A Case of Tracheoesophageal Cyst in the Posterior Mediastinum

Un-Jun Hyoung¹, Ki-Sup Chung¹, Young-Yun Park² and Kwang-Kil Lee²

We experienced a case of a tracheoesophageal cyst in the posterior mediastinum of a three-year-old girl, who complained of cough and fever. We confirmed this case by computed tomography and pathologic examination after surgical resection. A brief review of the literature is presented.

Key Words: Tracheoesophageal cyst

A large number of histologically different cysts and tumors arise from many anatomical structures located within the mediastinum but the incidence is lower than that in other parts of the body (Oldham 1971).

Mediastinal cysts and tumors are classified into two groups, congenital and acquired. Congenital types are epidermoid cysts, dermoid cysts, pericardial celomic cysts, bronchogenic cysts, esophageal cysts, gastroenteric cysts, cystic lymphangiomas, teratomas and rarely, mixed tumors. Acquired types are cysts or tumors due to parasites and hematomas, leukemias and lymphomas (Bower et al. 1977) (Table 1). The usual locations of common and rare mediastinal masses are presented in Table 2. Mediastinal cysts are less frequent than tumors, and mixed cysts are so extremely rare that only a few cases have been reported in the world (Le Roux 1960).

This report is a case of a three-year-old female who was confirmed pathologically with a tracheoesophageal cyst in the posterior mediastinum, in which the two components of esophageal and bronchogenic cysts were mixed.

CASE

A three-year-old female patient was admitted to our hospital with chief complaints of cough for 2

Table 1. Classification of the mediastinal mass embryologically (cited from Bower et al. 1977)

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Acquired</th>
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</thead>
<tbody>
<tr>
<td>Epidermoid cyst</td>
<td>Cysts due to parasite</td>
</tr>
<tr>
<td>Dermoid cyst</td>
<td>Leukemia</td>
</tr>
<tr>
<td>Pericardial celomic cyst</td>
<td>Lymphoma</td>
</tr>
<tr>
<td>Bronchogenic cyst</td>
<td>Hematoma</td>
</tr>
<tr>
<td>Esophageal cyst</td>
<td></td>
</tr>
<tr>
<td>Gastroenteric cyst</td>
<td></td>
</tr>
<tr>
<td>Cystic lymphangioma</td>
<td></td>
</tr>
<tr>
<td>Teratoma</td>
<td></td>
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<tr>
<td>Mixed cyst and tumor</td>
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</tbody>
</table>

months and fever for 5 days. There was no contributory past and family history.

On physical examination, she was mentally alert with the following vital signs: body temperature 38.0°C, pulse rate 118/min, and respiratory rate 24/min. Her developmental and nutritional status was good but she looked very ill at the time. The chest wall expanded symmetrically without retraction. Breath sounds were coarse and there were moist rales on the right lung field. Her heart and abdomen were normal.

The chest X-ray revealed a round mass in the posterior mediastinum, 5.0cm in diameter, with no evidence of calcification (Fig. 1). The esophagogram revealed displacement of the esophagus to the right side in the lower portion (Fig. 2) and the computerized tomogram of the chest revealed a 5.0×4.0cm sized, sharply defined, round cystic mass in the left side of the posterior mediastinum. It was extended into
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Table 2. Usual locations of a mediastinal mass (cited from Bower et al. 1977)

<table>
<thead>
<tr>
<th>Location</th>
<th>Anterosuperior</th>
<th>Middle</th>
<th>Posterior</th>
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</thead>
<tbody>
<tr>
<td>Common</td>
<td>Teratoma</td>
<td>Lymphoma</td>
<td>Neurogenic tumor</td>
</tr>
<tr>
<td></td>
<td>Thymus</td>
<td>Lymph nodes</td>
<td>Duplication</td>
</tr>
<tr>
<td></td>
<td>Hyperplasia</td>
<td>Granuloma</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Cyst</td>
<td>Bronchogenic cyst</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lymphangiosarcoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Lymphangioma</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hemangioma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rare</td>
<td>Substernal thyroid</td>
<td>Pericardial cyst</td>
<td>Pheochromocytoma</td>
</tr>
<tr>
<td></td>
<td>Thymic tumor</td>
<td></td>
<td>Anterior meningocele</td>
</tr>
</tbody>
</table>

**Fig. 1.** Chest PA and left lateral chest showing an approximately 5.0cm sized lobulated mass like shadow which does not obliterate the left cardiac border.

the middle mediastinum and displaced the left main bronchus (Fig. 3).

On the 7th hospital day, an aspiration biopsy was done with a 20-gauge aspiration needle. The pathologic findings of aspirates revealed many macrophages and other inflammatory cells, but malignant cells were not seen.

On the 10th hospital day, the patient was transferred to the Department of Chest Surgery for an operation. A 10.0×7.0×7.0cm sized, round, smooth surfaced cyst was found between the IVC and right hilum. It was attached to the left lower bronchus and the esophageal longitudinal muscle was inserted in the wall of the cyst, but there was no connection with the esophageal lumen. The inner surface was similar to the inner esophageal surface grossly and mucinous material was found in the cyst. Microscopic findings revealed that it was lined by ciliated pseudostratified columnar epithelium and contained cartilage plates and seromucinous glands in its wall (Fig. 4). In the other site, the stratified squamous epithelium lined the cyst, and beneath it, two muscle layers were present (Fig. 5).
Fig. 2. Esophasogram showing the displacement of the esophagus to the right side at the lower portion.

