Case Report

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Ciliated Foregut Cyst and Accessory Spleen in the Pancreas: A Case Report and Literature Review

췌장에서 발생한 부비장과 동반된 섬모성 전장낭: 증례 보고와 문헌고찰

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Ciliated foregut cyst is a relatively rare disease; thus, most reports are in the form of case studies. This benign cyst is usually found in the mediastinum and account for approximately 20% of all mediastinal masses. However, it is rarely found in the hepatobiliary and peripancreatic regions. Approximately 20 cases of ciliated foregut cysts involving the pancreas have been reported in the Enlgish literature. Here, we present a case of ciliated foregut cyst that occurred in the tail of the pancreas in a 29-year-old female. The patient's ultrasonography, CT, and MRI findings are presented, along with a review of the literature.

Index terms Pancreas; Cyst; Magnetic Resonance Imaging; Spleen

INTRODUCTION

Ciliated foregut cyst was first reported by Friedreich in 1857 (1), and then changed to its present name by Wheeler and Edmondson in 1984 (2).

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Ciliated foregut cysts are benign congenital cystic lesions, which probably result from abnormal development of the primitive foregut. Their occurrence in the hepatobiliary system can be explained by the fact that the tracheobronchial tree, esophagus, liver, and pancreas arise from the primitive foregut during fetal development. The tracheobronchial tree arises from the ventral foregut, whereas the esophagus, stomach, liver, and pancreas arise from the dorsal foregut. Foregut cysts arising in the liver and pancreas purportedly originate from a detached outpouching of the primitive foregut and are then sequestered by the liver and pancreas during embryonic development (3).

With the widespread use of diagnostic imaging, including CT and MRI, incidentally identified pancreatic cystic lesions are increasing. Pseudocysts are the most common non-neoplastic pancreatic cysts and occur secondary to trauma or pancreatitis. Other types of non-neoplastic cysts, including retention, congenital epithelial, lymphoepithelial, and enterogenous cysts, are rare and always benign (4).

Ciliated foregut cysts are rarely found in the pancreas, and their radiological findings are worth reporting. Herein, we present the case of a 29-year-old female with an incidentally noted cystic mass in the pancreatic tail.

CASE REPORT

A 29-year-old female was referred to our institution for the evaluation of an incidentally detected pancreatic lesion. The lesion was detected when the patient underwent an abdominal CT scan for the evaluation of lower abdominal pain and surgery for acute appendicitis. She had no clinical symptoms at the time of her visit, and no history of chronic alcoholism, acute or chronic pancreatitis, biliary lithiasis, or abdominal trauma. However, the patient had a history of type 1 diabetes mellitus. Except for an elevated carbohydrate antigen (CA) 19-9 level of 441 U/mL (normal range: 0–27 U/mL), other laboratory results, including pancreatic enzyme and liver function tests, were within the normal range.

