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Case Report

Gallbladder agenesis in a patient with choledochoduodenal fistula, report of an exceptional case

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ABSTRACT

Vesicular agenesis refers to the congenital absence of the gallbladder, however, vesicular agenesia is a very scarce condition worldwide, so there is little information available on its clinical presentation and surgical data. This is a 79-year-old male patient, with a history of diabetes mellitus of long evolution, without a surgical history, who comes for colic pain of 15 days of evolution in right hypochondrium, as well as unquantified fever and jaundice, during its initial evaluation in the emergency room para clinics were requested observing leukocytosis, hyperbilirubinemia and elevation of liver enzymes, ultrasound of liver and bile ducts reported common anatomical situation gallbladder, occupied entirely by multiple hyperechogenic images, configuring sign of W.E.S. and common bile duct of 7.2 mm, concluding gallbladder scleroatrophic and vesicular agenesia during trans-surgery. Vesicular agenesis is an extremely rare condition that is mostly diagnosed during surgery, which can lead to erroneous diagnosis and unnecessary surgery in patients with symptoms including biliary colic, choledocholithiasis with or without cholangitis, and an ultrasound showing a scleroatrophic gallbladder. Surgeons In situations where there is clinical evidence consistent with biliary colic in a context of gallbladder agenesis, and the symptoms persist without finding any other cause, a surgical approach to release adhesions could be considered; since, as observed, this may result in improved symptoms, although the explanation is not yet completely clear.

Keywords: Liver, Agenesis, Gallbladder, Surgery, Leukocytosis

INTRODUCTION

Vesicular agenesis refers to the congenital absence of the gallbladder, however, vesicular agenesia is a very scarce condition worldwide, so there is little information available on its clinical presentation and surgical data.[1]

Ther vesicular agenesis is rare, no precise prevalence has been established in the general population. Most studies report isolated cases or case series, which makes it difficult to obtain accurate data, however, they have been reported in some of these series, an incidence ranging between 0.01% and 0.075% (10-75 per 100,000 inhabitants).[1] The reported incidence based on autopsy findings is approximately one case per 6334 live births, or approximately 0.035% to 0.3%, it is usually diagnosed during the transoperative.[2,3]

In any case, if, during surgery the gallbladder is not found, the surgeon must take the necessary measures to ensure the safety of the patient. This may include careful examination of the area for possible gallbladder remnants, gallbladder ectopia, as well as evaluation of the bile duct. The rarity of this association is an exceptional event, since we do not find in the literature the concomitant presentation with choledoco-duodenal fistula.

Biliary fistulas are a rare entity, of which common bile duct-duodenal fists report the lowest incidence, usually congenital or acquired, as a result of inflammatory or traumatic processes, gallstones, previous biliary surgery, infections and inflammatory diseases of the biliary tract. These conditions can weaken the wall between the common bile duct and duodenum, allowing the formation of an abnormal connection.

The clinical presentation is very varied and may become asymptomatic. complications may occur, however, cholangitis being more frequent, followed by biliary ileus, by the impact of a lithum at the level of the ileocecal valve, or present as Bouveret syndrome due to obstruction at the level of the duodenum.[2,4,9] A clinical case of a patient with acute cholangitis and choloduodenal fistula in which vesicular agenesis was diagnosed during the trans operative procedure, also describes its management, evolution and a review of the medical literature.

CASE REPORT

This is a 79-year-old male patient, with a history of diabetes mellitus of long evolution, without a surgical history, who comes for colic pain of 15 days of evolution in right hypochondrium, as well as unquantified fever and jaundice, during its initial evaluation in the emergency room para clinics were requested observing leukocytosis, hyperbilirubinemia and elevation of liver enzymes, ultrasound of liver and bile ducts reported common anatomical situation gallbladder, occupied entirely by multiple hyperechogenic images, configuring sign of W.E.S. and common bile duct of 7.2 mm, concluding gallbladder scleroatrophic (Figure 1). with high risk for choledocholithiasis by criteria of ASGE 201917 and with data of mild acute cholangitis according to criteria of Tokyo 18 with significant adjacent inflammatory process, so endoscopic retrograde cholangiopancreatography (ERCP) was performed observing normal intrahepatic bile duct, extrahepatic dilated 10 mm in its greater portion, balloon catheter was scanned with removal of abundant stones of 3 to 8 mm and bile sludge, finding a choledooduodenal fistula, (Figure 2) which was handled conservatively, evolved favorably, so it was decided to discharge and schedule for elective laparoscopic cholecystectomy a month later.

