Mediastinal Masses in Children

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Residents in Radiology

The mediastinum is the most common location of chest masses in the pediatric population. Mediastinal masses may be nonvascular or vascular masses and represent congenital anomalies, infections, benign and malignant neoplasms, and pseudomasses. In both asymptomatic and symptomatic children with mediastinal masses, imaging plays a crucial role in making the diagnosis and developing a treatment plan. As in adults, mediastinal masses in pediatric patients are placed in one of three mediastinal compartments (anterior, middle, posterior) on the basis of the lateral chest radiograph (Fig. 1 and Table 1). Further characterization can be made with ultrasound, CT, and MRI. Combining the characteristic imaging appearances with clinical information (age, physical examination findings, and laboratory analysis) often can provide a precise diagnosis.

Anterior Mediastinal Masses

For practical purposes and to facilitate the diagnosis, anterior mediastinal masses in children can be classified on the basis of their density into the three categories: solid, fatty, and cystic lesions.

Solid Lesions

Prominent thymus (pseudomass)—The thymus is a bilobed encapsulated organ located anterior to the great vessels and pericardium. Its primary function is the maturation of T-lymphocytes. In infancy and childhood (until about age 5 years), the thymus has a quadrilateral shape with convex margins. After this time, the thymus gradually becomes more triangular and the margins straighten. By age 15 years, the margins of the thymus should be straight or concave.

On chest radiographs, the normal thymus is not visible in adolescents and adults. In normal infants and young children, however, the thymus may be very prominent and mistaken for a true mass (Fig. 2). However, the classic appearance of the “sail” sign, the morphologic appearance of a nautical sail, confirms that the thymus is normal (Fig. 3). Indentations produced by anterior ribs on the soft thymus (“wave” sign) are another normal pattern. Ultrasound, CT, or MRI can confirm the presence of a prominent but normal thymus. Even though CT is not typically used to evaluate the thymus, it is important to realize that this organ may appear prominent and suggest an anterior mediastinal mass to the unwary eye. Indeed, such a pseudomass may be the source of referral to a pediatric center when the chest CT study of an infant or young child is interpreted by an adult radiologist (Fig. 4). Cross-sectional imaging shows the thymus as a homogeneous soft-tissue structure without calcification or compression on the airway or vascular structures. The soft and pliable consistency of this organ can be confirmed at fluoroscopy because the normal thymus changes contour and size with respiration (Fig. 5).

A retrocaval thymus is a normal variant in which there is posterior extension of the thymus between the superior vena cava (SVC) and great arteries (Fig. 6). On chest radiography, a retrocaval thymus may produce a confusing appearance that mimics a true mediastinal mass or right upper lobe collapse. Cross-sectional imaging shows that the retrocaval thymus is contiguous with the anteriorly positioned normal thymic tissue and forms a single structure, with homogeneous attenuation or signal that is similar to the rest of the thymus. An ectopic cervical thymus, which results from arrested migration along the thyropharyngeal duct, may be found from the mandibular angle to the thoracic inlet. In this condition, imaging shows a mass of
homogeneous attenuation that is similar to that of the normally positioned thymic tissue.

Thymic hyperplasia—Thymic hyperplasia, or thymic rebound, refers to enlargement of the gland after atrophy from severe illness or medications (chemotherapy, corticosteroid treatment). After chemotherapy, the thymus atrophies in about 90% of cases. The thymic gland grows back over the next several months, and the volume of the rebounding thymus may exceed the baseline value.

In addition to CT evaluation of the size and shape of the thymus (Figs. 7A and 7B), MRI also can show a hyperplastic thymus, which is typically fatty and can be distinguished from the soft-tissue component of more ominous entities, such as lymphoma. Thymic rebound is seen routinely on follow-up PET studies of patients with neoplasms and should not be confused with a pathologic condition (Fig. 7C). A standardized uptake value of more than 4 on PET suggests malignancy, although there can be substantial overlap of benign and malignant processes.

Thymoma—Thymomas are epithelial neoplasms containing a variable amount of lymphocytes. Although representing about 20% of mediastinal tumors, thymomas are infrequent in the pediatric population, accounting for only 1–2% of mediastinal tumors. Thymomas may be discovered incidentally, although about one third of patients have symptoms related to local compression or invasion. About 40% of patients with thymomas present with a paraneoplastic syndrome, such as hypogammaglobulinemia; red cell aplasia; or, most commonly, myasthenia gravis.

