

Agnesis of gallbladder: A diagnostic dilemma

**Atul Kumar Mittal, Dhananjay Saxena, Raju Kadam, Prashant Garg,
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ABSTRACT

Introduction: Agnesis of gallbladder is a rare (13–65 cases/100,000) anomaly, in which about 23% patient presents with symptoms of biliary disease. In these patients, ultrasonography (USG) abdomen frequently falsely reveals shrunk or contracted gallbladder and sometimes non- visualization of gallbladder (GB) in GB fossa. Basis of these misinterpreted reports these patients undergone unnecessary surgery and may encounter iatrogenic biliary tract injuries and portal injuries because of excessive dissection to find out the absent gallbladder and ectopic gallbladder.

Case Report: A 40-year-old female attended surgical outdoor with complain of pain right hypochondrium and dyspepsia since last four years patient followed-up with USG abdomen which was suggestive of chronic cholecystitis with cholelithiasis, and common bile duct (CBD) was normal in diameter on the basis of clinical symptoms and USG findings patient admitted and planned for laparoscopic cholecystectomy. On laparoscopy after removing the adhesions, exploration done up to the porta but gallbladder could not be visualized.

On postoperative day-1 patient was sent for MRCP with MRI abdomen. On MRCP gallbladder and cystic duct were not visualized. CBD was normal in caliber. Liver and pancreas were normal. Hence the diagnosis of agnesis was made.

Conclusion: Agnesis of gallbladder is an unusual anomaly in which about 23% presents as biliary disease. These patient frequently undergone surgery because of misinterpreted reports of USG abdomen, ERCP and CT abdomen. So, in cases with ultrasonographic diagnosis of scleroatrophic or non-visualization or suspicious of ectopic gallbladder and absence of wall echo shadow (WES) triad or double arc shadow, when non-visualization of gallbladder is present during laparoscopy or open exploration intraoperative cholangiogram, intraoperative ultrasound and postoperative MRCP or Endoscopic ultrasound (EUS) can help in the diagnosis of agnesis or ectopic gallbladder.



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CASE REPORT

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ABSTRACT

Introduction: Agnesis of gallbladder is a rare (13–65 cases/100,000) anomaly, in which about 23% patient presents with symptoms of biliary disease. In these patients, ultrasonography (USG) abdomen frequently falsely reveals shrunken or contracted gallbladder and sometimes non-visualization of gallbladder (GB) in GB fossa. Basis of these misinterpreted reports these patients undergone unnecessary surgery and may encounter iatrogenic biliary tract injuries and portal injuries because of excessive dissection to find out the absent gallbladder and ectopic gallbladder. **Case Report:** A 40-year-old female attended surgical outdoor with complain of pain right hypochondrium and dyspepsia since last four years patient followed-up with USG abdomen which was suggestive of chronic cholecystitis with cholelithiasis, and common bile duct (CBD) was normal in diameter on the basis of clinical symptoms and USG findings patient admitted and planned for laparoscopic

cholecystectomy. On laparoscopy after removing the adhesions, exploration done up to the porta but gallbladder could not be visualized.

On postoperative day-1 patient was sent for MRCP with MRI abdomen. On MRCP gallbladder and cystic duct were not visualized. CBD was normal in caliber. Liver and pancreas were normal. Hence the diagnosis of agnesis was made. **Conclusion:** Agnesis of gallbladder is an unusual anomaly in which about 23% presents as biliary disease. These patient frequently undergone surgery because of misinterpreted reports of USG abdomen, ERCP and CT abdomen. So, in cases with ultrasonographic diagnosis of scleroatrophic or non-visualization or suspicious of ectopic gallbladder and absence of wall echo shadow (WES) triad or double arc shadow, when non-visualization of gallbladder is present during laparoscopy or open exploration intraoperative cholangiogram, intraoperative ultrasound and postoperative MRCP or Endoscopic ultrasound (EUS) can help in the diagnosis of agnesis or ectopic gallbladder.

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INTRODUCTION

Agnesis of gallbladder is a rare (13–65 cases/100,000) anomaly, about 23% patient presents with symptoms of

biliary disease [1, 2]. In these patients, ultrasonography (USG) abdomen frequently falsely reveals shrunk or contracted gallbladder and sometimes non-visualization of gallbladder in GB fossa [3]. Due to these misinterpreted reports, patients undergone unnecessary surgery, and may encounter iatrogenic biliary tract injuries and portal injuries, due to excessive dissection to find out the absent gallbladder and ectopic gallbladder [4]. Sometimes conversion to open exploration needed when an injury to biliary tract is suspected which adds morbidity to the patient. Preoperative imaging like MRCP and EUS should be considered [5]. And when such condition is encountered during intraoperatively, intraoperative cholangiography and intraoperative ultrasound can be done to rule-out agenesis and ectopic gallbladder [5].

