AUDIT OF MANAGEMENT OF ADULT CHOLEDORHAL CYSTS

Ammad Ud Din Nasir, Muhammad Shoaib Khan, Anas Bin Saif*, Qasim Butt, Hanif Abbasi
Pak Emirates Military Hospital/National University of Medical Sciences (NUMS) Rawalpindi Pakistan, *FTC Kotli Pakistan

ABSTRACT

Objective: To analyze the complexity and diversity of type, surgical management and complications in adult choledochal cysts presenting to a Hepatobiliary unit of a tertiary care hospital.

Study Design: Case series.

Place and Duration of Study: Pak Emirates Military Hospital Rawalpindi, from Jan 2017 to Dec 2019.

Methodology: All the patients diagnosed with choledochal cyst and underwent surgical procedure between Jan 2017 to June 2019 were analyzed. The clinical features, types, operative procedure and outcomes of the patients in terms of post-operative morbidity and mortality were assessed. Calvin Dindo classification was used to assess complications.

Results: A total of 17 patients were studied, out of which, 13 were females (76.5%) and 4 were males (23.5%) with mean age of 37.41 ± 16.96 years. There were 13 type I (76.5%), 2 type IVa (11.8%) and 1 type II (5.9%) choledochal cysts according to Todani’s classification. Extrahepatic cyst excision with a Roux-en-Y hepatico-jejunostomy was performed on all 17 patients. There was no mortality in the series. Post-operative complications occurred in 4 (23.5%) patients, 2 were grade II, 1 was grade I and 1 grade III according to Clavin Dindo scale. Long-term follow up revealed recurrent cholangitis in 3 (17.6%) patients which settled on antibiotic regime. The median follow up time was 1.5 years.

Conclusion: The total extra-hepatic excision with Roux-en-Y hepaticojejunostomy is the treatment of choice for adult choledochal cyst. The procedure has low morbidity and very low mortality.

Keywords: Choledochal cyst, Hepatobiliary disease, Obstructive jaundice.


INTRODUCTION

Choledochal cyst is a cystic dilatation of the biliary system. It is among the rarest congenital anomalies and is estimated to be present in 1 in 13,000 to 1 in 2 million live births.1 Although it is considered to be common in children, yet 25% of choledochal cysts present later in life.3,4 In adults, it is often associated with complications making the diagnosis and subsequent treatment more difficult.5 Todani and his colleagues devised a method for classifying choledochal into five subtypes, which is widely accepted. The most common type is Type-I that is fusiform dilatation of common bile duct.6

Choledochal cyst is more common in Asian population with about half of the reported cases are from Japan.4 It is four times more common in female population.7 The classical triad of pain right hypochondrium, upper abdomen mass and jaundice is present in only a third of adult patients. The most dreadful complication of choledochal cyst is development of cholangiocarcinoma and the risk increases with age. Other complications include cholangitis, pancreatitis and choledocholithiasis. Ultrasonography is the first line investigation modality to see the biliary tree with sensitivity of 71-97% in diagnosing choledochal cyst. Computed tomography is performed to delineate the intra and extra-hepatic anatomy and evaluate the surrounding relationships of cyst. The gold standard for diagnosing choledochal cyst is Magnetic Resonant Cholangio Pancreatography (MRCP) with sensitivity of 90-100%.8

Initially choledochal cysts were treated by drainage procedures (cysto-jejunostomy, cystoduodenostomy) but they have been abandoned due to high complication rates and stricture formation.4 Currently complete excision of the cyst with Roux-en-Y hepaticojejunostomy is the recommended treatment of choice.9 It is pertinent to mention that complete excision of cyst does not abolish risk of carcinoma completely.10

The rationale of this study was to analyze the presentation and types of choledochal cysts in adult Pakistani population, demographic distribution, radiological and laboratory investigations and to evaluate the type of surgical procedures done and compare short and long-term outcomes in terms of morbidity and mortality.

Correspondence: Dr Anas Bin Saif, House No. 10, Street no. 14, Sector G, DHA Phase-II, Islamabad Pakistan
Received: 13 Jun 2020; revision received: 26 Nov 2020; accepted: 01 Dec 2020

METHODOLOGY

This case series was conducted at Pak Emirates Military Hospital Rawalpindi after approval of Ethical Committee (lt no. A/28/1), from January 2017 to June 2019. Sample size was calculated by WHO sample size calculator keeping the confidence interval of 95% and prevalence of choledochal cyst to 0.1%. The data was collected by non-probability convenient sampling technique.

