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Case Report

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Gall bladder agenesis: a surgeon's dilemma

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ABSTRACT

Congenital agenesis of gall bladder is a rare anomaly which poses a diagnostic dilemma to the surgical fraternity. Ultrasonography, which is the standard investigation of choice in gall bladder diseases often gives a mistaken diagnosis of cholelithiasis in the background of a shrunken gall bladder with hyperechoic shadows suggestive of gall stones. Intra-operatively this poses a surprise to the operating surgeon who fails to locate gall bladder in its normal anatomic position as well as any of the ectopic sites. The controversy in further course of management whether to abandon the procedure and follow-up with a post-operative imaging such as MRCP or to convert the laparoscopic procedure into an open surgery has been a debate for the surgeons. Most of the literature favours the first approach, thereby reducing the morbidity associated with the surgery. In this case report we present a 42-year-old male who presented with symptoms of gall bladder disease to a peripheral surgical centre and how we approached the intra-operative dilemma once gall bladder was not visualized.

Keywords: Gall bladder, Agenesis, MRCP, Cholecystectomy

INTRODUCTION

Congenital agenesis of gallbladder (CAGB) is a well-recognized, but rare congenital abnormality affecting females more than males in ratio 3:1.[1] The reported incidence of CAGB vary from 0.013 to 0.075%.[2-5] 70-80% of population are asymptomatic throughout their lifetime. Even in the absence of gallbladder, some affected individuals present with the classical clinical picture of gall bladder disease including biliary colic. The reason behind this is poorly understood.[1] The classical presentation of a gallbladder disease coupled with the limitation of standard abdominal ultrasound to convincingly diagnose agenesis of gall bladder creates a diagnostic dilemma for surgeons.[6] Hence most of agenesis is only diagnosed intraoperatively and confirmed with post-operative magnetic resonance cholangiopancreatography (MRCP).[7] In this case report we discuss a case of 42-year-old male who presented with right upper quadrant pain which was confirmed on ultrasonography as gall-stone disease and later found to have gall bladder agenesis intraoperatively.

CASE REPORT

42-year-old gentle man presented to a peripheral surgical centre with complaints of right hypochondrial pain which was associated with dyspepsia and intolerance to fatty meal of three years duration. The pain was intermittent, non-radiating and was not associated with any vomiting, jaundice, abdominal distension or fever. Familial history was non-contributory. Physical examination of the abdomen was essentially within normal limits with no Murphy's sign or palpable lump. Radiological evaluation using sonography of the abdomen revealed a contracted gall bladder with multiple calculi. Lumen could not be identified with normal CBD. A radiological diagnosis of contracted gall bladder with wall echogenic shadow triad

suggestive of multiple calculi was given, with CBD diameter of 5 mm. His haematological as well as biochemical parameters including liver function tests and Alkaline phosphatase was within normal limits.

With the symptomatology and sonographic evidence of gall stone disease, patient was taken up for Laparoscopic Cholecystectomy under general anaesthesia. Intra-operatively gall bladder could not be identified in its usual location. Right lobe of liver was lifted for better visualization. All probable ectopic locations were inspected for the presence of gallbladder, failing which the procedure was abandoned and decided to investigate further using MRCP.

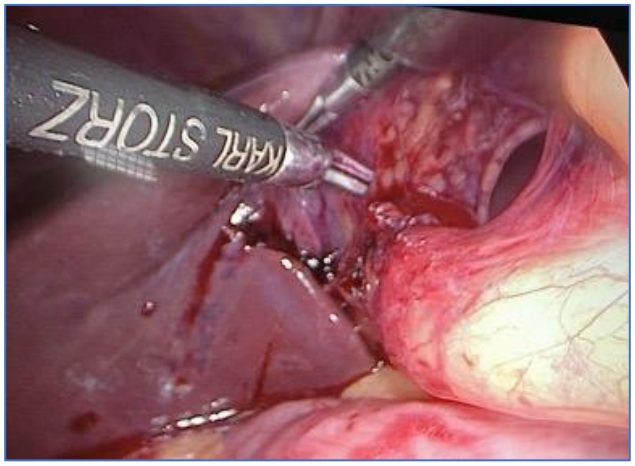

Figure 1: Falciform ligament take-down.

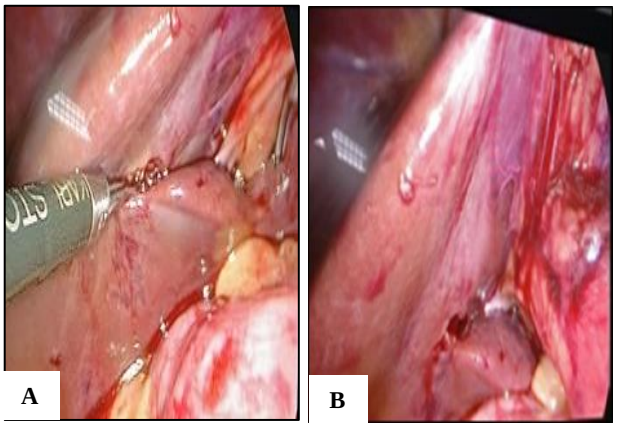

Figure 2 (A and B): Scanning under surface of Liver for ectopic gallbladder.

