ORIGINAL ARTICLE

Characteristics of Choledocal Cyst Patients in Bandung, Indonesia : Single Centre Experience

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ABSTRACT

Introduction: Choledochal cyst (CC) is a congenital cystic dilation of bile duct. Although considered a benign disorder, but CC has various complications like cholangitis, cholelithiasis, pancreatitis and malignant degeneration. The characteristic of this malformation are still not well documented in our region. We aimed to describe the characteristic of CC patients in Hasan Sadikin Hospital Bandung, Indonesia. **Method:** The medical records of patients that were diagnosed with CC between 2014 and 2017 were reviewed. We retrospectively collected demographic data, clinical symptoms, diagnostic imaging and surgical data. **Results:** Fifteen patients were diagnosed as CC, predominantly were girls (12 patients). The mean age was 63,47 months old (range from 7 to 144 months). Jaundic was found in all patients, followed by abdominal pain in 12 patients (80%) and abdominal mass in 9 patients (60%). The laboratory results showed signs of bile obstruction in 5 patients (33,3%). Diagnostic imaging using ultrasonography revealed 5 patients (33,3%) with CC type IV, whereas type I in 6 patients (40%). MRI was performed in 8 patients (53,3%) and CT Scan was performed in 5 patient (33,3%). Eleven patients (73,3%) underwent cyst excision and Roux-en-Y hepaticojejunostomy with liver biopsy. **Conclusion:** Our data suggested that CC predominantly found in girls, and abdominal pain was the most common symptom. Ultrasonography, MRI, and CT are imaging modalities use to diagnose CC. Surgical treatment should be done includes total cyst excision dan biliodigestive reconstruction.

Keywords: Characteristic, Choledochal cyst, Obstructive jaundice

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INTRODUCTION

Choledochal cysts (CC) are a congenital cystic dilatation of the biliary tract. First described by Vater and Ezler in 1723, CC are commonly found in Asia (1). The incidence of CC ranges from 1 in 13.000 individuals in Japan to 1 in 100.000 to 1 in 150.000 individuals in Western countries (1,2,3). Females are predominant for this disease. Nearly 75%-80% of CC are diagnosed in childhood (1,2,3,4). Although the exact cause is still unknown, an anomaly of pancreaticobiliary union are proposed to be the etiology (2,5). The junction of common bile duct (CBD) and pancreatic duct outside the duodenum leads to a long common channel, cause the exposure of biliary epithel to reflux of pancreatic enzymes may contribute to CC formation (1,2,5).

Most anomalies will present in childhood with obstructive jaundice or abdominal pain. The classic triad of pain, jaundice and a palpable mass not commonly found, which result in delays of diagnosis (4,5,6). CC can be associated with severe complications such as cholangitis, cholelithiasis, pancreatitis, cyst perforation, liver failure and malignancy (7,8). Alonso-Lej and colleagues proposed the first CC classification in 1959, but the widely accepted classification was reported by Todani and colleagues in 1977. Five types of CC are described and classified: type I is dilatation of the choledochus (80% to 90% of all CC), type II shows diverticular dilatation, type III is a choledochocele, type IV is a multiple cyst, and type V shows single or multiple dilatations of intrahepatic bile duct. The common diagnostic method is ultrasound, thus the use of computed tomography (CT) scan and the recent advances in magnetic resonance (MR) imaging have provided a new cholangiopancreatography technique, MR cholangiopancreatography (MRCP) (8). The aim of this study is to describe the characteristics of choledochal cyst patients in Hasan Sadikin Hospital Bandung, Indonesia.

MATERIALS AND METHODS

We conducted a retrospective review of hospital medical records for patients admitted to the pediatric surgery division with the diagnosis of a choledochal cyst from January 2014 to December 2017. We did not require the Ethical approval since we did not publish any personal data. A total of fifteen patients were included

and we collected the following data: age of the time of presentation, sex, clinical symptoms, laboratory tests, imaging diagnostic examination, surgical technique used for treatment, and complications. We excluded two patients due to lack of preoperative medical records data. We analyzed data with descriptive statistical analysis by calculating mean, median, and percentage.

