Intrapulmonary Bronchogenic Cysts: Computed Tomography, Clinical and Histopathologic Correlations

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Background/Purpose: Bronchogenic cysts (BCs) are usually located in the mediastinum and they occur less commonly in the lung parenchyma. This study investigated the findings from computed tomography (CT) images, clinical presentation and histopathologic findings of intrapulmonary BCs.

Methods: From the last 7 years, the CT images of 20 patients (12 females, 8 males; mean age, 38.8 ± 21.7 year; median age, 34 years) with intrapulmonary BC were available. Contrast-enhanced CT findings were characterized and correlated with clinical presentation and histopathologic findings (using Fisher’s exact tests).

Results: The majority of intrapulmonary BCs were subpleural in location (55%), in the lower lobes (60%), symptomatic (80%), and in adults (90%). Three CT patterns were identified: cyst with content of fluid attenuation (9 patients), cyst with air and fluid content (9 patients), cyst with content of soft tissue attenuation (2 patients). Preoperative diagnosis of intrapulmonary BC was correct in only 20% using the CT criteria of cysts with fluid attenuation and without anomalous blood supply. Cysts with air component were significantly larger than those without air component (p = 0.0452), but cyst size and air component were not correlated with clinical presentation. Surrounding infiltration or thick wall on CT were significantly correlated with the presence of any clinical symptom (p = 0.014) or fever (p = 0.042). CT findings of surrounding consolidation, ground glass opacity or thick wall were significantly correlated with chronic inflammation or pneumonic change on histopathology (p = 0.0008).

Conclusion: There is a wide spectrum of intrapulmonary BCs that have CT findings that are correlated with clinical presentations and histopathologic findings. [J Formos Med Assoc 2007;106(1):8–15]

Key Words: bronchi, clinicopathologic features, computed tomography, cyst, lung

Bronchogenic cyst (BC) is a congenital lesion and represents one of many bronchopulmonary malformations. BC presumably represents supernumerary lung buds from the primitive foregut. BCs occur more frequently in the mediastinum around the tracheobronchial tree when they form early or within the pulmonary parenchyma if they develop later.1–4 Intrapulmonary BCs typically present as thin-walled cystic lung parenchymal lesions with water content and are reported in about 15–23% of all BCs.3,4 Intrapulmonary BCs are not infrequently detected by ultrasound at the prenatal or neonatal stage.3–5 The clinical presentations of intrapulmonary BCs are variable from those of...
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incidental radiologic findings without symptoms to abnormal radiographic findings associated with clinical symptoms. Variable radiologic presentations of intrapulmonary BCs were also reported from well-defined cysts with water attenuation to complete air-filled cysts or cysts with air-fluid levels. Although radiographic, computed tomography (CT), magnetic resonance imaging (MRI) findings of intrapulmonary BCs have been described, the number of CT studies are few. In this study, we conducted a retrospective review of the CT images of a case series of 20 intrapulmonary BCs and correlated their CT patterns with the clinical presentations and histopathologic findings.

Patients and Methods

Intrapulmonary BC was defined as a BC that was located within the lung parenchyma and confirmed intraoperatively and by histopathology. Between January 1995 and December 2002, 27 patients had pathologically diagnosed intrapulmonary BC in our hospital. Twenty (12 females, 8 males; mean age, 38.8 ± 21.7 year; median age, 34 years; age range, 4 months to 76 years) of the 27 patients had chest CT available. Among the 20 patients, there were 18 adults, one 15-year-old boy and one 4-month-old male infant. Seven of the 20 patients were older than 50 years of age. All patients underwent surgical resection, including lobectomy (15 patients) and partial wedge resection (5 patients). All fulfilled the diagnostic criteria of intrapulmonary BC. The intrapulmonary location of the cysts was confirmed during surgery and by histopathologic evidence of bronchial glands, cartilage, smooth muscle and, occasionally, calcifications and/or ossification foci. The CT studies were performed without and with intravenous contrast enhancement in three patients and only with intravenous contrast enhancement in the remaining 17. CT studies were performed in the following ways: using contiguous 6 mm sections at 6 mm intervals (13 patients) (C150, Imatron; GE Healthcare, Milwaukee, WI, USA); 10 mm sections at 10 mm intervals (4 patients); and 5 mm sections at 5 mm intervals (3 patients) (HiSpeed; GE Healthcare). All CT images with contrast enhancement were reviewed.