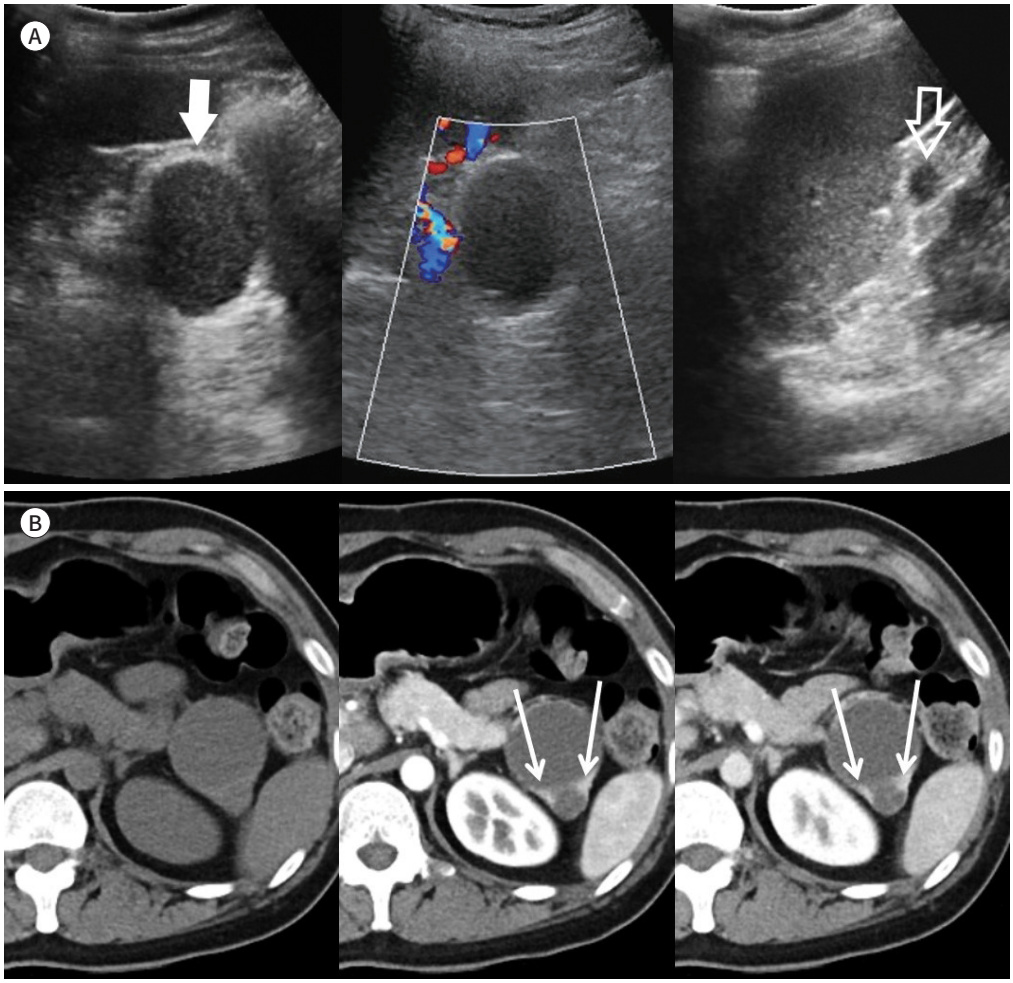
On abdominal ultrasonography, the lesion appeared as an approximately 4.1 cm sized, thick-walled, anechoic cystic lesion with internal echogenic debris and a 1.6 cm multiseptated cystic lesion in the tail portion of the pancreas (Fig. 1A). Vascular flow was not observed in any of the lesions.

The CT and MRI revealed a well-defined 4.6 cm unilocular cystic lesion and a 1.5 cm multiseptated cystic lesion in the tail portion of the pancreas. A small solid component was located between the two cysts. On pre-contrast imaging, the two cystic lesions showed lower attenuation than the normal pancreatic parenchyma. Contrast-enhanced CT images showed a small, homogenously enhanced solid component between the two cystic lesions (Fig. 1B). No calcifications were observed within the cysts.

On the MRI scan using a 3T imaging system (MAGNETOM Vida, Siemens Healthineers, Erlangen, Germany), the cystic component was hypointense on T1-weighted image (T1WI) and marked homogenous hyperintense mass with thin hypointense wall on T2-weighted image (T2WI).

The solid component was isointense on T1WI and slightly hyperintense on T2WI, relative to normal pancreatic parenchyma. On a dynamic contrast-enhanced scan using gadoxetic acid, the cystic component reveal mild peripheral rim like wall enhancement and the solid

Fig. 1. A 29-year-old female with ciliated foregut cyst and accessory spleen in the pancreas.
A. Abdominal US images show a 4.1-cm thick-walled, anechoic cystic lesion (thick arrow) with internal echogenic debris without intratumoral vascularity and a 1.6-cm multiseptated cystic lesion in the tail region of the the pancreas (empty arrow).
B. Axial dynamic CT pre-contrast (left), arterial (middle), and portal venous (right) phase images show a 4.6-cm unilocular cystic lesion and a 1.5-cm multiseptated cystic lesion in the tail portion of pancreas. On the pre-contrast image, two cystic lesions show lower attenuation than that of the normal pancreatic parenchyma. Contrast-enhanced CT images show small, homogeneously enhancing, solid components (arrows) between two cystic lesions.



