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Case Report

Choledochal cyst or gallbladder duplication? Diagnostic laparoscopy for chronic abdominal pain in a 10 year old boy

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ABSTRACT

Structural abnormalities in close proximity with the gallbladder fossa such as gallbladder duplications could be misrepresented as choledochal cysts, duodenal duplication cysts or gallbladder adenomyomas on sonography. We present a case of a ten year old boy who presented with a chronic abdominal pain of a three month duration and nausea and vomiting. Ultrasound and magnetic resonance cholangiopancreaticoography (MRCP) was reported by radiology as a possible 2.7 x 1.7 x 1.6 cm choledochal cyst with high differential consideration for a gallbladder duplication. The patient underwent a diagnostic laparoscopy and a grossly normal gallbladder was visualized with no gross visualization of cystic biliary tree dilatation. The gallbladder was successfully resected laparoscopically following a meticulous dissection of the triangle of calot. Pathology showed evidence of a septate gallbladder with a common cystic duct and no evidence of cholelithiasis. The patient is two years post -laparoscopic cholecystectomy and remains symptom-free. Successful laparoscopic resection of a duplicate gallbladder requires a pre-operative diagnostic delineation of the biliary tree anatomy to rule out other possible differentials.

Keywords: Gall bladder, Duplication, Laparoscopic, Cholecystectomy

INTRODUCTION

Duplicated gallbladder is rare and easily confused with choledochal cysts and duodenal duplications. Distinguishing these two conditions preoperatively is important as their management modalities are different.

CASE REPORT

A ten year old African boy resident with both parents in New York City presented at the Emergency Department of our facility with acute onset of a chronic right upper quadrant abdominal pain of a three month duration. The pain had started a few hours prior to presentation, was of sharp intensity and unrelated to meals. He had multiple episodes of vomiting. His mother denied any fever, weight loss, recent travel or jaundice. Vital signs on initial presentation include a blood pressure of 112/82 mmHg, pulse of 88/min Respiratory rate of 14/min temperature of 99.5°F. Physical examination revealed no jaundice or palpable mass. He was admitted to the pediatric surgery service for further evaluation. Admission complete blood count was normal, hepatic panel showed aspartate aminotransferase was 16 mg/dl, alanine aminotransferase was 20 mg/dl, alkaline phosphatase was 269 mg/dl. Total bilirubin and direct bilirubin was within normal limits and amylase and lipase levels were normal.

Abdominal ultrasound (Figure 1) on admission showed normal gallbladder wall thickness and no peri-cholecystic fluid. Adjacent to the gallbladder was a cystic structure measuring approximately 2.7 x 1.7 x 1 .6 cm, concerning for a choledochal cyst. Images on MRI/MRCP (Figure 2 and 3) done a day following admission showed multiple communicating cysts of varying sizes around the gallbladder - possibly representing choledochal cysts or a multiple - septated gallbladder. There was no evidence intra-hepatic or extra-hepatic biliary duct dilatation.

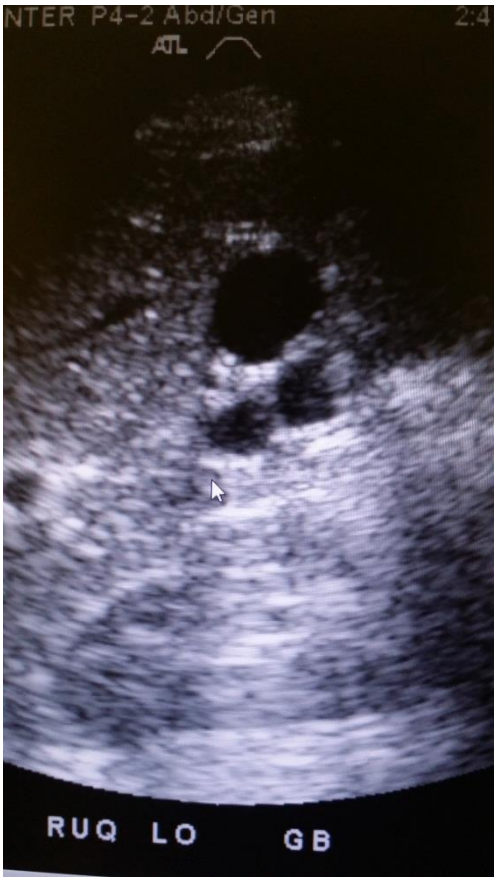

Figure 1: Arrow pointing to multiple cystic structures on gallbladder ultrasound suspicious for choledochal cysts.

