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Prospective study of clinical presentation and management of choledochal cyst

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Abstract

A prospective study of 18 cases of choledochal cysts admitted in a tertiary care hospital is hereby presented. The aims and objectives were to study the clinical presentations and management of these cases. Materials and methods included thorough history and examination , investigations and appropriate operative management of these cases. Mean age of incidence of choledochal cyst was 17.24 yrs and the incidence of classical triad of symptoms(pain abdomen, jaundice and mass) was 33.3%. USG was done in all cases and ERCP/ CTS were done if necessary. Type-I choledochal cyst was commonest presentation while Type- II, III and V were not encountered. Associated complications were present in 60% although incidence of malignancy was zero in our series as against 3-8% in others. Surgical excision of cyst is the main modality of treatment, more so in our population because of poor follow up. Complication rate after surgery is 60% which is high. The present study is limited to patients presenting to a tertiary care hospital hence the incidence and other statistics cannot interpolated to general population for which a large series is needed.

Introduction

Choledochal cyst is defined as an isolated or combined congenital dilatation of the extra hepatic or intrahepatic biliary tree. Vater and Elizer first described the anatomical details of choledochal cyst in 1723(1). Incidence is more frequent in Asians particularly Japanese with more preponderance in females.

Todani et al has described the widely accepted classification of choledochal cysts in 1997 (2), as follows :

TYPE	DESCRIPITION	INCIDENCE
1	Extra hepatic bile duct cystic dilation. 1a. Cystic dilation. 1b. Segmental or focal dilation. 1c. Fusiform or diffuse dilation.	80-90%
2	Diverticulation of extra hepatic bile ducts.	2.5-4%
3	Choledochocele	1.4-5%
4	Multiple cystic dilation of biliary tree. 4a. Intra and extra hepatic bile duct cysts. 4b. Multiple extra hepatic bile duct cysts.	11-19%
5	Caroli's disease 5a. Single /Multiple intrahepatic cysts. 5b. Bile duct cysts without extrahepatic involvement.	1%

Bile duct cysts typically present in infancy and childhood. However in nearly 20% of patients the diagnosis is delayed until adulthood , in whom there is an increased rate of associated biliary pathology. In children the classical findings include a right upper quadrant abdominal mass, jaundice and abdominal pain. In adult it is complicated, so the diagnosis is often confused with benign disease of biliary tract or pancreas.

The diagnosis should always be established preoperatively primarily by ultrasonography followed by cholangiography, to know the associated complications, probable etiology and to delineate the exact anatomy. Other investigation modalities are C.T Scanning , TC99m scanning ,HIDA scanning , MRCP , ERCP and PTC.

The most common complications are recurrent cholangitis , pancreatitis , gallbladder diseases ,carcinoma of biliary tract, cirrhosis with portal hypertension ,portal vein thrombosis and intrahepatic abscess.The risk of cholangiocarcinoma increases with age and is 18% in adults(4).

The management depends on the type of cyst and its associated complications. Choledochal cyst excision with reconstruction via biliary enteric Roux-en-y anastomosis rather than cyst drainage alone, has become the treatment of choice for most types. However type III and type IV choledochal cysts are managed conservatively with stenting and sphincterotomy,

Complications of surgery although very rare include intra-operative bleeding , injury to major vascular structures and

postoperative biliary leak and pancreatic complications.

We hereby present our series of 18 cases of choledochal cysts treated in tertiary care hospital, along with the review of literature.

Aims and objectives

The aim of the study was to analyse cases of choledochal cysts presented to department of surgery in a tertiary care hospital .

1. To study sex, age incidence, clinical presentations and complications of choledochal cysts.

2. To study the diagnostic modalities and surgical management of choledochal cyst.

Materials and Methods

Materials

The study included 18 patients of choledochal cysts in various age

groups who presented to department of surgery .

