CASE REPORTS

A Large Cystic Tumor with Bile Duct Communication Originating Around the Hepatic Hilum

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Abstract

Biliary cystic tumors are rare neoplasms occurring in the liver and less frequently in the extrahepatic biliary system. Recently, biliary cystic tumors in the liver are thought to be divided into a biliary mucinous cystic neoplasm and intraductal papillary neoplasm of the bile duct. We report a case of a large cystic tumor originating around the hepatic hilum which had luminal communication with the bile duct. A 74 year-old-woman underwent abdominal ultrasonography for a routine checkup. It revealed a large cystic tumor in the liver. CT scan and MRI showed a multilocular cystic tumor about 12 cm in diameter with a mural nodule occupying the medial and anterior segment of the liver. Intraoperative cholangiography showed a communication between the cystic tumor and the bile duct. Central bisegmentectomy of the liver and extrahepatic bile duct resection was performed. A papillary tumor existed in the common hepatic duct and was connected with the cystic tumor in the liver. The tumor was mostly composed of noninvasive papillary adenocarcinoma with adenoma components, and was associated with focal microinvasion of adenocarcinoma. Ovarian-like stroma was not observed. This lesion was diagnosed as a cystic variant of intraductal papillary neoplasm of the bile duct. The patient is alive with no recurrence for 18 months since the surgery.

Keywords

Biliary cystic tumor – bile duct communication – intraductal papillary neoplasm of the bile duct (IPN-B)

Introduction

Biliary cystic tumors are rare neoplasms occurring in the liver and less frequently in the extrahepatic biliary system [1, 2]. In practice, the most commonly encountered primary cystic tumors of the liver are biliary cystadenoma and cystadenocarcinoma, which constitute less than 5% of cystic lesions of the liver [2-6].

Recently, biliary cystic tumors, which have been diagnosed as biliary cystadenoma or cystadenocarcinoma, are thought to be divided into a biliary mucinous cystic neoplasm (MCN) and intraductal papillary neoplasm of the bile duct (IPN-B) such as the cystic tumors in the pancreas with regard to tumor stroma and luminal communication between tumors and bile ducts [2, 5-12].

We report a case of large cystic tumor originating around the hepatic hilum. In this case, we could observe the luminal communication between the tumor and the bile duct by direct cholangiography. We diagnosed the lesion pathologically as IPN-B.

Case Report

A 74-year old woman underwent abdominal ultrasonography for a routine checkup without any significant symptoms. It revealed a large multilocular cystic tumor in the liver (Fig. 1A). It had a mural nodule on the wall (Fig. 1B). The CT scan (Fig. 2) and MRI showed a multilocular cystic tumor about 12 cm in diameter occupying the medial and anterior segment of the liver. The tumor protruded into the hepatic hilum and had a mural nodule which was well...
enhanced with contrast medium. The result of the laboratory examination showed no liver dysfunction or jaundice and showed increased serum levels of CEA (6.5 mg/ml) and CA19-9 (74 U/ml).

On operation, the multilocular cystic tumor protruded to the hepatic hilum and the surface of the medial and anterior segment of the liver (Fig. 3A). Intraoperative cholangiography showed a filling defect of the right anterior duct and stenosis of the common hepatic duct by the papillary tumor. The right posterior duct was joined directly to the common hepatic duct, and had no involvement with the tumor. As little contrast medium flowed out from the bile duct to the intra-cystic space, a communication between the cystic tumor and the bile duct was suggested (Fig. 3B).

Before hepatectomy, puncture and aspiration of the content of the cystic tumor was performed for preventing the rupture of the tumor during hepatectomy. About 300 ml yellowish mucinous fluid was aspirated. During hepatectomy, the tumor extending to the common hepatic duct was observed (Fig. 3C). Central bisegmentectomy of the liver and extrahepatic bile duct resection was performed.

A papillary tumor about 2.5 cm in diameter existed in the common hepatic duct and was connected with the cystic tumor in the liver. The right anterior duct was not identified and was suggested to be occluded by the papillary tumor (Fig. 4). Microscopically, both the papillary tumor (Fig. 5A) and the cystic tumor (Fig. 5B) were composed of papillary proliferation of dysplastic tumor cells with mucin hypersecretion. These lesions were mostly

Fig 2. A. CT scan showing a multilocular cystic tumor about 12 cm in diameter occupying the medial and anterior segment of the liver. GB; gallbladder. B. The tumor protrudes into the hepatic hilum and has a mural nodule which is well enhanced with contrast medium (arrow).

