Cervical Bronchogenic Cysts

Usual and Unusual Clinical Presentations

Natacha Teissier, MD; Monique Elmaleh-Bergès, MD; Latifa Ferkdadji, MD; Martine François, MD; Thierry Van Den Abbeele, MD, PhD

Objective: To discuss the clinical, radiologic, and histopathologic characteristics of cervical bronchogenic cysts.

Design: Retrospective case study using a pathologic database at our institution.

Setting: Pediatric hospital.

Patients: Eight patients with cervical bronchogenic cysts were identified in the past 13 years (January 1994 to December 2007).

Main Outcome Measures: The patients' clinical presentations and surgical procedures are described.

Results: Two cervical bronchogenic cysts were located on the cervical anterior midline, 5 were anterolateral substernal, and the other was paraspinal. One corresponded to an intralaryngeal and extralaryngeal cyst. One was associated with an ectopic thymus. No patient had been diagnosed as having a bronchogenic cyst before surgery. No major surgical complications were noted. There was no relapse after surgery.

Conclusions: Although rare, cervical bronchogenic cysts are difficult to differentiate clinically from other cystic cervical masses because their location, radiologic characteristics, and evolution can mimic those of any other cervical mass. Cervical cysts are usually a pathologic finding, showing respiratory-type epithelium, cartilage, mucinous glands, and smooth muscle fibers. They result from abnormal development of the tracheobronchial tree. Some atypical locations or associations may be explained by embryologic origin. The curative treatment consists of complete surgical resection. To our knowledge, this study represents the largest pediatric series published about cervical bronchogenic cysts.


BRONCHOGENIC CYSTS ARE BENIGN CONGENITAL ABNORMALITIES OF THE EMBRYONIC FOREGUT. These cysts are frequently thoracic or mediastinal but may be cervical or abdominal. More than 70 cases have been reported in the literature, but the preoperative diagnosis is difficult to make. Cervical locations may suggest a diagnosis of thyroglossal duct cyst, lymphatic vascular malformation, dermoid cyst, branchial cleft cyst, or cervical thymic cyst. Clinical and radiologic criteria rarely allow a definitive preoperative diagnosis. Pathologic findings are usually necessary to differentiate these cysts from other cervical cysts. Pathologic analysis shows respiratory-type epithelium, cartilage, mucinous glands, and smooth muscle fibers.

We identified 8 cases of cervical bronchogenic cysts at our institution. Three patients had atypical presentations and are described herein. One cyst was located in the postero-cervical region, the second was associated with an ectopic thymus, and the third was intralaryngeal and extralaryngeal. Details of the other cysts are discussed.

METHODS

A 13-year (January 1994 to December 2007) retrospective case review was performed at our pediatric institution. Patients were selected from a pathologic database using the key words bronchogenic cyst in a search, which revealed 23 cases, 8 of which involved a cervical location. We identified 16 cysts that were thoracic or abdominal. Another 7 cysts had features similar to those of bronchogenic cysts but did not show cartilage formation and were excluded from the series. A cyst was considered bronchogenic if the pathologic examination identified respiratory-type epithelium, cartilage, mucinous glands, and smooth muscle fibers. All cases were histopathologically confirmed before inclusion in the series.

Two girls and 6 boys having cervical bronchogenic cysts were diagnosed at birth and up to age 3 years. Two patients manifested dyspnea and failure to thrive. One 2-month-old child coughed and had dysphagia. The other patients had an asymptomatic painless cervical mass. One mass was considered a thyroid cyst.

The children were assessed using computed tomography. In 1 child, magnetic resonance imaging was also performed to better appreciate the structure of the associated ectopic...
thymus. All except 1 cyst were unilocular. The cysts ranged from 0.5 to 3 cm in diameter.

Surgical removal was usually performed a few days or months after discovery of the cyst. The surgical excision was complete in all cases, and no major complications were noted after surgery. No nerve damage was identified. We have encountered no relapses during follow-up. The clinical characteristics of each case are summarized in the Table.

