ORIGINAL RESEARCH PAPER

General Surgery

KEY WORDS: Choledochal cyst, Stricture, Cholangitic abcess, Roux En Y Hepatico jejunostomy, Cholecystectomy

TYPE 1 CHOLEDOCHAL CYST – A CASE REPORT

Dr. K. Senthil kumar — Assistant professor, Institute of general surgery, Madras medical college and Rajiv Gandhi government general hospital, Chennai.

Dr. K. Surendar — Post Graduate , Institute Of General Surgery, Madras Medical College And Rajiv Gandhi Government General Hospital, Chennai.

Prof. P. Thangamani — Professor, Institute of general surgery, Madras medical college and Rajiv Gandhi government general hospital, Chennai.

Prof. R. Kannan — Director, Institute of general surgery, Madras medical college and Rajiv Gandhi government general hospital, Chennai.

ABSTRACT

We report a case of a 20 year old female patient presented with complaints of right upper abdominal pain, vomiting and jaundice for one week. Patient had similar complaints since childhood. On examination, patient is icteric and abdominal examination revealed tenderness in right hypochondrium and epigastrium with localised guarding. Preliminary blood investigations showed elevated WBC counta and Liver function test showed elevated total and direct bilirubin. USG Abdomen showed Choledochal cyst with distal benign stricture. Contrast CT of Abdomen and MRCP abdmoen showed Choledochal cyst with cholangitic abscess and distal benign stricture. Preoperatively patient was hydrated with in fluids and started on broad spectrum antibiotics for one week. Electively Excision of Choledochal cyst with Roux enY Hepatico jejunostomy and cholecystectomy was done. Post operatively patient was stable, jaundice subsided and no further complaints. She was discharged one week after surgery.

Case report:

History: A 20 yrs old female came with chief complaints of of right upper abdominal pain, vomiting and jaundice for one week. Patient had similar complaints since childhood. Abdominal pain was intermittent, colicky, dull aching type , non-radiating , aggravated by food intake, relieved by medications. Patient had low grade **Fever** for one day.

On Examination: Patient was Icteric. Epigastric tenderness and Right hypochondrial tenderness present with localised guarding.

Blood Investigations: Total counts elevated (>10000 /uL).Differential counts – **Neutrophilia**. **Total Bilirubin (5.8 mg/dl) elevated and Direct bilirubin(3.5mg/dl) elevated.**ALP mildly elevated (151 IU/L)

USG ABDOMEN:Dilated Common Bile Duct (max.diameter 5cm) noted upto distal part of CBD upto the pancreatic head with abrupt cut-off . Possibility of **Choledochal cyst with distal benign stricture**

CECT ABDOMEN(IV CONTRAST): Common bile duct is grossly dilated measuring 5.51cms max.diameter and length – 10 cms with Multiple focal dilatations noted in bilateral Common hepatic duct. Multiple hypodense lesions with surrounding areas of contrast enhancement in segment 6,7,8 of liver largest measuring 1.5 x 1.3 cms in segment 7 - evolving abscess.

IMPRESSION : Choledochal cyst with cholangitic abscess (Fig.1)

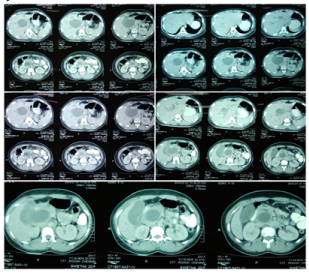

Fig.1.CECT Abdomen showing Dilated Extra hepatic biliary radicals

MRCP findings: CHD and CBD are grossly dilated and there is abrupt changing calibre before the distal end.T2 hyper intense lesion in segment VIII and ill defined areas of hyper intensities in right lobe of liver.

IMPRESSION: Type 1 Choledochal cyst with evolving cholagitic abscess (Fig.2)

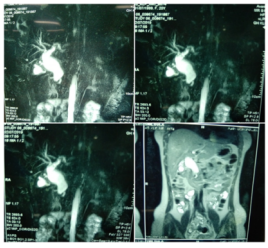

Fig.2.Type 1 Choledochal cyst

Treatment: Patientt was hydrated well with **iv fluids**. **Broad spectrum antibiotics** was given for one week. Total counts decreased and Patient symptomatically improved. Hence planned for **Excision of Choledochal cyst with Roux en Y Hepatico jejunostomy and cholecystectomy.**

Surgery Done : Excision of the cyst with roux en y hepatico jejunostomy and cholecystectomy.

