CLINICAL PRESENTATION AND MANAGEMENT OF ADULT CHOLEDOCHAL CYST: A RETROSPECTIVE REVIEW

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INTRODUCTION

Choledochal cyst is a rare congenital disease of the biliary tree with an incidence ranging from one in 13,000 to one in two million¹. Almost 60% present before the first birthday² but adult presentation is not a rarity³. The commonest theories for its causation are weakness in the wall of the bile ducts³⁻⁴, obstruction of the distal choledochus⁵, combination of obstruction and weakness⁶ and reflux of pancreatic enzymes into the CBD secondary to an anomaly of the pancreatico-biliary junction⁷⁻¹³. The most widely accepted and found cause is said to be the last theory. All these can explain the causation of type I, III and IV anomalies, but not type II and V, in which the CBD is normal. Perhaps genetic causes may be responsible for it¹⁴⁻¹⁶.

Choledochal cyst is more common in females¹⁴ and asians¹⁵. The risk of malignancy is always there in the intra and extra-hepatic bile ducts of patients with choledochal cyst. This risk appears to be age related and even complete cyst excision does not confer complete immunity¹⁷⁻²³. Adults usually present with the complication of the disease and the classic triad of pain, jaundice and a palpable mass is found only in 30% of patients²⁴. Ultrasound is the best initial investigation that is diagnostic in most of the patients. Complete cyst excision was possible in 69.56% of the patients. Complete cyst excision followed by Roux-en-Y common hepatico-jejunostomy was performed in 30.43% and common hepatico-dudenostomy in 26.08%.

Conclusion: The most common mode of clinical presentation of choledochal cyst was pain right hypochondrium with raised serum bilirubin and ALT levels. The commonest surgical procedure was complete cyst excision with Roux-en-Y common hepatico-jejunostomy.

Key Words: Choledochal cyst, Biliary diseases, Biliary bypass procedures

ABSTRACT

Objectives: To determine the mode of presentation and commonest surgical procedures done for adult choledochal disease in a tertiary care hospital.

Methodology: A retrospective review conducted in Lady Reading Hospital, Peshawar, from January 1995 to January 2005 with one year of follow up. All patients above 13 years of age with choledochal cyst were considered for the study. Mode of clinical presentation and commonest surgical procedures were recorded.

Results: Of the total 23 patients collected, 08 (34.78%) were males and 15 (65.22%) were females. Regarding mode of clinical presentation, pain right hypochondrium with raised serum bilirubin and ALT levels was found in 21.73% of patients. Complete cyst excision was possible in 69.56% of the patients. Complete cyst excision followed by Roux-en-Y common hepatico-jejunostomy was performed in 30.43% and common hepatico-dudenostomy in 26.08%.

Conclusion: The most common mode of clinical presentation of choledochal cyst was pain right hypochondrium with raised serum bilirubin and ALT levels. The commonest surgical procedure was complete cyst excision with Roux-en-Y common hepatico-jejunostomy.

Key Words: Choledochal cyst, Biliary diseases, Biliary bypass procedures
CLINICAL PRESENTATION AND MANAGEMENT OF ADULT CHOLEDOCHAL CYST: A RETROSPECTIVE REVIEW

METHODOLOGY

It was a retrospective review conducted in Lady Reading Hospital from January 1995 to January 2005 with one year of follow up. All patients above 13 years of age with choledochal cyst were considered for the study. Patients who underwent operation on their choledochal cyst in the past were excluded from the study (6 patients). All patients were diagnosed on abdominal ultrasound and ERCP/MRCP was done in most of the patients. Patients were admitted to our general surgical ward and preliminary investigations like Hb, blood urea/creatinine, blood sugar, urine RE, LFTs, PT/APTT (INR), CXR, ECG, Serum electrolytes and Blood cultures were done.

Mode of clinical presentation and commonest surgical procedures were recorded. The aim of surgery was to excise the cyst completely where possible followed by some drainage procedure preferably Roux-en-Y chledocho-jejunoanstomy. After prophylactic intravenous ceftriaxone 2 grams BD, Metronidazole 400 mg TDS and IV fluids for rehydration, patients were explored via right subcostal incision that can be converted to roof top incision if wider exposure was needed. The desired surgical procedure was done. 3/0 PDS or Vicryl was used as a suturing material for anastomosis (single layer and interrupted). Simple soft tube drain was place near the biliary anastomosis and nasogastric decompression was done in all patients.