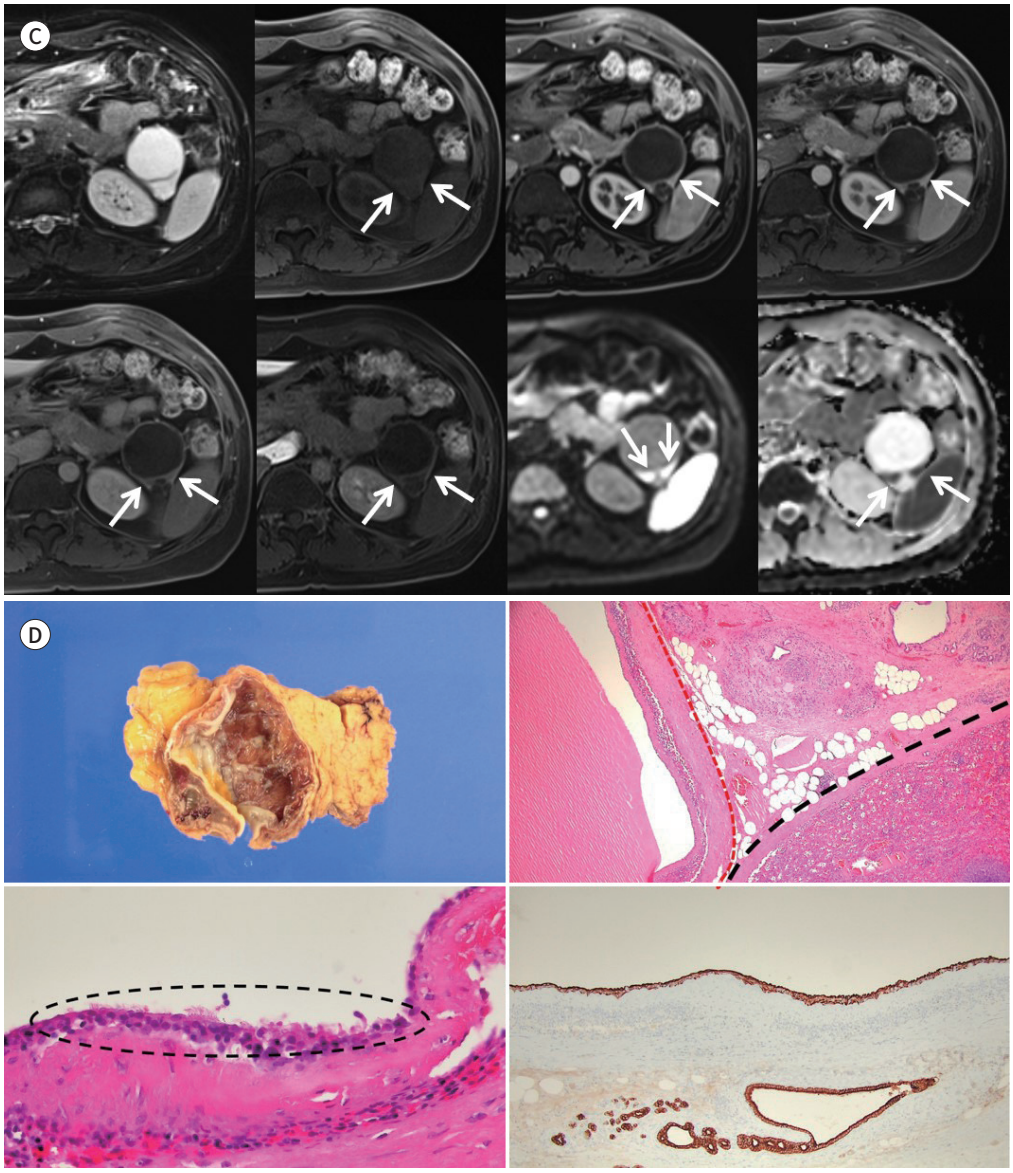
component was homogeneously well-enhanced. The solid component showed hyperintensity on diffusion-weighted imaging (DWI) at b = 800 s/mm^2 and a low apparent diffusion coefficient map (Fig. 1C).

