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Congenital Absence of Gallbladder

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Nine surgically proven congenital absence of gallbladder (CAGB) cases were reviewed. All of them had one or more kinds of biliary symptom. Tests such as abdominal ultrasonography, intravenous or oral cholecystography and even endoscopic retrograde cholangiography not only failed to predict CAGB but misleadingly indicated other similar conditions. Only the abdominal computed tomography (CT), performed on one patient, enabled the accurate diagnosis of CAGB. All the patients underwent abdominal exploration, and CAGB was confirmed by the meticulous dissection of the entire extrahepatic biliary tree and the operative cholangiography. Five patients had concomitant biliary pathologies responsible for their symptoms, but four patients had isolated CAGB. CAGB is a rarely encountered condition for a clinican, but extensive diagnostic work-ups including abdominal CT should be performed in all situations where CAGB is suspected. Thus unnecessary exploration can be avoided in the isolated CAGB case.

Key Word: Congenital absence of gallbladder

The biliary system shows relatively frequent anomalies; some (Stolkind, 1939) have reported incidences of anomalies in as many as thirty per cent of patients, so a surgeon should take care to determine any anomalous structure during a biliary operation.

But the congenital absence of gallbladder, not a part of extrahepatic biliary atresia (CAGB) is rare. Its incidence has been reported as about 0.01 to 0.04 % (Bennion *et al.* 1988). Fortunately most cases have been known to be asymptomatic and are found incidentally during extrabiliary operations or autopsies (Stolkind *et al.* 1929; Rabinovitch *et al.* 1958). But symptomatic CAGB has also been reported. Theoretically, conservative treatment is adequate for CAGB unless other biliary diseases are present but most symptomatic CAGB patients are subjected to an abdomin exploration because no imaging modality can predict CAGB definitely. Moreover, the nonvisualization of gallbladder disclosed by biliary imagings is thought to be an obstruction of cystic duct which is also the pathognomonic finding of acute cholecystitis.

Upon exploration of the abdomen, we discovered the absence of a gallbladder, without any preoperative information, and we thought the diagnostic dilemma and adequate treatment plans should be discussed for this rare condition.

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PATIENTS AND METHODS

Nine cases of CAGB without extrahepatic biliary atresia were confirmed by exploration at Yonsei University Hospital from 1969 to 1988. Their medical records were retrospectively reviewed with regard to symptoms, method of preoperative diagnosis and its accuracy, and, finally, the association between their operative findings and symptoms. All the patients had no other associated anomaly.

RESULTS

Clinical Manifestations

All the patients complained of one or more kinds

of the biliary symptoms. Jaundice was noted in five patients, and five complained of epigastric pain. Four complained of the righ upper quadrant pain, but signs of peritoneal irritation were absent. One patient expectorated bile-tinged sputum with the typical biliary colic (Table 1).

Table 1. Subjective symptoms presented by patients

Jaundice	5
Epigastric pain	5
Right upper quadrant pain	5
Bile tinged sputum	1

Preoperative Diagnosis

All the patients underwent ultrasonographic examinations but, in any case, CAGB could not be predicted by the ultrasonography: In five, the sonographic impressions were regarded as "gallbladder cannot be found due to bowel gases"; in two, "normal gallbladder"; and in antother two, "cholecystitis with gallbladder stones". Four patients underwent oral cholecystography, and two underwent intravenous cholecystography. The results were "nonvisualization of gallbladder implying an acute cholecystitis". Endoscopic retrograde cholangiography (ERC) was performed in two patients, and its also did not document the absence of gallbladder but could yield negative findings for other more common conditions. The abdominal CT scan was performed in one case and the possibility of absence of gallbladder was therefore predicted (Table 2).

Operative Findings

After meticulous dissections, the extrahepatic biliary trees were completely identified and the possibility of intrahepatic gallbladder was excluded by the operative cholangiography. The gallbladder was absent in each patient. The cystic duct was also absent except in one patient who had a cord-like outpouching identified by the cholangiography. All the patients had otherwise intact extrahepatic biliary trees, but four patients had common duct stones and/or intrahepatic duct stones. For those patients, the choledocholithotomy followed by T-tube choledochostomy was performed.

A juxtaampullary choledochocele interfering with the biliary outflow was found in one patient and a Roux-en-Y choledochojejunostomy for a choledochoenteric bypass was necessary. In the patient who complained of a bile-tinged sputum, a direct communication between intrahepatic bile ducts and the right main bronchus was found, and both ends of the fistula were separately ligated.

