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

Nadeem Ali Shah[1], Ainul Hadi[2], Tahira Mehreen[3], Mohammad Kalim[4], Mussarat Hussain[5], Inayat Shah[6], Juweria Abid[7]

[1, 3-7] Department of Surgery, Lady Reading Hospital, Peshawar - Pakistan.
2 Department of Surgery, Hayatabad Medical Complex, Peshawar - Pakistan.
Address for correspondence:
Dr. Nadeem Ali Shah
Assistant Professor,
Surgical D unit, Department of Surgery, Lady Reading Hospital, Peshawar - Pakistan.
Email: northstarlrh@yahoo.com
Date Received:
May 04, 2016
Date Revised:
January 18, 2017
Date Accepted:
January 28, 2017

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

This article may be cited as: **Shah NA, Hadi A, Mehreen T, Kalim M, Hussain M, Shah I, Abid J. Clinical presentation and management of adult choledochal cyst: A retrospective review. J Postgrad Med Inst 2017; 31(1): 51-5.**

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 million[1]. Almost 60% present before the first birthday[2] but adult presentation is not a rarity[3]. The commonest theories for its causation are weakness in the wall of the bile duct[3-5], obstruction of the distal choledochus[6], combination of obstruction and weakness[7] and reflux of pancreatic enzymes into the CBD secondary to an anomaly of the pancreatico-billiary junction[8-13]. 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 it[14-16].

Choledochal cyst is more common in females[14] and asians[15]. 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 immunity[17-,23]. Adults usually present with the complication of the disease and the classic triad of a pain, jaundice and a palpable mass is found only in 30% of patients[24]. Ultra sound is the best initial investigation that is diagnostic in most of the patients[25]. ERCP has a high rate of success and can map out the pancreato-billiary junction along with the whole of the biliary tree before surgery[26-29]. MRCP has now replaced ERCP, in neonates[30], children[31], adults[32] and even fetuses[33]. Other imagining investigations have a role in special circumstances and are not routinely done.

Complete cyst excision where possible, followed by roux en Y common hepatico-jejunostomy is the standard treatment for the extra hepatic cysts. The treatment for intra hepatic cysts is partial hepatectomy if localized and liver transplantation if widely distributed in the liver[17,18]. The rationale of the study was to find out the mode of presentation of choledochal cyst in adults and to assess the type of surgical procedures done and their short and long term success in a tertiary care hospital.

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 cholidocho-jejunostomy. 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 cholangiocarcinama. 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%) of patients. Complete cyst excision followed by Roux-en-Y common hepatico-jejunostomy was performed in 7 (30.43%) and common hepatico-dudenostomy 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-jejunostomy (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-deudinostomy 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-appropriat 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 cholidocal 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 interntional litrature reports that 80% of such patients will have re-

Table 1: Modes of presentation of choledochal cysts (n=23)

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

* Complications of the disease.

** With stones in the choledochus

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

S.No	Type of Cyst	No of pts.	%age
1	Todani Type I	15	65.21%
2	Todani Type II	2	8.69%
3	Todani Type III	1	4.34%
4	Todani Type IV	3	13.04%
5	Todani Type V	2	8.69%

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

S.No	Surgical Procedure	No of pts	%age
1	Cyst Excision with Roux-en-Y Common hepatico-jejunostomy	7	30.43%
2	Cyst Excision with Common hepaticodudenostomy	6	26.08%
3	Cyst Excision with Loop Common hepaticojejunostomy and Enteroenterostomy	3	13.04%
4	Cystodudenostomy (cyst not excised)	3	13.04%
5	Cystojejunostomy(cyst not excised)	3	13.04%
6	External Biliary Drainage (cyst not excised)	1	4.34%

current symptoms[18,19] and 69% will subsequently need revisional surgery[5,20-21]. In our study 17.39% of patients underwent revisional surgery within one year of their follow up.

One (4.34%) patient was operated in emergency department with biliary peritonitis due to suspected traumatic rupture of the GB. At laparotomy he was found to have ruptured Todani type I choledochal cyst. This is an uncommon finding in adult as compared to infants. One (4.55%) patient was referred from medical ward where he was admitted for a bout of hematemesis and was later diagnosed to have a Todani type I choledochal cyst along with liver cirrhosis and portal hypertension. This patient was not operated and was treated conservatively on numerous occasions in the one year follow-up period.

One patient had advance cholangio carcinoma, with metastasis in the liver and obstructive jaundice. Choledochal cyst is more common in Asian and in females[34-36]. 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[2,5]. 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[5,11,37]. We also encountered the same type to be the most common (65.22%), as shown in table 2. This was followed by Type IV cysts (13.04%) .

