

Biliary Cystadenoma and Cystadenocarcinoma: CT and Sonographic Findings

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Biliary cystadenomas and cystadenocarcinomas are rare cystic neoplasms, usually intrahepatic in location, that are characterized pathologically by a multilocular appearance. We report the CT and sonographic findings in eight cases of biliary cystadenoma and three cases of biliary cystadenocarcinoma and correlate them with the surgical and pathologic findings. CT showed internal septa in eight of the 10 multiloculated lesions, whereas sonography showed septa in all five cases in which it was used. CT in two of the three cystadenocarcinomas showed thick and coarse mural and septal calcifications as well as large solid soft-tissue masses in one. CT showed mural soft-tissue nodules in the single case of a unilocular cystadenocarcinoma.

Sonography and CT usually show the multilocular nature of biliary cystadenomas and cystadenocarcinomas. The presence of mural or septal nodules, discrete soft-tissue masses, and possibly thick and coarse calcifications increase the likelihood of a cystadenocarcinoma.

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Biliary cystadenoma is a rare cystic neoplasm occurring primarily in middle-aged women. Although usually benign, it tends to recur after subtotal excision and can develop into a malignant cystadenocarcinoma [1, 2].

The most striking feature of the gross pathologic specimen of a biliary cystadenoma or cystadenocarcinoma is its multiloculated appearance. Even in the exceptional tumor where the lesion appears to be primarily unilocular, gross or microscopic loculi can be identified in the cyst wall [2].

Scattered case reports and small series describing the CT and sonographic appearances of these lesions suggest that they have a characteristic appearance [3-9]. Most reports describe a septated, multiloculated cystic mass with varying degrees of mural and septal thickening and nodularity. We report eight patients with biliary cystadenomas and three patients with biliary cystadenocarcinomas in whom the appearance of the tumors on CT and sonography was correlated with the surgical and pathologic features.

Materials and Methods

Eleven patients with a surgically proved diagnosis of biliary cystadenoma or cystadenocarcinoma who had undergone CT and/or sonography were included in a retrospective study. The cases were collected from several different medical centers. No attempt was made to include all of the patients with these lesions seen at these institutions.

There were 10 women and one man, with a mean age of 55 years (range, 26-76 years old). Abdominal pain and/or distension was the major clinical symptom. Duration of symptoms ranged from 2 weeks to 5 years. In two patients a liver "cyst" had been partially excised 5 and 30 years earlier, respectively. In three additional patients there had been multiple episodes of percutaneous aspiration of upper abdominal or liver "cysts" over a period ranging from 4 months to 5 years.

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CT and sonography were performed on a wide variety of instruments. CT units with scanning times of 5 sec or less were used. Four patients had both CT and sonography, six patients had only CT, and one patient had only sonography. Of the 10 patients who had CT, five studies were performed only with IV contrast enhancement, two only without IV contrast enhancement, and three both before and after contrast enhancement. A variety of methods was used to inject the IV contrast agents.

At surgery a large cystic mass was found in or contiguous with the liver in each case. The cyst was totally excised in four cases and partially excised in two others. Hepatic lobectomy or segmentectomy was performed in the remaining five patients. One of the patients undergoing hepatic lobectomy had had a partial resection several weeks earlier. Mucinous fluid was present in all or some portions of the lesion in most patients. A final histologic diagnosis of biliary cystadenoma was made in eight patients and a diagnosis of biliary cystadenocarcinoma in three patients.

A pathologic diagnosis of biliary cystadenoma was made when the cyst wall and septa were lined primarily by tall columnar epithelium similar to the lining of normal bile ducts. Three cases were diagnosed as papillary cystadenocarcinoma: in all three there was evidence of malignant papillary epithelium coexisting with other regions of benign columnar epithelium. The malignant portion typically consisted of hyperchromatic columnar cells with prominent nucleoli and frequent mitoses, forming disordered papillary projections and pleomorphic glands. The thick and coarse calcifications shown on CT in the two cystadenocarcinomas were confirmed on histologic examination. Cal-

cification was not mentioned in the patient with a benign cystadenoma in whom CT showed a single focus of thin calcification.

