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Ciliated Hepatic Foregut Cyst Presenting With Cirrhosis of Liver: Case Report of a Rare Entity

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Abstract

Ciliated hepatic foregut cysts are uncommon developmental abnormalities, often presenting as incidental finding. We report a case of 27 year old female who presented with a large ciliated hepatic foregut cyst, and subsequent obstructive symptoms, leading to biliary type liver cirrhosis. A partial hepatectomy specimen was received showing a 55 x 40mm cyst attached to the inferior aspect of left lobe. Histopathology showed a cyst lined by pseudostratified ciliated columnar epithelium with discontinuous smooth muscle in the wall and fibrosis. Background liver showed cirrhosis.

Keywords: Ciliated hepatic foregut cyst; Liver cirrhosis; Partial hepatectomy

Introduction

Ciliated hepatic foregut cysts are, to date, considered a rare cystic developmental abnormality, arising from the foregut. Its name was introduced in literature by Edmondson and Wheeler, in their iconic paper titled "Ciliated Hepatic Foregut Cyst [1], which described a solitary cyst of liver with pseudostratified ciliated columnar epithelium. They postulated that the esophageal and bronchial parts of the embryonic foregut can get separated from the main body, and become subsequently entrapped in the concurrent developing liver, explaining the intra-abdominal location of this cyst. One of the peculiar clinical characteristics of this cyst is that it frequently presents in adulthood, in spite of its embryonic origin [2]. While they are frequently small and are incidental findings during surgery [3], they can enlarge throughout life and present with obstructive symptoms [4], similar to our case. Ciliated Hepatic foregut cysts are now increasingly diagnosed, with over 100 cases reported at present [5]. The vast majority behave in a benign fashion with only a few showing squamous metaplasia, and even fewer progressing to squamous cell carcinoma as an aftermath of metaplasia [6]. Here we present a case of ciliated hepatic foregut cyst in a 27 year old female with obstructive symptoms, and subsequent development of liver cirrhosis.

Case Report

A 27 year old female presented to the Outpatient of Hepatobiliary Pancreatic Department with obstructive jaundice in January 2025. She had a previous history of laparoscopic cholecystectomy, converted to open surgery, for Mirrizi syndrome, and had undergone choledochoduodenostomy 5 months ago. Her Liver function tests showed raised direct bilirubinemia (9.11mg/dl; laboratory reference values 0.02 to 0.3mg/dl), GGT (676 U/L; laboratory reference values <40 U/L), AST (70.6U/L; laboratory reference values 0-32U/L), ALT (52.6U/L; laboratory reference values 0-35U/L), and ALP (813U/L; laboratory reference values 35-104 U/L). Total bilirubin was 10.1 mg/dl against laboratory reference values of up to 0.9mg/dl. Her CECT scan abdomen with tri-phasic protocol reported a benign looking cystic lesion in left lateral segment measuring 5 x 4cm and dilated intra and extrahepatic biliary channels with narrowed choledochoduodenostomy site. MRCP reported a multilobulated intrahepatic cyst in left lobe of liver as shown in Figure 1. Mild dilatation was reported in intrahepatic biliary tree; extrahepatic biliary tree was normal. Intraoperatively, a definitive communication with the underlying biliary tree was not identified.

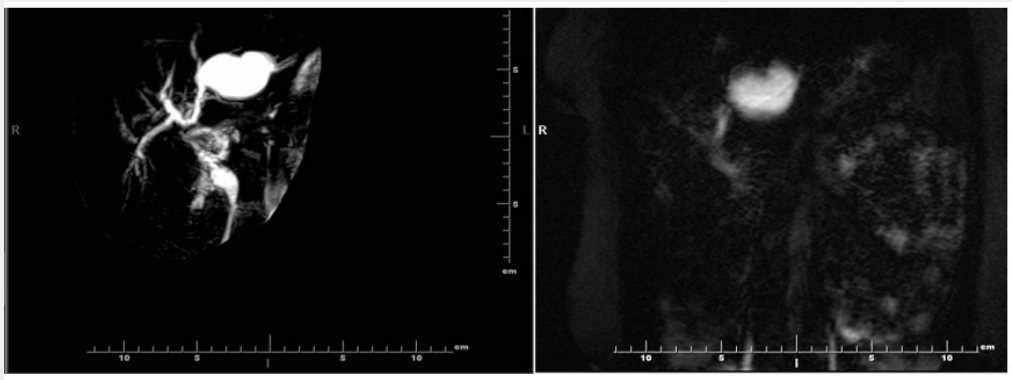

Figure 1: MRI Cholangiopancreaticography showing multi-lobulated inter-hepatic cyst, left lobe of liver.

Figure 2: Gross showed cyst with demarcation from liver tissue with a smooth inner lining and thin wall.

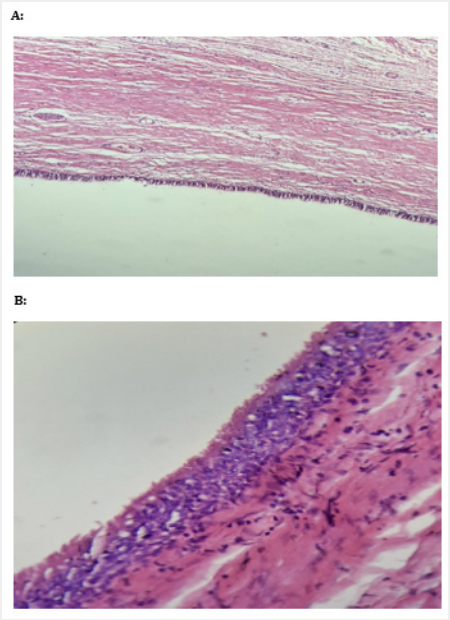

Figure 3 (A & B): Showing ciliated pseudostratified lining of the cyst wall, with conspicuous cilia are seen. The underlying tissue shows fibroconnective tissue, a discontinuous layer of smooth muscles and fibrous tissue at periphery by H&E staining at 400X.

