Ann Hepatobiliary Pancreat Surg 2016;20:201-203
https://doi.org/10.14701/ahbps.2016.20.4.201

Case Report

Cholangiocarcinoma in choledochal cyst after cystoenterostomy: how a mistreated choledochal cyst can progress to malignancy

HyungJoo Baik, Yo-Han Park, Sang Hyuk Seo, Min Sung An, Kwang Hee Kim, Ki Beom Bae, Chang Soo Choi, Sang Hoon Oh, and Young Kil Choi

Department of Surgery, Busan Paik Hospital, College of Medicine, Inje University, Busan, Korea

This case report presents an unusual case of cholangiocarcinoma arising nearly 35 years after cystoduodenostomy for choledochal cyst. The patient visited our hospital with dyspepsia and studies revealed bezoar within the choledochal cyst caused by bile and food reflux. The patient underwent pancreaticoduodenectomy and a biopsy revealed adenocarcinoma, stage IIB. After 19 months, the patient has no recurrence to date and has recovered well. This case shows that proper surgical management and meticulous, long-term follow-up is imperative for patients with congenital choledochal cyst. **(Ann Hepatobiliary Pancreat Surg 2016;20:201-203)**

Key Words: Choledochal cyst; Cholangiocarcinoma; Pancreaticoduodenectomy

INTRODUCTION

A choledochal cyst is a cystic dilatation of the biliary tree and a well-known cause of biliary tract malignancies.[1-3] It is a relatively rare disease; incidence rates ranging from 0.32% in Asia and 1 in 13,000 to 1 in 2 million births in western countries.[3,4] Most cases are diagnosed during early childhood, but between 20% to 30% of cases are first detected in adults as choledochal cyst either causing symptoms or found incidentally during imaging for an unrelated cause.[4] It has been reported that only a small number of patients who underwent surgical management for choledochal cyst had progression to biliary malignancy.[5] The incidence of cancer in patients with primary choledochal cyst is 9.9%, whereas the incidence of cancer development after cyst excision is 0.6%.[6] Herein, we present a case of a patient who had been treated for choledochal cyst in an alternative manner with no follow-up, which then progressed to cholangiocarcinoma.

CASE

A 46-year-old woman visited Busan Paik Hospital Hepatobiliary clinic with dyspepsia and right, upper quadrant, abdominal pain. The patient had a history of a cystoduodenostomy nearly 35 years ago. There was a palpable mass in her abdomen in the right, upper quadrant. Initial laboratory findings were as follows: total bilirubin 0.5 mg/dl, aspartate transaminase/alanine transaminase 16/14 IU/L, alkaline phosphatase 276 U/L, alpha-fetoprotein 2.01 ng/ml, carcinoembryonic antigen 0.756 ng/ml, carbohydrate antigen 19-9 11.70 U/ml, and hepatitis B surface antibody-positive. Abdomino-pelvic computed tomogram (APCT) scan and magnetic resonance cholangiopancreaticography (MRCP) showed moderate central dilatation of the intrahepatic duct with pneumobilia and a large, cystic, mass-like lesion with internal echogenicity and probable bezoar formation within a choledochal cyst of 6.8 cm×8.5 cm×5.0 cm in size (Fig. 1). There was also mild proximal pancreatic duct dilatation with lobulated mass suggestive of intraductal papillary mucinous neoplasm. Her esophagogastroduodenoscopy showed an

Received: May 9, 2016; **Revised:** July 15, 2016; **Accepted:** July 28, 2016
Corresponding author: Chang Soo Choi
Division of Hepatobiliary Surgery, Department of Surgery, Busan Paik Hospital, College of Medicine, Inje University, Bokji-ro 75, Busangjin-gu, Busan 47392, Korea
Tel: +82-51-890-6352, Fax: +82-51-898-9427, E-mail: changmd2000@naver.com

