International Surgery Journal
Ramos BG et al. Int Surg J. 2024 Aug;11(8):1347-1350
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: https://dx.doi.org/10.18203/2349-2902.isj20242130

Case Report

Gallbladder agenesis associated with dextrocardia in an adult patient: case report

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Received: 23 June 2024
Accepted: 17 July 2024

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ABSTRACT

The aim was to disseminate information about gallbladder agenesis and its association with some congenital malformations, in this case, with dextrocardia. The case report of a patient with dextrocardia and diagnosis of cholecystolithiasis (by ultrasound) who presented with agenesis of the gallbladder is presented. The patient was scheduled for laparoscopic cholecystectomy, however, during the surgical procedure, the absence of a gallbladder was evident. Gallbladder agenesis is a rare anatomical variant and its association with cardiac malformations in adult patients is uncommon.

Keywords: Gallbladder agenesis, Dextrocardia, Cholecystolithiasis

INTRODUCTION

Agenesis of the gallbladder is the rarest common congenital malformation of the bile duct and can affect up to 0.007-0.9% of patients undergoing cholecystectomy.[1,2] To date, very few cases have been reported worldwide (less than approximately 500).[3] It is thought to result from an interruption in the embryonic development of the liver.[4] Patients generally present with symptoms similar to those of biliary colic or biliary pathology, and the diagnosis is made as an incidental finding during surgery.

This condition is a diagnostic challenge due to the vague symptoms and non-specific ultrasound findings in the preoperative period, hence the importance of its study with the aim of disseminating important aspects to expand the knowledge about this disease.

CASE REPORT

A 51-year-old woman, with a history of dextrocardia, reported recurrent episodes of right upper quadrant abdominal pain, nausea, and occasional vomiting requiring multiple hospitalizations for analgesic treatment (Figure 1). Laboratory studies were ordered and were within normal parameters. Ultrasound of the liver and bile ducts revealed a partially assessable small gallbladder with a calcified wall projecting a posterior acoustic shadow, intrahepatic bile ducts without dilatation, and a 3 mm common bile duct. A diagnosis of a probable cholecystolithiasis with a scleroatrophic vesicle was made and a laparoscopic cholecystectomy was scheduled (Figure 2). The surgery failed to identify the gallbladder or any area of fibrosis in the gallbladder bed. A systematic laparoscopy in search of an ectopic site was perfomed without success. Therefore, it was deciding to stop the procedure to avoid disruption of the bile duct or adjacent organs (Figure 3). An additional study was

requested (magnetic resonance cholangiography), which showed the absence of a gallbladder was identified, confirming the suspected diagnosis (Figure 4).

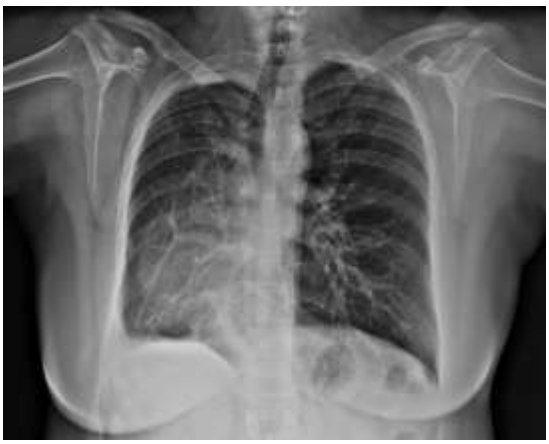

Figure 1: Anteroposterior chest X-ray where dextrocardia is identified.

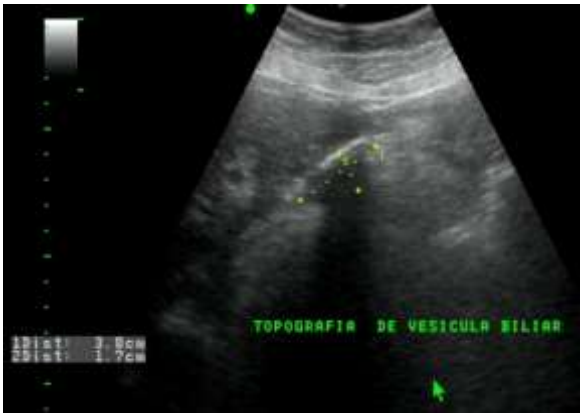

Figure 2: Preoperative ultrasound image showing an image of 3.8×1.7 cm in the topography of the gallbladder, with posterior acoustic shadow interpreted as a probable scleroatrophic gallbladder.

Figure 3: Laparoscopic image showing the hepatic bed with the absence of the gallbladder, identifying only the common hepatic duct that goes directly to the liver.

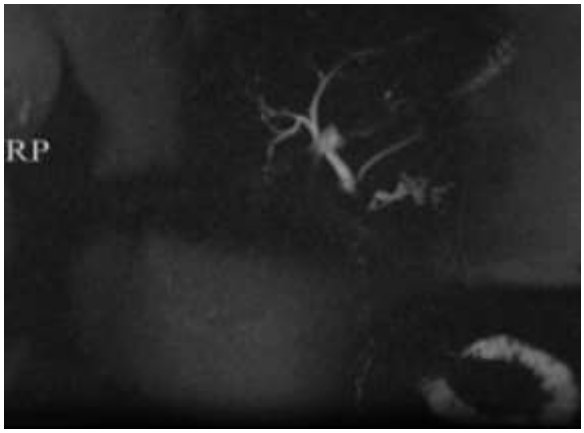

Figure 4: Magnetic resonance cholangiopancreatography where the absence of the gallbladder is identified, confirming the diagnosis of gallbladder agenesis.

