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Management of Choledochal Cyst: Experience from A Tertiary Care Center of Nepal

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ABSTRACT

Introduction

Choledochal cysts are infrequent congenital cystic dilation of the biliary tract. The aim of this study is to analyze the clinicopathological profile and short-term operative outcomes of patients with choledochal cysts.

Methods

This is a retrospective study of 32 consecutive patients of choledochal cyst who underwent multidisciplinary management in last two and half years at Tribhuvan University Teaching Hospital, Kathmandu, Nepal.

Results

A total of 32 patients, 9 males and 23 females were operated. The average age at diagnosis was 24.67±16.4 years (range from 2 to 56 years). The most common presenting symptoms were pain 31(96.88%), jaundice 10(31.25%) and mass 5(15.63%). Triad of pain, jaundice and mass was present in 4(12.5%). Transabdominal Ultrasonography (100%) was the initial diagnostic modality followed by Magnetic resonance cholangiopancreatography (MRCP) (68.75%), and contrast enhanced computed tomography (CECT) (31.25%). Endoscopic retrograde cholangiopancreatography (ERCP) was done for stent placement in 3 (9.38%) patients with severe cholangitis. Type IVA (37.5%) was the most common type of CC followed by type IC (31.23%), type IB (15.65%), type IA (12.5%) and type IVB (3.12%). Abnormal pancreaticobiliary duct junction was observed in 3 (9.38%) patients. All patients underwent open cyst excision with Roux-en-Y hepaticojejunostomy (HJ). The overall morbidity was seen in 6 patients (18.75%). There was no mortality. None of our patient had cholangiocarcinoma on pathological examination.

Conclusion

Choledochal cyst was common in young females. Type IC and IVA choledochal cyst were the most common types and majority of them were symptomatic. Cyst excision with Roux-en-Y hepaticojejunostomy was the commonest surgical treatment modality and had excellent perioperative outcome

Keywords
Choledochal cyst, cyst excision, Roux-en-Y hepaticojejunostomy

INTRODUCTION

Choledochal cysts (CC) are infrequent congenital cystic dilation of the biliary tract, with an incidence of 1:1000 in Asian population.[1] Choledochal cysts are four times more common in females and are usually diagnosed in infants and young children within the first decade of life. Anomalous pancreaticobiliary duct union (APBDU) as stated in Babbitt's theory is the most accepted hypothesis for CC.[2] The classic triad of abdominal pain, right upper quadrant mass, and obstructive jaundice is unusual and are most commonly seen in pediatric patients.[3] Cholelithiasis, common bile duct stone, cholangitis, pancreatitis, portal hypertension, and liver function test abnormalities are common in patient with CC.[3] CC are usually diagnosed on ultrasound scan of abdomen and pelvis done as a part of routine investigation. Magnetic resonance cholangiopancreatography (MRCP) is emerging as a highly sensitive, safe, and non-invasive diagnostic preoperative technique for CC.[4]

Todani and colleague's modification of Alonso-Lej types is most validated classification for CC used in general practice.[2] Kimura and Komi classification of APBDU are most used classification system in CC.[5,6]

The management of CC is surgical. Type I and IV CC management consists of complete extrahepatic bile duct cyst excision and restoration of bilioenteric continuity.[7] Diverticulectomy followed by primary CBD closure and endoscopic sphincterotomy are treatment of choice for type II and type III CC respectively.[8] Type V (Caroli's disease) management consists of liver resection or Orthotopic Liver Transplant (OLT).[9] This study was conducted with the aim of analysing the presentation, diagnosis and treatment of CC at tertiary care center, Tribhuvan University Teaching Hospital, Kathmandu, Nepal.

METHODS

This is a retrospective study conducted in Department of Gastrointestinal and General Surgery, Tribhuvan University Teaching Hospital, Kathmandu, Nepal. All cases of Choledochal Cyst diagnosed and managed from July 2017 to December 2019 were included. Ethical approval to conduct the study was obtained from Institutional Review Committee of Institute of Medicine, Tribhuvan University.

All patient that underwent surgical treatment for CC were included in the study. The presenting complaints, laboratory investigations, imaging modalities, surgical procedures, postoperative morbidity, and mortality and follow up till 6 months were analysed. Histopathology report of surgical specimen were collected from Pathology Department database system. Follow up of patient were done on outpatient department and by telephone questionnaire. Microsoft Excel for Mac version 16.42 was used for descriptive statistical analysis and calculated mean, median, percentage and standard deviation where appropriate.

RESULTS

Out of total 32 patients who underwent surgery for CC, there were 9 male and 23 females. The mean age was 24.67±16.4 years, the range being 2-56 years. Pain was the most common symptom present in 31 patients (96.88%), followed by jaundice in 10 patients (31.25%) and mass at right hypochondrium in five patients (15.63%). Classical triad was present in four patients. Ten patients had cholelithiasis, six patients had choledocholithiasis. Patients presenting with cholangitis and pancreatitis were five and three respectively (Table 1).

