DIAGNOSTIC VALUE OF CT SCAN IN DIFFERENT TYPES OF CHOLEDOCHAL CYSTS IN PAEDIATRIC PATIENTS

Zaheer Mustafa[1], Salahuddin Arbi[2], Muhammad Fiaz[3], Mehfooz ur Rehman[4]

ABSTRACT

Background: Choledochal cysts (CDC) are congenital dilatation of hepatic biliary tract, and various investigations are used for its diagnosis. **Objective**: To evaluate the type of choledochocal cyst by CT study of abdomen for prompt decision of surgical intervention. **Methodology:** This descriptive study was based on retrospective analysis of the data of children upto 16 years of age diagnosed as having choledochal cyst on CT scan. This study was conducted from 1st January 2006 to 31st March 2008, in children Hospital & Institute of child health, Lahore. **Results**: Total 30 pediatric patients of choledochal cysts were diagnosed on CT study of abdomen. Out of these patients, 21 patients (70%) were females and 9 (30%) were males. 15 patients (50%) out of 30 had type I CDC cyst and 14 patients (47 %) had type IV cyst where as only one patient (3 %) had type II cyst. None of patient had type III or type V cyst. **Conclusion**: CT scan is an important diagnostic toll to delineate different types of choledochal cysts.

Key Words: Computed Tomography, biliary tree, choledochal cysts.

INTRODUCTION

Choledochal cysts (CDC) are congenital dilatations of intra- and/ or extra hepatic biliary tree.[1,2] Traditionally there are five varieties of CDCs that are classified according to their location in the biliary tree by Todani.[1,3,4]

The disease has higher incidence of diagnosis during first decade of life as well as has predominance in females. Early diagnosis of CDC is important to over come its complications such as cholangitis, acute pancreatitis and cholangio-carcinoma. CDC also has association with biliary atersia and hepatic fibrosis. Early diagnosis and early surgical intervention greatly reduces or perhaps eliminates the risk of cholangio-carcinoma.[5,6] The most common presentation of disease is classically described triad of right upper quadrant pain, jaundice and palpable abdominal mass.[7,8] Young children usually presents with abdominal distension and jaundice.[3] The objective of present study was to evaluate the types of choledochocal cysts by using CT scan.

PATIENTS AND METHODS

This descriptive study was conducted in Children Hospital and Institute of Child Health Lahore from 1st January 2006 to 31st March 2008. All children upto the age of 16 years of both sexes were included in this study. Patients were referred for CT scan of abdomen from OPD as well as in patient departments with various complaints such as abdominal distension, jaundice, palpable mass or unexplained abdominal pain. We analyzed patient demographics, presentation, type of cyst and other associated anomalies. A series of 30 patients with above criteria was found and included in the study. The data was entered and analyzed in SPSS version 10.

Image Acquisition

Axial images of abdomen were acquired on 4 slice CT scanner Volume Zone made of Siemens company. 5 mm slices were obtained without and with I/V contrast. Coronal images were also reproduced in selected cases.

RESULTS

A total of 30 patients of various ages were discovered having CDC from 5 months to 16 years of age. Among 30 patients, 21 (70 %) were females and 9 (30%) were males. Patients were sub divided into two groups-group A less than two years and group B, 2 to 16 years. Various types of CDC and distribution of male and female patients is given in table I.

Table I: Types of choledochal cyst diagnosed on CT image

Type of CDC	Male	Female	Total
Type I	4	11	15
Type II	--	1	1
Type III	--	--	--
Type IV	5	9	14

1.Department of Radiology, Sheikh Zayed Meddical College /Hospital, Rahim Yar Khan.
2.Department of Ophthalmology, Sheikh Zayed Medical College /Hospital, Rahim Yar Khan.
3.Department of Radiology, Surriya Azeem Hospital, Lahore.
4. Department of Radiology, Sharif Medical College, Lahore.

