대한췌담도연구회지 1998; 3: 150~158

Choledochal Cyst Associated with Anomalous Union of Pancreaticobiliary Duct (AUPBD) Has a More Grave Clinical Course Than Choledochal Cyst Alone

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= 국문초록 =

췌담관 합류이상 유무에 따른 담관 낭종 환자의 임상적 비교 연구

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김명환 · 명승재 · 김홍자 · 심기남 · 임병철
유교상 · 박은택 · 서동완 · 이성구 · 민영일

연구배경 및 목적: 담관낭종은 선천성으로 담관의 확장을 보이는 질환으로 췌담관 합류이상과 자주 동반된다. 이 췌담관 합류이상은 담관 낭종 생성의 한 요소로 생각되고 있다. 그러나 췌담관 합류이상의 유무에 따른 담관 낭종 환자의 임상적 특성에 대해서는 잘 알려져 있지 않다. 이 연구의 목적은 담관낭종 환자의 임상상을 췌담관 합류이상의 유무에 따라 나누어 비교하고 췌담관 합류이상의 의미를 고찰하고자 하는 것이다. **대상 및 방법**: 1990년 8월부터 1996년 12월까지 5037예의 ERCP시행예 중 52예의 담관 낭종이 발견되었다. 52예 중 44예에서는 췌담관 합류부를 조영할 수 있었다. 이들 증례를 합류이상 동반예(n=28)와 합류이상을 동반하지 않은 예(n=16)로 나누어 임상적 특성을 비교하였다. 또한 합류이상 동반예에서는 Kimura의 분류에 따른 자료의 분석도 시도하였다. **결과**: 췌담관 합류이상은 28예 (64%)에서 동반되어 있었다. Todani의 분류에 따르면, 합류이상은 type I과 type IV에서만 발견되었다. 담석의 발생은 합류이상 유무에 따라 차이가 없었으나 급성 염증은 합류이상 동반군에서 의미 있게 자주 발생하였다(26/28, 93% vs. 5/16, 31%)($p<0.01$). 악성종양은 합류이상 동반군에서 (9/28, 32%) 동반하지 않은 군에 비해 (0/16, 0%)($p<0.05$) 더 자주 발생하였다. 췌장질환(즉, 췌관결석, 췌장염, 췌장암)은 28예의 합류이상 동반군에서 12예에서 발생하였으나(43%) 합류이상을 동반하지 않은 군에서는 16예중 1예에서만 발생하였다(6%)($p<0.05$). **결론**: 췌담관합류이상은 담관낭종의 발생에 원인적 요소로 작용할 뿐만 아니라 임상경과, 외과적 수술의 계획, 또는 예후에도 영향을 미치는 것으로 보인다. 담관낭종의 증례들에서는 췌담관 합류이상을 찾으려는 노력이 필요할 것으로 생각된다.

핵심단어: Anomalous union of pancreaticobiliary duct, Choledochal cyst, Prognosis

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INTRODUCTION

Choledochal cyst is a congenital dilatation of the bile duct. This congenital disease is relatively rare in western countries and more than two thirds cases were reported in Japan.[1] Anomalous union of pancreaticobiliary duct (AUPBD) has been regarded to be the etiological factor of the choledochal cyst.[2] However, choledochal cyst is not always associated with AUPBD and the presence of AUPBD without choledochal cyst has been increasingly recognized recently, probably because of the advances in hepatobiliary imaging techniques. Therefore some authors suggests that these two anomalies should be considered separately.[3] There have been many reports about choledochal cyst or AUPBD, but their cases were mainly infants or neonates. Moreover, cholangiography was obtained by percutaneous transhepatic route or intraoperatively.[4] Hence, the pancreaticobiliary junction might not be fully evaluated in previous series.

While most of the choledochal cysts were observed in infants, it can be found in adults.[5] All of our cases had choledochal cyst diagnosed in adulthood and the presence or absence of AUPBD was confirmed by endoscopic retrograde cholangiopancreatography (ERCP). The number of the cases was 44, the largest series collected from single institution except the ones from Japan. Moreover, there were rare reports in English literature which compared clinical features of choledochal cyst according to the presence of AUPBD. The purpose of our study was to compare the clinical characteristics of 44 cases with adulthood choledochal cyst according to the presence or absence of AUPBD. Furthermore, we tried to clarify the significance of AUPBD in choledochal cyst patients.

