[症例報告]Multiple Cystadenomas of the Liver : Report of A Case

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Multiple Cystadenomas of the Liver : Report of A Case

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ABSTRACT

A case of multiple cystadenomas of the liver in a 62-year-old woman is described. The patient had an 8-year history of multiple liver cysts. On admission, CT revealed a papillary projection into the cyst of the posteroinferior segment (S_7) of the cysts and its cyst fluid tumor marker levels was abnormally elevated.

At surgery, multiple unilocular and multilocular cysts with a number of nodules were found with extra-hepatic growth and our tentative diagnosis was macroscopicaly advanced adenocarcinoma. Yet, curative resection was thought not to be possible. Cystectomy of the lateral lobe which protruded into the stomach and biopsy of cysts with nodule in anteroinferior segment (S_5) were then done, and ethanol or Mitomycin C was injected into the remaining cysts. The diagnosis of congenital cysts with cystadenomas was confirmed histologically. There should be a careful followed-up of the patient because the congenital cyst and biliary cystadenoma of the liver sometimes develop combined cystadenocarcinoma.

INTRODUCTION

Despite recent advances in diagnostic imaging, it is very difficult to differentiate cystadenoma from cystadenocarcinoma because of morphological similarity. We present a case of multiple cystadenomas of the liver with suspected biliary cystadenocarcinoma showing a papillary projection in a part of the cysts and with a discussoin of differential diagnosis of cystadenoma from cystadenocarcinoma.

CASE REPORT

A 62-year-old female with cysts of the liver and kidneys has been seen in our clinic since

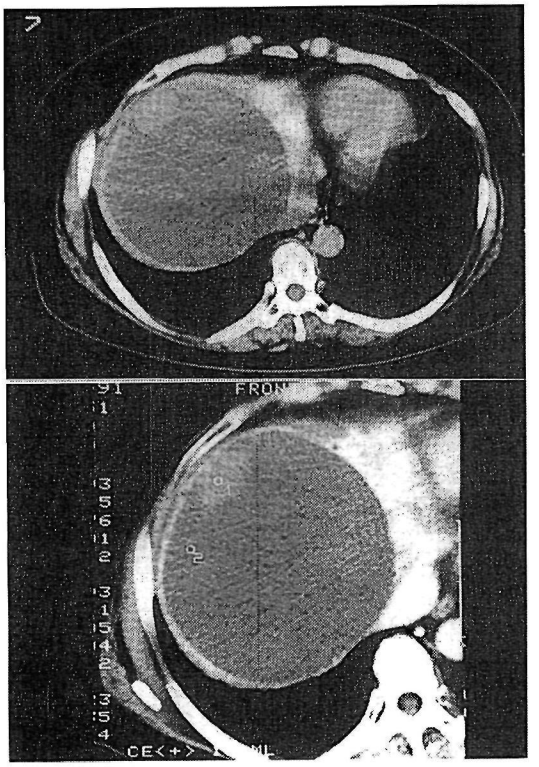

Fig. 1 Abdominal CT scans showing an unilocular cystic lesion with a papillary projection in the segment 7 (top) and it was not enhanced (bottom).

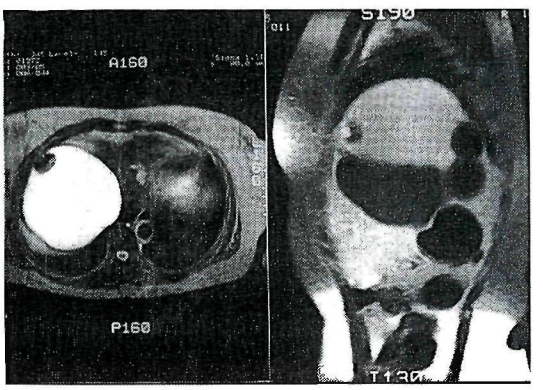

Fig. 2 MRI demonstrating a papillary projection as a low intensity nodule at both T1 and T2 intensity image.

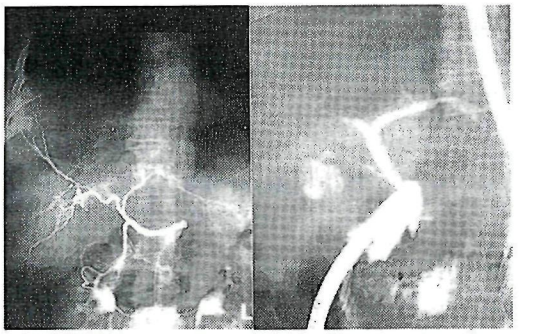

Fig. 3 Hepatic angiogram (left) and ERC (right) showing a huge mass lesion with stretching and attenuation of the hepatic arterial and biliary branches.

