

A Choledochal Cyst Resulting in Obstructive Jaundice in a Case with Gallbladder Agenesis: Report of a Case and Review of the Literature

Abdullah Büyük¹, Cüneyt Kırkıl², Koray Karabulut², Selim Sözen³, Salih Demirelli³

¹Department of General Surgery, Faculty of Medicine, Dicle University, Diyarbakır, Turkey

²Department of General Surgery, Faculty of Medicine, Fırat University, Elazığ, Turkey

³Department of General Surgery, Elazığ Research and Educational Hospital, Elazığ, Turkey

ABSTRACT

Co-existence of gallbladder agenesis and choledochal cyst is a very rare congenital anomaly of the extrahepatic biliary system. The literature presents only five cases with this combination. Herein, we report the sixth case. Gallbladder agenesis is usually asymptomatic, while choledochal cyst often presents symptoms before adolescence. This is the report of a 42-year-old female patient with gallbladder agenesis and choledochal cyst leading to obstructive jaundice. Radiological diagnosis of choledochal cysts is not difficult. However, in cases with gallbladder agenesis, cystic dilatation in the choledochus may be misdiagnosed as mislocation of the gallbladder or contracted and/or sclero-atrophic gallbladder.

Key word: Gallbladder agenesis, choledochal cyst, obstructive jaundice

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Introduction

Agenesis of the gallbladder (AGB) and choledochal cyst (CC) are among the rare conditions of congenital extrahepatic biliary tree anomalies. AGB results from the failure of the cystic bud to develop in the 4th week of intrauterine life. It normally develops from the caudal part of the hepatic diverticulum (1, 2). CCs are single or multiple dilatations of the intrahepatic or extrahepatic biliary tree. Here, we report choledochal cyst complications presenting as obstructive jaundice in a patient with gallbladder agenesis. This combination is very rare and a review of literature revealed only five such cases (3, 4).

Case Report

A 42-year-old woman was admitted to the emergency department with the complaints of unrelenting pain in the right upper abdomen, post-prandial nausea, vomiting and loss of weight for the previous 2 months. She had a medical history of severe and colicky right upper abdominal pain accompanied by intolerance to fatty foods for the previous 2 years. Moreover, she had jaundice for 10 days. On the abdominal examination, no masses or organomegaly was noted, but there was mild tenderness in the epigastric and right upper quadrant areas without rebound or rigidity. In the clinical examination, the patient was jaundiced and cachectic.

The results of the laboratory tests were as follows: white blood cells: $10.2 \times 10^3/\text{mCL}$, hemoglobin: 12.3 g/dL, blood

urea nitrogen: 16 mg/dL (normal range: 1-50 mg/dL), creatinine: 0.6 mg/dL (normal range: 0.5-1.2 mg/dL), aspartate aminotransferase (AST): 337 U/L (normal range: 1-38 U/L), alanine aminotransferase (ALT): 339 U/L (normal range: 1-41 U/L), alkaline phosphatase (ALP): 385 U/L (normal range: 35-129 U/L), gamma-glutamyltranspeptidaz (GGT): 830 U/L (normal range: 5-61 U/L), total bilirubin 7.4 mg/dL (normal range: 0-1.2 mg/dL), and direct bilirubin: 5.2 mg/dL (normal range: 0-0.2 mg/dL).

Ultrasonography (US) showed minimal dilatation of intrahepatic bile ducts. The gallbladder was located in the hilum of the liver, not in its normal anatomic location, and multiple millimetric stones were detected. The abdominal computerized tomography (CT) revealed an ectopic gallbladder containing tiny stones (Figure 1). In the endoscopic retrograde cholangiopancreatography (ERCP), intrahepatic bile duct dilatation, ectopic gallbladder with location anomaly of the ductus cysticus were visible. In the same session, bile duct stone extirpation was performed using ERCP.

The patient underwent laparotomy, during which the gallbladder was not found despite a meticulous search of the abdomen, either by direct vision or by the use of the intraoperative ultrasound probe. On the other hand, a Type 1 CC was detected based on the Todani classification of choledochal cysts (Figure 2) (5). The diagnosis was confirmed through intraoperative cholangiography (Figure 3). Cyst excision with Roux-en-Y hepaticojejunostomy was performed. Histologically, the wall of the common bile duct was composed of chronically inflamed fibrous tissue with no evidence of malignancy.