Complete cyst excision and restoration of bile flow by Roux-en-Y common hepatico-jejunostomy is said to be the gold standard against which all other treatments are to be compared[38]. This is now also carried out laproscopically with complication rate equivalent to that of open surgery[39,40]. However complete cyst excision does not confer immunity against the emergence of carcinoma in the remaining bile ducts. The risk of cancer is age related and appears to be high in elderly[17-23]. Greatest prevalence of malignancy appears to be in Todani type I, IV and V[18, 20]. 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 table 1.

Type II cyst was treated by simple excision as recommended. Now a days laparoscopic excision is also carried out with equivalent results[41]. Type III cyst was treated by cholidocho-duodenostomy with no complications noted in 1 year of follow up, although the best treatment of such cysts is un-roofing of the choledochocele so that both the ducts drain directly in to the duodenum[27,42]. 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-dudenostomy 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[24,29,43-45].

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

1. Olbourne NA. Choledochalcysts. A review of the cystic anomalies of the biliary tree. Ann R Coll Surg Engl 1975; 56:26-32.
2. Liu CL, Fan ST, Lo CM, Lam CM, Poon RT, Wong J. Choledochal cysts in adults. Arch Surg 2002; 137:465-8.
3. Suita S, Shono K, Kinugasa Y, Kubota M, Matsuo S. Influence of age on the presentation and outcome of choledochal cyst. J Pediatr Surg 1999; 34:1765–8.
4. Bismut H, Krissat J. Choledochal cyst malignancies. Ann Oncol 1999; 10: 94-8.
5. Flanigan PD. Biliary cysts. Ann Surg 1975; 182:635-43.
6. Spitz L. Experimental production of cystic dilation of the common bile duct in neonatal lambs. J Pediatr Surg 1977; 12: 39-42.
7. Kato T, Asakura Y, kasai M. An attempt to produce choledochal cyst in pupies. J Pediatr Surg 1974; 9:509-13.
8. Babitt DP. Congenital choledochal cysts: new etiology concept based on anomalous relationships of the common bile duct and pancreatic bulb. Ann Radiol 1969; 12: 231-40.
9. Kato T, Hebguchi T, matsuda K, Yoshino H. Action of pancreatic juice on the bile duct: pathogenesis of congenital choledochal cyst. J Pediatr Surg 1981; 16:146-51.
10. Miyano T, Suruga K, Sadu K. Abnormal choledoch-pancreatico ductal junction related to the etiology of infantile obstructive jaundice disease. J Pediatr Surg 1979; 14:16-26.
11. Okada A, Nakamura T, Higaki J, Okumura K, Kamata S, Oguchi Y. Congenital dilatation of the bile duct in 100 instances and its relationship with anomalous junction. Surg Gynecol Obstet 1990; 171:291-8.
12. Todani T, Tabuchi K, Watanabe Y, Kobayashi T. Carcinoma arising in the wall of congenital bile duct cysts. Cancer 1979; 44:1134-41
13. Yamashiro Y, Miyano T, Suruga k, Shimomura H, Suda K, Matsumoto M et al. Experimental study of the pathogenisis of choledochal cyst and pancreatitis, with special refrences to the role of bile acids and pancreatic enzymes in the anomalous choledoch-pancreatico ductal junction. J Pediatr Gastroenterol Nutr 1984; 3:721-7.
14. Lenriot JP, Gigot JF, Segol P, Fagniez PL, Fingerhut A, Adloff M. Bile duct cysts in adults: a multi-institutional retrospective study. French Associations for Surgical Research. Ann Surg 1998; 228:159–66.
15. Powell CS, Sawyers JL, Reynolds VH. Management of adult choledochal cysts. Ann Surg 1981; 193:666–76.
16. Jan YY, Chen HM, Chen MF. Malignancy in choledochal cysts. Hepatogastroenterology 2000; 47:337–40.
17. Yamaguchi M. Congenital choledochal cyst. Analysis of 1433 patients in the Japanese literature.Am J Surg 1980;140:653–7.
18. Chaudhary A, Dhar P, Sachdev A. Reoperative surgery for choledochal cysts. Br J Surg 1997; 84:781–4.
19. Swisher SG, Cates JA, Hunt KK, Robert ME, Bennion RS, Thompson JE, Roslyn JJ et al. Pancreatitis associated with adult choledochal cysts. Pancreas1994; 9:633–7.

20. Nagorney DM, McIlrath DC, Adson MA. Choledochalcysts in adults: clinical management. Surgery1984; 96:656–63.

21. Yamaguchi M. Congenital choledochal cyst. Analysis of 1433 patients in the Japanese literature.Am J Surg 1980; 140:653–7.

22. Tadani T, Watanabe Y, Toki A, Urushihara N. Carcinoma related to choledochal cyst with internal drainage operations. Surg Gynecol Obstet 1987; 164:61-4.