Inclusion Criteria: All the patients with age more than 18 years with characteristic symptoms, signs and radiological confirmed diagnosis of choledochal cyst were included in the study.

Exclusion Criteria: Patients with other causes of obstructive jaundice, pain abdomen or with malignant disease were excluded from the study.

All the patients underwent ultrasonography and Magnetic Resonant Cholangio Pancreatography, while 2 patients also had CT scan to rule out suspected pathology other than choledochal cyst. A proforma was designed to including demographic features of the patients, initial signs and symptoms, relevant laboratory results, radiological studies, per-operative findings and post-operative recovery. Follow up was done up to maximum period of two years for post-operative morbidity. Analysis of various signs and symptoms, operative findings, co-morbidities and relevant investigations was done. Histological and radiological co-relation were also carried out to find out the particular type of choledochal cyst. The types of choledochal cysts, were assigned according to the classification of Todani et al. Calvin Dindo 2 classification was used to assess complications.

The demographic data was analyzed using Statistical Package for the Social Sciences (SPSS) version 21 and mean with standard deviation was calculated for age. Frequency and percentages were calculated for ordinal variables such as symptoms, signs and radiological findings.

RESULTS

There were 17 patients that were included in the study out of which 13 were females (76.5%) and 4 were males (23.5%). Age of patients ranged from 18-70 years with mean age of 37.41 ± 16.96 years. Four (23.5%) patients were over 50 years of age. Five (29.4%) patients had some co-morbidity, out of which 3 (17.6%) had diabetes mellitus and 2 (11.8%) had more than one disease. The main complaint of patients was pain right hypochondrium present in 14 (82.4%) patients followed by jaundice in 3 patient (17.6%) while 2 patients had classical triade of fever jaundice and mass as shown in Figure.

Figure: Frequency of signs associated with choledochal cysts (n=17).

Important positive abdominal examination findings included tenderness right hypochondrium in 14 cases (82.4%), mass in upper abdomen in 3 patients (17.6%). One (5.9%) patient had previous cholecystectomy and one (5.9%) had endoscopic retrograde cholangiopancreatography. There were deranged liver function tests in 5 (29.4%) cases showing obstructive pattern (raised Alkaline Phosphatase and bilirubin). Three (17.6%) patients had raised white cell count depicting presence of infection. Ultrasonography was performed in all cases followed by Magnetic Resonant Cholangio Pancreatography (MRCP) which diagnosed choledochal cyst accurately. The final diagnosis of choledochal cyst was made per-operatively and on histopathological report of specimen. Follow-up data was obtained via a review of patients’ medical records and through follow-up visits. Histopathological confirmation of the diagnosis was obtained in all cases. There was type I choledochal cyst in 13 (76.5%) cases, whereas type IVa cyst was present in 3 cases (17.6%) and one (5.9%) case had type II cyst. Fifteen (88.2%) patients had some sort of complication of choledochal cyst, 4 had stone in common bile duct, 4 had cholangitis and 5 had obstructive pattern of liver enzymes while 1 had stricture in distal part and 1 patient had repeated bouts of pancreatitis as shown in Table-I.

**Table-I: Complications associated with choledochal cysts in adults (n=17).**

<table>
<thead>
<tr>
<th>Complications</th>
<th>n (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biliary-lithiasis</td>
<td>4 (23.5)</td>
</tr>
<tr>
<td>Cholangitis</td>
<td>4 (23.5)</td>
</tr>
<tr>
<td>Stricture</td>
<td>1 (5.9)</td>
</tr>
<tr>
<td>Pancreatittis</td>
<td>1 (5.9)</td>
</tr>
</tbody>
</table>
All patients underwent excision of common bile duct and Roux-en-Y Hepatico-jejunostomy after optimization. 5/0, and 4/0 PDS was used t-enteroent stomy was done about 60 cm below the loop with 3/0 PDS continuous. There was no mortality. However 4 patients (23.5%) developed postoperative complications. Two patients (11.8%) developed bile leaks and one patient (5.9%) had basal atelectasis while one had seroma (5.9%). These have been classified according to Calvin dindo 2 classification in Table-II. The median postoperative length of hospital stay was 8 days (range 6-12 days).

