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Small Intestinal Adenocarcinoma Arising at the Anastomotic Site after Kasai Operation for Biliary Atresia: A Case Report and Literature Review

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Biliary atresia is an obliterative cholangiopathy of unknown etiology. Hepatic portoenterostomy, in which obliterated extrahepatic bile ducts are resected and bile flow is restored, known as Kasai operation, is performed within 3 months after birth. While this operation enhances long-term survival of patients, the occurrence of primary malignant hepatic tumors has been increasing. We report a case of small intestinal adenocarcinoma arising at the anastomotic site after Kasai operation. A 49-year-old man, who underwent Kasai operation for biliary atresia when he was 2 months old, experienced rapidly progressive jaundice and liver dysfunction. Deceased-donor liver transplantation was performed for liver failure. Macroscopically, there was a white-yellow tumor located at the anastomotic site of hepatic portoenterostomy of the resected liver. Pathological examination revealed a well-differentiated adenocarcinoma with some Paneth cells in the neoplastic lesion. Immunohistochemically, the tumor cells were negative for cytokeratin 7 (CK7) but positive for cytokeratin 20 (CK20) and a homeobox domain-containing transcription factor (CDX2). Mucin expression in tumor cells was negative for mucin 1 (MUC1) and mucin 6 (MUC6) and positive for mucin 2 (MUC2) and mucin 5AC (MUC5AC). The pathological diagnosis was small intestinal adenocarcinoma originating from the jejunum. The patient was discharged 48 days after the operation. The patient had not experienced recurrence at 10 months after the operation. This is the first report of small intestinal adenocarcinoma arising at the anastomotic site after Kasai operation for biliary atresia. Special care should be taken for the patients after Kasai operation with acute progressive jaundice and liver dysfunction because there is a possibility of malignancy in their native liver.

Keywords: biliary atresia; Kasai operation; liver transplantation; malignant tumor; small intestinal adenocarcinoma
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Introduction

Biliary atresia (BA) is an idiopathic cholangiopathy that develops in neonates and causes the progressive inflammatory obliteration of extrahepatic bile ducts. Untreated BA will lead to biliary cirrhosis and liver failure in the first or second year of their lives. Hepatic portoenterostomy, known as Kasai operation, is necessary for survival. Kasai operation was first developed for the treatment option of BA in 1950 and significantly improved patients' prognosis. However, after Kasai operation, half of the patients require liver transplantation (LT) before adulthood because of liver failure, hepatopulmonary syndrome, portal hypertension, and repeated cholangitis (Kasahara et al. 2017; Nio 2017; Miyagi et al. 2022b). There are some reports of primary malignant hepatic tumors after Kasai operation. Hepatocellular carcinoma (HCC) is the most common, followed by cholangiocarcinoma and hepatoblastoma (Van

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Wyk et al. 1972; Tamura et al. 1993; Kohno et al. 1995; Tatekawa et al. 2001; Azuhata et al. 2003; Taat et al. 2004; Brunati et al. 2007; Hol et al. 2008; Hadžić et al. 2011; Aggarwal et al. 2012; Kim et al. 2012; Vera et al. 2012; Fukuda et al. 2013; Yoon et al. 2014; Arai et al. 2016; Nio et al. 2019; Uno et al. 2020). Here, we report the first case of small intestinal adenocarcinoma arising at the anastomotic site after Kasai operation for BA.

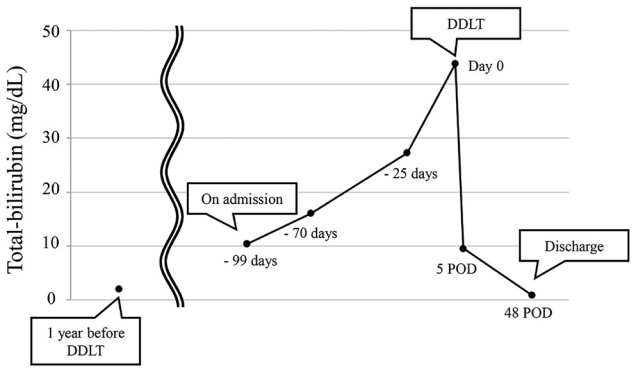

Fig. 1. Preoperative time course changes of the serum level of total bilirubin.
One year before liver transplantation (LT), the serum level of total bilirubin was 2.1 mg/dL. On admission, the serum level of total bilirubin level increased to 10.4 mg/dL and dramatically increased to 43.8 mg/dL just before LT. It immediately decreased to the normal level after the operation. DDLT, deceased-donor liver transplantation; POD, post operative day(s).

