A Case of Intrahepatic Cholangiocarcinoma with Atypical Multinodular Image Findings

<Case Report>

Noriko Nishikawa[1)], Tadashi Katsuramaki[1)], Toru Mizuguchi[1)], Sachiko Kimura[2)], Aya Sasaki[2)], Takayuki Nobuoka[1)], Akihiro Nui[1)], Yasutoshi Kimura[1)], Tomohisa Furuhata[1)], Koji Yamaguchi[1)], and Koichi Hirata[1)]

[1)]First Department of Surgery, [2)]Department of Clinical Pathology, Sappro Medical University School of Medicine South1, West 16, Chuo-Ku, Sapporo, Hokkaido 060-8543, Japan

ABSTRACT

We report the imaging findings of an atypical case of cholangiocarcinoma. The patient was a 59-year-old man , in whom a multicystic liver mass was found on the left lobe of the liver by US. However, the images obtained from contrast CT showed small nodules had gathered and a low density mass in the early phase and peripheral enhancement in the late phase. It seemed to be liver echinococcosis because a group of small nodules appeared on the CT image. An extended left lobectomy of the liver was carried out. The pathological finding was cholangiocarcinoma.

Key words : Cholangiocarcinoma, Image findings, Hepatectomy.

INTRODUCTION

The incidence of intrahepatic cholangiocarcinomas, malignant tumors that offer a poor prognosis, is rising worldwide[1)]. The development of imaging techniques such as ultrasound sonography (US), computed tomography (CT), and magnetic resonance imaging (MRI) may contribute to an increase in the detection rate of the tumors, however, a satisfactory consensus on making a clinical diagnosis has not yet been achieved[2)]. Intrahepatic cholangiocarcinoma grows along the bile ducts and is elongated, speculated, or branchlike. Bile ducts are dilated because of obstruction by the tumor, expansion-sloughed tumor debris, or an excessive amount of mucin. On US, CT, or MR cholangiography, the bile ducts of the involved hepatic segment or hepatic lobe appear dilated. An intraductal mass can appear as an echogenic mass on US and as an enhancing soft-tissue mass on CT, and most intrahepatic cholangiocarcinomas are therefore well depicted. If such a mass does not appear, however, it is difficult to distinguish the condition from metastasis and liver flukes. We report the imaging findings of an atypical case of cholangiocarcinoma.

Correspondence should be addressed to:
Koichi Hirata, M.D., Ph.D.
First Department of Surgery, Sapporo Medical University School of Medicine
South 1, West 16, Chuo-ku, Sapporo, Hokkaido, 060-8543, Japan
E-mail:jtakagi@sapmed.ac.jp
Tel.: +81-11-611-2111 (ext. 3281) Fax: +81-11-613-1678

A Case Report

A 59-year-old man undergoing an annual medical examination was found by US to have a multi-cystic liver mass on the left lobe of the liver. The patient was referred to the university hospital for an elective surgery. The serum laboratory tests (Table 1) showed slight elevations of alkaline phosphatase and gamma glutamyl transpeptidase. In addition, tumor markers, including CEA and CA19-9 levels, were elevated. On the other hand, the serum antigen of echinococcosis was negative by the Western blot method. The images obtained from contrast CT showed that small nodules had gathered. Dynamic CT images showed a low-density mass in the early phase and peripheral enhancement in the late phase (Fig. 1). This finding is atypical for intrahepatic cholangiocarcinoma and had caused us to suspect liver echinococcosis. Magnetic resonance images showed a highly intensified mass in T2 images, which are inconsistent with echinococcosis (Fig. 2). The final clinical diagnosis was cholangiocarcinoma (CCC) based on the composite clinical criteria.

Table 1 : Serum biochemical analysis

TP (6.5-8.0 g/dl)	6.3 g/dl
ALB (3.7-5.2 g/dl)	3.9 g/dl
AST (11-39 IU/L)	33 IU/L
ALT (5-40 IU/L)	19 IU/L
LDH (119-229 IU/L)	440 IU/L
T-bil (0.2-1.2 mg/dl)	0.7 mg/dl
D-bil (0.0-0.3 mg/dl)	0.2 mg/dl
γ-GT (9-70 IU/L)	239 IU/L
ALP (110-370 IU/L)	393 IU/L
CEA (0-5.9 ng/ml)	53.0 ng/ml
CA19-9 (0-37 ng/ml)	2529.0 ng/ml

TP: total protein, ALB: albumin, AST: asparste aminotransferase, ALT:alanine aminotransferase, LDH: lactate dehydrogenase, T-bil: total bilirubin, D-bil: direct bilirubin, γ-GT: γ-gultamile transpeptitase, ALP: alkaline Phosphatase, CEA: carcinoembryonic antigen, CA19-9: carbohydrate antigen 19-9.

