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Gallbladder Agenesis: A Case Report of an intraoperative diagnosis

Agnesia da Vesícula Biliar: Relato de Caso de um diagnóstico intra-operatório

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ABSTRACT

Introduction: Agenesis of the gallbladder is a rare congenital anomaly occurring in 10 to 65 per 100 000 people. Patients are usually asymptomatic and the diagnosis is made as an incidental finding during another abdominal surgery or at autopsy. The presence of biliary symptoms and a misleading ultrasound study can lead to unnecessary surgery. **Case Presentation:** We present a 43-year-old female proposed to laparoscopic cholecystectomy for suspected symptomatic gallbladder lithiasis in the clinical and radiological studies prior to surgery. During operation the gallbladder was not identified and the procedure was terminated for further diagnostic procedures. The post-operative study with magnetic resonance cholangiopancreatography confirmed the diagnosis of gallbladder agenesis. **Conclusion:** The difficulty in diagnosing the absence of gallbladder in the preoperative period can be explained by the infrequency of the condition and consequent low index of suspicion for agenesis when interpreting imaging findings. Surgeons must be aware of this condition and when gallbladder isn't visualized at laparoscopy decide between convert surgery to open procedure or abort surgery and continue study with other imaging techniques. The latter is a good option, allowing a better characterization of biliary anatomy as well as avoiding morbidity of a laparotomy.

Key Words: Gallbladder, agenesis, laparoscopic cholecystectomy, case report.

RESUMO

Introdução: A agnesia da vesícula biliar é uma anomalia congénita rara que ocorre em 10-65/100 000 pessoas. Os doentes são geralmente assintomáticos e o diagnóstico é feito como um achado incidental durante outra cirurgia abdominal ou na autópsia. A existência de sintomatologia biliar e uma ecografia não esclarecedora podem ser a causa de uma cirurgia desnecessária. **Caso Clínico:** apresentamos o caso de mulher de 43 anos proposta para colecistectomia laparoscópica por suspeita de litíase biliar sintomática na avaliação clínica e radiológica prévia à cirurgia. No intra-operatório a vesícula biliar não foi identificada e terminou-se a cirurgia para execução de outros procedimentos diagnósticos. O estudo pós-operatório com Colangiorrressonância confirmou o diagnóstico de agnesia da vesícula biliar. **Conclusão:** A dificuldade no diagnóstico de agnesia da vesícula biliar no período pré-operatório pode ser explicada pela raridade da situação e consequente baixo índice de suspeição ao interpretar os achados imagiológicos. Os cirurgiões devem conhecer esta entidade e quando a vesícula biliar não se visualiza na laparoscopia, decidir entre a conversão do procedimento ou terminar a cirurgia e continuar o estudo com outros meios imagiológicos. Esta parece ser uma boa opção, permitindo não só uma boa caracterização da anatomia biliar como evitar a morbilidade de uma laparotomia.

Palavras Chave: vesícula biliar, agnesia, colecistectomia laparoscópica, caso clínico.



INTRODUCTION

Although biliary system variants are extremely frequent, isolated congenital agenesis of gallbladder is an extremely rare entity with an estimated incidence of 10-65 per 100 000 people. The first reports of a gallbladder agenesis date from 1701 and 1702 by Lemery and Bergman. Some associations with other congenital abnormalities were reported, mainly involving genitourinary, gastrointestinal, cardiovascular systems or skeleton and abdominal wall defects.^{1,2,3}

Fifty percent of the patients are asymptomatic and the diagnosis is made as an incidental finding during another abdominal surgery or at autopsy.⁴ A quarter to a half of patients will develop duct stones and present biliary colic or other forms of biliary tract disease, requiring some form of intervention. The presentation is usually between 2nd and 5th decade of life, with female to male ratio of 3:1. Despite all available imaging studies, this entity is usually not diagnosed preoperatively and patients undergo unnecessary surgery with increased risk of iatrogenic injury.^{1,2}

It is our purpose to alert other surgeons for this unusual congenital biliary anomaly as well as to highlight the importance of careful dissection during surgery even with normal prior image studying.

