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# One Case of Common Bile Duct Cancer Mimicking Cystic Neoplasm of the Pancreas, Arising 9 Years after Excision of a Choledochal Cyst

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A 42-years-old woman had undergone operation for choledochal cyst with gallbladder cancer 9 years ago. Pathology revealed a polypoid mass in the gallbladder with liver infiltration as poorly differentiated adenocarcinoma. Computed tomography, magnetic resonance cholangiopancreatography, and endoscopic ultrasound showed a newly developed suspected solid nodule in the peripheral portion of cystic lesion in the pancreas head. She underwent a pylorus preserving pancreaticoduodenectomy for the suspected mucinous cystic neoplasm of the pancreas. Pathology revealed poorly differentiated adenocarcinoma. The remnant choledochal cyst had developed to cholangiocarcinoma, which mimicked cystic neoplasm of the pancreas.

**Key Words:** Choledochal cyst; Pancreatic cyst; Cholangiocarcinoma

## INTRODUCTION

Choledochal cysts are rare congenital or acquired cystic dilations of the intra- or extrahepatic bile ducts and dilatation of duct.<sup>1</sup> It is well accepted that a choledochal cyst is a premalignant state.<sup>2</sup> Cancer usually occurs within the cyst, and many authors postulated that malignancy occurs at the site of bile stasis, irritation, and inflammation.<sup>3</sup> Thus, as well as the prevention of later development of cancer in cyst, complete excision of the cyst and cholecystectomy with reconstruction is recommended as the mainstay of treatment. Herein, we report a rare case of common bile duct (CBD) cancer arising from a remnant cyst, 9 years after the excision of a choledochal cyst.

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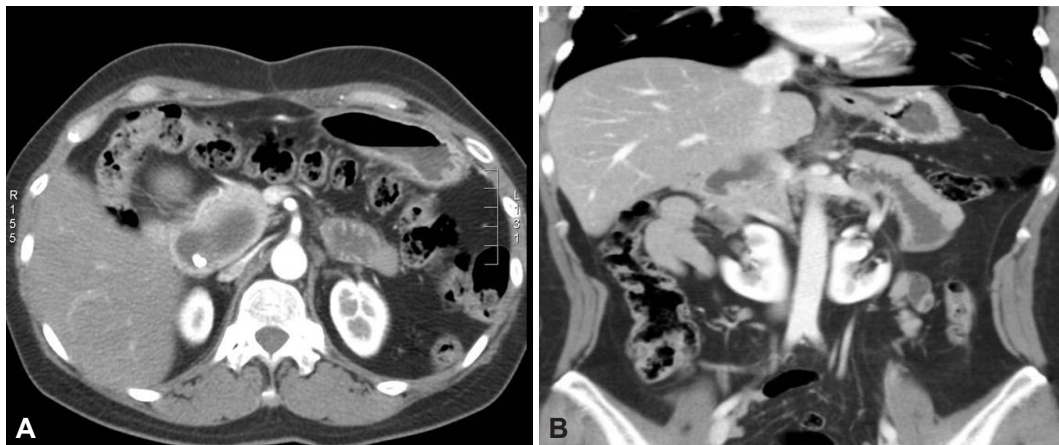
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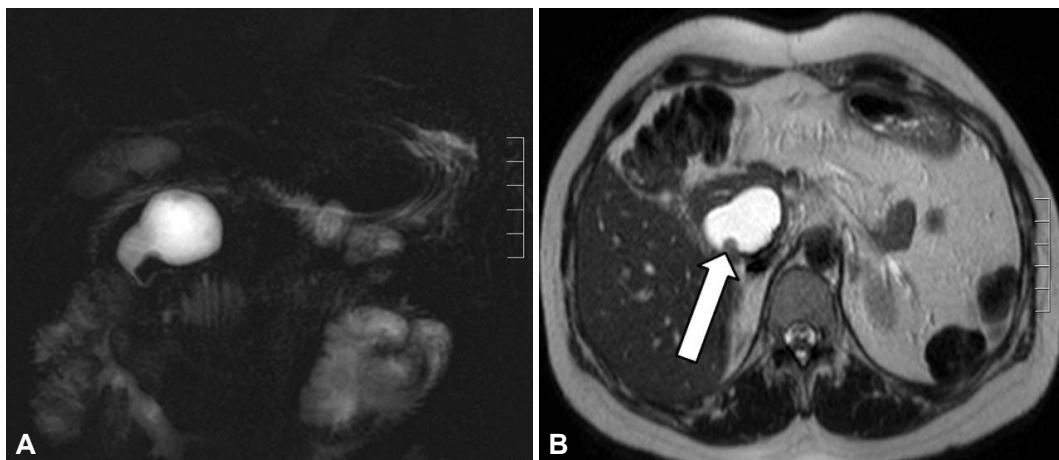
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## CASE REPORT