1984. In 1989, from the result of gastrointestinal screening, she was suspected of having gastric submucosal tumor and it was also demonstrated by computed tomography (CT) and ultrasonography (US) that she had enlarged liver cysts. The submucosal tumor was affected by liver cysts protruding into the stomach. She was admitted to the 1st Department of Internal Medicine in the Ryukyu University Hospital for further examination of the liver cysts. The CT and US revealed a papillary projection in one of the cysts. Cystography with percutaneous puncture and angiography suspected biliary cystadenocarcinomas and she was referred to the 1st Department of Surgery.

The physical examination revealed no abnormal abdominal findings. The hematological examination and blood chemistry tests including carcinoembryonic antigen (CEA), CA19-9, and alpha-fetoprotein (AFP) were almost within normal limits. Preparatory capacity test of the liver showed hepaplastin test (HPT)80%, ICG-Rmax 4.630mg/kg/m, R-15 6.5% and KICG 0.182. The CT and US revealed multiple liver cysts in both lobes and one cyst in the posteroinferior segment (S_7) occupied a large area, its diameter being about 10 cm. The cyst had a papillay projection of the same density as the

liver (Fig.1). The magnetic resonance image (MRI) showed a low intense papillary projection of the cyst wall at both T1 and T2 intensity (Fig.2).

Cystadenoma or cystadenocarcinoma was suspected from CT, US and MRI findings. The hepatic angiogram revealed a huge avascural lesion with stretching and attenuation of hepatic branches. Endoscopic retrograde cholangiography (ERC) showed also stretching and attenuation of the bile duct. Bile ducts were not communicated with the cystic lesions (Fig.3)

Percutaneous puncture and aspiration cytology of the S_7 cyst was class 1, but there was a high level of tumor marker cyst fluid (CA19-9: 13937.0 U/ml, CEA : 28.7ng / ml, CA125 : 11000U/ml). Based on these findings, we suspected a malignant transformation of the cystadenoma that subseaquently led us to perform surgery (Fig. 3).

Operative findings

Liver cysts were mulitlocular and were growing extrahepatically in all lobes. There were many nodules and we suspected malignancy in most of them, so we decided against curative resection(Fig.4). Only the cysts of the left lateral segment which protruded into the stomach, and small cysts with an anteroinferior segment (S_5) nodule were resected for pathological examination. Transhepatic ethanol injection therapy was administered to the remaining cysts and 2mg of Mitomycin C (MMC) was injected into S_7 cyst to prevent intraabdominal dissemination. The cyst fluid was serous and yellowish except that of S_7, whitch was brown in color, probably due to bleeding by puncture. The amount of aspirated fluid for each ranged from 40ml to 550ml. The tumor marker (CA19-9) of S_7 cyst fluid was 10,055 U/ml and of the anteriosuperior segment (S_8) cyst fluid was 706,210 U/ml. The cytological assessment of both was Class 1.

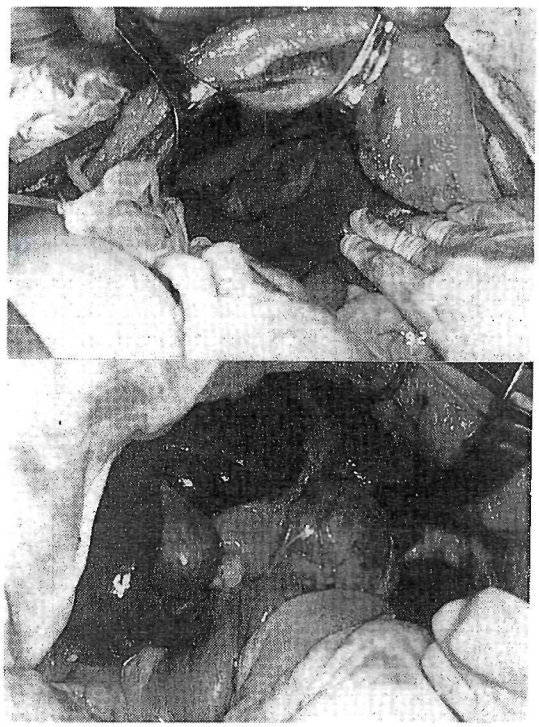

Fig. 4 Macrophotogrphs at surgery showing multiple cystic lesions with nodules.

