AGENESIS OF GALL BLADDER : A CASE REPORT

ABSTRACT

Background: Agenesis of the gall bladder is a rare condition, majority of which are usually diagnosed during intraoperative procedure because of low incidence of suspicion and high rate of false positive results for cholecystitis on ultrasound imaging.
Methodology:A female, 25 years old patient presented with right upper quadrant pain, whose clinical features and abdominal ultrasound suggested chronic calculous cholecystitis. After pre-operative evaluation and preparation, she was planned for laparoscopic cholecystectomy.
Results: The patient underwent elective operative intervention on 17th June 2023. Intra-operatively, the liver and the extra-hepatic biliary tree were thoroughly inspected, but no gall bladder was detected anywhere in the undersurface of the liver. Decision was made to stop the procedure avoiding any further exploration thereof. Post-operatively the patient was subjected to magnetic resonance cholangiopancreatography (MRCP), which confirmed agenesis of gall bladder.
Conclusion: The accuracy of abdominal ultra-sonography (USG) in the diagnosis of gall bladder disease is not always 100% and a second USG screening or MRCP has got a definite role to play in doubtful cases.

Keywords: Agenesis, Gall bladder, MRCP

1. INTRODUCTION

Developmental abnormalities of the gall bladder are relatively rare. Agenesis of gall bladder is three times more common in females than males.[2,3] While half of the patients are asymptomatic, the other half typically presents with symptoms akin to biliary colic or cholecystitis with symptoms such as dyspepsia, abdominal pain, nausea, vomiting, intolerance to fatty food and so on.[2,4] Most cases are sporadic (around 70%) and there is very little literature on familial links.[1] Since its first description by Lemery and Bergman in 1701 and 1702 respectively, a number of cases have been published, with a reported incidence of 0.01 to 0.06%.[8] combined with the rarity of the condition, the diagnosis is infrequently made preoperatively, and so the patient undergoes unnecessary operative intervention. Intraoperatively the risk of iatrogenic injury is higher, and so the associated morbidity of the procedure is greater.[9] Upper abdominal ultrasound is a first line imaging for the diagnosis of cholelithiasis with a sensitivity of 97%, but in gall bladder agenesis, the sensitivity decreases to 61%,[3,24] and erroneous report of cholelithiasis or shrunken gall bladder suggestive of chronic cholecystitis is often made. Due to these misinterpreted reports patients undergo unnecessary surgery and may encounter iatrogenic biliary tract injuries and portal injuries, due to excessive dissection to find out the absent gall bladder.[12] Sometimes conversion to open exploration needed when an injury to biliary tract is suspected which adds morbidity to the patient. Preoperative magnetic resonance cholangiopancreatography (MRCP) and endoscopic ultrasound (EUS) should be considered. And when such condition is encountered during intraoperatively, intraoperative cholangiography and intraoperative ultrasound can be done to rule out agenesis and ectopic gall bladder.[13]

2. CASE REPORT

A 25-year-old woman presented to the hospital with history of right upper abdominal pain with a similar kind of episode on and off since months. She was otherwise fit and healthy with no other medical or surgical history. In the out

patientdepartment (OPD), she was haemodynamically normal, and she had right upper quadrant abdominal tenderness. Her blood tests show normal liver function tests (LFTs). She had an ultrasonography of the whole abdomen. The ultrasonography gave an impression of a contracted gallbladder. There was no definite wall hyperaemia and probe tenderness was present. She was diagnosed with acute on chronic cholecystitis and antibiotics were commenced. Symptoms improved so she was subsequently discharged with plan for an elective laparoscopic cholecystectomy in the next 4 weeks. Intraoperatively, there was no gallbladder seen. The examination of right and left subhepatic space, and portal region was done. The anatomy looked unremarkable other than the absence of a gallbladder. Another surgeon was called into theatre for a second opinion who agreed with the findings or lack thereof and the procedure was converted to open cholecystectomy. Even after opening the abdomen, no gall bladder was seen and hence the intended operation was stopped. The patient was discharged home on recovery and planned for outpatient MRCP and repeat LFT. On MRCP no gallbladder or cystic duct was identified and was consistent with congenital agenesis. Common bile duct (CBD) was normal with no evidence of biliary stricture or choledocholithiasis. The repeat LFT was normal. She was reviewed in outpatients following these investigations. She remained well postoperatively with no further symptoms. She was discharged with surgical follow-up.