All patients underwent abdominal ultrasonography. Magnetic Resonance Cholangiopancreatography (MRCP) (Figure:1) and Contrast Enhanced Computed

Table 1. Demographics and patient characteristics (n=32)

Variables	Frequency (%)
Age (years)	
Mean	24.67±16.4
Range	2-56
Gender	
Male	9 (28.13%)
Female	23 (71.87%)
Associated symptoms*	
Pain	31 (96.88%)
Jaundice	10 (31.25%)
Mass	5 (15.63%)
Triad	4 (12.5%)
Associated complications	
Cholelithiasis	10 (31.25%)
Choledocholithiasis	6 (18.74%)
Cholangitis**	5 (15.63%)
Pancreatitis	3 (9.39%)
Diagnostic investigations	
Ultrasound	32 (100%)
MRCP	22 (68.75%)
CECT***	10 (31.25%)
ERCP****	3 (9.38%)

** One patient diagnosis was incidental findings during routine abdominal ultrasound*
*** Three patient had severe cholangitis*
**** Two patient had both MRCP and CECT. One patient was referred with CECT from other center and other patient had CECT on the background of hepatolithiasis*
***** ERCP was performed only for therapeutic placement of stent in three patients with severe cholangitis.*

Table 2. Frequency of type of choledochal cyst (n=32)*

Type	Frequency (%)
Type IA	4 (12.5%)
Type IB	5 (15.65%)
Type IC	10 (31.23%)
Type IVA	12 (37.5%)
Type IVB	1 (3.12%)

**Classification was based on radiological findings (radiological findings were consistent with operative findings)*

Table 3. Surgical treatment of choledochal cyst (n=32)

Type	Frequency (%)
Excision of choledochal cyst plus Roux-en-Y hepaticojejunostomy	30 (93.76%)
Lilly's procedure with Roux-en-Y hepaticojejunostomy (HJ)	1 (3.12%)
Excision of choledochal cyst with extraction of right anterior sectorial duct stones plus Roux-en-Y hepaticojejunostomy	1 (3.12%)

Tomography (CECT) was done as diagnostic test in 22 (68.75%) and 10 (31.25%) patients respectively. Endoscopic Retrograde Cholangiopancreatography (ERCP and stenting) was done in three patients with severe cholangitis (Table 1).

The cysts were categorised according to Todani's classification. Thirty seven percent belonged to type IVA, 31.23% to type IC, 15.65% to type IB, 12.5% to type IA and 3.12% to type IVB cysts (Table 2). APBDU was detected on three patients. All patients with APBDU were females, two patients had type 2B and one patient had type 2A APBDU. Komi classification was used for categorisation of APBDU.

Complete surgical excision (Figure 2 and 3) of the extrahepatic bile duct and restoration of biliary enteric communication with Roux-en-Y hepaticojejunostomy (HJ) was possible in 31 patients (Table 3). In one patient cyst was adhered to the portal vein. It was excised by Lily's technique.[10] Four patients developed superficial Surgical Site Infection (SSI) and two patients developed hospital acquired pneumonia during early postoperative period. The overall morbidity was 18.75%. There was no mortality. The histopathology of excised cyst was reported as choledochal cyst in all cases. All cases are on regular follow up.