The location, size and morphology (including the components of the cysts or adjacent lung parenchyma) of all intrapulmonary BCs were classified as subpleural, mid-lung and perihilar, depending on where the major part was. The distribution of the intrapulmonary BC was also recorded depending on which lung lobe was located. CT images with contrast enhancement were categorized according to the cystic content: type 1 cysts had fluid attenuation content only; type 2 cysts had both air and fluid attenuation content; type 3 cysts had soft tissue attenuation content isodense to chest wall muscle and cystic wall. Fluid attenuation content was defined as the cyst having lower attenuation than that of enhancing chest wall muscle based on visual perception on hard copies in window setting level – 500 HU to −750 HU and width 1000 HU to 1500 HU for lung parenchyma, and level 20 HU to 35 HU and width 300 HU to 500 HU for mediastinum. Wall thickness was defined as “thin” if the thickest part of the cystic wall was ≤3 mm on enhanced CT, while a “thick” wall was considered if the thickest part was >3 mm. It was subcategorized into “a” and “b” for each individual type, where “a” indicated a thin-walled cyst with clear interface and no infiltration of the surrounding lung parenchyma and “b” indicated a thick cystic wall or with infiltration of the surrounding parenchyma. Surrounding lung parenchymal change of intrapulmonary BC was defined in terms of consolidation and/or ground glass opacity. Emphysema was recorded based on available CT images because it was reported to be an associated finding of intrapulmonary BC in a previous study. The clinical history, preoperative diagnosis and all major clinical symptoms including
productive cough, dry cough, blood-tinged sputum, hemoptysis, chest pain, dyspnea, fever, chills, were reviewed based on the medical records. Pathologic specimens of the cysts were reviewed by an experienced pulmonary pathologist. The pathologic findings of surrounding lung parenchyma were also recorded.

All analyses were performed using SAS version 8.2 (SAS Institute Inc., Cary, NC, USA) for Microsoft Windows. A p value <0.05 was considered statistically significant. Group comparisons were conducted using Fisher’s exact tests for proportions and general linear models for continuous variables. Correlations between types and sizes of intrapulmonary BCs and clinical symptoms were performed with Fisher’s exact test. Correlations of CT patterns, including the presence of air or surrounding parenchymal change, with each symptom and the presence of any symptom were performed. Histopathologic findings of surrounding lung parenchymal change (pneumonia or inflammation) were correlated with CT evidence of surrounding consolidation, ground glass opacity or thick wall (category b).

### Results

**CT findings and patterns of intrapulmonary BCs**

The Table shows the distribution of CT patterns. Nine patients (45%) had type 1 cyst, nine (45%) had type 2 cyst, and two (10%) had type 3 cyst. Among the type 2 cysts, the percentage of air component ranged from little (5%) to predominantly air-filled cysts (15%), but the majority (25%) had an approximately equal amount of air and fluid component. The mean size of intrapulmonary BC was 6.9 ± 4.6 cm (range, 1.2–20 cm). Type 2 cysts with air component (9.6 ± 4.9 cm) were significantly larger in size (p = 0.0452). Calcified spots of the cystic wall were found in two patients (10%), including one type 1 (Figure 1) and one type 2 intrapulmonary BC. In type 3 cysts, wall thickness could not be evaluated. There were multiple locules of intrapulmonary BC in nine patients and a single locule of intrapulmonary BC in 11 patients. Consolidation and ground glass opacity were found adjacent to or surrounding the cysts in 11 patients (55%). There were eight cysts (40%)