3. DISCUSSION

The liver and gall bladder develop in the fourth week of intrauterine life.[6] The gall bladder originates from the cystic bud as a ventral outgrowth from the caudal region of the foregut. The vacuolation of hyperplastic epithelium starts at about the seventh week during which the gall bladder and cystic duct develop a lumen.[7] Though the exact pathogenesis is unknown, gall bladder agenesis is thought to be due to the failure of the cystic bud to develop further or to the failure of vacuolation.[6]
Gall bladder agenesis is a rare congenital anomaly characterized by the absence of the gall bladder in conjunction with a normal bile duct system.[10] The prevalence range is 0.007 to 0.13 % with an incidence of 0.007 to 0.027% in surgical cases and 0.04 to 0.13% in autopsy reports.[10]
Agenesis of gall bladder is explained by two developmental theories:[14,15]

1. Failure of hepatic diverticula to develop into gall bladder
2. Failure of recanalization of cystic duct and gall bladder.

Agenesis of gall bladder may present as:[16]

1. Neonates with multiple fetal anomalies (15 – 16%): in these patients, agenesis usually diagnosed on autopsy because of death in perinatal period due to associated GIT, GUT, CVS anomalies.
2. Asymptomatic (35%): in these patients, agenesis recognized at autopsy and during laparotomy for other cause.
3. Symptomatic (40 – 60%): In these patients agenesis is discovered on USG abdomen, MRCP, EUS, and during laparoscopy for evaluation of (colicky) pain in right hypochondrium (90%), dyspepsia, vomiting.

Agenesis of gall bladder is associated with congenital syndromes such as cerebrotendinousxanthomatosis, G-syndrome, Klippel-Feil syndrome, trisomy 18 and some cases reported after thalidomide therapy.[17-20]

When a patient presents with symptoms of biliary colic or upper abdominal pain, the first investigation of choice would be an abdominal ultrasound. In order to allow the gall bladder to distend and to reduce the amount of gas present in the bowel and assist in distinguishing bowel loops from an absent gallbladder, the ultrasound should be performed after fasting. The “WES” triad (wall-echo-shadow sign - demonstration of gallbladder Wall, Echo of stone, and acoustic Shadow) has been proposed to differentiate between a contracted gallbladder with gallstones and bowel loops.[5,7]

MRCP and Endoscopic Retrograde Cholangiopancreatography (ERCP) provide excellent alternatives to open exploration and intra-operative cholangiography.[11] Though ERCP contributes little in diagnosis of agenesis because non-visualization of gall bladder is interpreted as cystic duct obstruction.[22] If the diagnosis is made pre-operatively or the symptoms continue postoperatively, a possible conservative treatment for this symptomatic group includes smooth muscle relaxants, and if this fails, sphincterotomy.[11]
Preoperative MRCP should be considered in cases of USG diagnosis of non-visualization of gallbladder.[23] Other diagnostic modality includes EUS, intra-op ultrasound and selective arteriography can be used for agenesis. But their availability is less.[22] There were some cases reported in which AGB was diagnosed preoperatively and the operation was avoided.[21]

4. CONCLUSION

When non-visualization of gallbladder is present during laparoscopy or open exploration intra-operative cholangiogram, intraoperative ultrasound and postoperative MRCP or endoscopic ultrasonography (EUS) can help in the diagnosis of agenesis or ectopic gallbladder.

CONSENT

As per university standard patient written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

Takenfrominstitutionalethicscommittee.

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