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GALLBLADDER AGENESIS: AN ANATOMICAL SURPRISE

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ABSTRACT

We report the case of a patient with a rare congenital anomaly of the bile ducts: gallbladder agenesis. This rare congenital anomaly is often asymptomatic and undetected during life. In our patient, admitted for acute lithiasic angiocholitis complicated by liver abscesses, agenesis of the gallbladder was suspected on ultrasound and confirmed by surgical exploration.

INTRODUCTION

Gallbladder agenesis is a rare biliary tract anomaly characterized by the congenital absence of the gallbladder (GB) associated or not with the absence of the cystic duct.[1] It is most often asymptomatic and not detected during life.[2] According to M. Bouche et al.[3] in 23% of cases, patients with this anomaly present with symptoms such as liver colic. However, the usual radiological work-up in the presence of hepatic colic rarely evokes this diagnosis, rather interpreting the images observed as a sclero-atrophic vesicle.[3] We report, through this observation and the review of the literature, the epidemiological, anatomical and therapeutic particularities of this anomaly.

Patient ET Observation

Patient information: A 58-year-old diabetic patient on insulin therapy, hypertensive on ACE inhibitor, never operated, was admitted to the emergency room for right hypochondriac pain evolving in a feverish context for 72 hours.

Clinical findings: Clinical examination on admission found a conscious patient, hemodynamically and respiratory stable, febrile at 39°C, tachycardic at 92 beats/min, with slightly discolored conjunctiva. On abdominal examination, there was tenderness in the right hypochondrium and a negative shake sign.

Diagnostic approach: The biological work-up revealed a normocytic normochromic anemia at 10.7 g/dl, and a biological inflammatory syndrome with a CRP at 150 mg/dl and a hyperleukocytosis at 10680 elts/mm3 with a predominance of neutrophilic polynuclear cells. Abdominal ultrasound followed by an abdominal CT scan with injection of contrast product revealed several liver abscesses straddling segments V, VII, and VIII, of which the largest measured 12.5 cm. The main bile duct (MBD) was dilated to 14 mm without visible obstruction, and the gallbladder was not seen (Figure 1).

Therapeutic intervention: Echoguided drainage of the abscess was first performed, bringing back 1200 cm3 of chocolate pus. A probabilistic antibiotic therapy was then started with metronidazole (1.5g/dr) and ceftriaxone (2g/dr), with saline rinsing every 6h. In front of the appearance, on the 3rd day after the drainage, of intense abdominal pains diffuse to all the abdomen with generalized abdominal defense and the increase of white blood cells, an imaging was made objectifying the persistence of abscessed collections of the segments VI, VII and VIII, containing air bubbles opposite to the distal end of the drain, with the presence of a small to moderate intraperitoneal effusion at the peri-hepatic and pelvic levels and enhancement of the peritoneal sheets.

The patient was then referred to the operating room because of the strong suspicion of acute generalized peritonitis, and the surgical exploration, performed by median laparotomy, revealed the presence of a purulent effusion at the level of the cul de sac of Douglas and at the level of the parieto-colic gutters in relation to intra-peritoneal leaks around the radiologic drain. The gallbladder was not found and the MBD was dilated and lithiasic. The surgical procedure consisted of an abundant warm saline lavage, followed by a wide drainage of the abdominal cavity, after removal of the radiological drain and surgical drainage of the hepatic abscess. Subsequently, an endoscopic retrograde

cholangiopancreatography confirmed the dilatation of the VBP to 18 mm upstream of multiple lacunar images. Sphincterotomy with extraction of all stones with the balloon ensured the vacuity of the main bile duct (Figure 3).

Follow-up and results: The postoperative course was simple and the patient was discharged from the hospital within a week of the procedure (figure 3).

