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Adult Type 4a Choledochal Cyst

Ranjith Babu R
Department of General Surgery, K.A.P. Viswanathan Government Medical College

Abstract : Choledochal cyst ,a rare congenital anomaly is an anomalous dilatation of the biliary tree It is the dilatation of either intra or extrahepatic biliary system or both. Most of the reported cases in the world come from Asia and about two-third of cases are reported from Japan. It occurs in approximately 1 in 1,50,000 live births. According to modified todanis classification it is categorized into six types. Among this type 1 was present in 80-90percent cases and type 4 choledochal cyst was present in 8-10 percent of cases only. We are reporting a case of type 4A choledochal cyst in 30 years female.

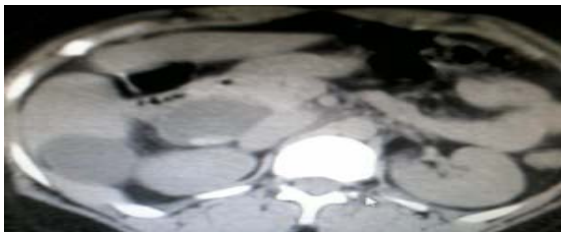
Keyword : Choledochal cyst, hepaticojejunostomy, Todani

INTRODUCTION:

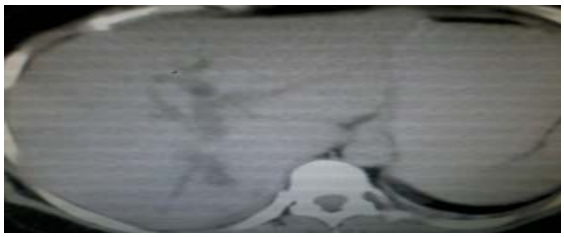
Choledochal cysts are rare congenital, but not familial, anomalies of the intrahepatic or extrahepatic biliary tract. Cystic dilatation may affect every part of the biliary tree and may occur singly or in multiple numbers. The incidence in the population is 1:100000 to 1:150000 . The clinical classification, which describes five different types and subtypes, was revised in 1977 by Todani and colleagues. The leading symptoms include cholestatic jaundice and abdominal pain. A palpable abdominal mass occurs in less than 20% of the cases. In adults, chronic and intermittent abdominal pain is the most common symptom. Recurrent cholangitis and jaundice may also occur.

CASE HISTORY:

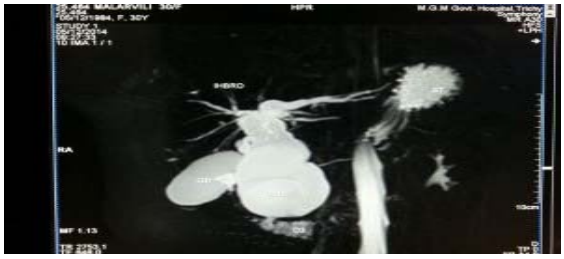
30 years old female patient admitted with h/o abdominal pain & vomiting for three days duration. Pain was in the right hypochondrium, dull aching and was not radiating. No h/o fever. She had similar complaints in the past which resolved by itself. Pt had no other complaints. On general examination she was moderately built ,afebrile and not icteric. Abdomen examination – Soft, rt hypochondric tenderness present, no guarding /rigidity. Bowel sounds present. Blood investigations showed elevated liver enzymes. OGD scopy showed normal study. USG ABDOMEN:Distended GB, grossly dilated CBD with fusiform dilation of mid and distal CBD and multiple small CBD calculi. CT ABDOMEN: Mild IBHR dilation ,sacular and fusiform dilation of distal CBD. choledochal cyst with IBHR dilatation. MRCP: Gross cystic dilation of intra and extra hepatic biliary radicles with multiple calculi within CBD.choledochal cyst type 4A.



