CASE REPORT

Biliary cancer developed after the reparative surgery for congenital choledochal cyst: a case report and review of the literature

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Received: 12 October 2016 / Accepted: 6 November 2016 / Published online: 12 November 2016
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Abstract The incidence of bile duct cancer developed in the patients with congenital choledochal cysts (CC) with a history of reparative surgery is not well known. We experienced a case developed choleductal cancer 45 years after reparative surgery. She underwent reparative surgery with cyst excision and hepatic bile duct duodenal anastomosis at 1 year of age. She developed the symptoms of jaundice, anorexia, and dull pain in the right upper part of the abdomen at 47 years of age. The carcinoma arose from the dilated proximal bile duct anastomosed with the duodenum. Cholestasis and regurgitation of duodenal fluids seemed to have influenced the development of cancer in this patient. We additionally reviewed seven cohort studies concerning the incidence of biliary carcinoma after surgery for congenital choledochal cysts. The incidence of biliary cancer developed after surgery was 2.2 ± 2.5 (ranged 0–6.5)%. We also reviewed 33 Japanese case reports cited in Japan MEDLINE from 1986 to 2015. Regarding the CC types according to the Totani's classification, 12 were type I and 14 type IVa, 1 was type II and 6 were unknown type. The rate of coexistence of pancreaticobiliary maljunction (PBM) was 92%, (22/24; other 9 cases were not documented), and biliary cancer

arose from the proximal stump of the reparative surgery in 68% (17/25) and from the distal stump in 32% (8/25) of cases. These findings suggested that the reflux of pancreatic juice due to PBM only partially explained the carcinogenesis of CCs. Intensive follow-up of such patients throughout their lives is necessary to avoid cancer death even after standard reparative surgery.

Keywords Congenital choledochal cyst · Biliary cancer · Reparative surgery

Background

Choledochal cysts (CCs) are a rare congenital biliary duct abnormality, first described by Vater in 1948 [1]. The incidence of CCs was approximately 1/13,000 which is much more frequent than in Western country, where the incidence rate is reported 1/100,000 [2].

Most patients with CCs are accompanied with the pancreaticobiliary maljunction (PBM) at approximately 90% and frequently develop biliary cancer [3–5]. The rate of developing malignant disease can be reduced with standard reparative operations such as cholecystectomy, bile duct resection and bile duct reconstruction [6], although internal drainage fails to mitigate the risk of malignant change [7]. However, there are several reports of cases that had underwent reparative operation and developed biliary cancer after the long year follow-up. The incidence and features of bile duct cancer developed in the patients with CCs with a history of reparative surgery are not well known. We herein report a case of choleductal cancer 45 years after the reparative surgery. We are conducted a literature review of cancer development in operated cases with CCs.

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Case report

A 47-year-old female were referred to our hospital for the jaundice and anorexia in 2015. She underwent reparative operation of common bile duct duodenal anastomosis at 1 year of age. The type of CC and the extent of the resected bile duct were unknown. She had previously been treated for left breast cancer by breast-conserving therapy at 42 years of age, and was subsequently treated with hormone therapy for 5 years. She had no family members with bile duct disease, nor any history of bile duct infection or other disease.

Her hight and weight were 161 cm and 60 kg, respectively. And she had symptoms of jaundice, anorexia, and dull pain in the right upper part of the abdomen. The laboratory data at admission showed elevated values of T-Bil at 15 mg/dl (D-Bil 11.4 mg/dl) and AST 129 U/l, ALT 116 U/l, ALP 1965 U/l, and LDH 175 U/l. The serum tumor markers of CEA and CA19-9 were not elevated with the value of CEA 4.1 ng/ml and CA19-9 2.0 U/l, respectively.

