

Case Report
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## A foregut cyst mimicking a cystic pancreatic tumor—report of a case

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### Abstract

We reported a rare case of foregut cyst with bronchial and small bowel epithelium. In this case there was a difficulty in differentiating from pancreatic tumor. The patient was a 30 year-old female with a complaint of left abdominal pain. Abdominal ultrasonography, CT scan and endoscopic ultrasonography revealed a tumor that was located in the body and the tail of the pancreas. And solid parts and cystic parts were present within the tumor. The elevation of serum CA19-9 level was observed. Endoscopic retrograde cholangio-pancreatography did not show abnormalities in the pancreatic duct. With the findings of the imaging examinations stated above, firstly, solid-pseudopapillary tumor of the pancreas was suspected. Therefore, we performed laparotomy, but there was no continuity between the tumor and the pancreas. And the resection of the tumor was carried out without difficulties.

Histological examination of the resected specimen revealed both bronchial and small bowel epithelium, in the absence of ectodermal structures. The diagnosis was a foregut cyst, which is a type of benign developmental anomalies arising in the primitive foregut. Immunohistochemical staining of CA19-9 showed a positive staining in the stratified squamous epithelium, bronchial epithelium and the fluid within the cystic structures. Cystic developmental anomalies of the foregut are mostly seen cranial to the diaphragm, and it is rare to see such anomalous cystic lesions caudal to the diaphragm. In conclusion, although foregut cyst of the location in this case is rare, it should be considered as one of the differential diagnosis of the retroperitoneal organs including pancreas.

(Key words : foregut cyst, CA19-9, retroperitoneum)

### Introduction

Foregut cyst is a developmental anomaly arising in abnormal budding of the primitive foregut during the third to seventh weeks of gestation.<sup>1)2)</sup> Primitive foregut gives rise to

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broncho-pulmonary organs, esophagus, stomach, proximal small intestine, liver and pancreas.<sup>3)</sup> A common developmental anomaly of the foregut is bronchogenic cyst, which is located mostly above the diaphragm.<sup>4)</sup> Foregut cyst is usually used as a term that includes less differentiated developmental anomalies of the foregut. Foregut cyst of the location in this case is rare, and its preoperative diagnosis is not always possible because of non-specific findings on imaging examinations. We experienced a case of foregut cyst, which was situated adjacent to the pancreas. The preoperative imaging diagnosis in this case was a pancreatic tumor because the tumor appeared to exist in continuity to the pancreas. In addition to the diagnostic difficulty on imaging examinations, serum CA19-9 was higher than normal level. In this case report, we discussed the diagnostic aspects of the tumor, and also the rarity of the location of foregut cyst in this case by reviewing the previous reports.

### Case Report

The patient was a 30 year-old female with a complaint of left abdominal pain. The past history and the family history were unremarkable. In June 2004, the patient began to feel a dull pain on her left abdomen. She was seen in a local clinic, where no abnormalities were found except for a mild elevation of serum amylase. Eventually, the symptom disappeared. Ten months later, the patient recognized a recurrent pain on her left abdomen and visited a local hospital. CT scan and abdominal ultrasonography revealed a tumor measuring 50 mm in diameter that was located in the body and the tail of the pancreas. She was referred to our hospital for further examinations and treatment.

On admission, her height and weight were 157.7cm and 39.9kg respectively. There were no abnormal findings on physical examinations, and no tumors were palpable in the abdomen. Laboratory data of the blood and the urine were normal except for a high serum CA19-9 level of 947U/ml. The levels of other tumor markers such as CEA and alpha-fetoprotein were normal. The plain abdominal X-ray showed fine calcifications in the left upper abdomen. There was no intestinal gas shadow in the upper left quadrant of the abdomen suggesting compression of the bowel by the tumor. Abdominal ultrasonography showed a mass measuring 64x35 mm with a combination of high echoic solid areas and low echoic cystic areas (Toshiba SSA-270A, 3.5MHz Toshiba Japan) (Fig. 1). In the cystic areas, septum like structures was depicted. The