Case Presentation

The patient was a man who underwent Kasai operation for BA 2 months after birth. The postoperative course was good, and he only experienced several episodes of cholangitis in his adulthood without admission. When he became 49 years old, the sudden onset of jaundice and the progressive elevation of hepatobiliary enzymes occurred. He was hospitalized with cholangitis, and his jaundice and liver dysfunction progressed rapidly. Serum total bilirubin was 10.4 mg/dL (reference range 0.4-1.5 mg/dL) on admission and dramatically increased up to 43.8 mg/dL (Fig. 1). The elevation of other hepatobiliary enzymes and the decrease of synthetic capacity of the liver was detected. The elevation of carcinoembryonic antigen (CEA) and protein induced by vitamin K absence or antagonist-II (PIVKA-II) was also observed (CEA 7.0 ng/mL, reference range < 5.0 ng/mL; PIVKA-II 12,351 mAU/mL, reference range < 40 mAU/mL).

Enhanced computed tomography (CT) showed the rapid progression of intrahepatic bile duct dilation within two months after the onset of jaundice (Fig. 2A, B), and the abdominal X-ray showed the appearance of massive ascites (Fig. 2C, D). There was no apparent stone or mass detected by CT or magnetic resonance imaging. LT was necessary

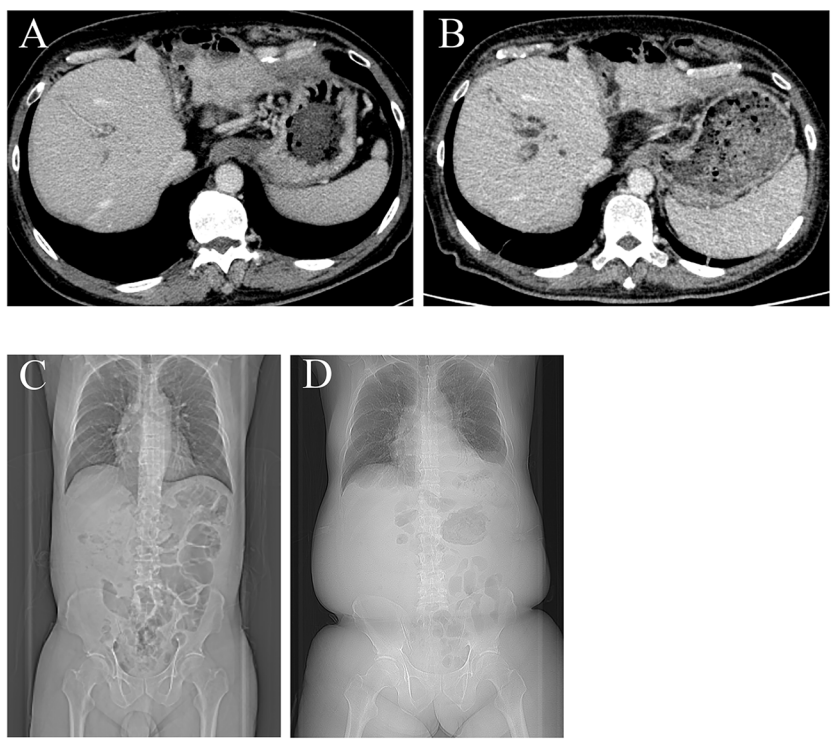

Fig. 2. Preoperative time course changes of imaging findings.
(A, B) Enhanced computed tomography imaging on admission (A) and before liver transplantation (LT) (B). Rapid progression of dilatation of the bile duct was observed. (C, D) Abdominal X-ray on admission (C) and before LT (D). Massive ascites was observed just before LT.

