Journal of Zhejiang University SCIENCE
ISSN 1009-3095
http://www.zju.edu.cn/jzus
E-mail: jzus@zju.edu.cn

Ciliated hepatic foregut cyst: report of first case in China and review of literature

CAI Xiu-jun (蔡秀军)†, HUANG Di-yu (黄迪宇), LIANG Xiao (梁 霄),
YU Hong (虞 洪), LI Wei (李 伟), WANG Xian-fa (王先法), PENG Shu-you (彭淑牖)
(*Department of General Surgery, Sir Run Run Shaw Hospital, Zhejiang University, Hangzhou 310016, China*)
†E-mail: cxjzu@hotmail.com

Received May 21, 2003; revision accepted Sept. 18, 2003

Abstract: Objective: To report the first case of ciliated hepatic foregut cyst in China, and review of literature to introduce the characteristics of this disease for doctors to recognize this disease. Method: Report the clinical procedure of diagnosis and treatment for the first case of ciliated hepatic foregut cyst in China, and to review the embryologic genesis, incidence, clinical manifestation, radiologic features and therapeutic principle of this disease. Results: We performed the resection for ciliated hepatic foregut cyst under laparoscopy; the patient recovered well after the procedure. Conclusion: Ciliated hepatic foregut cyst is quite rare clinically, belongs to non-parasitic, solitary and unilocular cystic lesion, is always less than 4 cm in diameter, mostly seen in the left lobe, and has the tendency of malignant change. It should be removed as soon as diagnosed.

Key words: Liver, Ciliated foregut cyst, Laparoscope, Hepatectomy
Document code: A **CLC number:** R774.31

INTRODUCTION

Cystic lesion of liver is generally divided into parasitic and non-parasitic ones. Ciliated hepatic foregut cyst (CHFC) is non-parasitic, and first used by Wheeler and Edomondson (1984) to describe the hepatic cystic lesion arising from the embryologic foregut. Ciliated foregut cyst mostly occurs in the tracheobronchial tree and esophagus, and is rarely found in the liver. According to the foreign literature, there were 72 cases reported, none in China. Recently we performed a laparoscopic hepatectomy for a mass in the left lobe. The postoperative pathologic report was CHFC, the first case reported in China.

CLINICAL MATERIALS

The patient was male, 30 years old. His chief complaint was dull pain in right upper abdomen for 5 months. He had no fever or chilling, no nausea or vomiting, no icterus. P.E.: no jaundice, no palpable enlarged superficial lymph nodes, and no any significant signs in the abdomen. Total bilirubin was 0.7 mg/L, direct bilirubin 0.2 mg/L, albumin 4.2 g/L, AFP, CEA and CA19-9 were all negative. B-Ultrasound detected a mass about 4 cm in diameter in the IV segment of liver. CT scan showed a round low-density and like siege lesion, with clear margin, in the above mentioned segment of the liver, and could be slightly enlarged, so solid mass of left lobe was suggested. We performed a laparoscopic hepatectomy of the mass in the left lobe under general anesthesia on Jan 17, 2002. During the operation, we found a protruding cystic mass in the IV segment of the left lobe, with a clear margin, and

a thick, intact cyst wall. We removed the mass completely along the liver tissue 2 cm away from the mass. The procedure was smooth; intraoperative blood loss was 20 ml, the operative lasted 90 mins. The specimen was a 6×6×5 cm unilocular cyst, and had thick yellow fluid in it, and a thick, smooth cystic wall. Postoperative pathologic examination showed the wall of the cyst was composed of fibrous smooth muscle tissue, and dilated cyst-like bile ducts were found in the peripheral tissue; the inner layer of the cyst was pseudostratified columnar epithelium. The final pathologic report revealed it was a CHFC.