Our initial differential diagnoses included pancreatic cystic tumors with solid components, such as neuroendocrine tumor, or solid pseudopapillary neoplasm with cystic degeneration and pancreatic cystic neoplasms, such as mucinous cystic neoplasm (MCN) or serous cystadenoma.

The patient underwent a robotic distal pancreatectomy. The final histopathological diagnosis was ciliated foregut cyst of the pancreas with an intrapancreatic accessory spleen. Grossly, the resected specimen showed two intrapancreatic cystic masses measuring 4.1 cm × 3.5 cm and 1.6 cm × 1.1 cm, respectively and the two cysts are communicated by adhesion. Micro-

scopic findings revealed relatively the connected cysts along with mildly inflamed pancreas and accessory splenic tissue. The cysts were lined with a pseudostratified ciliated columnar epithelium, thin subepithelial connective tissue and fibrinous capsule. No cytological atypia

Fig. 1. A 29-year-old female with ciliated foregut cyst and accessory spleen in the pancreas (Continued).
C. Axial MRI scan T2-weighted image (1st), pre-contrast T1-weighted image (2nd), arterial phase (3rd), portal venous phase (4th), 3-minute transitional phase (5th), hepatobiliary phase images (6th). The cystic component shows a bright high signal on T2-weighted image and hypointensity on pre-contrast T1-weighted image. On dynamic contrast enhanced scan using gadoxetic acid, solid components (arrows) show homogeneous enhancement. Diffusion-weighted image (7th) and apparent diffusion coefficient map (8th) reveal diffusion restriction of the solid component and the cystic components are communicated.
D. Grossly, the resected specimen shows two relatively well-circumscribed cystic masses (left upper). The specimen consists of well-demarcated cysts (left side of red dot), inflamed pancreas (center), and an intrapancreatic accessory spleen (right side of black dot) (H&E stain, × 40) (right upper). The cysts are lined by ciliated columnar epithelium (black dot), loose connective tissue and fibrous capsule. No cytologic atypia is observed (H&E stain, × 400) (left lower). The lining epithelial cells were positive for cytokeratin 7 immunohistochemical stain (× 100) (right lower).
H&E = hematoxylin and eosin



or goblet cells were observed. On immunohistochemistry, the lining epithelial cells and pancreatic ductal cells were positive for cytokeratin 7 (CK 7), but negative for D2-40 and anti-mesothelial antibodies (Fig. 1D). Ten days later, the patient was discharged without any complications. Postoperatively, serum CA 19-9 level decreased to 62.2 U/mL after 3 months.

This case report was approved by our Institutional Review Board, which waived the requirement for written informed consent (IRB No. DAUHIRB-22-116).

DISCUSSION

Ciliated foregut cysts are lined by a pseudostratified ciliated epithelium and may contain goblet cells. Epithelial cells are immunoreactive for CK 7, thyroid transcription factor-1, and CA 19-9. In addition to ciliated epithelium, the presence of respiratory glands or cartilage defines a bronchogenic cyst. Similarly, a duplication cyst contains a gastrointestinal mucosal lining, and the presence of two muscle layers indicates esophageal differentiation. A cyst with ciliated columnar epithelium without other additional features, as in our case, is properly defined as a "ciliated foregut cyst" (3). Each type of foregut cyst had similar CT findings with characteristic locations. On CT, all three types of cysts appeared as solitary, well-defined, and unilocular cystic masses with fluid attenuation.