RESULTS

We studied a total of fifteen patients, 80% were female (n=12) and 20% were male (n=3), the female to male ratio was 4:1. The average age of presentation was 63.47 months old (range from 7 to 144 months) (Table I).

The most common clinical manifestation was jaundice in all of patients (n=15), followed by abdominal pain, present in 80% of patients (n=12). Palpable abdominal mass was present in 60% of patients (n=9), while acholic stool was found in 46,6% patients (n=7). Fever due to acute cholangitis present in 40% of patients (n=6); 3 patients (20%) were nausea and vomiting when admitted to hospital, while 2 patients (13.3%) had anorexia at the first assestment (Table I).

Table I: Demographic data of patients

Demographic data	Total	Percentage (%)
Sex:		
Male	3	20.0
Female	12	80.0
Age (mean 63.47 months old):		
0 – 12 months old	2	13.3
1 – 2 years old	2	13.3
2 – 5 years old	3	20.0
>5 years old	8	53.3

The laboratory results showed elevated alkaline phosphatase and gamma-glutamyltransferase (GGT) as well as bilirubin levels as a marker for biliary stasis only in 33.3% patients (n=5). Three patients (20%) had increased bilirubin levels with normal alkaline phosphatase and GGT level and the other 3 patients (20%) showed increased alkaline phosphatase with normal level of bilirubin serum (Table II).

Abdominal ultrasonography (USG) was done in all patients (n=15) as an early diagnostic approach. Of these, 12 patients were confirmed the type of CC in this first evaluation. In five patients computed tomography (CT) scan of the abdomen was indicated, demonstrating choledochal dilatation type IV. MR imaging was used in 8 patients, it was indicated after abdominal ultrasonography. According to Todani classification, type I (Fig.1) was recorded in 40% patients (n=6), type IV (Fig. 2) was in 53.2% patients (n=8) and type II in 3 patients (20%) (Table II).

Table II: Clinical Symptoms and imaging results

Clinical Symptoms :	Total	Percentage (%)
Abdominal Pain	12	80.0
Jaundice	15	100.0
Palpable intraabdominal mass	9	60.0
Acholic stool	7	46.6
Anorexia	2	13.3
Fever	6	40.0
Nausea and vomiting	3	20.0
Imaging cyst types		
USG		
Type I	6	40
Type II	5	20
Type IV	3	33.3
MRI		
Type IV	8	53.2
CT Scan		
Type IV	5	33.3

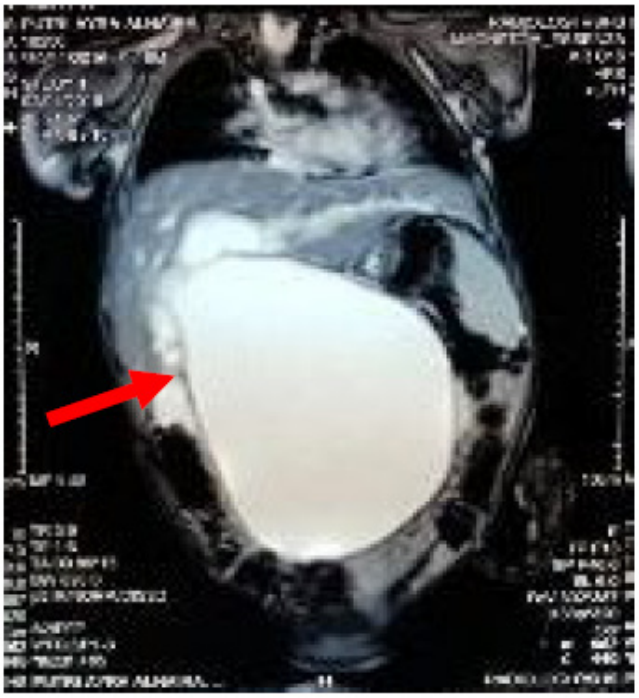

Figure 1: Type I of choledochal cyst in an 1-year-old female. Coronal MRI image shows fusiform dilatation of the common bile duct (arrow), with normal intrahepatic ducts.