DISCUSSION

Ciliated foregut cyst mostly occurs in the mediastinum, and is commonly generated from the tracheobronchial tree and esophagus, and is less frequently found in the liver. This lesion is always solitary, unilocular and less than 4 cm in diameter. Its wall is composed of 4 layers: pseudostratified columnar epithelium with admixed mucous cells, loose subepithelium connective tissue, one to three smooth muscle layers and an outer fibrous capsule (Wheeler and Edmondson, 1984; Terada *et al.*, 1990). Friedreich described the first case of CHFC in 1857, and postulated it was congenital, arising from embryological abnormality at that time. Jones suggested that it was a certain type of non-parasitic liver cystic lesion. Wheeler and Edmondson (1984) first used the name CHFC to describe this disease as related articles kept on increasing from then on, so doctors have gradually recognized this disease.

The embryological origin of CHFC is still uncertain, although most authors advocated its arising from the embryological foregut, also arising from the foregut are the primitive pharynx, esophagus, stomach, part of the duodenum beyond the opening of the common bile duct, liver, biliary system, pancreas, tracheobronchial tree and lungs (Gray and Skandalakis, 1972). By the end of 3 weeks of gestation, the ventral cranial foregut dilates to form a laryngobronchial diverticulum, from which are developed trachea, bronchi, bronchioles and pulmonary bud. At the beginning of the 4 weeks of gestation, the ventral caudal foregut develops a process to form hepatic diverticulum from which are developed liver and biliary tract. The primitive diaphragm separates the pleural and peritoneal cavity. This separation was not complete initially, and formed two canals between the pleural and peritoneal cavity, that allows the growing of lung buds. By 8 weeks of gestation, the pleura and peritoneum fuse together and the canals close. But at that time, abnormal bronchioles could emigrate from the pleural cavity to the peritoneal cavity before the closure of the canals, and then is surrounded by the endoblast of the hepatic diverticulum, and formed the cystic mass finally. The left lobe is the major part of the primitive liver, which may explain why most CHFCs locate in the left lobe.

This disease is slightly predominant in the male, and common outbreak age is approximately 50. Except for occasionally dull upper abdominal pain, most patients have no significant symptoms, and most cases were diagnosed by imaging, autopsy or surgical exploration. There was one case reported to be characterized by portal vein compression which caused portal hypertension and splenomegaly (Harty *et al.*, 1998); one case had bile duct compression which caused jaundice (Dardik *et al.*, 1964), and another case resulted in malignant transformation into squamous cell carcinoma (Vick *et al.*, 1999a; 1999b). Most cases located in the left lobe, but some cases located in the right lobe and attached to the gallbladder. The CHFC cyst is commonly unilocular and solitary, contains mucoid or viscous material.

Kadoya and Shoenut (Kadoye *et al.*, 1990; Shoenut *et al.*, 1994) found that this disease always appeared as a solitary lesion with clear margin in the radiological image. B-ultrasound always reveals hypoechpic or anechoic lesion. Plain CT scan reveals slightly protruding liver lower attenuation lesion without contrast enhancement. MRI presents high signal intensity in T2 weighted image, and that was postulated to be caused by protein components in the cystic fluid, but the T1 weighted image was quite different. MRI is considered to be helpful for distinguishing this disease from other oligvascular

solid tumors.

Terada *et al.*(1990) found neutral, acid, carboxylated and sulfate mucin. The wall stained positively for various cytoketatins and other epithelial markers such as CEA, EMA. Electron microscope revealed the presence of cilia in the epithelium of the inner wall, and numerous mucus containing granules contained in the goblet cells. Benlolo and Peltier (Benlolo *et al.*, 1996; Peltier *et al.*, 1993) also found the cyst contained neutral and acid mucin; the latter found the epithelium stained negative for EMA. Chatelain *et al.*(2000) identified the endocrine cells present in the epithelium had immunoreactivity with chromogranin and calcitonin. That is quite commonly the case of endocrine cells in the epithelium of the respiratory tract.

