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A CASE OF CHOLEDOCHAL CYST - ON TABLE SURPRISE AND MANAGEMENT OF POST OPERATIVE COMPLICATION

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Abstract :
A 28yrs old female presented with abdominal pain at right hypochondrium vomiting for 1wk. USG CECT-abdomen reported as hydatid cyst. On laparotomy it is surprisingly found to be choledochal cyst. Complete excision of cyst and roux-en-y hepaticojejunostomy done. Post operatively patient develops bile leak which is treated conservatively with draining. Then she developed pancreatic leak which is treated with pancreatic duct stunting. Patient discharged in good condition and on follow up.Choledochalcyst is a rare condition and in this case its presentation in adult, importance of imaging, management, post-operative complication and its management.

Keyword :
choledochal cyst, todani classification, roux-en y hepaticojejunostomy, pancreatic fistula

INTRODUCTION:
Choledochal cyst is a rare case found between 1 in 100,000 & 1 in 150,000 populations[1]. Although frequently diagnosed in infancy or childhood, as many as one half of the patients have reached adulthood when diagnosed. Adults commonly present with jaundice or cholangitis. Less than one half of patients present with the classic clinical triad of abdominal pain, jaundice, and a mass. Ultrasonography or CT scanning will confirm the diagnosis, but endoscopic, transhepatic, or MRCP is required to assess the biliary anatomy and to plan the appropriate surgical treatment.

CASE REPORT:
A 28 years old female patient was admitted with complaints of abdominal pain in right hypochondrium for a duration of 1 week which was pricking in nature and was associated with nausea & vomiting. On examination the patient had mild tenderness in her right hypochondrium as well as epigastrium. The patient also had a hepatomegaly extending upto 3cm below right costal margin. Ultrasound abdomen revelaed a hydatid cyst involving both lobes of liver of size 7cm X 6cm. Contrast enhanced CT of the abdomen also showed a hydatid cyst of the same size occupying segments 3, 4, 6, 7.(fig:1)

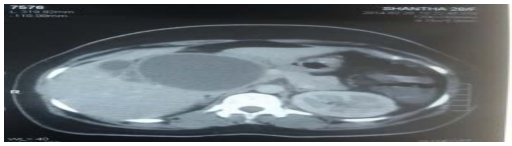

Fig:1 CECT showing the cystic lesion in liver

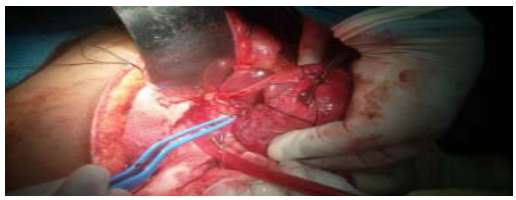

Fig:2 intraoperative view of choledochal cyst

The patient was then taken up for laparotomy only to find a choledochal cyst type 1 and not a hydatid cyst.(fig:2) The ontable surprise was well tackled and a complete excision of the cyst along with cholecystectomy and roux - en - y hepaticojejunostomy was done.(fig:3)

POST OPERATIVE COMPLICATION AND ITS MANAGMENT:
The patient developed post operative bile leak on 6th post operative day which was evident from the bile leak through the main wound. It is The anastamotic leak was confirmed by the free bilious collection as seen in the contrast CT. A minilaparotomy was done and patient was managed with two foley's catheter in main wound and one more in right flank drain site on 7th post operative day. Around 1000ml bilious fluid was drained for 5 days and the drain fluid reduced to around 300ml in next 7 days. The drain fluid turned into a opalescent fluid. Drain fluid amylase was done and it was 1,36,400 IU/L thus confirming a pancreatic fluid leak probably due to pancreatic fistula. Endoscopic Pancreatic Stent[2] was placed with pigtail stent on 23rd post operative day and the drain reduced to minimum levels when it was removed one by one on 26th & 27th post operative days. Patient was discharged on 30th POD and is on follow up.

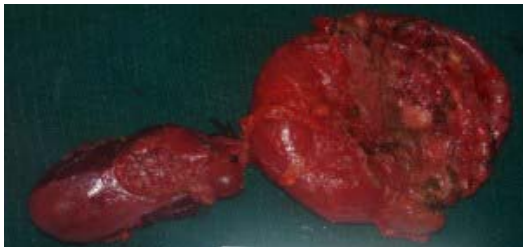

Fig:3 postoperative specimen showing the excision of the choledochal cyst along with cholecystectomy

DISCUSSION

Choledochal cysts are focal or diffuse dilatations of the biliary tree that are believed to be congenital abnormalities. The female: male ratio is between 3:1 and 4:1. The incidence of choledochal cysts varies. The incidence in Asia is as high as 1 in 1000. In Western countries, the incidence is 1 in 13,000 to 1 in 150,000.

Todani Classification of Choledochal Cysts [3]

Type I- Classic cystic dilatation of the common bile duct, most common type. It comprises 50–85% of all biliary cysts; subdivided into three. I-A (cystic), I-B (fusiform) and I-C (saccular) Type II- Simple diverticulum of the extrahepatic biliary tree, comprising less than 5% of all cysts & located proximal to the duodenum. Type III- Cystic dilatation of the intraduodenal portion of the extra hepatic common bile duct also known as a choledochocele comprise approximately 5%. Type IV- Involve multiple cysts of the intrahepatic and extrahepatic biliary tree. It is subdivided into two types. IV-A (both intrahepatic and extrahepatic cysts), IV-B (multiple extrahepatic cysts without intrahepatic involvement), type IV-A is the second most common type of biliary cyst (30–40%). Type V- Isolated intrahepatic biliary cystic disease, also known as Caroli's disease -associated with periportal fibrosis or cirrhosis, can be multilobar or confined to a single lobe

Pathogenesis

The exact cause of choledochal cysts remains unclear. Several theories are proposed.

