

Primary biliary cystadenocarcinoma perforating the duodenum and left intrahepatic biliary tree - mimicking a hydatid cyst

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Abstract: We report the case of a 76-year-old woman with biliary cystadenocarcinoma perforating the left biliary tree and exhibiting intratumoral gas bubbles resulting from invasion of the duodenum. The clinical history included subfebrile temperatures of 3 months duration, and pains associated with an abdominal mass in the right upper quadrant. Blood tests showed leucocytosis, and radiological studies revealed the features of a partially calcified septated tumor with nodular components combined with multiple gas-fluid levels, mimicking an infected hydatid cyst. Intraoperative ultrasonography, cholangiography and frozen section histology were necessary to prove the malignant nature of this cystic tumor. Provided that complete resection with strict adherence to oncological precepts is possible, the prognosis of cystadenocarcinoma is better than in hepatocellular or cholangiocellular carcinoma.

Bile duct eystadenocarcinoma is a rare, malignant cystic tumor lined by mucus-secreting columnar epithelium with papillary infoldings (1). Its basic histological features are similar to those of cystadenocarcinoma of the ovary and pancreas. Grossly, cysts are multilocular and the cyst wall is irregularly thickened with or without tumor infiltration into the adjacent parenchyma. It is common to see a transition from benign single layered columnar epithelium to malignant epithelial growth in histological specimens of this neoplasm. However, differentiating between a benign cystadenoma and carcinoma is often difficult in tumors not invading the adjacent parenchyma. On ultrasonography, the tumor is a globular or ovoid thick-walled mass, often containing multiple septations or papillary infoldings. On computer tomography it appears as a low density mass that contains mural nodules or internal septations. Adenocarcinoma or cholangiocarcinoma arising in a liver with cystic disease is often mistaken for cystadenocarcinoma. Incorrect diagnosis as a benign cystic disease such as hydatid

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cyst has been rarely reported (2). Although cyst puncture and sclerosing treatment procedures are recommended in benign cysts of the liver (3-5), we strongly endorse total surgical resection of cysts with unclear dignity. We report a case of a cystic liver tumor displaying gas-fluid levels, which was radiologically diagnosed first as an echinococcal cyst.

Case report

In October 1996, a 76-year-old female patient with a suspected hydatid cyst of the left liver lobe was admitted to our department. The patient's history disclosed that she had been suffering from an enlarging abdomen and subfebrile temperature for the past 3 months. On admission laboratory data were within normal limits except for mild leucocytosis of 12.1 G/l and an elevated axillary temperature of 37°C. Echinococcal antibodies, hepatitis markers and tumor markers were negative. Abdominal ultrasound showed a multilocular cystic

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tumor in the left liver lobe with a maximum diameter of 14 cm. The lesion contained septations, nodular tumorous components, areas of calcification within the septa and a thickened tumor wall. Furthermore, white echoes, implying intratumoral gas bubbles, which are common in infected hydatid cysts, were seen. A computer tomographic scan confirmed tumor calcifications and intratumoral air-fluid levels (Fig. 1). Enhanced computed tomography showed a well marginated, septated, multilocular cystic mass 14 x 11 x 10 cm in size. The cystic mass displaced the duodenum anteriorly, with signs of gut perforation by the tumor. The portal triad was displaced dorsally without evidence of bile duct dilatation or portal vein invasion.

Intraoperatively, we discovered a large cystic tumor occupying the left liver lobe, with inflammatory infiltrates extending into the pars superior duodeni. Furthermore, tumor penetration into the left intrahepatic biliary system was suspected on the basis of routinely performed intraoperative ultrasonography and cholangiography. Since the cystic mass macroscopically appeared to be neoplastic in nature, intraoperative frozen section was done to clarify the dignity of the tumor. A histopathological examination revealed adenocarcinomatous tissue. The cystic neoplasm was resected by left hepatic lobectomy (Fig. 2) and the duodenal invasion was locally resected and closed with single layer sutures. The bile duct system was rinsed with normal saline and the choledochal duct was drained with a T-drain, which is our usual procedure with ruptured inflammatory cysts (6, 7). The postoperative course was without complications, and to date (19 months) the patient, is doing well without any evidence of tumor recurrence.