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Address for Correspondence: Dr. Abdullah Büyük, Department of General Surgery, Faculty of Medicine, Dicle University, Diyarbakır, Turkey
Phone: +90 412 248 80 01 E-mail: azboyuk@hotmail.com



Figure 1. Millimetric densities compatible with stone formations in the liver hilus

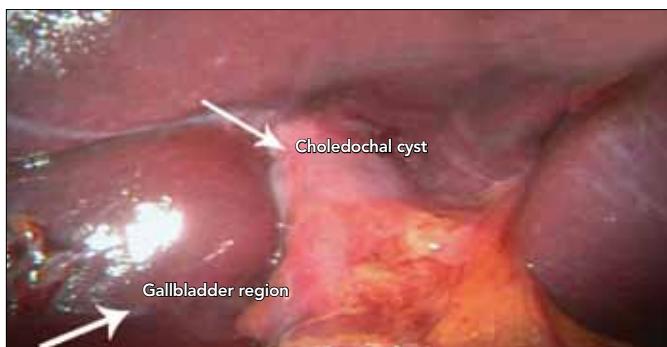


Figure 2. Gallbladder agenesis and choledochal cyst

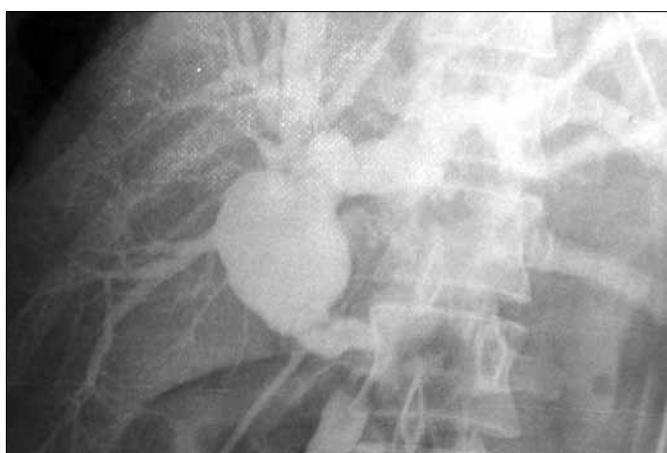


Figure 3. Gallbladder agenesis and choledochal cyst on the cholangiography

Table 1. Classification of choledochal cysts

Type I	Dilatation of hepatic and common bile ducts (comprising 80-90% of cases)
Type II	Diverticulum of the common bile duct
Type III	Intraduodenal common bile duct dilatation (termed choledochocele)
Type IV -a	Intrahepatic and extrahepatic bile duct dilatation
Type IV- b	Multiple extrahepatic cysts
Type V	Intrahepatic bile duct dilatation (as in Caroli's Disease)

The patient had cholangitis on the 60th postoperative day and was treated medically. In the 8-month follow-up period, the patient was symptom free.

Discussion

Congenital anomalies of extrahepatic biliary systems are rare. These anomalies may be isolated or combined (3). Bergman (6) was the first author to describe this anomaly in human beings in 1701. AGB is a rare congenital biliary anomaly with an incidence of 0.01-0.06%. In a recent review of autopsies, the incidence of AGB has been reported to be about 1/6334 live births (7).

Despite having no characteristic symptomatology, possible mechanisms of AGB symptoms have been described as primary duct stones, biliary dyskinesia, or non-biliary disorder. While many adults with AGB remain asymptomatic for life, about 23% of affected individuals present with biliary symptoms, including 90% with right upper quadrant pain, 60% with nausea and vomiting, 37% with food intolerance, 35% with jaundice, and 30% with dyspepsia (3).

The diagnosis of AGB is difficult with routine investigations and may be misleading, particularly in cases of contracted or ectopic gallbladder. Considering these reasons, in 1967 Frey et al. (8) suggested laparotomy as the only method of diagnosis for AGB. Similarly, in our patient, absence of gallbladder could not be diagnosed preoperatively, but it was determined during the laparotomy. This was evaluated as an abnormal location of the gallbladder because of the presence of a CC. The AGB is usually diagnosed incidentally during investigation or surgery for another disease. Tabibian et al reported a co-existence of AGB and CC in a patient who underwent surgery for duodenal obstruction. We determined similarly a AGB coexisting with CC in a patient who underwent surgery for obstructive jaundice. If the diagnosis of AGB is made during surgery, the surgeon must prove AGB by thoroughly examining the most common sites for ectopic gallbladder, which are intrahepatic, retrohepatic, on the left side, within the leaves of the lesser omentum or within the falciform ligament, retro-duodenal, retropancreatic and retroperitoneal (9).

Choledochal cyst (CC) is another congenital anomaly of the biliary tree. It was first defined by Vater and Ezler in 1723 (3). Whereas CC is a rare medical condition with an incidence of 1 in 100.000-150.000 live births in the western population, it is remarkably higher in Asian populations with a reported incidence of 1 in 1000, and about two thirds of cases in Japan (10). CC is more common in women, with a male to female ratio of 1:3 to 4 (11). Alonso-Lej et al. established the first classification

system for CC in 1959, followed by Todani et al. modifying this system in 1977. Now, the classification system of Todani et al is the most commonly used by clinicians (Table 1) (5).

