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General Surgery

# GALLBLADDER AGENESIS – A RARE CASE REPORT

**KEY WORDS:** gall bladder agenesis, biliary colic, cholecystitis, choledocholithiasis

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**ABSTRACT**

Gallbladder agenesis is a rare congenital entity. The incidence is around 1 per 6500 live births. Around 50-70% patients are asymptomatic and the rest are symptomatic with symptoms mimicking biliary colic. Right upper quadrant ultrasound (US) is usually either misleading or inconclusive. Also advanced diagnostic studies such as hepatobiliary iminodiacetic acid (HIDA) scan and endoscopic retrograde cholangio-pancreatography (ERCP) may show non-visualization of the gallbladder and erroneously lead providers to a diagnosis of cystic duct obstruction rather than Gallbladder Agenesis. So some patients are only finally diagnosed intraoperatively. Surgery in these patients becomes risky because unnecessary dissection while looking for the non-existent gallbladder can result in injury of the biliary tree, hepatic vasculature, or small bowel. Therefore, clinicians should keep Gallbladder Agenesis on their differential diagnosis list and imaging modalities such as magnetic resonance cholangiopancreatography (MRCP) should be obtained when other tests are inconclusive. We report a 48-year-old female presenting with chronic symptoms consistent with biliary colic and an equivocal US reported as cholelithiasis. She underwent laparoscopy during which the absence of the gallbladder was noted.

## INTRODUCTION

The absence of the gallbladder, also known as gall-bladder agenesis (GA), is a very rare anatomic anomaly. The incidence has been reported to be of less than 1 per 6500 live births [1]. Most patients suffering from GA remain asymptomatic lifelong.

Hence, the majority of the cases are found either at autopsy or incidentally while undergoing diagnostic imaging or surgery. However, about 23 to 50 % [2,3] of patients can present with symptoms of right upper quadrant pain that can be mistaken for cholecystitis or symptomatic cholelithiasis.

Routine preoperative ultrasound (US) can be misleading and patients can be subject to unnecessary surgical procedures. We report a case of GA in a 48-year-old female who presented with symptoms consistent with symptomatic cholelithiasis and an equivocal US, whose gallbladder was absent on surgical laparoscopy. Postoperative MRCP confirmed the diagnosis of GA.

### Case Presentation

A 48-year-old female patient presented to OPD with abdominal pain for 10-12 months which was intermittent, colicky involving right upper quadrant. The pain usually exacerbated after food intake and fasts for about an hour. It was not associated with fever, chills, nausea, vomiting, jaundice, and changes in the color of her stool and urine. She denied any alcohol intake or any substance abuse. Her physical examination was only remarkable for obesity. There was no icterus.

On examination, there was no tenderness in the right upper quadrant and no palpable organomegaly, hernia, or mass. Laboratory work up which included a complete blood count, comprehensive metabolic panel, and liver function tests was done, all of which were within normal limits. An abdominal ultrasound did not demonstrate a well-delineated gallbladder.

There was increased echogenicity on the proximal edge of the gallbladder fossa compatible with a Wall Echo Shadow (WES) sign suggestive of either large calcified or multiple small calcified gallstones in the gallbladder. Murphy's sign on ultrasound was negative and the common bile duct measured around 6 mm.

Based on the clinical history and the ultrasound findings, the diagnosis was presumed to be symptomatic gall stones and the patient was taken to laparoscopic cholecystectomy. The abdominal cavity was entered by using standard laparoscopic technique, but the gallbladder was not appreciated. The right lobe of the liver was elevated but the gallbladder was not visualized.

Additional dissection was performed in order to clear the liver plate but still no gallbladder was seen (Figure 1). No surgical clips or signs of prior surgery in the right upper quadrant were identified. Close inspection of both lobes of the liver as well as the falciform ligament was performed but no aberrant location of the gallbladder was identified. Intra-operative Indocyanine green induced gamma probe studies also couldn't identify gall bladder. At that point the procedure was terminated.

A magnetic resonance cholangio pancreatography (MRCP) was performed to evaluate the patient's anatomy in an outpatient setting that confirmed the absence of the gallbladder. The patient was discharged home and recovered as expected. She continues to experience intermittent right upper quadrant pain similar to that which she experienced prior to surgery.