PATIENTS AND METHODS

From August 1990 to December 1996, 52 cases (1.03%) were diagnosed as having choledochal cyst out of 5,037 ERCP referrals. The diagnosis of choledochal cyst was made as a localized non-proportional dilatation of bile duct after exclusion of tumor, stone, or inflammation as a cause of the dilatation.[6] All of the patients in our series were more than 16 years of age. Of the 52 choledochal cyst cases, we selected 44 cases, of which the pancreaticobiliary junction was clearly visualized.

Choledochal cyst was classified as I, II, III, IVa, IVb, V according to Todani's classification. Type I is cystic or diffuse dilatation of extrahepatic bile duct, type II is a diverticulum in the extrahepatic bile duct, type III is choledochocele, type IVa is multiple cystic dilatation of intra- and extrahepatic bile duct, type IVb is multiple cystic dilatation of extrahepatic bile duct, and type V is multiple cystic dilatation of intrahepatic bile ducts (Caroli's disease).[7] AUPBD was defined as anomalous union of pancreaticobiliary duct system at a distance >15 mm from the papilla of

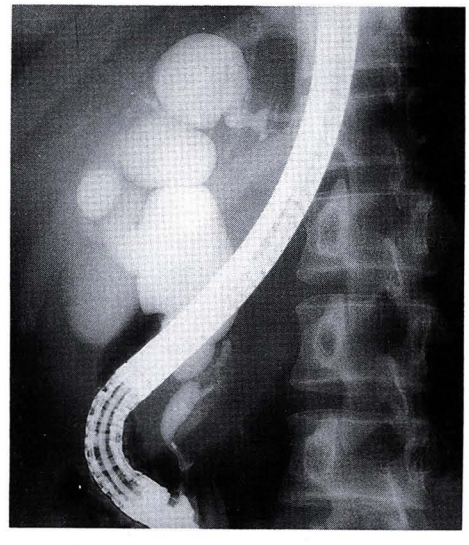

Fig. 1. Endoscopic retrograde cholangiopancreatogram of choledochal cyst (Todani type IVa) and anomalous union of pancreaticobiliary duct (Kimura type I). Pancreatic duct is partially visualized due to incomplete filling.

Vater.[8] This anomaly was divided into type I (Fig. 1, 2) and II (Fig. 3) according to Kimura's classification. Type I AUPBD looks as though the pancreatic duct joins the bile duct, which is the major duct, whereas in type II, it looks as though the bile duct joins the pancreatic duct, which is the major duct.[8]

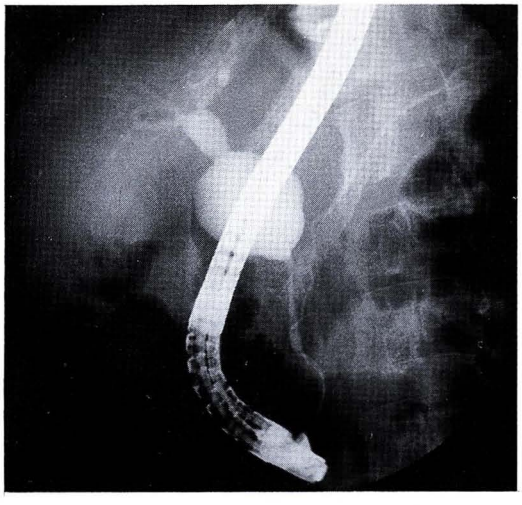

Fig. 2. Endoscopic retrograde cholangiopancreatogram of choledochal cyst (Todani type IVb) and anomalous union of pancreaticobiliary duct (Kimura type I).

