Clinical Case Report

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A five year CT surveillance of ciliated retroperitoneal foregut cyst resembling a cystic pancreatic lesion

A case report

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Abstract

Introduction: Ciliated retroperitoneal foregut cysts are rare, and to obtain a preoperative definitive diagnosis of this condition is relatively difficult. In addition, the exact mechanism and formation of ciliated retroperitoneal foregut cysts remains unknown.

Case presentation: Here, we report a case of a 47-year-old woman who presented with an unusual shaped cystic lesion associated with a patch of solid components between the pancreas and the left kidney, initially misdiagnosed as a cystic pancreatic lesion 5 years previously to presentation at our clinic. During the past years, reports relating to the surveillance of these lesions described that their shapes progressively change while its volumes remain invariably unchanged. We did not observe this phenomenon in any literatures to our knowledge. The patient was diagnosed with ciliated retroperitoneal foregut cyst with remote hemorrhage, after the laparoscopic surgery.

Conclusions: Ciliated retroperitoneal foregut cysts have characteristic manifestations. From this case, we summarized that cysts in the retroperitoneum, associated with a changing shape of the lesion, highly suggest the diagnosis of foregut cysts, which are safe under long-term surveillance.

Abbreviations: CT = computed tomography, MRCP = magnetic resonance cholangiopancreatography.

Keywords: computed tomography, follow-up study, foregut cyst, retroperitoneal space

1. Introduction

Ciliated foregut cysts are rare congenital aberrations, originating from the abnormal budding of the embryonic foregut. These cysts are often less than 4 cm in diameter, with a unilocular and smooth-walled appearance.[1,2] Ciliated foregut cysts typically occur in the mediastinum; however if they appear below the diaphragm, typically they occur in the liver, and rare in the retroperitoneal space.[1,3,4] There have been very few reported cases of ciliated retroperitoneal foregut cysts, none of which, to our knowledge, have reports of long-term surveillance. There natural history and growth pattern are poorly understood. Therefore, we describe a case with a 5-year follow-up, which we hope can bring to light useful insights into the growth pattern of these rare cysts, and put forward suggestions relating to their treatment.

1.1. Consent

The retrospective case report was approved by the patient, as well as the Review Board of Institute of Second Affiliated Hospital of Zhejiang University School of Medicine.

2. Case report

A 47-year-old woman was admitted to the Second Affiliated Hospital of Zhejiang University School of Medicine (September 14, 2017) with the chief complaint of discovering cystic pancreatic lesion 5 years previously. At the time when the patient underwent stomach polyp extirpation, at a local hospital (unclear) 5 years ago, she was diagnosed as suffering from a cystic pancreatic lesion. This diagnosis was coincidentally made upon performing a conventional abdominal CT scan. She occasionally suffered from abdominal distension after satiation without nausea, vomiting, or abdominal pain. The result of the physical examination was unremarkable. Laboratory results including pancreas function and tumor markers were all within normal ranges.

Several contrast-enhanced abdominal CT examinations were performed during the 5 years at our hospital. In general, CT imaging revealed an irregularly shaped cystic lesion, located in the left upper retroperitoneal region, which had a very close relationship with the pancreas from the view of the transverse section alone. Furthermore, it was misdiagnosed as a cystic pancreatic lesion for the first 3 times due to lack of the