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Grossly, as shown in Figure 2, a partial hepatectomy specimen was received, with an attached cyst measuring 55 x 40 x 35mm. Liver capsule showed cirrhotic nodules. Cyst, on opening was filled with mucoid fluid and was multiloculated, with maximum wall thickness of 1mm. There was no connection with underlying biliary channels, and the interface between liver and cyst could grossly be defined. We postulate that the dilatation of intrahepatic biliary tree would have been the result of mechanical obstruction caused by the cyst. Histopathology (Figure 3a & 3b) showed a cyst with well-developed pseudostratified columnar lining, and very focal cuboidal/low columnar metaplasia (Figure 4). The wall showed areas of fibrosis and a discontinuous smooth muscle layer, highlighted by SMA. Scattered inflammatory cells were also seen. Background liver parenchyma (Figure 5) showed "garland" type of cirrhotic nodules, in keeping with obstruction-associated cirrhosis.

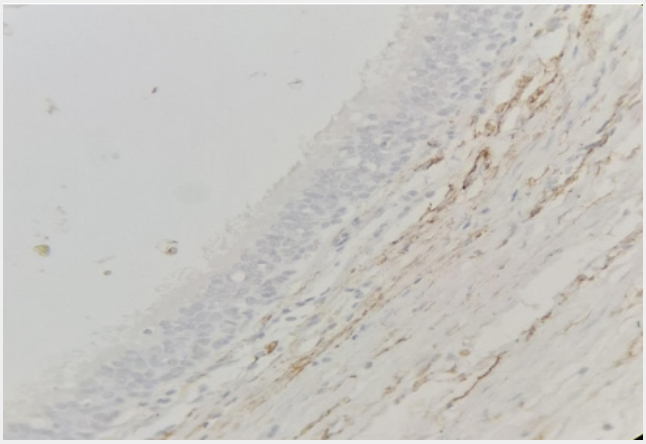

Figure 4: Showing smooth muscle actin stain, highlighting discontinuous smooth muscle cells in the wall, 400X magnification.

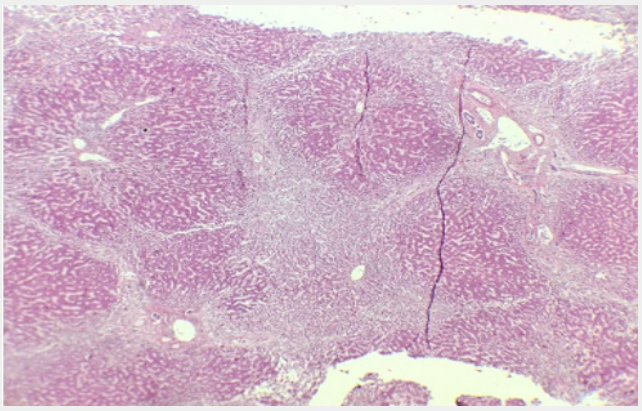

Figure 5: Showing background liver tissue and nodule formation with bridging cirrhotic bands (Batts Ludwig stage 4), 2X magnification.

Discussion

Ciliated hepatic foregut cysts are too rare to have established epidemiological data [7], but this may also be attributed to the fact that these are usually incidental findings, and may be present undiagnosed in a larger portion of population than we know. Ciliated hepatic foregut cysts are postulated to arise from orphaned parts of embryonic foregut that become entrapped in liver during caudal migration. Ciliated hepatic foregut cysts have a higher prevalence in men, and usually involve medial segment of left hepatic lobe [8]. They are generally small, averaging around 3cm, with a few reaching up to 12cm [9]. In our case report, the cyst was 55mm (5.5cm) in the largest dimension. Another variation seen in our case was the multiloculation of the cyst; these cysts are often reported to be unilocular [10]. Uniloculation, to the best of our knowledge, is not an absolute diagnostic requirement. Ciliated Hepatic Foregut cysts are recognized histologically by 4 layers. From inside to outside as shown in Figure 3 & 4, these are as follows: a pseudostratified ciliated columnar lining; sub-epithelial connective tissue; smooth muscle layer; and outer fibrous/fibro collagenous capsule.

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Because of their distinct microscopic appearance, ciliated hepatic foregut cysts pose little diagnostic challenge with almost nonexistent differential diagnosis. Diagnosis may be suggested by radiological findings, but microscopy remains the gold standard. It is also worthwhile to note that there is no specific serological marker for ciliated hepatic foregut cyst. Ciliated hepatic foregut cysts are reported as benign cysts. It is, however, imperative to note that they have a small, albeit a very real, risk of malignant transformation [11]. Squamous cell carcinoma may arise following a metaplasia-dysplasia pathogenesis pathway [12]. For this reason, cysts are removed surgically. Current literature review does not support long term follow up for these patients, since there are no reported cases of recurrence after excision [13].

Conclusion

Ciliated hepatic foregut cysts are uncommon, but should be considered in the differential diagnoses of hepatic cysts. They can undergo malignant transformation to squamous cell carcinoma. Surgical excision is the treatment of choice.

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