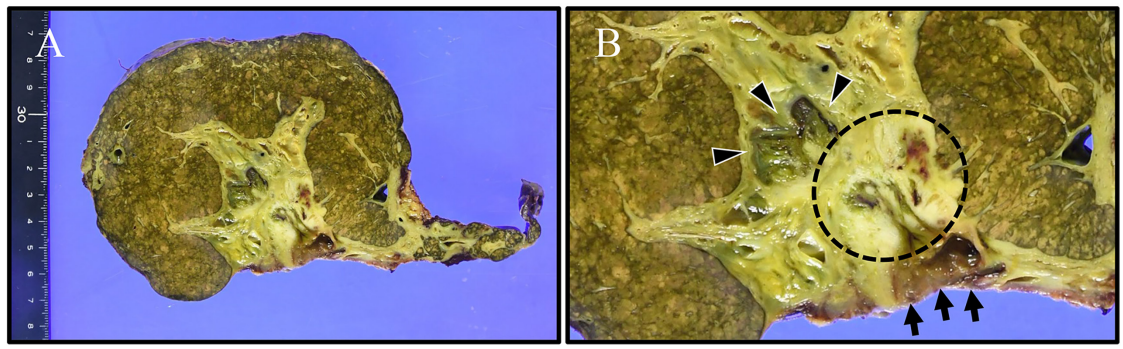

Fig. 3. Macroscopic findings of the resected liver.
(A) The atrophy of the left lobe and biliary cirrhosis was observed. (B) A white-yellow tumor with indistinct boundaries (inside of the dotted line) was located at the anastomotic site of the hepatic portoenterostomy (arrows, jejunal loop; arrowheads, bile duct).

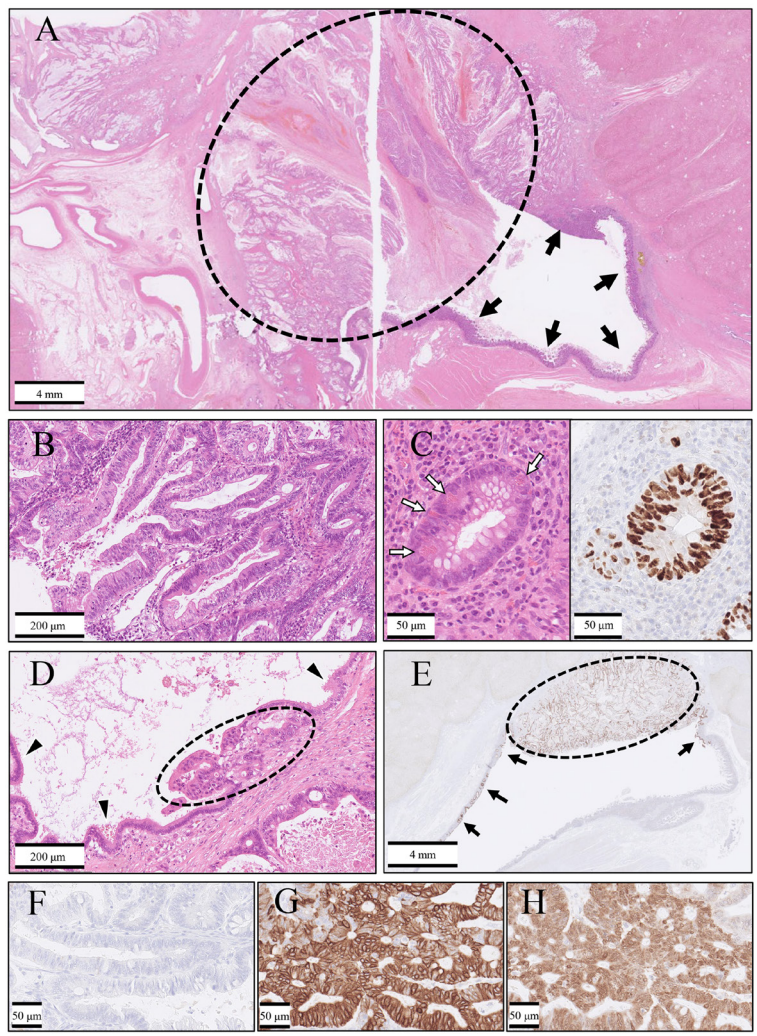

Fig. 4. Pathological findings of the lesion.
(A) Predominant localization of the lesion (inside of the dotted line) at hepatic portoenterostomy (arrows, jejunal loop) revealed by hematoxylin and eosin (H&E) staining. (B) Observation of well-differentiated adenocarcinoma. (C) H&E staining (left side) and p53 immunostaining (right side) of the identical tumor gland obtained from serial sections. Paneth cells in the neoplasm (white arrows). (D) Carcinoma cells (inside of the dotted line) displaced the bile duct epithelium (arrowheads). (E) p53-positive carcinoma in situ (arrows) in the jejunal loop continuously linked to the advanced lesion (inside of the dotted line). (F, G, H) Immunohistochemistry reveals CK7-negative staining in carcinoma cells (F). Diffuse positivity for CK20 (G) and CDX2 (H).