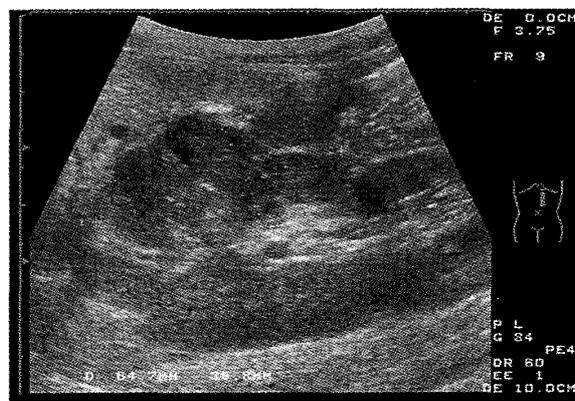


Fig. 1 Ultrasonography demonstrates a combination of high echoic solid areas and low echoic cystic areas.

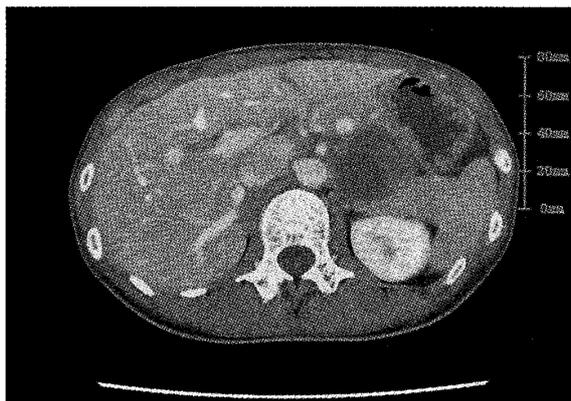


Fig. 2 Abdominal CT showed septum-like structures in the cystic parts of the tumor with calcified foci.

mass was contiguous to the pancreas tail, and there were no signal of blood flow on Doppler ultrasonography. Endoscopic ultrasonography (Olympus UMQ200, 7.5MHz, Olympus Japan), which was performed with the probe in the stomach, showed a tumor measuring 50 mm in maximum diameter situated in the body and the tail of the pancreas. The tumor was mostly round with an uneven and unclear margin. There was a low echoic area around the margin of the tumor, and papillary solid parts and cystic parts with septum-like structures were present within the tumor. Abdominal CT showed a tumor measuring 60x50x45 mm on the dorsal part of the pancreas body, and there was no clear boundary between the tumor and the pancreas. There was a thick capsule surrounding the tumor. Similar to the findings of the ultrasonography, there were both solid and the cystic parts within the tumor. And there were septum-like structures in the cystic parts of the tumor. Calcified foci were seen in the tumor. On venous injection of the contrast media, there was no enhancement effect in the tumor (Fig. 2). Endoscopic retrograde cholangio-pancreatography did not show abnormalities in the pancreatic duct.

With the findings of the imaging examinations stated above, firstly, solid-pseudopapillary tumor of the pancreas was suspected. Therefore, a resection of the body and tail of the pancreas including the tumor was scheduled. Upon laparotomy, the tumor existed caudal to the pancreas body and cranial to the left renal vein. There was no continuity between the tumor and the pancreas. Arterial blood supplies were seen both from superior mesenteric artery and the splenic artery. The dissection of the tumor from the surrounding structures was performed without difficulty, and the tumor was completely resected.

The resected tumor measured 65 mm in maximum diameter. The cut surface of the tumor showed both solid parts and cystic parts. The cystic parts were divided by septum like structures (Fig. 3).

Histological examination showed that the cystic structures were mostly lined by stratified squamous epithelium with scattered keratinizations. There were no appendages of the skin. Bronchial epithelium and small bowel epithelium were present in some areas of the cystic structures. The findings were diagnostic of foregut cyst (Fig. 4). There were fibrosis and smooth muscle of the bowel wall in the solid part of the tumor.

