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A Case of Biliary Cystadenocarcinoma

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Biliary cystadenocarcinoma is a very rare cystic tumor that arises in the liver or, less frequently, in the extrahepatic biliary system. It has been shown to arise in congenital liver cysts, bile ducts, biliary cystadenoma, in the context of fibropolycystic disease and in the hepatoduodenal ligament. Common presenting symptoms include an abdominal mass, local pain, nausea, jaundice, fever or occasional ascites. Some patients are asymptomatic, the lesion being an incidental finding at autopsy or surgery. Approximately 50 cases have been reported in the literature.

We report a case of biliary cystadenocarcinoma in a 63-year-old man with a review of the literature.

Key Words: *Biliary cystadenocarcinoma*

INTRODUCTION

Biliary cystadenocarcinoma is a very rare cystic tumor and constitutes less than 5% of intrahepatic cysts of biliary origin[1]. Along with the recent advances in diagnostic imaging techniques, approximately 50 cases have been reported in the literature[1-13].

We report a case of biliary cystadenocarcinoma in a 63-year-old man with a review of the literature.

CASE REPORT

A 63-year-old man was readmitted to our hospital for further evaulation of hepatic cystic lesion. One year prior to admission, he had visited a local hospital due to pain in the right upper quadrant of the abdomen and fever. Abdominal sonographic finding showed about a 10cm-sized cystic nature space occupying lesion in the right lobe of the liver(Fig. 1) and pus-like material was aspirated. He was referred to our hospital for diagnosis and treatment. On admission, his temperature was 38.0℃, pulse 80 per minute, respiration 20 per minute. The blood pressure was 110/70mmHg. The laboratory tests were as follows: leukocyte, 12,900/mm^3; hemoglobin, 12.9g/dl; total bilirubin, 4.6mg/dl; AST, 39IU/L; ALT, 74IU/L; Alkaline phosphatase, 590IU/L; GGT, 90IU/L. HBsAg and anti-HCV were all negative. Antiamebic antibody (IHA) was positive as 1:512 titer. Urinalysis and stool examination revealed within normal limits. The patient refused further study and was treated a as liver abscess. After discharge, he did not follow up with any specific problem for about 1 year. He visited to know the progress of the previous hepatic leision. He had anorexia and weight loss of over 1kg during 1 month. Upon readmission, his temperature was 37.0℃, pulse 80 per minute, respiration 20 per minute. The blood pressure was 120/80mmHg. There was no pathologic lesion in his eyes, ears, nasal or oral mucosa. On auscultation of chest, breathing sounds were normal, and the heart sound was regular without murmur. Three finger breadth palpable liver was felt on examination of the abdomen.

The results of a routine examination showed a hemoglobin of 13.3g/dl, hematocrit of 39%, a

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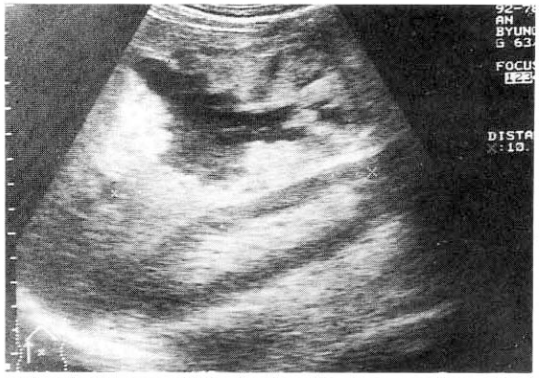

Fig. 1. Abdominal ultrasonography shows papillary of various shapes within the cystic lesion of the liver.

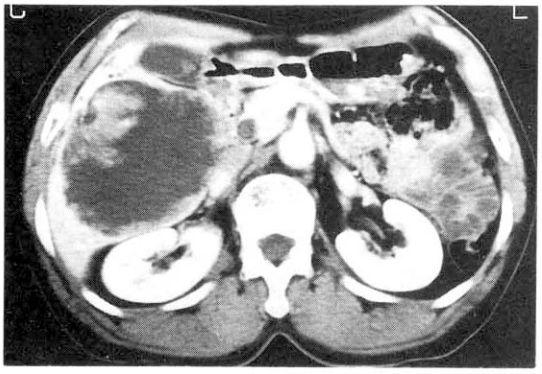

Fig. 2. Abdominal CT demonstrates a large cystic lesion within abundant papillary projections in the liver.

leukocyte count of 11,600/mm^3(neutrophil 67%, lymphocyte 27%, monocyte 6%) and a platelet count of 273,000/mm^3. Liver function test showed as follows: total bilirubin, 0.9mg/dl; direct bilirubin, 0.6mg/dl; AST, 29IU/L; ALT, 13IU/L; alkaline phosphatase, 70IU/L; GGT, 38IU/L. Antiamebic antibody(IHA) was negative as below 1:64 titer. Urinalysis and stool examination revealed within normal limits. All tumor markers were within normal limits; AFP, 3.1ng/ml; CEA, 0.4ng/ml; CA 19-9, 8.96U/ml and CA 125, 40.1U/ml.