During the surgical procedure, diagnostic laparoscopy was performed, finding an plastron epiploon dependent, liver and bile duct, adheciolysis was performed with electrocoagulation and choledoco-duodenal fistula was observed, which was dismantled without observing leakage data, the hepatic bed was focused without observing the gallbladder, so that when it was not found in the usual position, it is reviewed in possible ectopic locations such as the left liver lobe, major omentum, minor and suprahepatic without visualizing it, so it is decided to perform longitudinal choledotomy and transoperative cholangiography (Figure 3), observing preserved intrahepatic bile duct, extrahepatic dilated up to approximately 15 mm, without filling defects, with adequate contrast passage to the duodenum, unidentified gallbladder, bile duct (Figure 4), so it is carried out coledocorraphy with monocryl and it is placed drainage to Morrison's space, it enters the floor of general surgery where it is observed adequate postoperative evolution and it is decided to leave after three days with adequate tolerance to the oral route, without data of biliary leakage, follow-up was carried out a week in an outpatient clinic where stitches and drainage were removed, ultrasound of the liver and bile ducts was requested, and no gallbladder was reported in the usual topography, with dilated intrahepatic bile duct (Figure 5) that did not merit management, was revalorized a month later in the office, where he referred asymptomatic so he discharged our service.

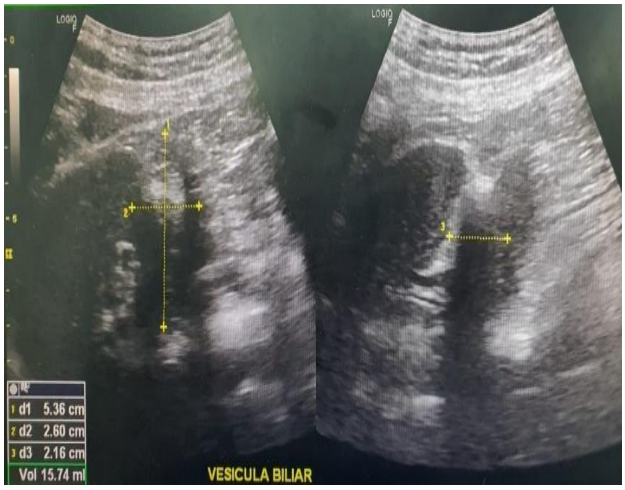

Figure 1: Ultrasound of the liver and bile ducts showing hyperechogenic images, with acoustic shade, and which was reported as WES phenomenon, concluding the scleroatrophic gallbladder.

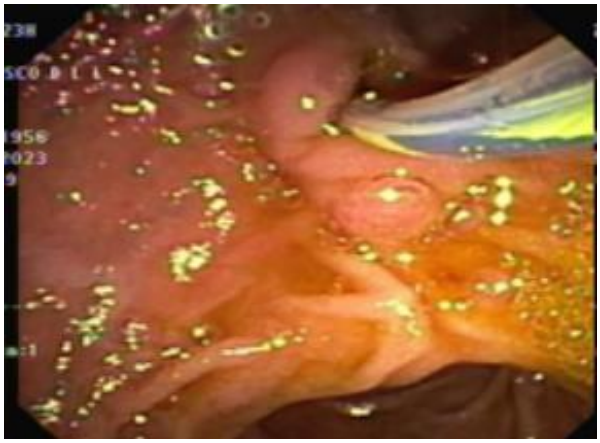

Figure 2: Endoscopic retrograde cholangiopancreatography in which the greater papilla is observed without alterations, with hydatophilic guidance cannulating the choledochoduodenal fistula.