Total cyst excision combined with biliodigestive anastomosis had become the treatment of choice in our institution. Only eleven patients (73.3%) were performed cyst excision with Roux-en-Y hepaticojejunostomy, one patient (6.6%) died due to acute cholangitis lead to septic condition and 2 patients refused any surgical procedure. All patients whom underwent surgery, had liver biopsy also. One patient had external drainage prior cyst excision with Roux-en-Y hepaticojejunostomy. Eight patients had the cyst size less than 10 cm in diameter, and 3 patients had bigger size of cyst. Two patients (13.3%) had postoperative complication, and need required reoperation due to leakage of biliodigestive anastomosis. All pathology results suggested no malignant transformation.

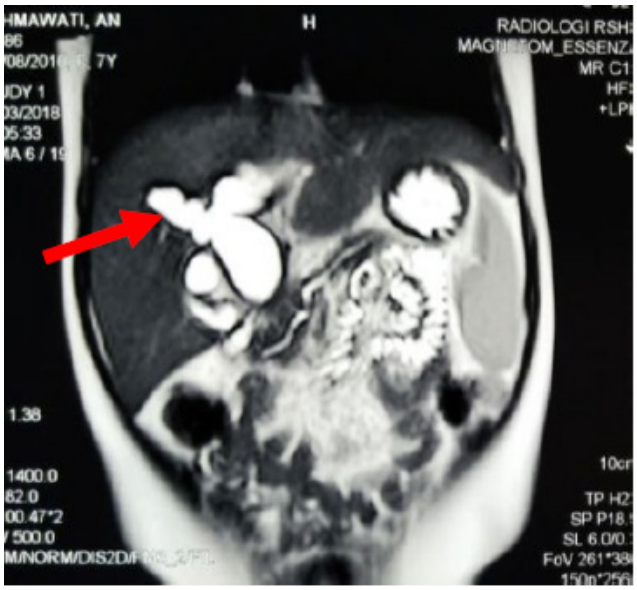

Figure 2: Type IV of choledochal cyst in a 9-years-old female. Coronal MRI image shows cystic intrahepatal ductal dilatation (arrow). Left intrahepatal duct sized 1.7 cm and right intrahepatal duct sized 1 cm.

DISCUSSION

Choledochal cysts (CC) are congenital dilatation of the biliary tree.1,2 The etiology is not well explained, but the early theory proposed by Babbit DP in 1969 suggest that an abnormal junction between the biliopancreatic duct build a common channel that allows reflux of pancreatic secretion into the biliary tract thus causing increased pressure with subsequent ductal dilation (2,3,9,10). Other congenital anomalies such as double common bile duct, sclerosing cholangitis, hepatic fibrosis, pancreatic cyst, annular pancreas and cardiac anomalies have been associated with CC (11). In our study we did not search for any maljunction between the biliopancreatic duct neither the other anomalies.

Most studies noted that CC are commonly found in female than male. Female to male ratio range from 2.5:1 to 4:1, which remain relatively rare in the West compare to Asia countries such as China and Japan although real statistics on prevalence still not exist (5). However in Mexican literature some authors reporting different proportions. Orozco-Sanchez et al, 1997, reported 16:1 ratio and Gallardo-Meza et al, 2010, reported 2:1 ratio (12,13). Although the Asian population is more affected than any other ethnicity but Wiseman K et al, 2005, reported series of CC in North America which have similar statistically significant results in the presentation, management or outcome between Caucasians and Asians. Female to male ratio also found of 4:1. In developed countries up to 15% of all choledochal malformations are detected before birth and this rate is expected to rise. In this study, there is no patient diagnose antenatally. All CC diagnosis were made after birth with only two patients diagnosed at the age less than one year. The age at which the diagnosis was made, with the majority of patients in the first decade of life, similar with other reports (8,13).

