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AGENESIS OF GALL BLADDER: A CLINICORADIOLOGICAL DILEMMA

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ABSTRACT

Agenesis of gall bladder is a rare congenital biliary abnormality which is rarely diagnosed preoperatively. Most of these patients present with typical symptomatology of gallstone disease which adds to the diagnostic confusion. Its clinical significance lies in the fact that this clinical entity should always be considered in patients with contracted gall bladders with stones both preoperatively as well as intraoperatively to avoid surgical catastrophes. Here we present such a case in a young male who was diagnosed to have this clinical entity only intraoperatively

Keywords: gall bladder, stone, clinical.

Introduction

Congenital absence of the gall bladder is a rare anomaly which may occur alone or in association with other malformations.' Earlier studies have described a variety of defects associated with agenesis of the gall bladder, but in the majority of reported cases this anomaly is found alone. Establishment of a preoperative diagnosis using noninvasive imaging modalities permits to avoid a risky surgical procedure

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Case report

A 25 year old male patient was admitted to our surgical unit as a case of chronic pain in upper part of abdomen. Ultrasonography of abdomen revealed the presence of gall stone disease with contracted gall bladder containing multiple calculi and normal common bile duct. After routine preoperative investigations, the patient was taken up for laparoscopic cholecystectomy. Intraoperatively, gall bladder could not be identified, common bile duct was prominent and the structures at the porta hepatis could be clearly visualised (intraoperative photographs figure 1,2 enclosed).There were adhesions of liver to the anterior abdominal wall for which the adhesiolysis was done. A thorough search for the gall bladder at the ectopic sites was done. A presumptive diagnosis of gall bladder agenesis was considered in this patient and the procedure was aborted. Postoperatively, patient was

managed conservatively and discharged in stable condition. In view of the intraoperative findings, the patient underwent repeat ultrasound abdomen at our institute which corroborated with our intraoperative diagnosis of gall bladder agenesis (Figure 3,postop ultrasound abdomen picture enclosed).

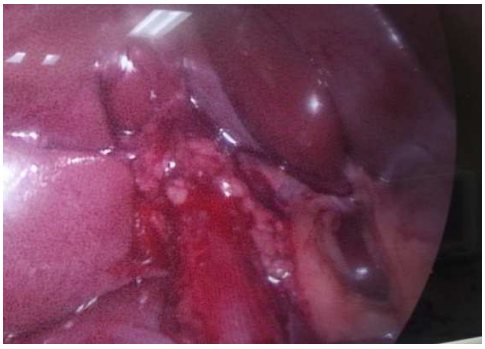

Figure 1: Intraoperative photograph showing structures at porta hepatis with absence of gall bladder

