CASE REPORT

Laparoscopic Excision of a Ciliated Hepatic Foregut Cyst

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ABSTRACT

Ciliated hepatic foregut cysts are uncommon solitary cysts of the liver that originate from the embryologic foregut. Clinically and radiographically, these lesions can be difficult to distinguish from neoplasms. Recent reports have demonstrated that ciliated hepatic foregut cysts may undergo dysplastic progression, supporting the argument to excise these cysts when discovered. Fewer than 100 cases have been described in the literature since the first description of a ciliated hepatic foregut cyst in 1857. We present a patient who recently underwent laparoscopic excision of a ciliated hepatic foregut cyst, review the literature, and propose the rationale for attempting removal of these cysts via a laparoscopic approach.

Key Words: Hepatic cyst, Foregut cyst, Ciliated hepatic foregut cyst.

INTRODUCTION

The ciliated hepatic foregut cyst (CHFC) is a rare, solitary, usually benign cyst recognized by its ciliated pseudostratified columnar epithelial layer. Given the distinct epithelial appearance and the histologic similarity to bronchial and esophageal cysts, the CHFC is generally considered to arise from a remnant of the embryologic foregut. Friedreich originally described CHFC in 1857, but the term was not coined until 1984 by Wheeler and Edmondson.[1,2] In the last 150 years, approximately 100 cases have been reported in the literature worldwide, only 4 of which describe laparoscopic excision.[3–7] We report the case of a patient who recently underwent successful laparoscopic excision of a CHFC and discuss the typical findings associated with this cyst as well as the rationale for attempting laparoscopic removal of these lesions.

CASE REPORT

A 55-year-old male with a past medical history significant for coronary artery disease, hyperlipidemia, well-controlled gastroesophageal reflux, and a remote right nephrectomy for trauma presented with an asymptomatic gallbladder mass that had increased in size over the last year on serial abdominal ultrasonography. During a workup for abnormal liver function tests, a rounded hyperechoic mass appeared to be within the fundus of the gallbladder, with a year's interval increase from 1.5x1.4 cm to 1.8x1.7 cm **(Figure 1)**. No evidence was present of cholelithiasis or biliary ductal dilatation. Subsequent contrast-enhanced CT revealed an ovoid lesion measuring 2.5x1.7 cm in the region of the gallbladder fundus. The mass did not enhance postcontrast and appeared to be well circumscribed without evidence of hepatic invasion. The patient's physical examination was unremarkable. Laboratory analysis, including repeat liver function tests, was normal. Given the appearance on CT, a solid tumor could not be ruled out. Therefore, a laparoscopic cholecystectomy with en-bloc excision of the mass was planned. In addition, the patient was consented for possible segment IVb-V liver resection should intraoperative frozen section demonstrate malignant changes in the mass.

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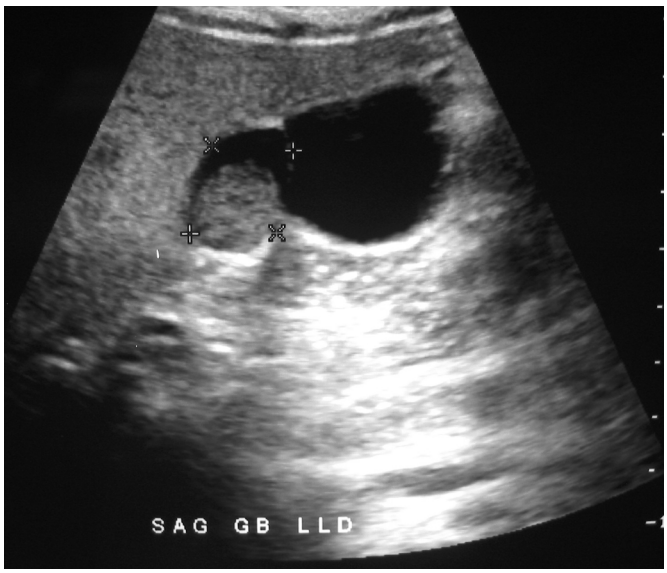

Figure 1. Right upper quadrant US demonstrating the hyperechoic lesion in the region of the gallbladder fundus.

