ORIGINAL ARTICLE

OUTCOME OF CHOLEDOCAL CYSTS IN ADULTS — EXPERIENCE AT TERTIARY CARE CENTRE

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ABSTRACT

Objective: To find out the outcome in terms of post operative recovery, and mortality for choledochal cyst in adults in our setup.

Materials and Methods: This was a case series study done in Surgical-A ward at Khyber Teaching Hospital, Peshawar, Pakistan. The study was done from January 2014 to December 2016 (i.e three years period). A total of 22 diagnosed cases of choledochal cyst above 18 years of age were included in the study. Sex, age, symptoms, surgical management and postoperative morbidity and mortality were noted. Data was recorded in proforma and results were drawn accordingly.

Results: A total of 22 diagnosed adult cases of choledochal cyst were included in the study. Age of the study sample ranged from 18 years to 29 years (mean age 22 years ± 2.7 SD). There were 6 (27%) males, and 16 (73%) females. Male to female ratio was 1: 2.6. About 18 (81%) patients presented with recurrent upper abdominal pain, 20 (90%) cases presented with jaundice, and 5 (22%) cases had fever. Ultrasound examination provided the diagnosis in 21 (95%) cases and magnetic resonance cholangiopancreaticography was done in all patients. All the cases were subjected to the surgical repair. Anastamosis through hepaticojejunostomy (bloomguard technique) was done in 21 (95%) cases, and complete cyst excision was done in only 1 (5%) patient. The histopathological report of the specimen was recorded. Only 1 (5%) case had diagnosis of cholangiocarcinoma on histopathological examination. Out of 22 patients, only 1 (5%) patient died in post operative period owing to acute liver failure as there were deranged liver function test and Hepatitis C status was positive in the patient. In the remaining 21 (95%) cases, post operative recovery was uneventful and were discharged accordingly.

Conclusion: Surgical repair through cyst excision and hepaticojejunostomy, if done properly and in expert hands ensures uneventful post operative recovery and low mortality.

Key Words: Choledochal cyst, Ultrasonography, cholangiocarcinoma, anastomosis, post operative period.

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INTRODUCTION

Choledochal Cyst is a rare congenital disorder of the biliary tract, in which there is localized cystic dilatation of the biliary tract[1,2,3]. It is commonly seen in infants and children[4,5]. About 60% of the cases present within the first year of life[6,7]. Although it presents in childhood, it may also present for the first time in adults[6,7]. Choledochal cysts are rare in general population[8]. Worldwide, the incidence of choledochal cyst is 1 case per 100,000 live births[9,10]. Choledochal cysts are more common in Asian countries[11,12]. The reported incidence of choledochal cysts in the Asian countries is 1 in 13000 live births[13]. Choledochal cysts are four times more common in females as compared to males[14].

Choledochal cysts are often asymptomatic[9]. About 80% of the patients present before the age of 10 years, but presentation can be at any age[15]. Only 30% patients present with the classic triad of jaundice, recurrent abdominal pain and upper abdominal mass[16]. Most of the patients present with one or two of the above

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symptoms[9]. Usually, the infants present with jaundice and upper abdominal mass. Adult patients present with jaundice, abdominal pain and fever. Choledochal cysts obstruct the bile ducts and thus cause retention of the bile in the biliary tree. This causes jaundice and an enlarged liver. Choledochal cyst predisposes to development of cholangiocarcinoma in the biliary tract[4]. The risk of development of malignancy in the biliary tract increases with increasing age[16]. The reported incidence of cholangiocarcinoma in patients with choledochal cyst ranges from 9-28%[17].

