Intrahepatic Biliary Cystadenoma Masked by Adjacent Multiple Simple Hepatic Cysts: a case report

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Intrahepatic biliary cystadenoma is a rare disease with only a few reports in the literature. On the other hand, simple hepatic cysts are frequently encountered in daily practice. Unless finding internal septa or mural nodules, we regard the well-defined intrahepatic cystic lesions with homogenous content as simple hepatic cysts. Here we report a 70-year-old woman with multiple, simple hepatic cysts, one of which was biliary cystadenoma. Ultrasound (US) failed to detect the simple cyst mimicker amongst the hepatic simple cysts for one year. Finally abdominal multidetector computed tomography (MDCT) detected the subtle thin internal septa and focal irregular wall, a key feature for the diagnosis of biliary cystadenoma.

Key words: Biliary cystadenoma; Hepatic cyst

Intrahepatic biliary cystadenoma is a rare disease with only a few reports in the literature [1-3]. The characteristic findings of intrahepatic biliary cystadenomas are hypoattenuated, well-defined cystic lesion associated with thick, fibrous capsule, internal septa, mural nodules, and rarely capsular calcification, keys features for the differentiation of simple cysts [1-5].

Similar to popular playing of “finding the differences” in similar pictures, it may be difficult to identify a hepatic cystic tumor within background of simple hepatic cysts. We report such an interesting case of biliary cystadenoma that mimicked a simple hepatic cyst, masked by adjacent multiple simple hepatic cysts. The correct diagnosis was not made until after a year, when MDCT was employed for diagnosis. The imaging findings on US and MDCT, and differential diagnosis of the hepatic cystic lesions were discussed.

CASE REPORT

A 70-year-old woman complained of suddenly epigastralgia and increasingly severe right upper abdominal pain associated with abdominal fullness. Thus she was brought to our emergency service for help. US was performed and showed multiple simple cysts in liver, the largest measuring 10.8 cm in maximum diameter with homogenous content and thin wall. Then, she was diagnosed gastrointestinal disturbance with regular follow up in our OPD. One year later, she still complained of repeat RUQ abdominal pain but denied nausea, emesis, fever, jaundice, dysphagia, or anorexia. Because of her family history of hepatoma, she underwent regular US follow-up which showed growing hepatic cysts, the largest measuring at least 15 cm in maximum diameter and with homogenous content. The largest cystic lesion could not be detected completely by US because of its size. The patient decided to admission for further evaluation.

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Preliminary laboratory analyses revealed a slightly elevated serum carcinoembryonic antigen (CEA) level of 104.03 ng/ml (normal, <5 u/ml) and CA199 level of 24342 u/ml (normal, <30 u/ml). Serum bilirubin, hepatic enzymes, and coagulation parameters were normal, as was alpha-fetoprotein. Because of extraordinary high levels of CEA and CA199, a careful search for possible malignancy was performed. Abdominal MDCT with and without contrast enhancement was performed and demonstrated multiple hepatic cystic lesions in both lobes of liver (Fig. 1). Most of them were typical simple hepatic cysts with homogenous content, regular thin wall and smooth contour similar to the findings on US except the largest one (measuring $12 \times 14\text{cm}^2$ in size) in the right hepatic lobe. Owing to the use of MDCT of upper abdomen obtained with a 2-mm section thickness and a 1-mm interval, it was easy to identify the thin internal septa about 3mm in this lesion and focal irregular surface, which were both enhanced and not typical findings of simple hepatic cysts (Fig. 2, 3). Unlike the other peripheral hepatic simple cysts which were easily diagnosed, the possibility of cystic tumor for the largest one was considered by abdominal MDCT findings.

Because of progressive growth of the lesion and persisting right upper abdominal pain of the patient, percutaneous transabdominal aspiration was performed under US guidance for pain relief and pathologic proof. About 200 ml dark-brown fluid was aspirated. The cytological report only showed many degenerated red cells and further investigation was warranted. After a few days, the cystic lesion was refilled again and the size was increased about 16 cm in diameter which strongly implied the possibility of a cystic tumor. Partial hepatectomy of right hepatic lobe was performed and a huge cystic lesion measuring 15 cm was resected. The cystic lesion was filled with mucinous fluid. The pathological

Figure 1. Abdominal MDCT with contrast enhancement showed multiple cystic lesions with similar appearance in the right lobe of liver.

Figure 2. a. Pre-contrast abdominal MDCT showed thin internal septum (arrow) about 3mm in diameter of the largest cystic lesion. b. Post-contrast abdominal MDCT showed enhanced thin internal septum (arrow) about 3mm in diameter of the largest cystic lesion.
examinations revealed single layer of cuboidal epithelium with surrounded dense collagenous tissue of the lesion without evidence of malignancy. The final diagnosis was intrahahepatic biliary cystadenoma.

