Retroperitoneal Bronchogenic Cyst Mimicking Pancreatic Cystic Lesion

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Retroperitoneal bronchogenic cyst is detected extremely rarely and often masquerades as other diseases. Here, we report 2 cases of retroperitoneal bronchogenic cyst mimicking pancreatic mucinous tumor. Histologically, both cysts were composed of ciliated respiratory-like epithelium with abundant mucin content, smooth muscle bundles and mature cartilage, compatible with the diagnosis of retroperitoneal bronchogenic cyst. In addition to these 2 cases, another 42 retroperitoneal bronchogenic cysts reported in the English literature were collected for review and analysis. Twelve (28%) were located over the peripancreatic area. Just over half (51%) of them were asymptomatic. No accurate preoperative diagnosis could be made for any of the lesions. About a third (33.3%) of the peripancreatic retroperitoneal bronchogenic cysts masqueraded as pancreatic cystic lesions. [*J Chin Med Assoc* 2006;69(11):538–542]

Key Words: bronchogenic cyst, pancreatic cyst, retroperitoneal

Introduction

Bronchogenic cyst is one of the few types of congenital anomalies of foregut cysts during embryonic development. It is formed by integration of abnormal budding and pinching of the tracheobronchial tree from the 3rd week through to the 7th week of intrauterine life.[1] Bronchogenic cysts occur most commonly in the posterior aspect of the superior mediastinum near the carina and is less frequently found in extrathoracic locations such as skin and subcutaneous tissue, pericardium and intradiaphragm.[2–4] Bronchogenic cyst in a retroperitoneal location is an extremely rare occurrence. To date, only 42 cases of retroperitoneal bronchogenic cyst have been reported in the English literature.[5–14] One of the earliest reports was in 1953, by Miller et al.[5]

Here, we present 2 cases of retroperitoneal bronchogenic cyst mimicking pancreatic cystic lesion. In addition to these 2 cases, the other 42 retroperitoneal bronchogenic cysts reported in the English literature were collected for review and analysis, with particular emphasis placed on cyst location relating to the pancreas and preoperative diagnosis.

Case Reports

Case 1

A 69-year-old male patient had undergone subtotal gastrectomy with Billroth-II gastrojejunostomy reconstruction for perforated peptic ulcer 30 years ago, and thymectomy and adjuvant radiotherapy for malignant thymoma 2 years ago. He presented with right upper quadrant discomfort of 1 month's duration. Physical examination was unremarkable. Ultrasound and computed tomography (CT) of the patient's abdomen revealed a mass measuring 4 × 2.5 cm at segment V of the liver, which we suspected to be hepatocellular carcinoma. Another cystic mass lesion, measuring 4 cm in diameter and with homogeneous low attenuation on unenhanced CT, was found at the left anterior pararenal space, just above and attaching to the pancreatic body. Contrast-enhanced CT showed no contrast enhancement of the cystic mass, with imperceptible wall (Figure 1A). Angiography revealed only a faint tumor stain within the cystic lesion. Based on a collective review of the results of these diagnostic studies, the preoperative diagnosis was pancreatic mucinous tumor or pseudocyst