Chest and abdominal X-ray revealed no abnormality. Abdominal CT demonstrated huge cystic lesions measuring 10×10cm in right liver and having focal irregular wall thickening with

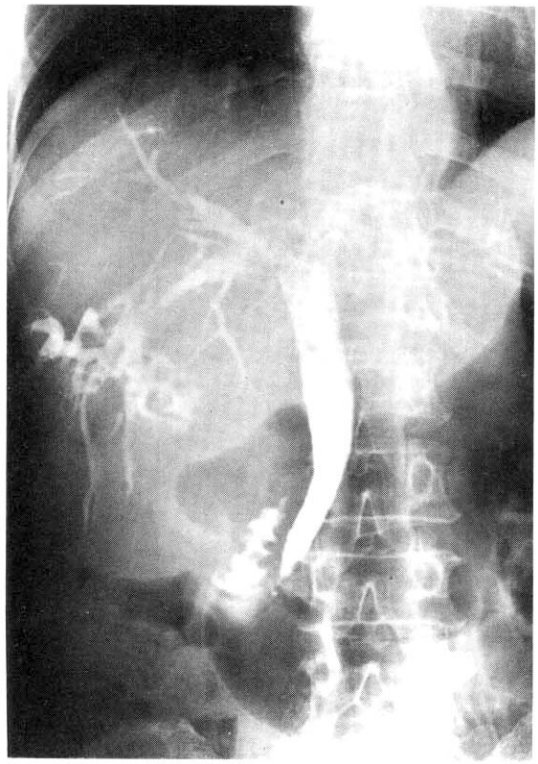

Fig. 3. ERCP reveals mildly dilated common bile duct and some filling defect in right intrahepatic duct. The cystic lesion is connected with right intrahepatic duct and some filling defect is seen in cystic lesion.

internal papillary projections(Fig. 2). Endoscopic retrograde cholangiography(ERCP) revealed mild dilated common bile duct and some filling defect in right intrahepatic duct. The cystic lesion was connected with right intrahepatic duct and some filling defect was seen in the cystic lesion(Fig. 3). Mucin-like material was drained by balloon catheter after sphincterotomy. Celiac angiography showed a large cavitary lesion which is supplied by hepatic artery(Fig. 4). Surgery was carried out for definitive diagnosis. On laparotomy, a huge hepatic mass was occupying in segment V and the cystic mass contained a portion of the right hepatic duct. The aspirated material from the cyst was found to be the same as the mucinous material drained endoscopically. He underwent right hepatic segmentectomy(segment V) with cholecystectomy.

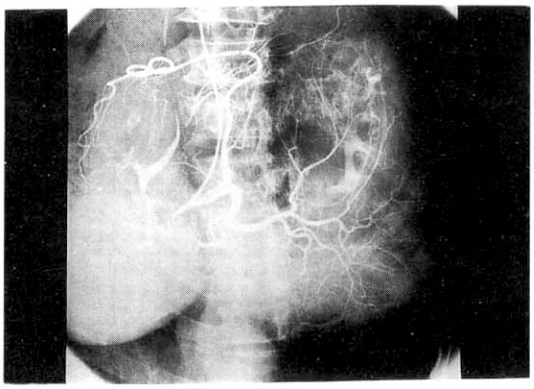

Fig. 4. Celiac angiography shows a large cavitary lesion which is supplied by hepatic.

Fig. 5. Postoperative gross specimen shows gray brown colored, 10.0×8.0×5.0cm-sized, well demarcated cystic lesion.

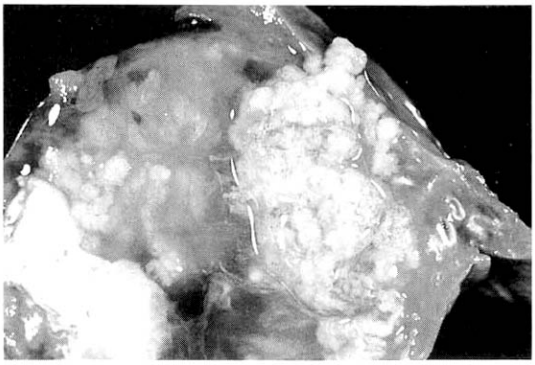

Fig. 6. The cut resected specimen dicsloses unilocular cystic lesion with multiple papillary proliferations projectiog from the wall and massive transparent mucinous fluid in it.

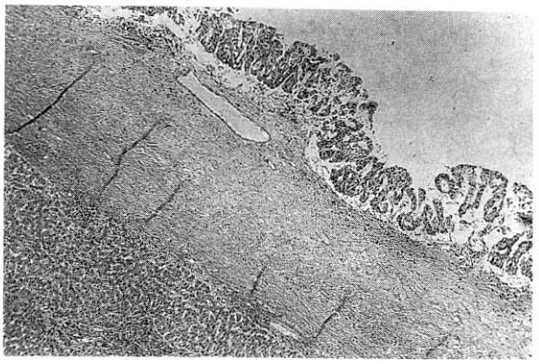

Fig. 8. Microscopic finding shows papillary proliferation of atypical columnar cell with mucin production in the cystic area (H & E, ×100).