At the time of surgery, general anesthesia was induced, and the patient was placed in a supine position. The operation was undertaken as in routine laparoscopic cholecystectomy. Port placement was initiated with a 10-mm infraumbilical trocar followed by two 5-mm ports inferior to the right costal margin along the anterior and midaxillary lines. Adhesions between the anterior abdominal wall and the liver were taken down sharply so that the gallbladder hilum could be visualized. A 10-mm VersaStep port was then placed in the epigastrium. Upon lateral and cephalad retraction of the gallbladder, a well-encapsulated mass was identified in the gallbladder fossa **(Figure 2)**. Cholecystectomy was undertaken with identification of hilar structures followed by ligation and transection of the cystic duct and artery. To isolate the mass, the gallbladder was dissected from the liver bed by using a dome-down technique. No apparent vessels or bile ducts directly communicated with the mass from within the hepatic parenchyma. The mass was then circumferentially dissected from the liver bed, taking a thin margin of subcapsular tissue en-bloc with the specimen **(Figure 3)**. Frozen section demonstrated a benign-appearing cystic mass. The operation was then completed in standard fashion with hemostasis, irrigation, and port-site closure. The patient was awakened and recovered in the postanesthesia care unit. He was informed of the findings and extent of operation and was discharged the same day. To date, the patient has done well without postoperative complications or recurrence of the mass.

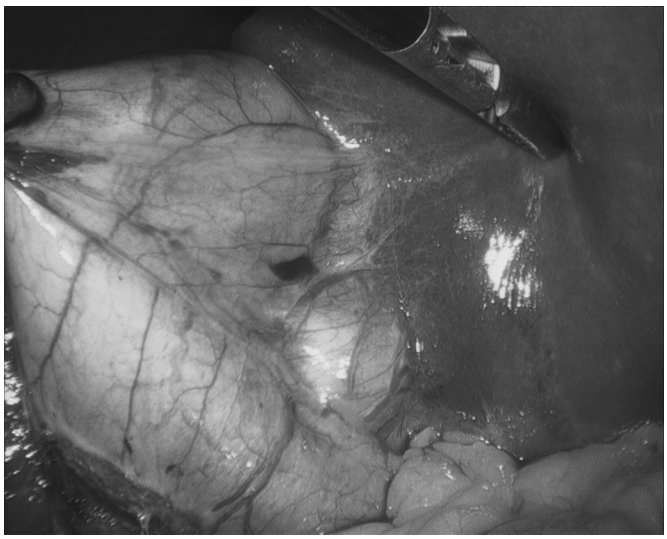

Figure 2. Laparoscopic view of the CHFC *in situ* in the gallbladder fossa, prior to dissection.

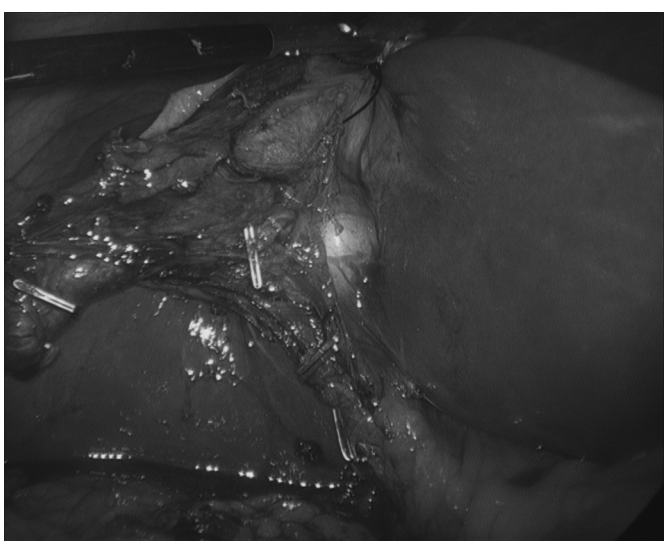

Figure 3. Laparoscopic view of CHFC between the gallbladder fossa and hepatic parenchyma, following ligation and transection of the cystic duct and artery.