The commonly performed investigations in patients of choledocal cyst include white cell count, liver function tests , and abdominal ultrasound[14]. Magnetic resonance cholangiopancreaticography (MRCP) is done when ultrasound finding are inconclusive[17]. The white blood cell count is raised, and Liver Function Tests are abnormal. Abdominal ultrasound is the investigation of choice for the diagnosis of choledochal cysts[15]. It is the best initial investigation that is diagnostic in most of the patients[16]. Abdominal CT scan and MRI scan may sometimes be used to see the surrounding structures. Percutaneous transhepatic cholangiography (PTC) or endoscopic retrograde cholangiopancreatography (ERCP) helps in the identification of the associated extrahepatic or intrahepatic strictures[18].

Choledochal cysts are treated by surgical repair. Complete cyst excision and Roux en Y Hepaticojejunostomy are the surgical procedure of choice[19]. However, complete cyst excision does not ensure that carcinoma will not develop in the remaining bile ducts[6]. The risk of developing carcinoma in the biliary tract increases as the age of the patient increases.

Patients who have cholangitis are first treated with broad-spectrum antibiotics first. Once the fever settles, they are then surgically operated[20]. The rationale of the study was to analyse the experience of management and outcome of choledochal cyst in adults in our setup tertiary care hospital.

MATERIAL AND METHODS

This case series was conducted at Surgical A unit of Khyber Teaching Hospital, Peshawar, Pakistan. The study was done from January 2014 to December 2016. All patients above 18 years of age with choledochal cyst were included in the study. All patients were diagnosed on abdominal ultrasound, and MRCP was done in all patients. The surgical procedures (hepaticojejunostomy and cyst excision) was done. Patients were observed for 72 hours post operative period for development of any complications or mortality. Surgery was considered successful if there was no bile leak, hemorrhage, and if there was successful healing of bypass, and no episode of cholangitis. The biopsy report of the cyst specimen was also viewed in all patients. Clinical presentation, surgical procedures performed, any morbidity or mortality, and report of the specimen biopsy were recorded in the proforma, and the results were drawn accordingly.

Results were analysed using SPSS version 18. Mean and standard deviation were calculated for quantitative variables e.g. age. Frequencies and percentages were calculated for qualitative variables like e.g gender.

RESULTS

A total of 22 diagnosed cases of choledocal cyst, above 18 years of age were included in the study. Age of the study sample ranged from 18 years to 29 years (mean age 22 years ± 2.7 SD) The data of the study sample is shown in the Table 1.

Table 1: Profile of adult patients with choledochal cyst

Variables	Number of patients & % ages
Gender	
Male	6 (27%)
Female	16 (73%)
Clinical presentation	
Abdominal pain	18 (81%)
Jaundice	20 (90%)
Fever	5 (22%)
Diagnostic modality	
Ultrasound scan	21(95%)
MRCP	22 (100%)
Surgical procedure performed	
Hepaticojejunostomy	21 (95%)
Cyst excision	1 (5%)
Post operative events	
Uneventful	21 (95%)
Any complications	0 (0%)
Death	1 (5%)
Histopathology findings of the excised specimen	
No malignancy	21 (95%)
Malignancy	1 (5%)

DISCUSSION

Choledochal cyst is a rare abnormality of the biliary tract, that is commonly seen in infants and children. It occurs mainly in the females. Choledochal cyst

in adults is not very common and accounts for only about 20% of cases in literature[21]. The present study showed that choledochal cyst was common in females as compared to males (i.e 73% versus 27% respectively). This is in accordance with literature[5,9,11,9]. In a local study done by Shah and colleagues , it was found that choledochal cyst was common in females (65.22% vs. 34.78% respectively)[6].

The surgical procedures performed on patients of choledochal cyst depended upon the type of cyst, choice and experience of the surgeon[6]. If the surgeon is less experienced, he may end up in inadequate procedure and the patient may develop post operative complications like hemorrhage and bile leak from the anastomosis[6]. In the present study roux en y hepaticojejunostomy was performed in 95% cases and complete cyst excision was done in 5% cases. None of the patients developed procedure related post operative complications. In a study done by Shah and collegues in 2017, it was experienced that in 30% of the cases, choledochal cyst could not be resected because of cyst infection and cholangitis[6]. In those cases, the cyst was bypassed to the duodenum (in 13% cases), a jejunal loop (in 13%) or was drained to exterior via T-tube (in 4.%)[6]. Resection and drainage procedure was done later[6].