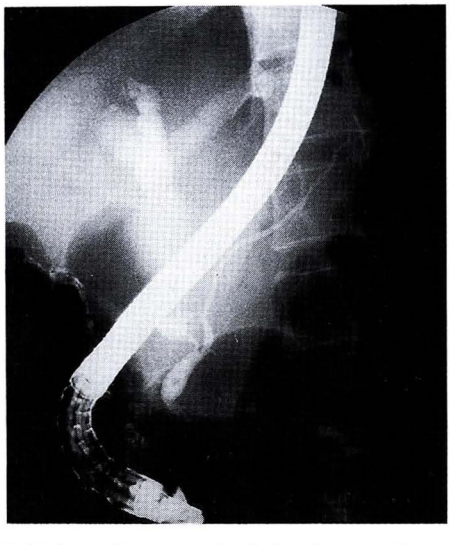

Fig. 3. Endoscopic retrograde cholangiopancreatogram of choledochal cyst (Todani type IVa) and anomalous union of pancreaticobiliary duct (Kimura type II).

The cases with choledochal cyst were divided into those associated with AUPBD (n=28, AUPBD-present group) and those without (n=16, AUPBD-absent group) and clinical characteristics were compared between two groups. Furthermore, in AUPBD-present group, characteristics were also compared according to Kimura's classification of AUPBD. The angle of the pancreaticobiliary junction was measured as viewed frontally. Statistical analysis was made by Fisher's exact test and Mann Whitney U test.

RESULTS

Of the 44 choledochal cyst cases, 17 cases had type I cysts, 1 had type II, 6 had type III, 18 had type IVa, 1 had type IVb, and 1 had type V. AUPBD-present group was 28 (64%), while AUPBD-absent group 16 (36%). AUPBD was observed only in type I and IV patients, whereas it was not shown in type II, III, V patients (Table 1). Age distribution of choledochal cyst was 7 in 16～19 years, 7 in 20～29 years, 11 in 30～39 years, 7 in 40～49 years, 5 in 50～59 years, and 7 in 60～69 years. Those were 15 males and 29 females (M : F ratio, 1 : 1.9)(Table 2).

Comparing the characteristics according to the

Table 1. Type of choledochal cyst by Todani's classification

Type	Cyst + AUPBD (n=28)	Cyst − AUPBD (n=16)	Total
I	12	5	17
II	0	1	1
III	0	6	6
IVa	15	3	18
IVb	1	0	1
V	0	1	1

AUPBD: anomalous union of pancreaticobiliary duct

Table 2. Age distribution of choledochal cyst

Age (yrs)	Cyst+AUPBD (n=28)	Cyst−AUPBD (n=16)	Total
16~19	5	2	7
20~29	6	1	7
30~39	8	3	11
40~49	4	3	7
50~59	4	1	5
60~69	1	6	7

AUPBD: anomalous union of pancreaticobiliary duct

AUPBD-association, female cases were more observed in both groups, whereas the mean age of AUPBD-present group was 49.2 years, younger than that of AUPBD-absent group although it was not statistically significant (Table 3). Gallstone diseases were associated in 18 (41%) patients with choledochal cyst (n=44). The location of the gallstones was 11 in cyst, 5 in gallbladder, and 2 in intrahepatic duct. Pancreatic stones were shown in 2 (5%) patients. Acute inflammation was observed in 31 (70%) cases. They were cholecystitis (n=12), cholangitis (n=8), and pancreatitis (n=11). Malignant neoplasm occurred in 9 (20%) cases: gallbladder in 3, common bile duct in 4, and pancreas in 2 cases. All the cancers in common bile duct arose from the cyst wall and were adenocarcinoma pathologically.

The difference in the incidence of associated diseases according to the presence of AUPBD was as follows. The incidence of gallstone disease in AUPBD-present group did not differ from that in AUPBD-absent group, while acute inflammation occurred more frequently in AUPBD-present (26/28, 93%) than in AUPBD-absent group (5/16, 31%)($p<0.01$). Malignant neoplasm developed only in AUPBD-present group (9/28, 32%), more often than in AUPBD-absent group (0/16, $p<0.05$)(Table 3). Pancreatic disorders (pancreatic stone, pancreatitis, or pancreatic cancer) occurred in 12 of 28 cases (43%) in AUPBD-present group, whereas only in 1 of 16 (6%) cases of AUPBD-absent group ($p<0.05$, Table 3).