On gross examination of the specimen, the gallbladder was intact without gallstones or inflammatory changes. An adjoining unilocular 2x0.5-cm cyst containing thick white mucoid fluid had no communication with the lumen or wall of the gallbladder. Histological analysis demonstrated an inner layer of ciliated stratified columnar epithelium with scattered goblet cells surrounded by loose connective tissue, a discontinuous smooth muscle layer, and an outer fibrous capsule **(Figure 4)**. These findings were consistent with a ciliated foregut cyst of hepatic origin

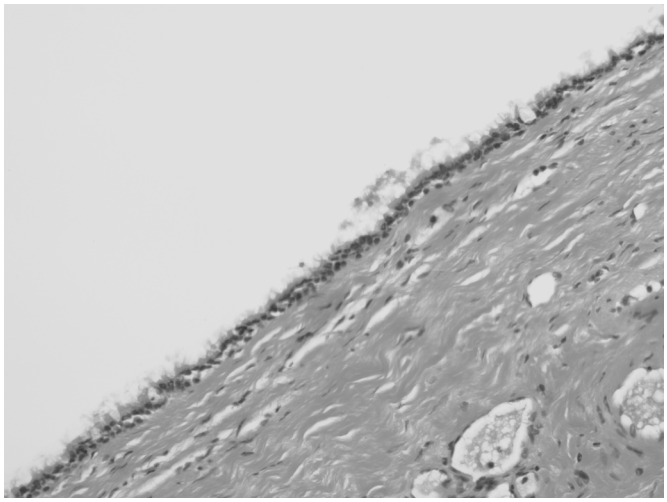

Figure 4. Photomicrograph of the ciliated hepatic foregut cyst wall, demonstrating the characteristic pseudostratified ciliated epithelium, subepithelial connective tissue, smooth muscle layer, and outer fibrous capsule with hematoxylin-eosin staining, 200X magnification.

without evidence of epithelial dysplasia or invasive malignancy.

DISCUSSION

The ciliated hepatic foregut cyst is a rare but increasingly reported solitary cyst of the liver. There appears to be an historical bimodal distribution of CHFC cases in the literature from the late 19th century and again in the last 20 years. Vick et al[8] suggested that the former cluster of reports could be associated with a time period during which autopsies were more routinely performed; the more recent peak is likely driven by increased detection from the more frequent use of radiologic imaging modalities. In addition, a preponderance of publications can be found in the Japanese literature since 1988. These cases are likely the result of more aggressive reporting rather than a true increased prevalence in this population.[3,8–10]

It is difficult to determine the true prevalence of CHFCs because they are generally asymptomatic. Instead, CHFCs commonly present after incidental discovery on routine radiologic imaging, during surgical exploration, or at autopsy. An approximation of CHFC prevalence can be derived from a study by Sanfelippo's group of 88,000 abdominal operations at the Mayo Clinic in which the incidence of solitary cystic hepatic lesions was 0.09%.[8,11] CHFCs may be more common than previously reported, if labeled only as a benign simple hepatic cyst.