Methods

1. The *Clinical features* considered are - Abdominal pain, Jaundice, Fever, Abdominal mass and Upper gastrointestinal bleeding.
2. The *Investigations* done are – Routine investigations, Liver function tests, Serum amylase and *Imaging studies* - USG, ERCP, C.T Scan and MRCP.
3. *Management* – The medical management included preoperative stenting of the patient to reduce jaundice levels. The surgical management included Complete excision of cyst with Roux-an-Y-hepatico jejunostomy and Lili's modification in one case.
4. *Complications* following surgery like primary haemorrhage and injury to adjacent structures were looked for.
5. *Histopathological* examination was done of all resected specimens particularly to exclude malignancy.
6. *Statistical analysis* was done along with comparision of data with other studies.

Observations and results

1. *Age incidence*

In present series age group is extending from 7 years to 56 years .There are 15 patients between 0-20 years of age. The mean age is 17.24 years.

2. *Sex incidence*

The study includes 14 female and 4 male patients.

3. *Incidence of clinical features*

All patients in the present series had abdominal pain which was recurrent, episodic, dull aching type in right hypochondrium and epigastric region. Jaundice was seen in 14 patients and Fever in 11 patients. The triad of abdominal pain, jaundice and mass was present in 6 patients. Four patients presented with choledocholithiasis and two with pancreatitis.

The provisional diagnosis before investigations in most patients was either acute or choronic cholecystitis or obstructive jaundice due to CBD stones. In most patients ,choledochal cyst was considered as a differential diagnosis.

4. *Imaging studies*

Ultrasonography of abdomen was done in all patients and 16 cases were diagnosed as choledochal cyst. Further investigation with ERCP, MRCP or CT scan abdomen were done to confirm and define anatomical variations and extent of choledochal cyst. ERCP diagnosed choledochal cyst in 8 patients ,In 2 patients CBD stricture was noted at the lower end and Stenting was done for 3 patients. In most cases surgery was done 3-5 days after ERCP, which should actually be done within 24 hours. 4 patients were subjected to MRCP because of their age and 2 were subjected to CT scan abdomen to delineate morphology and extent of choledochal cyst.

5. *Lab investigations*

11 patients showed raised serum alkaline phosphate levels, 6 patients showed hyperbilirabinaemia, 2patients showed raised SGPT level and 3patients showed high serum amylase indicating possible pancreatitis.

6. *Surgical management*

Preoperative management included antibiotics , stenting, vitamin K injection and bowel preparation. Preoperative stenting as done in 3 patients In all patients cholecystectomy was done. In type-I cases complete excision of choledochol cyst with roux-en-Y-end-to side hepaticojejunostomy was done. In one Type-I with dense adhesions to cyst wall ,Lilly's modification was done. In Type IV cases extrahepatic cysts were completely excised and hepaticojejunostomy was done. T tube was kept in five cases for whom 'T' tube cholangiogram was done on 10th post operative day and T-tube was completely removed in 6 weeks .The post operative stay of patients varied from 5 to 18 days with an averag e of 9 days.

7. Complications of surgery:

Anastomotic leak was seen in 4 cases, Wound infection was seen in 3 cases, 1 case had pancreatic leak ,1 case developed DVT and 1 patient developed shock post operatively and succumbed to death. Significant intraoperative bleeding was not seen in our series.

8. *Type of cysts in present series*

This was determind by imaging studies and surgical exploration.16 patients had type I cyst ,specifically type Ic type.2 patients had type IV cyst while Type II,III and V cyst were not encountered

9. *Complications of cyst*

Cystolithiasis is seen in 3 patients,CBD stones seen in 4 patients, GB stones seen in 1 patient andPancreatitis seen in 2 patients.

Associated hepaticobiliary malignancy is not seen in present series.

Discussion

Choledochal cysts are rare congenital disorders causing dilatation of extrahepatic and rarely intrahepatic bile ducts many of which can present in adult. We present herewith a prospective study of 18 cases admitted in our tertiary care hospital.

• *Sex Variation* - The sex incidence in present series is 3.5:1,which is comparable to the series of Atkinson et al (43). Chijiiwa etal(41) shows high female preponderance ,while other series (39,40)show almost equal incidence(1.2:1).

• *Age Presentation* - The mean age in the present series is 17.24 years, while age group ranges between 7-56 years .In most of other series (25,39)the average age is higher ,as only adults were considered.

