



Cystic Mediastinal Masses in Children

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15.1 Introduction

Mediastinal masses in children are rare, consisting of a varied group of entities including congenital, infectious, and neoplastic lesions. According to the size of the mass, we can observe both small asymptomatic lesions and large lesions that may cause airway compression. Patients with large mediastinal mass typically present with respiratory symptoms. Both benign and malignant tumors occur in the mediastinum and include neurogenic tumor, lymphoma, germ cell tumor, thymoma, lipoma, etc.

Main cystic lesions of the mediastinum include bronchogenic cyst, thymic cyst, cystic lymphangioma, and esophageal duplication cyst.

Large cystic tumors are more likely to cause airway compression in children than adults because of the small size and more compressibility of the trachea. For this reason, prompt management of these lesions is necessary to avoid respiratory impairment [1].

Thoracic tumors can be incidentally diagnosed without clinical symptoms or can be associated

with various signs and symptoms, such as fever, cough, pneumonitis, chest pain, and a chest wall mass, or symptoms related to compression of adjacent structures such as the airway, esophagus, or superior central venous system. Compression of airway or mediastinal vasculature can be a life-threatening complication. Diagnostic evaluation usually starts with chest radiography. However, computed tomography (CT) (Fig. 15.1) and/or magnetic resonance imaging (MRI) is usually needed. CT is most indicated for parenchymal lesions, while MRI better investigates soft-tissue and cystic lesions, vascular anatomy, and posterior mediastinum tumors [2].

Standard treatment for cystic thoracic tumors in children consists of surgical resection. The traditional approach foresees thoracotomy or sternotomy (Fig. 15.2), but thoracoscopic technique is well established, both for diagnosis and treatment of mediastinal masses, and may be particularly useful in some cases, like benign neurogenic tumors of the posterior mediastinum [2].

15.2 Thymic Cyst

Thymic cysts are uncommon among the pediatric population, consisting of about 1% of all mediastinal masses. They can be divided into congenital and acquired lesions [3]. The median age of presentation is between 2 and 15 years, although they rarely present in adults [4].

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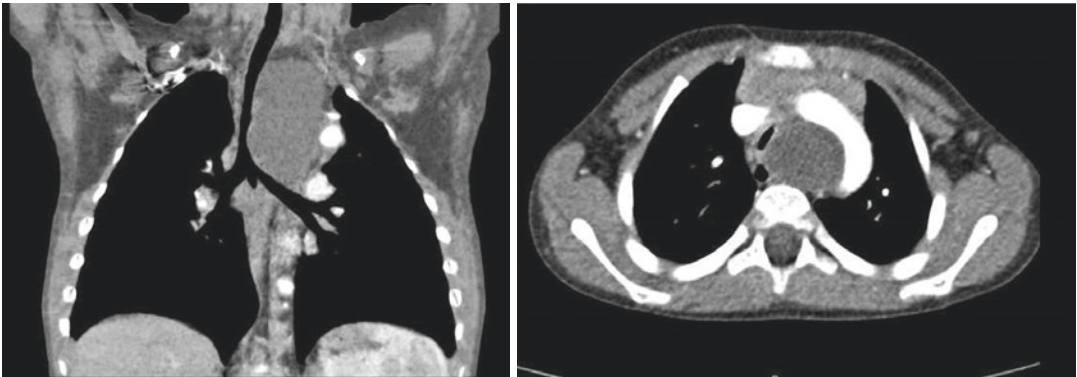


Fig. 15.1 mediastinal cyst, corresponding to a foregut duplication cyst: coronal and axial CT scan