### Table. Clinical features and computed tomography patterns of 20 patients with intrapulmonary bronchogenic cysts*

<table>
<thead>
<tr>
<th></th>
<th>Type I, simple cyst (n = 9)</th>
<th>Type II, cyst with air (n = 9)</th>
<th>Type III, soft tissue mass (n = 2)</th>
<th>Total (n = 20)</th>
<th>p†</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sex, male/female</strong></td>
<td>3M/6F</td>
<td>5M/4F</td>
<td>0M/2F</td>
<td>8M/12F</td>
<td>0.538</td>
</tr>
<tr>
<td><strong>Age, yr</strong></td>
<td>37.9 ± 17.6</td>
<td>36.9 ± 25.9</td>
<td>51.5 ± 27.6</td>
<td>38.8 ± 21.7</td>
<td>0.705</td>
</tr>
<tr>
<td><strong>Symptoms</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chest pain</td>
<td>3 (33)</td>
<td>2 (22)</td>
<td>0 (0)</td>
<td>5 (25)</td>
<td>1.000</td>
</tr>
<tr>
<td>Dry cough</td>
<td>1 (11)</td>
<td>1 (11)</td>
<td>0 (0)</td>
<td>2 (10)</td>
<td>1.000</td>
</tr>
<tr>
<td>Productive cough</td>
<td>3 (33)</td>
<td>4 (44)</td>
<td>0 (0)</td>
<td>7 (35)</td>
<td>0.818</td>
</tr>
<tr>
<td>Blood-tinged sputum</td>
<td>1 (11)</td>
<td>2 (22)</td>
<td>1 (50)</td>
<td>4 (20)</td>
<td>0.420</td>
</tr>
<tr>
<td>Hemoptysis</td>
<td>1 (11)</td>
<td>3 (33)</td>
<td>0 (0)</td>
<td>4 (20)</td>
<td>0.733</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>1 (11)</td>
<td>2 (22)</td>
<td>0 (0)</td>
<td>3 (15)</td>
<td>1.000</td>
</tr>
<tr>
<td>Fever</td>
<td>3 (33)</td>
<td>3 (33)</td>
<td>0 (0)</td>
<td>6 (30)</td>
<td>1.000</td>
</tr>
<tr>
<td>Chills</td>
<td>2 (22)</td>
<td>0 (0)</td>
<td>0 (0)</td>
<td>2 (10)</td>
<td>0.574</td>
</tr>
<tr>
<td>No symptoms</td>
<td>3 (33)</td>
<td>0 (0)</td>
<td>1 (50)</td>
<td>4 (20)</td>
<td>0.153</td>
</tr>
<tr>
<td>Leukocytosis</td>
<td>0 (0)</td>
<td>2 (22)</td>
<td>1 (50)</td>
<td>3 (15)</td>
<td>0.142</td>
</tr>
<tr>
<td><strong>Size, cm</strong></td>
<td>4.5 ± 3.1</td>
<td>9.6 ± 4.9</td>
<td>5.2 ± 0.2</td>
<td>6.9 ± 4.6</td>
<td>0.045‡</td>
</tr>
</tbody>
</table>

*Data are presented as mean ± standard deviation or n (%); †p values are from statistical comparison among the groups of patients with different CT patterns; ‡p < 0.05.
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with wall thickness > 3 mm and 10 cysts (50%) with wall thickness ≤ 3 mm. Fistula was found between cyst and bronchus in only two patients during surgery: one was visible on CT while the other was not visible on CT but found during operation.

The majority of the intrapulmonary BCs were of subpleural location (55%), followed by mid-lung (30%) and perihilar (15%). The distributions of intrapulmonary BCs were in the right upper lobe (2 patients), right middle lobe (1 patient), right lower lobe (8 patients), left upper lobe (4 patients), left lower lobe (4 patients), and right middle and upper lobe (1 patient). Lower lobes (60%) were the most common location. Nine patients had type 1 intrapulmonary BC (4 type 1a, 5 type 1b) (Figure 1), nine patients had type 2 intrapulmonary BC (3 type 2a, 6 type 2b) (Figure 2).

Figure 1. A 22-year-old woman with a perihilar type 1a intrapulmonary bronchogenic cyst associated with a dense calcified nodule in the right middle lung lobe.

Figure 2. (A, B) A 48-year-old woman with a type 2 intrapulmonary bronchogenic cyst (BC): (A) axial-enhanced computed tomography scan shows a cyst with surrounding mild consolidation and heterogeneous water attenuation content with entrapped air pockets; (B) microscopically, one can see that the BC is lined by ciliated columnar epithelium with chronic inflammation and fibrous cystic wall (hematoxylin & eosin; original magnification, 33×). (C) A 19-year-old man with type 2 intrapulmonary BC with dominant air component that was diagnosed as an infected bulla before operation.
and two patients had type 3 intrapulmonary BC (1 type 3a, 1 type 3b) (Figure 3). Two type 1a, one type 1b and one type 2b intrapulmonary BC were correctly diagnosed before surgery. The remaining two type 1a intrapulmonary BCs were diagnosed incorrectly as metastatic tumors (Figure 4) due to round shape and history of colon cancer, and pulmonary sequestration due to coexistence of a large pulmonary sequestration in the left lower lobe and a small BC near the left hilum. All three type 2a intrapulmonary BCs were diagnosed as bulla or bullae due to a large amount of air component (Figure 2C). The other five type 2b intrapulmonary BCs were considered lung abscesses due to air in the thick-walled cyst and surrounding lung parenchymal consolidation. Both two type 3 intrapulmonary BCs were considered to be lung tumors before operation due to soft tissue attenuation of cystic content (Figure 3). The overall percentage of preoperative correct diagnosis of intrapulmonary BC was 20% (4/20). Surrounding regional hypoattenuation and vascular attenuation that were compatible with CT findings of pulmonary emphysema were found in two patients (1 type 2 cyst, 1 type 3 cyst).

