



**ORIGINAL RESEARCH PAPER**

**Hepatobiliary Surgery**

**A RARE PRESENTATION OF CHOLEDOCHAL CYST IN ADULT.**

**KEY WORDS:**

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**ABSTRACT**

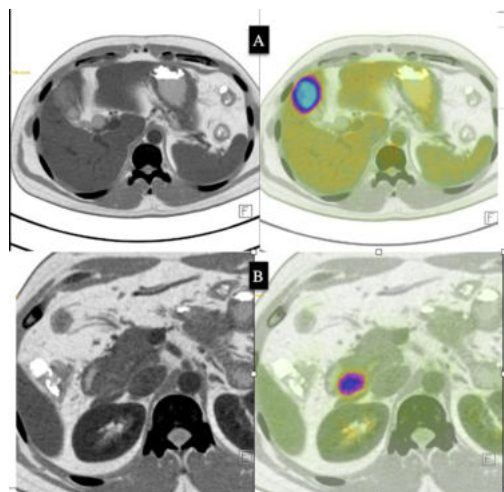
Choledochal cyst (CC) is a relative rare condition in adult population. Only 20% of CC presents in adult population, of these 80% presents with additional hepatobiliary pathology (1,2). With widespread usage of cross sectional imaging, more cysts are likely being identified incidentally in asymptomatic individuals. The development of cancer arising from cysts increases from <0.7% in first decade of life to >11.4% after 2nd decade of life. Anatomic abnormality pancreaticobiliary maljunction (PBM) is a noted etiologic factor for cyst development and even for development of cancer in cysts. We report a rare adult presentation of choledochal cyst with underlying abnormal pancreaticobiliary maljunction with gall bladder carcinoma.

**CASE REPORT**

34 years old male, resident of port Blair with no co morbid illness presented to us with vague right upper quadrant pain for 2 months. Physical examination was normal. Biochemical parameter were within normal limits. Ultrasound abdomen was showing Asymmetrical gall bladder wall thickening in the fundus with dilated common bile duct with non dilated intrahepatic bile ducts.. Patient was further evaluated with MRCP and Computed tomography Triple phase. CT is suggestive of Heterogeneously enhancing soft tissue density lesion involving the fundus and body of gall bladder with suspicious focal loss of fat plane with liver. There is Fusiform cystic dilatation of common bile duct (3.5 cm) and common hepatic duct noted with abrupt narrowing in distal common bile duct. Intra Hepatic Biliary Radicals no dilation. Periportal and posterior pancreaticoduodenal lymph nodes enlarged. Suggestive of malignant pathology of gall bladder with choledochal cyst. MRCP ( Image 1) Suggestive of No inter hepatic radical dilation, Right & left Hepatic ducts 9 mm each, Cystic duct dilated 10 mm, Common bile duct grossly dilated 30 mm, with smooth tapering seen at distal third, Type I choledochal cyst. Gall bladder is partially distended with asymmetrical wall thickening seen at fundus region. Focal lesion of size 2.1\*2.7cm is seen at the posterior aspect of pancreatico duodenal groove, likely Enlarged Lymph node. Pancreatic duct is normal in size, abnormal long common channel noted with complex type of PBM. CA 19 9 was 3.5 U/ml. Whole body FDG PET CT scan (Image 2) was done for staging purpose, which revealed no evidence of metastasis. With the diagnosis of Choledochal cyst with Gall bladder malignancy patient underwent surgical excision. Intraoperatively Bile was aspirated from choledochal cyst and amylase levels were estimated, which were 2496 U/L (Suggestive of PBM). Radical cholecystectomy with complete excision of the choledochal cyst and Roux en y Hepaticojejunostomy with hepatoduodenal ligament lymph nodes, posterior pancreaticoduodenal nodes were dissected (Image 3). Post operative course was uneventful. Final histopathology was suggestive of Moderatly differentiated adenocarcinoma of gall bladder. pT3 pN1, Margins not involved. Lympho vascular invasion and perineural invasion are present. 2 out of 10 Lymph nodes were positive. Choledochal cyst shows no e/o dysplasia or malignancy. Resected liver shows 2 infiltrative nodules. Post recovery patient received adjuvant chemotherapy. Patient had loco regional recurrence after 10 months of surgery. To the date of publication patient is alive receiving 2nd line of chemotherapy.