• *Presenting Symptoms* - In the present series ,pain abdomen is present in all patients whereas Cholangititis with fever is seen is 61% of cases while hepatomegaly seen in 22%.The classical triad presentation of pain, mass and jaundice in this series is 33.3%,which is higher when compared to other published data(39,41).The presentation of choledochal cyst as mass abdomen in our series is 38% while jaundice was seen in 77%.In Jesudason et al (42) and Atkinso et al (43) series ,the classical tried of pain ,mass and jaundice are not at all seen, but pain in abdomen is the most common presentation in most of the series (41,42).

• *Diagnostic Modalities* - Ultrasonography provides adequate information about the intra and extra hepatic biliary tree and is an extremely useful investigation(42). In present series all patients were subjected to USG abdomen with diagnostic accuracy of 90% and 5 patients were operated based on the USG findings alone. ERCP has 100%accuracy in diagnosing choledochal cyst and its associated anomalies (eg APBDJ), but is associated with complications like pancreatitis and cholangitis. MRCP is gaining ground against ERCP, as it is non invasive ,but accuracy in diagnosing anomalies is less compared to ERCP. Demonstration of stagnation of bile by HIDA scan in a dilated bile duct in the absence of intraductal calculi or biliary obstruction has been reported as a diagnostic sign(42) .

• *Laboratory Investigations* - They have a supportive role in assessing complications associated with choledochal cysts. In the present series, 4 patients had normal LFT while others had abdominal LFT with 3 patients showing raised serum amylase in addition. In the series of Jesudason et al (42), liver function tests were normal in 0.5% of the patients ,while serum bilirubin and alkalin phosphatase were elevated in 28% and 61.4% respectively. In the series of Atkinson et al (43)abnormal liver function tests were recorded in 69%.

• *Surgery* - The ideal treatment of choledochal cyst is its total excision . In type I and V, this involves complete excision of the bile duct from the confluence of the hepatic duct proximally up to the pancreatico biliary junction

distally and restoration of biliary–enteric communications by a Roux-en-Y hepatico -jejunostomy(42). In situations where the intensity of fibrosis precludes safe periductal or pericystic dissection, it is advisable to follow Lilly's technique(42) i.e. mocosectomy. Treatment options for choledochal cysts of type I and IV are considered optimal in the present series and the compared series(41,42). Anastomotic leak and wound infection are the common post operative complications (42,43). Post operative cholangitis was seen in 40% cases in the series of Atkinson et al (43) caused by the development of anastomotic stricture.

• *Associated complications of cyst* - The total percentage of complications associated with the cyst at presentation in this series is 55.5% which is slightly higher when compared to the western series but is same as compared to Indian series (39,42,43).The high rate in the present series may be due to high rate of gallbladder disease and cholangitis associated with choledochal cyst .In the present series the incidence of malignancy and associated cirrhosis is zero, where as in western series the malignancy rate is 28%(5,39).The complications of cystolithiasis is more common in this series . Jesudason et al series(42) also found stones associated with choledochal cyst and gall bladder where as Atkinson et al series(43) found pancreatitis associated with choledochal cyst.

• *Type of cyst* - Type I cyst is the commonest type of cyst in this series ,about 88% which is slightly higher than other series, while the incidence of type IV cyst is 12%,which is comparable to other series .The incidence of type II, III and V cysts is 0% in the present series where as in western series it is 13%,4% and 0.5%respectively. Jesudason et al series shows 1.8% incidence of type V cyst(7,42).

Conclusion

Choledochal cyst is an uncommon entity and needs appropriate evaluation for proper treatment. We have presented a prospective study of 18 cases of choledochal cysts managed in a tertiary care hospital with satisfactory outcome of the operative treatment which is the mainstay of treatment for the same. Majority of the cases were of type 1 in our series and complete excision of the cyst was done in most of the cases. However as the present study is limited to patients presenting to a tertiary care hospital, the

incidence and other statistics cannot interpolated to general population for which a large series is needed.

Conflict of Interests

The authors' declare that there are no conflicts of interests.

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