Intra-operative Findings: Fusiform dilataion of extrahepatic biliary tree upto distal end of CBD. Type 1 choledochal cyst. Hepatic duct confluence is normal(Fig.3,4,5)

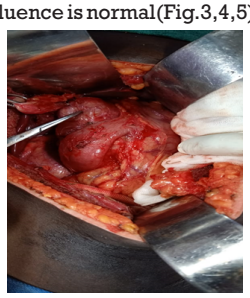

Fig.3.Fusiform dilatations of EHBR

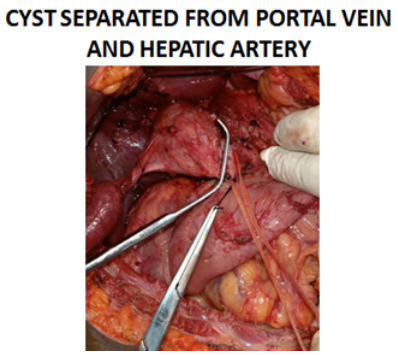

Fig.4.Choledochal Cyst

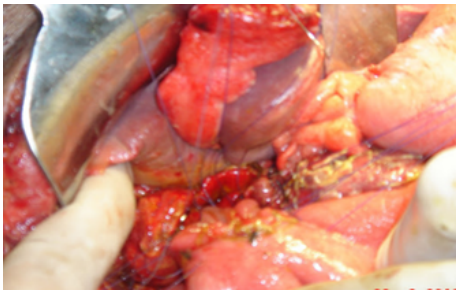

Fig.5.Roux En Y Hepatico jejunostomy

DISCUSSION:

CHOLEDOCHAL CYST: Cysts of the biliary tree are rare - 1/100,000.Common in Asian descent.**Three to eight times more common in women than in men.It is a Premalignant condition** – hence surgical intervention is necessary. It is commonly diagnosed in infancy, but many present in adulthood. Presence of **anomalous pancreaticobiliary junction (APBJ)** is seen in up to **90%**.Pancreatic duct and biliary tree fuse to form a **long common channel** before passage through the duodenal wall **(>15mm).**APBJ is located outside the duodenal wall,hence the normal sphincter does not work. **Pancreatic secretions reflux into the biliary tree causing inflammation, ectasia and dilatation of duct and fibrosis. Pancreatic duct has higher secretory pressures** than the biliary tree, exocrine pancreatic secretions reflux up into the bile duct. Pancreatic enzymes, lysolecithin, secondary or unconjugated bile acids, injure the epithelium of the biliary tract , induce **metaplasia and malignant change**

TODANI'S CLASSIFICATION OF CHOLEDOCHAL CYST:

- **Ia** - Gallbladder arises from the choledochal cyst with e**Type I** – Fusiform dilation of extrahepatic biliary tree
- **Type** xtrahepatic biliary tree appears dilated
- **Type Ib** - Isolated dilation of the most distal aspect of the CBD, with the rest of the extrahepatic biliary tree appearing normal
- **Type Ic** - Smooth, fusiform dilation of the CHD and CBD along with pancreaticobiliary malunion
- **Type II** - Supra duodenal diverticulum of CHD or CBD
- **Type III** - Intraduodenal diverticulum of the distal CBD or choledochocele
- **Type IVa** - dilation that extends from the CBD and CHD into the intrahepatic bile ducts
- **Type IVb** - multiple dilations of the extrahepatic biliary tree, with an uninvolved intrahepatic biliary tree
- **Type V** - Caroli disease, dilatations of intrahepatic ducts only - solitary or diffusely in all segments
- The frequency of bile duct cyst is - **type I cyst (79%)** followed by type IV (13%), then type III (4%), and type II (2.6%)

Clinical presentation: *Recurrent epigastric or right hypochondrial pain, abdominal tenderness, fever, and mild jaundice* Abdominal mass - associated malignancy. Long-standing disease - chronic injury to the liver - cirrhosis. Cholangitis, pancreatitis, hepatic fibrosis, malignancy. Premalignant condition - cholangiocarcinoma The incidence of malignancy - 10% to 30%

Imaging:

USG – initial investigation to identify extent of dilatation. USG in Type 1 cyst -**irregular hypoechoic segmental dilation** of the extrahepatic bile duct . Focal duct wall thickening or **nodularity - suspicion for cancer.** CECT abdomen can diagnose choledochal cyst if suspected. MRCP is used for classification of cyst

TREATMENT:

Type I cysts - complete surgical excision, cholecystectomy, and Roux-en-Y hepaticojejunostomy

Type II cysts - excised entirely and in the presence of an APBJ, **biliary-enteric diversion by Roux-en-Y hepaticojejunostomy**

Type III cysts - transduodenal endoscopic drainage. In duodenal or biliary obstruction, **transduodenal excision of cyst or sphincteroplasty**

Type IVa cysts - intrahepatic extension involving only one lobe can be treated with **partial hepatectomy and reconstruction**

Type IVb cysts affecting only the extrahepatic bile ducts are managed similarly to type I cysts, with **excision and hepaticojejunostomy.**

Type V Caroli disease - **liver transplantation**

CONCLUSION:

Choledochal cyst may present with recurrent attacks of right upper quadrant pain and jaundince since childhood. Early surgical intervention is necessary as it is a premalignant condition. Patient must be advised to have regular follow-up for early detection of malignancy.

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