Immunohistochemical staining of CA19-9 showed a weakly positive staining in the stratified

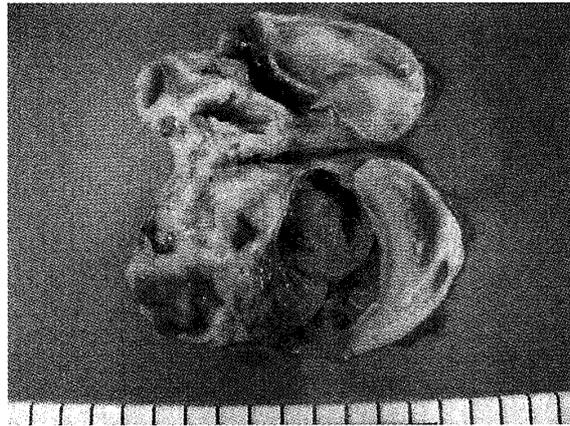


Fig. 3 The cut surface of the tumor showed both solid parts and cystic parts, and the cystic parts were divided by septum like structures.

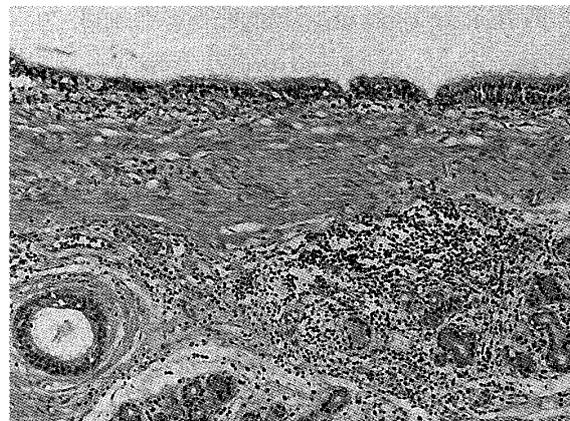


Fig. 4 Histological examination showed stratified squamous epithelium, bronchial epithelium and small bowel epithelium.

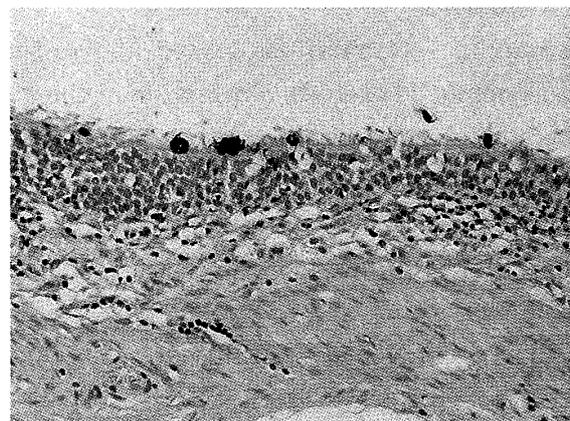


Fig. 5 Immunohistochemical staining of CA19-9 showed a weakly positive staining in the bronchial epithelium.

squamous epithelium, bronchial epithelium and the fluid within the cystic structures (Fig. 5). The small bowel epithelium was negative for CA19-9 staining.

The post operative course was uneventful and the patient was discharged on the 8th day after the operation. The serum CA19-9 level measured one month after the operation was 47 U/ml,

which was much lower than the preoperative level, nearly reaching the normal range.

## Discussion

Preoperative diagnosis of the tumor in the present case was solid-pseudopapillary tumor of the pancreas (SPT). The reasons for the preoperative diagnosis were as follows. Firstly, the patient was relatively young and female, secondly, CT and ultrasonography showed findings of a pancreatic tumor, thirdly, there was no continuity between the tumor and the pancreatic duct, and lastly, the tumor consisted of cystic parts and the solid and papillary areas on endoscopic ultrasonography. We did not perform angiography in this case. However, if it were done, it would not have been diagnostic, because both SPT of the pancreas and foregut cyst are usually avascular or hypovascular. In a retrospective observation of the abdominal CT in our case, we could not see what is called a "beak sign" between the pancreas and the cystic tumor. The retrospective observation on CT led to a possible diagnosis of extra-pancreatic tumor.