Table 3. Differences of clinical characteristics and associated diseases according to the presence of AUPBD

	Cyst+AUPBD (n=28)	Cyst−AUPBD (n=16)
Age(mean+SD,yrs)	36.5±14.1	49.2±12.2
Sex(M : F)	9 : 19	6 : 10
Stone		
Cystolithiasis	5	6
Gallbladder stone	2	3
Intrahepatic stone	1	1
Pancreatic stone	2	0
	(n=10)	(n=10)
Acute inflammation		
Cholecystitis	10	2
Cholangitis	6	2
Pancreatitis#	10	1
	(n=26)*	(n=5)
Malignancy		
GB cancer	3	0
CBD cancer	4	0
Pancreatic cancer	2	0
	(n=9)**	(n=0)
Pancreatic diseases	12(43%)**	1(6%)

AUPBD: anomalous union of pancreaticobiliary duct; GB: gallbladder; CBD: common bile duct; *$p<0.01$; **$p<0.05$, compared to that of cyst-AUPBD, by Fisher's exact test; #3-fold or more elevation of amylase level associated with acute abdominal pain, two cases with pancreatic stones were included

According to Kimura's classification, AUPBD (n=28) was divided into type I (n=12) and II (n=16), but we could not find any difference in associated diseases between two groups (Table 4). The maximal diameter of common bile duct was higher in type II AUPBD, but it was not statistically significant. However, the angle between the biliary and pancreatic duct was higher in type II (85.1°) than in type I (45.2°)($p<$

Table 4. Differences of clinical characteristics and associated diseases according to the type of AUPBD

	AUPBD	
	Type I (n=12)	Type II (n=16)
CBD diameter	30.5±3.4 mm	42.5±3.7 mm
Length of CC	22.7±1.5 mm	22.8±1.7 mm
Angle*(°)	45.2±15.4	85.1±16.2#
Type of choledochal cyst		
Type I	5	7
Type IVa	6	9
Type IVb	1	0
Stone		
cystolithiasis	3	2
GB stone	1	1
IHD stone	0	1
pancreatic stone	0	2
Acute inflammation		
cholecystitis	5	5
cholangitis	2	4
pancreatitis	4	6**
Malignancy		
GB cancer	2	1
CBD cancer	2	2
pancreatic cancer	0	2
Pancreatic diseaes	4(33%)	8(50%)

AUPBD: anomalous union of pancreaticobiliary duct; CBD: common bile duct; CC: common channel; GB: gallbladder; IHD: intrahepatic duct; *angle which was formed when common bile duct and pancreatic duct joined; #$p<0.05$, compared to that of type I, by Mann Whitney U test; **two cases with pancreatic stones were included

0.05, Table 4).

DISCUSSION

At the Childrens Hospital of Los Angeles, choledochal cyst was diagnosed in 0.5 patient per year.[10] However, in our institution (3rd referral center), we experienced 53 cases during 6 years (incidence of about 9 per year). Our data suggest that choledochal cyst is more prevalent in Korea than in western countries. It also implies that the incidence of choledochal cyst may be higher not only in Japan but in other oriental countries than in western countries.[11] Since the earlier report by Babbit et al.[12] about frequent association of AUPBD with choledochal cyst, AUPBD has been regarded as an etiological factor of choledochal cyst.[2] However, the rate of AUPBD association with choledochal cyst was from 33% to 100% according to the reports.[4,13~15] It might be due to the difference in the characteristics of selected cases. According to our results, AUPBD was associated only with type I and IV choledochal cyst (Table 1). Therefore, the rate of AUPBD can be affected by the number of cases with type I or IV. Furthermore, Todani et al.[6] sub-classified type I choledochal cyst into type Ia, Ib, Ic, and suggested that AUPBD may not be associated in type Ib. This also implies that the association rate of AUPBD can be influenced even by the sub-classification in type I choledochal cyst. Moreover, Matsumoto et al.[16] divided choledochal cyst into childhood-type and adulthood-type. Association of AUPBD was observed in almost 100% of childhood-type, whereas less frequently found in adulthood-type. The difference of association rate in many reports including ours may be explained by the difference in the characters of cases included.