Histological study demonstrated a radically resected middle-grade eystadenocarcinoma of the in-

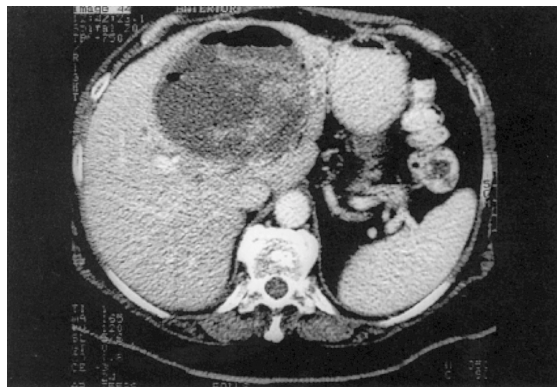


Fig. 1. Computed tomography scan showing mural calcifications and gas bubbles within the tumor.

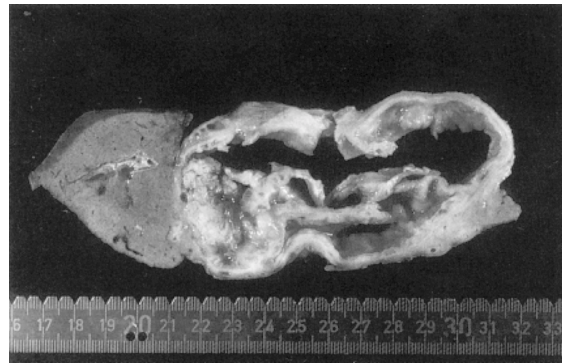


Fig. 2. Resected specimen with rest of normal liver tissue (left liver lobe).

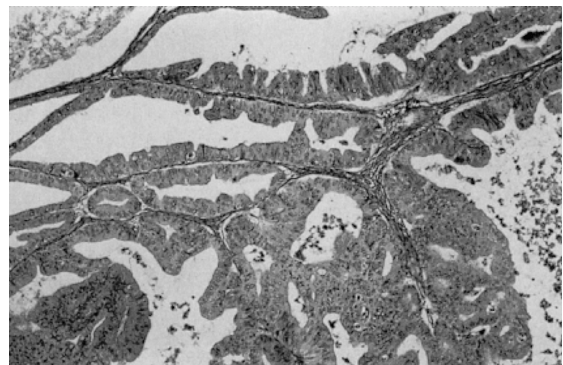


Fig. 3. Hematoxylin-eosin stained section of the cystadenocarcinoma (original magnification x100)

trahepatic bile duct with mesenchymal ovarian-like stroma (Fig. 3). The tumor had spread into neither the liver parenchyma nor the lymph nodes.

Discussion

Biliary cystadenocarcinoma, first documented by Willis, Richmond and Tompson et al. (8-10), is a rare malignant epithelial tumor of the liver. Takayasu reported an incidence of 0.41% among all hepatic malignant epithelial tumors, including autopsy cases (11). The etiology of cystadenomas of the liver remains unclear, but they are possibly due to an enteric malformation of the embryonic foregut (12). Immunohistochemical methods support the theory that biliary cystadenoma evolves from primitive hepatobiliary stem cells (13). The neoplasm usually occurs in middle-aged women, but reported cases range from a 2-year-old girl with cystadenoma up to an 84-year-old woman with cystadenocarcinoma (14). The histological finding of areas of benign epithelium in the majority of cys-

denocarcinomas favors the theory that these tumors arise in previously benign cystadenomas with mesenchymal stroma (incidence less than 5%), which are reported to have a clear malignant potential (12).

In contrast to the literature, we observed a higher incidence of cystic neoplasms during the last 14 years. Among the 64 cystic liver tumors treated at our institution, we diagnosed 30 hydatid cysts, 13 simple cysts, 7 cases of polycystic disease, 2 binary cystadenomas, and 2 biliary cystadenocarcinomas. This represents a 6.25% incidence of cystic neoplasms and a 3.125% incidence of biliary cystadenocarcinoma. Between 20% and 30% of liver abscesses, whether pyogenic or parasitic in nature, appeared septated or multilocular on CT imaging, frequently with intratumoral calcifications and gas bubbles (15, 16). The ultrasound findings in biliary cystadenomas and cystadenocarcinomas were multilocular cystic masses with associated septations and nodular tumor components. In contrast hydatid cysts were usually presented with thicker tumor walls and intratumoral septations and displayed round or oval daughter cysts.

The presence of intratumoral septations in a cystic tumor with a hemorrhagic cystic content strongly suggests benign cystadenoma, whereas the same features with an additional nodular tumor component are more likely to be a malignant cystadenocarcinoma. In our experience, fine-needle aspiration biopsy is associated with sampling error and needle tract tumor seeding (17). Definitive frozen section diagnosis always remains difficult and is sometimes impossible in cystic tumors (18). In our case, better prognosis may be expected, because the tumor proved to be an intracystic noninvasive middle grade adenocarcinoma without angioinvasion (19, 20). We recommend complete surgical resection of cystic neoplasms with strict adherence to oncological precepts (21). Provided that these criteria are respected, the prognosis of this rare cystic tumor is better than in other hepatic neoplasms.