CASE REPORT

A 40-year-old female attended surgical outdoor with complains of pain right hypochondrium and dyspepsia since last four years. Patient evaluated with USG abdomen which was suggestive of chronic cholecystitis with cholelithiasis, and CBD was normal in diameter. LFT's were within normal limit. On the basis of clinical symptoms and USG findings patient admitted and planned for laparoscopic cholecystectomy.

On laparoscopy findings were:

1. Omentum and colonic loops were densely adherent to the inferior surface of liver (Figures 1A–B).
2. After removing the adhesions, exploration done up to the porta but gallbladder could not be visualized (Figure 2A–B).
3. Further exploration was done to rule out the ectopic gallbladder but gallbladder could not be visualized at those ectopic sites also.
4. CBD was normal in diameter.
5. Procedure terminated at this stage and decided to do postoperative MRCP in spite open conversion to prevent iatrogenic bile duct injuries.

On postoperative day-1 the patient was sent for MRCP with MRI abdomen. In MRCP findings, gallbladder and cystic duct were not visualized. CBD was normal in caliber. Liver and pancreas were normal (Figures 3A–B). Hence the diagnosis of agenesis was made.

In postoperative period patient sent for ERCP sphincterotomy. Patient followed-up for three months and she was comfortable without any episode of pain and dyspepsia. Followed up last month patient is doing well. final diagnosis was agenesis of gall bladder.

DISCUSSION

Agenesis of gallbladder was first reported by Lemery and Bergman in 1701 and 1702 respectively [2]. The

incidence of agenesis in surgical cases is (0.007–0.027%) and in autopsy reports (0.04–0.13%) [6, 7]. Gallbladder develop late in first month of intrauterine life from distal part of hepatic diverticular bud of the foregut. Agenesis of gallbladder is explained by two developmental theories [8, 9]:

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

Agenesis of gallbladder may present as [10]:

- (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 patient agenesis discovered on USG abdomen, MRCP, EUS and

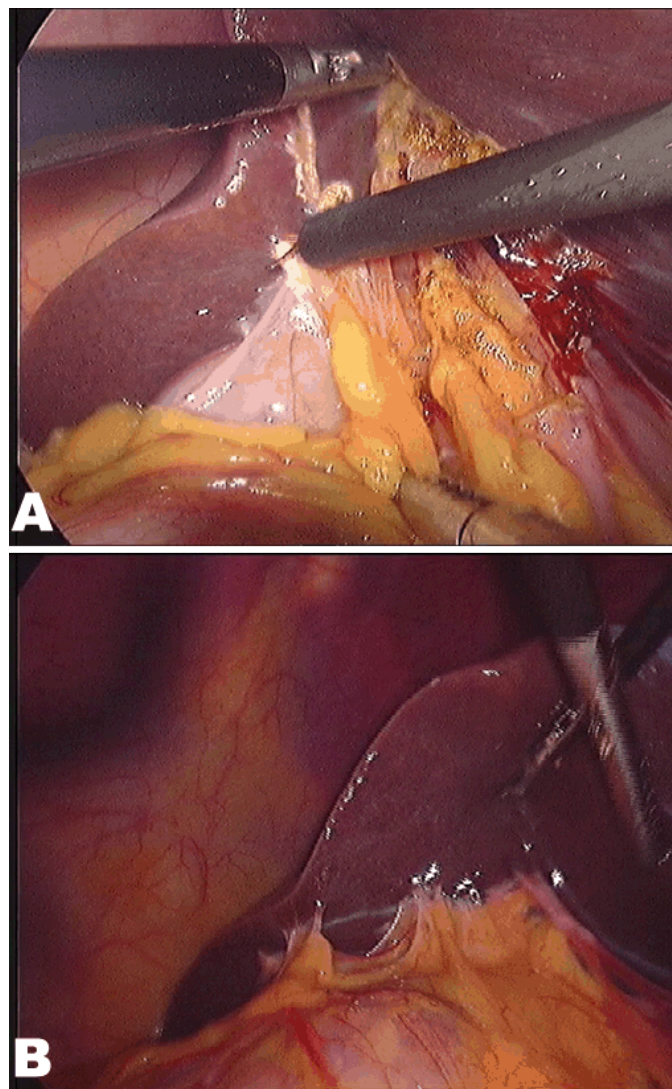


Figure 1 (A, B): Omentum and colonic loop adherent to inferior surface of liver.

during laparoscopy for evaluation of (colicky) pain right hypochondrium (90%), dyspepsia, vomiting.