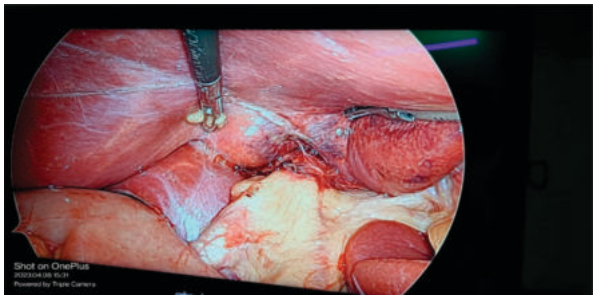

**Figure 1: Laparoscopic picture showing inferior surface of liver with no gall bladder.**

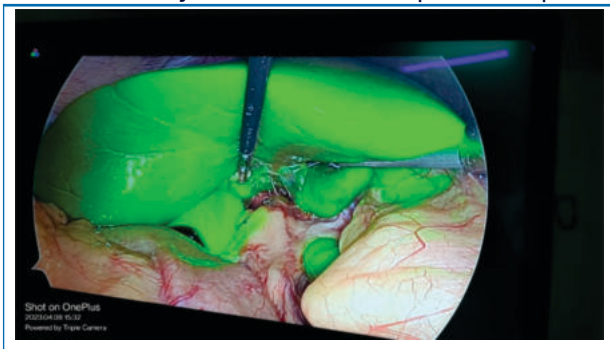


**Figure 2: Indocyanine green induced gamma probe enabled laparoscopic picture showing inferior surface of liver with absent gall bladder.**

## DISCUSSION

Gallbladder agenesis is a rare congenital entity. The prevalence ranges from 0.007 to 0.013 percent [4]. The autopsy reports have not shown a difference in frequency between females and males, but the incidental cases of GA found during surgical procedures have been reported to have a 3:1 female predominance [5]. Gallbladder agenesis was first reported in the medical literature in 1702 by Bergman [6]. Gall bladder and cystic duct arises from the caudal bud of hepatic diverticulum along with the ventral pancreas at fourth week of intra uterine life [7]. Although the exact pathogenesis of GA is unknown, there are two main hypotheses. The first hypothesis postulates that the superior division of the caudal bud of the hepatic diverticulum fails to develop into the cystic duct and gallbladder, possibly due to an aberration in the development of the vessels surrounding the caudal bud [8]. Consequently, GA is frequently found in patients with other gastrointestinal and cardiovascular abnormalities [9]. The second theory states that GA occurs due to a failure of recanalization of the gallbladder and cystic duct and in these cases are noted without any association [5].

The presentation of GA is varied. In 1988, Bennion et al. classified GA patients into three groups, according to their presentation. The first group patients are those with associated other congenital anomalies. These patients usually die secondary to those anomalies rather than GA. The second group are the patients in whom GA is found in postmortem and they were asymptomatic in their entire life. A third group are the patients presenting with gastrointestinal symptoms which resulted in the intraoperative diagnosis of GA [1]. Amongst patients with GA, 50 to 70 percent are asymptomatic [2,3]. The remaining patients usually present with symptoms similar to acute cholecystitis or biliary colic like chronic right hypochondriac pain, dyspepsia, jaundice and intolerance to food [10]. These symptoms might be due to associated sphincter of oddi dysfunction which can be explained by the common origin of gall bladder and the sphincter. Sphincter of Oddi dysfunction may predispose to biliary stasis resulting in patient jaundice, elevated liver functions tests, and even common bile duct stones [11].

Diagnosing GA before patients undergo unnecessary surgery can be challenging. In fact, a significant proportion of patients with GA are only diagnosed intraoperatively [9,12-14]. Surgery in this patient population carries risk. When the gall bladder is not found in its normal anatomical position intra operatively especially when there was a high suspicion of biliary disease, other areas should be explored. The falciform ligament, the leaves of the lesser omentum, intrahepatic and retrohepatic locations must be inspected [4]. This might cause damage to the liver, biliary tree, small bowel, and hepatic vessels. Intraoperative ultrasound and cholangiography can be done to localize the ectopic gall bladder. Though US is the initial investigation done to evaluate a biliary disease, in these cases its often misleading [15]. GA is usually misdiagnosed as contracted gall bladder in US [16]. Hepatobiliary iminodiacetic acid (HIDA) scan and endoscopic retrograde cholangio-pancreatography (ERCP) can also be misleading in the setting of GA [17,18]. In these methods, it is interpreted as obstruction of the cystic duct which is consistent with a diagnosis of cholecystitis, rather than as absence of the gallbladder [12]. MRCP can diagnose patients with GA prior to surgery [5,19]. Because it does not rely on the passage of contrast for visualization of the biliary tree, it is not affected by biliary stasis. Therefore, MRCP should be considered when the diagnosis is uncertain.

## CONCLUSIONS

Gallbladder agenesis is a rare condition which can present with symptoms mimicking of cholecystitis and symptomatic cholelithiasis. As the surgery carries risks and has no benefits, it is important to keep gall bladder agenesis on their list of differential diagnoses, especially when the gallbladder is not clearly delineated on pre-operative US. In patients where pre-operative US imaging is equivocal, an MRCP should be obtained prior to proceeding to the OR for exclusion of GA.

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