Correspondence: Dr. Zaheer Mustafa. Assistant Professor, Department of Radiology, Sheikh Zayed Medical College / Hospital, Rahim Yar Khan.
Phone. 0300-9670650, **Email**: zaheermustafa1@gmail.com

Results showed that type I CDC was the commonest variety (50%) while type IV was second commonest variety (47%). Among patients having type 1V variety, type 1V A was more common. Only one case (3%) of type II variety was seen. None of the case was seen having type III or type V cysts. Two cases of type I CDC, both females also have multiple congenital anomalies including that of kidneys.

Common presentation was abdominal distension and jaundice in younger children below the age of 2 years. While older children commonly presented with abdominal pain.

DISCUSSION

Choledochal cysts are relatively rare congenital disorders of biliary tree, usually discovered in infancy or childhood. They are congenital cystic dilatations of any portion of the bile ducts, but most often occur in the main portion of the common bile duct.[1] There are many theories about the etiology of the bile duct cysts. The common channel theory proposed by Babbit et al is the most widely accepted theory.[5] According to their theory, there is an abnormal pancreatobiliary duct union that allows reflux of pancreatic secretions into the biliary system during a critical stage of its development. The common bile duct and pancreatic duct normally unite within the sphincter of Oddi to form a common channel which opens into the middle portion of the duodenum. An anomalous pancreatobiliary junction has two features that are relevant to formation of choledochal cysts. One is that the union of pancreatic duct and common bile duct is located far from the duodenum, creating a long common channel. The other is the angle of this junction. Consequently chemical and enzymatic destruction of the ductal wall, leads to cystic dilatation. However, the theory of malformation of the pancreatobiliary ductal junction does not explain the occurrence of choledochal cysts in the presence of a normal pancreatobiliary ductal union. An alternative hypothesis is that choledochal cysts represent a spectrum of embryonic malformation of the pancreatobiliary system, one of which may be an anomalous junction.

A widely accepted classification of choledochal cysts based on anatomical location and cholangiographic morphology of the choledochal cysts was developed by Todani.[3] Todani and colleagues described 5 types of choledochal cysts: Type I is the most common type (80-90%). It is subdivided into type IA (cystic dilatation of the common bile duct), type IB (segmental dilatation of the common bile duct) and type IC (fusiform dilatation extending to common hepatic duct). Type II is rare (2%) and is a true diverticulum anywhere in the extrahepatic duct. Type III cyst, also referred as choledochocele, is also rare (1,4-5%) and confined to the common bile duct within the duodenal wall. Type IV cyst is more common (19%) and may be further subdivided into type IVA, which involves both the intra- and extrahepatic bile ducts, and the less common type IVB, where only extrahepatic cysts are observed. Type V, or Caroli's disease, includes single or multiple intrahepatic bile cysts. In our study, it is found that type I CDC is the most common variety where as type IV is next most common variety. In a research the changing presentation of choledochal disease an incidental diagnosis" showed that 64 % of patients have type I CDC.[6] While in our study incidence of type I CDC was 50 %.[6] Another study showed that incidence of type I cyst is 90 % mainly seen in girls/women.[7]

Histopathologically, choledochal cysts have a fibrotic wall varying in thickness from a few millimeters up to 1cm. The cyst wall consists of dense collagenous connective tissue with occasional elastic fibers and smooth muscle bundles. A complete epithelial lining is usually absent. However, scattered columnal or cuboidal epithelium can be found. An inflammatory reaction is often present.[8]

Usually, choledochal cysts appear clinically as abdominal pain, jaundice and a palpable abdominal mass.[9,10] Intermittent jaundice and an abdominal mass are the most common findings in infants. . Complications of choledochal cysts include cholecystitis, recurrent cholangitis, biliary stricture, choledocholithiasis, recurrent acute pancreatitis and malignant transformation into cholangio-carcinoma.[1,9,11] The frequency of biliary malignancy increases with age.

