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Agenesis of the gallbladder as a rare misdiagnosis

Case Report

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Abstract: Anatomic anomalies of the biliary tract are not uncommon, but gallbladder and cystic duct agenesis is rare, with an incidence of 0.01% to 0.04% and a frequency of 0.016% at autopsy. It is usually asymptomatic and discovered incidentally. Although this congenital anomaly is infrequent, it may be encountered in clinical practice; thus, the surgeon should be aware of the associated problems. A correct preoperative diagnosis of this congenital anomaly is difficult to establish because of the nonspecific nature of the symptoms and the relative inaccuracy of the currently available diagnostic tests. Here we report a patient with a preoperative false diagnosis of cholelithiasis that was found on laparoscopy and open surgery to be agenesis of the gallbladder.

Keywords: *Aganesis • Gallblader • Cystic duct*

1. Introduction

Gallbladder agenesis is an extremely rare embryologic abnormality and is often encountered during cholecystectomy [1-4]. Even if biliary agenesis is diagnosed during surgery, the surgeon may not be aware of the accompanying pathologic conditions, and a delay in the proper patient guidance may result. Biliary agenesis could be misdiagnosed radiologically as cystic duct obstruction or acute cholecystitis or chronic cholecystitis in patients in whom the gallbladder is not seen during ultrasonographic examination, since shrunken and scarred gallbladder tissue is difficult to visualize in chronic inflammatory conditions [4,5]. In this report, we describe such a case of gallbladder agenesis that was diagnosed both at surgery and during post-surgical MRI scanning after a prior false diagnosis of cholelithiasis and chronic cholecystitis.

2. Case report

A 35-year-old woman was admitted to our department having had intermittent upper abdominal pain for 2 months. Her blood pressure was 160/80 mm Hg, and her body temperature was 36.5ºC. Her abdomen was soft and flat, with tenderness in the upper quadrants. Hematologic laboratory, blood chemistry, and renal function tests were unremarkable. Serum levels of aspartate aminotransferase, alanine aminotransferase, total bilirubin, g-glutamyltransferase, and alkaline phosphatase, which indicate liver function, were not elevated. An abdominal ultrasound showed an area with hyperechoic boundaries, which was interpreted as an inflamed gallbladder with thick, irregular walls and lithiasis, and she was referred for laparoscopic cholecystectomy. Surgery was performed, but with the laparoscopic approach, we failed to detect the gallbladder in its anatomic position, and the anatomy of the biliary tract was also unclear (Fig. 1B). Despite a thorough intraoperative ultrasonographic examination of the intrahepatic (left-sided areas within the lesser omentum), retroperitoneal, retrohepatic (within the falciform ligament), retroduodenal, and retropancreatic areas, the gallbladder was not found. We then decided to convert the procedure to an open laparotomy, but the gallbladder was again not detected in any of the possible ectopic positions. A white scar extended through the surface of the right lobe of the liver (Fig. 1A). The patient was diagnosed as having agenesis of the gallbladder. The operation was completed without resection. Her postoperative course was uneventful. Magnetic

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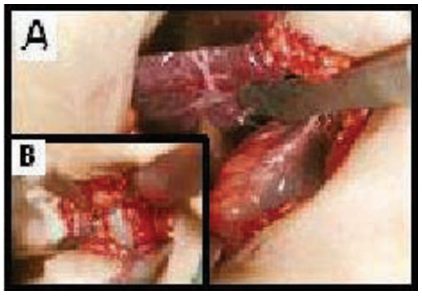
Figure 1. A) Intraoperative view of gallbladder bed; B) Intraoperative view of choledoc.



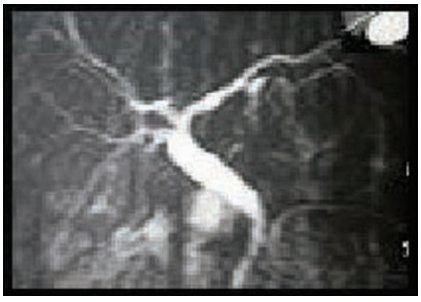
resonance cholangiopancreatography revealed dilated extrahepatic ducts without stones in the common bile duct, thus providing concrete evidence of gallbladder agenesis (Fig. 2).