Thymomas are typically classified as noninvasive or invasive thymoma. Noninvasive thymomas tend to have well-defined margins because they do not extend beyond their fibrous capsules (Fig. 8). In contrast, an invasive thymoma does extend beyond its fibrous capsule (Fig. 9), tending to spread locally to invade adjacent mediastinal structures and the chest wall. An invasive thymoma also may spread contiguously along the pleural surface, usually unilaterally, and often recurs after surgical removal.
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On chest radiographs, thymomas most often appear as oval anterior mediastinal masses that usually project on one side of the chest. They are often seen best on the lateral image, obliterating the retrosternal clear space. There may be thin linear peripheral capsular calcification. Invasive thymomas can present with pleural nodules or masses. CT confirms an oval or lobulated enhancing anterior mediastinal mass and may show thin capsular calcification. Cystic regions and necrosis are seen in about 30% of cases, particularly in larger tumors. Signs of invasion include irregular borders; obliteration of mediastinal fat planes surrounding mediastinal vascular structures; pericardial thickening; and extension to the chest wall, pleura, or diaphragm. On MRI, a thymoma typically has high signal intensity on T2-weighted images, but this modality usually does not provide more diagnostic information than does CT.

Encapsulated and invasive thymomas are histologically identical. Because the diagnosis of invasive thymoma is therefore based on visualization of gross or microscopic evidence of extension through the capsule, there must be surgical excision of the entire mass.

Thymic carcinoma—Thymic carcinoma is an aggressive epithelial carcinoma that is histologically characterized by malignant features of nuclear atypia, numerous mitotic figures, and necrosis. Rare in the pediatric population, thymic carcinomas usually occur in the fifth or sixth decades of life. Almost all affected patients are symptomatic at presentation, usually with chest pain and constitutional symptoms such as weight loss, fatigue, and night sweats.

Thymic carcinomas usually present as large irregular anterior mediastinal masses with aggressive local spread and mediastinal vascular invasion (Fig. 10). They often contain calcification and enhance heterogeneously with areas of necrosis. Thymic carcinoma has a poor prognosis because there is aggressive local growth and distant metastatic disease is common. Treatment options include neoadjuvant chemotherapy to improve the resectability of the tumor and postoperative radiation therapy.

Lymphoma—Lymphoma is the most common anterior mediastinal mass in children. Although Hodgkin lymphoma typically occurs before age 10 years, non-Hodgkin lymphoma is common in both the first and second decades of life. Hodgkin lymphoma, which is histologically characterized by the presence of Reed-Sternberg cells, has a good prognosis, with an approximately 90% cure rate. Non-Hodgkin lymphoma is due to clonal proliferation of either T or B cell origin. Lymphadenopathy from mediastinal lymphoma may cause cough, dyspnea, dysphagia, hemoptysis, or SVC syndrome. Nonspecific B symptoms, which are more common with aggressive lymphomas, are fever, weight loss, and night sweats.

About 85% of Hodgkin lymphomas show intrathoracic involvement at presentation, most commonly affecting the anterior superior mediastinal lymph nodes (Fig. 11). The nodes rarely calcify before treatment, and approximately 5% calcify after therapy. In addition to enlarged lymph nodes or nodal conglomerations, Hodgkin lymphoma may manifest as multiple pulmonary nodules and multifocal consolidation; about 15% of patients have pleural effusions.

In non-Hodgkin lymphoma, about 50% of cases show intrathoracic involvement (Fig. 12). The anterior and posterior mediastinal nodal chains are equally involved. Associated thoracic findings include pulmonary nodules that may cavitate, airspace consolidation, and diffuse interstitial thickening. Pleural involvement in non-Hodgkin lymphoma may manifest as effusions or pleural masses.
Fig. 2—Prominent but normal thymus in 1-week-old boy with cough and possible pneumonia. 
A, Frontal chest radiograph shows prominent thymus (arrows). There is no mass effect on trachea and mainstem bronchi. 
B, Lateral chest radiograph shows prominent but normal thymus (T) in anterior mediastinum, with no mass effect on trachea.

Fig. 3—Radiograph shows thymic sail sign in 4-day-old girl with respiratory distress. Note normal thymus (arrow) with shape similar to nautical sail.

Fig. 4—Prominent but normal thymus, which is incidental finding in 1-month-old boy who underwent CT examination for pulmonary sequestration. 
A, Contrast-enhanced axial CT image shows prominent but normal thymus (arrows), characterized by homogeneous attenuation and without mass effect on adjacent mediastinal structures. 
B, Contrast-enhanced coronal CT image again shows prominent but normal thymus (arrows), with no mass effect on adjacent great vessels or heart.
Chest radiographs in lymphoma show an anterior mediastinal mass with obliteration of the retrosternal clear space or mediastinal widening (Figs. 11 and 12). Close inspection of the trachea is valuable in differentiating an anterior mediastinal mass from normal thymus in younger children because even a prominent thymus should not exert mass effect on or displace the trachea.

On CT, lymphomatous involvement of the thymus often causes homogeneous enlargement of the gland. However, larger nodal conglomerates often become heterogeneous, with hypodense and cystic areas suggestive of necrosis (Fig. 13). MRI is not typically used to evaluate mediastinal lymphoma because CT is superb at depicting the anatomic extent of disease. PET is often added to CT both at the time of initial staging and to follow the response to treatment.