The medical records and imaging studies of the 11 patients were reviewed. From the medical charts of each patient we reviewed the discharge summaries, surgical reports, pathologic reports, and reports of all CT scans and sonograms. One of the authors reviewed the CT scans and sonograms of each patient and compared the retrospective interpretation with the initial report; no major discrepancies were apparent. The CT and sonographic findings were then compared with the clinical, surgical, and pathologic data.

Results

CT scans and sonograms showed a large cystic liver mass in 10 patients; an upper abdominal cyst in or adjacent to the liver was noted in the other patient. The size of the mass ranged from 5 to 27 cm in greatest diameter. No masses were localized to the bile ducts. In 10 of the 11 patients the imaging studies indicated a lesion more complex than a simple cyst: one or more septa were shown in nine patients and mural nodules were shown in the other. In the four patients who had both CT and sonograms the findings agreed in three and disagreed in one: septa shown on both examinations were confirmed in three patients, whereas in the other patient septa shown by sonography were not visualized on CT and

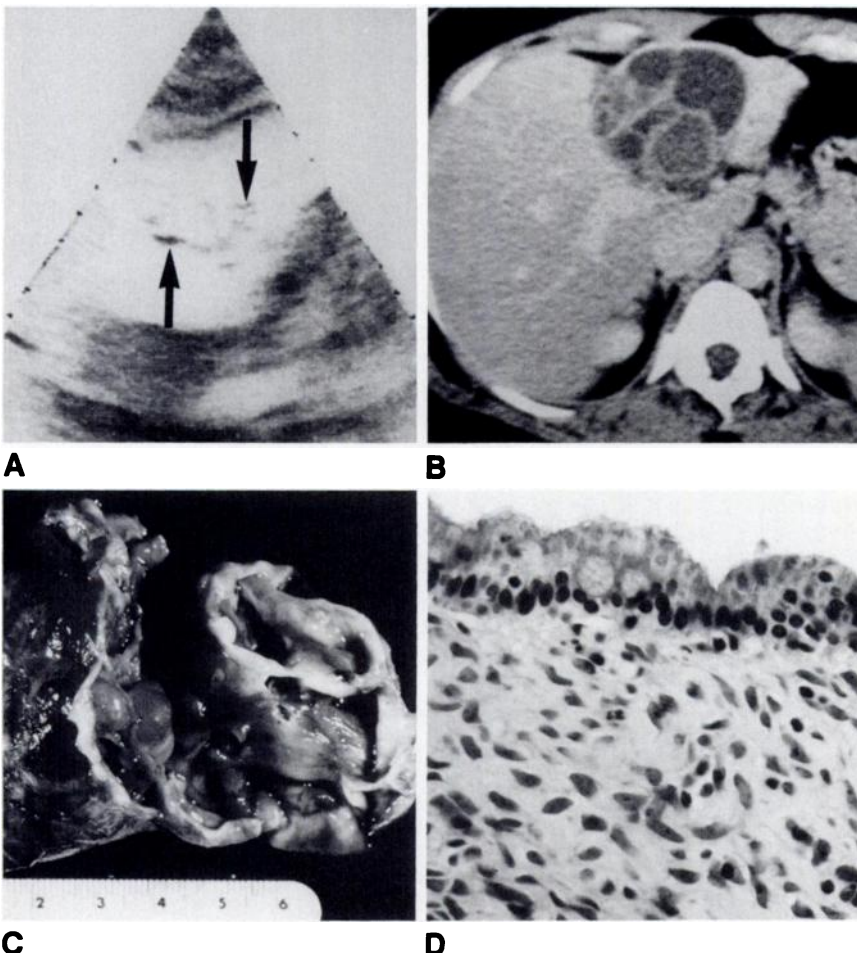


Fig. 1.—Biliary cystadenoma.
A, Sonogram shows cystic mass with thin echogenic septa (arrows).
B, CT scan shows multiloculated cyst in medial segment of left lobe. Note density of some loculations is higher than others.
C, Multiple septa and loculations are seen in unroofed half (right side) of gross pathologic specimen.
D, Microscopic section of typical septum shows benign tall columnar epithelium (top) overlying cellular stroma (H and E, $\times 500$).

were denied on the pathology report. The gross morphologic features of the complicated cysts were described in the CT and sonographic reports, but a histologic diagnosis usually was not offered. Specifically, the possibility of biliary cystadenoma or cystadenocarcinoma was mentioned only in a minority of the cases.

