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Liver Pathology

Ciliated hepatic foregut cyst

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INTRODUCTION

Ciliated hepatic foregut cyst (CHFC) is a rare developmental disorder of the liver. Diagnoses of CHFC has become frequent during the past two decades due to technological progress and increased use of various imaging modalities.[1] Clinically and radiologically, this lesion can be difficult to distinguish from the other cystic neoplasms of the liver. In this issue, we present a case of CHFC and discuss the typical histopathologic findings.

CASE SUMMARY

A 65-year-old man was admitted to our hospital for the evaluation of an asymptomatic hepatic cyst detected incidentally during a health checkup. Serological tests for hepatitis B and hepatitis C virus were negative. All other parameters were normal. On contrast-enhanced computed tomography (CT) imaging, a 1.5 cm-sized, oval-shaped, low attenuated cystic lesion was demonstrated in hepatic segment IV, just beneath the hepatic surface. Internal attenuation of the lesion was slightly higher than that of simple cyst, possibly suggesting internal mucoid fluid or high protein content (Fig. 1).

PATHOLOGIC FINDINGS

Surgical exploration revealed a 1.5×1.2 cm-sized, well-circumscribed, unilocular cyst (Fig. 2A). The cyst was filled with brownish mucoid fluid. Histologically, the cyst was lined by pseudostratified ciliated columnar epithelium with scattered goblet cells (Fig. 2B). No mitotic figures were observed in the epithelial cells and there was no evidence of pleomorphism or nuclear atypia, suggesting malignant tumor. Masson's trichrome stain demonstrated distinct four layers of this cyst (Fig. 2C): 1) an inner layer of pseudostratified ciliated columnar epithelium with scattered goblet cells, 2) an underlying loose connective tissue, 3) a smooth muscle layer, and 4) an outer capsule layer consisting of dense fibrous tissue. Immunohistochemistry for alpha-smooth muscle actin highlighted the presence of smooth muscle layers in the cyst wall (Fig. 2D). Thirty-seven months after surgery the patient remained well and a

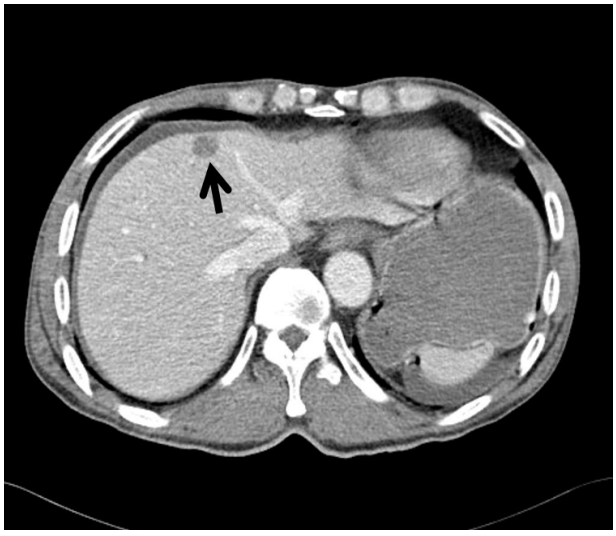

Figure 1. Portal venous phase of contrast computed tomography showing a 1.5 cm-sized, oval-shaped, low attenuated cystic lesion in hepatic segment IV (arrow).

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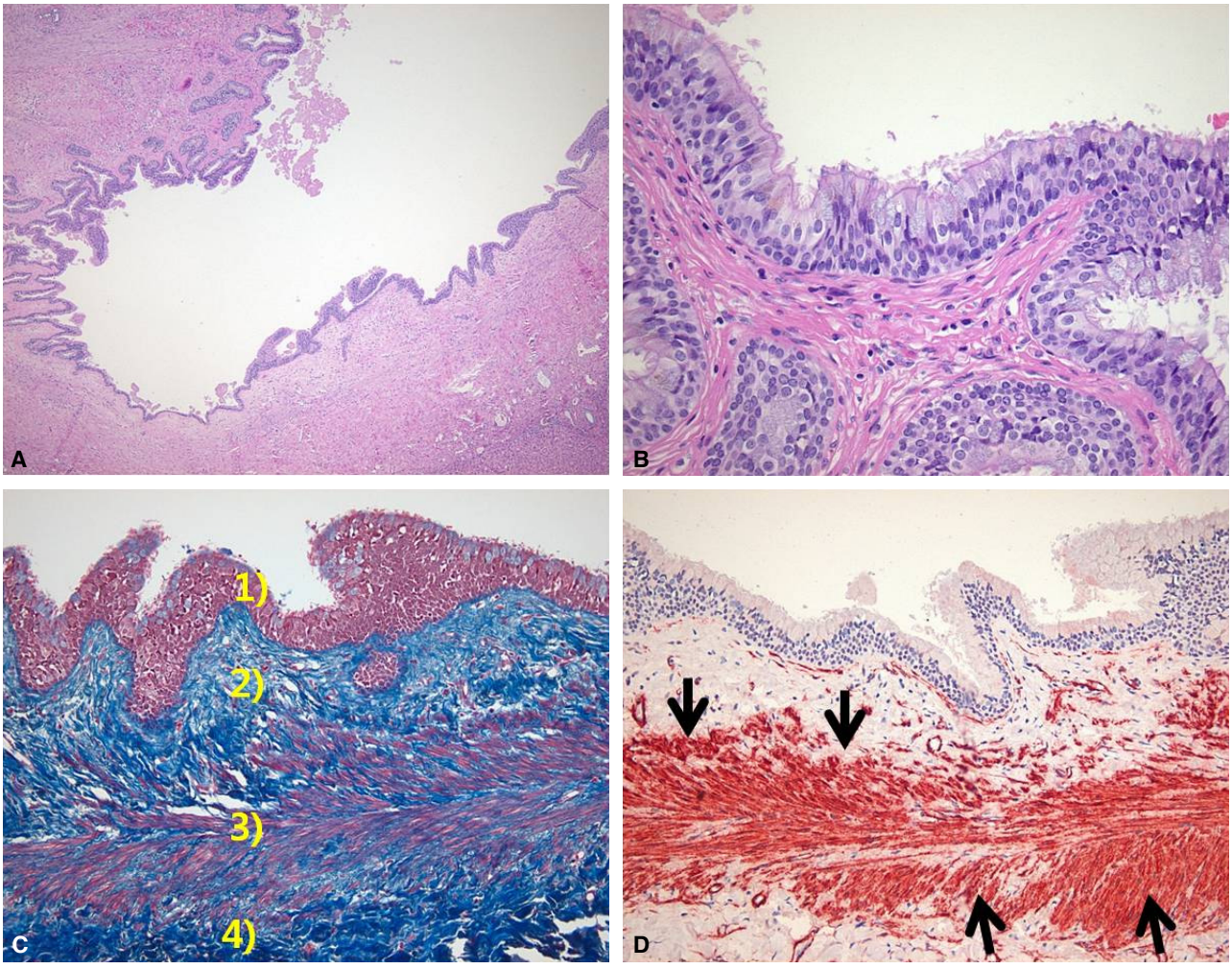