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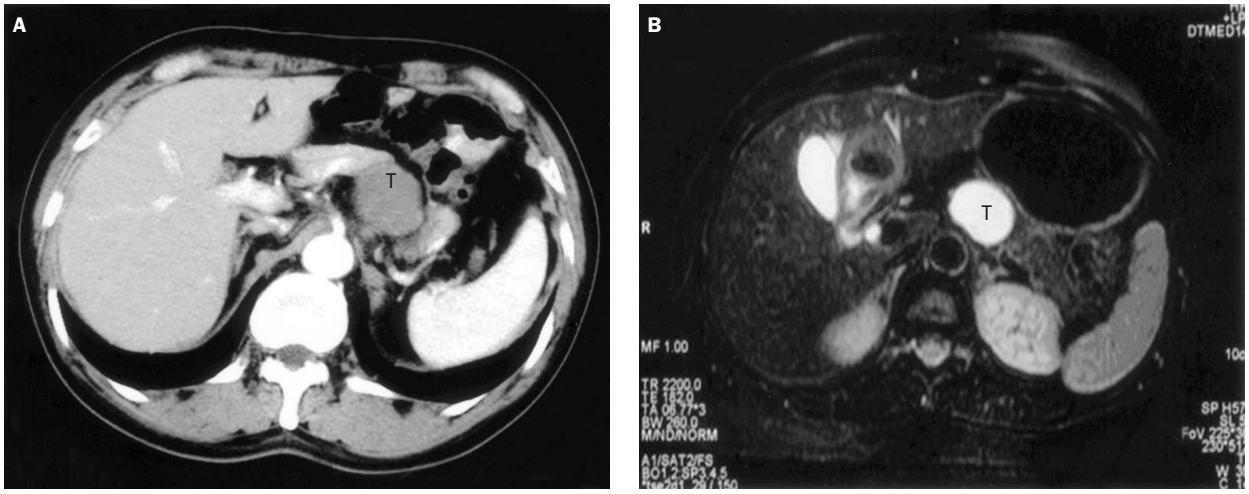

Figure 1. (A) Contrast-enhanced computed tomography shows a thin-walled water-attenuated cystic lesion (T) in the pancreatic tail region. (B) Axial T2-weighted fat-suppressed magnetic resonance imaging demonstrates a cystic lesion (T) with homogeneous high signal intensity close to or adjacent to the pancreatic body.

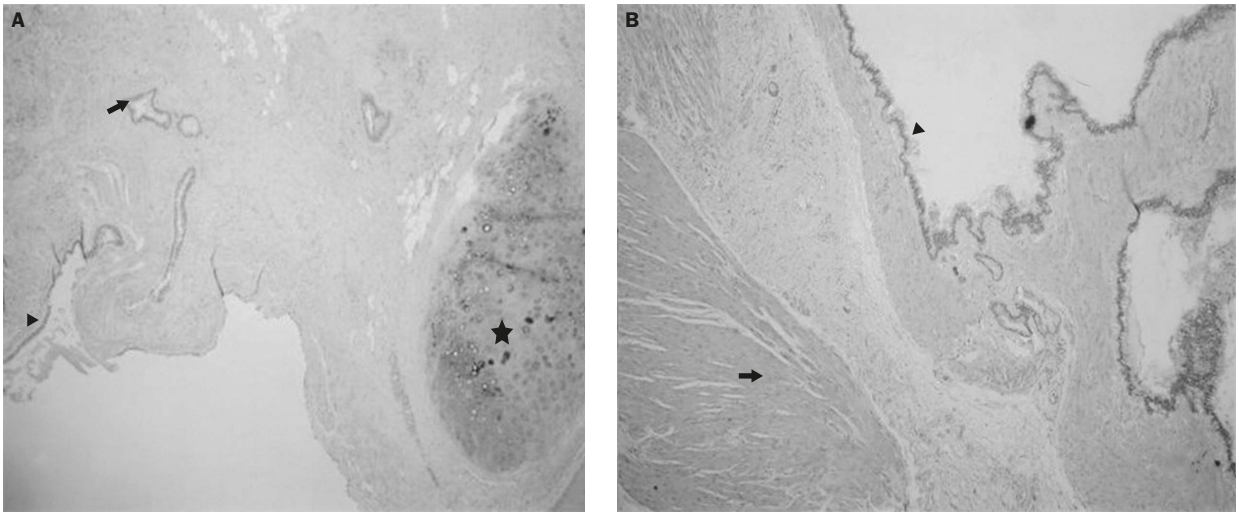

Figure 2. Microscopic appearance of retroperitoneal bronchogenic cyst. (A) The cystic lining is composed of a thin layer of epithelium (arrowhead) covering loose connective tissue and some bronchial glands (arrow) and cartilage (star) (hematoxylin & eosin, 10×). (B) A detail of the cystic wall with respiratory-type ciliated columnar epithelium (arrowhead), containing scattered smooth muscle fibers (arrow) (hematoxylin & eosin, 40×).

