Gross mural cartilage in a congenital bronchogenic cyst: MRI features

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ÖZET
Konjenital bronkojenik kist olgusunda makroskobik kıkırdak dokusu: MRG bulguları

Doğum sonrasında kardiyak üfürümü olan hasta ekokardiyografi ile incelendi. Rastlantısal olarak, üst mediastende kalp ile ilişkisi olmayan 1 cm çapında kistik alan saptandı. Manyetik rezonans görüntüleme (MRG)'de üst mediastende ve alt servikal bölgede 4 x 4.5 x 8 cm boyutunda kist izlendi. Ameliyat esnasında, MRG'de kistin duvarına yapışmış sert kartilaj dokusu ayırt edilebilmiştir. Bronkojenik kist ile birlikte bu şekilde gross kartilaj dokusu daha önceki çalışmalarda rapor edilmemiştir. Sonuç olarak bronkojenik kistler, duvarında gross kartilaj dokusuna sahip olabilir ve ekstratorasik boşluklara uzanarak, teratom ve kistik lenfanjiyomları taklit edebilir.

Anahtar Kelimeler: Bronkojenik kist, kartilaj, manyetik rezonans görüntüleme, mediastinal kist.

SUMMARY
Gross mural cartilage in a congenital bronchogenic cyst: MRI features

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An infant who had a cardiac murmur after the delivery was evaluated with echocardiogram. A 1 cm cystic area was incidentally identified in the upper right mediastinum, that was not related with the heart. A 4 x 4.5 x 8 cm cystic mass in the superior mediastinum and lower neck was demonstrated on the magnetic resonance imaging (MRI). At surgery, a small solid structure corresponding to the nodule seen on MRI was identified, which was firm and cartilaginous in nature and attached to the wall of the cyst. Grossly visible cartilage in association with a bronchogenic cyst has not been previously reported. In conclusion, the bronchogenic cysts can have grossly visible cartilage in their wall and extend to extrathoracic spaces in a contiguous fashion, mimicking other entities such as teratomas and cystic lymphangiomas.

Key Words: Bronchogenic cysts, cartilage, magnetic resonance imaging, mediastinal cysts.
Mediastinal foregut cysts make 21% of all primary mediastinal tumors in children (1,2). Computed tomography (CT) and magnetic resonance imaging (MRI) are very helpful for identifying and characterizing the cyst and showing its extent. We report a case of bronchogenic cyst with cartilage in its wall that was demonstrated radiologically, surgically and pathologically.

CASE REPORT

An infant who had a cardiac murmur after the delivery was evaluated with echocardiogram. The echocardiogram showed a patent ductus arteriosus, which later closed spontaneously. A 1 cm cystic area that was not related with the heart was incidentally identified in the right upper mediastinum, and further assessment was recommended.

Figure 1. T1-weighted coronal (a), T2-weighted coronal (b), postcontrast T1-weighted coronal (c) and axial (d) MRI of the mediastinum and neck show a large cystic mass measuring 8.3 x 4.5 cm. The mass is in the right superior mediastinum to the right of the aortic arch and posterior to the brachiocephalic vein and superior vena cava. It has low signal intensity on the T1-weighted image (a), and high signal intensity on the T2-weighted image (b). The heterogeneous signal on the T2-weighted image (b) is secondary to pulsations of adjacent vessels. On post-contrast images, the mass demonstrates an eccentric enhancing nodule attached to the medial wall (arrows).
An MRI was performed and showed a 4 x 4.5 x 8 cm cystic mass with lobulated contours in the superior mediastinum and lower neck. The mass was located in the right side of the mediastinum and extended into the lower neck. There was no vascular encasement but marked compression of the superior vena cava and trachea was noted (Figure 1). The mass was entirely cystic except for a 7 mm nodule on its superomedial wall. This nodule demonstrated mild enhancement with gadolinium. A resection of the mass was performed via a right thoracotomy and supraclavicular approach. At surgery, a small solid structure corresponding to the nodule seen on MRI was identified, which was firm and cartilaginous in nature and attached to the wall of the cyst. After an uneventful postoperative course, the patient was discharged home in stable condition.

The histopathologic examination was consistent with bronchogenic cyst and the nodule represented mature cartilage. A mature teratoma was ruled out.

DISCUSSION

Congenital mediastinal foregut cysts develop as a result of a budding or branching abnormality of the primitive foregut and have three subgroups:
1. Bronchogenic cysts (54-63%),
2. Esophageal duplication cyst, and
3. Enteric cysts (1,2).

Bronchogenic cysts are lined with columnar respiratory epithelium and filled with mucoid material. Mucous glands, cartilage and muscular elements are frequently found in the cyst wall. The typical locations of bronchogenic cysts are pericarinal (52%), paratracheal (19%), esophageal wall (14%), and retrocardiac (9%) regions (2). Stridor and dysphagia are the most common presenting symptoms. Esophageal duplication cysts contain no cartilage and are lined by gastrointestinal tract epithelium (1,2). Enteric cysts may be connected to the meninges through a midline defect and are frequently associated with vertebral anomalies. Significant overlap of histologic features between these subgroups may render a specific diagnosis impossible, when the cyst is named as an undifferentiated foregut cyst (2-5).

Differential diagnosis of a cystic mediastinal mass in the newborn period includes foregut cysts, cystic hygroma, thymic cyst, teratoma and cystic neurogenic tumors (1-3). Distinction among these entities is primarily based on the location and histologic findings, and a specific preoperative diagnosis may not always be possible.

Our case was unique in that a cartilaginous nodule mimicking a mass was seen on the MRI, which made us consider neoplastic entities such as a mature teratoma and cystic schwannoma in the differential diagnosis. Presence of cartilaginous and other respiratory elements in the cyst wall is the histologic hallmark of the bronchogenic cyst but even microscopic examination does not show cartilage in all cases (6-10). Grossly visible cartilage in association with a bronchogenic cyst has not been previously reported.

This case was also unusual in that the cyst involved both the mediastinum and the neck. Cystic lymphangiomas are known to extend from one compartment to the next, but contiguous intrathoracic involvement by bronchogenic cyst has not been reported before. Approximately 80% of the bronchogenic cysts are intrathoracic and mediastinal in location. Extrathoracic bronchogenic cysts are known to occur in the neck, abdomen and retroperitoneum, but these are not associated with an intrathoracic cyst (2).

The appearance of the bronchogenic and other foregut cysts on imaging studies can be variable. Large cysts with septations and/or lobulations, as seen in this case, are not uncommon (2,11,12). Interestingly, majority of the bronchogenic cysts show soft tissue-like attenuation on CT studies. This is apparently due to the high protein content of the mucoid cyst fluid and may lead to confusion of the bronchogenic cyst with solid tumors (11). On MRI, likewise, high T1 signal may be observed secondary to protein in the cyst fluid (11). In our case, no such difficulty was encountered since the cyst demonstrated fluid signal on all pulse sequences. Occasionally, fluid-fluid levels may be present within the cyst on MRI or CT studies (2,11,12), which was not seen in our case.
In conclusion, bronchogenic cysts can have grossly visible cartilage in their wall and extend into extrathoracic spaces in a contiguous fashion, mimicking other entities such as teratomas and cystic lymphangiomas.

REFERENCES