Cause of pain in symptomatic patients includes biliary dyskinesia, adhesion in the GB fossa or periportal adhesions, remnant cystic duct stone and choledocholithiasis. ERCP sphincterotomy and adhesiolysis resolved the pain in these patients [11]. Agenesis of gall bladder is associated with congenital syndromes cerebrotendinous xanthomatosis, G-syndrome, Klippel–Feil syndrome, trisomy 18 and some cases reported after thalidomide therapy [12–15]. On laparoscopy if gallbladder is not visualize in GB fossa, dissection should be carried out up to the porta and usual sites for ectopic gallbladder, which are intrahepatic, retrohepatic, left sided, or within the falciform or lesser omentum, to prove the agenesis of gallbladder [8]. But sometimes this dissection is lead to major biliary tract injuries. So, excessive dissection and open exploration is avoided. On USG and CT abdomen diagnosis of agenesis is limited by bowel gas artifacts due to adhesions at liver bed which makes shadowy opacities, periportal tissue,

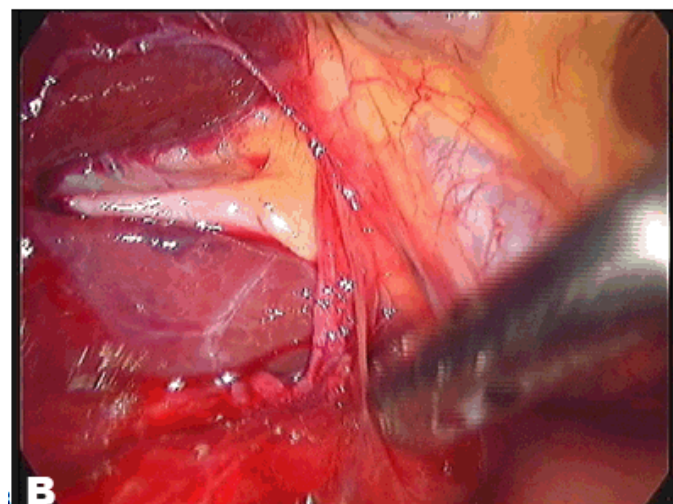
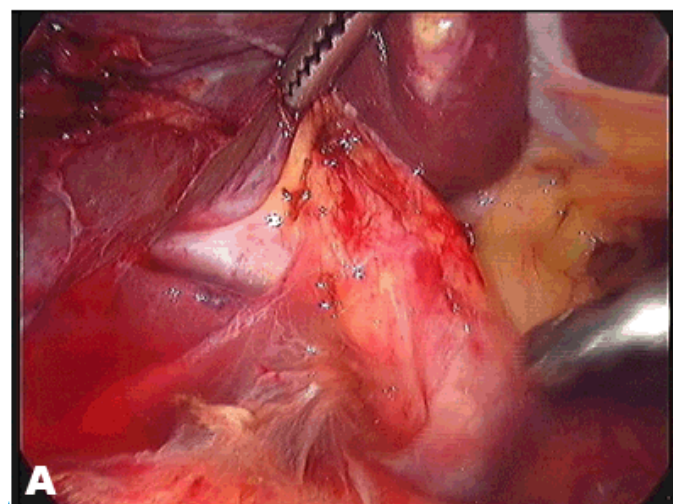
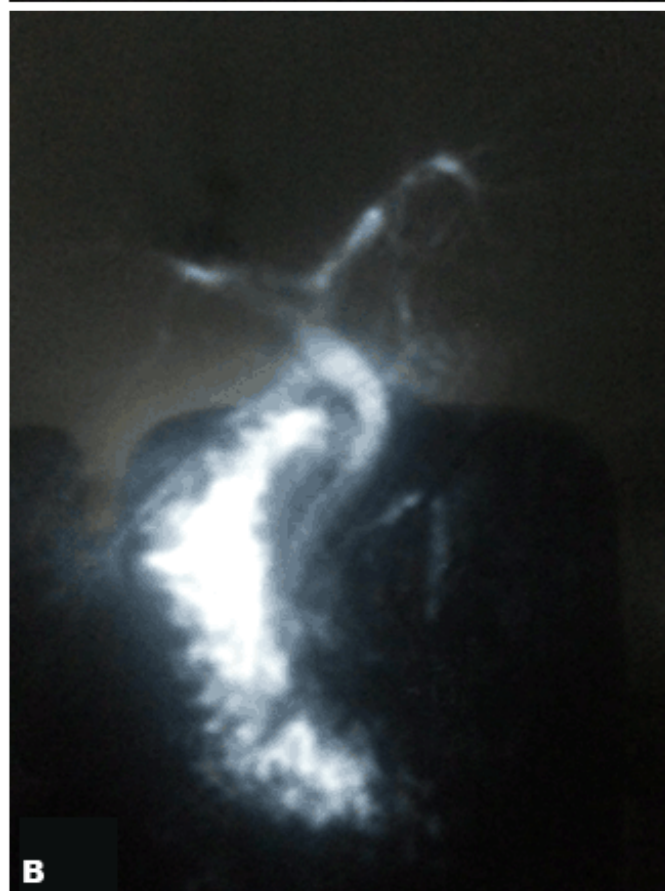
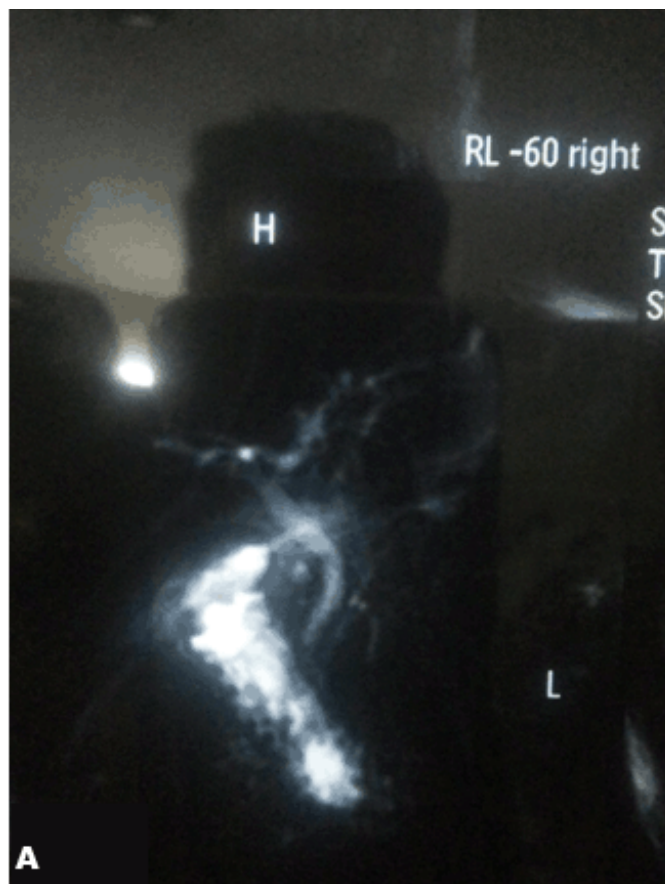


