Case Report

Diagnostic dilemma in a large choledochal cyst

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Abstract

Choledochal cysts are rare congenital anomalies that have a variable presentation with occasional diagnostic and therapeutic dilemmas.

Our case is a 14-year female presented with recurrent abdomen pain with confusing diagnostic findings, suggestive of a large choledochal cyst (22X29X17cm) initially managed by percutaneous tube biliary drainage and later underwent excision and HPE revealing non-malignant findings.

The choledochal cyst should always be a differential diagnosis for any patient presenting with an abdominal mass and should be managed accordingly.

Keywords: Choledochal cyst; Hepaticojejunostomy; Percutaneous biliary drainage

Introduction

Choledochal cysts are congenital conditions involving cystic dilatation of bile ducts and were described for the first time by Vater in 1723.[1] The incidence is one case per 1,00,000 to 1,50,000 live births with 75% presenting at childhood and 25% at adulthood. The ratio of incidence in male to female is 3-4:1.[2]

Case report

A 14-year female presented with recurrent abdominal pain and distension for 7-8 months and jaundice for 2weeks. The patient gives no history of fever, nausea vomiting or altered bladder habit. On examination, the child was afebrile but was pale and icteric. The abdomen was soft, with fullness and mild tenderness at the right hypochondrium and epigastric region. And a large cystic mass that moved with respiration at the right hypochondriac region extending up to the right iliac fossa. A provisional diagnosis of a hydatid cyst of the liver was made.

On further workup, her hemoglobin was 5.6gm/dl and her total serum albumin was 1.9gm/dl. Her total bilirubin was 3.9mg/dl with direct bilirubin 1mg/dl with alkaline phosphatase 114U/l. Her renal function test and bleeding profiles were within normal limits. Ultrasonography of the abdomen showed huge cystic swelling at the subhepatic region of around 20 X 20 X 19 cm; a suspicion of hydatid cyst was made but they could not define the origin of the cyst. Magnetic resonance cholangiopancreatography (MRCP) findings were also confusing with hydatid cyst and choledochal cyst as a differential diagnosis. Her contrast-enhanced computerized tomography (CECT) findings suggested of giant (22.3X29.5X17.2 cm- 4050cc) type I choledochal cyst with moderate intrahepatic biliary radicals dilatation (IHBRD) with folded GB and intrahepatic Phrygian cap. Figure 1. ERCP was attempted twice but failed to cannulate the ampulla because of anatomical distortion. Her hemoglobin was corrected and PTBD drain was kept for relieving jaundice. After 4 weeks of nutritional build-up, her hemoglobin was 12.1gm/dl and the albumin level was 3.4gm/dl. She underwent exploratory laparotomy.

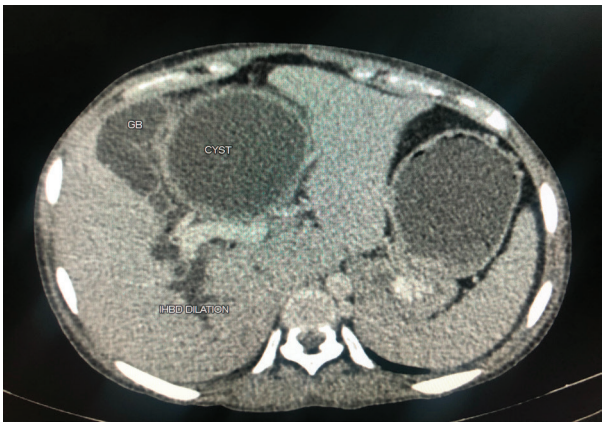

Figure 1: CECT showing Liver, Gallbladder and the Cyst

At laparotomy, the cyst was seen collapsed with the biliary drain in situ, with cystic duct entering the upper end of the cyst and the gallbladder was contracted. Figure 2. The total excision of the choledochal cyst was done followed by Roux-en-Y hepaticojejunostomy. Figure 3. Her postoperative recovery was uneventful and she was discharged on 7th postoperative day.