### Table. Clinical Characteristics of 8 Cases of Cervical Bronchogenic Cysts

<table>
<thead>
<tr>
<th>Case No./Sex/Age at Discovery of Cyst</th>
<th>Symptoms</th>
<th>Location of Cyst</th>
<th>Particularity</th>
<th>Pathologic Findings in Addition to Respiratory-Type Epithelium</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/2 mo</td>
<td>Cough, dysphagia</td>
<td>Pretracheal</td>
<td>Necessity of tracheoplasty</td>
<td>Cartilaginous nodules, mucinous glands</td>
</tr>
<tr>
<td>2/M/1 y</td>
<td>. . .</td>
<td>Left suprasternal</td>
<td>. . .</td>
<td>Cartilaginous nodules, seromucinous glands, and smooth muscle fibers</td>
</tr>
<tr>
<td>3/M/3 y</td>
<td>. . .</td>
<td>Right suprasternal</td>
<td>Intrathyroid cyst</td>
<td>Cartilaginous arches, seromucinous glands</td>
</tr>
<tr>
<td>4/M/2 wk</td>
<td>Dyspnea</td>
<td>Right suprasternal</td>
<td>Adherence to the trachea</td>
<td>Cartilaginous nodules, seromucinous glands, and smooth muscle fibers</td>
</tr>
<tr>
<td>5/F/1 mo</td>
<td>Dyspnea</td>
<td>Anterolateral to the trachea, descending to the carina</td>
<td>. . .</td>
<td>Cartilage, seromucinous glands, and smooth muscle fibers</td>
</tr>
<tr>
<td>6/F/1 mo</td>
<td>. . .</td>
<td>Posterior neck mass</td>
<td>Adherence to C2</td>
<td>Cartilaginous rings, mucinous glands</td>
</tr>
<tr>
<td>7/M/At birth</td>
<td>. . .</td>
<td>Laterocervical</td>
<td>Association with an ectopic thymus</td>
<td>Cartilaginous nodules, mucinous glands</td>
</tr>
<tr>
<td>8/M/2 wk</td>
<td>Stridor</td>
<td>Intralaryngeal and extralaryngeal</td>
<td>Necessity of a laryngoplast</td>
<td>Cartilaginous nodules, mucinous glands</td>
</tr>
</tbody>
</table>

Abbreviation: Ellipsis, not applicable.

Surgical excision of the mass was performed through a 4-cm transverse incision at the apex of the cyst. Dissection was simple except for a fibrous tract adherent to the lateral mass of the second cervical vertebra. Its content was mucinous and thicker than expected in cystic lymphangioma. Pathologic examination revealed a bronchogenic cyst bordered with respiratory epithelium rich in mucinous glands. The conjunctive wall was characterized by a hyaline cartilage segment.

### CASE 6

Case 6 in the Table was 1-year-old girl who had an 11-month history of an asymptomatic soft posterocervical mass located just below the posterior border of the mastoid. There had been no increase in size or inflammatory event. Ultrasonography identified 2 adjacent cysts, with the largest measuring $47 \times 23 \times 17$ mm. Magnetic resonance imaging confirmed the absence of intracranial communication and the presence of a liquid signal of the cyst and identified a small membrane at the posterior part of the cyst (Figure 1). The radiologist concluded that the small membrane was cystic bilocular lymphangioma.

Surgical excision of the mass was performed through a 4-cm transverse incision at the apex of the cyst. Dissection was simple except for a fibrous tract adherent to the lateral mass of the second cervical vertebra. Its content was mucinous and thicker than expected in cystic lymphangioma. Pathologic examination revealed a bronchogenic cyst bordered with respiratory epithelium rich in mucinous glands. The conjunctive wall was characterized by a hyaline cartilage segment.

### CASE 7

A 4-month-old boy was referred to our institution from Martinique for evaluation of 2 right laterocervical masses discovered at birth. The masses were clinically asymptomatic. The upper cervical mass, located just below the parotid, was smooth and painless. The lower one, located at the upper part of the thyroid gland, seemed more cystic. Ultrasonography confirmed the existence of the 2 structures. The lower mass in the thyroid region measured $31 \times 23 \times 23$ mm and pushed inwardly on the right thyroid lobe and the carotid artery and outwardly on the jugular vein. It contained membranes, and its aspect suggested cystic lymphangioma. The other mass was located above and was between the carotid artery and the jugular vein and extended up to the submaxillary gland. Its echostructure was comparable to that of the thymus gland, with the thymus being in its normal position, suggesting an ectopic thymus. Magnetic resonance imaging confirmed the presence of 2 masses and their locations (Figure 2). During the surgical procedure, the dissection planes of the lower cervical mass were not as clear as those of the ectopic thymus because there was a fibrous shell around the cyst without any clear fibrous tract. The cyst had a mucous content (Figure 3).
Pathologic examination confirmed the association of an ectopic thymus and a bronchogenic cyst. The cysts contained the characteristic structures, including mucous glands, hyaline cartilage, and respiratory-type epithelium (Figure 4).