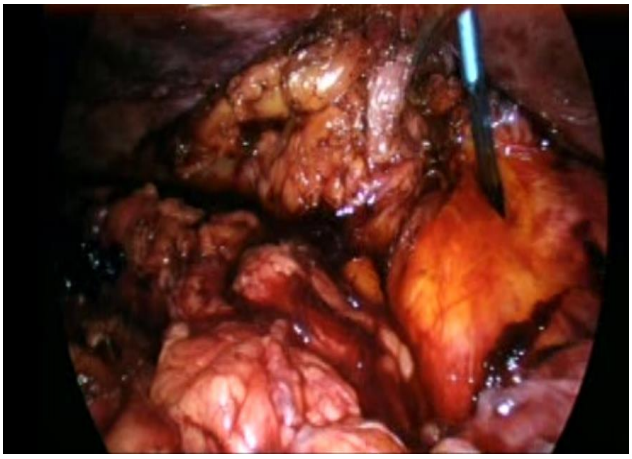

Figure 3: Laparoscopic vision of dissected plastron, dilated common bile duct and absence of gallbladder in hepatic bed, with feeding tube introduced in common bile duct, prior to cholangiography.

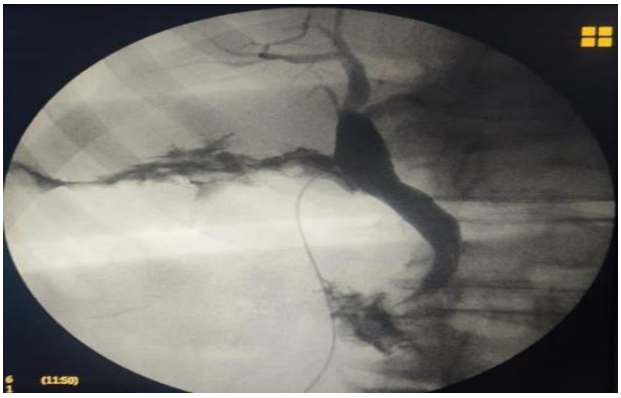

Figure 4: Transoperative cholangiography, after dismantling of choledochoduodenum fistula, where intrahepatic bile is observed without alterations, dilated extrahepatic up to 15 mm, without filling defects, with adequate passage from contrast medium to duodenum and leakage at choledotomy site, as well as absence of gallbladder.

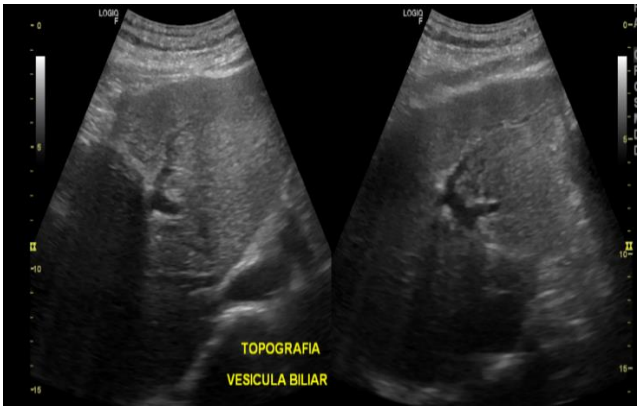

Figure 5: Ultrasound of the liver and bile ducts, after a surgical procedure, where the gallbladder is absent in the usual topography, with dilation of the intrahepatic bile duct.

DISCUSSION

There is no exact date of publication in the medical literature describing for the first time vesicular agenesia, however, the first cases reported are attributed to Lemery in 1701 and Bergman in 1702, and since then there are about 430 reports of new cases, however, the number is increasing as new publications come to light.[2,4]

Incidence varies between 0.01% and 0.075% (10-75 per 100,000 inhabitants).[1] The reported incidence based on autopsy findings is approximately one case per 6334 live births, or approximately 0.035% to 0.3%.

Women are most often affected (3:1) and are usually diagnosed in the second or third decade of life.

It may be associated with other congenital defects in 40-65%, such as trisomy 18, thalidomide malformations, cerebrotendinous xantomatosis, inheritance not linked to sex with variable penetrance and alterations of the primitive intestine, cardiopulmonary defects, in addition to genitourinary abnormality; however, these alterations were not found in our patient.[9,12]

Regarding the clinical presentation of vesicular agenesis, three main groups have been described: 1) those with multiple fetal abnormalities detected perinatally (15%), 2) asymptomatic cases found at autopsy or incidentally during surgery and 3) symptomatic cases that usually occur in the third to fourth decades of life (50%).[9]