Contrasted computed tomography (CT) at the time of the breast operation showed pneumobilia and dilatation of the intrahepatic bile duct, and the finding of common bile duct duodenal anastomosis (Fig. 1a). CT at the most recent admission revealed further dilation of the hepatic duct, a solid tumor at the hepatic hilum measuring 4.0 cm in diameter, and swelling of the lymph nodes of the hepato-duodenal ligament (Fig. 1b). The margin between the tumor and pancreas head was well defined, suggesting that the tumor arose from the upper common bile duct anastomosed to the duodenum. Magnetic resonance cholangiopancreatography (MRCP) showed a dilated intrahepatic

bile duct, and PBM, but the remnant of the dilated bile duct was unclear to define (Fig. 2).

The opened orifice of the bile duct duodenal anastomosis was identified at the protruding bulb by duodenal fiberscope. The patient underwent percutaneous transhepatic choleductal drainage (PTCD) and laparotomy after the T-Bil level dropped to 2.89 mg/dl.

The operative findings were as follows: the hepatic hilum tumor measuring 5.0 cm in diameter had invaded to the liver and enlarged lymph nodes were seen in the hepato-duodenal ligament and anterior surface of the pancreas head. Therefore, we performed pancreatoduodenectomy and partial resection of the liver to achieve R0 resection. Billroth 2 (modified Child) reconstruction was done.

The macroscopic findings showed that a tumor measuring 50 mm × 30 mm invaded to the liver and duodenal anastomosis (Fig. 3a, b). The pathological findings indicated moderately differentiated adenocarcinoma (Fig. 4a) of nodular-infiltrating type with lymph vessel invasion (Fig. 4b), vascular invasion (Fig. 4c) and perineural invasion (Fig. 4d). The pathological TNM classification was pT3a (liver and duodenum) pN1 M0 Stage B. Additionally, immunohistochemical staining showed over-expression of P53 protein in several tumor cells and a high labeling index of Ki67. No staining of P53 protein was noted in the epithelial cells of the common bile duct close to the cancer and PBM.

Although the post-operative course was uneventful, she developed liver metastases and peritoneal dissemination, 6 months after the operation. Despite treatment with gemcitabine and S1, she died of progressive disease, 14 months after the diagnosis of bile duct cancer.

Fig. 1 Computed tomography at the time of breast operation showed pneumobilia and dilatation of intrahepatic bile duct, and the finding of common bile duct duodenal anastomosis (a). CT at admission

revealed further dilated hepatic duct, and solid tumor at the hepatic hilum measuring 4.0 cm in diameter (b)

Fig. 2 MRCP showed dilated intrahepatic bile duct, and PBM (circled), and the remnant of dilated bile duct was unclear to define

Discussion and review of the literature

CC is closely associated with PBM which can allow the pancreatic juice to reflux into the common bile duct, and related with carcinogenesis [7]. The frequency of PBM in patients with CCs is reported to be approximately 90% [4, 5].

Many reports have demonstrated that internal drainage without resection of CCs carries a high morbidity rate and an increased risk of biliary tract cancer with increasing age. The incidence of cancer associated with CCs and PBM was reported to be 16.2% in Japan [8], and 11.4% in a review of the global literatures [2].

However, the cause of carcinogenesis in such case has not been well elucidated, and pancreatic juice reflux into the biliary duct, combined with cholestasis and repeated

infections, is considered to cause the malignant change of the biliary epithelium [9, 10].

An increasing trend in p53 level and Ki-67 values from metaplasia to biliary intraepithelial neoplasia to carcinoma has been reported [11]. In the present case, immunohistochemistry revealed over-expression of P53 in only some cancer cells, and on a few epithelial cells of the bile duct and PBM close to the tumor. A high Ki67 labeling index approximately 90% of Ki67 was seen in cancer cells. The remnant of CC was not identified in pathological examinations.

The incidence of biliary cancer associated with CC ranges from 8.4 to 17.5% tending to increase with age [5]. However, the incidence of biliary cancer in these patients after reparative surgery is unclear. We summarized the findings of seven reports concerning the follow-up study (Table 1) [8, 12–17]. The incidence of biliary cancer developed after surgery was 2.2 ± 2.5 (ranged 0–6.5) %. However, the follow-up periods varied, making it difficult to definitively determine the incidence rate. Among these reports, Ohashi et al. reported that the cumulative incident rates after 15, 20, and 25 years were 1.6, 3.9, and 11.3%, respectively [15].