Although AUPBD was considered as an etiological factor of choledochal cyst,[2,12] the pathogenetic mechanism of choledochal cyst may not be explained solely by AUPBD because AUPBD was not found in all of the choledochal cyst cases. Our data also showed that AUPBD was not always associated with choledochal cyst cases (28/44, 64%). The increasing reports of AUPBD without choledochal cyst supports the notion.[17]

In AUPBD, a union of the pancreatic and biliary

ducts is located outside the sphincter of Oddi. Therefore, two-way regurgitation occurs. Pancreatic juice refluxes into the common bile duct, or bile regurgitates into the pancreatic duct because the action of the sphincter muscle does not functionally affect the union.[18] Because the intraductal pressure is generally higher in pancreatic duct than in bile duct, pancreatic juice regurgitates into biliary tract resulting in the pancreatic enzyme activation and subsequent recurrent inflammation. These may give rise to metaplastic and finally malignant change of the biliary epithelium.[19] Furthermore, after recurrent inflammation of the bile duct, the pressure in the bile duct rises and bile may reflux into the pancreatic duct causing various pancreatic disorders including acute or chronic pancreatitis, pancreatic stone, or pancreatic cancer.[18]

The incidence of malignant diseases in choledochal cyst is said to be 2.5%~15%, 15 times greater than the control population without choledochal cyst.[20] In our series, cancer developed in 9 of 44 (20%). The incidence in our series might be higher than in other reports because all of our cases were in adulthood. It is well known that the incidence of cancer in choledochal cyst increases with age.[21] Moreover, cancer developed only in AUPBD-present group, which implies that AUPBD may be more important contributing factor than the choledochal cyst itself. Flanigan et al.[22] pointed out that only 57% of cancers occurred in choledochal cyst were located in the cyst wall and the rest of cancers developed in bile duct other than cyst wall. Moreover, Nagorney et al.[23] also suggested that malignant neoplasm developed in choledochal cyst is not always located in the cyst wall. In one of their cases, cancer developed in the remaining bile duct after complete cyst excision. These data implies that risk factors causing cancer in choledochal cyst are more than the choledochal cyst itself. The cancers developed in our series were located in gallbladder (n=3), common bile duct (n=4), and pancreas (n=2). Only 4 cases occurred in cyst wall. Recent reports of gallbladder cancer in AUPBD cases without choledochal cyst suggests that AUPBD is more important for the carcinogenic process than the choledochal cyst itself.[24] In 35 cases of choledochal cyst described by Yoshida et al., 8 cases developed cholangiocarcinoma.[25] AUPBD was associated with all of these 8 cases, which supports our notion that AUPBD may be the major contributing factor for the cancer development. Suda et al. examined AUPBD in 34 bile duct cancer patients, 24 gallbladder cancers, and 171 controls without biliary disease.[26] They observed AUPBD in 8 of 34 cholangiocarcinoma, 4 of 24 gallbladder cancer, but none in controls. They suggested that AUPBD is one of the pathogenetic factors in biliary malignancy.

In our results, acute inflammatory condition such as cholecystitis, cholangitis, or pancreatitis was more prevalent in AUPBD-present than in AUPBD-absent group. The younger age in AUPBD-present group suggests that patients in this group might visit hospital earlier because of more severe symptoms.

One of the factors worth mentioning is pancreatic diseases associated with choledochal cyst. There had been several reports about the association of pancreatitis, pancreatic stone, or pancreatic cancer in choledochal cyst.[20,27~29] In the previous reports, however, they did not analyzed the data considering AUPBD. In one of Japanese report, acute pancreatitis occurred in 30 (17%) of the 176 cases with AUPBD.[30] Activated pancreatic enzymes after entering the biliary tract may cause cholangitis, gallstone, and cholangiocarcinoma.[31] Likewise, these enzymes may reflux back into the pancreatic duct and cause various pancreatic disorders such as acute or chronic pancreatitis, and pancreatic cancer.[32] In our results, pancreatic disorders developed more frequently in AUPBD-associated group (Table 3).