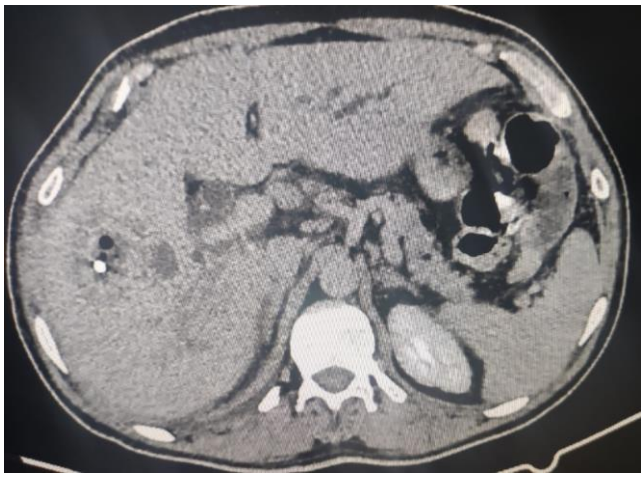

Figure 1: CT scan section, showing a hepatic abscess with a drain within it, no individualization of the gallbladder.

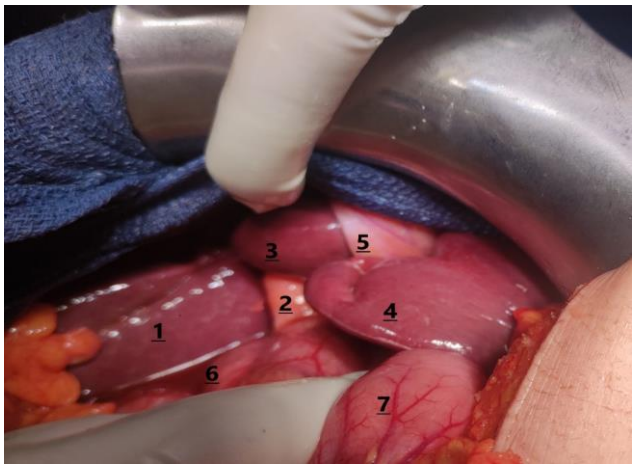

Figure 2: Intraoperative image confirming the absence of the gallbladder. 1: segment 5, 2: hepatic pedicle, 3: segment 4 of liver, 4: left liver, 5: round ligament, 6: duodenum, 7: stomach.

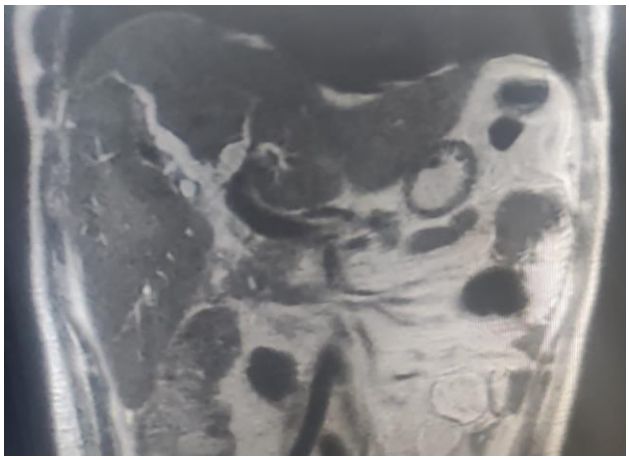

Figure 3: Frontal MRI section showing dilatation of the MBD, and IHBD, without individualization of the gallbladder.

