Case Report

A Case Report of Extrahepatic Biliary Cystadenoma Treated by Simple Cystectomy

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We present a relative rare case of extrahepatic biliary cystadenoma communicated with biliary system. A 75-year-old man consulted with low grade fever up and abnormal liver function results were demonstrated by routine blood examination. In abdominal ultrasound and helical triple phase computed tomography scan, a cystic septated lesion, measuring 3 cm in diameter, was found. And endoscopic retrograde cholangiography showed mucinous filling defects in the common bile duct, which was not noted malignant findings by histological examination. Intraoperatively, the cystic lesion was identified to connect with left hepatic biliary duct. Simple cystectomy was indicated to be enough, since a frozen section of the mass stump was negative of mucinous epithelium and malignant area. No evidence of cellular atypia or tissue invasion to suggest malignancy was found by pathological examination. With immunohistochemical study, positive cells were noted for Ki-67 in the epithelium of cystic wall, while they were negative for p53 and CA19-9.

Key Words: extrahepatic biliary cystadenoma, extrahepatic biliary cystadenocarcinoma, immunohistochemical study

Introduction

The extraordinary detection of liver cyst is one of the most common experiences in screening examination for the patients with hepatobiliary pancreatic complications, since it is occurred in approximately 0.1% of the population\(^1\). Of them, extrahepatic biliary cystic tumors have been reported to represent less than 5% of all intrahepatic cysts of bile duct origin\(^2\) and also usually occur in middle-aged women\(^3,4\). Cystic lesion of the hepatobiliary pancreatic field represents a wide spectrum of disease ranging from simple benign cysts, to potentially malignant biliary cysts, to de novo malignant cystic tumors\(^5\). Although the mechanism of carcinogenesis in cystadenocarcinoma remains unknown, it has been reported to arise by malignant transformation of cystadenoma\(^5,6\). Since a curative resection provides the best chance of longterm survival in patients with cystadenocarcinoma\(^7\), the decision for operation on detection should be attempted whenever hepatic functional reserve is adequate to tolerate. However, it might be difficult to make a definitive diagnosis of a cystic mass preoperatively, especially it represents at extrahepatic state. The consideration about the strategic management, therefore, is always necessary to be proposed. Here, our recent experience is reported, since the case with the presence of communication between cystic lesion and biliary tract is also relatively rare.

Case Report

A 75-year-old man presented to his local physician with low grade fever up. Routine investigations demonstrated abnormal liver function test results, and he was referred for a
hepatopancreatobiliary specialist opinion. Clinical examination was entirely normal. Levels of hepatobiliary enzymes were slightly elevated, as follows: glutamic oxaloacetic transamylase (GOT), 135 IU/l; glutamic pyruvic transaminase (GPT), 130 IU/l; and total bilirubin, 1.8 mg/dl (normal, 0.2–1.2 mg/dl). The other biochemical blood examination levels and tumor markers, carcinoembryonic antigen (CEA) and carbohydrate antigen (CA) 19-9, were within the normal range. Abdominal ultrasound (US) and helical triple phase computed tomography (CT) scan revealed the presence of a cystic septated lesion, measuring 3 cm in diameter, at the hepatic hilum on anterior of caudate lobe (Fig. 1A). The mass pressed portal vein (B).

Endoscopic retrograde cholangiography demonstrated the filling defects (white arrow) in the common bile duct and slightly described cystic lesion (curved arrow). Selective abdominal angiography showed no tumor brushing and portal vein pressured by the cystic mass.

In the view under operation, the cystic lesion was identified to connect with left hepatic biliary duct (white arrow). Cholangiography demonstrated mucinous filling object in the common bile duct (Fig. 2), which included no malignant cells histologically, indicating to be benign cystic tumor, cystadenoma. Selective abdominal angiography demonstrated mucinous filling object in the common bile duct (Fig. 2), which included no malignant cells histologically, indicating to be benign cystic tumor, cystadenoma. Selective abdominal angiography demonstrated mucinous filling object in the common bile duct (Fig. 2), which included no malignant cells histologically, indicating to be benign cystic tumor, cystadenoma.
phy showed no tumor brushing and portal vein pressured by the cystic mass.