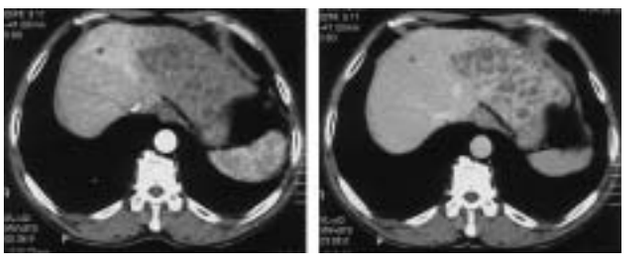

Fig. 1 Abdominal computed tomography with intravenous contrast performed upon admission to the university hospital. The tumor in the left liver lobe was low density without enhancement in the early phase (A). Subsequently, the tumor showed gradual enhancement in the late phase (B).

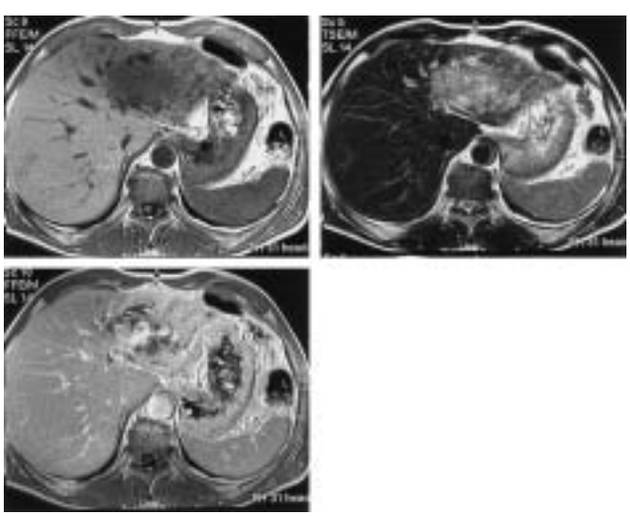

Fig. 2 Magnetic resonance image taken upon admission to the university hospital. The tumor appeared as a low-intensity mass on the T1-weighted image (A) and as a high-intensity mass on the T2-weighted image (B). The tumor was enhanced by contrast medium (C).

An extended left lobectomy of the liver was carried out uneventfully, and the patient was discharged from the hospital at postoperative 14 days. On a gross specimen, the tumor was firm and whitish-gray because of its large amount of fibrous stroma. The margin of the tumor was lobulated and multinodal. There was a multi-in-

trahepatic metastatic tumor around the main tumor. The pathological finding was adenocarcinoma of the CCC, which was consistent with the clinical diagnosis. The main tumor was considered to be on a bile duct in the left lobe, and small nodules had metastasized to the whole left lobe of the liver (Fig. 3). Furthermore, the middle of the tumor region had a cystic formation that was responsible for the images resembling liver echinococcosis.

