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Gallbladder Agenesis: Two Cases Report and Brief Literature Review

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Abstract: Agenesis of the gallbladder and the cystic duct is one of the rarest congenital abnormalities of the biliary system. Almost half of the patients develop common duct stones and 23% of them manifest signs and symptoms that mimic biliary colic. We present two cases of gallbladder agenesis. The first case is of a woman presenting symptoms of biliary colic. Laparoscopy failed to reveal either gallbladder or cystic duct. The procedure was continued to further search for ectopic sites of gallbladder. A gallbladder agenesis was suspected and then confirmed via post-operative magnetic resonance cholangio-pancreatography, who also objectified a cystic dilatation and septum of the main bile duct. The second case is of a woman with symptoms of biliary colic. A first abdominal ultrasonography objectified a "scleroatrophic" gallbladder; on the second ultrasonography a gallbladder agenesis was suspected and later confirmed via magnetic resonance cholangio-pancreatography. We report through these cases our experience with regard to the challenges associated with the diagnosis and management, and a brief review of the literature of this rare pathology.

Key words: Gallbladder agenesis, congenital biliary tract abnormalities, magnetic resonance cholangio-pancreatography.

1. Introduction

Gallbladder agenesis is a rare congenital abnormality with a reported incidence of 0.007-0.027% in surgical series and 0.04-0.13% in autopsy series [1]. This rare pathology presents predominantly in women (ratio 3:1), and results from either failure of the cystic bud to develop into the gallbladder and cystic duct in utero or failed recanalization of the cystic duct and gallbladder in the first month of life in utero [2]. Its presentation is variable; it may be asymptomatic in 35% of patients, later being discovered incidentally during abdominal surgery or at autopsy. It is present with other fatal fetal anomalies in 15%-16%, and is accompanied by typical symptoms in 50% [1, 2]. The clinical symptoms suggestive of gallbladder agenesis are common to gallbladder diseases. They include right upper quadrant pain, fatty food intolerance, nausea, and jaundice, making it a convincing mimic of cholecystitis and biliary colic [2]. Routine preoperative ultrasound (US) can be misleading and showing a possible "scleroatrophic" gallbladder, and patients can be subject to unnecessary surgical procedures. This diagnosis is in doubt, however, when patients have no previous history of cholecystitis. In this case, magnetic resonance cholangiopancreatography confirms the diagnosis, by showing the absence of the gallbladder. We present two clinical cases of gallbladder agenesis from our practice.

2. Case Report

2.1 Case 1

A 24-year-old woman, without any medical pathologic history, admitted to the emergency department for intermittent right upper quadrant pain described as sharp, evolving since two weeks, and

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associated with nausea and vomiting. On examination, the patient had normal vital signs and mild abdominal pain in the upper abdomen. Laboratory tests were normal for white blood cell count of 9,400 cells/L with normal differential percentages and platelet count of 324,000 cells/L. She had a normal basic metabolic panel, bilirubin, aspartate transaminase (AST), alanine transaminase (ALT), and lipase. She underwent an ultrasound exploration, which was interpreted as "scleroatrophic" gallbladder with multiple gallstones, but without sonographic Murphy's sign, gallbladder wall thickening, pericholecystic fluid, or intra or extrahepatic biliary duct dilatation. A second ultrasound examination was practiced before surgical intervention. It showed no gallbladder in the gallbladder fossa. A magnetic resonance cholangio-pancreatography was performed, which confirmed the gallbladder agénesis diagnosis (Fig. 1).

2.2 Case 2

A 41 year-old woman without a medical history, came to the emergency department for right upper quadrant pain and jaundice, associated to nausea and vomiting. There was not any other medical or surgical history or family history of gastrointestinal diseases. On examination, she had normal vital signs and mild abdominal pain in the upper abdomen. Laboratory tests show a high level of white blood cells, bilirubinemia and transaminase with normal blood levels of lipase and basic metabolic panel. The patient underwent an ultrasound examination, which was interpreted as "scleroatrophic" gallbladder with multiple gallstones associated to biliary duct dilatation, but without gallbladder wall thickening. A diagnosis of scleroatrophic gallbladder with common bile duct stone was evoked. The patient was selected for a laparoscopic cholecystectomy. During the surgical intervention, there was no visible gallbladder, but the surgeon identified a moderate dilatation of the common bile duct without intraluminal stone. Subsequent cholangio-pancreatic MRI was consistent with gallbladder agenesis, showing dilatation of the common bile duct with an intraluminal septum, associated to hilar and periampular stenosis. There were also liver signal abnormalities on hypo signal on T1 ponderation and hyper signal on T2 weighted sequences, with marginal enhancement after gadolinium administration (Fig. 2). No bile duct stone was diagnosed. An echo-endoscopic exploration was performed and no common bile duct stone was detected. The patient was operated for the second time; the gesture consisted on a bilio-digestive bypass with hepatic and biliary biopsies, who diagnosed liver and bile duct sarcoïdosis.

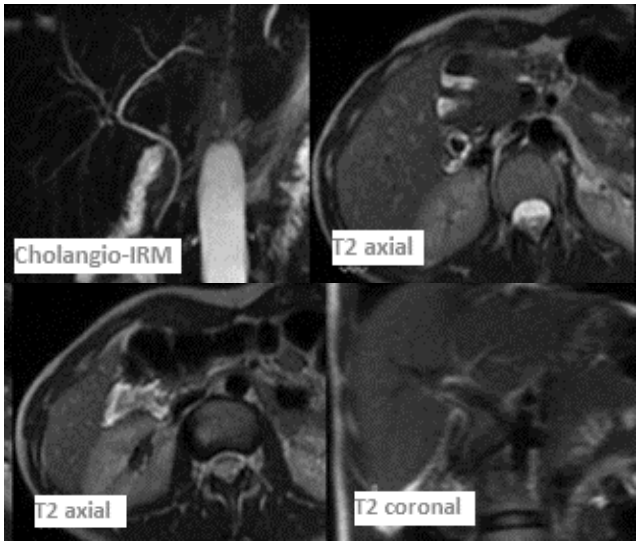

Fig. 1 Magnetic resonance cholangiopancreatography showing normal liver anatomy, absence of the gallbladder and the cystic duct, no common bile duct dilatation. Absence of the gallbladder in the cystic fossa on axial and coronal T2 sequences.