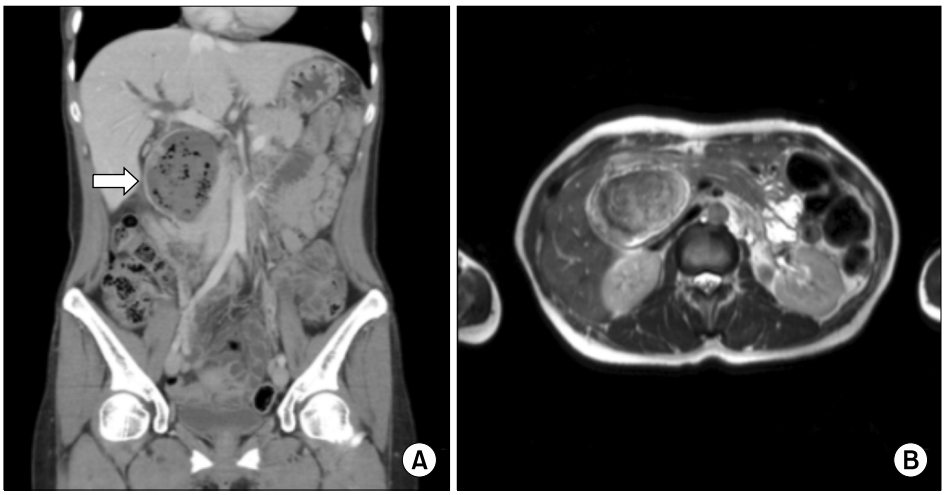

Fig. 1. Imaging studies of the patient. (A) Patient's abdomino-pelvic computed tomogram shows dilatation of intrahepatic duct, pneumobilia, and a 6.8 cm×8.5 cm×5.0 cm-sized bezoar (arrow). (B) Magnetic resonance cholangiopancreaticography also represents cystic dilatation of the extrahepatic bile duct.

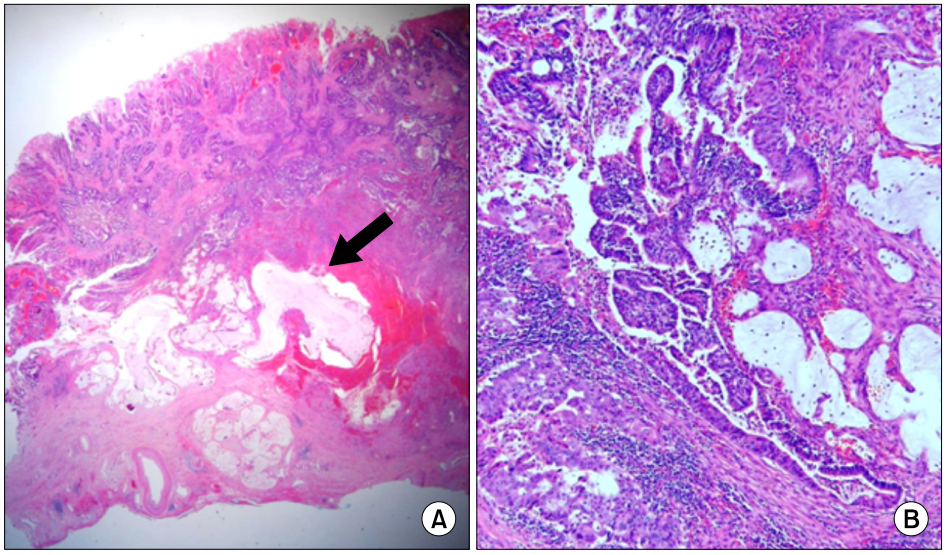

Fig. 2. Microphotographs of the specimen. (A) The common bile duct wall is thickened with tumor invasion and shows extracellular mucin pools (arrow, HE, ×10). (B) Tumor reveals glandular architecture and mucin formation (HE, ×100).

opening, at the second portion of the duodenum,with gastritis.

The initial plan for the operation was to undergo revision of the cystoduodenostomy and/or cyst excision with a hepaticojejunostomy. Because of the bezoar-like mass in the cyst, the patient was in a fasting state for 2 days prior to surgery, hoping that the mass would be dismantled. An incision was made via the previous right paramedian scar. Underneath the scar there was a severe adhesion so adhesiolysis was performed. The previously performed cystoduodenostomy was identified with multiple lymph node enlargement near the superior mesenteric vein and stomach. A small portion of the common bile duct was excised for frozen biopsy, which revealed adenocarcinoma; moreover, cystic dilatation had extended to the pancreas because of an anomalous pancreaticobiliary ductal union (APBDU). Thus, a pancreaticojejunostomy was inevitable for complete excision of the cyst and proper management for cholangiocarcinoma. Within the cyst, there was a mass: a conglomeration of food material with bile. A cholecystectomy, subtotal gastrectomy, pancreaticojejunostomy, and gastrojejunostomy were performed. The patient's extrahepatic bile ducts were unusually low-lying, so the surgeon performed a ductoplasty, conjoining the left and right extrahepatic bile ducts side to side, making it easier to carry out hepaticojejunostomy.