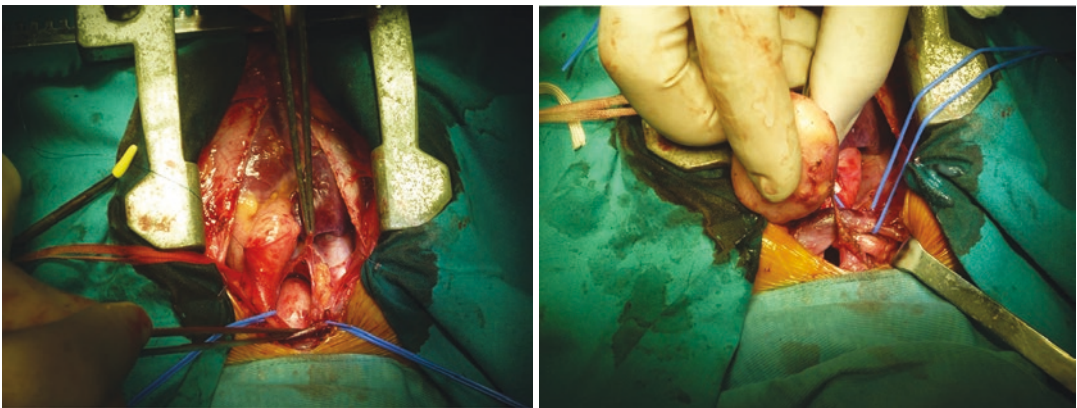


Fig. 15.2 surgical approach through sternotomy; removal of the cyst

Congenital cysts consist of a remnant of the thyropharyngeal duct, while those acquired may develop after radiotherapy in case of lymphoma or as inflammatory cysts seen in the context of autoimmune disorders; furthermore, they may also arise after thoracotomy procedures or may be associated with thymic tumors that can cause distortion or compression of normal thymic parenchyma [3].

Cysts may occur anywhere along the thyropharyngeal duct, from the pyriform sinus to the anterior mediastinum, but they are typically found in the lateral infrahyoid neck, with an intimate association with the carotid sheath, connected to the mediastinal thymus directly or by a fibrous cord.

Thymic cysts are usually asymptomatic; however, symptoms like dysphagia, respiratory dis-

stress, or vocal cord paralysis may be observed in the case of large lesions [4].

Thymic cysts are usually composed of a single large dominant cyst, smooth and thin-walled, and may have mural nodularity; they are lined by ciliated epithelium, with the presence of lymphocytes, thymic tissue, cholesterol crystals, and Hassall corpuscles within the wall [2]. Larger thymic cysts may present as dumbbell-shaped cervicothoracic masses, with the cyst passing through the thoracic inlet from the lower lateral neck into the superior mediastinum [4].

When a thymic cyst is suspected, ultrasonography (US), CT, and MRI are useful imaging investigations. US may be performed to characterize the fluid-filled mass, although CT and MRI have superior sensibility in showing the extent of the lesion and its relationship with the mediastinal

structures [2]. Needle aspiration has been an alternative proposed technique for making the diagnosis, but examination of surgically resected specimens is mandatory for diagnosis. Differential diagnoses of cervical thymic cyst include thyroglossal duct cyst, branchial cleft cyst, laryngocele, lymphovascular malformations, benign tumors (dermoid and epidermoid cysts), and malignant tumors [3].

Radical surgical excision allows an excellent prognosis [2].

15.3 Bronchogenic Cyst

Bronchogenic cysts are the result of an abnormal budding of the tracheal diverticulum during the formation of the foregut. They are more frequent in males and can be located anywhere along the tracheoesophageal tree, although they tend to occur near the carina or right paratracheal regions. Intrapulmonary location accounts for about 20% of the cases [4, 5]. Bronchogenic cysts may be considered a part of a spectrum of anomalies, linked to anomalous lung and foregut development. These sometimes share some characteristics and histologic findings and include congenital cystic adenomatous malformation (CCAM), bronchopulmonary sequestrations, congenital lobar emphysema, bronchogenic cysts, esophageal duplication cysts, and neurenteric cysts. Some authors have proposed the term bronchopulmonary-foregut malformations to enclose all these anomalies [2].

In many cases, bronchogenic cysts are asymptomatic, and the discovery is incidental on chest radiography, or as a space-occupying lesion or as a cyst with an air-fluid level. They become symptomatic concurrently with the impairment of the airway or airway/lung infection. Airway obstruction is more typically seen in infants, while infection tends to present in older children. Symptoms related to airway alterations include stridor, wheezing, cough, nasal flaring, retractions, and intermittent cyanotic spells. Symptoms related to infection include fever, cough, hemoptysis, and recurrent pneumonia. Communication with the airway is rare, but they may tightly close to it.

There may also be associated vertebral anomalies [6]. CT demonstrates in most cases a spherical nonenhancing mass of variable attenuation with sharp borders. An air-fluid level is rarely present if there is open communication with the airway [5].

Visualization of respiratory epithelium on histology sample allows the definitive diagnosis of bronchogenic cyst [4]. The cysts are lined with respiratory tract ciliated columnar epithelium or cuboidal epithelium containing mucous glands [5–7].

Cysts are typically radically excised for diagnosis when discovered. Infectious complications are thought to be frequent enough to indicate elective excision. If a cyst presents acute infection, surgery should be delayed to start antibiotic treatment [2–8]. The procedure could be performed either by video-assisted thoracic surgery or by lateral thoracotomy, depending on the complexity of the cyst and the experience of the surgeon [2–5].