Postoperative evaluation by MRCP revealed features suggestive of agenesis of gall bladder with anomaly of biliary tree. Gall bladder and cystic ducts were not visualized. The right anterior and posterior hepatic duct were joining left hepatic duct at porta to form common hepatic duct which was further joining main pancreatic duct to form CBD and draining in to major papilla.

Patient was discharged on post-op day five and still experience intermittent pain upper abdomen. He is been managed conservatively with antacids and life style modifications.

DISCUSSION

The embryological development of the biliary tree starts from the fourth week of intrauterine life with the liver, gallbladder, and biliary system developing as a ventral outgrowth from the caudal part of the foregut. This hepatic diverticulum as it grows splits into two parts, one representing the primordium of the liver, and the second, the primordium of the gallbladder and cystic duct. A vacuole and a stalk develop from pars cystic by the seventh week, representing the gallbladder and the cystic duct respectively. Gall bladder being a hollow organ in the initial stages, is followed by proliferation of its epithelium determining a phase in which its cavity is temporarily cancelled. The vacuolation of its epithelium again transforms it into a hollow organ. Failure of this developmental process at any stage results in agenesis of the gallbladder.[8,9] The occurrence of the pathology is most often sporadic; however familial occurrence of CAGB has also been documented in literature, often associated with hereditary forms such as congenital syndromes, trisomy 18 and even associated with thalidomide therapy.[10-12] It has even been reported as a heritable trait.[5]

This rare anomaly was first reported in humans by Lemery in 1701.[13] Mostly diagnosed during surgery or autopsy, the incidence in general population is believed to be around 10-65 per 100000 population.[14] Literature cites a classification system by Bennion et al in 1988 wherein he divided CAGB in to three groups (i) multiple foetal malformations (30%) (ii) asymptomatic (31.6%) (iii) symptomatic (56.6%).[4] The symptomatology in group (iii) include jaundice, right upper quadrant pain, dyspepsia, vomiting and intolerance to fatty food.[5]

The preoperative diagnosis of agenesis of gall bladder is complicated by the fact that there is no conclusive evidence regarding the accuracy of imaging techniques for this anomaly. The standard investigation of choice in a case of gall bladder pathology is ultrasonography. However mostly preoperative imaging shows “shrunken or contracted gall bladder and hyperechoic shadows which are mistaken for gallbladder stones” leading to a diagnosis of cholelithiasis. The mistake here leads to operative intervention, which was repeated in our case too.[6,15,18] Malde algorithm published in 2010 suggests further imaging modality according to accuracy is MRCP, CT, ERCP and endoscopic ultrasound according to availability if gall bladder is not visualized properly in sonography.[18] Of all the investigation in cases of doubtful gall bladder, MRCP is the test of choice due to the fact that an ectopic gallbladder as well as anomalies of biliary system can be identified on MRCP.[9,19]

Most of the case reports in this subject state that due to rarity of the incidence and absence of a conclusive preoperative diagnosis, most patients end up undergoing surgical intervention. However, a controversy exists among surgeons with regard to the intra-operative decision once gallbladder is not found in its normal location, whether to convert to laparotomy from laparoscopy or abandon the procedure. A large group of surgeons are of the opinion that conversion to open is unnecessary as laparoscopic view provide a better visualization of all the ectopic sites of gallbladder and a prolonged surgery may be more traumatic to the patient. The abandoned surgery can always be followed up with a MRCP which can establish the diagnosis.[6,15,17,20-22] We also followed this consensus and once thorough examination of all possible location of gallbladder was carried out, procedure was abandoned and a post-operative MRCP was done to confirm the diagnosis of gallbladder agenesis.

CONCLUSION

Gallbladder agenesis is a rare but well recognized congenital anomaly. Low suspicion and a preoperative sonological diagnosis create a diagnostic dilemma and surprise in the intra-operative period. Clinical and radiological parameters mimic cholelithiasis making the patient land up in unwanted surgical procedure. We recommend going ahead with a confirmatory imaging modality, preferably MRCP if the pre-operative sonography is not conclusive in making a diagnosis of gallbladder disease. Radiologist and clinicians keeping this entity in mind, should proceed for a better imaging modality whenever the gallbladder is improperly visualized in routine imaging methods, in patients with or without biliary-type of pain and also in patients with other known abnormalities/agenesis. A conservative approach in managing such patients should be taken up by the treating surgeons.

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