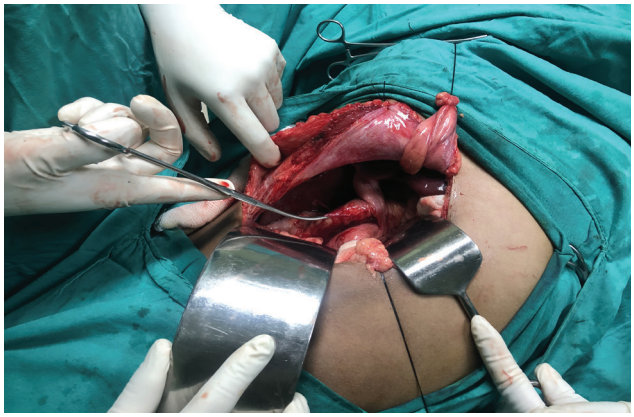

Figure 2: Intraoperative finding of the cyst which was already decompressed after PTBD drain

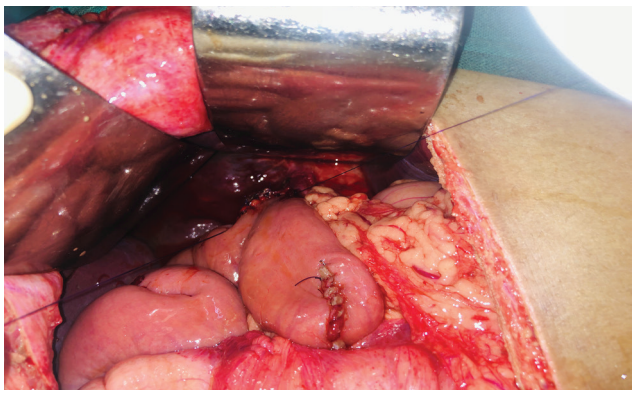

Figure 3: Final picture of the Roux-en-Y hepaticojejunostomy

Pathology confirmed the choledochal cyst and showed no malignant change.

Discussion

Choledochal cyst is a rare clinical entity that is defined as congenital dilatation of any portion of the bile duct.[4] Among many hypotheses, the Babbit's theory is one of the accepted theory for its cause, according to which, the long common channel allows mixing of the pancreatic secretions and bile for longer than usual, activating pancreatic enzymes.[4,5] The activating enzymes then cause inflammation and destruction in the wall of the biliary tract, causing dilatation.[5]

The widely accepted classification system for choledochal cyst is the Todani classification; a modified version of Alonso-Lej classification.[3] Type I is the most common (80-90%) and is subdivided into Ia (cystic dilatation of extrahepatic bile ducts), Ib (extrahepatic distal focal segmental biliary dilatation), Ic (extrahepatic fusiform dilatation). Type II is extrahepatic biliary diverticula, type III is choledochocele, type IVa is multiple cystic dilatations of the intrahepatic and extrahepatic bile duct, type IVb multiple cystic dilatations of the only extrahepatic bile duct, type V is intrahepatic dilatation of bile duct (Caroli's disease) .[3]

The triad of abdominal pain, jaundice, and an intraabdominal palpable mass are known as classical presentation, but this triad is present in only 20% of cases.[9] Pathologically, the choledochal cyst is devoid of epithelium and the wall consists of fibrous tissue, they fail to contract to lead to poor emptying causing bile stasis.[6]

The first line investigation to diagnose choledochal cyst is ultrasonography however, MRCP and CECT are diagnostic as well as helpful for delineating relevant anatomy.[8, 10] Here the surprising thing was that, though MRCP is the diagnostic modality of choice for choledochal cyst, in our case CECT was diagnostic, maybe because it was a large cyst.

The complications of choledochal cyst include recurrent cholangitis, calculus disease and cholangiocarcinoma.[7] Surgery is the mainstay of treatment once diagnosed. Type I cysts need resection and reconstruction of the bile duct by means of Roux-en-Y hepaticojejunostomy.[6] Type II cyst can usually be excised, the defect in the CBD being repaired by primary suture over a T-tube.[8] Type III may be repaired by transduodenal sphincteroplasty.[7] Type IV and V may need partial or complete hepatic resection followed by hepatic transplantation.[7] The prognosis for extrahepatic choledochal cyst with appropriate resection and reconstruction is usually good.[8]

Such a large choledochal cyst is very rare and may present a diagnostic dilemma. Sometimes even investigations may confuse more than enlighten.

Conclusion

The diagnosis of a choledochal cyst can be challenging sometimes. The choledochal cyst should always be a differential diagnosis for any patient presenting with an abdominal mass.

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