Table 1. Primary malignant tumors after hepatic Kasai operation for biliary atresia.

Tumor type	Small intestinal adenocarcinoma	Hepatocellular carcinoma	Cholangiocarcinoma	Hepatoblastoma
Number of reports	1	17	6 (3 PCCs and 3 ICCs)	2
Age (range)	49	7 (0-39)	23.5 (13-63)	2 and 4
Sex (M:F)	1:0	7:10	2:4	0:2
Elevation of tumor markers (%)	1 (100%)	7 (41%)	3 (50%)	2 (100%)
Diagnosis before operation	None	15	3 (One PCC and 2 ICCs)	2
The number of LT	1	11	4	1
Follow-up period, month (range)	10	18 (2-72)	9.5 (6-180)	Not addressed
Mortality	0	5	4	1
References	The current case	Van Wyk et al. 1972 Tamura et al. 1993 Kohno et al. 1995 Tatekawa et al. 2001 Azuhata et al. 2003 Brunati et al. 2007 Hol et al. 2008 Hadžić et al. 2011 Aggarwal et al. 2012 Kim et al. 2012 Yoon et al. 2014 Arai et al. 2016	Vera et al. 2012 Fukuda et al. 2013 Yoon et al. 2014 Arai et al. 2016 Nio et al. 2019 Uno et al. 2020	Tatekawa et al. 2001 Taat et al. 2004

ICC, intrahepatic cholangiocarcinoma; LT, liver transplantation; PCC, perihilar cholangiocarcinoma.

for lifesaving because of the progressive jaundice and liver failure (The Model for End-Stage Liver Disease score was 30 and Child-Pugh classification was categorized as class C). Deceased-donor LT (DDLT) was performed. The recipient's abdominal cavity was highly adhesive with clear ascites. The recipient's liver was removed, and the liver graft was transplanted. With gross observation, his left lobe of the liver was atrophic, and the cut surface revealed biliary cirrhosis (Fig. 3A). A white-yellow tumor measuring 30 mm in diameter with indistinct boundaries was observed at the anastomotic site of the hepatic portoenterostomy (Fig. 3B).

The pathological examination revealed well-differentiated adenocarcinoma containing Paneth cells, and the tumor was located mainly in the anastomotic site of hepatic portoenterostomy (Fig. 4A-C). On the hepatic side, the tumor invaded the hepatic hilum and the hepatic parenchyma. The tumor cells displaced the bile duct epithelium, while intraductal papillary neoplasm of bile duct was not observed (Fig. 4D). On the jejunal side, the tumor invaded the muscularis propria at its deepest point, and p53-positve carcinoma in situ was also observed continuously linked to the advanced lesion (Fig. 4E). To determine the origin of the tumor, immunohistochemical staining was performed. The tumor cells were diffusely positive for cytokeratin 20 (CK20) and a homeobox domain-containing transcription factor (CDX2), whereas negative for cytokeratin 7 (CK7) (Fig. 4F-H). In addition, the cells were diffusely positive for mucin 2 (MUC2), partially positive for mucin 5AC (MUC5AC), and negative for mucin 1 (MUC1) and mucin 6 (MUC6). From these results, the pathological diagnosis was small intestinal adenocarcinoma originating from the jejunal loop, pT4 cN0 cM0 Stage IIB (UICC-TNM, 8th edition) (Brierley et al. 2017). Although CT-guided drainage was performed for the intra-abdominal abscess, the postoperative course was uneventful and the patient discharged 48 days after the operation. No recurrence has been detected for 10 months after the operation without any adjuvant chemotherapy.