CASE 8

A 2-week-old 2-kg neonate was referred because of increasing stridor with feeding difficulties. Flexible laryngoscopy demonstrated a left-sided intraluminal cyst obstructing 75% of the laryngeal diameter, as well as reduced mobility of the left vocal fold. Cervical ultrasonography identified a voluminous cyst extending from the left subvocal cord region through or around the thyroid cartilage to the suprathyroid region. The left thyroid lobe could not be located, suggesting a thyroid teratoma or a fourth branchial anomaly. However, endoscopy performed under general anesthesia did not reveal a piriform sinus fistula, and the patients plasma level of $\alpha$-fetoprotein was normal. Complete surgical removal of the cyst required an open laryngeal procedure with careful dissection under microscopic guidance from the extralaryngeal left region through the thyroid cartilage to the subglottic region. The cyst contained a mucinous liquid. The recurrent nerve was identified using the microscope. Conchal auricular cartilage was used to cover the lateral wall of the larynx. Pathologic examination confirmed the presence of respiratory-type epithelium, smooth muscle fibers, and cartilage.

COMMENT

To our knowledge, this case study represents the largest pediatric series published about cervical bronchogenic cysts. Mehta et al described a series of 24 patients 18 years or younger with bronchogenic cysts, only 4 of which had a cervical location. An important retrospective study covering 20 years was performed by Hsieh et al among 331 pediatric patients with cervical cysts. They found that the most common pediatric cysts were thyroglossal duct cysts (55%), followed by cystic hygromas (25%), and branchial cleft cysts (16%), while bronchogenic cysts composed less than 1% of the whole series. Only 1 thymic cyst was noted, and 9 cases were unclassified. The slight male predominance is in accord with our series' findings (6 boys and 2 girls).

Congenital bronchogenic cysts result from an anomaly of tracheobronchial development. The tracheobronchial tree develops at days 24 to 36 of gestation from an initial median bulge that appears on the ventral wall of the pharynx. At days 28 to 30, the lung buds elongate, forming the primary bronchi. Progressive subdivision of the lung buds allows growth of several segmental bronchi. This development continues during the first 2 years of life and beyond.

Abnormal budding of the bronchial tree may lead to the development of bronchogenic cysts. These cysts are usually intrathoracic in the anterior mediastinum or around the hilum. Some authors believe that the cyst may also originate from a preformed cyst of the thoracic cav-
Radiologic findings can be useful to differentiate bronchogenic cysts from other cervical cysts but may not always confirm the diagnosis. Ultrasonography will identify a unilocular fluid-filled cystic mass. Computed tomography will reveal an encapsulated mass as showing no contrast enhancement. The fluid within a bronchogenic cyst is a mixture of water and proteinaceous mucus. However, the content is variable, explaining differences in attenuation obtained on computed tomography. Furthermore, biochemical analysis of the fluid has revealed the presence of calcium. Magnetic resonance imaging has shown that high attenuation was not due to calcium but to mucus and proteinaceous debris. Indeed, magnetic resonance imaging may be helpful to delineate a bronchogenic cyst based on an increased signal in T2-weighted images. McAdams et al. established radiologic guidelines to aid in the diagnosis of bronchogenic cysts, including the presence of a well-defined thin smooth wall suggesting a cyst and homogeneous areas of high attenuation on computed tomography that do not enhance with administration of contrast material. Magnetic resonance imaging may be useful to differentiate high-attenuating cysts on computed tomography from soft-tissue masses. Bronchogenic cysts are usually isointense or hyperintense to cerebrospinal fluid in all pulse sequences. McAdams et al underscore the importance of analysis of the cyst wall to rule out a malignant component. These elements may help orient the diagnosis before surgery. However, it can be difficult to obtain good-quality magnetic resonance images in young children without sedation. Therefore, computed tomography before surgery may suffice because this examination outlines all adjacent vital structures.

Surgery allows a complete cure of bronchogenic cysts and is the treatment of choice in children. The surgical risk is usually low, and excision allows confirmation of the diagnosis. Percutaneous catheter drainage and sterile alcohol ablation have been performed in selected high-risk patients, particularly in adults, but seem to have no place in pediatric therapeutics, in contrast with other entities such as lymphangiomas. Infection, hemorrhage, or neoplasia within the cyst are potential risks of late complications.

Malignant transformations have been reported in adults. These include adenocarcinoma, leiomyosarcoma, anaplastic carcinoma, and mucoepidermoid carcinoma but have not been reported in children.

Compared with other cervical masses, bronchogenic cysts are rare. Their clinical presentation is classically a cervical noninflammatory mass manifesting during childhood. Although radiologic examination has become more common and radiologic criteria more defined, diagnosis of a bronchogenic cyst is seldom made before surgical resection, and histologic examination may be the only way to confirm its nature. In children, surgery remains the best therapeutic option and is mandatory when symptoms appear such as dysphagia or dyspnea due to compression of the trachea. Recur-
REFERENCES


