

Accessory gallbladder in an intrahepatic location mimicking a cystic tumor of the liver

A case report

Ji-Hye Won, MD^a, Seo-Youn Choi, MD^{a,*}, Hae Kyung Lee, MD^a, Boem Ha Yi, MD^a, Min Hee Lee, MD^a, Min Jung Jung, MD^b

Abstract

Background: Double gallbladder (GB) is a rare congenital anomaly of the biliary system characterized by the presence of an accessory GB.

Clinical Findings: A 38-year-old female presented with a history of right upper quadrant (RUQ) pain. Computed tomography (CT) showed a lobulated cystic mass involving the center portion of liver. Magnetic resonance imaging (MRI) additionally revealed a tubular structure of T2 bright signal intensity (SI), which connected the cystic lesion of the liver to the bile duct. Preoperative endoscopic retrograde cholangiopancreatography (ERCP) confirmed the connection between the intrahepatic cystic lesion and the left main hepatic duct. We regarded as a cystic mass with biliary communication, such as a cystic intraductal papillary neoplasm of the bile duct (IPNB) or localized Caroli disease. The patient underwent partial hepatectomy for the cystic mass of the liver and a final diagnosis of chronic inflammation of an intrahepatically located accessory GB was made.

Conclusion: We report a case of an accessory GB in an intrahepatic location mimicking a cystic mass such as cystic IPNB or localized Caroli disease of the liver showing a cystic mass with biliary communication.

Abbreviations: ALP = alkaline phosphatase, CRP = C reactive protein, CT = computed tomography, ERCP = endoscopic retrograde cholangiopancreatography, GB = gallbladder, GGT = gamma-glutamyl transferase, IPNB = intraductal papillary neoplasm of the bile duct, MRI = magnetic resonance imaging, RUQ = right upper quadrant, SI = signal intensity, US = ultrasound, WBC = white blood cell count.

Keywords: case report, gallbladder, liver neoplasm, magnetic resonance imaging, multidetector computed tomography

1. Introduction

Double gallbladder (GB) is a rare congenital anomaly of the biliary system characterized by the presence of an accessory GB.^[1] In double GB, the duplicate organs typically adjoin each

other, and the accessory GB may be adjacent to the normal organ in the GB fossa. Although the incidence of ectopically located accessory GBs remains unclear, accessory GBs might remain intrahepatic, in subhepatic locations, or within the gastrohepatic ligament.^[2–4] Preoperative diagnosis of this anomaly is especially important to prevent repeated surgery and unexpected postoperative complications in patients. Here, we report a case of an accessory GB in an intrahepatic location mimicking a cystic mass of the liver on computed tomography (CT) and magnetic resonance imaging (MRI) with magnetic resonance cholangiopancreatography (MRCP). The findings were regarded as a cystic mass with biliary communication, such as a cystic intraductal papillary neoplasm of the bile duct (IPNB) or localized Caroli disease.

2. Case report

A 38-year-old female presented with right upper quadrant (RUQ) pain, which she had for a duration of 2 weeks. She had no serious medical or surgical history and grew up in normal residential and social environments. Her vital signs were unremarkable. Physical examinations revealed mild tenderness in the RUQ abdomen. Laboratory values revealed an elevated erythrocyte sedimentation rate (ESR) and gamma-glutamyl transferase (GGT) associated with normal values of white blood cell count (WBC), C reactive protein (CRP), total bilirubin, transaminases, and alkaline phosphatase (ALP). An ultrasound (US) of the upper abdomen was performed for the first step of the imaging examination, and acute cholecystitis was suspected with findings

Editor: Abd Elrazek Mohammad Ali Abd Elrazek.

Funding: This work was supported by the Soonchunhyang University Research Fund.

Ethical statement: This study was approved by the institutional review board of the Soonchunhyang University Hospital. It has been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments. The institutional review board of this university waived the need to obtain informed consent.

The authors have no conflicts of interest to disclose.

^a Department of Radiology, ^b Department of Pathology, Soonchunhyang University, College of Medicine, Bucheon Hospital, Bucheon, Korea.