The final pathology report proved the presence of adenocarcinoma: moderate differentiation arising in the choledochal cyst, invading to surrounding adipose tissue (pT2a), accompanying severe inflammatory infiltration, and one metastatic lymph node out of seven excised lymph nodes, stage IIB (Fig. 2). It has been nineteen

months since the patient had her surgery and she is going through oral doxifluridine chemotherapy without any complications or recurrences to date.

DISCUSSION

Choledochal cysts are congenital anomalies of the bile ducts, which are subdivided into five different categories. The most common types are I and IVa, the dilatation of the extrahepatic bile duct and both the extrahepatic and intrahepatic bile ducts, consecutively. Asians and women tend to have a higher incidence than Caucasians and men, though clear reasons for these tendencies are yet to be elucidated.[2] In many cases, diagnosis is made early during childhood in 80%; however, due to the advance of imaging techniques, some diagnoses are incidentally made in adulthood.[4] Symptom triads are abdominal pain, jaundice, and an abdominal mass, but only rarely do these symptoms coexist. Treatment of choice is a total cystectomy and Roux-en-Y hepaticojejunostomy; less frequently, cystoenterostomy has been used as an alternative method.[1] The standard form of surgery prevents subsequent complications such as pancreatitis, cholangitis, portal hypertension and malignancy. The risk of cholangiocarcinoma in a choledochal cyst is as high as 20-30% in early adulthood if no surgical intervention is performed. On the other hand, cholangiocarcinoma following a choledochal cyst resection is relatively rare, with an incidence rate of 0.7-6%.[5]

The exact pathophysiology of cholangiocarcinoma in a choledochal cyst is still unknown, but it is suspected that the reflux of pancreatic juice is the main cause, promoting chronic inflammatory changes and carcinogenesis.[3] This patient had a Type I choledochal cyst and, due to a previous cystoduodenostomy, not only pancreatic juice but also food material had refluxed into the biliary tract, causing bezoar formation and eventually malignancy. Previous studies have emphasized the necessity of long-term follow-up for patients who have had a cyst excision, since it is known that the risk of biliary malignancy in the remnant bile duct increases more than 15 years after surgery, as in the case of our patient.[5] For those who had a cystoenterostomy,[7,8] it is mandatory that they have regular check-ups since the incidence rate of malignancy or complications such as cholangitis, hepatolithiasis, and pancreatitis could be as high as 70%.[1]

In conclusion, it should be emphasized that the standard treatment of choledochal cyst is complete cyst excision and hepaticojejunostomy. If for some reason this is not possible, a cystoenterostomy can be an alternative treatment, but meticulous life-long follow-up is essential for early detection and management of complications.

REFERENCES

1. Khandelwal C, Anand U, Kumar B, Priyadarshi RN. Diagnosis and management of choledochal cysts. Indian J Surg 2012;74: 401-406.
2. Sallahu F, Hasani A, Limani D, Shabani S, Beka F, Zatriqi S, et al. Choledochal cyst - presentation and treatment in an adult. Acta Inform Med 2013;21:138-139.
3. Kim SH, Kim HW, Kang DH, Kim MD, Lee JH, Lee JH, et al. A case of intrahepatic cholangiocarcinoma associated with type iv choledochal cyst. Korean J Gastroenterol 2012;60: 123-127.
4. Franko J, Nussbaum ML, Morris JB. Choledochal cyst cholangiocarcinoma arising from adenoma: case report and a review of the literature. Curr Surg 2006;63:281-284.
5. Ohashi T, Wakai T, Kubota M, Matsuda Y, Arai Y, Ohyama T, et al. Risk of subsequent biliary malignancy in patients undergoing cyst excision for congenital choledochal cysts. J Gastroenterol Hepatol 2013;28:243-247.
6. Lee SE, Jang JY, Lee YJ, Choi DW, Lee WJ, Cho BH, et al. Choledochal cyst and associated malignant tumors in adults: a multicenter survey in South Korea. Arch Surg 2011;146: 1178-1184.
7. Ong J, Campbell W, Taylor MA. Metastatic cholangiocarcinoma following choledochal cyst excision: an unusual cause of abdominal pain in a 35-year-old female. Ulster Med J 2013;82: 21-22.
8. Ono S, Fumino S, Shimadera S, Iwai N. Long-term outcomes after hepaticojejunostomy for choledochal cyst: a 10- to 27-year follow-up. J Pediatr Surg 2010;45:376-378.