DISCUSSION

The exact cause of choledochal cyst is not known. One explanation can be due to the abnormal pancreaticobiliary junction (APBJ) which causes backlash of pancreatic juice into the common bile duct (CBD). This anomaly is frequently encountered in choledochal cyst.11-13

Adult Choledochal cysts mostly present with complicated clinical and pathological features that alter surgical management as seen in study by Nagoney et al,14 this complex presentation also seen in present study. Similarly, most choledochal cysts occur in women, also seen in present study with 76.5% patients being females in our study. This high incidence in female population was also seen in studies of Hewitt et al, Stain et al.16-19 The present study also confirms that adult choledochal cyst patients usually have vague signs and symptoms. The most common clinical presentation of adults with choledochal cyst was non-specific abdominal pain in 82.4% of our patients followed by cholangitis and jaundice similar to previous studies reporting nonspecific abdominal pain occurring in 70% of patients.16-19 A palpable mass is uncommon as observed by O’Neil in his study20, similarly in present study, only 3 patients with palpable mass.

The symptoms of choledochal cyst are sometimes indistinguishable from those of biliary calculus diseases. In adults, the choledochal cyst is present alongside some other hepatobiliary conditions (e.g. biliary lithiasis pancreatitis and malignant disease) resulting in requirement of an individualized therapeutic approach.

Abdominal Ultrasound and CT scan usually delineate the cyst. There could be an atypical pancreatic-biliary union; such anomalous junctions can be delineated by ERCP.21 In present study, ERCP was done in only one patient du before referral to our center. We did not do ERCP in any of our patients it can lead to severe pancreatitis in patients with choledochal cyst.

Choledochal cyst can be associated with pancreatitis as seen in one of our patients.16 The commonest type of cyst was type 1 which has also been confirmed in other studies.22 A rather dreadful and well-recognized complication of choledochal cyst is the development of cancer namely cholangiocarcinoma. It is pre-sumed to develop in patients who went undiagnosed. Similarly, tumor is also seen to form in a cyst that was mistreated by cyst enterostomy. The incidence of carcinoma in choledochal cyst ranges from 2.5-15.6%.23 However, in our series none of the patient had malignancy on histopathology. This absence of malignancy is histopathology specimen was also observed in other studies.22 The treatment of cancer developing due to choledochal cyst merits radical excision of the biliary tract as the disease is not limited to the cyst. The cancer can be present in other areas of hepatobiliary system such as the gallbladder, pancreas and liver.24-25

The gold standard treatment recommended by most centers around the world is total extrahepatic cyst excision with Roux en-Y hepaticojejunostomy.21 In our study, same procedure was adopted, 4/0 PDS was used for the anastomosis, like in other studies.22 Cystoenterostomy is no longer recommended. Like other studies, we also found that hepaticojejunostomy is not only safe but also prevents morbid issues as residual cyst, bile stasis, recurrent cholangitis, formation of de novo stone, pancreatitis and carcinoma of bile duct. 16,17,19 It is also recommended that adults diagnosed on screening or incidental must undergo complete excision.25 In this series, 3 (17.6%) out of 17 patients with type I cysts who had undergone a hepaticojejunostomy developed cholangitis. This result is comparable to overall literature which quotes incidence of 18% post-operative cholangitis if the recommended surgery is undertaken in a specialized hepatobiliary unit.22

Table-II: Types of post-operative complications according to clavin dindo grades (n=4).

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Type of Cyst</th>
<th>Age(years)</th>
<th>Gender</th>
<th>Complication</th>
<th>Clavin Dindo Scale</th>
<th>Management</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Type I</td>
<td>50</td>
<td>Female</td>
<td>Minor leak</td>
<td>Grade II</td>
<td>Conservative</td>
</tr>
<tr>
<td>2</td>
<td>Type IVa</td>
<td>47</td>
<td>Male</td>
<td>Major leak</td>
<td>Grade III</td>
<td>Redo-anastomosis</td>
</tr>
<tr>
<td>3</td>
<td>Type I</td>
<td>65</td>
<td>Female</td>
<td>Basal atelectasis</td>
<td>Grade II</td>
<td>Conservative</td>
</tr>
<tr>
<td>4</td>
<td>Type I</td>
<td>38</td>
<td>Female</td>
<td>Seroma</td>
<td>Grade I</td>
<td>Conservative</td>
</tr>
</tbody>
</table>

CONCLUSION

After experience of surgical management of 17 patients, we can safely conclude that adult choledochal cyst should be treated by total extrahepatic cyst excision and this is a safe procedure with low morbidity and mortality. However, cases are best managed in a specialized tertiary surgical units for satisfactory post-operative outcome.

Conflict of Interest: None.

Authors’ Contribution

AUDN: Data collection, data analysis, MSK: Data collection, ABS: Data collection, analysis, QB: Supervisor, HA: Data analysis.

REFERENCES