1.Congenital.
2.Weakness of the wall of the bile duct.
3.Obstruction of the distal choledochus.
4.Combination of obstruction and weakness.
5.Anomaly of the Pancreatico Biliary Junction (APBJ), accounts for about 60-80% of cases of choledochal cyst.

Anomalous pancreaticobiliary junction

The abnormal junction of the pancreatic duct [4] and common bile duct occurs outside the duodenal wall to form a long common channel (>15 mm). This allows reflux of pancreatic juices into biliary system, leading to increased intraductal pressure and inflammation. The anomalous junction is often associated with a choledochal cyst or a biliary tract carcinoma and predisposes to acute pancreatitis. Endoscopic retrograde cholangiopancreatography (ERCP) allows accurate diagnosis of the anomalous junction, but ERCP is invasive. Multidetector CT (MDCT) is a recently developed technology that has provides detailed information on the pancreaticobiliary ductal anatomy. Treatment is surgical correction

Choledochal cyst-Presentation

The classic triad of presentation is pain, jaundice, and abdominal mass. Triad is found in only a minority of children at the time of presentation. Infants commonly present with elevated conjugated bilirubin (80%), failure to thrive, or an abdominal mass (30%). In patients older than 2 years of age, chronic and intermittent abdominal pain(50-96%), Intermittent jaundice and recurrent cholangitis are also common (34-55%), abdominal mass is less common (10-20%), Pancreatitis(20%), biliary lithiasis(8%). Intermittent jaundice and recurrent cholangitis are also common, as is pancreatitis.

Diagnosis

The diagnosis of a choledochal cyst requires a high level of suspicion. Ultrasound or CT scan imaging can be especially helpful in suggesting the presence of a choledochal cyst. Magnetic Resonance Cholangio Pancreatography (MRCP) may also be helpful in the diagnosis of choledochal cyst. Cholangiography is considered the gold standard for diagnosis of choledochal cysts. Percutaneous transhepatic cholangiography (PTC) or endoscopic retrograde cholangiography (ERC) is typically performed on adults and larger children, while intraoperative cholangiography may be done on infants and small children. Cholangiography can demonstrate areas of cystic dilatation, the presence of stones, and excludes complete obstruction of the bile duct. It is also effective in demonstrating the presence of pancreaticobiliary maljunction.

Why is it important to operate?

Cholangiocarcinoma: they have 20-30 fold higher risk than general population. The risk of cholangiocarcinoma in the first decade of life is only 0.7%[5]. This increases to 14% at 20 years and increase even further throughout life. Cholangiocarcinoma may develop in all kinds of cysts but type I and IV cysts are associated with a higher incidence, even after cyst excision. Complete excision of these lesions is recommended as early as possible, preferably before puberty, to decrease the chance of developing cancer. It is also associated with an increased risk of gallbladder malignancy. Prophylactic cholecystectomy is also advised in all patients with either pancreaticobiliary maljunction or choledochal cyst.

Management

The operative management of choledochal cysts [6] should first consist of careful exploration of the patient. Midline incision laparotomy, the initial step should be searching for possible metastatic disease. Once metastatic disease has been excluded, management of the choledochal cyst depends upon the type. Type I cysts - complete cyst excision with cholecystectomy and reconstruct the bile duct by hepaticojejunostomy or hepaticoduodenostomy, although Roux-en-Y hepaticojejunostomy is by far the most commonly used technique. Long-term results are generally excellent, especially with type I cysts. Type II cysts treated with simple cyst excision. CBD should be closed transversely (avoid stricturing) Type III cysts excision uncommon (low malignancy rate). Endoscopic sphincterotomy (symptomatic), Transverse duodenotomy for resection. Type IV-A extrahepatic is managed similarly to type I cysts with extrahepatic biliary resection, cholecystectomy, and biliary reconstruction. Regarding intrahepatic ducts in type IV-A, surgery should be individualized depending on lobes affected, strictures or stones, cirrhosis or malignancy. If intrahepatic cysts are localized into one lobe, hepatic lobectomy is the preferred approach. For diffuse intrahepatic disease, liver transplantation would be considered. Type IV-B cysts are managed similarly to type I cysts with regard to extrahepatic biliary resection, cholecystectomy, and biliary reconstruction. Type V (Caroli's disease): If unilateral or segmental with cirrhosis: resection of the involved parenchyma. In the absence of cirrhosis or malignancy, Roux-en-Y hepaticojejunostomy with bilateral transhepatic Silastic stents may be indicated to improve biliary drainage (stents left for 6-12 months). Patients with Caroli's disease and liver failure needs liver transplantation.

Conclusion

Choledochal cysts require proper diagnosis. Appropriate imaging and reporting is mandatory. Treatment is to

address associated symptoms, risk of malignancy, and disease progression. The majority of cases of biliary cysts (type I and IVA) can be treated effectively with cyst resection, cholecystectomy, and biliary reconstruction. Surgery decreases the risk of subsequent cancer, It require long-term surveillance for recurrent cholangitis, intrahepatic stones, pancreatitis, postoperative biliary strictures, and malignancy. Patient may develop post operative complications like anastamotic bile leak and pancreatic fistula. Bile leak can be managed by proper drainage and monitoring and pancreatic fistula can be managed by endoscopic pancreatic stenting.

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