DISCUSSION

A large number of histologically different cysts and tumors arise from the many anatomical structures located within the mediastinum, whether neoplastic or inflammatory in condition (Oldham 1971). The incidence of cysts and tumors in the mediastinum is mostly congenital. The most common type is the dermoid cyst and the incidence of cysts originating from the foregut embryologically accounts for 10 percent.

Kirwin et al. (1973) used the term ‘cystic foregut derivative’ which includes a group of different lesions which have been called bronchogenic cysts, tracheobronchial cysts, esophageal cysts, gastric cysts, enterogenous cysts, and tracheoesophageal cysts.

Embryologically, congenital cysts originate from the notochord which begins to appear in the third week of embryonic life. Occasionally the notochord
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Fig. 4. Microscopic findings: The cyst is lined by ciliated pseudostratified columnar epithelium and contains cartilage plates and seromucinous glands in its wall.

Fig. 5. Microscopic findings: Stratified squamous epithelium lined the cyst. Beneath it, two muscle layers are present.
splits so that a gap is left in the dorsal tissue mass between the two halves of the notochord (Fig. 6). Through this gap, the ventrally situated yolk sac or gut anlage may be herniated and become adherent to the dorsal ectoderm or skin anlage. Bently and Smith (1960) named this phenomenon 'split notochord syndrome'. It may give rise to a variety of abnormal endodermal remnants, depending on which part of the dorsal endodermal fistula persists. The lesions which may occur are a dorsal enterocutaneous fistula, a dorsal sinus, and a posterior mediastinal cyst (enteric cyst). The respiratory system develops as a median ventral diverticulum of the foregut. From this diverticulum arises the tracheobronchial tree and the respiratory epithelium of the alveoli. Abnormal budding of the bronchial tree may give rise to abnormal cystic structures with or without a communication with the bronchial tree. These cystic structures are called bronchogenic cysts. After the fourth week, the embryonic esophagus elongates fairly rapidly. The lining epithelium perforates, converting the esophagus into an almost solid tube. At about six weeks, vacuoles develop within this solid tube. These vacuoles gradually coalesce to form the esophageal lumen. One vacuole may persist, however, giving rise to an intramural esophageal cyst.

Only a few cases of tracheoesophageal cysts have been reported in the world. And it is known that it is the most rare type of the cyst of the mediastinum (Laipply 1945; Guilley 1937; Adams and Thornton 1943; Cassel et al. 1950). The pathogenesis of the tracheoesophageal cyst is unknown, but there are two theories. The one is that sequestration of entodermal cells in the region of the primitive laryngotracheal groove would result in a cyst containing tracheal and esophageal elements (Abell 1956). The other theory is that a tracheoesophageal fistula could become closed off at each end and a mixed cyst could result, which could then be termed tracheoesophageal cyst (Maier 1948). Some of the paraesophageal bronchogenous cysts that have been reported may be of this type.

The collected series of mediastinal masses indicate that an infant or child is more likely to have symptoms than an adult. This may be due to the fact that children more often have malignant lesions (Ruebusch et al. 1973). It may also be due to the smaller thoracic capacity in children, which makes any lesion more likely to become symptomatic. In addition to size and invasiveness, symptoms depend on the location of the mass. The most common lesions in children are near the trachea or in the spinal canal, and therefore give symptoms at an early stage of development (Richard et al. 1977). Many patients have mild symptoms of cough, fever, wheezing, or bronchitis. But some cases needed intubation or tracheostomy due to airway obstruction (Haller et al. 1969).

Despite the inaccessibility of mediastinal structures, physical examination showing Homer's syndrome, increased deep tendon reflexes, or leg weakness can be informative in the diagnosis.

A variety of radiological techniques to investigate the mediastinum are available, but posteroanterior (PA) and lateral chest roentgenograms alone are often sufficient for diagnosis. In addition to localizing the lesion, plain chest X-ray films can give indirect evidence of the type of mass. Teratoma may show calcification in the chest roentgenogram. A barium swallow examination may help delineate a mediastinal mass, and esophageal displacement is sometimes demonstrated even when a mass cannot be appreciated on plain X-ray films. A myelogram is needed in patients with spinal cord symptoms, or when thoracotomy reveals foraminal tumor extension without spinal cord symptoms. A rare cause for a posterior mediastinal mass is anterior meningocoele, and this can be well delineated by a myelogram. The other diagnostic methods for mediastinal masses are mediastinoscopy, fungal titer and skin test, vanillymandelic acid level determination or bone marrow studies. A recent report states that approximately 50 percent of thoracic duplications contain ectopic gastric mucosa and can be diagnosed by sodium pertechnetate Tc99m scan (Ferguson et al. 1973). But the most available method for diagnosis of a mediastinal mass is computerized tomography of the chest or aspiration biopsy of the mass guided by ultrasonography (Goldwin et al. 1977; Livesay et al. 1979).

On histopathologic study of the tracheoesophageal cyst, the tracheal elements were obvious but the esophageal elements were part of this cyst.
unilocular, contained a mucilaginous fluid, and the wall varied in thickness. It was lined by ciliated pseudostratified columnar and stratified squamous epithelium. The smooth muscle layer consisted of two components. The one consisted of two muscle layers like the esophagus and the other consisted of one layer as in the trachea. And cartilage, bronchial and esophageal glands were seen in the wall of the cyst. In the absence of bronchial glands and cartilage, ciliated epithelium may not indicate a respiratory component. Ciliated epithelium is found in the fetal and adult esophagus and sometimes is observed in intra-abdominal gastrojejunal cysts (Evans 1929).

Treatment of the tracheoesophageal cyst involves removal of the cyst by operation, as for any other cyst of the mediastinum.

REFERENCES

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