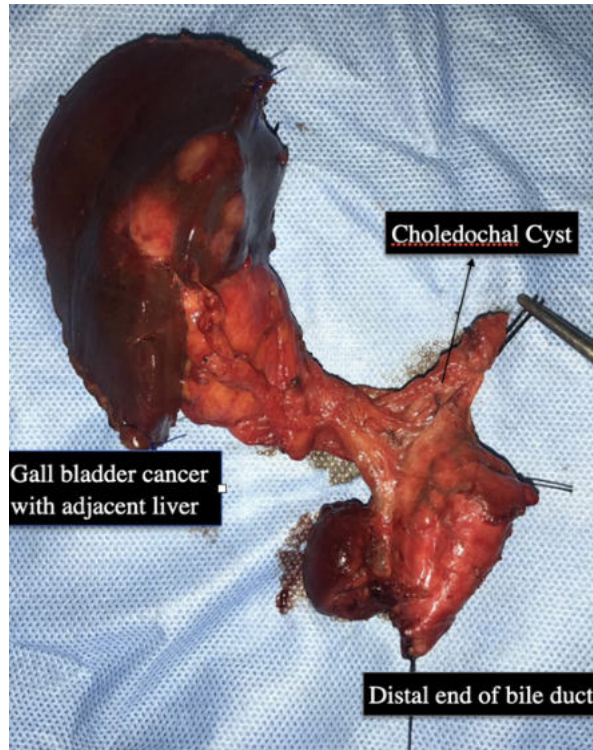


**Image 1 - MRCP - Grossly Dilated (30 mm) Common bile duct, with smooth tapering seen at distal third. Gall bladder is Partially distended with asymmetrical wall thickening seen at fundus region. Pancreatic duct is Normal in size. Complex Pancreaticobiliary maljunction Noted.**



**Image 2 - Whole body FDG PET CT scan. A- Axial section**

image showing gall bladder fundus lesion infiltrating the liver and corresponding FDG avidity. B - Axial section showing enlarged posterior pancreaticoduodenal lymph node and corresponding FDG avidity.



**Image 3 - Resected specimen. Radical cholecystectomy with lymphadenectomy and choledochal cyst excision.**

**DISCUSSION:**

Choledochal Cyst is a relatively rare condition in adult population with females being more commonly affected. Only 20% of CC are diagnosed after 2nd decade (1, 2). 80% of adult CC have additional pathology ranging from cholelithiasis, calculous cholecystitis, malignancy to cirrhosis with portal hypertension. Multiple etiological theories have been put forward, most commonly accepted one is abnormal pancreatic biliary maljunction (3). According to Todani classification, Type I and IV a ( Todani classification) CC are associated with underlying PBM. PBM can be manifested with or without CC (4). CC is a pre malignant condition, and association of PBM increases the risk of developing malignancy. In Japanese nation wide study of adult population, (5) biliary cancer was seen in 22% of adult patients with PBM and CC. These patients were 15-20 years younger and had lower incidence of gall stones. (5,6) Of these biliary cancers most commonly cancer occurred in gall bladder (62%) followed by bile duct cancer (32%). Pancreatic juice regurgitates into the biliary tract through a long common channel, Bile mixed with regurgitated pancreatic juice produces substances that are hazardous to the biliary epithelium, including activated pancreatic enzymes, The mixture of bile and pancreatic juice produces hazardous substances that stagnate in a dilated bile and gallbladder (3). These hazardous substances then irritate the epithelium, resulting in hyperplasia that progresses to carcinoma through multiple molecular changes. In Japanese cohort association of PBM increases the incidence of Gall bladder malignancy(5). Primary presentation of choledochal cyst with malignancy and underlying PBM has an advanced stage of presentation. Even after surgical excision lifelong follow up is recommended.

**CONCLUSION -**

Adult choledochal cyst with underlying PBM patients needs to be evaluated for underlying malignancy. Any lesion in bile

duct or gall bladder should be properly staged before surgical planning. In our case, patient had advanced nature of presentation. Even after excision life follow up is recommended.

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