Choledochal cysts can lead to significant morbidity and mortality if not diagnosed early and treated promptly.[12] Hence early detection followed by surgical intervention is an important factor to reduce the risk of complications especially Cholangio-carcinoma.[1,5,13] The life time risk of CDC associated Cholangiocarcinoma is as high as 26 % and rate of

occurrence increases with age. Once found, the current treatment of choice is surgical excision.[14] From a surgeon's standpoint, preoperative imaging evaluation is essential for planning the operative approach to choledochal cysts. Anatomic details on the pancreatico-bile duct union and the exact morphology of the duct are important factors for the surgeon to consider when determining the appropriate surgical procedure. Surgeons are also concerned about the exact location of the pancreatic duct, the site of entry into the duodenum, and the length of the common channel. Since malignancy may develop in the remaining stump after resection, surgeons prefer to resect the cyst as low as possible without causing injury to the pancreatic duct and the common channel.[4,15]

CONCLUSION

Choledochal cysts are uncommon biliary lesions. Early detection of CDC and followed by surgical intervention is an important factor to reduce the risk of complications particularly clolangio-carcinoma. CT scan is an important adjunct in evaluating choledochal cysts to delineate the anatomy of lesion and the surrounding structures which greatly help to decide surgical procedure.

REFERENCES

1. J. S. de Vries, S. de Vries, D. C. Aronson, et al., "Choledochal cysts age of presentation, symptoms and late complications related to Todani s classification". Journal of Pediatric Surgery, 2002. 37(11).1568-73.
2. M. Yamaguchi, "Congenetal choledochal cyst analysis of 1433 patients in the Japanes literature," The American Journal of Surgery,1980, 140 (5), pp. 653-57.
3. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts. Classification, operative procedures, and review thirty-seven cases including cancer arising from choledochal cyst. Am J Surg. 1977;134:263- 69.
4. J. Singhavejsakul and N. Ukarapol, "Choledochal cysts in children epidemiology and outcomes," World Journal of Surgery, 2008, 32(7) pp. 1385-88.
5. Babbit D., Starshak R, Clemett A. Choledochal cyst: a concept of etiology AJR. 1973;119:57-62.
6. J. Singham, D. Schaeffer, E. Yoshida, and C. Scudamore, "Choledochal cysts Analysis of disease pattern and optimal treatment in adult and paediatric patients" HPB,2007. 9 (5), pp. 383-87.
7. B. H. Edil, L. Cameron, S. Reddy, et al., "Choledochal cyst disease in children and adults a 30- year single institution experience," Journal of American College of Surgeons,2008, 206 (5) pp 1000-1005.
8. A. Chaudhary, P. Dhar, A. Sachdev, et al., "Choledochal cystsdifferences in children and adults," British Journal of Surgery, 1996, 83 (2), pp. 186-88.
9. P. A. Lipsett, H. A. Pitt, M. Colombani, J. K. Boitnott, and J. L. Cameron, "Choledochal cyst disease: a changing pattern of presentation," Annals of Surgery, 1994, 220 (5) pp. 644-52.
10. Visser BC, Suh I, Way LW, Kang SM. Congenital choledochal cysts in adults. Arch Surg. 2004;139:855-62.
11. Crittenden S, Mckinley M. Choledochal cyst-clinical features and classification. Am J Gastroenterol. 1985;80:643-47.
12. Ok Hwa Kim, Hong Jun Chung, Bying Gil Choi. Imaging of the choledochal cyst. RadioGraphics. 1995;15:69-88.
13. Iwai N, Yanagihara J, Tokiwa K, Shimotake T, Nakamura K. Congenital Choledochal Dilatation with emphasis on pathophysiology of the biliary tree. Ann Surg. 1992;215:27-30.
14. Oldham KT., Hart MJ, White TT. Choledochal Cysts presenting in late childhood and adulthood Am Surg.1981;141:568-571.
15. Yoshida H, Hai Y, Minami M, Kokudo T, Ohtomo K, Kuroda A. Biliary malignancies occurring in choledochal cysts. Radiology.1989;173:389-92.