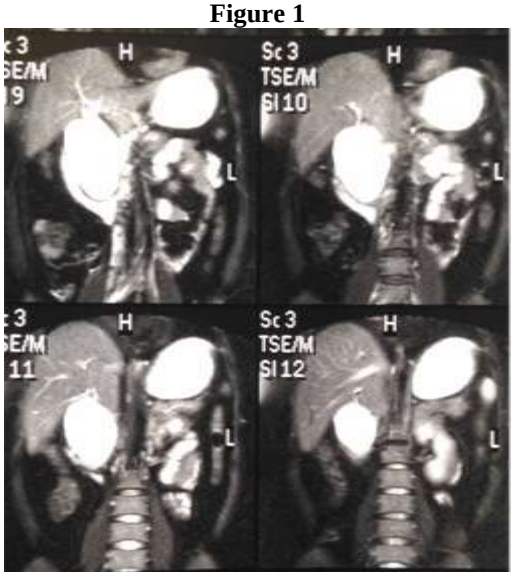

Figure 2

5. Conclusion

Choledochal cysts are congenital anomalies bile duct leading to cystic dilatation of extrahepatic and /or intrahepatic biliary radicles. Most of the cases presented before puberty. Pain in abdomen is the most common clinical presentation. However the classical triad of pain abdomen, jaundice and mass per abdomen may not be found. Ultrasonography of

abdomen can be used as a single modality for diagnosis of choledochal cyst. Complete cyst excision with cholecystectomy followed by biliary reconstruction using Roux-en-Y hepaticojejunostomy, is the treatment of choice. Choledochal cyst disease has good outcome, if operated, with less postoperative complications.

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