* Correspondence: Seo-Youn Choi, Department of Radiology, Soonchunhyang University, Bucheon Hospital, 170 Jomaru-ro, Wonmi-gu, Bucheon-si, Gyeonggi-do 420-767, South Korea (e-mail: sychoi@schmc.ac.kr)

Copyright © 2016 the Author(s). Published by Wolters Kluwer Health, Inc. All rights reserved.

This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial-No Derivatives License 4.0 (CCBY-NC-ND), where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially without permission from the journal.

Medicine (2016) 95:44(e5293)

Received: 22 August 2016 / Received in final form: 30 September 2016 /

Accepted: 11 October 2016

<http://dx.doi.org/10.1097/MD.0000000000005293>

including distended GB, diffusely thickened GB wall, sludge, and a sonographic Murphy sign. In addition, as an incidental finding, a lobulated anechoic mass with hyperechoic debris in a dependent area was discovered in the central portion of the liver, just above the GB fossa. On CT, a 5.3-cm-sized lobulated cystic mass involving the central liver with fluid–fluid levels and amorphous high-density materials was found in a dependent portion, suggesting either hemorrhage or sludge (Fig. 1A–C). Furthermore, on MRI, a tubular structure of T2 bright signal intensity (SI), which connected the cystic lesion of the liver to the bile duct, was additionally suspected, and the wall of the cystic lesion seemed to be more thickened than in the case of a simple cyst (Fig. 1D–E). Consequently, the findings were regarded as a cystic mass with biliary communication, such as a cystic IPNB or localized Caroli disease. Preoperative endoscopic retrograde cholangiopancreatography (ERCP) confirmed the connection between the intrahepatic cystic lesion and the left main hepatic duct (Fig. 1F). Based on these findings, the patient underwent cholecystectomy for acute cholecystitis and partial hepatectomy for the cystic mass of the liver. On pathologic examination of a surgical specimen, the cystic lesion of the liver was revealed to have chronic inflammation, and was lined with simple columnar epithelium and smooth muscle layer, which is a characteristic manifestation of chronic cholecystitis (Fig. 1G). Only at that point was the hepatic lesion confirmed to represent chronic inflammation of an intrahepatically located accessory GB. Additionally, the main GB (located in the normal GB fossa) was also confirmed as having chronic cholecystitis with Rokitansky–Aschoff sinuses. The patient was discharged after surgical resection without complications,

3. Discussion

Double GB is a rare congenital anomaly of the biliary system associated with an accessory GB occurring in about 1 of 4000

births. It is a result of a rare embryonic diverticulum of the hepatic duct, which occurs during the fifth or early in the sixth week of embryologic development.^[1] The classifications of Boyden and Harlaftis are widely used for categorization of various congenital abnormalities of GBs. According to Harlaftis classification, type 1 abnormalities comprise a split primordial group of GBs, subdivided into septate, V-shaped and Y-shaped types, similar to the vesica fellea divisa (bilobed GB) and Y-shaped type of vesica fellea duplex (true duplication), which are designated in Boyden classification system. Type 2 abnormalities involve an accessory GB that is an H or ductular type, which entails 2 separate GBs together with cystic ducts connecting the organs to a common bile duct (as in the H-shaped type of the vesica fellea duplex of Boyden), and a trabecular type that contains accessory cystic ducts connecting to the left or right hepatic duct. In addition, type 3 abnormalities include any abnormalities that cannot be categorized into the aforementioned 2 types, not being classified by Boyden et al. Our case represents the trabecular type of accessory GB with an accessory cystic duct inserting into the left hepatic duct.^[1,5]

Ectopic positions of the GB are of great clinical significance because ectopic positions potentially create repeated surgeries and unexpected complications in patients. Although various locations of accessory or malposed GBs have been reported, the exact incidence of ectopic locations of accessory GBs remains unclear. Ectopic GBs might be located in positions that are intrahepatic, subhepatic, or within the gastrohepatic ligament, wherein the accessory organ is close to the main GB.^[2–4] To the best of our knowledge, only a few case reports of intrahepatic GB have been reported. Accessory organs can be detected incidentally by US and radionuclide scanning, or by complications such as gallstones and cholecystitis, and are ultimately confirmed by follow-up cholecystectomy.^[6,7,8]