DISCUSSION

Vesicular agenesis was first described by Lemery in 1701 and by Bergman in 1703.[5,6] Since then, only nearly 400 cases have been published in the medical literature.[7] It is defined as a congenital anomaly characterized by the absence of the gallbladder without atresia of the extrahepatic bile duct.[3]

The incidence ranges from 0.007-0.009%, is more common in females than males by a ratio of 3:1 (2.8), and generally occurs in third or fourth decade of life. It is estimated that approximately 15-30% of patients with gallbladder agenesis have other organs with malformations.

The etiology is still unknown, however, the occurrence has been observed in several family members, suggesting certain genetic factors, but family predisposition is only determined when this pathology occurs in more than two generations.[7,8]

The embryonic development of the liver and gallbladder begins in the third week of gestation with the appearance of the hepatic diverticulum, which, as it grows, connects to the intestine to form the extrahepatic biliary tract, followed by a ventral invagination occurs that will give rise to the gallbladder around the seventh gestational week.[9,10] Any interruption before the gallbladder has formed will culminate in agenesis of the gallbladder and cystic duct.[4,7,11]

Associations with other congenital defects have been described in up to 40-65%, such as trisomy 18, cerebrotendinous xanthomatosis, abnormalities in different organs (imperforate anus, tracheoesophageal fistula), cardiopulmonary, and even genitourinary defects, which are rare in adult patients.[8]

The clinical picture is the same as in those patients with cholelithiasis or cholecystitis, with the presence of pain in the right upper quadrant (90%), nausea or vomiting (66%), oral intolerance (37%) and jaundice (35%), meteorism (29%), dyspepsia (28%), fever (27%), weight loss (26%) and anorexia (16%).[8] The explanation for the symptoms in this pathology is secondary to a dysfunction of the sphincter of Oddi, which may favor biliary ectasia and stone formation in the common bile duct causing biliary dyskinesia with painful spasms.[4]

It has been described that 25-50% of patients with this pathology have choledocholithiasis, secondary to biliary stasis.[12,13] When jaundice occurs, it is usually due to choledocholithiasis with or without ascending cholangitis.[14]

Generally, patients present with abdominal pain similar to cholecystitis or cholelithiasis. Imaging studies often show scleroatrophic gallbladder or cholecystolithiasis.

According to Bennion, patients can be classified into three large groups based on symptoms: symptomatic patients (50%), asymptomatic patients (35%), and those with other fetal anomalies (15%) of which up to 40-70% are associated sphincter of Oddi obstruction and common bile duct cysts.[15,16] It has also been shown that there are other anomalies in extrabiliary organs such as cardiac defects (54%), and gastrointestinal and genitourinary defects.[8]

Ultrasound is the study of choice for various biliary pathologies, with high sensitivity and specificity (95-98%), using the WES triad (E: gallbladder wall, E: stone echo, S: acoustic shadow).[7] Preoperative diagnosis by ultrasound is very rare, as most ultrasounds in patients with gallbladder agenesis report cholecystolithiasis, which can be confused by periportal tissue, subhepatic peritoneal folds, calcified liver lesions, and duodenal artifacts and due to the presence of acoustic shadows caused by intestinal gas.[8,13,17]

Some authors suggest performing intraoperative cholangiography and even performing wide dissections to exclude ectopic gallbladders, which are usually located in the falciform ligament and the left hepatic lobe, but this may increase morbidity. Therefore, magnetic resonance cholangioresonance (gold standard) should be considered as a confirmatory diagnostic method in cases where ultrasound fails to identify the gallbladder and there is a high index of suspicion, prior to surgery.[3,4] However, most cases are diagnosed during surgery, requiring prolonged surgical exploration that may end in complications.

In 1967, Frey used a series of steps in orden to make a reliable intraoperative diagnosis. He recommended dissection of the extrahepatic bile duct (inclunding the common bile and hepatic ducts) and the perfomance of intraoperative cholangiography to demonstrate the absence of the gallbladder and to exclude an acute or chronic inflammatory process or fibrosis of the cystic plate.[17-19]

There is still controversy regarding the best surgical approach in patients with agenesis, due to the probability of injury to the bile duct and/or other organs.

Most articles recommend expectant management with patient monitoring, without any other type of treatment. For symptomatic patients without cholestasis, supportive care (antispasmodics) should be provided and for patients with cholestasis, ERCP plus sphincterotomy is suggested.[7] Improvement of symptoms after sphincterotomy has also been reported in patients with persistent symptoms who have failed previous treatments.[20,21] However, there are other therapeutic options that are usually used if the patient's symptoms persist, such as biliary diversion (choledochoduodenostomy) or biliary dilatation.

CONCLUSION

Gallbladder agenesis is a rare diagnosis, which is generally made during surgery. Therefore, it is important to make a differential diagnosis with other hepatic and biliary duct pathologies. It is also relevant to mention that magnetic resonance cholangiography should be requested in case of diagnostic doubt since it allows to make a definitive diagnosis, avoiding unnecessary surgical procedures, reducing the risk of complications (bile duct disruption) and mortality for patients.

ACKNOWLEDGEMENTS

The authors thank Dr. Adrián Ferrer Jiménez for the support provided to publish this interesting clinical case.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Ramos BG, Jiménez AF, Trejo MA, Becerra JMR. Gallbladder agenesis associated with dextrocardia in an adult patient: case report. Int Surg J 2024;11:1347-50.