near the pancreas, and hepatocellular carcinoma at segment V of the liver. The patient underwent exploratory laparotomy with resection of both lesions. The cystic lesion measured $7 \times 3.5 \times 2$ cm in size with mucin content, and was located in the retroperitoneum just above the pancreas body and tail. Pathologic diagnosis of the hepatic tumor confirmed hepatocellular carcinoma, but microscopic examination of the retroperitoneal cyst revealed a ciliated respiratory-like epithelium with abundant mucin content, smooth muscle bundles and mature cartilage, compatible with bronchogenic cyst (Figure 2A).

Case 2

A 54-year-old female patient was referred to our institution for the evaluation of a pancreatic cystic lesion over the pancreatic body, which had been incidentally discovered by abdominal ultrasound during work-up study of an episodic fever event. The abdominal sonogram showed a cystic mass with internal echogenic septum over the superior border of the pancreatic body. Axial magnetic resonance imaging (MRI) identified a thin-walled cystic mass measuring 4 cm, with regular margin, in the left anterior pararenal space, just cranial and attaching to the pancreatic body and

close to the upper lesser curvature of the stomach, with low signal intensity on T1-weighted MRI, hyperintensity when compared to cerebral spinal fluid on T2-weighted MRI, and no enhancement on post-gadolinium T1-weighted fat suppression gradient echo MRI (Figure 1B). Serum tumor markers including carbohydrate antigen 19-9 and carcinoembryonic antigen were all within normal ranges. Under the tentative diagnosis of pancreatic mucinous tumor, surgery was performed. Intraoperative exploration revealed an isolated $3 \times 3 \times 1$ cm retroperitoneal cyst without adhesion to adjacent organs. In addition, it was loosely attached to the superior border of the body of the pancreas, and contained viscous yellow fluid. Microscopically, the cyst was lined with respiratory epithelium and muscular tissue, and was compatible with bronchogenic cyst (Figure 2B).

Table 1. Location of retroperitoneal bronchogenic cyst in the 44 cases (inclusive of the 2 patients in this report) in the English literature[5–14]

Location	*n* (%)
Peripancreatic	12 (28)
Anterior pancreas	1 (2)
Superior pancreas head	2 (5)
Superior pancreas neck	1 (2)
Superior pancreas body	3 (7)
Superior pancreas tail	5 (12)
Nonpancreatic*	32 (73)
Right crus of diaphragm	5 (11)
Left crus of diaphragm	7 (16)
Superior left adrenal gland	19 (43)
Superior right adrenal gland	1 (2)

**Retroperitoneal bronchogenic cyst not near the pancreas.*

Discussion

Bronchogenic cysts arise from an abnormal budding of the tracheobronchial analog of the primitive foregut. When the attachment persists, the cyst is usually found near or associated with the tracheobronchial tree or the esophagus. If a complete separation occurs, the cyst may migrate to other unusual locations. A retroperitoneal location is extremely rare, and the exact mechanism of its migration is still unknown. Sumiyoshi et al[6] hypothesized that the thoracic and abdominal cavities are linked by the pericardioperitoneal canal in an early embryonic stage. When the canal is divided into 2 respective cavities by fusion of the pleuroperitoneal membranes (the future component of the diaphragm) at the end of the 6th week of intrauterine life, abnormal buds of the tracheobronchial tree are pinched off by these membranes and migrate into the abdomen, resulting in a retroperitoneal bronchogenic cyst. Another less likely alternative hypothesis explains aberrant differentiation of foregut-derived cysts originating intra-abdominally.[7]