We also reviewed 33 cases of biliary cancer developing after reparative surgery in the Japanese literature cited in the Japanese MEDLINE databases (from 1986 to 2015), including the present case (Table 2).

Concerning gender, female was more often affected than male around at 2–3 times (female 22, male 8, unknown 3). The types of CCs according to Totani's classification were 12 type I, 14 type IVa, 1 type II, and 6 of unknown type (Table 2).

The mean ages ($\pm$ standard deviation) at the time of reparative surgery and at the diagnosis of biliary cancer

Fig. 3 The macroscopic findings showed tumor measuring 50 mm $\times$ 30 mm invaded to the liver and duodenal anastomosis (circled)

Fig. 4 Pathological finding (HE stain) indicated moderately differentiated adenocarcinoma nodular-infiltrating type (**a** $\times 200$) with lymph vessel invasion (**b** $\times 100$), vascular invasion (**c** $\times 400$), and perineural invasion (**d** $\times 200$)

Table 1 Incidence of bile duct cancer developed after reparative surgery for congenital cysts

Author	Country	Year (Refs)	No. of cases ^a	Analyzed cohort	No. of patient developed bile duct cancer	Incidence of cancer (%) developed after the surgery
Ishibashi	Japan	1997 [8]	39	Single institute	0	0
Kobayashi	Japan	1999 [12]	46	Single institute	3	6.5
Ono	Japan	2010 [13]	53	Single institute	0	0
Takeshita	Japan	2011 [14]	144	Single institute	1	0.7
Ohhashi	Japan	2012 [15]	94	Single institute	4	4.3
Xiao-doing	China	2014 [16]	200	Single institute	1	0.5
Soares	USA	2015 [17]	394	Multi-institute	13	3.3

^a Number of patients without cancer at the surgery

were 23 ± 18 (ranged 1–61), 41 ± 16 (ranged 11–73) years, respectively. On comparing between type I and IVa, the mean ages of surgery were 35 ± 20 vs 13 ± 16 ($p = 0.01$), and developing cancer was 51 ± 11 vs 32 ± 12 ($p = 0.05$) years (Table 2). The patients with type IVa CCs underwent reparative surgery and developed

biliary cancer at a significantly younger age than the patients with type I CCs.

The rate of coexistence of PBM was 92%, (22/24; other 9 cases were not documented) and biliary cancer arose from the proximal stump of reparative surgery in 68% (17/25) and from the distal stump at 32% (8/25) of cases. These

