University Journal of Surgery and Surgical Specialities

ISSN 2455-2860 2019, Vol. 5(5)

A RARE CASE OF CHOLEDOCHAL CYST PRESENTING IN ADULT.

VIJAYANAND A ANNAMALAI
Department of General Surgery, THANJAVUR MEDICAL COLLEGE

Abstract : Choledochal cyst is essentially a disease of paediatric age group (80 percent of cases), though rarely presents in adults. We are presenting this case of choledochal cyst in an adult female of 40 years of age, for its rarity of occurrence and complications associated with excision of the cyst. This is the only case of choledochal cyst operated in our college in the acaemic year of 2012 - 2013. The incidence of choledochal cyst is 1 in1,00,000 to 1,90,000 in western countries .Choledochal cyst in adults should be dealt with caution because most of adult cases are associated with complications and association of cyst related malingnancy in adults is high.

Keyword : Benign hepatobiliary diseases, Choledochal cyst, Todani classification, Treatment options.

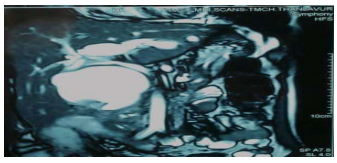
INTRODUCTION:

Choledochal cysts (CDCs) are uncommon, yet well documented congenital dilations of the intra- and/or extra-hepatic biliary tree that can carry significant morbidity if not recognized and treated early. Early detection and treatment of CDCs is an important factor in the overall lifetime occurrence of cholangiocarcinoma1 . The lifetime risk of CDC associated cholangiocarcinoma is as high as 26% in some studies, and importantly, the rate of occurrence increases with age.

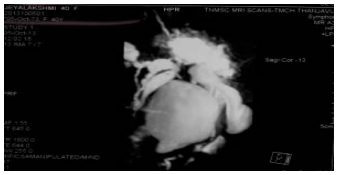
CASE DETAILS:

A 40 year old female patient was admitted with complaints of right hypochondrial pain, nausea, vomiting and intermittent fever for a period of 15 days, with no history jaundice. No history of similar episodes in the past or childhood. On examination patient had mild grade fever, no clinically detectable icterus, right hypochondrial tenderness and no palpable mass or organomegaly. Initial presentation was that of acute cholecystitis, so patient was started on i.v antibiotics and analgesics, while meantime basic investigations were done which showed subclinical hyperbilirubinemia. USG abdomen showed a 15*8 cm cystic lesion occupying the entire supraduodenal part of the common bile duct and common hepatic duct, reported as choledochal cyst and gall bladder was found to be normal, with no detectable calculi.

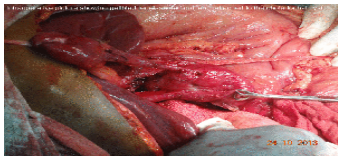
Patient was further evaluated with MRCP (Magnetic Resonance Cholangio Pancreaticogram) found to have fusiform shaped Todani's type I choledochal cyst. Oesophagogastroduodnoscopy showed normal upper digestive system and ampulla of Vater. ERCP could not be done due to non-availability. Patient planned for total excision of the cyst along with cholecystectomy and bilioentric continuity to be facilitated by Roux-en-Y hepatico jejunostomy and patient was evaluated for anaesthetic fitness. Patient underwent a successful complete excision of the choledochal cyst through a bilateral subcostal incision (roof top incision) with two separate anastomosis of the right and left hepatic ducts with Roux-en-Y loop as right and left Roux-e-Y hepaticojejunostomy .



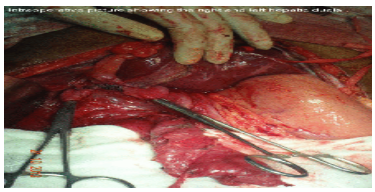
MRI SHOWING THE Cyst.



MRCP -3D recontsruction of cyst.



INTRAOPERATIVE PICTURE- SHOWING THE CYST.



INTRAOPERATIVE PICTURE- showing the right and left hepatic duc

Initial postoperative days were uneventful, but on 5th post-operative day patient developed increased bilious drainage from the right abdominal drain tube. Anastamotic leak was suspected and patient was kept nil per oral and nutritional support was given by total parentral nutrition. Drain through the fistula reduced in a period of 5 days and secondary suturing was done and patient discharged well with the advice to have regular followup. This patient was operated successfully under the financial assistance provided by the Tamilnadu Chiefminister's comprehensive health care scheme.

DISCUSSION:

Choledochal cyst is a surgical problem of paediatric age group, only 20% presents in the adult population2 and are complicated by increased rate of associated hepatobiliary pathology. First case was reported by **Douglas in 18523**. Then **Alonso-Lej** and **Todani et al** classified choledochal cyst into three types and five types respectively. A female preponderance4 is clearly noted in the ratio of 4: 1. The most accepted theory is the **anamolous arrangement of the pancreaticobiliary ductal junction5**(Babbit ,1969), which causes reflux of the pancreatic juices into the biliary tree . (CBD-12cm of water6; Pancreatic duct-22.2cm of water7), leading to inflammation, ectasia, and ultimately, to dilation. and abnormal angle of fusion2 between the pancreatic and bile duct, with increased length of the common channel . Most cases are diagnosed incidentally, when we evaluate the patient for upper abdominal pain. The triad of pain, fever and jaundice were noted in most of the cases. If an abdominal mass is present, should raise the suspicion of cyst associated malignancy. The ideal diagnostic tool to diagnose this short of congenital anamoly is MRCP and it is the gold standard8. Other investigations may contributing factor to the diagnosis.