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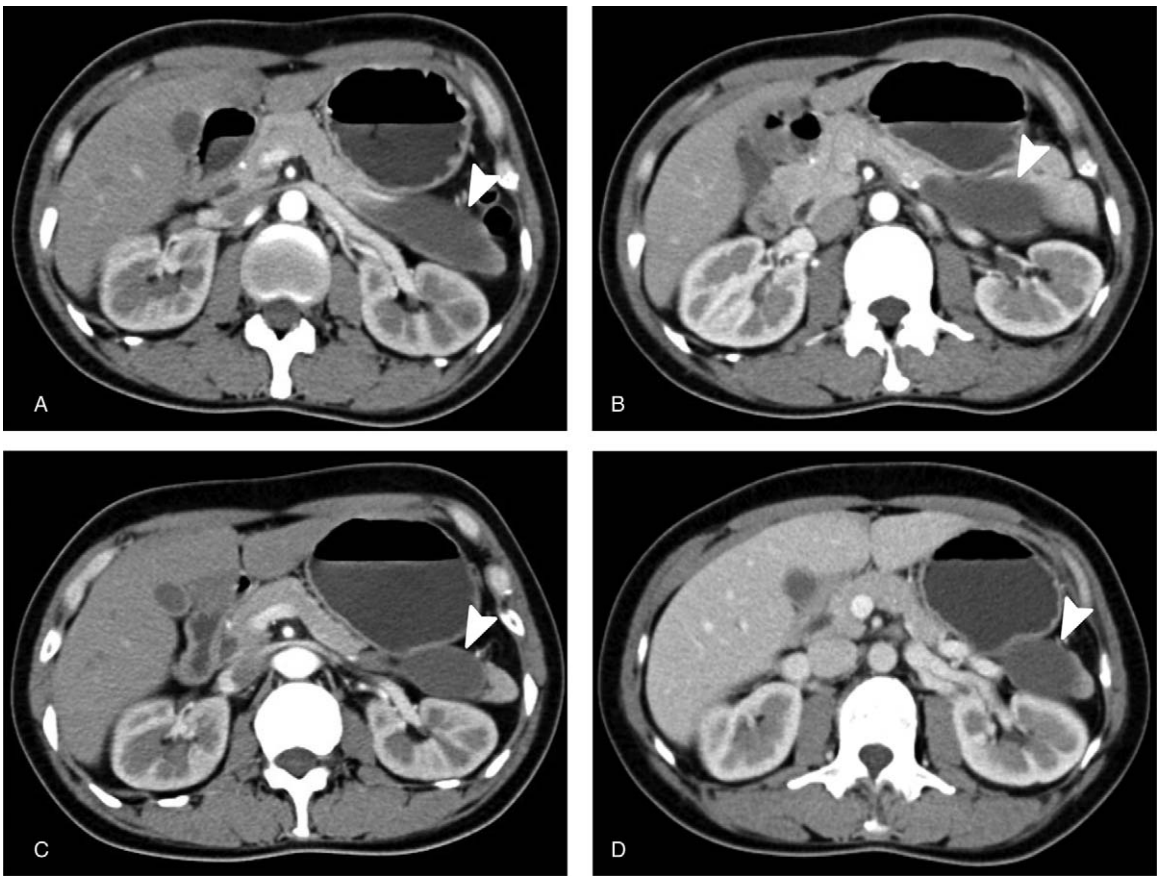

Figure 1. Contrast-enhanced CT during 5 years of surveillance. 2012-10-26 (A); 2013-06-17 (B); 2014-09-16 (C); 2017-09-07 (D). (A–D) Four images at different periods of time manifested as an irregularly-shaped cystic lesion without enhancement, which had a very close relationship with pancreas from the view of the transverse, and it was misdiagnosed with a cystic pancreatic lesion in the first 3 times. During follow-up, the shape of the lesion significantly changed, which may be a distinctive expression of the ciliated foregut cyst. CT = computed tomography.

multiplanar reconstruction (Fig. 1A–C). However, the boundaries between the pancreas and the lesion, which were deeply embedded in the retroperitoneal space, were well defined on the coronal reconstruction (Fig. 2A). The lesion was located adjacent to the pancreas, which was compressed and displaced downward. After the mentioned years of follow-up, the shape of the lesion significantly changed, while the volume did not significantly change. This was observed with the use of volumetric software on CT (Fig. 1A–D). The progressive changes to the volume, according to time, were as follows: 9.78 cm^3 (2012), 9.82 cm^3 (2013), 9.95 cm^3 (2014), and 10.02 cm^3 (2017), respectively. The lesion was much larger than usual, measuring 9.8 cm × 3.5 cm, with homogeneous water attenuation, and the average Hounsfield unit (HU) was 18.9, without any enhancement. Meanwhile there was slight enhancement representing a small area of solid components. No secondary infection, oppression symptoms, or perforation had occurred in surveillance.

Prior to surgery, magnetic resonance cholangiopancreatography (MRCP) was performed in order to confirm the relationship between the cystic lesion and the pancreatobiliary tract system; and the lesion did not communicate with either of them. Furthermore, on T2-weighted coronal imaging, the lesion exhibited a homogeneous high-intensity signal with a strip of low-intensity signal, which possibly indicated the presence of remote hemorrhage (Fig. 2C). Notably, the shape of the lesion was different from the CT scan performed 10 days prior, which pointed out the smooth outer surface and flowing fluid within the cyst (Fig. 2A and B).

Laparoscopic surgery was performed (September 20, 2017), and the gross specimen demonstrated a pearly white translucent cyst originating from the retroperitoneum, above the neck of the pancreas. Histopathology revealed that the cysts wall was abundant in smooth muscle, lined by pseudo stratified ciliated columnar epithelium, without any mucus glands or cartilage (Fig. 3). The remaining part of the cystic wall revealed epithelial shedding that was replaced by old bleeding and fibrous tissue hyperplasia. Postoperative histopathological examination confirmed the diagnosis of ciliated retroperitoneal foregut cyst with remote hemorrhage.

The patient was uneventful and discharged home 8 days after the surgery.