CASE PRESENTATION

A 43-year-old woman presented to the outpatient clinic with complaints of right upper abdominal pain for 6 weeks. She was medicated with Lisinopril 5mg once a day for hypertension. No past history of abdominal surgery. The pain was described as dull and crampy, sudden in onset with associated nausea and anorexia. Symptoms worsened with meals, mainly with fatty food, and lasted for 45-60 minutes after each meal. The physical examination was unremarkable, as well as the blood-work, mainly in what concerns to liver-functions tests. Abdominal ultrasound showed a shrunken fibrotic gallbladder with posterior acoustic shadowing suggesting lithiasis and a normal biliary tree. The

patient was scheduled for laparoscopic cholecystectomy. Intraoperatively, the gallbladder, the cystic duct and the cystic artery were not observed in the gallbladder fossa (Figure 1) or other possible ectopic sites, despite meticulous inspection of the supra-mesocolic abdominal cavity. The supraduodenal common bile duct was inspected and didn't seem to have any abnormality. It was decided not to convert the procedure nor to perform an intraoperative cholangiography due to the good visualization of the biliary tract. The postoperative period was uneventful and the subsequent magnetic resonance cholangiopancreatography (MRCP) confirmed the diagnosis of agenesis of the gallbladder (Figure 2) and ruled out an ectopic gallbladder. The patient underwent endoscopic study of the gastrointestinal tract with upper and lower endoscopy, echocardiography and genitourinary tract ultrasound without identification of other congenital malformation.

After surgery the patient achieved a good control of symptoms with butylscopolamine 10mg taken up to three times a day and remains asymptomatic.

DISCUSSION

Although gallbladder agenesis is rarely diagnosed in living patients, it is found with higher incidence in autopsy based studies (up to 90 per 100 000 people).

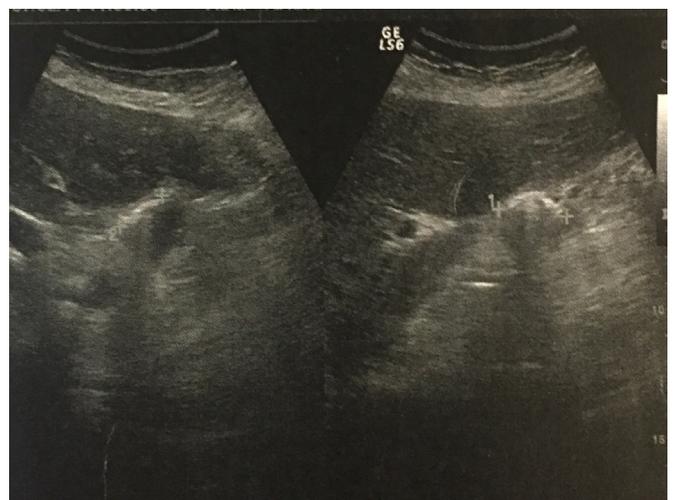


FIGURE 1: Intraoperative image showing absence of the gallbladder (A- supraduodenal biliary tree; B – duodenum).



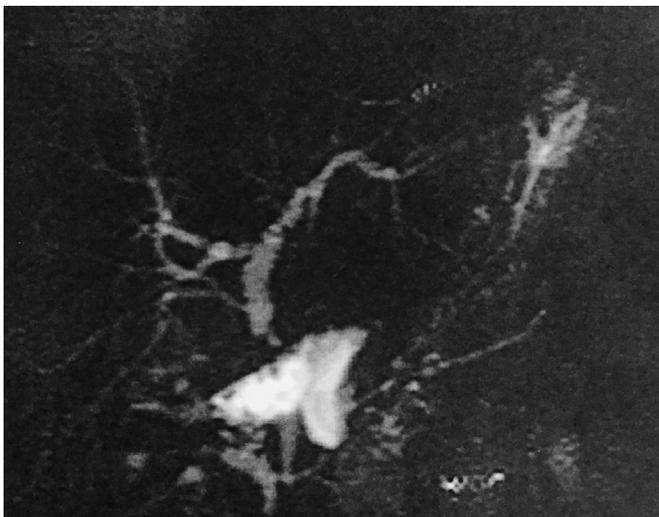


FIGURE 2: MRCP (Fig. 2) showing left (Fig. 2A), right (Fig. 2B) and common hepatic (Fig. 2C) ducts with absence of the gallbladder and cystic duct; the common bile duct is non-dilated.