Fig 3. A. The multilocular cystic tumor protruded to the hepatic hilum and the surface of the medial and anterior segment of the liver. GB; gallbladder. B. Intraoperative cholangiography showed filling defect of the right anterior duct and stenosis of the common hepatic duct by the papillary tumor. The right posterior duct joined directly to the common hepatic duct, and received no involvement of the tumor. As little contrast medium flowed out from the bile duct to the intra-cystic space, a communication between the cystic tumor and the bile duct was suggested. C. During hepatectomy, the tumor extending to the common hepatic duct was observed. LHD: left hepatic duct, CHD: common hepatic duct, RPD: right posterior duct.
Cystic tumor with bile duct communication originating around the hepatic hilum

Fig 4. A. A papillary tumor about 2.5 cm in diameter (arrow) in the common hepatic duct connected with the cystic tumor (arrowhead) in the liver. The right anterior duct was not identified and suggested to be occluded by the papillary tumor.
B. Schema of this case. The papillary tumor originated around the root of the right anterior duct and the cystic tumor existed continuously in the medial and anterior segment of the liver. C.T.: cystic tumor, P.T.: papillary tumor, LHD: left hepatic duct, CHD: common hepatic duct, RPD: right posterior duct

Fig 5. A. Photomicrograph of the papillary tumor (H-E x100). The tumor composed of papillary proliferation of dysplastic tumor cells with mucin hypersecretion. B. Photomicrograph of the cystic tumor (H-E x100). The tumor was histologically the same lesion as the papillary tumor. These lesions were mostly composed of noninvasive adenocarcinoma.

Concerning this case, the lesion formed a multilocular cystic tumor in the liver with a papillary tumor on the surface of the liver. The lesion was diagnosed as IPN-B on histopathological examination. The postoperative course was uneventful. The patient is alive with no recurrence for 18 months since the surgery.

Discussion

Recently, biliary cystic tumors in the liver are thought to be divided into two groups with regard to tumor stroma (presence or absence of ovarian-like stroma) and luminal communication between tumors and bile ducts [2, 5-12]. Biliary cystic tumors with ovarian-like stroma typically occur in females, have no luminal communication between tumors and bile ducts, and in most cases are adenomas. In contrast, biliary cystic tumors without ovarian-like stroma occur in both genders, have luminal communication between tumors and bile ducts, and more frequently are adenocarcinomas. Such tumors are thought to correlate cystic tumors arising from pancreas. The former are the biliary counterpart of MCNs, and the latter are one of the IPMNs of the pancreas. The latter are now named IPN-Bs.

This tumor entity is thought to include intraductal growth types of intrahepatic cholangiocarcinoma (IG-ICC), biliary papillomatosis and so-called biliary cystadenoma and cystadenocarcinoma with bile duct communication. IPN-B arises in the intrahepatic, and extrahepatic bile ducts, and shows histologically well differentiated adenocarcinoma, borderline lesion, and low grade malignancy. IPN-B shows focal invasion, and some of the advanced cholangiocarcinoma might be derived from IPN-B. Yeh et al [13] classified IPN-B pathologically into four types. Type 1 is lined by biliary epithelium of low-grade dysplasia; type 2, lined by that of high-grade dysplasia; type 3, by in situ and microinvasive adenocarcinoma; and type 4, lined by papillary lesions with stromal invasion of adenocarcinoma. IPN-B type 3 and type 4 correspond to IG-ICC.

Concerning the prognosis of patients with IPN-B, no disease-related deaths have occurred in patients with type 1 and 2. One-, three-, and five-year survival rates were 100%, 33%, and 17% in type 3, and 56%, 32%, and 14% in type 4, respectively. There was no lymph node metastasis in patients with type 1, 2, and 3; whereas there were 7 of 59 patients (12%) with lymph node metastasis at the hepatoduodenal ligament in type 4. Survival rate with lymph node metastasis in patients with type 4 was significantly worse than that without lymph node metastasis. All patients with type 3 and 4 with positive malignant section margin died of the disease within 2 years following the operation. An aggressive surgical attitude toward IPN-B is recommended because of rarer lymph node metastasis, higher resectability rate, and promising long-term survival of IPN-B compared with those of non-IG-ICC. Nanashima et al [14] and Paik et al [15] also reported relatively good prognosis after surgery and recommended accurate preoperative assessment and aggressive surgical resection.

Concerning this case, the lesion formed a multilocular cystic tumor in the liver with a papillary tumor on the surface of the liver. Immunohistochemically, the tumor cells were mostly positive for MUC5AC, partially for MUC2 and MUC6, but negative for MUC1 and CK20. Finally this lesion was diagnosed as IPN-B.

The postoperative course was uneventful. The patient is alive with no recurrence for 18 months since the surgery.
wall. A communication between the cystic tumor and the bile duct was confirmed. Microscopically, both the papillary tumor and the cystic tumor were composed of papillary proliferation of dysplastic tumor cells with mucin hypersecretion. These lesions were mostly composed of noninvasive adenocarcinoma with adenoma components, and were associated with focal microinvasion of adenocarcinoma. Ovarian-like stroma was not observed. These were characteristic findings of a cystic variant of IPN-B. This case was categorized into IPN-B type 3 according to Yeh’s classification [13].

This tumor might be diagnosed as a biliary cystadenocarcinoma according to the traditional classification. Biliary cystic tumors in the liver need to be divided to “intraductal papillary neoplasm of the bile duct” or “biliary mucinous cystic neoplasm”, as the cystic tumors in the pancreas. Recently, several reports [2, 7-17] show the clinical, pathological and radiological features of biliary cystic tumors. Surgical results and prognosis of them are being gradually revealed [13-15]. Further analyses of cases would be useful for establishing diagnostic criteria and optimal treatment strategy such as for cystic tumors in the pancreas [18].

References