Short term success of surgery was no bile leak and healing of the bypass procedure. Long term success was the absence of anastamotic stricture, no episode of cholangitis and no emergence of cholangiocarcinoma. They were followed monthly for 12 months. MRCP was done in all patients at 6 and 12 months of follow up visits.

RESULTS

A total of 23 patients; 8 (34.78%) males and 15 (65.22%) females were enrolled in the study. Regarding mode of clinical presentation, pain right hypochondrium with raised serum bilirubin and ALT levels was found in 5 (21.73%) of patients. The modes of presentation are shown in table 1. The various types of cyst encountered are shown in table 2.

The surgical procedures done are shown in table 3. Complete cyst excision was possible in 16 (69.56%) patient. Complete cyst excision followed by Roux-en-Y common hepatico-jejunoanstomy was performed in 7 (30.43%) and common hepatico-duodenostomy in 6 (26.08%). Cyst excision was not possible in 7 (30.43%) patients. In 7 (30.43%) of the cases of Roux en Y hepatico-jejunoanstomy (RYHJ), the drain was dry within 1 week of surgery and there was no case of anastamotic breakdown or stricture formation at follow up MRCP. Only 1 (04.34%) patient of RYHJ encountered two episodes of cholangitis needing admission and IV antibiotics. However, MRCP in the follow up period showed no stricture or dilation of intra hepatic ducts.

Cyst excision with choledocho-deudenostomy (CD) was done in 6 (26.08%) of patients. In this procedure drain was also dry within one week of surgery and there was no anastomotic leak. However the incidence of cholangitis was in 3 (50%) patients in the follow up period of one year duration needing admission and IV antibiotics. Post-op MRCP at 6 months showed no stricture at anastomotic site.

All the 3 patients with loop common hepatico-loop jejunostomy and entero-enterostomy suffered from recurrent episodes of cholangitis in the post-operative period. Cysto-jejunostomy and cysto-deudenostomy experienced repeated episodes of cholangitis and all needed excision of the cyst and RYHJ at some stage of their one year follow up. One patient of external biliary drainage underwent cyst excision and RYHJ 03 months after their first surgery. There was no operative or post-operative deaths upto one year of follow up in any patient of the study.

DISCUSSION

Most of the choledochal cysts presented as complications of the disease like infection or stones in the choledochal cyst. Their presentation did not follow any classical patterns and all experienced abdominal pain and most had episodes of cholangitis. Asymptomatic cases that were accidentally diagnosed on ultrasound were only 8.69%.

The commonest type of choledochal cyst was Todani type I that was found in 65.21%. Todani type I is a commonly found cyst on ultrasound as it can be picked up easily by most of the ultrasound specialists. They rest of the types are deceptive and need ERCP or MRCP. This often results in delayed diagnosis or end up in in-appropiat upper abdominal surgery.

The various surgical procedures offered depended upon the type of cyst, surgeon preference and expertise. In less experienced hands, when cyst presents as emergency, often end up in inadequate procedures. In 30.43% of the cases choilodal cyst could not be resected because of cyst infection and cholangitis. In these cases it was bypassed either to duodenum (13.04%), a jejunal loop (13.04%) or was drained to exterior via T-tube (4.34%) as shown in table 3. These cases however can always be re-assessed latter when stable and considered for proper resection and drainage procedure as was done in our study. The only problem with this group of treated patients is that they tend to stop coming for follow up once discharged. The intemtional literature reports that 80% of such patients will have re-
Table 1: Modes of presentation of choledochal cysts (n=23)

<table>
<thead>
<tr>
<th>S. No.</th>
<th>Mode of Presentation</th>
<th>No. of pts</th>
<th>%age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Epigastric Pain with raised SBR and ALT**</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>2</td>
<td>RHC Pain with raised SBR and ALT**</td>
<td>5</td>
<td>21.73%</td>
</tr>
<tr>
<td>3</td>
<td>Epigastric Discomfort with raised SBR and ALT**</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>4</td>
<td>RHC Discomfort with raised SBR and ALT**</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>5</td>
<td>Chronic Cholecystitis*</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>6</td>
<td>Mass RHC with discomfort**</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>7</td>
<td>Cholangitis (pain, fever and rigors)*</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>8</td>
<td>Asymptomatic</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>9</td>
<td>Acute Pancreatitis*,**</td>
<td>1</td>
<td>4.34%</td>
</tr>
<tr>
<td>10</td>
<td>Cirrhosis with Portal hypertension*</td>
<td>1</td>
<td>4.34%</td>
</tr>
<tr>
<td>11</td>
<td>Collangiocarcinoma*</td>
<td>1</td>
<td>4.34%</td>
</tr>
<tr>
<td>12</td>
<td>Traumatic rupture with Peritonitis*</td>
<td>1</td>
<td>4.34%</td>
</tr>
</tbody>
</table>