Discussion

BA is an idiopathic cholangiopathy of neonates and early infancy characterized by obstruction of the extrahepatic bile ducts, and its incidence rate is about 1/10,000 live births (Kasahara et al. 2017; Nio 2017). Kasai operation is performed for BA within 3 months, and the earlier the operation is performed, the better the prognosis and native liver function would be (Jimenez-Rivera et al. 2013). As the prognosis of the patients after Kasai operation improves, the occurrence of postoperative primary malignant hepatic tumors has been increasing. Although there are some reports of intrahepatic malignancy after Kasai operation, there is no other report of small intestinal adenocarcinoma originating from the jejunal loop at the anastomotic site.

It is sometimes difficult to determine the origin of the tumor of hepatic hilum. In our case, the tumor was located mainly in the bile duct and caused obstructive jaundice, so cholangiocarcinoma was highly suspected at first (Fig. 3). On pathological examination, the tumor contained Paneth cells, which are observed in the small intestinal tissue. In addition, the tumor continued from the surface of the jejunum to hilum of the liver. Immunohistochemical staining

also showed that the tumor was CDX2-positive, CK7-negative, and CK20-positive staining patterns, which indicates the diagnosis of intestinal tumors (Park et al. 2007). MUC1-negative, MUC2-positive, and MUC5AC-positive expression patterns also align with the characteristics of intestinal tumors (Lau et al. 2004).

Small intestinal adenocarcinoma is a rare malignant tumor, and its incidence rate is reported to be less than 0.001% (Li et al. 2016). Inflammatory bowel diseases such as ulcerative colitis and Crohn's disease have been reported as the risk factors for small intestinal adenocarcinoma (Goodman et al. 2013). In this case, the chronic inflammation at the anastomotic site caused by repeated cholangitis could be associated with developing the tumor after Kasai operation. On the other hand, intrahepatic tumors including HCC and intrahepatic cholangiocarcinoma, could be caused by cholestatic liver cirrhosis (Arai et al. 2016).

In this case, the tumor was coincidentally found by the pathological examination after LT. Retrospectively, there were two preoperative findings suspicious for the presence of the tumor. Firstly, the obstructive jaundice and the dilatation of the intrahepatic duct had progressed rapidly in a short period (Fig. 1). Secondly, the level of CEA, which is a nonspecific tumor marker for small intestinal adenocarcinoma, was elevated (Chen and Vaccaro 2018). There are some reports that balloon-assisted endoscopic retrograde cholangiopancreatography (ERCP) was useful for obstructive jaundice after Roux-en-Y reconstruction (Liu et al. 2017). Although the success rate of ERCP after Kasai operation is low, there are some reports that ERCP was effective in controlling cholangitis after Kasai operation (Liu et al. 2017; Hyun et al. 2018). Therefore, the balloon-assisted ERCP could be one of the options for controlling obstructive jaundice and the diagnosis of the tumor in our case. Table 1 shows the summary of the reports searched by Pubmed with the keywords "Biliary atresia," "Kasai operation" and "Malignant tumor," and the references of those reports.

The tumor markers were also elevated in half of the cases of malignancy after Kasai operation (Table 1). Since primary malignancies after Kasai operation occurred at younger age compared with natural populations, careful follow up should be done using imaging examinations and tumor markers.

LT for patients with malignancy is controversial. The effectiveness of LT for some malignancies, including HCC, hepatoblastoma, and hilar cholangiocarcinoma are reported (Mantel et al. 2016; Shimamura et al. 2019; Miyagi et al. 2022a; Srinivasan et al. 2023). In our case, the preoperative diagnosis was difficult because the imaging tests could not detect any tumor, and it was difficult to distinguish the obstructive jaundice from the jaundice caused by liver failure. The accuracy of preoperative diagnosis of the tumor located in the liver hilum was extremely low after Kasai operation (Table 1). We are using everolimus as an immunosuppressant, which was reported to reduce the risk of recurrence after LT for HCC (ALoun et al. 2023). Postoperative management of small intestinal carcinoma was unclear because of the small number of cases (Li et al. 2016). Further studies are warranted to establish the treatment and predict the prognosis of primary malignant tumors arising after Kasai operation.

In conclusion, we experienced a case of small intestinal adenocarcinoma arising at the anastomotic site after Kasai operation. The number of malignancies after Kasai operation is increasing because of the improving prognosis. Special care should be taken for the patient with rapidly progressive jaundice and the elevation of tumor markers.

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Conflict of Interest

The authors declare no conflict of interest.

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