15.4 Esophageal Duplication Cyst

Esophageal duplication cysts are discussed in the chapter related to the esophagus.

15.5 Lymphangioma

Lymphangiomas, well known as microcystic/macrocystic lymphatic malformations, are congenital lesions that are characterized by an abnormal proliferation of lymphatic and vascular tissue. They occur in about 1 in 6000 births, and only about 1% are limited to the thoracic cavity [2]. Large cervical lymphatic malformations, however, may extend into the mediastinum in 2% up to 10% of cases [2]. Most lymphatic anomalies present in the first years of life. They are often asymptomatic, and they may present with dyspnea when compressing the airway or other vital mediastinal structures [4].

These malformations are mostly composed of large lymphatic cysts; they can also consist of thicker tissue with more prominent vascular elements. Lymphatic tissue has a thin endothelial

lining and may also contain smooth muscle cells. Isolated thoracic lymphangiomas are usually found in the anterior mediastinum, although they can be found in other compartments and the lung tissue as well. Cervical lymphangiomas sometimes extend into the posterior mediastinum [2].

Thoracic lymphatic malformations are often incidental findings on chest imaging. Symptoms, when present, are secondary to airway, lungs, or other mediastinal structures compression by the mass. They include cough, stridor, dyspnea, dysphagia, hemoptysis, superior vena cava syndrome, and Horner syndrome. Infection or collection of chyle in the pleura or pericardium may occur. Intralesional bleeding with rapid enlargement of the mass and sudden onset of symptoms has been described [2].

Thoracic lymphangiomas appear as homogeneous masses on chest radiography. US shows a lesion that is primarily cystic, sometimes containing debris or other signs of recent bleeding.

A CT or MRI image could be visualized enhancement of internal septa and cyst wall, without enhancement of central portions. When a hemorrhage or infection occurs, proteinaceous material within the cysts can be produced, and consequently, lymphatic malformation appears as a more complex fluid collection instead of a simple fluid-filled cystic mass [4].

The optimal therapy for symptomatic mediastinal lymphangiomas is radical excision, but it is widely agreed that vital structures should not be sacrificed during the procedure. Because often pathologic tissue remains after surgery and some lesions are simply not resectable because they involve vital structures, alternative strategies for therapy have been advanced. Intralesional injection of sclerosing agents is often used as a primary or secondary therapy [2].

15.6 Pericardial Cysts

Pericardial cysts arise in case of mesodermal lacunae fail to coalesce during pericardium development. These cysts are typically located at the cardio-phrenic angle in the middle mediastinum and are thin-walled with a flat meso-

thelial lining. Pericardial cysts are usually found incidentally on chest images. Because of the benign histology and the absence of symptoms, some investigators believe that the approach of pericardial cysts is observation. However, others authors, for their potential symptomatology, such as inflammation, justify surgical excision. In fact, cardiac tamponade, attributable to hemorrhage or effusion, has been reported. US and CT usually confirm the diagnosis. Surgical excision is usually safe and can be performed by thoracotomy or a thoracoscopic approach [2].

15.7 Neurenteric Cyst

Neurenteric cysts are rare congenital lesions resulting from the abnormal separation of gastrointestinal tract from the primitive neural crest. They are most commonly located in the posterior mediastinum and the spinal canal can be involved. Congenital vertebral anomalies such as spinal dysraphism are classically associated with neurenteric cysts [2].

Neurenteric cysts can be detected in any age-group (usually discovered during the first 5 years of life) and can be found anywhere from head to abdomen, but they are often located in the posterior mediastinum [9].

In the pediatric population, one-third of the patient with mediastinal cysts remains asymptomatic, while two-third present with alterations of the respiratory system. These cysts are usually benign, but due to their size, they can cause compression of the adjacent structures [9].

In case of respiratory symptoms or distress, neurenteric cyst is suggested by a chest radiograph showing cervical or thoracic vertebral anomalies and a posterior mediastinal cyst. Both CT and MRI have great sensibility in diagnosing the condition. Histologically, both neural elements and gastrointestinal epithelium are typically seen [4].

Neurenteric cysts can present with a broad spectrum of signs and symptoms and can be life-threatening. When the gastric epithelium lines the cyst, hemorrhage, anemia, and pain can be the

principal symptoms. The majority of children with these cysts presents with central nervous system symptoms, such as back pain, sensory/motor deficit, or gait disturbances.

Complete surgical resection by thoracotomy or thoracoscopy of these lesions is feasible. If an asymptomatic or less symptomatic lesion is discovered, elective excision is recommended if operative risks are not too high [9].

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