The most common current presentation of a CHFC is as an asymptomatic lesion found incidentally on radiologic imaging. The most common symptoms include vague right upper quadrant pain, nausea, or vomiting.[6] Discovery of CHFCs has also been made in the setting of viral hepatitis or abnormal liver function tests. In addition, there are isolated reports of portal hypertension and splenomegaly caused by portal vein compression, a case involving the hepatic vein, obstructive jaundice from external compression of the common hepatic duct, and cyst communication with the gallbladder.[12–14] In one review, 17% of CHFCs were actually found during imaging workup of extrahepatic malignancies. Typically, the mean age at diagnosis is 50 years, ranging from 3 months to 82 years, with a slight male predominance.[8,15]

CHFCs are most often located in the left lobe of the liver with a strong predilection segment IVb. They are unilocular and avascular, usually lying just beneath the anterior surface of the liver in a subcapsular position.[8,9] Several cases have been reported in the right lobe of the liver as well as in the gallbladder wall.[15] The characteristic position of the lesion can potentially be explained by embryologic histogenesis. Freidreich[1] initially suggested a congenital origin, but currently it is suspected that the CHFC is a detached hepatic diverticulum or abnormal tracheobronchiolar bud that may have migrated caudally to be included with the liver during the early embryological development of the foregut.[15,16] The foregut differentiates into the oropharynx, esophagus, stomach, duodenum, liver, gallbladder, pancreas, tracheobronchial tree, and lungs through a series of diverticuli and budding. The liver appears in the fourth week of embryological development, with a dominant left lobe until the sixth to eighth week when the liver remodels with the growth of the right lobe. This differential lobar growth could explain the preferential localization of the CHFCs in the left lobe of the liver. Prior to the closure of the pleuroperitoneal canals, an abnormal tracheobronchiolar or esophageal bud could migrate caudally to be included in the hepatic diverticulum and mature into a CHFC while retaining many of the features of a bronchogenic or esophageal cyst. Bronchogenic cysts can be distinguished by the presence of mural cartilage; esophageal cysts can be distinguished by the presence of 2 distinct smooth muscle layers and ciliated or squamous epithelium.[10] Intestinal duplication cysts can also have a ciliated epithelial lining, but in contrast with CHFC, duplication cysts remain in continuity with the intestinal tract through its muscular layers or serosa.[15]

Ciliated hepatic foregut cysts grow to a mean diameter of 3 cm, with size ranging from 1 cm to 12 cm.[6,8,9] The cyst is characterized by its 4 layers: a ciliated pseudostratified columnar epithelium with mucin-containing goblet cells, loose subepithelial connective tissue, a smooth muscle layer, and fibrous outer capsule.[3,8,17] The ciliated epithelium is the hallmark of the CHFC. Neuroendocrine and mucus-secreting goblet cells within the epithelial layer are reminiscent of bronchial origin. Mucoid material contained within the cyst varies from clear serous fluid to milky white or brown mucoid material with abundant protein, lipids, and cholesterol, or calcium deposits.[18] Several pathologic reviews have also demonstrated small bile ductules within the cyst wall.[15]

As in this case, the diagnosis of CHFC is often confounded by the clinical and radiographic difficulty in distinguishing a benign CHFC from a neoplastic process. The variability in appearance on radiographic imaging is often attributed to the elements of the cyst contents. Ultrasonography usually shows an anechoic to slightly hypoechoic lesion that may contain spotty hyperechoic areas.[19–21] This case was notable for the homogeneous hyperechoic appearance of the cyst. On CT imaging, CHFCs are often hypodense compared with the surrounding hepatic parenchyma, but may be isodense or hyperdense. These cysts are uniformly unenhancing on contrast-enhanced CT. Although almost all cysts display a high-intensity signal on T2-weighted MRI, T1-weighted imaging has no characteristic signal intensity. The most common combination of imaging features is a unilocular hypoechoic mass on ultrasound that is hypodense on CT without septations or vascular enhancement. Approximately one third of CHFC cases have a solid tumor appearance on CT and noncystic characteristics on ultrasound.[9]