It is estimated that 25% to 50% of patients with gallbladder agenesis may develop choledocolithiasis secondary to bile duct stasis8, jaundice is due to choledocholithiasis associated with or without ascending cholangitis.[2] which explains the clinical presentation of our patient, when debuting with mild acute cholangitis, which merited drainage of the bile duct with ERCP, however during the procedure the presence of a choledocho-duodenal fistula was found, which is reported with an incidence of 5% of biloenteric fistulas (incidence of 0.15-8% in the presence of cholelithiasis), however, there are no reports of the presence of choldocho-duodenal fistula in the context of gallbladder agenesis.[9]

Ultrasound is the method of choice for the diagnosis of biliary pathology, for which the radiologist uses the triad WES (Wall, Echo, Shadow) which is estimated to have a sensitivity of 95-98% for the diagnosis of cholecystitis; some poorly diagnosed acoustic shadows are due to intestinal gas, duodenum artifacts, peritoneal subhepatic folds, small or atrophic vesicles, contracted on lithos, so in many cases of vesicular agenesia, is reported as scleroatrous gallbladder, making preoperative diagnosis more difficult.[11,13]

In the case of our patient, who underwent acute cholangitis, an important local inflammatory process was developed that was observed during the surgical

procedure when finding adhesions, plastron in the biliary tract, as well as the presence of the choledochoo-duodenal fistula, which was reported in ultrasound as a sclerotrophic gallbladder and according to the literature.[2,4,10]

In the literature review of Peloponissios et al covering the period 1960 to 2003, they indicate that all the cases analyzed were diagnosed during surgery or in the postoperative period, with only two exceptions.[12,14] Therefore, it is difficult to make a preoperative diagnosis reliably and, in most cases, the diagnosis is established during surgery or in the postoperative period.[2,4,15]

So far, there are no diagnostic or management guidelines for gallbladder agenesis, however in 1967, Frey proposed to meet certain criteria during the transoperative that consisted in evidencing the absence of inflammatory signs or fibrosis in the vesicular bed, convert to laparotomy and perform the exhaustive search for an ectopic gallbladder, intentionally searching for it intrahepatically, retrohepatically, in the left hemiabdomen, between the 2 layers of the minor omentum, in the sickle ligament, retropancreatically, retroperitoneal and in the anterior wall and if not found, a transoperative cholangiography with bile duct exploration should be performed if the common bile duct is dilated more than 2 cm or if choledocholololithiasis.[6,9,12]

In this case, we decided to continue using the laparoscopic approach because we could clearly and completely visualize the bile duct. Therefore, we consider that switching to a laparotomy would imply a greater risk of complications associated with the intervention, without providing significant additional information, however, finding the dilated bile duct and having a history of choledocholithiasis, we decided to perform transoperative cholangiography where we observed the integral biliary tree, as well as absence of the gallbladder, so the diagnostic dilemma was solved during the transoperative.

Most patients reported in the literature experience improved symptoms after surgery, even if it was not therapeutic. This is believed to be due to the release of periportal adhesions and the right hypochondrium during the gallbladder search, although it is unclear why the symptoms are resolved.

In our patient, deferrelaparoscopic cholecystectomy was indicated as part of the management for acute cholangitis, in accordance with the guidelines of Tokyo 18, however, since no gallbladder was found, it was decided to end the therapeutic algorithm.

In cases where symptoms associated with biliary colic occur and persist in the postoperative period, a conservative approach using smooth muscle relaxants may be considered. If this is not successful, it is suggested to perform a sphincterotomy.[11,12]

CONCLUSION

Vesicular agenesis is an extremely rare condition that is mostly diagnosed during surgery, which can lead to erroneous diagnosis and unnecessary surgery in patients with symptoms including biliary colic, choledocholithiasis with or without cholangitis, and an ultrasound showing a scleroatrophic gallbladder. Surgeons should be alert to the possibility of vesicular agenesis when faced with a difficult dissection, scleroatrophic gallbladder or anatomical variations during laparoscopic cholecystectomy. Transoperative cholangiography is useful to confirm the diagnosis, address other conditions, and prevent bile duct lesions. In situations where there is clinical evidence consistent with biliary colic in a context of gallbladder agenesis, and the symptoms persist without finding any other cause, a surgical approach to release adhesions could be considered; since, as observed, this may result in improved symptoms, although the explanation is not yet completely clear.

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