The pathogenesis, which could result in congenital absence or other malformations of the gallbladder, are poorly understood and probably multiple. Liver, gallbladder and cystic duct begin to develop in the second week of gestation as double ventral outgrowth of the endodermal tube. The first elongates and become the liver and the common bile duct. During the fourth week, it forms the more caudal outgrowth, the cystic bud, the primordium of the gallbladder and cystic duct. Failure during any stage of this process can result in congenital absence of the gallbladder.^{3,4} In the majority of situations, gallbladder agenesis is sporadic, but there are some described associations with genetic abnormalities, like trisomy 18, as well as inherited defects.^{3,5} Any anomaly during migration of gallbladder primordium will result in ectopic gallbladder, with described ectopic sites including intra-hepatic location, hepatic left lobe, posterior hepatic surface, lesser omentum, retroperitoneum as well as retropancreatic and retroduodenal areas.⁴

Some authors classify patients in three groups: asymptomatic ones with findings in necropsy or as a finding during surgery for other reason; symptomatic ones; and those with other severe congenital anomalies found in childhood.

As in our patient case the symptoms could include right upper quadrant pain, as well as, dyspepsia, jaundice, fatty food intolerance and nausea, which make differential diagnosis between gallbladder agenesis and choledocholithiasis, biliary dyskinesia or post-cholecystectomy syndrome, amongst others.

The symptoms reported by the patients with gallbladder agenesis are usually explained by biliary dyskinesia (with retrograde contraction of the Sphincter of Oddi), primary choledocolithiasis or extrabiliary symptoms.^{3,5}

Historically all cases were diagnosed during surgery or necropsy. Nowadays, with advance of imaging resources, this cases are more often diagnosed before any intervention. Usually abdominal ultrasound is the only imaging study done before cholecystectomy for lithiasis. However, as happened in this case, ultrasound can be misleading toward an atrophic or fibrotic gallbladder, which is the most frequent radiologic report seen in this patients. Pre-operative MRCP should be the imaging tool used as the next investigation step when US is not conclusive.⁵

Before, when the diagnosis was made during laparoscopy, the option to convert to open procedure was indicated, allowing a better exploration of abdominal cavity as well as the extra-hepatic biliary tree. The conversion doesn't have any benefit and adds morbidity to the procedure. Nowadays, the majority of authors agree that invasive procedures during surgery should be avoided, and the investigation should continue after surgery with other radiologic and endoscopic modalities. The MRCP, Endoscopic Ultrasound (EUS) and Endoscopic Retrograde Cholangiopancreatography (ERCP) provide good alternatives to explore the extra-hepatic biliary tree. The ERCP has little contribution to the diagnosis, because the non-visualization of the gallbladder can be interpreted as a cystic occlusion. In turn, it is a treatment option allowing sphincterotomy. The MRCP, as noninvasive imaging method, can be used to diagnosis gallbladder agenesis or to exclude ectopic gallbladder. It should be considered when US is unable to demonstrate the gallbladder.^{4,5}



Treatment for symptomatic patients can vary between medication with smooth muscle relaxants and invasive measures like sphincterotomy.

CONCLUSION

Despite being a well-recognized entity and with all advances in biliary tract imaging, the presence of biliary symptoms and a misleading ultrasound study can lead to unnecessary surgery.

Surgeons performing laparoscopic cholecystectomy must be aware of this anomaly to prevent iatrogenic injuries. In fact, newer minimally invasive imaging modalities provide surgeons good means to study the biliary anatomy as well as any anomaly. In cases of intraoperative diagnosis of gallbladder agenesis, it is better to stop the surgery and undergo imaging study. MRCP is the most accurate non-invasive imaging study to establish the diagnosis.

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