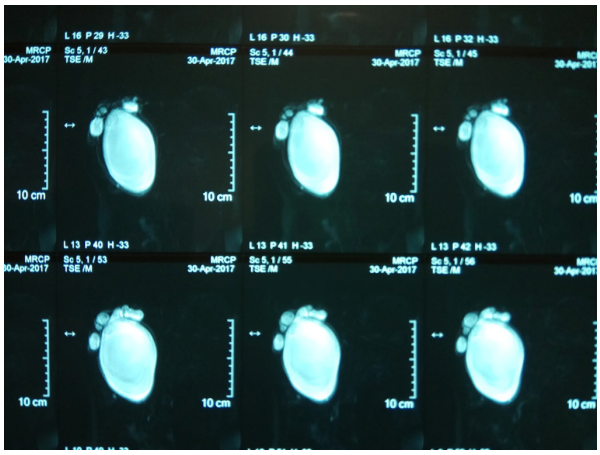

Figure 1. MRCP of choledochal cyst

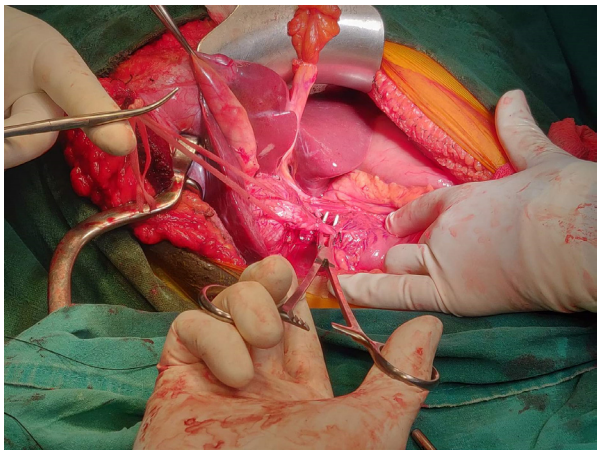

Figure 2. Intraoperative picture of type I choledochal cyst

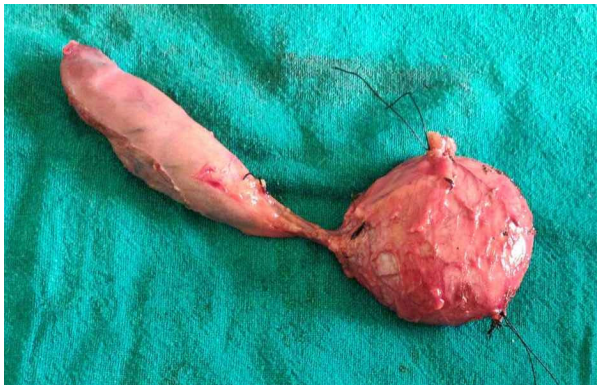

Figure 3. Specimen of type I CC

DISCUSSION

Choledochal cysts are common among Asian population.[1] Choledochal cysts are more common in females and are usually diagnosed within the first decade of life.[11,12] Contradicting to norm, only 34.38% of patient were under 15 years of age with mean age of 24.67±16.4 years. In this study CC was more common among females (71.87%) and male to female ratio was 1:3.9.

Choledochal cysts are believed to be congenital in origin; however, exact etiology remains unknown. Multiple etiologic theories have been proposed for the origin of choledochal cysts. The most widely accepted theory is that cystic dilatation of bile ducts is related to an anomalous pancreaticobiliary ductal union (APBDU). Anomalous pancreaticobiliary duct union (APBDU) as stated in Babbitt's theory is seen in 0-17% of all CC.[2] APBDU was diagnosed in three of our patients. All patients were females, two patients had Komi type 2B and one patient had Komi type 2A APBDU.

According to Singham, abdominal pain in pediatric and adult patients has an incidence ranging from 78% to 90%, jaundice and cholangitis being in 40 to 50%.[13] Similarly, in our study most of our patient presented with abdominal pain (96.88%) followed by jaundice (31.25%) and mass (15.63%). Only, 12.5% patient presented with classical triad. Samuel et al, stated the association of pancreatitis (38%) and cholangitis (14%) in their study.[14] In this study, patients had associated cholelithiasis (31.25%), choledocholithiasis (18.74%) and only 15.63 % and 9.38% patients had associated cholangitis and pancreatitis respectively.

CC is usually picked up by ultrasound scan of abdomen and pelvis (USG) done as a part of routine investigation. Well performed USG is very sensitive (71 to 97%) in the detection of CC.[15] All patients 16 years above (68.75%) underwent MRCP as definitive diagnostic tool. Most of the pediatric patient (31.25%) underwent CECT abdomen and pelvis as secondary diagnostic tool following USG. ERCP was performed in three patients for therapeutic stenting for severe cholangitis.

Alonso-Lej et al published the first systematic description of choledochal cysts, based on the clinical and anatomic findings in 96 cases.[15] Five types of CC are described and classified as per Todani classification of CC. Type I cysts are dilatation of the extrahepatic bile duct. Type I choledochal cysts make up about 50 to 80% of all choledochal cysts, type II 2%, type III 1.4 to 4.5%, type IV 15 to 35%, and type V 20%. In this study, type IVA was most common type (37.5%). Second common CC was type IC (31.23%) followed by type IB (15.65%), type IA (12.5%) and type IVB (3.12%). There were no cases of type III and type V in our study.

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.[16] Endoscopic sphincterotomy and cyst unroofing have become the treatment of choice for type III CC.[17] Surgical treatment of Caroli's disease type V ranges from resection if the disease is unilobar to liver transplantation when diffuse disease is detected. In this study, 31 patients underwent Excision of choledochal cyst plus Roux-en-Y hepaticojejunostomy. In one patient cyst was adherent to portal vein so, it was excised by Lily's technique.[10]

In the patients with CC, in addition to the 5% risk of gallbladder cancer, the risk of cholangiocarcinoma is 14%.[18]The histopathology of all our CC were benign. There were no cases of carcinoma gallbladder or cholangiocarcinoma.

CONCLUSION

Choledochal cysts was more common in young females. The most common presenting symptom was pain. All patient with CC were diagnosed using ultrasound abdomen and pelvis and MRCP and CECT abdomen and pelvis were done as an adjunct for proper delineation of biliary anatomy. Type IC and IVA were common types of CC. Surgical excision of the cyst with bilioenteric anastomosis was the procedure of choice and it had excellent early postoperative outcomes.

CONFLICT OF INTEREST

None declared.

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