DISCUSSION

Congenital absence of the gallbladder is rare, its incidence is variable according to authors and varies from 0.01 to 0.09% of live births. The study of autopsy reports does not show a gender predominance,[4,5] but in clinical reports the ratio of females to males is 3, probably because the symptomatology is similar to that of gallbladder disease, which is more frequent in females.[6]

Embryologically, the agenesis is secondary to a defect in the in-utero development of the caudal bud of the hepatic diverticulum, which appears around the middle of the 3rd week of gestation at the distal part of the foregut. More recently, it has been shown that genetic transcription factors are required for the development of the vesicle such as: Sox 17+, Pdx 1+, HNF6, HNF1b located on chromosomes 8, 13, 15 and 17 respectively.[8,9]

Agenesis of the bile duct is most often asymptomatic and undetected during life In this context, Bennion et al have established a classification that distinguishes 3 groups.[1,10]

a) Multiple fetal anomalies group (12.9%): these patients usually die in the perinatal period from their malformations, the absence of gallbladder is only found at autopsy. Cardiovascular malformations are the most common, followed by those of the gastrointestinal and genitourinary systems.

b) Asymptomatic group (31.6%): This group of patients without a gallbladder is most often discovered at autopsy or during an operation performed for another diagnosis. None of these patients have any biliary symptoms.

c) Symptomatic group (55.6%): This group corresponds to the age group 40-50 years, usually without other congenital anomalies.

As far as the clinical presentation of this pathology is concerned, the functional signs are not specific explaining the delay in diagnosis. They are dominated by pain of the hepatic colic type (54%), jaundice (27%) with lithiasis associated with dilatation of the bile ducts in 25 to 50% of cases.[5]

Vesicular agenesis is not always easy to establish because of the rarity of this anomaly and the failure of hepatobiliary imaging means to reach 100% sensitivity.[1] Ultrasound is the initial reference examination for the exploration of biliary pathology.[11] Its sensitivity is 95% in the diagnosis of vesicular lithiasis.[12] In most cases, the ultrasound report reported by the radiologist is a lithiasic scleratrophic vesicle. These false positives can be explained by the interposition of small intestines in the vesicular fossa, the interposition of periportal peritoneal folds or by foci of hepatic calcifications.[11,13] As another pathophysiological consequence of agenesis, we can find a dilatation of the choledochus which mimics a shrunken gallbladder to compensate for the absence of a gallbladder by taking on the function of bile storage.[10]

Currently, Bili-MRI is the gold standard for detecting gallbladder agenesis. It is a non-invasive imaging test that allows to find a gallbladder in an ectopic site (in the falciform ligament, the lesser omentum, in the pancreas, behind the duodenum, in the pyloric digestive wall, or even intrahepatically), and in the absence of a found gallbladder, to make the correct preoperative diagnosis of the agenesis.[14]

The therapeutic approach differs according to the circumstance of discovery (preoperatively or intraoperatively); whether or not it is associated with lithiasis of the main bile duct and whether or not there is an Oddi's Sphincter dysfunction.[1]

In the absence of a preoperative diagnosis, symptomatic patients are operated for cholecystectomy either by conventional or laparoscopic approach. During the operation, the various ectopic sites of the gallbladder are searched for with the help of intraoperative ultrasound if available. Intraoperative bile duct puncture cholangiography is not recommended due to the high risk of bile duct or portal vein injury during cholangiography. In all cases, postoperative bili-MRI should be performed to confirm the diagnosis.[14,16]

If the diagnosis is made preoperatively and the patients are asymptomatic, no treatment is required. In patients with biliary pain, the treatment consists of either medical treatment represented essentially by smooth muscle relaxants used to relieve discomfort, or endoscopic sphincterotomy.[1,17]

Concerning agenesis associated with choledocholithiasis, the treatment is the same as in the case of residual lithiasis, namely endoscopic sphincterotomy in the first instance and surgery in the case of a large stone with impossibility of extraction.[1]

CONCLUSION

Agenesis of the gallbladder is a very rare congenital condition, due to an anomaly of embryological development. Most often asymptomatic and of fortuitous discovery, it does not require any treatment, except in the case of complications as in our patient who presented with choledocholithiasis complicated by hepatic abscess. On the other hand, it must be evoked in front of a scleratrophic aspect of the gallbladder on ultrasound and must be confirmed by bili-MRI in order to avoid a useless surgical intervention to the patient.

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