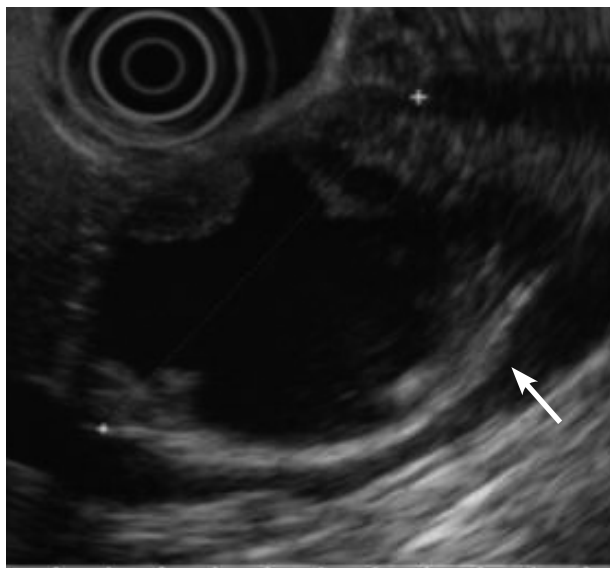
A 42-year-old woman was admitted to our hospital for the investigation of dyspepsia and abdominal discomfort which have lasted for 1 week. It was constant and was not associated with nausea, constipation, or diarrhea. Nine years ago, the patient had undergone operations for choledochal cyst with gallbladder cancer and pancreatic tail mass. Radical cholecystectomy (lymph node [LN] dissection, liver wedge resection), choledochal cyst excision and Roux-en-Y hepaticojejunostomy, distal pancreatectomy, and splenectomy for multinodular hard mass in the tail of the pancreas were performed. Pathology showed a polypoid mass with liver infiltration as poorly differentiated adenocarcinoma and a tail mass as chronic pancreatitis. The patient's stage was T2N0M0 (LN 0/16). The patient received four cycles of postoperative adjuvant systemic chemotherapy with 5-fluorouracil and cisplatin and there was no evidence of disease until her visit to our clinic. The patient had no history of diabetes mellitus, hyperlipidemia, and hypertension. She did not take any medications. The body temperature was 36.8°C, the pulse was 63 beats/min, and the respiration was 18/min. The blood pressure was 100/60 mm Hg. The patient was uncomfortable but not in acute distress. She did not



**Fig. 1.** (A) Enhanced computed tomography showed solid nodule in the peripheral portion of cystic lesion, 5 cm in diameter, in the pancreas head (axial view). (B) Coronal view.

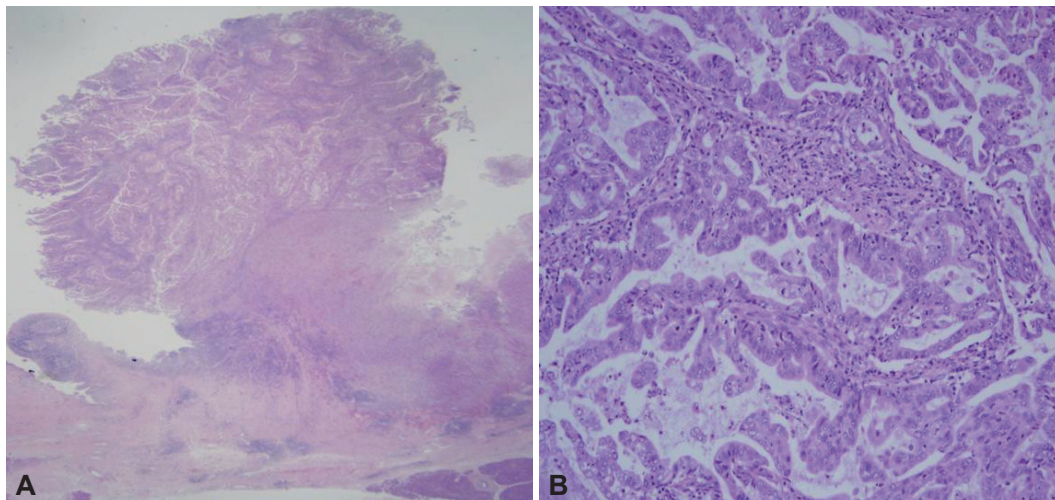


**Fig. 2.** (A) Magnetic resonance cholangiopancreatography representing cystic lesion in the pancreatic head, (B) a tiny solid nodule in the cystic lesion (white arrow).