CT patterns and clinical presentations of intrapulmonary BCs

The majority of patients with intrapulmonary BCs (80%) were symptomatic at initial presentation, including productive cough (7 patients), fever (6 patients), chest pain (5 patients), hemoptysis (4 patients), blood-tinged sputum (4 patients), dyspnea (3 patients), dry cough (2 patients), and chills (2 patients) (Table). Three patients with type 1 and one type 3 intrapulmonary BC were free of symptoms with incidentally found lung lesions on chest radiographs. Two of the six patients with fever were associated with leukocytosis and the other two with chills (Table). No significant correlation of different types of intrapulmonary BCs was found with sex, age, clinical symptoms, or the presence of leukocytosis (Table).

Six patients with intrapulmonary BC were found to have infection due to single or multiple pathogens, including Pseudomonas aeruginosa (2 patients), Klebsiella pneumoniae (2 patients), Aspergillus fumigatus (2 patients), and Streptococcus viridans (1 patient). One patient with type 1 intrapulmonary BC was diagnosed with acute myeloid leukemia and superimposed infection of the intrapulmonary BC with A. fumigatus.
One patient with a small type 1 intrapulmonary BC had coexistent large intralobar bronchopulmonary sequestration in the left lower lobe. Mild hemoptysis occurred in four patients (20%), including one patient with type 1b intrapulmonary BC and three with type 2b intrapulmonary BC. Two patients had documented fistulae between intrapulmonary BC and bronchial tree in surgery but only one had hemoptysis. Our analysis showed the significant correlation between the presence of any clinical symptom and the presence of surrounding infiltration (i.e. consolidation and/or ground glass opacity) or thick wall (category b) on CT images ($p=0.014$). Among these clinical symptoms, significant correlation between fever and category b was also noted ($p=0.042$). Patients with any clinical symptoms or fever had higher likelihood of lung parenchymal change surrounding intrapulmonary BC on CT.

All patients with type 2 intrapulmonary BC with the presence of air component (9/9, 100%) had at least one clinical symptom. In contrast, only seven of 11 intrapulmonary BCs without air component (63.6%) were symptomatic. However, the correlations of the presence of air component and the presence of any clinical symptom were not significant ($p=0.094$). Besides, the size of intrapulmonary BC was not correlated with clinical symptoms.

**CT patterns and histopathologic findings of lung parenchyma surrounding intrapulmonary BC**

The cysts were usually lined by ciliated columnar epithelium with variable acute and chronic inflammation. However, some of the components might be destroyed when infection is present for a long time. On histopathology, 12 of the 20 intrapulmonary BCs revealed chronic inflammation ($n=7$) (Figure 2) or pneumonic change ($n=5$) on adjacent lung parenchyma under microscopic evaluation. Among the 12 patients with histopathologic change in adjacent lung parenchyma, eight had thick wall, and 11 had surrounding consolidation or ground glass opacity on CT. In the eight patients without histopathologic change of surrounding lung parenchyma, only one had thick wall and none had surrounding consolidation or ground glass opacity on CT. The correlation of histopathologic findings (chronic inflammation or pneumonic change) of intrapulmonary BC with CT category b pattern was significant ($p=0.0008$).

**Discussion**

Intrapulmonary BC has a wide spectrum of imaging and clinical presentations. They are more common in the lower lung lobes, as shown also in our study. In this study, the subpleural region of the lower lobe (55%) was the most common location, followed by the mid-lung (30%) and perihilar (15%).