To date, approximately 20 cases of ciliated foregut cysts involving the pancreas have been reported (5). However, Gómez Mateo Mdel et al. (6) mentioned that upon review of the proposed classification mechanisms, only some cases could be classified as true ciliated foregut cysts, making this entity rarer. To date, only seven cases of true ciliated foregut cysts in the pancreas have been reported (3, 5-10). To our knowledge, there was no case report about coexistence of ciliated foregut cyst and accessory spleen in the pancreas. Most patients presented with abdominal pain; however, some were asymptomatic. They are mostly benign, and there have been no reports of recurrence following surgical removal. Unlike in our case, the CA 19-9 level was within the normal range in all cases.

Imaging findings of ciliated foregut cysts are mostly nondiagnostic. The reported pancreatic ciliated foregut cysts were small or up to 10 cm in size on imaging, and were located at the head, body, or tail of the pancreas. The cysts are generally solitary, well-defined, unilocular, or multilocular cystic masses. The cystic component usually appears anechoic with or without layering debris on ultrasonography and hypodense on non-enhanced CT.

In a case reported by Alessandrino et al. (7), MR cholangiopancreatography revealed a 2 cm, well-defined, cystic lesion in the head of the pancreas, which was mildly hyperintense with layering debris on T2WI, slightly hypointense on T1WI, not enhanced on the post-contrast images, and hyperintense on DWI. In that case, communication with the main pancreatic duct was noted, and the preoperative diagnosis was side-branch intraductal papillary mucinous neoplasm.

When we retrospectively reviewed our case, we found two cystic masses in the tail portion of the pancreas, and a solid component was located between them. Postoperative microscopic findings revealed relatively well-demarcated cysts with accessory splenic tissues. On contrast-enhanced CT and MRI, the solid component showed the same density and intensity as that of the spleen. The accessory splenic tissue shows hyperintensity on high-b-value DWI because

the spleen shows the most restricted diffusion among the upper abdominal organs. This explains the diffusion restriction by the solid component in our case. Unlike in previously reported cases, the cyst wall was relatively thick in our case and had fibrous capsule on histologically.

In the reviewed articles, ciliated foregut cysts of the pancreas were often diagnosed as MCN on imaging (3, 10). Based on imaging alone, pancreatic ciliated foregut cysts are difficult to differentiate from other cystic pancreatic lesions, such as MCN and pseudocysts. Hence, a pathological diagnosis is considered accurate and is obtained only upon definite histological evaluation of the surgical specimen. In our case, the presence of pseudostratified ciliated epithelium along the cyst wall was suggestive of a ciliated foregut cyst.

In conclusion, while imaging findings of ciliated foregut cysts of the pancreas are non-specific, when well-defined unilocular or multiseptated cystic masses of the pancreas occur, ciliated foregut cysts should be included in the differential diagnosis.

Author Contributions

Conceptualization, K.H.J., K.H., C.J., P.M.G.; data curation, K.H.J., K.H., L.K., C.J., P.M.G.; investigation, K.H.J., K.K.W.; methodology, L.K.; resources, K.H., P.M.G.; supervision, K.H., L.K., C.J.; validation, K.H.; visualization, K.H.J.; writing—original draft, K.H.J.; and writing—review & editing, K.H.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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췌장에서 발생한 부비장과 동반된 섬모성 전장낭: 증례 보고와 문헌고찰

김현진[1] · 권희진[1*] · 임경재[1] · 조진한[1] · 박민경[2] · 김관우[3]

섬모성 전장낭(ciliated foregut cut)은 드문 종양으로 주로 증례 보고 형태로 보고되었다. 이 양성 낭종은 대개 종격동에서 발견되어 전체 종격동 종양의 20%를 차지하지만 간 담도계와 췌장 주변 조직에서는 매우 드물다. 현재까지 췌장의 전장낭에 대한 영문 보고는 전 세계적으로 20건 정도이다. 저자들은 29세 여성에서 췌장 미부에 발생한 섬모성 전장낭의 사례를 초음파, 컴퓨터단층촬영, 자기공명영상 소견들과 함께 제시하고 문헌을 검토하고자 한다.

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