DISCUSSION

Biliary cystadenomas are rare benign tumors. The majority of biliary cystadenomas are entirely intrahepatic, and the rest of them arise from extrahepatic tissue and the gallbladder [1]. They predominantly occur in the middle-aged females and frequently cause abdominal pain and/or a palpable mass [1-4]. The most frequent symptoms and signs include right upper abdominal discomfort, hepatomegaly on physical examination, abdominal swelling, dyspepsia, nausea, and vomiting. They are all non-specific. Biliary dilatation is uncommon because they rarely communicated with the biliary ductal system and should raise a clinical suspicion of malignancy if present. Most of these tumors are macroscopically multilobulated and large, occasionally exceeding 10 cm in largest diameter with fluid content [1-5]. They may contain mucinous fluid produced by mucin-secreting cuboid or columnar epithelium [2-4]. It was previously proposed that cystadenomas were congenital and they were thought to develop from either an aberrant hamartomatous bile duct or from an ectopic remnant of embryonal gallbladder [1]. Recently, peribiliary glands distributed around the large bile ducts have also been suggested to be the origin [6]. Total surgical resection is the preferred treatment for biliary cystadenomas because of their premalignant nature. If they can be removed completely, the prognosis is excellent [1, 5], but the recurrence is possible.

The imaging characteristics of biliary cystadenomas and cystadenocarcinomas have been described [1-5, 7, 8]. The characteristic CT findings are multiloculated, well-defined cystic masses with internal septa and mural nodules. The internal septa and wall usually show enhancement after contrast enhancement. It may be so thin that it is barely recognized in CT study. US usually shows a cystic mass with multiple echogenic septa and/or papillary projections along the wall or septa [2-5, 8].

In our case, the cystic lesion was mistaken for a hepatic simple cyst by US because that: (a) the location of lesion was at hepatic dome which was difficult for US survey; (b) the cystic lesion was too large for US to detect the whole internal echogenicity; (c) the internal septa was too thin to detect. Under these circumstances, MDCT can provide more detailed images than US in identification of the internal septa and mural nodules even the lesion is only 1mm in size as well as differential diagnosis with simple hepatic cysts. In this case, the clue of significantly increased size strongly implied the possibility of a cystic tumor and the evidence of cystic tumor became more straightforward by using MDCT,
leading to the correct diagnosis.

The differential diagnosis for biliary cystadenoma include neoplasms (biliary cystadenocarcinoma, mesenchymal hamartoma, undifferentiated embryonal sarcoma, cystic hepatoma and cystic metastasis) and nonneoplastic disorders such as infectious disease (abscess, intrahepatic hydatid cyst) and developmental lesion (simple hepatic cysts and hematoma) [7].

It is difficult to differentiate cystadenoma from cystadnecarcinoma on CT or US. The presence of solid nodular masses or coarse calcifications along the wall or septa may indicate biliary cystadenocarcinoma [9]. Mesenchyma hamartoma [10] and undifferentiated embryonal sarcoma [11] are rare hepatic neoplasms that can have a multiloculated cystic appearance on CT and US, but their occurrence almost exclusively in children and teenagers differentiates them from cystic biliary neoplasms. A cystic hepatoma or metastasis can rarely simulate a unicocular cystadenoma or cystadenocarcinoma. Tumor maker will offer us more clues about the cystic lesion [7].

Liver abscess and hydatid disease of liver are the two entities most likely to be confused with biliary cystadnoma. Between 20% and 30% of liver abscesses, whether pyogenic or amebic in etiology, have a septated or multiloculated appearance on CT images [12]. Hydatic liver cysts caused by echinococcus granulosus are commonly well-defined hypoattenuating lesion with a distinguishable wall which mimic biliary cystadenoma. But coarse calcifications of the wall are present in 50% of cases, and daughter cysts are identified in approximately 75% of patients [7]. The diagnosis of both infectious diseases is usually easily made by combination of clinical and laboratory findings, including serologic tests and/or Gram stain and culture of cystic fluid obtained by percutaneous aspiration. Solitary simple hepatic cysts are sometimes shown on routine screening examinations of the upper abdomen. Unlike septa that are occasionally seen in otherwise benign-appearing renal cysts, in our experience, septa and mural nodule are less commonly shown within hepatic cysts discovered as an incidental finding on CT or sonography [7]. Subacute or chronic hematoma is similar to biliary cystadenoma. MRI is more suitable than CT for detection and characterization of hematoma and the clinical trauma history is also important.

In summary, multiple simple hepatic cysts seen on US may conceal a cystic tumor. If there are clinical features at all suggestive of something beyond a simple cyst, further evaluation is warranted. Abdominal MDCT, with its greater degree of resolution, is a reasonable next step in the evaluation of a suspicious cystic lesion.

REFERENCES

1. Ishak KG, Willis GW, Cummins SD, Bullock AA. Biliary cystadenoma and cystadenocarcinoma: report of 14 cases and review of the literature. Cancer 1977; 38: 322-338
偽裝在鄰近多發性單純肝囊泡的肝內膽管囊腺瘤：
一病例報告

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肝內膽管囊腺瘤（biliary cystadenoma）是一種很罕見的腫瘤，而單純性肝囊泡（simple hepatic cysts），卻是常常可見的。如果沒有看到內部分隔或是結節的話，我們很容易會把肝內囊狀腫瘤誤認為單純性肝囊泡。在此我們報導一位有多發性單純肝囊泡的七十歲女性，其中的一顆最後診斷為肝內膽管囊腺瘤。因為這顆囊狀腫瘤在超音波下很像單純性肝囊泡，所以一年後才被電腦斷層診斷出來。電腦斷層可以進一步看到很薄的內部分隔和局部不規則的邊緣進而做出正確診斷。

關鍵詞：膽管囊腺瘤；肝囊泡