Clinical symptoms can develop at any time; however, 80% of patients present symptoms before the age of 10 years. Only a minority of patients present with the classic clinical triad of abdominal pain, jaundice, and abdominal mass. The clinical presentation mostly depends on age, with abdominal pain as the most frequent presenting symptom in adults and jaundice in infants (11). Our patient, who was asymptomatic until the age of 42, had a choledochal cyst that caused episodic abdominal pain, jaundice, nausea, and vomiting for the previous 2 months.

Presentation, diagnosis, and prognosis of CC largely depend on its size and location, and its type. Our patient had coexisting CC and AGB; therefore, preoperative imaging methods were of limited use.

Complete cyst excision and Roux-en-Y hepaticojejunostomy is the standard procedure for Type I CC. Our patient underwent total cyst excision and hepaticojejunostomy with laparotomy.

Complications of CC, which are the result of stasis, include cholangitis, stone formation, recurrent pancreatitis, cirrhosis, and portal hypertension. Cholangiocarcinoma is another serious complication, with an even higher risk after drainage procedures (11). Particularly in case of Type I CC, to avoid the high risk of cholangiocarcinoma, complete cyst excision and Roux-en-Y hepaticojejunostomy has been advocated, as was performed in our patient. Operation should be performed as soon as the diagnosis of CC is established because of its serious complications, even when it is not symptomatic.

Radiological diagnosis of CC is not difficult. However, in cases with AGB, the cystic dilatation in the choledochus, as in our case, may be misinterpreted as location anomaly of the gallbladder (ectopic or in the hilum of the liver), or contracted and/or sclero-atrophic gallbladder, and it may lead to diagnostic challenges or misdiagnosis. Therefore, when the gallbladder is not detected in its normal location or size, AGB and CC coexistence should be kept in mind. In the determination of the etiology for jaundice and pancreatitis, particularly in young patients, the gallbladder anatomy should be well evalu-

ated and in the differential diagnosis, the potential presence of a congenital CC should be kept in mind.

Goel et al.(4) reported the fourth case of CC and AGB coexistence in 1994. Only one such case has been reported since 1994 to date (3). To our knowledge, the case reported here is the sixth case of CC and AGB coexistence.

Conflict of Interest

No conflict of interest was declared by the authors.

References

1. Fiaschetti V, Calabrese G, Viarani S, Bazzocchi G, Simonetti G. Gallbladder agenesis and cystic duct absence in an adult patient diagnosed by magnetic resonance cholangiography: report of a case and review of the literature. Case Report Med 2009;2009:674768.
2. Gotohda N, Itano S, Horiki S, Endo A, Nakao A, Terada N, et al. Gallbladder agenesis with no other biliary tract abnormality: report of a case and review of the literature. J Hepatobiliary Pancreat Surg 2000;7:327-30. [\[CrossRef\]](#)
3. Tabibian JH, Tabibian N, Aguet JC. Choledochal cyst complications presenting as duodenal obstruction in an 82-year-old patient with gallbladder agenesis. Dig Dis Sci 2009;54:184-7. [\[CrossRef\]](#)
4. Goel AK, Seenu V, Khosla NK, Gupta A, Sarda AK. Agenesis of gallbladder with choledochal cyst--an unusual combination. Trop Gastroenterol 1994;15:33-6.
5. Todani T, Watanabe Y, Narusue M, Tabuchi K, Okajima K. Congenital bile duct cysts: classification, operative procedure and review of 37 cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9. [\[CrossRef\]](#)
6. Latimer EO, Mendez FL, Hage WJ. Congenital absence of gallbladder: report of three cases. Ann Surg. 1947;126:229-42. [\[CrossRef\]](#)
7. Bennion RS, Thompson JE, Tompkins RK. Agenesis of the gallbladder without extrahepatic biliary atresia. Arch Surg 1988;123:1257-60. [\[CrossRef\]](#)
8. Frey C, Bizer L, Ernst C. Agenesis of the gallbladder. Am J Surg 1967;114:917-26. [\[CrossRef\]](#)
9. Bani-Hani KE. Agenesis of the gallbladder: difficulties in management. J Gastroenterol Hepatol. 2005;20:671-5. [\[CrossRef\]](#)
10. Singham J, Yoshida EM, Scudamore CH. Choledochal cysts: part 1 of 3: classification and pathogenesis. Can J Surg. 2009;52:434-40.
11. Chaturvedi A, Singh J, Rastogi V. Case report: Cholangiocarcinoma in a choledochal cyst. Indian J Radiol Imaging 2008;18:236-8. [\[CrossRef\]](#)