A final pathologic diagnosis of biliary cystadenoma was made in eight patients. In seven of these CT and/or sonography showed a septated multilocular cyst without gross nodules or soft-tissue masses (Figs. 1 and 2). A single focus of thin subtle calcification was seen in one of these seven cases. In the eighth patient, a discrete nodular echogenic structure associated with the internal septa was seen (Fig. 3). In three patients, all studied with CT only, the final pathologic diagnosis was biliary cystadenocarcinoma. In one, prominent soft-tissue mural nodules lined a unilocular cyst (Fig. 4). Thick, coarse mural and septal calcifications were seen in both multilocular cystadenocarcinomas, one of which also showed large soft-tissue masses within the cyst (Figs. 5 and 6).

Discussion

In order to analyze a series of patients with cystic biliary neoplasms we collected cases from several institutions using different CT and sonographic scanners and nonstandardized techniques. Although this is an unselected group representing all known patients from our institutions with a proven diagnosis of biliary cystadenoma or cystadenocarcinoma who underwent CT scans and/or sonography, this report is limited by its retrospective assessment of imaging features of a specific entity. In addition, the radiologic-pathologic correlation is based only on the surgical and pathologic reports, rather than the better approach in which the imaging studies are compared directly with the surgical and gross and micro-

scopic findings. This latter limitation may partially explain the few discrepancies between imaging and pathologic findings of internal septa.

Our series confirms that CT and sonography accurately demonstrate the internal morphologic features characteristic of cystic biliary neoplasms. Unequivocal septa and multilocularity within the cyst were shown in nine patients and confirmed by surgical and gross pathologic observations in seven of these. Of the other two patients, the cyst aspiration performed at surgery in one could have disrupted internal septa; in the other there was no specific comment about either the presence or absence of septa in the surgical and pathologic reports. Although not confirmed on this retrospective review, we believe it is likely that the highly characteristic appearance of septa on sonography in one case and on CT in the other correctly mirrored the internal morphology.

In one case imaged with CT but not sonography, multiple septa described in the surgical report were not visible on the CT scans. Overall we interpret the results of our study to indicate that CT failed to show definite or probable septa in two of 10 cases, whereas sonography correctly showed internal septa in all five cases in which it was used. As suggested in other reports, we believe that sonography is somewhat more sensitive than CT in the detection of septa in a cystic lesion [10].

Several previous reports of sonographic findings in benign cystadenoma have included a description of mural nodules and papillary projections in addition to the usual presence of septa within a liver cyst [4, 5]. It is difficult to explain what these sonographic nodules or projections represent: most authorities state that solid nodular projections, although common in biliary cystadenocarcinoma, are rare in cystadenoma [1, 2]. Even microscopically, papillary proliferation of benign epithelium is uncommon in cystadenoma. Only one of the cystadenomas in our series showed an unequivocal nodular mass on sonography.

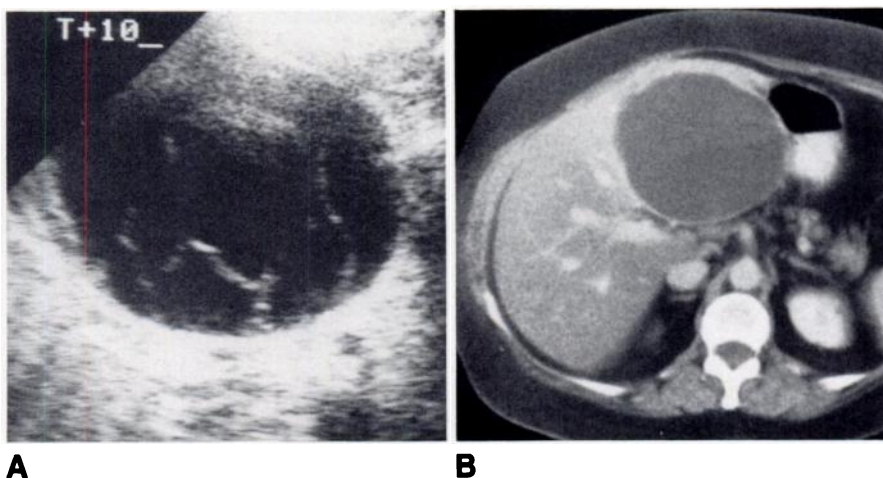


Fig. 2.—Biliary cystadenoma.