3. Discussion

A lesion with an associated cystic wall lined with ciliated columnar epithelium is known as a ciliated foregut cyst, which is almost a benign congenital abnormality. The diagnosis can be made by the presence of the pseudo stratified ciliated columnar epithelium as well as abundant smooth muscle, without the presence of cartilage tissue, which can be distinguished from bronchogenic cysts.[5,6]

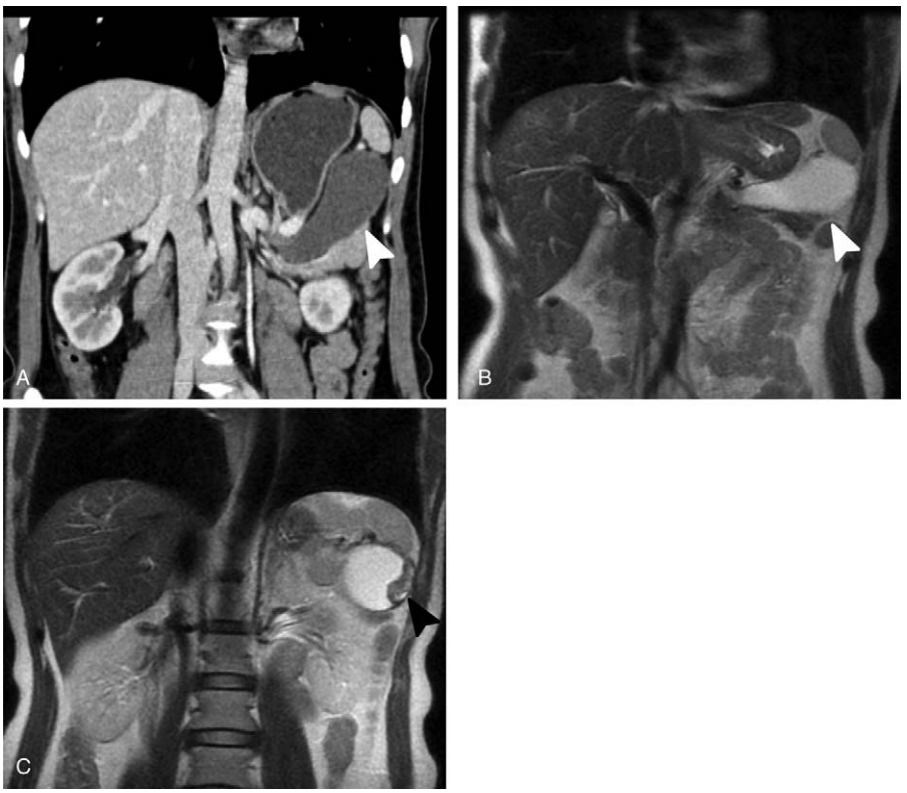

Figure 2. Coronal CT (2017-09-07) and T2WI of MRI (2017-9-18). (A) Coronal CT image showed a well-defined boundary between the ciliated foregut cyst and pancreas. The pancreas was compressed and displaced downward. (B) Just 11 days later, the shape of the cyst was changed, but the volume had no significant change in the imaging. (C) T2WI showed the lesion exhibited homogeneous high-intensity signal with a strip of a low-intensity signal, which may indicate the presence of remote hemorrhage. CT = computed tomography.

Embryologically, the foregut includes compositions of the endoderm and mesoderm, which results in the development of the pharynx, tracheobronchial tree, esophagus, stomach, intestine, and hepatobiliary system, presumably by migration.[2] Foregut cysts originate from the abnormal budding of the embryonic primitive foregut during the 3rd to 7th weeks of development, and can be classified into bronchogenic, enterogenous, or undifferentiated types. Furthermore, cysts with ciliated epithelium that lack other distinguishing characteristics, such as ciliated foregut cysts, belong to the undifferentiated classification.[7] Owing to the developmental origins of the cysts, foregut cysts usually occur in the mediastinum when a respiratory bud develops on the anterior aspect of the foregut that develops into the trachea and bronchi.[2,4] However, abnormal budding off the tracheobronchial tree may "pinch off" and migrate into the abdominal cavity through the pericardio-peritoneal canal on the 6th week of embryogenesis.[8] They are frequently located on the left upper side of the midline behind the stomach, especially near the adrenal gland and the peripancreatic region.[9] In the case we presented here, the lesion was closely located in the peripancreatic