Histologically both tracheal and small bowel epithelium were present in the cystic lesion with the absence of ectodermal and mesodermal structures such as appendages of the skin such as hair or nail. Such histological findings were considered compatible with foregut cyst and not with teratoma. Foregut cyst is a developmental anomaly that originate in abnormal budding of the primitive foregut during the third to seventh weeks of gestation. There are wide variations in the histological features of the foregut cyst. In the most frequent type, tracheobronchial elements were seen. Such a foregut cyst is usually diagnosed as bronchogenic or bronchopulmonary cyst, and is usually situated cranial to the diaphragm. The occurrence of bronchogenic cyst in the location of this case is rare. Haddadin et al reported that there were 20 such cases in the world literature at the time of their report in 2001.<sup>1)</sup> Takahashi reported a case of retroperitoneal bronchogenic cyst, and stated that there were 23 such cases in the Japanese literature.<sup>5)</sup> Kohzaki reported a case of ciliated foregut cyst of the pancreas mimicking teratoma.<sup>6)</sup> In that case ciliated pseudostratified epithelium was seen in the absence of cartilage and smooth muscle. In another variation of the foregut cyst, there are gastrointestinal epithelium or both gastrointestinal and bronchial epithelium. Such a type of foregut cyst is even rarer than the bronchogenic cyst, and is sometimes seen in the retroperitoneum.<sup>2)7)</sup>

Similar to the case reported by Kohzaki, we also needed to make a differential diagnosis between foregut cyst and teratoma and made the final diagnosis of foregut cyst because of the absence of ectodermal or mesodermal structures.

The serum CA19-9 level was higher than normal in our present case. The serum CA19-9 is elevated usually in pancreatic or bile duct tumors. However, it is reported that CA19-9 is present in normal epithelium of the salivary gland, pancreatic duct, bile duct, bronchus and the stomach.<sup>8)</sup>

There were reported cases of bronchogenic cysts in which serum CA19-9 level was elevated.<sup>6)9)</sup> However, the staining pattern of CA19-9 in the cysts was not presented. In our case, CA19-9 staining was positive in a part of the bronchial epithelium and in the stratified squamous epithelium of cystic structures. Although the stratified squamous epithelium lining of the cystic structure showed a positive staining for CA19-9, it is extremely unusual to see

CA19-9 production in the stratified squamous epithelium. There were CA19-9 positive accumulations within the cysts, and the positive staining of the stratified squamous epithelium might be secondary to the accumulation of the fluid. The cause of the CA19-9 positive fluid accumulation is still unclear, and the scattered staining of the bronchial epithelium might be secondary to the accumulation of the positively stained fluid within the cystic structure.

We reported a case of foregut cyst located ventral to the pancreas. Although such a location of foregut cyst is rare, we have to keep it in mind that foregut cyst should be considered as one of the differential diagnosis of the cystic lesions of the retroperitoneum.

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## 「嚢胞性膵腫瘍と鑑別困難であった foregut cyst の一例」

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## 要 約

術前診断に苦慮した foregut cyst の 1 例を経験したので文献的考察を加えて報告する。症例は 30 歳の女性、主訴は左側腹部鈍痛であった。腹部超音波、CT 検査、超音波内視鏡では膵尾部と連続する辺縁明瞭な腫瘤が存在し、内部は cystic part と solid part の混在が認められた。また、血液検査では CA19-9 の上昇がみられた。ERCP では膵管に拡張はなく嚢胞との交通も認めなかった。以上より膵臓原発の solid and pseudopapillary tumor を第一に考え、開腹手

術を行った。術中所見で腫瘍は膵尾部とは連続性がなく腹腔内腫瘍として腫瘍摘出術を行った。術後の病理組織診断は foregut cyst であった。また、病理組織では CA19-9 染色で重層扁平上皮、気管支上皮、嚢胞内容物に陽性所見が認められた。腹腔内にみられる foregut cyst は極めて稀であるが、鑑別診断として挙げる必要があると考えられる。

(キーワード: foregut cyst, CA19-9, retroperitoneum)

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