**Fig. 3.** Endoscopic ultrasound showed 5 cm diameter cystic lesion in pancreatic head, thick cystic wall, and multiple mural nodules in the cystic lesion (white arrow).

appear so ill, with no sclera icterus or jaundice. Bowel sound was normal, and there was neither tenderness nor guarding. The remainder of the examination revealed no abnormalities. White blood cell count was  $6,100/\text{mm}^3$ , hemoglobin was 12.0 g/dL, and platelet count was  $463,000/\text{mm}^3$ . Liver function tests revealed the following: protein 6.5 g/dL; albumin 4.0 g/dL; aspartate aminotransferase 18 IU/L; alanine aminotransferase 45 IU/L; alkaline phosphatase 45 IU/L; total bilirubin 0.7 mg/dL; and cholesterol 193 mg/dL. Amylase was 47 U/L, and lipase was 59 U/L. Tumor markers showed the following: carbohydrate antigen 19-9 17 U/mL (0 to 37); alpha-fetoprotein 1.8 IU/mL (0 to 20); and carcinoembryonic antigen 2.4 ng/mL (0 to 5). Based on her past medical history, we conducted computed tomography (CT) first, which detected suspicious solid nodule in the peripheral portion of cystic lesion in the pancreas head (Fig. 1). Magnetic resonance cholangiopancreatography and endoscopic ultrasound (EUS) detected the cystic lesion with mural nodule in the head of pancreas (Figs. 2, 3). The cyst with thick wall seemed in the pancreatic head, and



**Fig. 4.** Microscopic finding. (A) Polypoid nodule, invaded to the perimuscular connective tissue of pancreas. Acinar structure of pancreas was seen beneath the nodule. (B) Moderately to poorly differentiated adenocarcinoma surrounded by desmoplastic reaction (H & E stain,  $\times 100$ ).

multiple mural nodules appeared in it. The patient was referred to the department of surgery for operation. She underwent a pylorus preserving pancreaticoduodenectomy. At surgery, there was severe adhesion in the previous hepaticojejunostomy site. Roux-en-Y hepaticojejunostomy was in retrocolic state, remnant intrapancreatic distal CBD was dilated and two hard mass was in it. Duodenojejunostomy was performed. Cross sections of the excised specimen showed dilated CBD to 5 cm in the pancreatic head and inside of it, two nodular lesions were seen as  $1.3 \times 1.2 \times 1.0$  cm and  $0.7 \times 0.5 \times 0.5$  cm in diameter. Histologically, it was a poorly-differentiated adenocarcinoma, which was limited to the perimuscular connective tissue and which did not penetrate to other parts of the bile duct wall (Fig. 4). There was no LNs metastasis and all surgical margins were free of tumor. Pancreas and duodenum was free of carcinoma cells. At the time of writing, the patient was alive after 34 months without recurrence or metastasis.

## DISCUSSION

Choledochal cyst is rare disease, but has a clinical importance because of high incidence of biliary tract cancer, even in an asymptomatic case.<sup>4</sup> The overall risk of cancer has been reported to be 10% to 15%.<sup>5</sup> The carcinogenesis is unclear, it was suggested that metaplasia and dysplasia of biliary duct epithelial cells could develop to malignancy at the site of bile stasis, irritation, and inflammation.<sup>6</sup>

The importance of complete cyst excision is commonly understood to prevent the development of malignancies and other complications.<sup>7</sup> In one Japanese study, approximately 1% of cases developed to cholangiocarcinoma which was mainly originated from incomplete excision.<sup>8</sup>

There have been previous reports similar to our case in Eng-

lish and Korean literature. A PubMed search of all patient articles yield reports of a total of 25 patients in whom bile duct carcinoma developed after the choledochal cyst excision (Table 1).<sup>9-18</sup> These 26 patients, including our patient, consists of eight male and 18 female. They were aged from 16 to 70 years. The mean time from the cyst excision to the development of bile duct carcinoma was 9.1 years (range, 0.6 to 17.6). The type of choledochal cyst according to Todani's classification was type IV-A in 10 patients, type I in six patient, and not-described in 10 patients. In most of cases, carcinoma developed in the residual dilated bile duct, in the hepatic duct at anastomotic site, or in the dilated intrahepatic bile duct. In only five patients including our patient, the development of carcinoma was reported in the head of pancreas or intrapancreatic bile duct. However, mimicry of cystic neoplasm of pancreas had not been reported, except our patient.