Controversy remains as to whether the presence of a duplicate GB is a predisposing factor for gallstone formation and

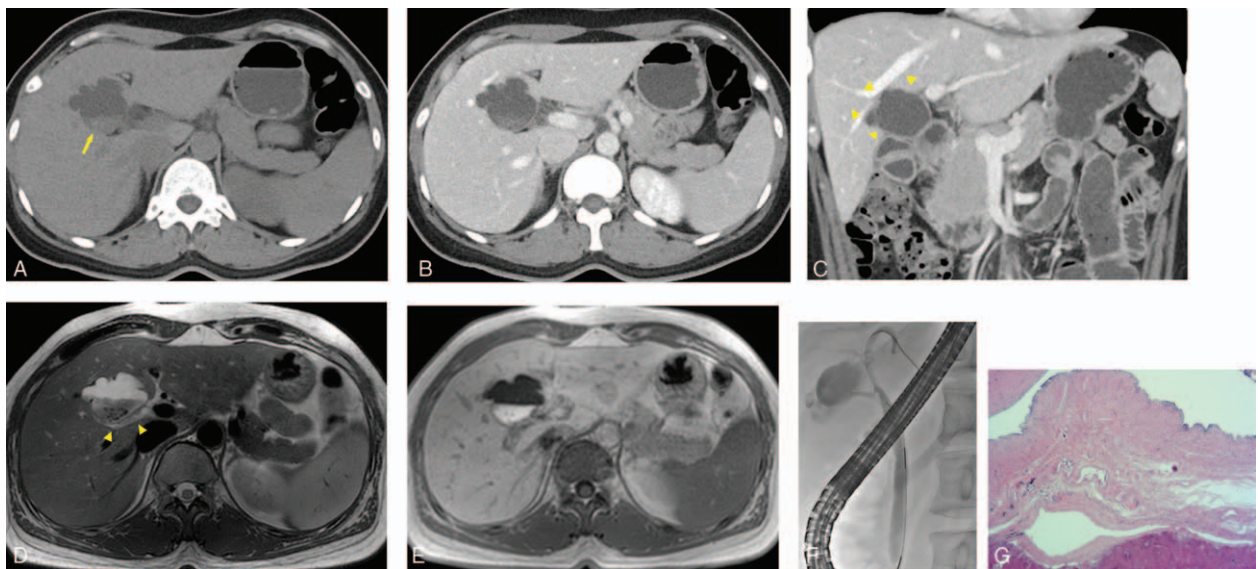


Figure 1. Abdomen CT (A–C), MRI (D and E), ERCP (F), and histopathology (G) of a double gallbladder with intrahepatic location in a 38-year-old female. (A–C) Precontrast (A) and contrast-enhanced abdomen CT axial (B) and coronal (C) scans show a 5.3-cm-sized lobulated cystic mass involving the central liver with fluid–fluid levels and amorphous hyperdense materials floating in a dependent portion (arrow in A). The cystic mass of the liver is located just above the original GB (arrowheads in C). (D and E) Axial T2-weighted (D) and contrast-enhanced T1-weighted (E) MRI reveal a tubular structure (arrowheads in D) connecting the thick-walled cystic lesion of the liver and the left hepatic duct. (F) Endoscopic retrograde cholangiopancreatography (ERCP) shows a connection between the intrahepatic cystic lesion and the left main hepatic duct. (G) The presence of a muscular layer with inflamed mucosa is focally identified on a surgical specimen of the cystic mass in the liver (H&E, ×40), which is a characteristic feature of an accessory gallbladder.

inflammation. Several previous reports suggest that malformations of the biliary system can be associated with hypokinesia of the GB.^[9,10] Although the underlying reason is not clear, degradation of the contractile function of an accessory GB in comparison with a normal GB would contribute to this result. Additionally, we surmise that this phenomenon would be further complicated when the location of the accessory GB is intrahepatic.

