ORIGINAL ARTICLE

Type I choledochal cyst: A single institution study

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Abstract

Background: Congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract known as a choledochal cyst (CC) are uncommon, yet well-documented. They carry significant morbidity if not recognized and treated early.

Aim: (1) To correlate with the clinical presentation of Type I CC, (2) To classify Type I CC into If and Ic.

Materials and Methods: Between 2009 and 2014, 20 cases of CC were managed. The clinical presentation, radiological findings, the gross and microscopy of the excised specimen (CC and gall bladder) and outcome were analyzed. All patients underwent cyst excision with Roux-en-Y hepaticojejunostomy (RYHJ) as the mode of treatment. CCs were classified based on King’s College hospital classification of choledochal malformation.

Results: The patients were divided into two broad categories: Children (n = 16) and adult group (n = 4). All children presented with fever and jaundice, whereas all adults presented with pain abdomen. Among adults only one had classical triad of pain abdomen, jaundice, and mass in right upper quadrant. Among children Type If and Ic were equal and in adults Type Ic was more common. Most of the cases had a good outcome except for one death due to complications of cirrhosis. Incidental malignancy in two adult cases were reported.

Conclusion: CCs have no classic clinical features. Ultrasonography is the major diagnostic tool. The best surgical treatment seems to be complete excision of the cyst and RYHJ.

Introduction

Congenital dilatation of the extrahepatic biliary tract with or without dilatation of the intrahepatic biliary tract, first described in 1852 by Douglas is known as the choledochal cyst (CC).[1] They are uncommon, yet well-documented and can carry significant morbidity if not recognized and treated early. Classically, this disease has a higher incidence of diagnosis during the first decade of life with a predominance in females of 3-4:1.[2] These are one of the common cause of cystic mass in the pediatric abdomen.[3] Diagnostic and therapeutic applications for CC have changed quickly in the last decades. Development of malignant tumors and biliary cirrhosis due to CC has increased the importance of management for the surgeons.[4] In both adults and children, total excision of the cyst with Roux-en-Y hepaticojejunostomy (RYHJ) is the treatment of choice.[5]

Materials and Methods

Between 2009 and 2014, 20 cases (age 2 months-51 years) of CC were managed. The clinical presentation, radiological findings, the gross and microscopy of the excised specimen (CC and gall bladder) and outcome were analyzed. All patients underwent cyst excision with RYHJ as the mode of treatment. CCs were classified based on King’s College hospital classification of choledochal malformation.[6]

Results

The patients were divided into two broad categories: Children (n = 16) and adult group (n = 4). Among children, five were <1 year of age and 11 were >1 year of age. All four adults were females. All children presented with fever and jaundice, whereas all adults presented with pain abdomen. Among four adults, two
had jaundice along with pain abdomen, and one had classical triad of pain abdomen, jaundice and mass in right upper quadrant. Clinical presentation of all children is shown in Table 1.

Pre-operative ultrasonography (USG) documented CC in all cases. Magnetic resonance imaging (MRI) done in two cases suggested Type Ic in adult and Type If in a child [Figure 1c and d]. Based on operative/gross findings, among children eight cases each of anatomical Type Ic and If [Figure 1a and b] were present whereas in adults Type Ic (3 cases) was common than If (1 case). Microscopic examination showed dilated common bile duct with mucosal lining which was absent or present in the patchy distribution. In one child aged 5 years associated changes of pyloric gland metaplasia were highlighted by periodic acid Schiff positivity and in an adult infiltrating adenocarcinoma were observed. Wall showed variable fibrosis in between smooth muscle bundles with mild to absent inflammatory infiltrate. Van Gieson and Masson trichrome special stains were used to demonstrate extensive fibrosis [Figure 1e]. Gallbladder excised along with CC showed no significant pathology except for one case of neuroendocrine carcinoma positive for synaptophysin on immunohistochemistry.

In children, 12 cases had good outcome, three had biliary peritonitis (one preoperatively and two following surgery).

Table 1: Presenting symptoms in children

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Children&lt;1 year (5)</th>
<th>Children&gt;1 year (11)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain</td>
<td>-</td>
<td>2</td>
</tr>
<tr>
<td>Jaundice</td>
<td>5</td>
<td>41</td>
</tr>
<tr>
<td>Clay-colored stools</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>Fever</td>
<td>5</td>
<td>11</td>
</tr>
<tr>
<td>Abdominal distension</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Vomiting</td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>

Figure 1: (a) Type Ic choledochal cyst (CC) in 4 years female child, (b) Type If CC in 4 years female child, (c) Magnetic resonance imaging (MRI) of 51 years female showing Type Ic CC, (d) MRI of 5 years female child showing Type If CC, (e) Masson trichrome stain depicting fibrosis of the wall in 4 years male child (×20) and a 2-year-old child died due to portal hypertension with associated cirrhosis. Two adults were lost to follow-up and two had malignancy.