23. Watanabe Y, Toki A, Tadani T. Bile duct cancer developed after cyst excision for choledochal cyst. J Hepatobiliary Pancreat Surg 1999; 6:207-

24. O'Neill JA Jr. Choledochal Cyst. Curr Probl Surg 1992; 29:361-410.

25. Yamaguchi M, Sakurai M, Takeuchi S, Awazu S. Observation of cystic dilatation of the common bile duct by ultrasonography. J Pediatr Surg 1980; 15:207-10.

26. Kasai M, Asakura Y, Taira Y. Surgical treatment of choledochal cyst. Ann Surg 1970; 172:844-51.

27. Jona JZ, Babbitt DP, Starshak RJ, LaPorta AJ, Glicklich M, Cohen RD. Anatomic observation and etiologic and surgical consideration in choledochal cyst. J Pediatr Surg 1979; 14:315-20.

28. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: Classification, operative procedures, and review of thirty seven cases including cancer arising from choledochal cyst. Am J Surg 1977; 134:263-9.

29. Todani T, watanabe Y, Fujii T, Toki A, Uemura S, Koike Y. Congenital cholidochal cyst with intrahepatic involvement. Arch Surg 1984; 119:1038-43.

30. Lin JTH YH, Chen YH, Ni YH, Lai HS, Peng SS. Magnetic resonance cholangiopancreatography diagnosed pancreatitis associated choledochal cyst: report of one case. Acta Paediatr Taiwan 2001; 42:363-6.

31. Yamataka A, Kuwatsuru R, Shima H, Kobayashi H, Lane G, Segawa O et al. Initial experience with non-breath-hold magnetic resonance cholangiopancreatography: a new non invasive technique for the diagnosis of choledochal cyst in children. J PediatrSurg 1997; 32:1560-2.

32. Zhong L, Yao QY, Li L, Xu JR. Imaging diagnosis of pancreato-biliary diseases: a control study. World J Gastroenterol 2003; 9:2824-7.

33. Wong AM, Cheung YC, Liu YH, Ng KK, Chan SC, Ng SH. Prenatal diagnosis of choledochal cyst using magnetic resonance imaging: a case report. World J Gastroenterol 2005; 11:5082-3.

34. Saxena R, Pradeep R, Chander J, Kumar P, Wig JD, Yadav RV et al. Benign disease of the common bile duct. Br J Surg 1988; 75:803-6.

35. O'Neill JA Jr, Templeton JM Jr, Schnaufer L, Bishop HC, Ziegler MM, Ross AJ 3rd. Recent experience with choledochal cysts. Ann Surg 1987; 205:533-40.

36. Nagata E, Sakai K, Kinoshita H, Hirohashi K. Choledochal cyst: complications of anomalous connection between the choledochus and pancreatic duct and carcinoma of the biliary tract. World J Surg 1986; 10:102-10.

37. Komi N, Tamura T, Miyoshi Y, Kunitomo K, Udaka H, Takehara H. Nationwide survey of cho- ledochal cysts. Analysis of coexistent anomalies, complications and surgical treatment in 645 cases. Surg Gastroenterol 1984; 3:69-73.

38. Shimotakahara A, Yamataka A, Yanai T, Kobayashi H, Okazaki T, Lane GJ et al. Roux-en-Y hepaticojejunostomy or hepaticoduodenostomy for billiary reconstruction during the surgical treatment of choledochal cyst: which is better?. Pediatr Surg Int 2005; 21:5-7.

39. Li L, Feng W, Jing-Bo F, Qi-Zhi Y, Gang L, Liu-Ming H et al. Laproscopic-assisted total cyst excision of choledochal cyst and Roux-en-Y hepaticoenterostomy. J Pediatr Surg 2004; 39:1663-6.

40. Martinez-Ferro M, Esteves E, Laje P. Laparoscopic treatment of billiary atresia and choledochal cyst. Semin Pediatr Surg 2005; 14:206-15.

41. Liu dc, Rodriguez JA, Meric F, Geiger JL. Laparoscopic excision of a rare type II choledochal cyst: a case report and reviewe of the literature. J Pediatr Surg 2000; 35:1117-9.

42. Josef VT. Surgical techniques and long-term results in the treatment of choledochal cyst. J Pediatr Surg 1990; 25:782-7.

43. Ohi R, Koike N, Matsumoto Y, Ohkohchi N, Kasai M. Changes of intrahepatic bile duct dilatation after surgery for congenital dilatation of bile duct. J Pediatr Surg 1985; 20:138-42.

44. Ohi R, Yaoita S, Kamiyama T, Ibrahim M, Hayashi Y, Chiba T. Surgical treatment of congenital dilation of the bile duct with special reference to late complications after total excisional operation. J Pediatr Surg 1990; 25:613-7.

45. Todani T, Narusue M, Watnabe Y, Tabuchi K, Okajima K. Management of congenetalcholidochal cyst with intrahepatic involvement. Ann Surg 1978; 187:272-80.

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.