A GHOST GALLBLADDER. AN UNUSUAL CASE OF GALLBLADDER AGENESIA FROM A CLINICAL AND MEDICO-LEGAL POINT OF VIEW

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ABSTRACT

Introduction*: Congenital agenesis of the gallbladder is an uncommon anatomical variation. This congenital disease is usually asymptomatic, although some affected individuals may have a clinical picture suggesting gallbladder disease.*

Case presentation*: A 31-year-old woman entered the emergency room with nonspecific abdominal symptoms, compatible with a gallbladder disease. The patient underwent many ultrasounds and an MRI cholangiography that did not display the gallbladder. Despite the negative results, doctors suspected lithasic cholecystitis, due to persistent symptoms, and suggested a laparoscopic cholecystectomy. However, after converting the surgery to laparotomy and performing an intraoperative cholangiography, surgeons diagnosed a gallbladder agenesis.*

Conclusion*: The present case raises two interesting medico-legal issues related to surgical indication and informed consent. Clinical knowledge of this case and the medico-legal implications can be useful to any healthcare professional in order to increase the safety of treatments and prevent adverse events.*

Keywords: *gallbladder agenesis, congenital abnormalities, informed consent, medico legal evaluation.*

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Introduction

Cystic duct and/or gallbladder congenital agenesis (GA) is an uncommon anatomical anomaly of the biliary system, which occurs in about 0.01%-0.13% of patients at birth (some authors report an incidence of 13-65 cases per 100,000 people or an incidence of 0.04-0.13% in the autopsies). In fact, to date, about 400 cases of GA have been described in the medical literature[1-2]. This disease depends on an alterated developmental of the embryonic ectodermal hepatic diverticulum during the fourth week of gestation and it is usually associated with other congenital malformations (extrahepatic bile duct atresia, absence of quadrate lobe, cardiovascular anomalies, etc.); the isolated absence of the gallbladder is possible, but very rare[3]. Most affected individuals remain asymptomatic for life, but 23% of cases have nonspecific symptoms, such as right upper quadrant pain, dyspeptic symptoms or abdominal discomfort, especially during the fourth or fifth decade of life. Due to its low frequency and nonspecific symptoms, it represent a diagnostic and therapeutic dilemma for surgeons. In fact, routine investigations often fail to diagnose GA, which are usually misinterpreted as cholecystitis with cystic duct obstruction or as a sclero-atrophic gallbladder, thus leading to unnecessary surgery. Furthermore, during laparoscopic gallbladder research, biliary or portal structures can be easily injured during the dissection process; in fact,

the absence of normal anatomical structures and the inability to pull the gallbladder to dissect the Callot triangle represent a risk of iatrogenic injury. In this regard, in case of incidental detection of a GA during a surgey, it is recommended to stop any further operations, in order to avoid any damage with surgical manipulation, and to perform a cholangio-MRI to define the anatomical situation.

We present here a real clinical case of GA treated in an Italian hospital in 2016 with the related medicolegal issues on the good clinical practice and informed consent.

Case report

In august 2016, a 31-years-old woman accessed to the Emergency Room for abdominal pain and, after a brief observation and analgesic therapy, was discharge. A few weeks later, the patient underwent two abdominal ultrasounds with two different diagnosis: the first reported that the gallbladder was not visible; the second described a poorly evaluated and contracted gallbladder, with some calculus formations inside.

A few months later, the patient had severe abdominal pain and twice accessed to the Emergency Room. During the second hospitalization, the patient presented continuous pain, Blumberg's and Murphy's signs were negative and peristalsis was valid. Therefore, she also underwent a surgical examination, which diagnosed recurrent episodes biliary colic, and a third abdominal ultrasound that did not find gallbladder or other alterations of the bile ducts. Based on these persistent symptoms, the patient was presumed to have symptomatic cholelithiasis and chronic cholecystitis. Therefore, a laparoscopic cholecystectomy for biliary stones was proposed to the patient and she was made to sign the relative informed consent form with these premises.

A laparoscopic cholecystectomy with a pre-operative cholangio-MRI, which did not detect the gallbladder, was planned (Figure 1), despite this, surgery is performed. During the laparascopic exploration of the abdominal cavity, the surgeons did not find the gallbladder in the usual place; therefore, in suspicion of an intrahepatic sclero-atrophy, they decided to convert the surgery into a laparotomy. After opening the peritoneum, they performed an intraoperative cholangiography, which confirmed the GA (Figure 2). A few days later, the patient underwent an eesophagogastroduodenoscopy that found an esophagitis with non-erosive antral gastropathy, compatible with previous abdominal symptoms. Therefore, the patient was discharged with reflux terapy. Subsequently, the patient opens a dispute for professional liability regarding the incorrect indication to surgery and the false information provided regarding the pathology in the informed consent.