Histologic characteristics of typical bronchogenic cyst include ciliated epithelium, cartilage and bronchial glands with mixed content of serous and mucous secretions.[8] Plain radiography, ultrasonography, and CT of the abdomen have been the primary imaging modalities for evaluating and defining the nature of these masses and their anatomic relations. Bronchogenic cysts appear rounded, well-circumscribed, and hypoattenuating without enhancement on CT.[8] Sometimes, hyperattenuation may suggest hemorrhage or noncalcified solid masses, which are caused by the presence of thick proteinaceous secretions in the lesion.[8] Marin et al[15] emphasized the usefulness of MRI for diagnosing bronchogenic cysts; intermediate to high signal intensity on T1-weighted images and markedly high signal intensity on T2-weighted images would indicate that the mass contains mucinous protein-rich fluid. It has been reported that bronchogenic cysts occur in both sexes in equal ratio, and in a wide age range.[8,13]

In 1997, Hsieh et al[11] reported the first case of retroperitoneal bronchogenic cyst in Taiwan. Since then, no Chinese literature relating to bronchogenic cyst has been found. In this report, we described our discovery of 2 cases of retroperitoneal bronchogenic cyst, which were diagnosed in 2001 and 2005, respectively. Adding these 2 cases to the rest of the cases in the current English literature,[5–14] a total of 44 cases of isolated retroperitoneal bronchogenic cysts have now been reported. Table 1 lists the locations of these 44 retroperitoneal bronchogenic cysts. About 1-third (28%) of retroperitoneal bronchogenic cysts are located in the peripancreatic region. Moreover, there is a tendency for these lesions to be discovered on the left side of the abdomen. Accordingly, the most common locations are as follows: (1) superior left adrenal gland (43%); (2) left crus of the diaphragm (16%); (3) superior pancreas tail (12%); (4) right crus of the diaphragm (11%); (5) superior pancreas body (7%). The reason for this phenomenon is still unknown, but we presume that it is related to the pathway of embryologic development.

Table 2 summarizes the characteristics of the 44 patients with retroperitoneal bronchogenic cyst. Mean patient age was 39.4 ± 3 years, with a wide age range of 10 weeks to 72 years. Cysts occurred equally in both sexes (M/F: 55.8%/44.2%). Mean cyst size was 6.3 ± 0.6 cm (range, 1.7–18.9 cm), and just over

Table 2. Characteristics of the 44 patients (inclusive of the 2 patients in this report) with retroperitoneal bronchogenic cyst in the English literature[5–14]

	Total ($n=44$)	Peripancreatic ($n=12$)	Nonpancreatic* ($n=32$)	p
Age (yr)				0.18
Median (range)	43 (0.2–72)	57 (0.2–69)	41 (0.3–72)	
Mean ± SEM	39.4 ± 3	45.8 ± 6.8	37 ± 3	
Sex (male:female)	25:19	5:7	20:12	0.21
Cyst size (cm)				0.05
Median (range)	5 (1.7–18.9)	7 (2–18.9)	5 (1.7–14)	
Mean ± SEM	6.3 ± 0.6	8 ± 1.4	5.6 ± 0.5	
Symptoms, *n* (%)				0.80
No	21 (51)	6 (14.6)	15 (36.6)	
Yes	20 (48.8)	5 (12.2)	15 (36.6)	
Epigastric pain	8 (19.5)	3 (7.3)	5 (12.2)	
Left flank pain	8 (19.5)	0	8 (19.5)	
Body weight loss	3 (7.3)	1 (2.4)	2 (4.9)	
Abdominal fullness/discomfort	2 (4.9)	1 (2.4)	1 (2.4)	
Back pain	2 (4.9)	0	2 (4.9)	
Left upper quadrant pain	1 (2.4)	0	1 (2.4)	
Nausea/vomiting	4 (9.8)	2 (4.9)	2 (4.9)	
Anorexia	1 (2.4)	0	1 (2.4)	

