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Case Report

Case Study and Bibliographic Review of Gallbladder Agenesis

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Abstract

Gallbladder agenesis (GA) is a rare congenital biliary system anomaly often associated with other congenital anomalies. Only about 400 cases of GA are reported in the literature.

Whilst most patients remain asymptomatic, about 50% present a symptomatology that resembles lithiasis biliary pain. The first examen in case of suspected biliary pathology is based on hepato-biliary ultrasound which is not very effective in the diagnosis of biliary tree malformation. GA is often mistaken for other hepato-biliary diseases, particularly sclero-atrophic gallbladder, which leads to unnecessary and sometimes dangerous surgical interventions.

We report the case of a 13-years-old female patient with chronic biliary-type pain in whom agenesis of the gallbladder was suspected on abdominal ultrasound and confirmed by Magnetic resonance cholangiopancreatography (MRCP).

Keyword: *Gallbladder; Agenesis; Cholangiopancreatography; Magnetic Resonance*

Introduction

The agenesis of the gallbladder and cystic duct represents one of the rarest anomalies of the biliary system.

To date, only about 400 cases of GA are reported in the literature [1,2].

This agenesis is attributed to an embryonic developmental abnormality, most likely related to a genetic mechanism. Most cases of GA are associated with other congenital anomalies. These are present mainly at birth and most of them are lethal during the first year of life. As a result, only patients with isolated GA are seen in adulthood [3].

Furthermore, the GA can occasionally be associated with other congenital anatomical anomalies or congenital syndromes such as cerebrotendinous xanthomatosis, Klippel-Feil syndrome, trisomy 18, and following exposure to thalidomide [2].

We report the case of a 13-years-old female patient with chronic biliary-type pain in whom agenesis of the gallbladder was suspected on abdominal ultrasound and confirmed by MRCP.

Patient Observation

A 13-year-old patient with no previous history. The patient presented with intermittent biliary pain, on a background of moderate and diffuse abdominal pain evolving for one year. The biological workup was normal, the liver workup showed no cytolysis or cholestasis. Her clinical examination was normal.

An abdominal ultrasound is performed as part of the exploration of his pain showed moderate hepatomegaly and the gallbladder was not visualized this examination. The diagnosis of gallbladder agenesis was suspected on this examination and given the rarity of the diagnosis; magnetic resonance imaging (MRCP) was performed (Figure 1). This allowed the diagnosis of gallbladder agenesis. The caliber of the main bile duct was at the upper limit of normal, without stones.

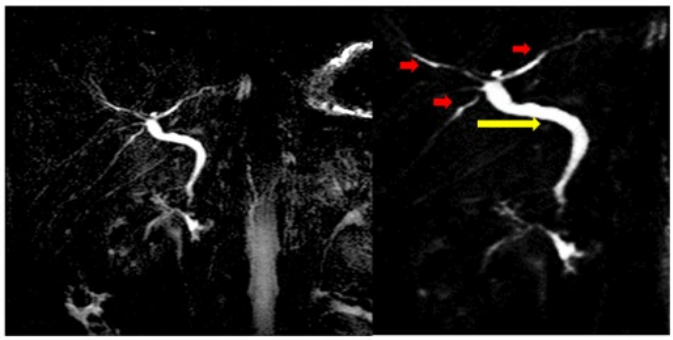

Figure 1: *Cholangio-MRI shows the absence of the gallbladder and cystic duct with normal calibre of the bile duct (yellow arrow) in its proximal part. The intrahepatic bile ducts (red arrows) are of normal calibre.*

Discussion

The gallbladder originates in the cystic bud, which develops as a ventral outgrowth of the caudal region of the foregut during the fourth week of life in utero. During the seventh week of development, vacuolation of the hyperplastic epithelium occurs. The gallbladder and cystic duct develop a lumen. It has been hypothesized that an abnormality at any stage of this process results in agenesis of the gallbladder [4] sometimes associated to other congenital malformations such as extrahepatic biliary atresia, imperforate anus, cardiovascular anomalies, intestinal malformations, etc. [5].

Inappropriate migration of the gallbladder during embryonic development leads to the formation of an ectopic gallbladder, usually intrahepatic and left, between the leaflets of the lesser omentum, in the falciform, retropancreatic and retroduodenal ligament [6].

For unclear reasons, despite the absence of the gallbladder, up to 50% of patients present with symptoms similar to biliary pain [7]. Some suggest that associated with sphincter Oddi dysfunction, may be the cause of biliary pain in these patients [8,9]. In other cases, the development of bile ductal or intrahepatic stones may be the cause [10].

In this context, Bennion., *et al*. [11] have established a classification that distinguishes 3 groups:

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

b) Asymptomatic group (31.6%): This group of patients without a gallbladder is most often discovered during the autopsy or during a surgery performed for other reasons. None of these patients have any biliary symptoms.

c) Group with clinical manifestations, symptomatic form (55.6%): this group corresponds to the age group 40 - 50 years, usually without other congenital anomalies.

The preoperative diagnosis of gallbladder agenesis is not always easy to establish because of the rarity of this anomaly. Ultrasound is the initial reference examination for the exploration of biliary pathology [12], however there are false positive cases of gallbladder agenesis that can be explained by the interposition of small intestines in the vesicular fossa [13], the interposition of peri-portal peritoneal folds [12] or by foci of hepatic calcifications [14]. Non-visualization of a gallbladder on ultrasound and CT scan requires confirmation of the diagnosis of gallbladder agenesis by a more powerful examination, such as Cholangio-MRI.

Endoscopic retrograde cholangio-pancreatography usually conclude to obstructions of the cystic duct, without reporting agenesis of the gallbladder [9].

Currently, Bili-MRI is the gold standard for detecting gallbladder agenesis [10,15]. It is a non-invasive imaging examination that allows to find a gallbladder in an ectopic site (in the falciform ligament, in the lesser omentum, in the pancreas, behind the duodenum, in the pyloric digestive wall, or even intrahepatically), and in its absence on this examination, it allows to make the positive diagnosis.

Conclusion

Agenesis of the gallbladder is the least common malformation of the bile ducts, due to an aberration of embryological development that may be associated with other congenital anomalies. It is mostly discovered isolated in adulthood. The diagnosis should be suspected when there is no visualization of the gallbladder on ultrasound or most often when there is a sclero-atrophic appearance. The MRCP allows to confirm the diagnosis with certainty.

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