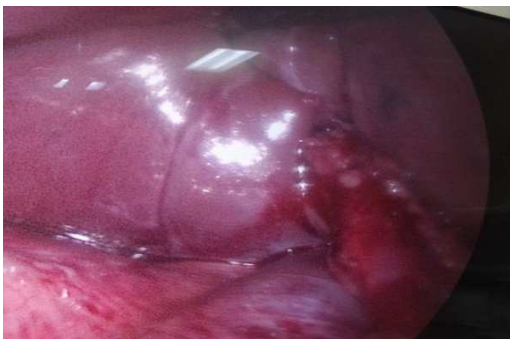

Figure 2: showing structures at porta hepatis

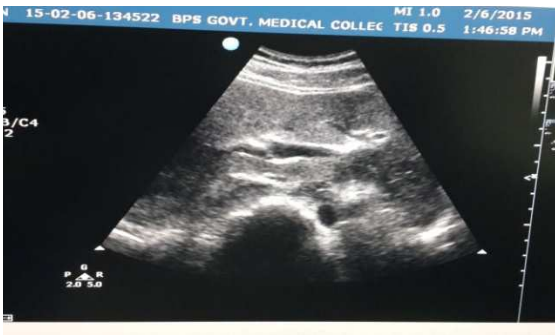

Figure 3: showing postoperative Ultrasound scan

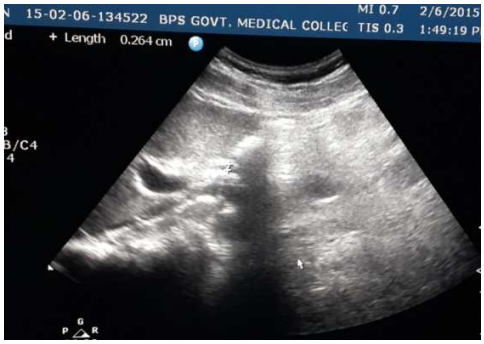

Figure 4: showing the excised fistula tract along with gall bladder

Discussion

Agenesis of gall bladder is a rare clinical entity with reported incidence of 0.01-0.04%. Female to male ratio has been found to be 3:1, however the incidence has been found to be the same in the autopsies[1]. It was first described by Leery in 1701. It may occur in isolation as well as in association with other congenital systemic anomalies like cleft palate, imperforate anus, musculo-skeletal defects, trachea-esophageal fistula, polycystic kidneys, congenital heart diseases like septal defects, syndromes like cerebrotendinous xanthomatosis, G syndrome,Klippel-feil syndrome, Trisomy 18, thalidomide abuse with an incidence of around 40-60%[2].It is usually found in association with extrahepatic biliary atresia in many cases.. It is believed to occur during third week of gestation when an unidentified insult leads to the failure of development of vessels on both sides of cystic bud, which may explain the association of systemic anomalies with this clinical entity. It is believed to have strong familial association and a non-sex linked heredity with variable penetration. Patients with agenesis of gall bladder may be classified in three groups based on their mode of presentation-A-asymptomatic (30-40%) who are found to have absent gall bladder as an incidental finding either radiologically or intraoperatively. B- Symptomatic group (45-55%) who present with dilated common bile duct with or without stones. C- Pediatric patients with systemic anomalies (15-20%) like cardiovascular, genitourinary and gastrointestinal anomalies leading to early mortality. It is believed that in absence of gall bladder, hepatic duct assumes the function of gall bladder and various pathologies like biliary dyskinesia, elevated sphincter of Oddi, basal pressure and element of cholestasis and infection of bile ducts with or without stone formation lead to various dyspeptic symptoms[3]. Patients with clinical entity may present with various dyspeptic symptoms like right upper quadrant pain, nausea, and vomiting, fatty food intolerance. Usually it is extremely difficult to diagnose this clinical entity preoperatively. USG whole abdomen is considered the diagnostic modality of choice for gallstones in 90-95% of patients but it is highly operator dependent and its accuracy is hindered by obesity, bowel gas and presence of artefacts or calcified lesions. Diagnosis of contracted gall bladder with stones has been found to be very difficult to diagnose on ultrasonography. Oral cholecystography and radionuclide scanning of the hepatobiliary tract have also been found useful in the evaluation of the function and detection of gall bladder pathologies and may show features of cystic duct obstruction providing a corroborative evidence of absent gall bladder. MRCP has been found to be the gold standard for diagnosis of this clinical entity as it helps in thorough imaging and evaluation of entire biliary tract[4]. It is non-invasive and can be used even in the cases of biliary stasis. Most of the cases of agenesis of gall bladder come to the clinical attention intraoperatively. If gall bladder is found to be missing in its normal anatomical position, one should look out for the ectopic location of gall bladder like falciform ligament, intrahepatic location, folds of lesser omentum , anterior abdominal wall,,retrohepatic, retroduodenal, retropancreatic and retroperitoneal areas[5-6].

Iatrogenic injury should be avoided during the extensive dissection while locating the gall bladder in the ectopic areas CBD must also be dissected and explored along its length from the confluence of rdshepatic ducts to its opening in duodenum especially in cases of choldocholithiasis and dilated CBD with diameter greater than 2cm. Any fibrous tissue in gall bladder fossa and porta hepatis should also be carefully explored. If still gall bladder is not identified, intraoperative cholangiography must be done to look for ectopic locations of gall bladder and stones in common bile duct which are commonly associated with this entity. if still the gall bladder is not located and cholangiogram is found to be normal, the operative procedure may be abandoned and a diagnosis of agenesis of gall bladder can be made.

Conclusion

Agenesis of gall bladder though rare, should always be considered in the differential diagnosis of contracted and ultrasonographically non visualised gall bladder in patients presenting with signs and symptoms of biliary tract disease

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Conflict of Interest: None

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