Figure 2. (A) Well-circumscribed, unilocular cyst with no solid tumor areas. (B) The inner cyst lining was composed of pseudostratified ciliated columnar epithelium with scattered goblet cells. (C) Masson's trichrome stain demonstrated four distinct layers of this cyst: 1) an inner lining epithelial cell layer, 2) underlying loose connective tissue, 3) smooth muscle layer, and 4) an outer fibrous capsule layer. (D) Immunohistochemistry for alpha-smooth muscle highlighted the presence of smooth muscle layer in the cyst wall (arrows). (A: hematoxylin and eosin (H-E), ×20; B: H-E, ×400; C: Masson's trichrome, ×200; D: alpha-smooth muscle actin, ×200).

follow-up CT scan showed no recurrent lesion.

DISCUSSION

CHFC is rare cystic disease of the liver. The term CHFC was coined to indicate the origin of this lesion as being a detached hepatic diverticulum or detachment and migration of buds from the esophageal and bronchial regions of the foregut and subsequent entrapment by the liver during the early embryonic development of the foregut.[1,2] It is found most commonly in segment IV of the left lobe just beneath the hepatic capsule, as in our case, whereas most other solitary cysts show a female predominance and greater occurrence in the right hepatic lobe.[3] The majority of patients are asymptomatic and the cyst is found incidentally. The clinical course of CHFC is essentially benign. However, a few CHFC patients have presented with carcinomatous transformation; these tumors were biologically aggressive.[1]

Although most CHFCs have typical imaging features, radiology alone is non-diagnostic. The various elements ranging from clear serous material to mucoid or viscous fluid contents confound imaging studies and often make definite diagnosis difficult. On ultrasonography, the most consistent findings in CHFCs are a well-delineated anechoic or slightly hypoechoic unilocular cyst. An enhanced CT usually displays these lesions as hypo-attenuating as in our case or isoattenuating relative to surrounding liver parenchyma. On magnetic resonance

imaging, all CHFCs are hyperintense on T2-weighted image, although they may be hypodense, isodense, or hyperintense according to the various cyst contents.[4]

If the imaging characteristic remains doubtful, further diagnostic study may be done using fine needle aspiration cytology (FNAC). The characteristic features of CHFCs on FNAC are the cellular smear with scattered benign-appearing ciliated columnar cells arranged in both tissue fragments and singly in a mucoid background. Other features supporting the diagnosis include the presence of fragment of smooth muscle, goblet cells, and macrophages in the cell population.[5] However, the absence of these features does not completely rule out the diagnosis, because in near one-quarter of adult cases, there is inadequacy of cellular material on aspiration.[1] Moreover, FNAC cannot differentiate CHFC from primary bronchogenic cyst of the liver because both possess a common lining epithelium. Histologically, the differential diagnosis of a hepatic cyst included simple cyst, hydatid cyst, biliary cystadenoma, cystadenocarcinoma, and bronchogenic cyst. The key histological features of CHFC are the presence of a four-layered cyst wall, which consists of a mostly inner lining of pseudostratified columnar epithelium followed by a layer of loose subepithelial connective tissue, smooth muscle layer, and an outer layer of dense fibrous tissue.[1-3] Presence of these unique histological features indicates CHFC with a reasonable degree of confidence, although a rare morphologic variant of CHFC without smooth muscle layer has reported.[6] The main differential diagnosis can be bronchogenic cyst because both cystic lesions share a common embryonic origin.[2] The presence of cartilage and respiratory glands in addition to ciliated columnar epithelium in the wall of the cyst defines the bronchogenic cyst in surgical specimens. Presence of malignant transformation in a few CHFC patients and its fatal clinical course emphasizes the need for total surgical resection. The subcapsular location and the presence of thick outer fibrous layer in the cyst wall makes it amenable to laparoscopic excision with minimal morbidity.[1,7]

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