A, Sonogram shows septated liver cyst.

B, CT scan shows unilocular cyst with no evidence of internal septa.



Fig. 3.—Biliary cystadenoma. Sonogram shows echogenic nodule (arrow) within septated multilocular cyst. This was the only discrete nodule or mass seen in the eight benign cystadenomas.

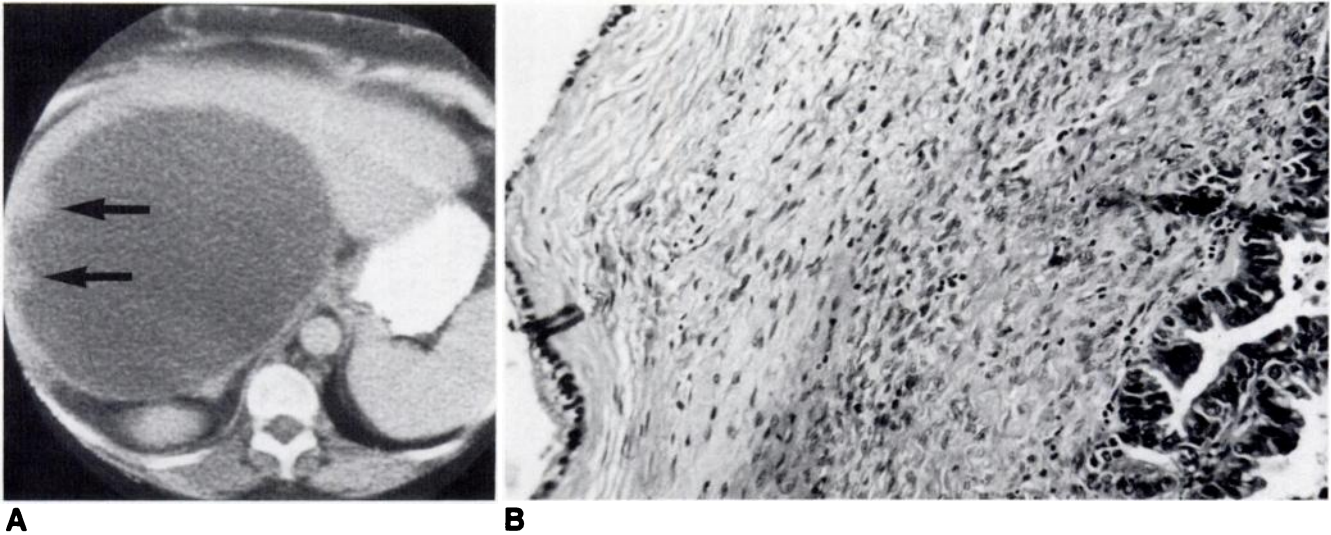


Fig. 4.—Biliary cystadenocarcinoma.

A, CT scan shows discrete mural nodules (arrows) lining unilocular hepatic cyst.

B, Histologic section shows both benign (left) and malignant (right) epithelium in the same mural nodule (H and E, $\times 250$).