These high incidence of malignancy and inflamma-

tory diseases associated with AUPBD also have therapeutic implications for choledochal cyst. In cases of choledochal cyst with AUPBD, cholecystectomy also should be performed in addition to cyst excision because the incidence of gallbladder cancer is very high. Moreover, surgical procedure for correction of AUPBD should be added. Biliary diversion from the pancreatic juice (pancreaticobiliary disconnection) may be needed for prevention of bi-directional reflux of pancreatic and bile juice.[23,24] In this regards, cholecystectomy along with the resection of dilated bile duct and the biliary diversion from pancreatic juice should be performed in cases with choledochal cyst and AUPBD. Komi et al.[35] subdivided AUPBD associated with choledochal cyst into several categories and suggested that pancreatitis could not be prevented by cholecystectomy, cyst excision and hepaticojejunostomy in certain subgroups. Furthermore, they suggested that in cases with AUPBD showing dilated common channel or accessory pancreatic duct, sphincteroplasty or pylorus preserving pancreatico-duodenectomy should be needed in addition to the previously mentioned procedures. Schreiber et al.[36] described that AUPBD may be observed as two clinical manifestations. One is the biliary tract disease such as acute cholecystitis, cholangitis, and cholangiocarcinoma. The other one is caused by stasis of pancreatic fluid due to anomalous drainage in the common channels leading to periductal and interlobular fibrosis as a histological sign of chronic pancreatitis. Thus, Schreiber et al. suggested that resection of the anomalous junction and hepaticojejunostomy with a Roux-en-Y anastomosis may resolve both pancreatic reflux into biliary system and stasis of pancreatic secretion. The claim by Schreiber et al. has something to do with that of Komi et al. suggesting pylorus-preserving pancreaticoduodenectomy may be recommended in certain cases of AUPBD.

AUPBD frequently associated with choledochal cyst may have an implication not only as an etiological factor but as an associated disorder leading to a grave clinical course. In this regard, we should make an effort to confirm the presence of AUPBD in patients with choledochal cyst. Moreover, adequate surgery may be required to prevent the occurrence of cancer. Cancer associated with choledochal cyst may often be in advanced stage when detected. Curative resection may be difficult.[20] Prevention, therefore, may be the best way if possible. AUPBD associated with choledochal cyst may be a very important factor that affects the clinical course, surgical planning, and prognosis.

SUMMARY

Background and Study Aims: Choledochal cyst is a congenital dilatation of the bile duct and frequently associated with anomalous union of pancreaticobiliary duct (AUPBD). AUPBD has been regarded to be the etiologic factor of choledochal cyst. However, the clinical significance of AUPBD in patients with choledochal cyst has not been clearly defined. The aims of our study were to compare the clinical features of patients with choledochal cyst according to the presence or absence of AUPBD, and to clarify the significance of AUPBD in choledochal cyst patients. **Patients and Methods:** From August 1990 to December 1996, 52 cases were diagnosed as having choledochal cyst out of 5,037 ERCP referrals. In 44 of 52 cases, the pancreaticobiliary junction was clearly visualized on cholangiopancreatography. We divided these cases into those associated with AUPBD (n=28, AUPBD-present group) and those without (n=16, AUPBD-absent group). Clinical features were compared between two groups. Furthermore, in AUPBD-present group, clinical data were also analyzed according to Kimura's classification of AUPBD. **Results:** AUPBD was associated with choledochal cyst in 28 (64%) cases.

According to Todani's classification of choledochal cyst, AUPBD was found only in type I and IV cases. The incidence of gallstone disease in AUPBD-present group did not differ from that in AUPBD-absent group, while acute inflammation occurred more frequently in AUPBD-present (26/28, 93%) than in AUPBD-absent group (5/16, 31%)($p<0.01$). Carcinoma developed only in AUPBD-present group (9/28, 32%), more often than in AUPBD-absent group (0/16, 0%)($p<0.05$). Pancreatic disorders (i.e. pancreatic stone, pancreatitis, or pancreatic cancer) occurred in 12 of 28 cases in AUPBD-present group (43%), while only in 1 of 16 cases in AUPBD-absent group (6%)($p<0.05$). **Conclusion:** AUPBD associated with choledochal cyst may have implication not only as a possible etiologic factor but also as an important factor that may affect the clinical course, surgical planning, and prognosis. In cases with choledochal cyst, we should make an effort to evaluate the presence of AUPBD.

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