

## **A case report : EXTRAHEPATIC BILIARY CYSTADNOCARCINOMA ARISING FROM COMMON HEPATIC DUCT.**

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### **ABSTRACT**

A case of extrahepatic biliary cystadenocarcinoma arising from the common hepatic duct is reported. The sonographic, CT and ERCP appearances correlated well with the pathological findings. The typical imaging characteristics of biliary cystadenoma and cystadenocarcinoma especially the sonography and CT can lead one to the correct preoperative diagnosis even the tumor arising from the extrahepatic bile duct.

### **INTRODUCTION**

Cystadenoma and cystadenocarcinoma are rare cystic tumors of biliary origin. They comprise of less than 5% of intrahepatic cystic lesions.<sup>1,2,3</sup> Biliary cystadenoma and cystadenocarcinoma may arise either from intrahepatic or extrahepatic bile duct as well as the gall bladder. However, most of them are located entirely within the liver.<sup>1,3,4</sup> Characteristic sonographic and computerized tomographic findings of intrahepatic biliary cystadenoma and cystadenocarcinoma were described as multiloculated or less often as uniloculated cystic mass lesion.<sup>2,3,6,7,8</sup> We report further on the typical imaging appearances of extrahepatic biliary cystadenocarcinoma arising totally from the common hepatic bile duct without contiguous intrahepatic bile duct component.

### **CASE REPORT**

A 42-year-old female patient presented with epigastric discomfort and mild intermittent jaundice for about 4-month duration. A large non-tender mass with firm to hard consistency was palpable in the right upper quadrant. No ascites nor palpable lymph nodes was revealed through physical examination. The hematogram and urine analysis were within normal limits. Minimal elevation of both direct bilirubin and total bilirubin as well as serum amylase were noted but the liver enzymes and alkaline phosphatase were

within the normal ranges.

Ultrasonography revealed a large, well-defined mass arising from common hepatic duct. It composed of multiple cystic spaces separated by septa. Several cysts contained internal echoes. Focal nodular thickening of septa was noted. The lesion caused proximal ducts dilatation and compressed upon the gall bladder. (Fig. 1 a, b)

CT confirmed the presence of a well encapsulated multiloculated cystic mass with variable attenuation values in the common hepatic duct causing proximal bile ducts dilatation. A coarse calcification was seen eccentrically within the wall. Nodular thickening of septa with enhancement of the septa and wall of the mass lesion was also observed. No other intrahepatic lesion, ascites or enlarged lymph nodes was detected. (Fig. 2 a, B)

ERCP demonstrated a large ovoid shape filling defect in the common hepatic duct with dilatation of both main intrahepatic ducts. The gall bladder was not opacified. (Fig. 3)

At laparotomy, a multiloculated cystic mass arising from the common hepatic duct was seen compressing the nearby gall bladder. Complete excision of the mass, cholecystectomy and hepaticojejunostomy were done. The patient recovered well after surgical treatment.

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ERCP

E = Endoscopic

R = Retrograde

CP = Cholangiopancreatography



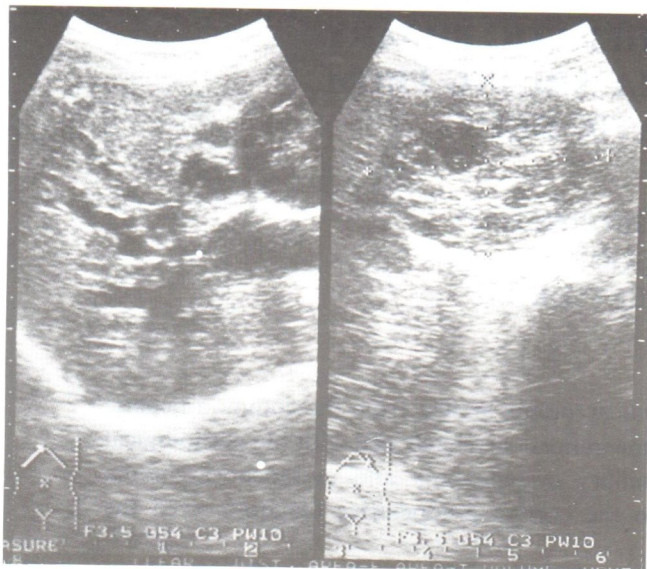


Fig. 1a . A large multiloculated mixed echoic mass was demonstrated with intrahepatic duct dilatation.

Grossly, the common hepatic duct was remarkably dilated. There was a large solid-cystic mass with nodulated free surface,  $6 \times 4 \times 4$  cm, arising from the lateroposterior wall. The residual mucosa was smooth in comparison to the tumor surface where focal hemorrhage was observed. (Fig. 4a) Cut sections showed a multiloculated lesion with abundant mucinous content and irregular fibrous septa. (Fig. 4b) Microscopically, there were numerous clefts and cystic spaces lined by mucin-producing high columnar epithelium which were occasionally pseudostratified. (Fig 5a) Pleomorphic nuclei with dysorientation and mitotic figures were frequent in the micropapillae and abortive tubules. (Fig 5b) Notably, small cysts with low columnar epithelia and hyperchromatic nuclei were observed in the fibrous septa and fibrotic wall of the hepatic duct. They were interpreted as stromal invasion. Calcification within the luminal mucin was focally observed. Neither vascular invasion nor distant metastases were seen.