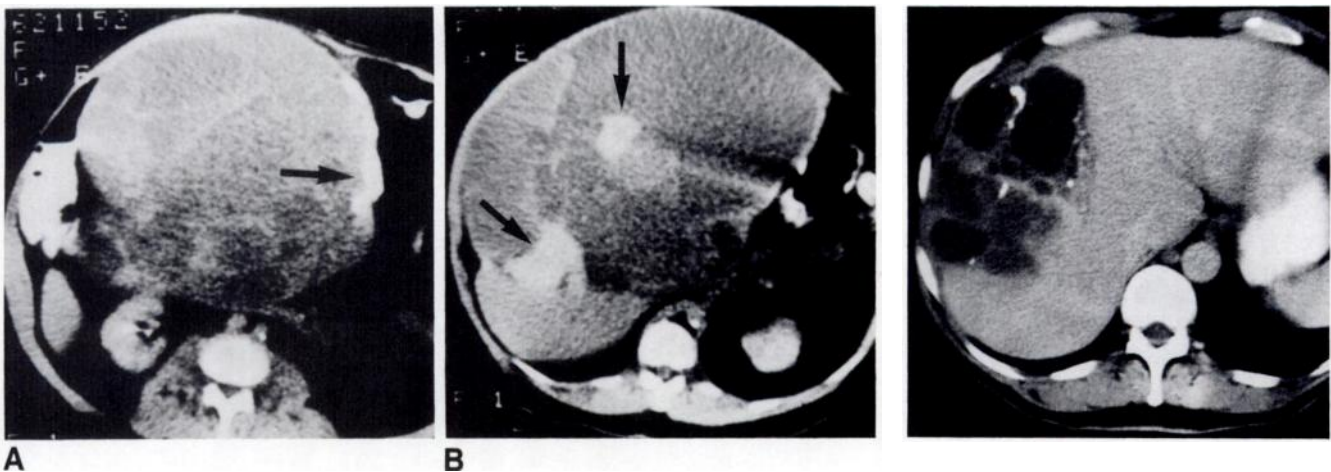


Fig. 5.—Biliary cystadenocarcinoma.

A, CT scan shows multiple septa poorly. Thick and coarse mural calcification is evident (arrow).

B, At a higher level, large, solid soft-tissue masses (arrows) are within septated cyst.

Fig. 6.—Biliary cystadenocarcinoma. CT scan shows coarse calcification of multiple thick septa.

Macroscopic nodules or solid elements were present on the CT scans in two of our three patients with biliary cystadenocarcinoma. These findings correspond to the grossly solid malignant tissue ranging up to 3.5 cm in diameter that Wheeler and Edmondson [2] described protruding from the internal cyst lining in their review of the pathologic features of malignant biliary cystadenocarcinoma. Multiple coarse and thick mural and septal calcifications were also shown on the CT scans of two of our three cases of cystadenocarcinoma, but a thin simple focus of mural calcification was also seen in one of our eight patients with cystadenoma. Although differentiation of cystadenoma from cystadenocarcinoma by imaging criteria may not be possible in many cases, our small series

suggests that the presence of solid nodular masses or coarse calcifications along the wall or septa in a multilocular cystic mass indicates a more likely diagnosis of biliary cystadenocarcinoma. Even if it is possible to distinguish biliary cystadenocarcinoma from cystadenoma by CT in some cases, the therapeutic implication of this distinction is minimal. The probable premalignant nature of biliary cystadenoma means that total surgical excision is the preferred treatment for both lesions. The essential point is that both neoplasms can almost always be differentiated from a simple hepatic cyst by CT and sonographic criteria.

Biliary cystadenoma must be differentiated from other cystic liver masses detected by CT and sonography. Solitary

simple hepatic cysts are sometimes shown on routine screening examinations of the upper abdomen. Although they are much less common than the ubiquitous renal cyst, similar care must be taken that the criteria for a simple cyst on CT and sonography are rigidly satisfied before other diagnoses are excluded. Unlike septa that are occasionally seen in otherwise benign-appearing renal cysts [11], in our experience septa are less commonly shown within hepatic cysts discovered as an incidental finding on CT or sonography. Mesenchymal hamartoma [12] and undifferentiated (embryonal) sarcoma [13] are rare hepatic neoplasms that can have a multiloculated cystic appearance on CT scans and sonograms, but their occurrence almost exclusively in children and teenagers differentiates them from cystic biliary neoplasms. A cystic hepatoma or metastasis [10] can rarely simulate a unilocular cystadenoma or cystadenocarcinoma.

Liver abscess and hydatid disease of the liver are the two entities most likely to be confused with biliary cystadenoma. Between 20% and 30% of liver abscesses, whether pyogenic or amebic in etiology, have a septated or multilocular appearance on CT images [14]. Hydatid liver cysts caused by *Echinococcus granulosus* are commonly multilocular in appearance because of the presence of daughter cysts within the original cyst [15]. The diagnosis of both infectious diseases is usually easily made by a combination of clinical and laboratory findings, including serologic tests and/or Gram stain and culture of cystic fluid obtained by percutaneous aspiration.

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