The differential diagnosis for the cystic mass of the liver with bile duct communication, as observed herein, could include a cystic variant of IPNB or localized Caroli disease. According to the WHO classification system, the former is characterized by the presence of an intraluminal papillary tumor with biliary mucinous cystic neoplasm or oncocytic epithelium without ovarian-like stroma.^[11] Rarely, mucin overproduction, recurrent hemorrhage, and neoplastic overgrowth of IPNB may cause cystic changes of IPNB.^[12] The work of Lloyd^[7] and Lim et al^[8] reports that 12 cases of cyst-forming IPNBs might arise from peribiliary glands showing diverticulum or balloon-like dilatation with a multilobulated appearance. The latter, also known as type V choledochal cyst in the Todani classification, is defined as cystic congenital cavernous ectasia of the biliary tract. Caroli disease usually involves saccular dilated intrahepatic cystic structures, which communicate with the biliary tree in ways that are either localized or diffuse. In our case, the intrahepatic cystic lesion seemed to have a thicker wall than the wall of a simple cyst, a lobulated appearance rather than a unilocular appearance, sludge, and communication with the biliary tree. Based on these imaging features, the accessory GB was mistaken for a cystic variant of IPNB or localized Caroli disease. Looking retrospectively at the imaging findings, an accessory GB in an intrahepatic location should in fact be included in the differential diagnosis for cystic lesions of the liver. Although it is rare, radiologists should consider the possibility of ectopic intrahepatic GB in differential diagnosis when a cystic lesion is discovered in the liver near the GB fossa.

We report a rare case of inflamed accessory GB with an intrahepatic location mimicking a cystic tumor of the liver. Although the incidence of double GB is extremely rare, when a cystic lesion of the liver with biliary communication is discovered near the GB fossa, we the possibility of double GB should be considered in addition to cystic neoplasms of the liver. Furthermore, it is conceivable that the knowledge of this anomaly may prevent repeated surgeries and unexpected complications in patients.

References

- [1] Boyden EA. The accessory gall-bladder-an embryological and comparative study of aberrant biliary vesicles occurring in man and the domestic mammals. *Am J Anat* 1926;38:177-231.
- [2] Cozacov Y, Subhas G, Jacobs M, et al. Total laparoscopic removal of accessory gallbladder: a case report and review of literature. *World Journal Gastrointest Surg* 2015;7:398-402.
- [3] Sahoo MR. Double gallbladder masquerading as a choledochal cyst: a case report. *Int J Case Rep Images (IJCRI)* 2013;4:283-6.
- [4] Gross RE. Congenital anomalies of the gallbladder: a review of one hundred and forty-eight cases, with report of a double gallbladder. *Arch Surg* 1936;32:131-62.
- [5] Harlaftis N, Gray S, Skandalakis J. Multiple gallbladders. *Surg Gynecol Obstet* 1977;145:928-34.
- [6] Schmammann J, Dent D, Mervis B, et al. Cholecystitis in an intrahepatic gallbladder. A case report. *S Afr Med J* 1982;62:1042-3.
- [7] Lloyd TV. Empyema of an intrahepatic gallbladder. *Clin Nucl Med* 1979;4:341.
- [8] Lim JH, Zen Y, Jang KT, et al. Cyst-forming intraductal papillary neoplasm of the bile ducts: description of imaging and pathologic aspects” *AJR. American Journal of Roentgenology* 2011;197:1111-20.
- [9] Hardoff R, Hardoff D. Scintigraphic evaluation of septated hypokinetic gallbladder in a teenager. *Clin Nucl Med* 1990;15:117-8.
- [10] Shapiro T, Rennie W. Duplicate gallbladder cholecystitis after open cholecystectomy. *Ann Emerg Med* 1999;33:584-7.
- [11] Bosman FT, Carneiro F, Hruban RH, et al. WHO Classification of Tumours of the Digestive System. WHO/IARC:World Health Organization; 2010.
- [12] Kakisaka T, Kamiyama T, Yokoo H, et al. An intraductal papillary neoplasm of the bile duct mimicking a hemorrhagic hepatic cyst: a case report. *World J Surg Oncol* 2013;11:111.