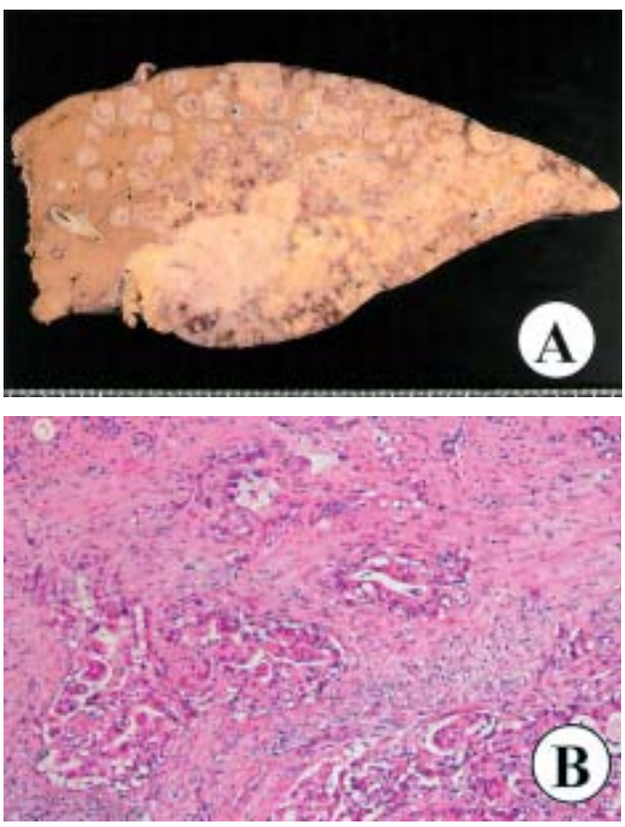

Fig. 3 Photograph of a gross pathologic specimen shows a whitish mass with extensive desmoplastic change. The main CCC tumor located at the hilar and the peripheral of the left liver lobules contained metastatic tumor cells (A). Hematoxylin–eosin staining of the tumor showed moderately differentiated adenocarcinomas. The character of the tumor represented tubular patterns with cystic formation (B).

DISCUSSION

The incidence of cholangiocarcinoma (CCC) has increased along with advances in diagnostic technology and awareness of this disease in addition to the biological development itself[1]. Although the etiology of CCC is not understood completely, chronic inflammation is considered to be a high risk factor. Therefore, chronic cholangitis, such as primary sclerosing cholangitis (PSC), Caroli's disease, and liver flukes, increases the risk of cancer development[2]. Molecular alterations of CCC have been reported in a number of signals associated with proliferation and apoptosis[3,4]. However, none of the molecular abnormalities has been specific for CCC. Furthermore, the mechanism by which one of the most malignant features in CCC is extension and infiltration along the periductal sheath, as in this report, is unknown. Therefore, it is very important to depict the clinical features of CCC to understand its biological features and develop a curative strategy.

An interesting feature in this case is how cancer cells took the place of hepatocytes without causing symptoms. There was a scarce blood supply for left liver parenchyma due to the obstruction of both the left hepatic artery and the protal vein. The parenchyma of the liver left lobe was nearly replaced by CCC. CCC may be more tolerated in the ischemic condition than hepatocytes. In this case, there was no symptom associated with bile obstruction because the hepatocytes no longer existed and no bile juice could be produced.

In Asian countries, the differential diagnosis of CCC and epidemic liver flukes may be difficult. Especially in Japan, CT images of echinococcosis can appear to be similar[5] to those obtained in cases of CCC, as shown in this report. Magnetic resonance images may give additional information to achieve a more precise diagnosis[2]. In this case, there seemed to be liver echinococcosis because the gathering of small nodules appeared on the CT image. CT imaging does not always provide an accurate diagnosis. Especially in the case of CCC, the tumor is not

enhanced by the contrast medium, which makes a differential diagnosis difficult. On the other hand, MRI can provide more detailed characteristics of the tumor. Therefore, MRI should be considered if the clinical diagnosis is CCC or liver echinococcosis. No specific serum tumor marker for CCC exists in laboratory tests. CA19-9, CEA, and CA125 levels may be increased by 85%, 30%, and 50%, respectively[2]. In any case, diagnosis of CCC should be achieved based on composite clinical features, including radiological images and serum laboratory tests.

Surgery is the exclusive strategy for the treatment of CCC. Surgical resection with a tumor-free margin has obtained five-year survival rates of 20% to 40%[2]. Since metastasis in the liver had already existed in this case, the possibility of recurrence soon after surgery is very high even with complete tumor resection. Therefore, adjuvant chemotherapy is necessary. However, the prognosis for patients with CCC is very bad[6] even with chemotherapy. A good survival rate can be obtained only if treatment is begun in the early stage of CCC. Therefore, the early detection of the cancer is very important. In advanced cases, a systematic study is required to identify patients who would benefit from treatment for CCC.

In summary, the differential diagnosis of CCC from liver flukes should be made through a composite diagnosis, including radiological studies and serum chemolaboratory tests. A prospective study should be designed to identify patients who would benefit from surgical resection.

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