Table 2 Literature review of 33 Japanese cases with CCs after reparative surgery

Case	Author	Year (Refs)	Age at reparative surgery	Gender	Type of CCs ^a	Acompany of BPM	Age at deveoping cancer	Site of cancer (P or D) ^b	Time interval (year)
1	Simoda	1992 [18]	1	F	IVa	+	11	nd	10
2	Yoshikawa	1986 [19]	1	F	IVa	+	14	nd	13
3	Kamizawa	1991 [20]	1	F	nd	nd	54	P	53
4	Kobayashi	2013 [21]	1	F	II	–	56	D	55
5	Fujisaki	1999 [22]	5	F	I	+	22	nd	17
6	Sugitou	1999 [23]	5	F	IVa	+	22	nd	17
7	Takadai	2008 [24]	9	M	IVa	+	32	nd	23
8	Shimamura	2009 [25]	10	M	IVa	+	44	P	34
9	Kumamoto	2014 [26]	12	F	IVa	+	40	D	28
10	Ohhashi	2013 [15]	12	M	I	nd	44	P	32
11	Chibana	2004 [27]	13	F	nd	nd	38	P	25
12	Ohhashi	2013 [15]	14	F	IVa	nd	27	D	13
13	Ohhashi	2013 [15]	15	F	I	nd	38	P	23
14	Yoshikawa	1986 [19]	15	F	IVa	+	27	P	12
15	Ishido	2010 [28]	15	F	I	–	28	P	13
16	Kobayashi	1999 [29]	15	F	IVa	+	35	P	20
17	Kobayashi	1999 [29]	16	F	IVa	+	18	P	2
18	Kitaguchi	2015 [30]	17	na	IVa	+	20	P	3
19	Suzuki	2015 [31]	20	M	IVa	+	46	P	26
20	Yamataka	1997 [32]	21	M	nd	nd	25	nd	4
21	Saito	2015 [33]	23	F	I	+	46	nd	23
22	Tsujiuchi	1991 [34]	28	F	IVa	+	36	P	8
23	Souda	1994 [35]	32	M	nd	nd	42	D	10
24	Eriguchi	2001 [36]	35	F	nd	nd	42	D	17
25	Goto	2001 [37]	42	F	I	+	52	P	10
26	Iwaisako	2007 [38]	46	F	I	+	54	D	8
27	Mizuguchi	2015 [39]	46	nd	I	+	69	D	33
28	Ohhashi	2013 [15]	50	M	I	nd	66	P	16
29	Sakamoto	2014 [40]	51	F	I	+	64	P	13
30	Yamomoto	1996 [41]	55	F	I	+	60	D	5
31	Sendo	2007 [42]	61	M	I	+	68	nd	7
32	Chuma	2014 [43]	61	nd	IVa	+	73	P	12
33	Present case		1	F	Unknown	+	46	P	45

nd not documented, P proximal bile duct, D distal bile duct

^a According to Totani's Classification

findings suggested that the reflux of pancreatic juice due to PBM only partially explained the carcinogenesis due to CCs. Furthermore, in contrast with the general incidence of CCs, where type I is three times more frequent than type IVa [17], the patients with type IV CCs developed carcinoma much more frequently than those with type I CCs. These facts suggested that the part of the CCs remaining after reparative surgery may serve an origin of carcinoma.

Of note, in the present case hepatic duct was anastomosed to the duodenum at 1 year of age and developed biliary cancer after a 45-year interval. The carcinoma arose

from the dilated proximal bile duct not from the pancreatic side of the residual bile duct or PBM. The carcinogenesis of this case should be considered distinguished from the case after the Roux-en Y jejunostomy. A dilated hepatic duct and pneumobilia suggested that cholestasis and regurgitation of duodenal fluids influenced the development of cancer in this patient. The biliary epithelia was immersed in the mixture of food, bile, pancreatic juice and intestinal fluid. This situation might be identical to that after internal drainage. Although the remnant CC was not noted on pathological examinations, we have another

possibility, in that the carcinoma developed from the remnant CC that had been anastomosed to the duodenum at the initial surgery.

Given the above, clinicians should understand the need for intensive follow-up of such patients throughout their life to avoid death from cancer even after standard reparative surgery. However, constitution of the follow-up program during very long period over 20 years seems to be difficult. Therefore, we propose the necessity of the following practices which are: education of the cancer risk for the patients and linking of the pediatrician to a physician who can check up adult patients. Furthermore, we hope the registration system constituted by Japanese Study Group on Pancreaticobiliary Maljunction (JSPBM) [44] to take a part of a life-long survey of these patients and to clarify the true incidence of carcinoma development after reparative surgery for CCs.

Conclusions

We experienced a 47-year-old female who developed biliary cancer 45 years after reparative surgery for CC. The cancer developed at the proximal side of the common bile duct anastomosed with the duodenum. Life-long survey of these patients is needed to avoid cancer death, even after reparative surgery.

Compliance with ethical standards

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration. Written informed consent was obtained from the patient for publication of this case and any accompanying images.

Conflict of interest The authors declare no conflicts of interest (COI).

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