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Case Report

Gallbladder Agenesis in Adults Presenting with Chronic Biliary Pain

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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 68-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).

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

Introduction

Gallbladder and cystic duct agenesis is a rare anomaly of the bile ducts; only a few cases described in the literature, not exceeding 400 cases [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 anatomical 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 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 68-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 68-year-old patient with a history of hypertension, diabetes, gout, and no history of abdominal surgery.

The patient presented with intermittent biliary pain, on a background of moderate and diffuse abdominal pain evolving for one year. The biological work-up was normal, the hepatic work-up did not show cytolysis nor cholestasis. Her clinical examination was normal.

A hepato-biliary ultrasound done in the context of exploration of his pain showed hepatic steatosis, moderate hepatomegaly and the gallbladder was not visualized during this examination. A CT scan confirmed the absence of individualization of the gallbladder with the presence of a small oblong structure of aerial content continuing with the gastroduodenal region. 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 (5 mm), with no stones.

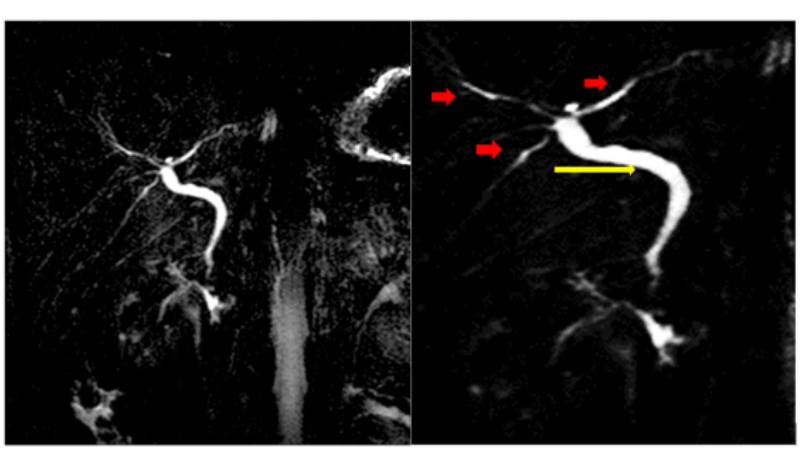

Figure 1: *MRCP shows the absence of the gallbladder and cystic duct with a normal caliber of the bile duct (yellow arrow) 3.8 mm in its proximal part. The intrahepatic bile ducts (red arrows) have a normal caliber.*

Discussion

The epithelium of the bile ducts takes origin from endodermal: the gallbladder and the common bile duct derive from the bile bud, during the fourth week of life in utero. during the seventh week of development, vacuolation of the endodermal bile bud is noted. thus, the gallbladder and the cystic duct develop a lumen. any abnormality at any stage of this process can cause cessation of gallbladder development [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 like biliary pain [7]. some suggest that biliary pain is secondary to sphincter Oddi dysfunction which can be seen in this context [8,9]. In other cases, the development of bile ductal or intrahepatic stones may be the cause [10].

Patients with gall bladder agenesis are classified into 3 categories [11]:

a) **Multiple fetal anomaly (12.9%):** Gallbladder agenesis is associated with multiple congenital malformations, mainly cardiovascular, gastrointestinal, and Genito-urinary. These malformations are often complex, leading to die during the perinatal period.

b) **Asymptomatic group (31.6%):** The diagnosis is made fortuitously during surgery for another reason. patients have no biliary symptoms.

c) **Group with clinical manifestations, symptomatic form (55.6%):** The diagnosis is often made in adulthood between 40 and 50 years old, the agenesis of the gallbladder is isolated without other associated malformations.

Gallbladder agenesis is a diagnosis poorly understood by clinicians because of its rarity.

Ultrasound is the first-line examination for the exploration of gallbladder [12], but false positive may exist following the interposition of the small intestine, peritoneal folds or the presence of hepatic calcifications in the gallbladder fossa [12-14]. 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, Cholangio-MRI is the gold standard for detecting gallbladder agenesis [10,15].

Cholangio-MRI remains the examination of choice for the positive diagnosis of gallbladder agenesis, and to diagnose an ectopic gallbladder (in the falciform ligament, in the lesser omentum, in the pancreas, behind the duodenum, in the pyloric, even intrahepatic digestive wall).

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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