* Complications of the disease.
** With stones in the choledochus

Table 2: Types of choledochal cysts encountered (n=23)

<table>
<thead>
<tr>
<th>S.No</th>
<th>Type of Cyst</th>
<th>No of pts</th>
<th>%age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Todani Type I</td>
<td>15</td>
<td>65.21%</td>
</tr>
<tr>
<td>2</td>
<td>Todani Type II</td>
<td>2</td>
<td>8.69%</td>
</tr>
<tr>
<td>3</td>
<td>Todani Type III</td>
<td>1</td>
<td>4.34%</td>
</tr>
<tr>
<td>4</td>
<td>Todani Type IV</td>
<td>3</td>
<td>13.04%</td>
</tr>
<tr>
<td>5</td>
<td>Todani Type V</td>
<td>2</td>
<td>8.69%</td>
</tr>
</tbody>
</table>

Table 3: Various surgical procedures done (n=23)

<table>
<thead>
<tr>
<th>S.No</th>
<th>Surgical Procedure</th>
<th>No of pts</th>
<th>%age</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Cyst Excision with Roux-en-Y Common hepatico-jejunostomy</td>
<td>7</td>
<td>30.43%</td>
</tr>
<tr>
<td>2</td>
<td>Cyst Excision with Common hepaticoduodenostomy</td>
<td>6</td>
<td>26.08%</td>
</tr>
<tr>
<td>3</td>
<td>Cyst Excision with Loop Common hepaticojejunos-tomy and Enteroenterostomy</td>
<td>3</td>
<td>13.04%</td>
</tr>
<tr>
<td>4</td>
<td>Cystoduodenostomy (cyst not excised)</td>
<td>3</td>
<td>13.04%</td>
</tr>
<tr>
<td>5</td>
<td>Cystojejunostomy (cyst not excised)</td>
<td>3</td>
<td>13.04%</td>
</tr>
<tr>
<td>6</td>
<td>External Biliary Drainage (cyst not excised)</td>
<td>1</td>
<td>4.34%</td>
</tr>
</tbody>
</table>

Current symptoms and 69% will subsequently need revisional surgery. In our study 17.39% of patients underwent revisional surgery within one year of their follow up.

One patient had advanced cholangio carcinoma, with metastasis in the liver and obstructive jaundice. Choledochal cyst is more common in Asian and in females. We also found a higher incidence in females as compared to males (65.22% vs. 34.78% respectively).
Studies show that nearly 80% of cysts in adults present with complications like cholangiocarcinoma, cystolithiasis, cholelithiasis, cholangitis and liver cirrhosis. Complications of choledochal cyst were found in 36.36% of patients in our study. The commonest type of cyst reported in literature is Todani type I, IV and V. We have observed a single case of cholangio-carcinoma with metastasis in the liver, in a 55 years old female having Todani type V cyst.

Type II cyst was treated by simple excision as recommended. Now a days laparoscopic excision is also carried out with equivalent results. Type III cyst was treated by choledocho-duodenostomy with no complications noted in 1 year of follow up, although the best treatment of such cysts is un-roofing of the choledocholecce so that both the ducts drain directly in to the duodenum. Type IV and V cysts were explored for cholangitis and stones in the biliary passages and after stones removal and wash of the biliary passages choledocho-duodenostomy was performed. This procedure brought a decrease in the frequency and intensity of cholangitis in these patients, although the standard treatment for such patients is either partial hepatectomy for localized disease or liver transplantation for the diffuse variety.

**CONCLUSION**

The most common mode of clinical presentation of choledochal cyst was pain right hypochondrium with raised serum bilirubin and ALT levels. The commonest surgical procedure was complete cyst excision with Roux-en-Y common hepatico-jejunostomy.

**REFERENCES**


CONTRIBUTORS

NAS conceived the idea, planned the study, and drafted the manuscript. TM, MK, MH, IS and JA helped acquisition of data. AH did statistical analysis and critically revised the manuscript. All authors contributed significantly to the submitted manuscript.