In this study, we proposed a new CT classification of intrapulmonary BC based on the attenuation of the cyst, the presence of air, the thickness of the cystic wall and surrounding lung parenchymal change. Type 1a cysts with a sharply defined thin-walled cyst and fluid attenuation content can be more easily diagnosed on enhanced CT when no systemic supplying vessel is found. Unfortunately, type 1a intrapulmonary BC (20%) was the minority type in our study. Type 2 intrapulmonary BC may be completely air-filled or with variable degrees of air–fluid level. Correct preoperative diagnosis of type 2 cysts may be difficult. Lung abscess could be the most common preoperative impression if there is thick cystic wall and pneumonic patch surrounding the cyst. Bulla could be impressed in the presence of a large amount of air in a thin-walled cyst. The content of BCs has a great variety of CT density, from thin watery liquid to hemorrhagic fluid, or to viscous mucoid material depending on whether there is hemorrhage, proteinaceous fluid or calcium. Soft tissue attenuation of the cystic content on precontrast study is not uncommon. Interestingly, type 3 cysts mimicked tumors on postcontrast CT images in our study. Theoretically, there should be no enhancement of the cystic content. It was presumed that the poor contrast between the cystic content
and wall of the type 3 cyst was due to inappropriate
timing of acquisition after bolus contrast injection
or poor enhancement of the wall and higher attenu-
ation of cystic content. The use of T2-weighted
MRI can reveal homogeneous high signal intensity
of the cystic content of BCs regardless of their CT
attenuation; hence, it makes the diagnosis of
type 3 intrapulmonary BCs easier. Unfortunately,
MRI is not routinely used in most circumstances.
Therefore, familiarity with the CT appearance of
intrapulmonary BCs is more important.

Emphysema of adjacent lung parenchyma in
adult intrapulmonary BC (100%) has been demon-
strated on thin-section CT. In contrast, there was
a very low incidence of pulmonary emphysema
(10%) in our study, although underestimation
might occur with thicker slice collimation. Further
study with thin-section CT is necessary to clarify
the findings.

Symptomatic intrapulmonary BCs were more
common than those without symptoms. It is not
uncommon for infants or children to
develop symptoms, such as fever and pneumonia.
Common clinical presentations included cough,
dyspnea, and no symptoms at all. Chest pain
and hemoptysis were only reported in adults,
while pneumonia was more common in children.
Productive cough and chest pain were the two most
common symptoms in our series. Hemoptysis in
patients with intrapulmonary BC was usually mild
and self-limited as in our study. Hemoptysis was
rarely severe and life-threatening in this entity.

From our analysis, CT findings of intrapul-
monary BCs explained their clinical presenta-
tions and were correlated with histopathologic
results. Consolidation and ground glass opacity
(category b) on CT were correlated with pneu-
monia or chronic inflammation on histopathol-
ogy. Patients with CT category b findings had
more clinical symptoms, especially with fever. The
size of intrapulmonary BCs was not correlated
with clinical symptoms. Cysts with air (type 2)
were larger than cysts without air. We presumed
that the presence of air in intrapulmonary BC
might be related to minute communication with
airways or superimposed infection of the cysts.

Interestingly, our results revealed that the presence
of air component (type 2) was not correlated with
clinical symptoms, although all type 2 cysts (9/9)
had at least one clinical symptom while only seven
of 11 cysts without air component were sympto-
matic. The small sample size in our study might
limit the statistical power in data analysis. It was
reported that a higher incidence of intrapulmonary
BC in infants and children was associated with
other malformations. However, only one adult
patient with intrapulmonary BC in our series had
associated intralobar pulmonary sequestration.

The differential diagnosis of pulmonary cystic
lesions with content of fluid attenuation should
include other foregut duplication cyst, congenital
cystic adenoid malformation, infected bullae,
bronchopulmonary sequestration, lung abscess
or lung tumor. By using CT or MR angiography,
demonstration of anomalous supplying artery to
the lung parenchymal lesion can establish the di-
agnosis of bronchopulmonary sequestration.

Surgical removal of BC has been considered to
be the treatment of choice with or without clinical
symptoms because of the potential complications
and risks, including infection, hemorrhage or
malignant change, of non-operated BC. The risks from surgery are minimal regardless
of whether it is thoracotomy or video-assisted
thoracoscopic surgery. Preoperative imaging
evaluation is important for guiding appropriate
strategy.

The major limitations of this study are the small
case number and retrospective review without
standard technique of CT study. Using our CT
classification of intrapulmonary BC, we demon-
strated the relationship between imaging and clinical
presentations or histopathologic findings, which has not been described in previous reports.

In summary, there is a wide spectrum of clinical
and imaging presentations in patients with intra-
pulmonary BC. Intrapulmonary BC with the
characteristic findings of thin-walled cyst with
water attenuation is in the minority. They may
look like lung abscesses, infected bullae or even
lung tumors. CT findings of intrapulmonary BC,
especially with surrounding lung parenchymal
change or thick cystic wall, are correlated with histopathologic findings of pneumonia and clinical symptoms, especially fever. Cysts with an air component were significantly larger than those without, but cyst size and air component of intrapulmonary BC were not correlated with clinical symptoms. Knowing the variety and relationship of CT patterns, histopathologic findings and clinical presentations of intrapulmonary BC is important for preoperative differential diagnosis and understanding this special disease entity.

References