CHFC was considered to be a benign liver disease arising from the embryological foregut. Vick *et al.*(1999a) reported 6 CHFC cases; reviewed related literature in 1999, did not find any malignant tendency of this disease; and suggested aspiration and injection of sclerosing agents for those asymptomatic lesions, and suggested operation for cases which developed jaundice or portal hypertension. But 5 months later, they reported another case that required operation, and intraoperative finding revealed malignant transformation to squamous cell carcinoma. The patient survived only 2 months after the operation. Because this lesion is potentially malignant, and is often confused with other hepatic cystic lesions, aggressive procedure should be applied for hepatic cystic lesions. The application of laparoscopic technique in liver surgery has become riper now; we have successfully completed 12 laparoscopic hepatectomies, and acquired initial experience. We suggest that for those solitary, low density lesions located in the left lobe, we should pay additional attention to exclude the possibility of CHFC, and aggressive surgery is indicated for those cases.

References

Benlolo, D., Vilgrain, V., Terris, B., Zins, M., Belghiti, J., Menu, Y., 1996. Imagerie des kystes ciliés hépatiques ou biliaires ă revêtement cilié. *Gastroenterol Clin Biol*, **20**(5):497-501.

Chatelain, D., Chailley-Heu, B., Terris, B., Molas, G., Le-Cae, A., Vilgrain, V., Belhiti, J., Degott, C., Flejou, J.F., 2000. The ciliated hepatic foregut cyst, an unusual bronchiolar foregut malformation: a histological, histochemical, and immunohistochemical study of 7 cases. *Hum pathol*, **31**(2):241-246.

Dardik, H., Glotzer, P., Silver, C., 1964. Congenital hepatic cyst causing jaundice: Report of a case and analogies with respiratory malformations. *Ann surg*, **159**:585-592.

Gray, S.W., Skandalakis, J.E., 1972. Embryology for Surgeons. *In*: the Embryologic Basis for the Treatment of Congenital Defects. Saunders, Philadelphia, p.217-383.

Harty, M.P., Hebra, A., Ruchelli, E.D., Schnaufer, L., 1998. Ciliated hepatic foregut cyst causing portal hypertension in an adolescent. *AJR Am j roentgenol*, **170**(3): 688-690.

Kadoya, M., Matsui, O., Nakanuma, Y., Yoshikawa, J., Arai, K., Takashima, T., Amano, M., Kimura, M., 1990. Ciliated Hepatic foregut cyst: radiologic features. *Radiology*, **175**(2):475-477.

Peltier, E., Leger-Ravet, M.B., Franco, D., Lemnigre, G., 1993. Kystes â revetêment cilié du foie. Deux cas. *Gastroenterol Clin Biol*, **17**(11):859-862.

Shoenut, J.P., Semelka, R.C., Levi, C., Greenberg, H., 1994. Ciliated hepatic foregut cysts: US, CT, and contrast-enhanced MR Imaging. *Abdom imaging*, **19**(2): 150-152.

Terada, T., Nakanuma, Y., Kono, N., Ueda, K., Kadoya, M., Matsui, O., 1990. Ciliated hepatic foregut cyst. A mucus histochemical, immunohistochemical, and ultrastructural study in three cases in comparison with normal bronchi and intrahepatic bile ducts. *Am j surg pathol*, **14**(4):356-363.

Wheeler, D.A., Edmondson, H.A., 1984. Ciliated hepatic foregut cyst. *Am j surg pathol*, **8**(6):467-470.

Vick, D.J., Goodman, Z.D., Deavers, M.T., Cain, J., Ishak, K.G., 1999a. Ciliated hepatic foregut cyst: a study of six cases and review of the literature. *Am j surg pathol*, **23**(6):671-677.

Vick, D.J., Goodman, Z.D., Ishak, K.G., 1999b. Squamous cell carcinoma arising in a ciliated hepatic foregut cyst. *Arch pathol lab med*, **123**:1115-1117.