The most common clinical symptom was jaundice in all patients, followed by abdominal pain, found in 80% cases (12 patients) and a palpable mass, presented in 60% cases (9 patients). We found fever as sign of acute cholangitis in 40% of cases (6 patients), acholic stool was present in 46.6% of cases (7 patients). The classic triad of choledochal cyst not constantly emerged conformity. Other study reported the classical clinical triad of jaundice, abdominal pain and a palpable mass can be found in about 20% of cases, with predominance in childhood (2,3,14). Some authors noted jaundice to be the main clinical manifestation, but other studies reported abdominal pain is the most prevalent symptoms. Jaundice should be more commonly seen in infants and abdominal pain in older patients (14,15).

Currently, ultrasonography (USG) is the most done imaging modality to diagnose choledochal cyst preoperatively, which has easy and less invasive approach for the diagnosis of biliary tract disease. It was the first complementary method used in all patients, and confirm the diagnosis of CC in this study, although not confirmed most of CC types (according to Todani). This is directly related to the operator's experience. Antenatal diagnosis of choledochal cyst is possible from second trimester of pregnancy, which widely use in developed countries, but not in this study (13,15).

In this study, 33.3% of cases (5 patients) had elevated alkaline phosphatase, gamma-glutamyltransferase (GGT) and bilirubin levels. GGT and bilirubin levels are commonly used as a marker for biliary stasis. It is possible that an elevated GGT indicates epithelial damage. All liver enzymes and total bilirubin showed a tendency to be increased in patients who progressed to CC related symptoms (13,16).

Abdominal CT was performed in 5 patients although the resolution of the images obtained low. Non-invasive test to define the anatomy of biliary tree include computed tomography (about 90% sensibility and specificity) and magnetic resonance imaging (MRI) (70-100% sensitivity and 90-100% specificity), all can be used as diagnostic approach, considering MRI as the first line in preoperative diagnosis to define biliary tract (2,14,16). The MRI has the advantage of simultaneously and effortlessly delineates both the biliary and pancreatic duct and therefore accordingly to the cholangiography morphology classify the images of Todani classification. Some authors concluded that preoperative imaging is unable to predict real intrahepatic involvement in CC accurately (17,18).

According to most literature, type 1 of CC is the commonest type overall. But in this study 53.3% (8

patients) diagnosed as CC type IV using MRI imaging. This results is very different from most literature, which probably because accompanied by a primary ductal stricture around the hepatic hilum and umbilicus , leads to either cystic or fusiform dilatation of intrahepatic duct. Although the cautious observation by direct observation, suitable cholangiograms and examination during surgery with operative cholangioscopy is essential to confirm the incidence of primary stricture (18,19).

A surgical approach is planned consisted of complete excision, cholecystectomy, and biliary tract reconstruction. Biliary tract reconstruction technique is not standardized, some still debating, the systematic review and meta-analysis by Narayanan SK et al, 2013 show that there were no statistically significant difference in biliary leak, cholangitis, anastomotic stenosis, surgical re-intervention and postoperative small bowel obstruction (19). In our cases series, eleven patients were underwent cyst excision with Roux-en-Y hepatojejunostomy anastomosis. Postoperative complication was 13.3% of cases (2 patients) required reoperation due to biliary leakage. Mortality rate was 6.6% of cases, a higher incidence than that Miyano et al and Hung et al, with no mortality (20).

The limitations of this study, the study was done in a retrospective matter, with small number of patients; standardized follow up is imperative for long-term conclusions. While, this represents the experience of a single institution experience, which have all possible bias in the surgical judgement.

CONCLUSION

Choledochal cyst is a rare congenital disease, with jaundice remain a warning sign for malformation of biliary tract. Diagnosis and treatment should be done early. The surgical approach still the treatment of choice, with total cyst excision and hepaticojejunostomy anastomosis.

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