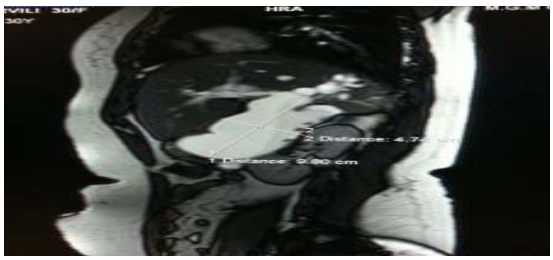
CT SHOWS GB AND CBD WITH STONES



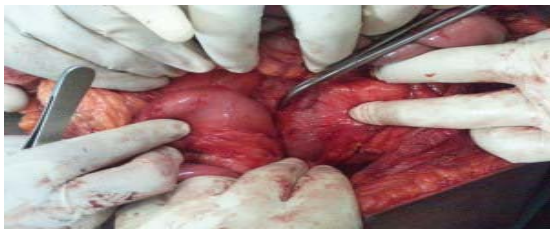
CT SHOWS DILATED IBHR



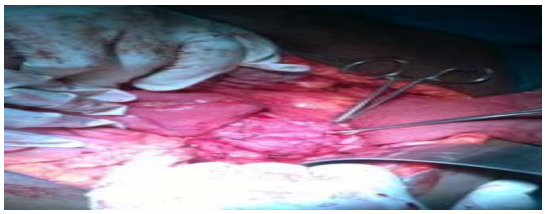
MRCP-CHOLEDOCHAL CYST WITH IBHR



MRCP-CHOLEDOCHAL CYST
PROCEDURE:
Patient was taken up for surgery Under epidural with general anaesthesia and supine posture, roof top incision was made .Thickened GB which was adherent to the undersurface of liver with 8-9cm fusiform dilation of CBD was seen. Cholecystectomy was done by funds first method.Choledochal cyst was separated from the surrounding adhesions and excision of dilated CBD segment done. Choledochotomy showed multiple stones in CBD. Since there was no hepatolithiasis, intrahepatic ductal strictures and hepatic abscess, we had done Roux-en-Y hepaticojejunostomy to restore continuity. Patient's post op period was uneventful. HPE showed cyst lined by low cuboidal epithelium, benign choledochal cyst. Post op liver enzymes were normal. Follow up USG showed normal liver, and pancreas, no collection.



INTRAOP PIC -DILATED CBD



CHOLEDOCHAL CYST
DISCUSSION:
Choledochal cysts are rare, congenital but not familial anomalies of the intrahepatic or extrahepatic biliary tract. Cystic dilatation may affect every part of the biliary tree and may occur single or in multiple numbers. The incidence in the population are 1:100000 to 1:150000 [1]. The clinical classification which describes five different types and subtypes, was revised in 1977 by Todani and colleagues [2].

· **Type I:** Most common variety (80-90%) involving saccular or fusiform dilatation of a portion or entire common bile duct (CBD) with normal intrahepatic duct.
· **Type II:** Isolated diverticulum protruding from the CBD.
· **Type III or Choledochocele:** Arise from dilatation of duodenal portion of CBD or where pancreatic duct meets.
· **Type IVa:** Characterized by multiple dilatations of the intrahepatic and extrahepatic billiary tree.
· **Type IVb:** Multiple dilatations involving only the extrahepatic bile ducts.
· **Type V:** Cystic dilatation of intra hepatic biliary ducts. Not the same etiology as caroli's disease.
· **Type VI:** An isolated cyst of the cystic duct is an extremely rare lesion. Only single case reports are documented in the literature. The most accepted classification system of biliary cysts, the Todani classification, does not include this lesion. Cholecystectomy with cystic duct ligation near the common bile duct is curative. The most common cystic dilatation is type I with diffuse or segmental fusiform dilatation of the common bile duct. This type accounts for 50 to 85% of cases. Type I cysts should be considered in the differential diagnosis of any patient with ductal dilatation.

