Congenital Absence of Gall Bladder

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Introduction

Although biliary system variants are extremely common, isolated congenital absence/agenesis of gall bladder (AGB) is extremely rare, with a reported incidence ranging between 0.013 and 0.075% [1]. More than 50% of the patients with AGB are symptomatic for biliary tract disease and require some form of surgical intervention [1]. In spite of all available diagnostic modalities pre-operative diagnosis of AGB may be illusive and present a surprise to the abdominal surgeon at laparotomy. We present a case of AGB who presented with features of obstructive jaundice and at operation was found to have AGB with choledocholithiasis.

Case Report

Forty year old serving soldier presented with recurrent episodes of jaundice, fever and pruritus of six months duration. Clinically he was found to have icterus. Liver and gall bladder were not palpable. Investigations revealed serum bilirubin of 6.7 mg%. SGOT 386 IU/L, SGPT 438 IU/L and Alkaline Phosphatase of 100 KA units. Ultra sonography (USG) of abdomen was reported as calculus cholecystitis with choledocholithiasis, common bile duct (CBD) measuring 8 mm. CECT abdomen was also reported as shrunken gall bladder, choledocholithiasis with mild dilatation of intra hepatic biliary radicles. MRCP done later was consistent with the USG and CECT findings except that the gall bladder was not well visualised. At operation the gall bladder was absent and the CBD was 3 cm in diameter with four large stones which were soft in consistency. The absence of gall bladder and cystic duct was confirmed by doing a per operative cholangiogram (Fig 1). A choledochoduodenostomy was done after clearing the bile duct. He had an uneventful recovery and has remained symptom free for the past six months. Pre and post operatively, patient was re-examined for associated congenital abnormalities like annular pancreas and congenital lumbar hernia but none was found.

Discussion

At the third week of foetal life ventral thickening of the endoderm at the distal end of the foregut forms the liver. The caudal proliferation of this endoderm will become the gall bladder and the cystic duct. Till date

413 cases of AGB have been reported in literature [1]. Patients with gall bladder agenesis are classified into three categories : (a) multiple foetal anomalies (12.9%), (b) asymptomatic (31.6%), (c) symptomatic (55.6%) [1]. Associated anomalies like annular pancreas and congenital lumbar hernia have been reported [2,3]. In confirming the diagnosis of an agenesis of gall bladder, it is necessary to exclude the abnormal locations which are intrahepatic, retrohepatic on the left side or within the lesser omentum or falciform ligament and retroperitoneal. Only about 50% of patients with AGB are symptomatic for biliary tract disease [1]. In spite of rapid advances in diagnostic modalities like USG, CECT, ERCP, hepato biliary imaging with various radioisotope compounds and MRCP, pre operative

Fig. 1 : Per operative cholangiogram showing absence of gall bladder

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diagnosis of AGB may be missed [1,4,5] as happened in our case. Even ERCP may fail to predict a AGB and may mimic an obstructed cystic duct [4]. All patients of AGB undergoing abdominal exploration for biliary tract disease pose a challenge to the abdominal surgeon to correctly identify AGB and avoid inadvertent injury / excision of the bile duct. AGB is confirmed at operation by meticulous dissection of the entire hepatic biliary tree and operative cholangiography [6]. In our case too, the final diagnosis was made after operative cholangiography. The importance of this anomaly lies in its recognition because it may be accompanied by choledocholithiasis, as was seen in our case. In such situations, choledocholithiasis must be treated with standard surgical procedures [6].

To conclude, agenesis of gall bladder is a rare condition but 50% of these are symptomatic. Preoperative imaging techniques may miss the diagnosis. Abdominal surgeons operating for biliary tract disease should be

aware of this rare condition to avoid injury to the bile duct.

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A somewhat advanced society has figured how to package basic knowledge in pill form.

A student, needing some learning, goes to the pharmacy and asks what kind of knowledge pills are available. The pharmacist says "Here's a pill for English literature." The student takes the pill and swallows it and has new knowledge about English literature!

"What else do you have?" asks the student.

"Well, I have pills for art history, biology, and world history," replies the pharmacist.

The student asks for these, and swallows them and has new knowledge about those subjects.

Then the student asks, "Do you have a pill for math?"

The pharmacist says "Wait just a moment", and goes back into the storeroom and brings back a whopper of a pill and plunks it on the counter.

"I have to take that huge pill for math?" inquires the student.

The pharmacist replied "Well, you know math always was a little hard to swallow."

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