Pathological findings

Microscopically, the cysts were multilocular and their internal surfaces were covered with columnal epithelium. Some cysts had a papillary projection (Fig.5,6). So the pathological diagnosis was polycystic liver (adult type) with multiple cystadenomas.

Clinical course

Pathological findings did not reveal malignancy but we still thought that the S_7 cyst had undergone malignant change, so we performed arterial infusion chemotherapy. We inserted a permanent catheter in the hepatic artery through the right femoral artery by an intraaortic injection catheter (Infuse-A-port), and 2 mg of MMC per a week was infused. A total of 10 doses of chemotherapy were performed at the out-patient clinic. During the chemotherapy, the patient was in good condition and there was no

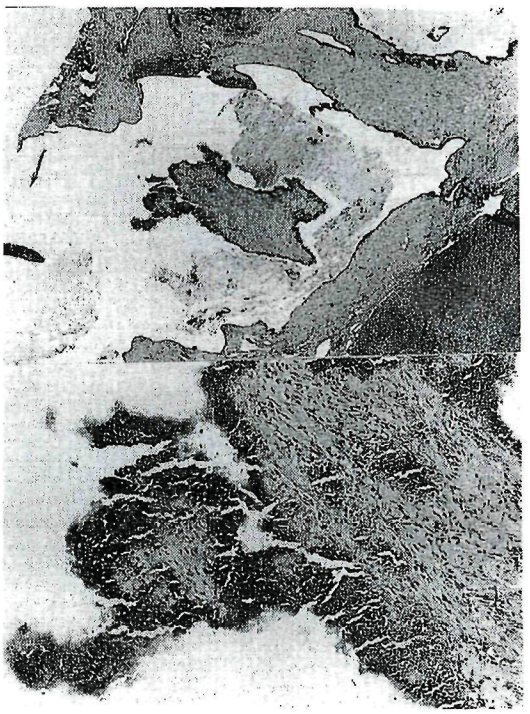

Fig. 5 Microphotographs of the cyst showing the cyst and a papillary projection (top : HE, ×2.5). The papillary projection consist of a dense stroma with stratified columnar lining epithelium(bottom:HE,×20).

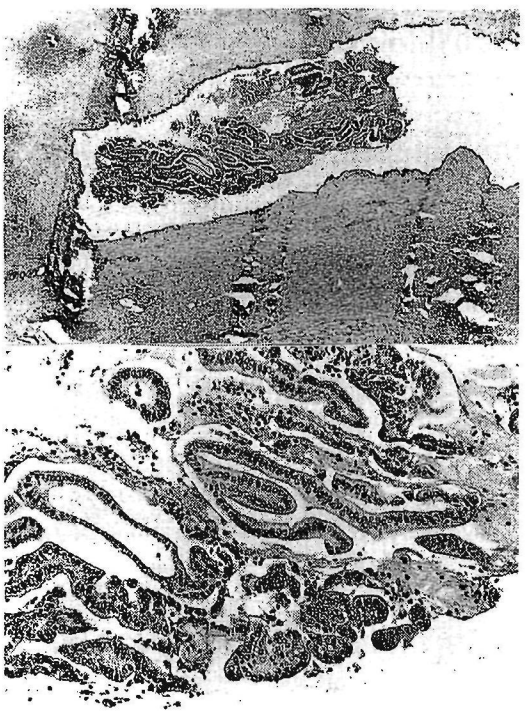

Fig. 6 Microphotographs of the other cyst showing the cyst with papillary adenoma (top :HE, × 2.5). (bottom:HE, ×40).

elevation of tumor markers. The papillary projection found on the last admission disappeared with CT and US (Fig.7).

Eleven months after the operation on Nov. 13th, 1992, enlargement of the residual cyst was found. The patient was readmitted and percutaneous transhepatic cyst drainage was performed in S_7 and S_8 lesions. The S_7 fluid showed a serous yellowish color. The chemical analysis were T-Bil 0.2mg/dl, CEA 9.9ng/ml, CA19-9 17.6U/ml. The S_8 fluid was muddy yellowish-brown in color with T-Bil 0.9mg/dl. The cytological diagnosis of both was class 1. The S_8 drainage tube was removed after the 10th sclerotherapy of 200 mg minocyclin hydrochloride. A biliary endoscopic biopsy of the S_7 cyst wall revealed the cyst to be connected with the biliary tract. Injection of minocyclin hydrochloride was therefore suspended. Both drainage tube were removed after the cyst fluid decresaed. The patient has been examined with no evidence of cyst growth at the out-patient clinic during follow up at the time of writing.