Discussion

CCs are more common in Asia, two-thirds of cases are from Japan.[6] The incidence of CCs was reported to be <1 in 13,000 to 2 million population.[6-7] Incidence in India is not available, except for one large series of 79 cases from West Bengal, India.[6] Occasional case reports share their experience.

CC is a rare but important cause of jaundice and abdominal pain. In contrast to adults, the classical presentation of jaundice, abdominal pain, and a right hypochondrial mass is rare in children.[9] In our study, pain abdomen and jaundice were the most common presenting symptoms in children and adults. None of the patients except for one adult had classical triad.

The classic triad was seen in <20% of cases. 85% of children and only 25% of adults presented with at least 2 features of the classic triad.[10]

Complications result from bile stasis, stone formation, recurrent superinfection and inflammation. Associated ascending cholangitis and pancreatitis give rise to symptoms in CCs. About 1-12% with spontaneous rupture have symptoms and signs of abdominal pain, sepsis, and peritonitis. The rupture has been hypothesized to be caused by mural fragility from chronic inflammation, increased ductal pressure due to distal obstruction or raised intr-abdominal pressure. At the junction of the cystic and common bile ducts, a site of poor blood flow, rupture is common.[11] In our study, biliary peritonitis was seen in three children, in one before surgery and the other two developed following surgery.

CC should always be considered in the differential diagnosis of jaundice, and a detailed anatomical investigation of the patient with high resolution fasting abdominal USG is frequently diagnostic.[9] In most patients, initial imaging of biliary tree by ultrasound will lead to the diagnosis. Computed tomography (CT) and MRI are useful modalities for diagnosis. Careful treatment decisions need to be made once diagnosed.[11] In the fetus during the prenatal period abdominal USG and CT are both commonly used to demonstrate CC.[4]

In a review of 79 cases, none of their patients were diagnosed antenatally. The youngest patient was 3-month-old male infant. The patients were admitted with abdominal pain in 68 cases (86%), jaundice in 33 cases (42%), lump in 24 cases (30%), and biliary peritonitis due to spontaneous rupture of CC in one patient. Classical triad of pain, lump and jaundice was seen in 17 cases (21.52%).[8]

None of our infants were diagnosed antenatally. The youngest patient was 2-month-old female infant. The patients were admitted with abdominal pain in 6 cases (2 children and 4 adult), jaundice in 19 cases (16 children, 3 adults) lump in one adult and biliary peritonitis due to spontaneous rupture of CC in one patient. Classical triad of pain, lump and jaundice was seen in one adult.
Diagnosis of CC is usually made in childhood.\textsuperscript{[12]} Diagnostic delay appears to be common. Hence, we saw in our setup, more cases in children >1 year of age, similar to Mishra et al.\textsuperscript{[13]} [Table 2].

The diagnosis is delayed in approximately 20% of cases, and these patients might be recognized first as adults with symptoms related to biliary tract pathology.\textsuperscript{[12]} All our adult cases also were identified after an investigation for biliary tract symptoms.

Based on cysts’ location, Todani et al. classified the CC into five types. The classification is accurate and allows pre-operative planning. Distribution of the different types of CC varied.\textsuperscript{[14]}

The subtypes have different etiologies, carcinogenicity, ideal imaging modalities, and optimal treatment strategies. Therefore, clustering all of them within the same disease modality, based solely on anatomy, seems simplistic.\textsuperscript{[15]}

The King George hospital classification utilizes the non-judgemental “choledochal malformation” with whatever descriptive term that suits the nature of the dilatation and has Types I-V. Thus, Type Ic refers to a classical cystic malformation while Type If describes a more fusiform appearance.\textsuperscript{[6]}

Table 2 compares our cases in children with Mishra et al.\textsuperscript{[13]} Table 3 compares our adult cases with Maheshwari P.\textsuperscript{[1]}

Management of CC depends on the type of cyst.\textsuperscript{[16]} Currently, complete cyst excision with RYHJ is widely accepted as the standard method of treatment for most CC\textsuperscript{[17]} and is the treatment of choice in Type I.\textsuperscript{[18]} All adult patients with Type I cysts\textsuperscript{[6]} and 59 of 76 children with Type I or IV cysts underwent radical cyst excision with RYHJ as a primary procedure.\textsuperscript{[9]} All our patients had complete cyst excision with RYHJ.