3. Discussion

Agenesis of the gallbladder is a rare anomaly. Theories for this condition include a failure of the development of the gallbladder anlage from the hepatic diverticulum, failure of recanalization, and breakdown of the suction field. It has been suggested that this condition represents an inheritable congenital developmental defect [6]. Gallbladder agenesis is usually asymptomatic, but 35% to 60% of the patients may have symptoms compatible with a biliary disorder like cholelithiasis [4,5]. However, it has been reported that patients with gallbladder agenesis will have at least one symptom suggestive of biliary tract disease during their lifetime, and bile duct stones will be found in 25% to 60% of these cases [6]. The symptoms and the increased frequency of gallstones has been attributed by some authors to biliary dyskinesia [7] and compared by others to a postcholecystectomy syndrome [8,9]. Choledocholithiasis has been reported to occur in approximately 18% to 50% of patients with gallbladder agenesis and invariably presents as jaundice [10]. In confirming the diagnosis of agenesis of the gallbladder, it is necessary to exclude abnormal locations of the organ, which include intrahepatic, retrohepatic, left upper quadrant sites or sites within the lesser omentum or falciform ligament and retroperitoneum.

The preoperative diagnosis is often difficult, especially if a dysplastic cyst is associated with the condition, because it simulates the gallbladder. Di Vita et al. reported agenesis of the gallbladder associated with lithiasis of the common bile duct [11]. This rare congenital malformation is frequently associated with other anomalies. Wilson and Deitrick suggested that absence of the gallbladder may be a familial trait [12]. Ultrasonography now makes it possible to detect the absence of the

Figure 2. Postoperative MRI view of the case.



organ in individuals without gallbladder disease.

The great difficulty in visualizing a contracted gallbladder with stones is well known [6]. According to Hammond *et al.* [13], there is always either a recognizable segment of wall or a thin rim of bile identifying the gallbladder. Those authors based that assertion on the reports of MacDonald *et al.* [14], and Raptopoulos *et al.* [15], which described the WES triad (demonstration of the gallbladder Wall, the Echo of the stone, and the acoustic Shadow) or the double arc shadow (two parallel, arcuate, echogenic lines separated by a thin anechoic space with distal acoustic shadowing). However, both the conditions of the examination and the examiner's experience may not always permit accurate appreciation of these features. Shadowy opacities misdiagnosed as stones can be due to intestinal gas artifacts or to other structures in close proximity, such as a calcified hepatic lesion or a surgical clip. Sarli *et al.* [3] recommend preoperative cholangiography and laparoscopic exploration followed by laparoscopic sonography as adequate modalities to diagnose gallbladder agenesis.

Magnetic resonance cholangiography (MRCP) is also a noninvasive and well demonstrated imaging method for the evaluation of the biliary tract [10]. Since contrast administration is not required to visualize the bile duct, MRCP is not compromised by biliary stasis and can thus demonstrate an absent or an ectopic gallbladder.

Because it is difficult to diagnose gallbladder and cystic duct agenesis preoperatively and this anomaly is almost always incidentally discovered at surgery, laparoscopy may be useless as well as risky. Gotohda *et al.* [5] stated that if the gallbladder is not visualized by imaging techniques, laparoscopy should be performed before laparotomy. However, laparotomy can be avoided and the risk of any possible complications reduced by the laparoscopic surgeon's awareness of the problems posed by this anomaly and a careful review of currently available diagnostic tests [16].

In conclusion, it is extremely difficult to make a correct diagnosis of gallbladder agenesis preoperatively. Thus, in a case with suspicious symptoms, the surgeon should

closely consult with the radiologist to determine whether the bladder stones are clearly visible in the presence of a thickened biliary wall or whether a sclerotic band is assumed to be associated with biliary stone disease. If there is any uncertainty, further radiologic tests should be promptly performed to avoid unnecessary surgery. Should this opportunity be missed, the surgeon should at least check for possible ectopic positions of the gallbladder and should offer the patient guidance regarding the likely accompanying medical anomalies.

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