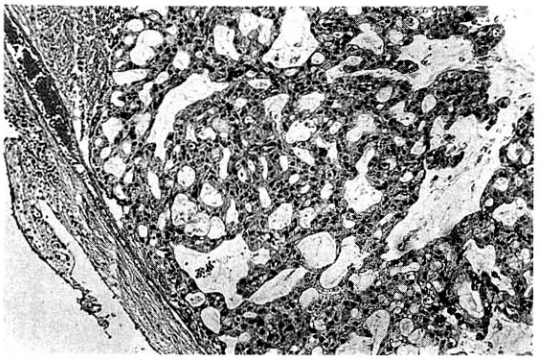

Fig. 7. The tumor is histologically composed of cuboidal tumor cells. The cyst wall is composed of thin fibrous tissue (H & E, ×40).

The surgical specimen contained a grayish-brown colored, well encapsulated unilocular cyst measuring 10.0×8.0×5.0cm(Fig. 5). On section, the internal cavity was filled with multiple papillary tumors and abundant mucinous fluid(Fig. 6). Histologic examination showed papillary proliferation of atypical cuboidal epithelium with mucin production in the cystic area. The tumor was determined as biliary cystadenocarcinoma(Fig. 7. 8).

The patient is doing well 2 year and 2 months after the operation and without a sign of recurrence.

DISCUSSION

Biliary cystadenocarcinoma is a very rare cystic tumor[1] and it has been reported to arise from congenital liver cysts[2-9], bile ducts[5,6], biliary cystadenoma[1-12], fibropolycystic disease[11] and hepatoduodenal ligament[13]. Approximately 50 cases have been reported in the literature[1-13]. Sexual distribution is nearly equal but it is slightly more frequent in women[14]. The mean age at presentation is the late fifties. The tumor size ranges from 3cm to 26cm in the largest diameter and, even though some patients are asymptomatic, most of them are large in size accounting for the symptoms and sign, such as abdominal distention, nausea, palpable abdominal masses and abdominal local pain. Some patients show jaundice, fever, hemobilia and, occasionally, ascites.

However, with recent progress in imaging diagnosis, cystadenocarcinoma has been diagnosed with relative ease and the rapid enlargement of the tumor may be a possible sign of malignancy, but the clinical diagnosis is difficult during the preopeative course. The typical ultrasonographic appearance is that of a globular or ovoid thick-walled cystic mass. Fluid or low level echoes may be seen within the cyst, a sonographic appearance similar to that of pancreatic or ovarian cystadenomas or cystadenocarcinomas[15]. CT may demonstrate a multilobulated cystic mass that contains septations, papillary projections and, sometimes mural nodules[16-18]. Our case was an unilocular cyst with internal papillary projections. ERCP and percutaneous transhepatic cholangiography demonstrate whether there are an intraluminal obstructing lesion and direct communication between the cyst lumen and the biliary tree, or not. In our case, ERCP showed that the cystic lesion was connected with the right hepatic duct and it suggests that the cyst has arisen in the bile duct. Angiography demonstrates a hypovascular mass in most instances or, sometimes, a small vascular blush along the periphery of the tumor as in our case[19].

Grossly, the tumor is usually large-size and consists of a solitary, well encapsulated, fluid-containin multilocular cyst. The cyst often contains hemorrhagic, bilous, mucinous or clear fluid. Wee et al[12] reported a case of biliary adenocarcinoma which was diagnosed preoperatively based on the fine needle aspiration cytologic findings and the imaging appearances. But, the results are usually negative and the chance of peritoneal seeding of malignant cells is present with such a procedure.

Histopathologically, the epithelium lining the cystic spaces is mostly nonciliated columnar to cuboidal mucin secreting epithelium and portions of transition from cystadenoma to cystadenocarcinoma tumor cells may be seen. Most epithelial tumor cells are positive on immunohistochemical staining with antibodies to cytokeratin, epithelial membrane antigen and carcinoembrionic antigen[14]. Biliary cystadenocarcinoma must be differentiated from benign biliary cystadenoma, cholangiocarcinoma and metastatic adenocarcinoma. It needs complete surgical resection for definitive diagnosis and treatment.

The prognosis is comparatively good with complete resection and generally seems to be much better than that for intrahepatic bile duct carcinoma[1]. Recently, Devaney et al[14] recognized that there was a close relation between morphological findings and prognosis. Two types of cystadenocarcinoma exist. One is seen in female patients and arise from a preexisting biliary cystadenoma, usually accompanied by ovarian stroma, and follows an indolent course. The other is found in the male, not associated with a preexisting biliary cystadenoma with ovarian stroma, is more aggressive and leads to death in over half the patients[14].

In conclusion, we report here a rare case of biliary cystadenocarcinoma in a 63-year-old man who was treated for liver abscess, although our case was unilocular, which is not typical.

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