Intraoperatively, the cystic lesion was identified to connect with left hepatic biliary duct (Fig. 3). Simple cystectomy could be performed without any events. Since a frozen section of the mass stump was negative of mucinous producing epithelium and malignant area, the operation was finished.

The gross macroscopic pathological findings were of a 3.2×2.8 cm multiloculated cystic lesion and the smooth inner wall without associated mass (A). Microscopic examination revealed the cystadenoma finding, which wall was lined by columnar epithelium demonstrating low papillary growth (B). (HE stain ×400) Immunohistochemical study showed positive for Ki-67 (C) and negative for p53 (D). (×400)

Discussion

Although the mechanism of carcinogenesis in cystadenocarcinoma remains still unclear, it has been evaluated to arise due to malignant
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transformation of benign cystadenoma. These cystic lesions usually represent solitary without connection from biliary tract, but only 37 cases have been reported to communicate with bile duct up to date\(^8\). According to this recent report, benign cystadenomas were noted in 21 cases and total tumor excision was performed in most patients (16 cases), which provided a good clinical course. Since uninvolved parenchyma or hilar structures is better to be preserved, the extended operation should be restricted just for cases with invasive findings\(^9\). By contrast, from high recurrence rate of extrahepatic biliary cystadenoma, Davies et al emphasized the importance of extended resection with bilioenteric reconstruction\(^10\). Indeed, since the malignant transformation from cystadenoma\(^4\) or late occurrence of cystadenocarcinoma after partial romove\(^11\) was reported, complete resection should be recommended. In addition, the secondary complications, such as rupture, hemorrhage or obstructive jaundice due to produced mucin, were apprehensive. In the present case, on the step of removal, simple cystectomy was considered to be enough for complete therapy. And finally, it was proved by the judgment based on frozen histological findings, that both malignant lesion and mucinous producing epithelium were not noted on the tumor stump. As a matter of course, to ensure the normal biliary epithelium on the edge of specimen, bile duct resection could be selected even for this case. Consequently, the follow up CT revealed no fluid reaction around removal area and no recurrence sign.

To select the most reasonable strategy, certain diagnosis should be estimated and then, secondary operation or chemotherapy might be necessary. However, in recent review, routine histological examination was reported to be difficult to distinguish cystadenoma from cystadenocarcinoma in 38% hepatobiliary pancreatic disease cases\(^12\). In addition, for 25% cases the diagnosis of cancer was established after the appearance of metachronous metastases. Routine analysis of genetic and proliferation markers, therefore, might be useful to estimate the decisions. The several markers for hepatobiliary pancreatic field have been evaluated, including CA19-9 or K-ras\(^{13,14}\), p53 tumor suppressor gene or apoptotic factors\(^{15,16}\). Among these common factors, Ki-67 is expressed in all phases of the cell cycle except G0, especially seen in G2/M, suggesting to be regarded an excellent marker for cell proliferation\(^17\) and also a prognostic value in cancer\(^18\). The immunohistochemical study showed the present case to represent negative for p53 and CA19-9, and positive for Ki-67 (Fig. 4C, D). To diagnosis the malignant tumor, p53 or CA19-9 was reported to be more critical than Ki-67 expression\(^18\). Therefore, the present cystic tumor might be concluded for benign tumor with cell cycle progression, such as inflammatory changes. Although additional therapy is not considered in the present case, the careful follow-up should be evaluated.

To decide reasonable strategy, another important action should be a definitive diagnosis to distinguish from mucin-producing malignant tumor. These tumors also exhibit mucin hypersecretion and often appear as a cystic lesion on imaging due to bile duct dilatation\(^19\). Since the cystic malignant mass showed hypovascularity, dynamic CT with angiography might not be useful. By contrast, the detections of solid lesion in the cystic wall, calcification and hemorrhagic fluid reveal helpful findings to diagnose as malignant mass\(^20\). In the present case, since the observations to suspect malignant tumor were not found preoperatively, minimum procedure was planed. The removed tumor lacked solid components, and was histologically classified as a benign tumor char-
acterized by epithelial development and papillary growth. Depending intraoperative findings, there is a possibility to decide an extended resection without any perplexity. To ease unnecessary burden on the patients, sufficient considerations for reasonable strategy should be evaluated.

References