**Retroperitoneal bronchogenic cyst not near the pancreas. SEM = standard error of the mean.*

Table 3. Preoperative diagnoses of the 44 patients (inclusive of the 2 patients in this report) with retroperitoneal bronchogenic cyst in the English literature[5–14]

	Total ($n=44$)	Peripancreatic ($n=12$)	Nonpancreatic* ($n=32$)
Adrenal tumor	9	1	8
Hemorrhagic cyst	5	0	5
Pancreatic mucinous tumor	3	3	0
Retroperitoneal cyst	3	1	3
Teratoma	3	0	3
Adrenal cyst	3	0	3
Neurogenic tumor	3	0	4
Pheochromocytoma	2	0	2
Pseudocyst	1	1	0
Post-traumatic process	1	0	1
Hamartoma	1	0	1
Leiomyoma	1	1	0
Paraganglion	1	0	1
Diaphragmatic tumor	1	0	1
Neurofibroma	1	0	1
Choledochal cyst	1	1	0
Lymphangioma	1	1	0
Not available	11	4	7

**Retroperitoneal bronchogenic cyst not near the pancreas.*

half (51%) had been found incidentally. Both of our cases were found incidentally; they had no significant symptoms or signs. Of the patients who presented with symptoms, the majority complained of epigastric pain (19.5%), left flank pain (19.5%), nausea/vomiting (9.8%), and body weight loss (7.3%). However, there was no significant difference between patients with peripancreatic- *vs.* patients with nonpancreatic-located retroperitoneal bronchogenic cysts in terms of age, sex, tumor size and symptoms.

A tentative preoperative diagnosis of retroperitoneal cystic masses depends on the location of the mass, the patient's age, and imaging characteristics. Table 3 shows the various preoperative diagnoses of the 44 cases. No accurate diagnosis was made preoperatively. In this literature review, the most frequent presumptive preoperative diagnoses were: (1) adrenal tumor; (2) hemorrhagic cyst; (3) pancreatic lesions including pancreatic pseudocyst and pancreatic mucinous tumor. Laboratory abnormalities were uncommon in patients with retroperitoneal bronchogenic cyst. There were some patients who had their metanephrine and catecholamines analyzed, which showed mild elevations. This phenomenon might be attributable to compression of the adrenal gland by the cysts, with secondary release of catecholamine from the gland. Pseudocyst and cystic mucinous tumor were also suspected preoperatively in 4 cases,[8,14] including our 2 patients. About 1-third (33.3%) of peripancreatic retroperitoneal bronchogenic cysts masquerade as pancreatic cystic lesions.

Malignancy arising in bronchogenic cysts has been reported only rarely, and most occur in the mediastinum and chest. Reported malignancies include adenocarcinoma, squamous cell carcinoma, fibrosarcoma, undifferentiated carcinoma, leiomyosarcoma, embryonal rhabdomyosarcoma, and anaplastic carcinoma.[13] So far, only 1 case of a retroperitoneal bronchogenic cyst attached to the ascending colon with associated adenocarcinoma has been reported.[13]

The treatment of retroperitoneal bronchogenic cyst is surgical removal via laparoscopy or laparotomy. Although most are asymptomatic, excision is recommended to establish a definite diagnosis, alleviate any symptoms and to remove the documented risk of malignant transformation.

In summary, among the 44 retroperitoneal bronchogenic cysts reported in the literature, there were no significant demographic differences with respect to age, sex, and presenting symptoms. Twenty-eight percent of retroperitoneal bronchogenic cysts are located over the peripancreatic region, and about 1-third (33.3%) of peripancreatic retroperitoneal bronchogenic cysts masquerade as pancreatic cystic lesions. Malignancy arising in bronchogenic cysts is rare, with only 1 case reported in the literature so far. The recommended treatment is surgical removal for definite diagnosis and symptom relief.

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