It is well known that the finding of mural nodules during EUS imaging may be helpful in identifying malignancy lesions, especially cystic tumor than pseudocyst.<sup>19</sup> In addition, if mural nodule  $>10$  mm was demonstrated in EUS feature, this can help distinguish between biliary intraductal papillary mucinous neoplasm and other cysts.<sup>20</sup> Therefore, malignant cystic neoplasm was strongly suspected in our patient according to her EUS feature.

In this case, CBD cancer had arisen from the remnant cyst 9 years after the excision of a choledochal cyst, and it mimicked mucinous cystic neoplasm of the pancreas. The present case is the first in the English-language literature.

In conclusion, complete excision is a crucial treatment strategy of choledochal cyst and careful long-term follow-up is important to detect a malignant biliary tract tumor at an early stage.

**Table 1.** Previously Reported Biliary Duct Carcinoma after Choledochal Cyst Excision

Author	Sex	Age, yr	Type <sup>a)</sup>	Age at cyst excision, yr	Interval to cancer development	Site of cancer	Pathology
Kelly et al. (1964) <sup>9</sup>	M	30	ND	30	4 mo	Head of pancreas	Cholangiocarcinoma
Thistlewaite (1967)	M	24	I	24	4 yr	Anastomotic site of HJ	Anaplastic adenocarcinoma
Gallagher et al. (1972) <sup>10</sup>	F	58	IV-A	51	7 yr	Intrahepatic bile duct	Cholangiocarcinoma
Chaudhuri (1982)	F	38	IV-A	21	17 yr	Intrahepatic bile duct	Squamous cell carcinoma
Nagorney et al. (1984) <sup>11</sup>	F	16	I	14	1 yr 8 mo	Hepatic duct bifurcation	Cholangiocarcinoma
Yoshikawa (1986)	F	27	IV-A	15	12 yr	Intrapancreatic bile duct	Cholangiocarcinoma
Rossi et al. (1987) <sup>12</sup>	M	32	IV-A	29	3 yr 4 mo	Hepatic duct bifurcation	Pleomorphic giant cell carcinoma
Rossi et al. (1987) <sup>12</sup>	F	61	I	48	13 yr	Intrahepatic bile duct	Adenocarcinoma
Yamamoto (1996)	F	60	I	55	5 yr	Anastomotic site of HJ	Adenocarcinoma
Joseph et al. (1996) <sup>13</sup>	M	29	IV-A	26	3 yr	Intrahepatic bile duct	Cholangiocarcinoma
Kobayashi (1999)	F	35	IV-A	10	19 yr 6 mo	Hepatic hilar bile duct	Adenocarcinoma
Kobayashi (1999)	F	24	IV-A	15	8 yr 8 mo	Hepatic hilar bile duct	Adenocarcinoma
Kobayashi (1999)	F	18	IV-A	17	2 yr 5 mo	Intrahepatic bile duct	Adenocarcinoma
Song et al. (1999) <sup>14</sup>	F	16	IV-A	12	4 yr	Intrahepatic bile duct	Squamous cell carcinoma
Goto (2000)	F	52	I	42	10 yr	Intrahepatic bile duct	Adenocarcinoma
Eriguchi et al. (2001) <sup>15</sup>	F	42	ND	25	17 yr	Head of pancreas	Intraductal papillary adenocarcinoma
Kallel (2006)	F	70	I	56	14 yr	Intrapancreatic bile duct	Adenocarcinoma
Shimamura (2007)	M	44	IV-A	10	34 yr	Intrahepatic bile duct	Adenocarcinoma
Ono et al. (2008) <sup>16</sup>	M	26	ND	5 mo	25 yr 7 mo	Intrahepatic bile duct	Adenocarcinoma
Cho et al. (2011) <sup>17</sup>	F	41	ND	39	1 yr 1 mo	Pancreas	Cholangiocarcinoma
Cho et al. (2011) <sup>17</sup>	M	65	ND	65	6 mo	Pancreas	Cholangiocarcinoma
Lee et al. (2011) <sup>18</sup>	F	70	ND	68	1 yr 1 mo	Head of pancreas	Cholangiocarcinoma
Lee et al. (2011) <sup>18</sup>	F	40	ND	45	4 yr 5 mo	Intrahepatic bile duct	Mucinous carcinoma
Lee et al. (2011) <sup>18</sup>	F	22	ND	2	20 yr	Extrahepatic bile duct	Cholangiocarcinoma
Lee et al. (2011) <sup>18</sup>	M	58	ND	56	1 yr 2 mo	Duodenum	Cholangiocarcinoma
Our patient (2011)	F	42	ND	33	9 yr	Head of pancreas	Adenocarcinoma

ND, not described; HJ, hepatic jejunostomy.

<sup>a)</sup>According to Todani's classification.

## Conflicts of Interest

The authors have no financial conflicts of interest.

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