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A case of biliary cystadenocarcinoma arising in the liver with a congenital retention of indocyanine green.

Kiyotoshi Gotoh* Eiji Konaga[†] Hitoshi Takeuchi[‡]

Shouhei Mano** Horishi Kohno^{††}

*Iwakuni National Hospital,

[†]Iwakuni National Hospital,

[‡]Okayama University,

**Iwakuni National Hospital,

^{††}Okayama University,

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Abstract

A case of biliary cystadenocarcinoma that occurred in a 45-year-old woman is reported. Ultrasonography and computed tomography clearly revealed papillary projections in the cyst of the liver. Percutaneous transhepatic cystography showed connection between the cyst and the common bile duct. The tumor was surgically resected and proved to be a mucinous papillary adenocarcinoma arising from a biliary cystadenoma. The patient is doing well 4 years after surgery. Interestingly, this is the first reported case of a biliary cystadenocarcinoma in the liver with markedly diminished excretion of indocyanine green.

KEYWORDS: biliary cystadenocarcinoma, indocyanine green

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A Case of Biliary Cystadenocarcinoma Arising in the Liver with a Congenital Retention of Indocyanine Green

Kiyotoshi Gotoh*, Eiji Konaga, Hitoshi Takeuchi, Shouhei Mano^a and Hiroshi Kohno^b

Department of Surgery, ^aDepartment of Pathology and ^bDepartment of Internal Medicine, Iwakuni National Hospital, Iwakuni 740, Japan

A case of biliary cystadenocarcinoma that occurred in a 45-year-old woman is reported. Ultrasonography and computed tomography clearly revealed papillary projections in the cyst of the liver. Percutaneous transhepatic cystography showed connection between the cyst and the common bile duct. The tumor was surgically resected and proved to be a mucinous papillary adenocarcinoma arising from a biliary cystadenoma. The patient is doing well 4 years after surgery. Interestingly, this is the first reported case of a biliary cystadenocarcinoma in the liver with markedly diminished excretion of indocyanine green.

Key words : biliary cystadenocarcinoma, indocyanine green

Imaging techniques are rapidly advancing, making evaluation of cystic lesions of the liver more facile. Biliary cystadenocarcinoma, a malignant cystic tumor, is very rare. We documented a case of biliary cystadenocarcinoma, the diagnosis of which was aided by ultrasonography (US) and computed tomography (CT). If cystic lesions with papillary projections are delineated in the liver by US or CT, current recommendations call for surgical resection of these cystic tumors, regardless of whether they are benign or malignant on histologic examination. Because biliary cystadenomas have the potential for malignant transformation. Our case shows such evidence.

A 45-year-old woman was admitted to Iwakuni National Hospital because of abdominal distension. On admission, the liver was palpable

two finger-breadths below the right costal margin; the patient was anicteric and without ascites. Biochemical tests were unremarkable, except for the indocyanine green (ICG) test, which revealed decreased excretion. Laboratory data were as follows: GOT, 13 IU; GPT, 9 IU; ALP, 5.2 KA. U; LDH, 200 Wro. U; total bilirubin, 0.9mg/dl; ICG R15-value, 99.8 % ICG K-value, 0.009; BSP (bromsulphalein) R 45-value, 10 %. From the discrepant results between ICG and BSP clearance tests, *i. e.*, an abnormal ICG-test and a normal BSP-test, we concluded this patient had a congenital disturbance of ICG excretion. Tumor markers, such as alphafetoprotein, carcinoembryonic antigen and cancer antigen 19-9, were within normal limits. Past history was significant for acute hepatitis at the age of 32. At this admission, hepatitis B surface (HBs-) antigen was positive, and HBs-antibody was negative.

* To whom correspondence should be addressed.

US disclosed a large, well-demarcated cystic tumor with papillary projections in the left lobe of the liver (Fig. 1-a). Enhanced CT also demonstrated a multilocular tumor and internal papillary projections in the largest cyst (Fig. 1-b). Celiac angiography showed prominent neovascularity (Fig. 2-a). Percutaneous transhepatic cystography (PTC) delineated the connection between the cyst and common bile duct (Fig. 2-b). A tentative diagnosis of biliary cystadenocarcinoma was made from these image findings; cytologic study

of PTC aspiration was negative.