References

1. ISHAK K G, WILLIS G W, CUMMINS S D, BULLOCK A A. Biliary eystadenoma and cystadenocarcinoma. *Cancer* 1977; 39: 322-38.
2. RUTLEDGE J N, PRATT M C, TAUPMANN R E. Biliary cysta-

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- denoma mistaken for an echinococcal cyst. *South. Med. J.* 1983; 76: 1575-7.
3. LEWIS W D, JENKINS R L, ROSSI R L, et al. Surgical treatment of biliary cystadenoma. A report of 15 cases. *Arch Surg* 1988; 123: 563-8.
4. SHORT W F, NEDWICH A, LEVY H A, HOWORD J M. Biliary cystadenoma. *Arch Surg* 1971; 102: 78-80.
5. CAHILL C J, BAILEY M E, SMITH M G M. Mucinous eystadenomas of the liver. *Clin Oncol (R Coll Radiol)* 1982; 8: 171-7.
6. MISCHINGER H J, WERKGARTNER G, CERWENKA H, et al. Management of hydatid disease of the liver-10 years' experience in a non-endemic area. *J Hep Bil Pancr Surg* 1997; 4: 315-21.
7. CERWENKA H, WERKGARTNER G, BACHER H, EL SHABRAWI A, MISCHINGER H J. Intrahepatic hematoma with secondary salmonella infection via biliary fistula. *Hepatogastroenterology* 1997;44:529-32.
8. WILLIS R A. Carcinoma arising in congenital cysts of the liver. *J Pathol Bacteriol* 1943; 55: 492-5.
9. RICHMOND H C. Carcinoma arising in congenital cysts of the liver. *J Pathol Bacteriol* 1956; 72: 681-3.
10. TOMPSON J E, WOLFF M. Intrahepatic cystadenoma of bile duct origin, with malignant alteration: Report of a case treated with total left hepatic lobectomy. *Mil Med* 1965; 130:218-24.
11. TAKAYASU K, MURAMATSU Y, MORIYAMA N, et al. Imaging diagnosis of bile duct eystadenocarcinoma. *Cancer* 1988; 61: 941-6.
12. WHEELER D A, EDMONSON H A. Cystadenoma with mesenchymal stroma (CMS) in the liver and bile ducts: a clinicopathologic study of 17 cases, 4 with malignant change. *Cancer* 1985; 56: 1434-45.
13. SIREN J, LÄRKKÄINEN P, LUUKKONEN P, KIVILUOTO T, KIVILAAKSO E. A case report of biliary cystadenoma and cystadenocarcinoma. *Hepatogastroenterology* 1998; 45: 83-9.
14. DEVANEY K, GOODMAN Z D, ISHAK K G. Hepatobiliary cystadenoma and cystadenocarcinoma. A light microscopic and immunohistochemical study of 70 patients. *Am J Surg Pathol* 1994; 18: 1078-91.
15. MISCHINGER H J, HAUSER H, RABL H, et al. Pyogenic liver abscess: studies of therapy and analysis of risk factors. *World J Surg* 1994; 18: 852-8.
16. SAFIOLEAS M, MISIAKOS E, MANTI C, KATSIKAS D, SKALKEAS G. Diagnostic evaluation and surgical management of hydatid disease of the liver. *World J Surg* 1994; 18: 859-65.
17. IEMOTO Y, KONDO Y, NAKANO T, TSUCHIYA K, OHTO M. Biliary eystadenocarcinoma with peritoneal carcinomatosis. *Cancer* 1981; 48: 1664-7.
18. KLIMPFINGER M, STEINDORFER P, HELLINGER P, et al. Probleme der intraoperativen Schnellschnittdiagnostik von Mammaläsionen-Analyse von 5479 konsekutiven Fällen. *Acta Chir Austriaca* 1997; 29: 118-22.
19. TSIFTSIS D, CHRISTODOULAKIS M, DEBREE E, SANIDAS E. Primary intrahepatic biliary cystadenomatous tumors. *Journal of Surg Oncol* 1997; 64 (4): 341-6.
20. NAKAJIMA T, SUGANO I, MATSUZAKIO, et al. Biliary cystadenocarcinoma of the liver: a clinicopathological and histochemical evaluation of nine cases. *Cancer* 1992; 69: 2426-32.
21. FONG Y, BRENNAN M F, COHEN A M, HEFFERNAN N, FREIMAN A, BLUMGART L H. Liver resection in the elderly. *Br J Surg* 1997; 84(10): 1386-90.