## DISCUSSION

Biliary cystadenoma and cystadenocarcinoma are rare, being slow growing tumors of biliary tract. They are assumed to be developed originally from aberrant hamartomatous bile ducts or ectopic rests of embryonic gall bladder.<sup>1,4,8</sup> They occur predominantly in middle aged women, with ages ranging from 19 to 71 years.<sup>2,3,4</sup> Most patients presented with epigastric discomfort and mass in right upper quadrant or epigastrium with or without fever and intermittent jaundice.<sup>1,3,7</sup> The

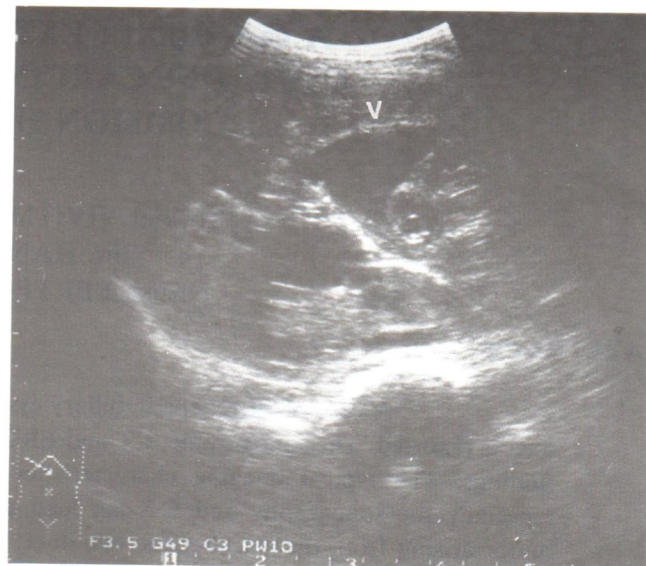


Fig. 1b . The mass was clearly seen arising from common hepatic duct (arrow).

mass is usually large at presentation.

The ultrasound appearance of cystadenoma and cystadenocarcinoma is of oval, cystic, anechoic mass with multiple septa. The cystic spaces occasionally have internal echoes suggesting infection or hemorrhage.<sup>3,6,7</sup> The mural nodules and papillary projections were seen arising from the walls and septae. These findings were commonly noted in biliary cystadenocarcinoma<sup>1,4,5</sup> but they could not be used as the indication of malignancy since they also have been described in benign cystadenoma as well.<sup>2,3,5,5,6,7</sup>

The CT of biliary cystadenoma and cystadenocarcinoma reveal multiloculated cystic areas with attenuation value ranging from 0 to 30 Hounsfield Units.<sup>3,6,10</sup> The variation of attenuation is due to various contents such as serous fluid, mucin, pus or cholesterol.<sup>3,5,10</sup> Enhancement of internal septations with intravenous contrast administration was also described.<sup>3,7</sup> Sonography is more sensitive than CT in the detection of septa in a cystic lesion. CT may detect calcifications in these lesions.<sup>5,9</sup> The presence of solid nodular masses or coarse calcifications along the wall or septa in a multilocular cystic mass suggested a more likely diagnosis of biliary cystadenocarcinoma.<sup>1,4,5</sup>

The sonographic and CT findings in our case are correlated well with the previous description of biliary cystadenocarcinoma even though it arises from the common hepatic duct. The other tumors which may arise from the extrahepatic bile duct such as choledochal cyst with or without malignant change,<sup>11,12</sup> hepatocellular carcinoma<sup>13</sup> and cholangiocarcinoma<sup>14,15</sup>