Figure 2: (A, B) CBD dissected up to the porta hepatis. Both right and left hepatic duct seen, but gallbladder and cystic duct are not visualized.

Figure 3: (A, B) MRCP films showing normal CBD and absence of cystic duct and gallbladder.

lipoma, liver hemangioma or migrated liver tissue [10, 16–18]. ERCP contributes little in diagnosis of agenesis because nonvisualization of gallbladder is interpreted as cystic duct obstruction [19].

MRCP is non-invasive and best method to delineate intrahepatic and extrahepatic biliary tract. Preoperative MRCP should be considered in cases of USG diagnosis of non-visualization of gallbladder [20]. Other diagnostic modality includes EUS, intra-op ultrasound and selective arteriography can be used for agenesis. But their availability is less [19]. There were some cases reported in which AGB was diagnosed preoperatively and the operation was avoided [16].

Agenesis of gallbladder is an unusual anomaly of which about 23% presents with biliary disease [2]. It is sometimes associated with anomalies of other system also and seems to be familiar inheritance [21]. These patient frequently undergo surgery because of misinterpreted reports of USG abdomen, ERCP and CT abdomen. So, in cases with ultrasonographic diagnosis of scleroatrophic or nonvisualization or suspicious of ectopic gallbladder and absence of WES triad or double arc shadow, Further preoperative investigation like MRCP and EUS should be done to rule out agenesis and ectopic gallbladder to avoid unnecessary surgery and iatrogenic biliary injuries.

CONCLUSION

When nonvisualization 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.

Author Contributions

Atul Kumar Mittal – Substantial contributions to conception and design, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Dhananjay Saxena – Substantial contributions to conception and design, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Raju Kadam – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Prashant Garg – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published.

Jeevan Kankaria – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published.

Rajkamal Jenaw – Substantial contributions to conception and design, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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