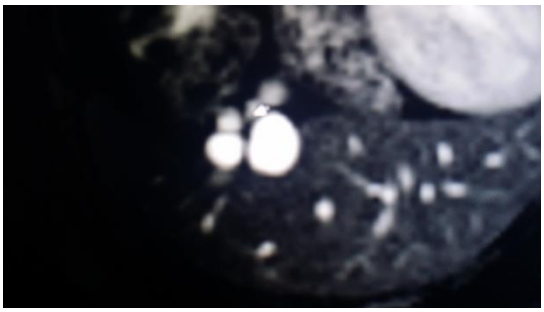

Figure 2: T2-weighted MRI image showing multiple cystic lesions around gallbladder and identifying a possible choeldochal cyst.

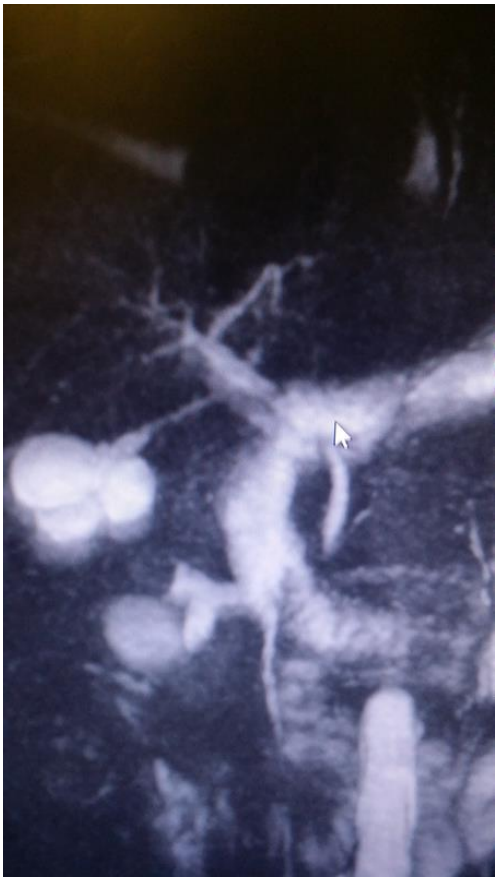

Figure 3: Magnetic resonance cholangiopancreaticography showing multiple cysts in the gallbladder area of varying sizes which communicate and could represent a choledochal cyst or a multi-septated gallbladder.

At this juncture, the patient was planned for a diagnostic laparoscopy and possible laparoscopic cholecystectomy or choledochal cyst resection with Roux-en-Y hepatico-jejunostomy. Access into the abdominal cavity was gained utilizing an umbilical 12 millimeter port, three additional 5 mm ports were placed in the right upper quadrant following which a grossly normal gallbladder was visualized and successfully resected after a thorough laparoscopic delineation of the triangle of calot.

The patient recovered uneventfully and was discharged 4 days postoperatively. Pathology revealed a singular cystic duct and two cystic gallbladder lumens with separate tunica muscularis and septation in one lumen. There was no evidence of cholelithiasis/cholecystitis.

Routine ultrasound 3 weeks following surgery showed a normal status post cholecystectomy, normal hepatic parenchyma and normal common bile duct diameter.