The differential diagnosis is often derived from the variable radiographic features but can be confirmed histologically. Commonly considered diagnoses include other unilocular hepatic lesions, such as simple hepatic cyst, parasitic (echinococcal) cyst, epidermoid (or endometrial) cyst, pyogenic abscess, intrahepatic choledochal cyst, mesenchymal hamartoma, hypovascular solid tumor, and hepatobiliary cystadenoma or cystadenocarcinoma.[3,20] Simple cysts are usually located in the right hepatic lobe and are more prevalent in women. Serologic tumor markers can be confounding, as elevated CA19–9 levels have been found in association with benign CHFC.[3,6] Evaluation of the cyst contents and lining by fine needle aspiration has been reported in 8 cases, and aids in distinguishing among many of the considered lesions, which are easily differentiated by their typical epithelial lining, cyst contents, surrounding stroma or parenchymal response, and immunohistochemical profiles.[3,17] An aspirate of hepatocytes admixed with ciliated cells and mucus cells distinguishes CHFC from other radiologically benign-appearing entities.[8,9,22]

Ciliated hepatic foregut cysts are generally considered benign non-neoplastic processes. However in the last decade, 3 cases of squamous cell carcinoma and 1 case of extensive squamous metaplasia arising from a CHFC have been reported.[4,18,23,24] Squamous metaplasia could be explained by the retention of matured bronchial or esophageal squamous epithelium from embryological development. The presence of dysplastic foci associated with squamous cell carcinoma could suggest progression from nondysplastic epithelium to dysplasia and carcinoma. Despite the ability of fine-needle aspiration biopsy to differentiate lesions based on epithelial appearance or cyst contents, sampling error may lead to an errant diagnosis of benign CHFC in the face of missed malignant transformation. To date, the major risk factor associated with diagnosing a CHFC with malignant transformation is size. All 3 cases of carcinoma had cystic masses >12 cm in diameter.[6]

The recommended treatment of CHFC described in the literature has varied from observation to aspiration to surgical excision. Given the difficulty of reliably diagnosing a CHFC on radiographic imaging, many CHFCs that are excised are not diagnosed until postoperative evaluation, as in our case. Imaging that suggests a lesion with features suspicious for malignancy, such as septations or focal wall thickening, can be preoperatively aspirated for cytological diagnosis. Several authors have also noted that aspiration and injection of a sclerosing agent can be utilized effectively to stop cyst growth and associated symptoms.[3,17] However, given the recent cases of CHFC malignant transformation, serial imaging may be required for those patients undergoing cyst sclerosis. Most authors agree that CHFCs should be surgically excised for cysts larger than 4 cm to 5 cm, symptomatic or enlarging lesions, asymptomatic lesions with wall abnormalities on imaging, or in patients with otherwise unexplained abnormal liver function tests.[5,6] To date, there has been no report of a cyst recurrence or metastasis after excision.[18]

Laparoscopic surgery is an ideal modality for benign or marginally dysplastic disease. Uncomplicated CHFCs are quite amenable to laparoscopic excision for several reasons. First, the small size and anterior subcapsular location allow for easy access to the cyst with minimal dissection to isolate the lesion. Second, the benign nature of the process allows

the cyst to be enucleated from is hepatic bed with little concern for adequate circumferential margins. Third, the CHFC traditionally has a thick cyst wall that permits easy handling with laparoscopic instruments.

The described case is a good representation of a typical presentation of an atypical lesion. Our patient was in his 50s and had an asymptomatic presentation with an incidental finding on right upper quadrant radiographic imaging. It should be noted that although the size and location of the cystic mass were characteristic of CHFC, this patient's cyst demonstrated imaging features that are less common for a CHFC, leading to the decision to excise the mass. By utilizing a laparoscopic approach to excise the CHFC, the patient benefited from an effective ambulatory procedure for the removal of a lesion with risk for malignant transformation. CHFC should be considered as a diagnosis for a superficially located unilocular left hepatic cystic mass that presents incidentally or asymptomatically, and laparoscopic excision should be considered as a first-line treatment to remove this benign cystic mass.

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