Type 4
Multiple communicating intra- and extrahepatic ductal cysts.
· second most common type of bile duct cysts (10%)
· subdivided into sub-types A and B
o **type 4a:** fusiform dilation of the entire extrahepatic bile duct with extension of dilation to the intrahepatic bile ducts
o **type 4b:** multiple cystic dilations involving only the extrahepatic bile duct
The leading symptoms include cholestatic jaundice and abdominal pain. A palpable abdominal mass occurs in less than 20% of the cases. In adults, chronic and intermittent abdominal pain is the most common symptom. Recurrent cholangitis and jaundice may also occur. A choledochal cyst is rarely symptomatic but should be considered if dilatation of the bile duct or the ampulla is demonstrated. The main diagnostic tool for detection of a choledochal cyst especially in childhood is ultrasonography. In adults, computer tomography can confirm the diagnosis; however, endoscopic retrograde cholangiography or magnetic resonance cholangiography are the most valuable diagnostic methods and can accurately show cystic segments of the biliary tree [3].Surgery is the treatment of choice for a choledochal cyst. Complete excision of all cystic tissue is recommended because of the risk of recurrent cholangitis and the high risk of malignant degeneration [4]. Excision of the cyst and reconstruction of the biliary tree by choledochal/hepato-jejunostomy with a Roux-en Y-loop is the standard procedure [5].

In comparison, simple congenital hepatic cysts are very common. Their incidence is 1:40 in the population and simple congenital hepatic cysts represent the most important differential diagnosis[6] These cysts are also rarely symptomatic. They are detected incidentally during an operation or by diagnostic measures for other conditions and generally do not require treatment [5]. If symptoms occur in the case of larger cysts, non-specific upper abdominal discomfort and a palpable abdominal mass are most common [7]. Symptomatic cysts can be treated by non-operative invasive intervention or by an operative procedure. Operative procedures comprise cyst fenestration, partial or total cyst resection, and hepatic resection. Laparoscopic cyst fenestration is the treatment of choice because it is a simple and effective procedure with a low mortality[5].Choledochal cysts are rare abnormities of the biliary tree and so may be frequently overlooked in the differential diagnosis. The non-specific symptoms of choledochal cysts, including pain in the upper abdomen and jaundice, are common in many other illnesses of the upper gastrointestinal tract. The clinical triad of jaundice, a palpable mass and abdominal pain occurs only in one-third of all patients. Abdominal pain is the prominent complaint in adults, which also led our

patient to seek medical attention. The choledochal cyst (1:100000) was easily mistaken, as may frequently happen, for a much more common solitary congenital liver cyst (1: 1000), especially if typical symptoms are absent in a large cyst [11,12]. Ultrasonography is usually the first examination and is very sensitive in the detection of cystic structures, but rather non-specific in identifying their origin. A computer tomography usually can give more information and modern techniques, including reconstruction, should allow for establishing the diagnosis. Endoscopic retrograde cholangiography or magnetic resonance cholangiography can precisely visualize the extrahepatic bile duct and these are the most specific diagnostic procedures. The treatment of a choledochal cyst has changed. In the past, a cysto-jejunostomy was the standard procedure. Currently, excision of the cyst and reconstruction by hepatojejunostomy is the standard therapy [13]. This case report also demonstrates the intraoperative difficulties in identifing a choledochal cyst. Retrospectively, an entire exploration, including elevation of the liver, should have been able to demonstrate a clear separation of the cystic structure from the liver. This intraoperative exploration should be performed and prompt any surgeon to dispute the preoperative diagnosis. Laparoscopic fenestration of a hepatic cyst is the appropriate approach. However, the finding of bile, and of even greater significance, two bile ducts, while possible, is so unusual for a hepatic cyst that it justifies an intraoperative re-evaluation by cholangiography. An intraoperative cholangiography in this case would have clarified the anatomy and pathology beyond any doubt.

Conclusion

This case demonstrates the diagnostic and therapeutical difficulties in the treatment of choledochal cysts. As Adult choledochal cyst ,the classical triad is rarely seen. Hence it should be included in one of the differential diagnosis of upper abdominal pain.

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