A treatment plan for newborns and infants is important because of the potential for complications later in life including biliary cirrhosis and portal hypertension.\textsuperscript{[17]} Cyst excision eliminates almost all the potential complications of CC and can be performed safely with no mortality and minimal morbidity even in small infants. There is some evidence that even in patients with biliary cirrhosis and portal hypertension, disease regression is possible after surgical treatment.\textsuperscript{[9]}

In our study, 12 children had good outcome, three had biliary peritonitis and a 2-year-old child died post-operatively due to portal hypertension with associated cirrhosis. In two post-operative deaths, due to overwhelming sepsis one had cirrhotic liver disease with portal hypertension, the other had deep icterus with the poor general condition.\textsuperscript{[13]}

The bile duct is composed of a fibrous tissue in lamina propria with few elastic fibers and lined by a single layer of tall columnar epithelium with few scattered mucus-producing goblet cells in the distal common bile duct. The epithelium in CCs subjected to increased biliary pressure, refluxed proteolytic enzymes and perhaps sepsis, initially shows changes of reactive hyperplasia but later tends to vanish leaving islands of abnormal mucosa in a denuded chronically inflamed fibrous wall.\textsuperscript{[19]} In our study, most of the cysts had patchy areas of epithelium and chronic inflammation, only one case had pyloric metaplasia.

Dysplasia and overt malignancy are age-related features. A number of different histological types of carcinomas have been reported; most are adenocarcinomas of the bile ducts or gallbladder.\textsuperscript{[19]} Among four adults in our study (age 22-51 years), two were lost to follow-up and one had carcinoma in the excised CC and the other in the gallbladder which was excised along with CC.

In the study of 36 cases of CC in the age range of 11-67 years metastasis was found in 40% of the cases (14/35). The most common type of metastasis was pyloric gland (13/14), followed by intestinal (5/14) and squamous metastasis (2/14). Carcinoma was identified in five cases (14.3%). The histologic type was adenocarcinoma in four cases and intraductal papillary neoplasm with high-grade dysplasia in one case. The background cystic lesions showed chronic inflammation and cholesterosis in 73.5% and 11% of the cases, respectively inflammation and cholesterosis were identified in more than 50% of the associated gallbladders; but the gallbladders all lacked dysplasia and carcinoma.\textsuperscript{[19]}

Pancreatic reflux of potent enzymes causing ulceration and increased epithelial turnover, recurrent cholangitis or the irritant effect of biliary tract stones might contribute for propensity to malignant change. As a result of early extensive local and regional spread at presentation; the 5 year survival even for those who undergo surgery is <40% and close to zero for the remainder.\textsuperscript{[16]} The adult patient aged 40 years with incidental detection of gallbladder neuroendocrine carcinoma is alive and the adult patient aged 43 years with carcinoma in CC was referred to oncology center. Both our patients did not have extensive local and regional spread.

Malignancy can be identified as a mass or a focal region of wall thickening on a CT scan. Once diagnosed, careful treatment decisions need to be made.\textsuperscript{[11]} Most cancer arose in the cyst itself.

### Table 2: Comparison of type of cysts in children

<table>
<thead>
<tr>
<th>Number and type</th>
<th>Mishra et al.\textsuperscript{[11]}</th>
<th>Our study</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of cases</td>
<td>Infants&lt;1 year (11) Children&gt;1 year (30)</td>
<td>Infants&lt;1 year (5) Children&gt;1 year (11)</td>
</tr>
<tr>
<td>Type Ic</td>
<td>9</td>
<td>3</td>
</tr>
<tr>
<td>Type If</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Type IVa</td>
<td>-</td>
<td>-</td>
</tr>
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</table>

### Table 3: Comparison of characteristics in adults

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Ic</th>
<th>If</th>
<th>Male</th>
<th>Female</th>
<th>Age (year)</th>
<th>Malignancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Our study</td>
<td>3</td>
<td>1</td>
<td>4</td>
<td>-</td>
<td>22-51</td>
<td>2</td>
</tr>
<tr>
<td>Maheshwari</td>
<td>4</td>
<td>-</td>
<td>3</td>
<td>1</td>
<td>23-75</td>
<td>1</td>
</tr>
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</table>
(50%) or in the gallbladder (46%) and only small percentage in the intrahepatic ducts (~2%); reflecting the gender disparity, most were in females. The presence of a CC changes the time of onset of bile duct malignancy, thus the usual age at onset is in the 3rd decade, 10-20 years sooner than bile duct cancer without cyst.\(^6\)

Surgeons favor complete cyst excision and hepaticoenterostomy. This separates the biliary tree from the pancreatic duct, thus ending the mixing of pancreatic and biliary secretions thought to be responsible for the pathogenesis of the disease; it also excises the damaged and presumably premalignant cyst tissue. If left in-situ, the risk of cancer in the retained cyst is as high as 50% and occurs 15 years earlier than primary cancer. Therefore, the cyst should be excised completely from the hepatic hilum to the pancreatic duct.\(^{16}\)

**Conclusion**

CCs have no classic clinical features. USG is the major diagnostic tool. Rarely, patients present with triad of symptoms in CC. In our study, all patients had jaundice as presenting symptoms, so CC needs to be considered in the differential diagnosis of jaundice. The best surgical treatment for Type I CC was complete excision of the cyst and RYHJ with good outcome in most of the patients. Carcinoma was incidentally diagnosed in adults.

**References**