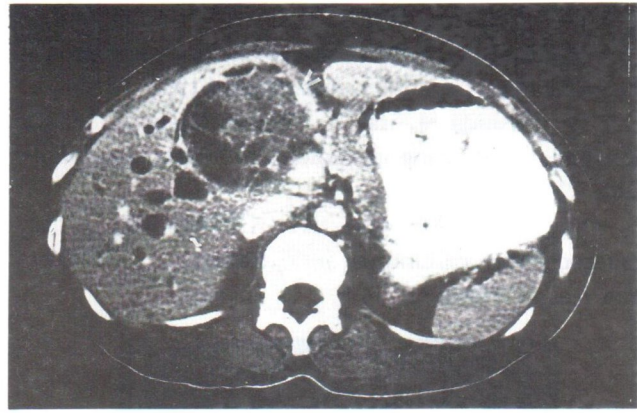
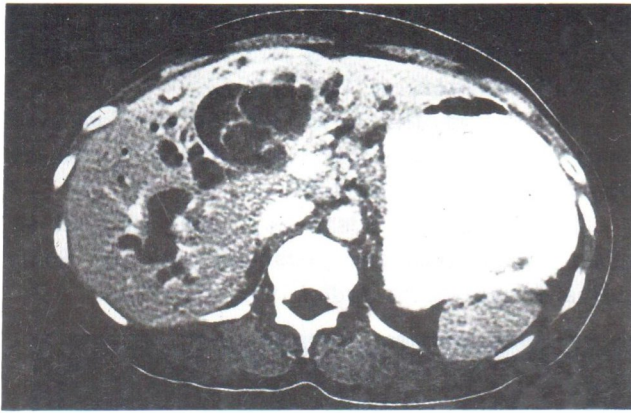


Fig. 2a . Axial CT showed a multiloculated cystic mass with variable attenuation contents, nodular thickening of septa and a coarse calcification (arrow) in the common hepatic duct. Intrahepatic duct dilatation was also noted.

were not presented as this multiloculated cystic character. Complete surgical removal of the tumor is the treatment of choice since this can relieve the biliary obstruction and patient's symptoms.<sup>1,7</sup> Most importantly, it provides histologic confirmation of benign or malignant nature that cannot be relied on the imaging characters.<sup>5,6</sup> Histologically, biliary cystadenomas appear well encapsulated and are composed of numerous cystic spaces lined with a single layer cuboid or columnar nonciliated, mucin-secreting epithelium. The cyst wall contains epithelium of biliary tract origin with a capsule of mesenchymal tissue.<sup>1,4</sup> Evidence of stromal invasion indicates malignancy.<sup>4</sup>

In conclusion, our case confirms that sonography and CT can accurately demonstrate the internal morphologic features characteristic of cystic biliary neoplasm even arising in the common hepatic duct.

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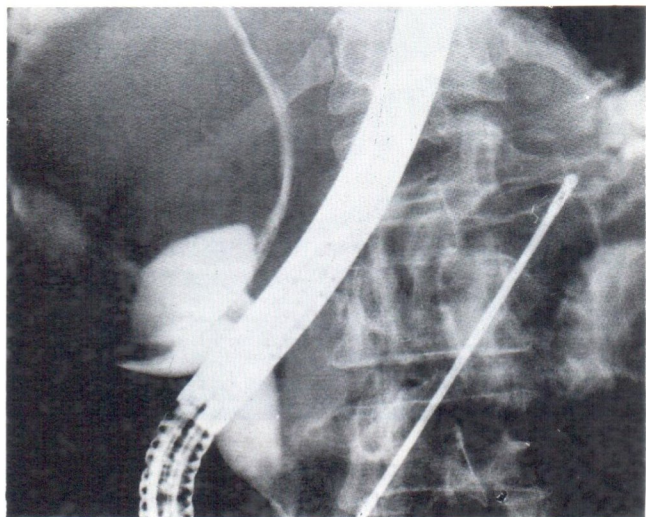


Fig. 3 ERCP demonstrated a large ovoid filling defect in the common hepatic duct.



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Fig. 4a . The tumor mass with nodulated surface and large area of hemorrhage arised from lateral wall of the common hepatic duct.

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Fig. 4b . The cut section disclosed multiloculation filled with mucin and separated by various thickness of septa.

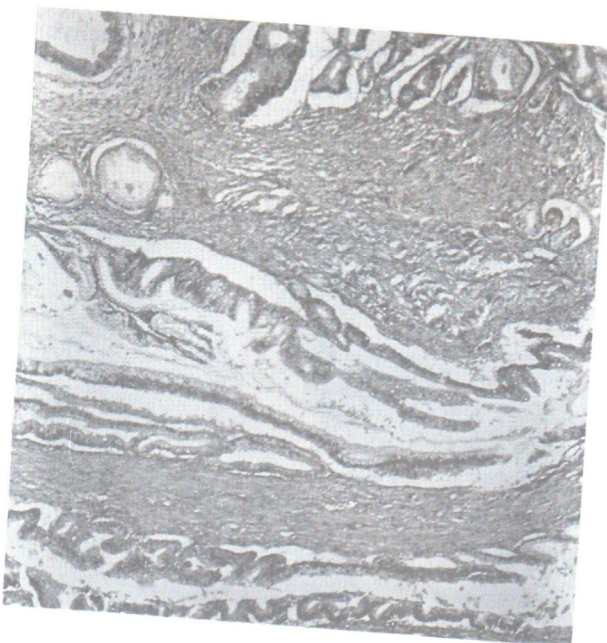


Fig 5a, b : Microscopically, the tumor mass consisted of abortive and well-formed tubular structures with stroma invasion. Formation of micropapillae with bizzare and dysorientated nuclei were prominent.