DISCUSSION

Gallbladder duplications and choledochal cysts are both rare congenital abnormalities of the gallbladder and

biliary tree primordium and are often difficult to differentiate clinically and radiologically. Gallbladder duplications are often a result of failure of extra-hepatic biliary ducts outpouchings to regress,[1] while choledochal cysts are more commonly attributed to an anomalous pancreatobiliary ductal junction (APBDJ) with reflux of pancreatic juice into the biliary tree and resultant ductal cystic dilation.[2]

Duplicate gallbladders has been reported in 1 per 12000 cholecystectomies and 1 in 4000 autopsies[3] while choledochal cysts have a much lower incidence of 1:100000-150000 with female predominance. Both gallbladder duplications and choledochal cysts are most often diagnosed before the age of ten.[1,2] The classic triad of abdominal jaundice, pain and a palpable abdominal mass is rarely seen in choledochal cysts[2] while abdominal pain from symptomatic cholelithiasis is the most frequent pathology associated with duplicated gallbladder. In our case, cystic dilations were seen on ultrasound and MRCP without evidence of gallstones or cholecystitis serving as a confounding factor for the etiology of the abdominal pain. Elevated alkaline phospahatase is a normal finding in children; but may also be in keeping with cholestasis from choledochal cysts as opposed to a gallbladder anomaly in the absence of stones on ultrasound. The absence of biliary tree dilatation on ultrasound and MRCP was also a minus for the diagnosis of choledochal cyst.

Ultrasonography is the initial investigative modality for both gallbladder and biliary tree disease with gallbladder duplications easily labelled as more sinister cystic intra-abdominal pathologies such as choledochal cysts or duodenal duplication cysts by sonography.[3,4] The MRCP modality is also useful in differentiating both conditions; both were of limited diagnostic benefit in our case.[2,4] For the surgeon considering both etiologies as possible differential diagnoses, a thorough evaluation of the biliary ductal system via MRCP or endoscopic retrograde cholangiopancreaticography (ERCP) is considered optimal pre-operative planning.[1-4] The higher diagnostic possibility of gallbladder disease as opposed to choledochal cyst; the adverse effects of ERCP such as radiation exposure and contrast exposure in a 10 year old boy coupled with the risk of cholangitis and pancreatitis resulted in the decision for a diagnostic laparoscopy as opposed to an ERCP.

Boyden described the A to F gallbladder duplications in 1926.[5] There are newer classifications such as the Harlaftis classification and its modifications.[1,6] A, B and C are partial duplications and classified into septate, fundic and body types. The septate type has a septum within the gallbladder while the fundic type is isolated to the gallbladder fundus and the body type has a split body. True double gallbladders are the D, E and F types of Boyden classification. Type D represents the Y-shaped gallbladders (in which both cystic ducts from a twin gallbladder merge into a Y-shaped bifid cystic duct before entering the common bile duct), E represents 2 separate cystic ducts and F represents bilateral gallbladders.[2] The Harlaftis classification into type 1 (split primordial) and 2 (accessory gallbladder) groups based on whether a common cystic duct is shared. Type 1 is either V shaped, Y-shaped or septate. Type 2 classifies accessory gallbladders into the ductular or trabecular types. The histologic class in our case was a type 1 septate gallbladder with a common cystic duct based on the Harlaftis classification.[6]

The modified Todani classification of choledochal cysts include types one to five with sub-classifications.[2,4] Type 1 is classified into A to C. Type A is cystic dilation of entire extra-hepatic biliary tree without intra-hepatic involvement. Focal extra-hepatic biliary tree dilation is seen in Type IB while Type IC is a fusiform dilation of the entire extra-hepatic biliary tree plus intra-hepatic duct involvement. Type II choledochal cysts represent a saccular diverticulum of the common bile duct and Type III choledochoceles are cystic dilations extending into the duodenum. Type IVA represent both intra-hepatic and extra-hepatic biliary ducts dilation while Type IVB represents multiple dilation of extra-hepatic biliary tree only. Type V (also known as Caroli's disease) represents multiple intrahepatic biliary ductal dilation.[2,4]

Surgery is recommended for choledochal cysts predominantly for its propensity for malignant transformation[2,4] whereas cholecystectomy is not indicated for incidental Gallbladder duplications.[1] Successful laparoscopic cholecystectomy has been described in cases of type 1 septate gallbladders as seen in our case[1] but higher rates of open surgery conversion are seen in difficult dissections with greater chance of ductal injury.

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