DISCUSSION

Recently with widely available diagnostic imaging(e.g.US,CT,MRI),it has become easier to detect cystic lesions. Cystic diseases of the liver are classified as non-tumor type and tumor type[1]. The former includes simple, multiple, parasitic and/or Caroli disease, and the latter includes cystadenoma and cystadenocarcinoma, and is rare. Only 28cases of cystadenoma have been reported in the Japanese medical literature[2-11], including congress summaries. Accord-

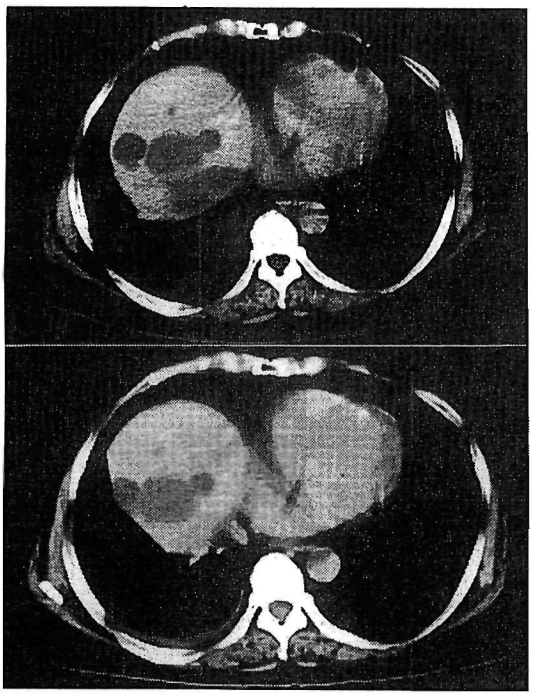

Fig. 7 Abdominal CT scans 3 months after surgery (top) and after the cyst drainage (bottom) showing decreasing size of the cyst and disappearance of the papillary projection.

-ing to Ishark[12], more than 80% of cystadenomas occur in women, and are seen in patients over 30 years of age. Most of the cases are single and in 60% of the cases, patients complained of abdominal symptoms. In about 90% of cases, an abdominal mass is palpable. In many cases,the mass is large, frequently exceeding 10 cm in diameter, and the description of several reports[4,9], are identical. It is characteristic of CT and US findings that more than 80% of cystadenomas are multilocular. Cases having thin internal septa without papillary proje-ction[9],and with papillary projection or mural nodules[11,13,14] are reported as cystadenoma. In cystadenoma, hepatic angiography reveals str-etching and attenuation of the intrahepatic branches, but do not show the tumor staining[3]. In our case, age, sex, clinical symptom, and angiographic findings led us to suspect cystadenoma. Others[5,6,15,16] report that congenital non-tumorous cyst produces serous fluid, but that cystadenoma and cystadenocarinoma produces mucinous fluid. Furthermore, tumor marker are elevated in both the tumor type cystic fluids.

Microscopic features of cystadenoma are reported as follows[17] : 1) lined by a singl layer of mucin producing tall columnal cells resting on a basement membrane, 2) a surrounding area of densely cellular stroma, 3) thick ring of collagen. When comparing the pathological feature of congenital cyst, cystadenomas, and cystadenocarcinomas [18,19], a differential diagnosis seems to be easy, because the cystic fluids, microscopic features of the capsule, stroma and epithelium were different from each other. Especially the cystadenoma may malignantly transform into cystadenocarcinoma in the papillary projection of the cyst[20]. So diagnosis must be carefully made.

On the other hand, some reported cases[21] of cystadenocarcinomas contained patholgically the carcinomatous congenital cyst and cystadenomas. Kawarada *et al* [21] analyzed 34 cases in Japan that had been reported as cystadenocarcinoma. They reported that 25 cases had malignant change in simple liver cysts, and 4 cases had malignant change in cystadenomas. They also pointed out that malignant change in a simple liver cyst was not so rare as reported earlier[21]. And Kasai *et al* [22] reported that 5.2% of liver cysts may transform into malignancy.

In our case, we have been following the patient for 8 years, and papillary projection in a liver cyst has been revealed. Only one other case similar to ours has been previously reported by Ohmagari *et al* [18]. The liver lesions in our case was pathologically diagnosed as multiple liver cyst with cystaednomas and was difficult to differentiate from cystadenocarcinoma with diagnostic imaging. We could not resect the cyst with papillary projection detected

by CT, but surgical specimens which was suspected of having undergone malignant change revealed no malignancy.

This patient must be carefully followed as an out-patient because of the possibility of malignant transformation[2] in the cyst.

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