In January 1988, the tumor was surgically resected by left lobectomy. There was no intraperitoneal dissemination, and lymphnodes were not metastasized. The surgical specimen contained multilocular cysts, packed with yellow mucinous material. Papillary projections were observed in the largest cyst (Fig. 3).

The histopathologic diagnosis was biliary cystadenocarcinoma. The cyst wall was made up of fibrous connective tissue contiguous with

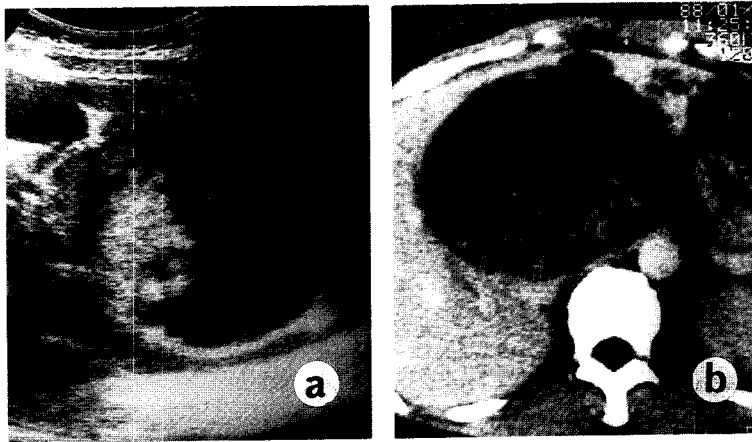


Fig. 1 Imaging diagnosis of the cystic tumor with papillary projections; (a) ultrasonography, (b) enhanced CT.

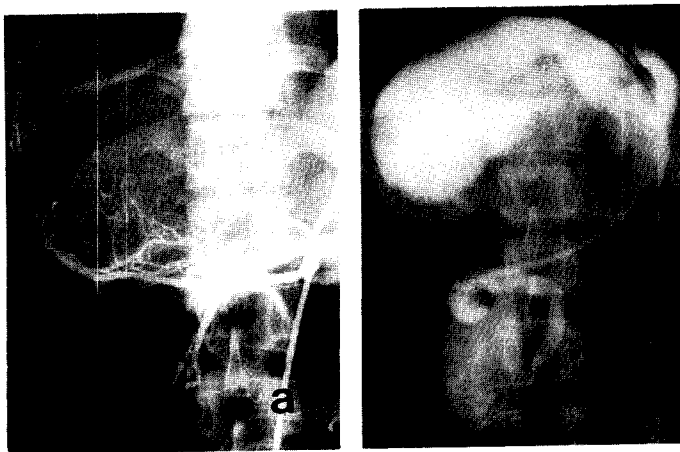


Fig. 2 Imaging diagnosis of biliary cystadenocarcinoma; (a) celiac angiography, (b) percutaneous transhepatic cystography.



Fig. 3 Gross appearance of surgically resected biliary cystadenocarcinoma.

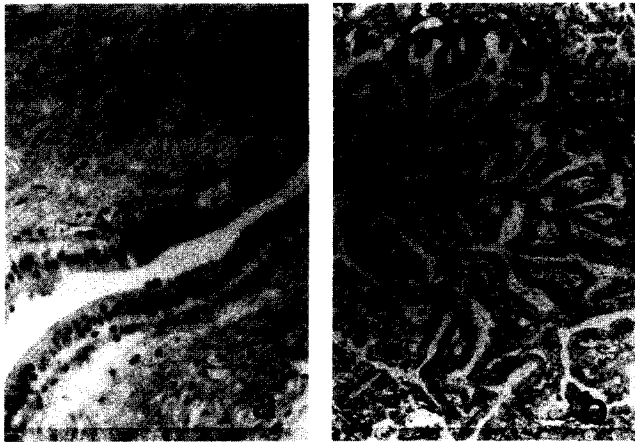


Fig. 4 Photomicrographs of biliary cystadenocarcinoma.

(a) cyst wall. It was lined by a layer of adenoma cells .