In paediatric cases ERCP- has added advantage because therapeutic papillotomy9 to relieve severe cholangitis can be done along with imaging of the biliary tree. Associated hepatobiliary pathology is found in more than 80% of adults with bile duct cysts, which include Cystolithiasis (2% to 72%) , hepatolithiasis , acute and chronic cholecystitis and choledocholithiasis , intrahepatic abscess, portal hypertension and pancreatitis(2% to 70%). Pseudopancreatitis**2** exaggerates the occurrence of pancreatitis which appears as peripancreatic and intrapancreatic low density in CT scan of abdomen, resembling fluid without other signs of pancreatitis .The pancreas may even appear swollen. Associated Hepatobiliary malingnancy2: (2.5% to 28%) includes cholangiocarcinoma, adenoacanthoma, anaplastic carcinoma, bile duct sarcoma, hepatocellular carcinoma, pancreatic carcinoma, and gall bladder carcinoma. Of these cholangiocarcinoma is most common and has 20 times greater chance of occurrence than general population. 57% of these tumours are intracystic in position. Etiology is said to be formation of carcinogens due to bile stagnation(unconjugated deoxycholate and lithocholate, or secondary bile salt) and denovo from abnormal cyst epithelium. Surgical excision of the cyst11 and bilioentric anastomosis is the definitive management of the choledochal cyst. Disadvantages -excision is cumbersome and long procedure which needs lot of expertise, Postoperative biliary leaks, fistulas, biliary stricture and obstruction. First laparoscopic repair of the bile duct cyst was reported in 1995 by Farello et al2, in a paediatric patient. In 2008 **Palanivel and collagues2** from India reported the largest series of laproscopic treatment of bile duct cysts, a series of 35 patients, of which 16 were adults. One clear advantage is the magnification provided by the laparosopy which defines the neovascularisation around the cyst, and aids in separation of posterior cyst wall from portal vein and in dissection and anastomosis in the hilar area.

Conclusion:

Choledochal cyst is rare entity in adults, most often associated with complications. Best mode of investigation is MRCP. Surgical excision is the best option of treatment available till now. Laparoscopic treatment is a developing field which needs till further evaluation and randomized control trails.

References:

1. Rajeev Dhupar, Brian Gulack, David A. Geller, J. Wallis Marsh, and T. Clark Gamblin, The Changing Presentation of Choledochal Cyst Disease: An Incidental Diagnosis, HPB Surgery (2009)
2. William R. Jarnagin, MD, FACS, Leslie H. Blumgart, BDS, MD, DSc(Hon), Blumgart's Surgery of the Liver, Biliary tract, and Pancreas, 5th ed ,Vol 1.
3. Michael Zinner Jr , Stanley Ashley, Maingot's Abdominal Operations, 12th ed.
4. Charles J. Yeo, MD, Jeffrey B. Matthews, MD, David W McFadden, MD, John H. Pemberton, MD and Jeffrey H. Peters MD, Shackelford's Surgery of the Alimentary Tract, 7th ed ,Vol 2.
5. Yamauchi S, Koga A, Matsumoto S, Tanaka M, Nakayama F-The American Journal of Gastroenterology [1987, 82(1):20-24]
6. Weiss HD, Kramann B, Wuttke V, Anacker H. Basic pressure in the pancreatic duct and in the bile duct , Med Klin. 1977 Mar 25;72(12):519-23.
7. Gerbail T. Krishnamurthy, Shakuntala Krishnamurthy -Nuclear Hepatology: A Textbook of Hepatobiliary Diseases.
8. F. Brunicardi, Dana Anderson, Timothy Billiar, David Dunn, John Hunter, Raphael E. Pollock**,** Schwartz's Manual of Surgery -8th edition.
9. De Angelis P, Foschia F, Romeo E, Caldaro T, Rea F, di Abriola GF, Caccamo R, Santi MR, Torroni F, Monti L, Dall'Oglio L. Role of endoscopic retrograde cholangiopancreatography in diagnosis and management of congenital choledochal cysts- J Pediatr Surg. 2012 May;47(5):885-8.
10. Park DH, Kim MH, Lee SK, Lee SS, Choi JS, Lee YS, Seo DW, Won HJ, Kim MY, Can MRCP replace the diagnostic role of ERCP for patients with choledochal cysts? Gastrointest Endosc. 2005 Sep;62(3):360-6.
11 . James S. Dooley , Anna Lok , Andrew K. Burroughs , Jenny Heathcote , Sherlock's Diseases of the Liver and Biliary system, 12th edition. April 2011 .

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