In four patients, no other pathologic condition was disclosed in the biliary tree (Table 3) and no specific procedure except the needle cholangio-

Table 3. Operative findings

Gallbladder status	
present	0
absent	9
Cystic duct status	
absent	8
hypoplastic	1
Extrahepatic biliary tree	
CBD or intrahepatic stones	4
Normal biliary tree	4
Choledochocele	1
Bronchobiliary fistula	1

Table 2. Preoperative diagnosis

Diagnostic Method ( ), Number of cases	Diagnosis			
	incomplete study	cholecystitis	normal GB	CAGB
Abdominal US[1] (9)	5	2	2	-
Oral GB[2] (4)	-	4	-	-
IVC[3] (2)	-	2	-	-
ERC[4] (2)	2	-	-	-
Abdominal CT (1)	-	-	-	1

1, ultrasound 2, oral cholecystography 3, intravenous cholecystography 4, endoscopic retrograde cholangiography

graphy was necessary.

DISCUSSION

Embryologically, the hepatobiliary system begins to develop about the third gestational week. The hepatic diverticulum is formed by an ectodermal outpouching of the distal end of the foregut, and as it grows the connection between gut and the hepatic diverticulum is narrowed. Small vacuolizations occur in the segment of the connection and they become the gallbladder and cystic duct. Intrahepatic biliary trees with both hepatic ducts while originate from the proximal portion of the hepatic diverticulum and gallbladder, cystic duct and the common bile duct are from the distal portion of the diverticulum (Blechschmidt, 1982). So, the probability of developing an isolated CAGB without extrahepatic biliary atresia is extremely low. Autopsy series have been reported with 0.035% to 0.3% incidences (Bennion *et al.* 1988) or less than one in 6,000 live births (Monroe 1959).

Because of CAGB's rarity, physicians and radiologists are unfamiliar with its condition and hesitate to document the condition of CAGB in spite of failure to filnd a gallbladder. In our series, all the patients underwent ultrasonographic examinations, but radiologists ascribed the findings to incomplete study in most cases. "Gallbladder stones" found in the sonography might be dilated common bile ducts and their stones. But the cause of the report of two normally appeared gallbladders can not be conjectured. Other studies, including oral and intravenous cholecystography and endoscopic retrgrade cholangiography, did not document the CAGB. Only computed tomography (CT) of the abdomen was able to accurately determine the condition. But the routine use of abdominal CT is not necessary for diagnosing cholecystitis so only in a case in which the gallbladder is not visualized on ultrasonography, should the CT of the abdomen be performed.

Once it has been discovered from a laparotomy that the gallbladder appears to be absent, surgeons should prove the agensis. A contracted gallbladder due to longstanding inflammation, an ectopic gallbladder and finally an intrahepatic gallbladder should be excluded (Sherson 1970; Langley *et al.* 1974). Needle operative cholangiography is essential and sufficient for exclusion of those differential points and the exploration of the common duct should be reserved only for cases in which stones are demonstrated on operative cholangiography and in cases of significant dilatation of the common bile duct.

The reasons for symptoms of CAGB is still obscure. If there is associated biliary pathology such as a common duct stone or intrahepatic duct stone, the mechanism of the symptoms is readily explained. But as in our series and others (Turkel *et al.* 1983; Rabinovitch *et al.* 1958; Bennion *et al.* 1988), biliary symptoms can arise from the isolated CAGB. Recently, it has been demonstrated that patients who suffer from recurrent episodes of biliary colic and possess an otherwise normal pancreatobiliary tree had a significant higher sphincter of Oddi resting pressure, as well as a significantly increased proportion of retrograde propagation of phasic muscular contraction when compared with healthy volunteers (Meshkinpour *et al.* 1984). This so called biliary dyskinesia is thought to be responsible for most cases of postcholecystectomy syndrome and isolated CAGB (Bennion *et al.* 1988). Because the biliary dyskineasia should be treated with medications such as muscle relaxants or anagesics, and even in the refractory cases, the endoscopic sphincter ablative procedures are sufficient for relief of symptoms, exploration of abdomen is not thought to be a reasonable treatment.

The CAGB is a rarely encountered condition to clinicians but extensive preoperative diagnostic work-ups including abdominal CT scan should be performed in all situations where the CAGB is suspected and unnecessary exploration should be avoided in the isolated CAGB case.

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