(b) papillary projections. They were composed of mucinous papillary adenocarcinoma cells.

non-cirrhotic, non-cancerous liver; it was lined by a layer of benign-appearing, columnar adenoma cells (Fig. 4-a). The papillary nodules protruding into the cyst cavity were composed of mucinous papillary adenocarcinoma cells (Fig. 4-b) with significant infiltration into the connective tissue wall. So this cystadenocarcinoma was thought to develop as a result of malignant trans-

formation of a cystadenoma.

The patient's post-operative course was unremarkable, and she is doing well 4 years after the operation.

Biliary cystadenocarcinoma is a rare tumor. According to Tomimatsu (1), approximately 40 cases have been reported in the English literature, up to 1985. And according to O'Shea (2), 32

cases have been reported, up to 1986. Takayasu (3) also reported an incidence of biliary cystadenocarcinoma in 0.65 % of operated cases and in 0.41 % of autopsy cases of the National Cancer Center Hospital in Tokyo. But, criteria of biliary cystadenocarcinoma differs somewhat among the authors, and it was difficult to study the clinical features of these tumors without unified criteria. Mizumoto (4) presented a new classification of adenocarcinoma with cystic formation in the liver. He advocated the generic term 'cystic adenocarcinomas in the liver' and divided them into three groups; (A) adenocarcinoma arising from congenital cyst, (B) cystadenocarcinoma arising from cystadenoma (malignant transformation of cystadenoma), and (C) cystadenocarcinoma arising *de novo*. Mizumoto argued the term 'biliary cystadenocarcinoma' should be used for only type B and type C tumors. Type A tumor was first reported by Willis (5) in 1943. In the tumor of this type, the cyst wall uninvolved with cancerous lesions is composed of normal epithelium. Type B tumor was first reported by More (6) in 1966. The cyst wall is composed of both adenoma and adenocarcinoma. Ishak (7), Woods (8), and Voigt (9) also reported the tumors of this type. Type C tumor was first reported by Sepulveda (10) in 1954. Reviewing Japanese literature, using Mizumoto's classification, 18 cases of Type A tumor were reported incorrectly as biliary cystadenocarcinoma. There were 17 cases of Type B tumors before 1990, which could be called true 'biliary cystadenocarcinoma'. Our case would be classified as a Type B tumor, too. It indicates the malignant transformation of cystadenomas. According to Ishak (7), who reported 6 cases of biliary cystadenocarcinoma arising from adenomas, this tumor is thought to originate in a bile duct. In our case, the connection between the cyst and the common bile duct was clearly shown by PTC. Interestingly, this is a very rare case; the liver of our patient had a constitutional excretory disturbance of ICG. Nambu and Namihisa (11) reported an incidence

of constitutional ICG excretory defect in 0.007 % of the population in Japan. So complicated case of biliary cystadenocarcinoma and the ICG excretory defect is uncommon. Data base search of MEDLINE (English literature) and JICST (Japanese literature) indicated that this is the first case of biliary cystadenocarcinoma occurring in such an abnormal liver, although we think this has no direct etiologic relation to the tumor.

CT and US are useful modalities for the diagnosis of biliary cystadenocarcinoma or cystadenoma. But the differential diagnosis between these two tumors still depends on histologic examination, because papillary projections of the cyst are commonly observed in benign cystadenoma as well (7).

Previously aspiration cytology was universally accepted; we do not recommend this because peritoneal seeding of cancer cells secondary to such cyst aspiration has been reported (12), and because aspiration cytology is often negative anyway (3). Indeed, aspiration cytology was performed initially in our patient prior to our consultation, though it was negative. Fortunately the cyst was punctured in the area of the liver tissue that was subsequently resected by us, not into the free peritoneal cavity. Aspiration cytology generally should be avoided in these patient due to the risk of peritoneal carcinomatosis.

The prognosis of biliary cystadenocarcinoma generally seems to be much better than that of intrahepatic bile duct carcinoma (3). However, Berjian (13) reported that 67 % of 18 such patients died within 2 years, and only 3 patients were doing well more than 3 years after resection. Our patient does well for 4 years, and is currently followed in our outpatient department.

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