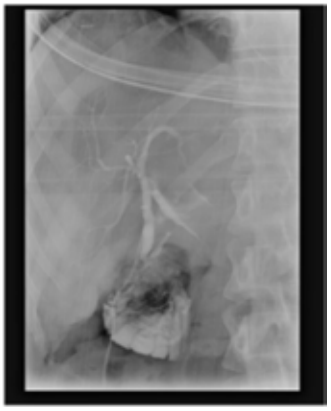

Figure 1: Preoperative RM: regular biliary tree and non-visualization of the gallbladder.

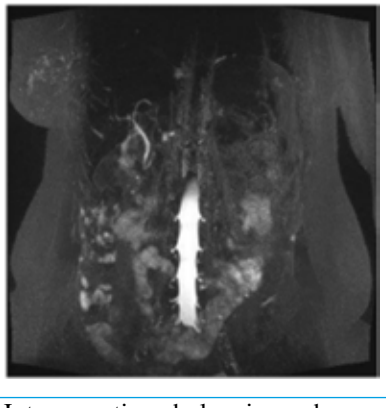

Figure 2: Intraoperative cholangiography: nonvisualization of the gallbladder

Discussion

The case concerns a suspected diagnosis of "recurrent biliary colic with cholecystitis" characterized by discontinuous and nonspecific abdominal pain without signs of cholestasis, inflammation or vomiting. Due to these symptoms, the patient underwent three abdominal ultrasounds, which were negative for gallbladder diseases, in particular only an ultrasound finds the colecisti and showed an uncertain presence of stones to investigate. The nonvisualization of the gallbladder, even with the execution of a cholangio-MRI (high visibility test) has been interpreted as a "scleroatrophic gallbladder" (an anatomic variant difficult to detect with ultrasound).

Because of these negative imaging results, it is difficult to understand why physicians have suggested performing a laparoscopic cholecystectomy and

confirmed the previous consent form for lithyasic cholecystitis[4], as also described in the intervention report.

In fact, the diagnostic tests had to be sufficient to diagnose the GA or, at least, to rule out inflammation or calculi of the gallbladder.

However, only subsequent laparoscopic abdominal exploration, laparotomy surgery and intraoperative cholangiography allowed the detection of GA, excluding any type of gallbladder disease, which was previously suspected. In connection with this sequence of events, some incorrect healthcare decisions can be highlighted.

The scientific literature describes that a high index of suspicion is needed in the interpretation of radiological images for GA. In fact, although ultrasound is not an accurate GA detection system[5], due to its low sensitivity, the inability to visualize the gallbladder should raise some doubts about its hypothetical disease. In addition, accidental detection of GA should be evaluated with cholangio-MRI, but not with surgery and, if GA is diagnosed during a laparoscopy, surgery should be stopped to prevent iatrogenic injuries[6-7].

In western countries in recent decades, we have witnessed an increase in disputes for medical professional liability in many specialist disciplines[8-12] with negative repercussions on medical assistance activities, for this reason it is necessary to act in order to prevent the occurrence of these events through the knowledge of some useful general indications to clinical practice[13-16].

In contrast, in the present case, surgeons have proposed and performed laparoscopic surgery based on a diagnostic suspect not supported by any imaging tests. In particular, the result of cholangio magnetic resonance imaging, which is the best non-invasive way to diagnose congenital alteration[17-19] has been ignored. In the subsequent dispute for malpractice it was noted above all that the information given to the patient was misleading in fact the state of the sensing had not been explained but the intervention for calculations in the gallbladder had been proposed when instead there was no evidence that the organ existed.

Therefore, the consent to the intervention was not valid because it was vitiated by false information. Furthermore, the conversion of the laparotomy was contrary to the consent given by the patient, who had been approved only for emergencies and was not looking for a missing organ.

In Italy, informed consent was recently regulated by Law 219 of 2017 which requires that the patient be given complete and updated information on his or her health status.

Conclusion

Gallbladder agenesis is a rare condition that can present nonspecific symptoms similar to other common pathological situations in internal medicine and surgery. It is necessary that all doctors who approach the suspicion of lithiasis cholecystitis are aware of this aspect as performing surgery in these cases, as well as not being useful, can be risky for anatomical damage and unnecessary scarring. In patients where US preoperative imaging is equivocal that an MRCP should be obtained and if this is negative one should not proceed with surgery. In addition to this it is necessary to remember that the patient must be provided with complete and truthful information and not only useful for signing consent.

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