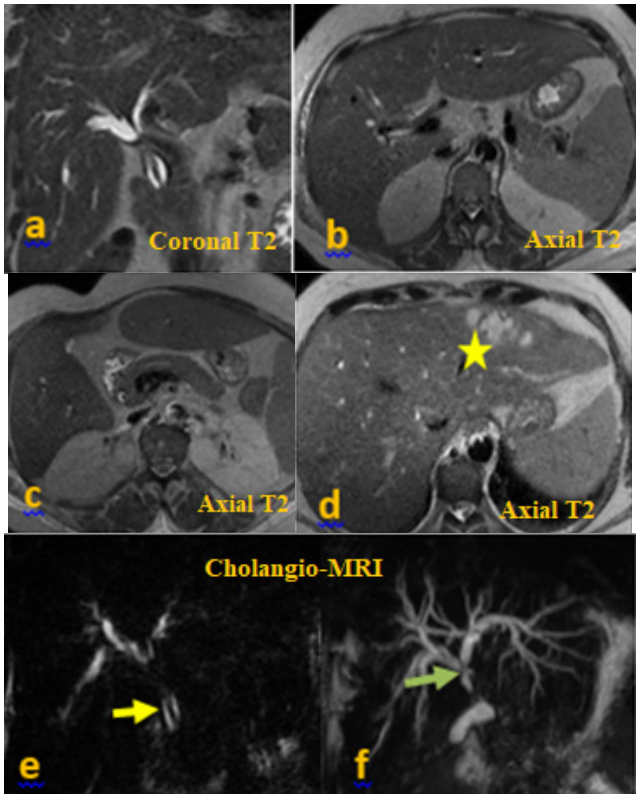

Fig. 2 Gallbladder agenesis: (a ,b, c) absence of individualization of the gallbladder and the cystic duct on coronal and axial T2 sequences. (e, f) Magnetic resonance cholangiopancreatography showing dilatation of the common bile duct with an intraluminal septum (yellow arrow), associated to hilar and periampular stenosis (green arrow). (d) Liver signal abnormalities hyperintense on T2 (yellow star).

3. Discussion

Gallbladder agenesis is an uncommon congenital entity [3]. The first case was diagnosed by Bergman in 1701. Since then, there were described about 500 similar clinical cases in the medical literature [4]. The diagnosis is often misread. Abdominal ultrasonography often evokes a cholecystitis, leading to unnecessary surgery as was in our second case [5]. This entity is related to an embryological abnormality happening on the 4th week of embryological period. It is due to a failure of ventrocaudal bud development from the hepatic diverticulum, or failure of gallbladder and cystic duct recanalization [5]. According to clinical symptomatology, cases are classified into three groups. The first group corresponds to asymptomatic patients; the diagnosis is made on autopsy. The second group of patients has classic symptoms of cholelithiasis such as biliary pain, vomiting, dyspepsia and/or jaundice. The last group presents other abnormalities involving cardiovascular and gastrointestinal systems; the diagnosis is often made in childhood [5]. In both cases, the patients were symptomatic with biliary colic and vomiting, characteristic of cholecystitis. During Gallbladder agenesis, the mechanism of biliary colic is unknown, but greater parts of authors consider that it is determined by dysfunction of Oddi's sphincter and biliary dyskinesia. The fact that pain is relieved after antispasmodic treatment strengthens this theory [4]. Abdominal ultrasonography is the preferred imaging technique for exploring bile ducts, but it is often misread in patients with gallbladder agenesis. Gas artifact, periportal tissue or subhepatic peritoneal folds lead to a wrong diagnosis of shrunken or contracted gallbladder [1]. This misleading diagnosis can lead to unnecessary surgery.

Magnetic resonance cholangiopancreatography (MRCP) is an interesting imaging technique that can

prevent unnecessary surgery [5]. The cholangio-MRI allows the biliary tree to be analyzed with precision and confirms gallbladder agenesis [4, 5]. It also eliminates an ectopic gallbladder (on the left of the falciform ligament, intrahepatic, retrohepatic, retroduodenal or retropancreatic) [6]. Abdominal CT has no place for confirming the diagnosis of gallbladder agenesis; however, according to some authors, it can be used in the absence of MRI [5]. The use of surgery to confirm diagnosis of gallbladder agenesis and to remove ectopic localizations may increase the risk of iatrogenic complications [5].

Our second patient presents in addition to the gallbladder agenesis, a septum of the main bile duct, staged stenosis of bile ducts and fleeting signal abnormalities of the hepatic parenchyma. Histological study of hepatic tissue and bile ducts biopsies revealed sarcoïdosis.

4. Conclusion

Gallbladder agenesis is a rare anomaly. It poses a diagnostic dilemma as it is usually diagnosed during a laparoscopic cholecystectomy due to misleading interpretation on ultrasonography which reveals usually a "scleroatrophic" gallbladder. However, cholangio-MRI allows exploring with precision the entire biliary tree and confirms agenesis. So, any suspicion of gallbladder agenesis must encourage carrying out a cholangio-MRI before any surgical gesture on the bile ducts, to decrease iatrogenic risk related to abusive operative gestures.

Conflicts of Interest

The authors declare that they have no conflict of interests.

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