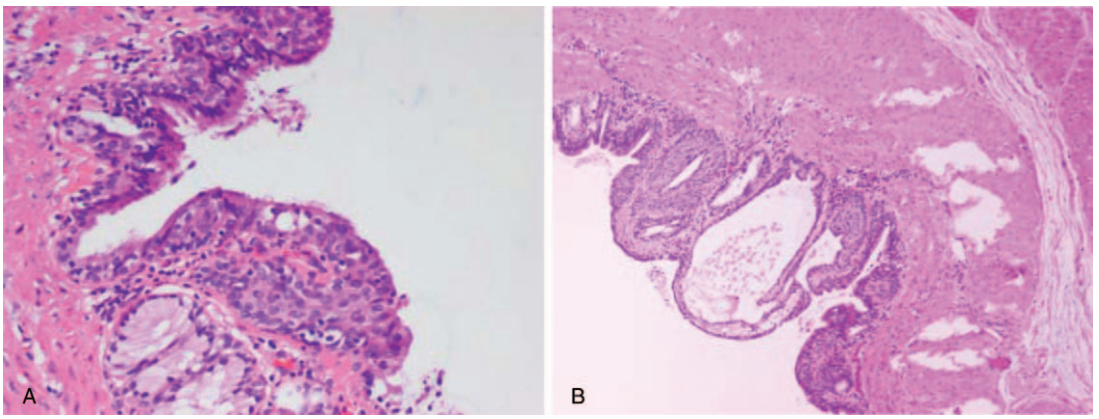

Figure 3. Hematoxylin and eosin stained sections from retroperitoneal cyst: ×100 (A); ×200 (B). (A) The cyst was lined by pseudo-stratified ciliated columnar epithelium, without any glands or cartilage. (B) The cyst wall was abundant in smooth muscle.

vicinity and was surrounded by the stomach and spleen, clearly observed by coronal CT imaging. However, it seemed to be derived from, or became incorporated in, the pancreas from the transverse section alone, which may be the reason behind it being the leading cause of misdiagnosing.

As seen in our case, ciliated retroperitoneal foregut cysts are able to grow to a large size asymptomatically owing to the considerable space that surrounds it. According to previous reports, most foregut cysts are reported to be less than 4 cm in size, and present with thin unilocular enhanced capsular walls, as fluid-filled masses, and with or without a soft tissue.[2,10] The case we presented in our study, reached up to 10 cm in size, and revealed a smooth unusual appearance, which is particularly rare. The patient did not experience any dramatic symptoms. Retroperitoneal foregut cysts are capable of becoming significant cysts, but not specific ones because of secondary infections, perforated cysts or compress the adjacent structures if they grow large enough.[10]

During the 5-year surveillance period, the appearance of the cyst had significantly changed, which hinted at the movement of the cyst. However, the exact mechanism and the formation of ciliated retroperitoneal foregut cysts remain unclear. Some researchers have postulated that the same mechanism that underlie the development of ciliated foregut cyst may be related to adjacent abdominal organs and the respiratory systems.[11] As for foregut cysts in the retroperitoneum, these cysts may arise from gastric or small intestinal epithelium. Under such circumstances, these cysts often adhere tightly to and recapitulate the construction of the native organ.[8] In our case, the cyst was abundant in smooth muscle and became close to the stomach, which may explain the mechanism of contraction. However, we know relatively little about the biological behavior of ciliated foregut cysts due to its rarity. Altering the shape of the cyst may be a distinctive expression of the ciliated foregut cyst, which can hint to its origin and formation.

Through these years of follow-up, we discovered that ciliated foregut cysts were benign slow-growing lesions that produce bleeding within the soft tissue. This may arise due to the congenitally underdeveloped vessels. Malignant degeneration has been reported in hepatic ciliated foregut cysts but not in retroperitoneal space.[12] Nevertheless, a possible destructive potential could not be entirely excluded. However, the management of asymptomatic ciliated retroperitoneal foregut cysts remains controversial.[13] Some suggest that immediate surgery is not recommended unless the patient develops symptoms. While others advocate that even a case of asymptomatic cysts that occur in young patients should be dealt with surgically due to potential risks of secondary complications.[14] Therefore, we would recommend surveillance with regular imaging when cysts are small or in older aged patients. Concerning young patients or large cysts, monitoring and minimally invasive surgical intervention are both recommended depending potential risks and secondary complications.

The strengths of this case report were that long-term surveillance and integrated clinical materials were obtained which helped us to conduct a fully analysis. However, there were limitations as part of this report. Multiplanar reconstructions of CT were not reconstructed in the first 3 follow-ups, which may have given us more information regarding the nature of the lesion. In addition, there was only one case, which is not sufficient to elucidate the underlying pathological mechanisms. Further studies are needed in order to provide a more detailed explanation.

In conclusion, we reported an unusual manifestation of a case describing a patient with a ciliated foregut cyst of the retroperitoneum, with 5 year surveillance. Making an accurate diagnosis of this condition preoperatively is very difficult; however, some particular symptoms can be helpful when diagnosing foregut cysts. From this case, we concluded that cysts in the retroperitoneal region, associated with an altered shape during follow-up, highly suggest a diagnosis of foregut cysts. These cysts are safe under long-term surveillance.

Author contributions

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Supervision: Xiu-Liang Zhu, Ri-Sheng Yu.
Writing – original draft: Hai-Yan Chen.

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