Studies show that in adult patients, about 70% of the cases present with complications like cholilithiasis, cholangiocarcinoma and cholangitis[5,9,11,12]. In the present study, 22% patients presented with fever, suggesting cholangitis. In a study done by Shah in 2017, 36% patients presented with cholangitis[6]. Low incidence of complication in our study may be due to early presentation of patients to the hospital or prompt diagnosis of the cases through ultrasonography, and early surgical treatment in our setup.

In the present study, none of the patient had post operative complications like biliary leak or hemorrhage. The surgical procedure was successful in all the cases. In a study done by Nag HH, about 5% patients showed leak of biliary tract anastomosis after the surgery[22]. That patient later died due to uncontrolled abdominal hemorrhage. Gadelhak showed that in his study about 23% cases developed post operative complications like hemorrhage and biliary leak from the anastamosis[23]. Low incidence of post opreative complications in the present study is attributed to better surgical procedure by the experienced surgeons in our setup.

It is suggested that about 15% of patients of choledochal cyst develop biliary tract malignancy. The risk of development of biliary tract malignancy in patients of choledochal cyst is 20 times greater than that of the normal population. It is also suggested that the risk of development of biliary tract malignancy is age related and it increases with increasing age[6,7]. Possible causes for development of malignancy include chronic inflammation process in the wall of biliary duct, stagnation of the bile, and disruption in the mucin-secreting glands of the bile duct[3]. In the present study, only 5% patient had cholangiocarcinoma on the biopsy of cyst specimen. In a study done by Shah and collegues, they also reported only one patient having cholangiocarcinoma[6]. However, Liu CL showed that in his study, about 30% cases of choledochal cyst had cholangiocarcinoma[7]. This is very high as compared to the present study. Gadelhak from Egypt analysed data of 50 adult patients over a period of 15 years, and found that only 2% cases had evidence of malignany in excised cyst[23].

In the present study, post operative mortality rate was 5%, i.e only one patient died after surgery. The patient was already hepatitis C positive and the liver function tests were deranged. In a study done by Liu CL, out of a total of 30 adult patients of chledochal cyst, none of the patient died[7]. So the mortality rate was 0% in his sudy[7]. In another study done by Naq HH, about 5% patients died due to intrabdominal hemorrhage[22]. Gadelhak from Egypt analysed data of 50 adult patients over 15 years period, and found that none of them died in early post operative period[23]. Duan X also had no mortality in his study on 31 patients during 5 years study period[24].

Therefore, timely surgical intervention of the choledochal cysts is the optimal treatment to avoid long term complications like malignant transformation[23] which results in poor prognosis[23].

LIMITATIONS

Moreover,the limitations we faced in our study were that of the patients being lost to follow up. Thus, the long term complications could not be measured in the operated cases.

CONCLUSION

The most common clinical presentation of choledochal cyst in adults is jaundice and upper abdominal pain. The roux–en- Y hepaticojejunostomy via bloomguard technique and cyst excision are safe surgical procedures for choledochal cyst, with low mortality and morbidity, if performed in expert hands.

RECOMMENDATIONS

Early surgical repair is warranted in adult choledochal cyst as there is increased risk of cholangiocarcinoma in such cases.

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AUTHOR'S CONTRIBUTION

Following authors have made substantial contributions to the manuscript as under:

Khan ZM: Study conception and design.
Muslim M: Acquisition of data.
Ahmed A: Analysis and interpretation of data.
Kaleemullah: Drafting of manuscript.
Ahmad N: Review of discussion.
Aurangzeb M: Critical revision and overall supervision.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.