About 80% of patients with Hodgkin lymphoma can achieve disease-free survival. The current preferred treatment involves combining chemotherapy medications, which not only provides a synergistic effect but also limits the toxicity of individual drugs. Children with both types of lymphoma are treated with radiation therapy and chemotherapy; bone marrow transplantation is also used in those with non-Hodgkin disease.

**Teratoma**—Germ cell neoplasms arise from collections of primitive germ cells that arrest in the anterior mediastinum on their migration to the gonads during embryologic development. Because they are histologically indistinguishable from germ cell tumors arising in the testes and ovaries, the diagnosis of a primary malignant mediastinal germ cell neoplasm requires exclusion of a primary gonadal tumor as a source of mediastinal metastases. Teratoma accounts for approximately 60% of all germ cell tumors in the mediastinum. Most are asymptomatic; however, when large, affected patients may present with respiratory distress due to airway compromise. There are two main types of teratoma: immature malignant teratoma and mature benign teratoma.

On chest radiographs, both immature and mature teratomas usually present as round and sharply margined anterior mediastinal masses. Calcification, which occurs in about 25% of teratomas, may be central, peripheral, or curvilinear. The hallmarks of both immature and mature teratoma are fat, fluid, and calcified components on CT or MRI. The presence of fat,
fluid, and calcified components within an anterior mediastinal mass in a pediatric patient can help differentiate a teratoma from other types of mediastinal masses (Fig. 14).

Another valuable diagnostic feature is the presence of solid tissue within the lesion, a feature more commonly seen in immature malignant teratomas than in mature benign tumors. Mature benign teratomas tend to displace rather than invade adjacent structures (the latter appearance being more suggestive of malignant immature tumors). Mature benign teratomas have an excellent prognosis after complete excision. Conversely, immature malignant teratomas must be treated with multiple modalities, including chemotherapy and radiation, in addition to surgery.

Fig. 8—Noninvasive thymoma in 17-year-old girl who presented with cough and abnormal chest radiographs. Contrast-enhanced axial CT image shows heterogeneously enhancing anterior mediastinal mass with well-circumscribed borders and mass effect on adjacent heart.

Fig. 9—Invasive thymoma in 18-year-old woman with shortness of breath and weakness. Contrast-enhanced CT image shows heterogeneously enhancing anterior mediastinal mass with focal area (arrow) extending to subpleural region. Follow-up study after mass removal showed development of subpleural tumor nodules.
Cystic Lesions

Thymic cyst—A thymic cyst is a rare fluid-filled mass that represents a cystic remnant of the thymopharyngeal duct. Although typically found in the lateral infrahyoid neck and intimately associated with the carotid sheath, a thymic cyst may occur anywhere along the thymopharyngeal duct from the pyriform sinus to the anterior mediastinum. It may also be connected to the mediastinal thymus directly or by a fibrous cord. The pathologic visualization of Hassall corpuscles in the cyst wall confirms the diagnosis. Often asymptomatic, when sufficiently large a thymic cyst may cause dysphagia, respiratory distress, or vocal cord paralysis. Most affected patients present between 2–15 years of age, although thymic cysts rarely may present in adults.

Thymic cysts are usually smooth and thin-walled and may have mural nodularity. There is usually a large dominant cyst, although the lesion may be multiloculated. Larger thymic cysts may present as dumbbell-shaped cervicothoracic masses, with the mass traversing the thoracic inlet from the lower lateral neck into the superior mediastinum (Fig. 15).

Contrast-enhanced CT and MRI show a thymic cyst as a nonenhancing low-density cystic mass. The MRI signal depends on the contents of the cyst fluid, with proteinaceous or hemorrhagic
Fig. 12—Non-Hodgkin lymphoma in 14-year-old girl with low-grade fever and weight loss for 3 months.  
A, Frontal chest radiograph shows large mediastinal mass (arrow).
B, Lateral chest radiograph shows location of mass (asterisk) within anterior mediastinum.
C, Contrast-enhanced axial CT image shows mildly enhancing anterior mediastinal mass (asterisk).

Fig. 13—Hodgkin lymphoma with areas of necrosis or cystic change in 12-year-old girl with chronic cough. Contrast-enhanced axial CT image shows large anterior mediastinal mass containing areas of low attenuation (asterisks) that likely represent areas of necrosis or cystic change.

Fig. 14—Mature teratoma in 3-year-old boy with respiratory distress. Contrast-enhanced axial CT image shows large anterior mediastinal mass containing areas of soft tissue, fat (asterisks), and calcification (arrow). Mass is causing significant mass effect on heart and atelectasis of right lower lobe.
material appearing hyperintense on T1-weighted images. Ultrasound may be performed to characterize the fluid-filled mass, although CT and MRI are superior for showing the extent of the lesion and its relationship to the mediastinal structures. The prognosis is excellent if the lesion is completely resected.

Lymphatic malformation—A lymphatic malformation is a lymph-containing cystic or multicystic structure lined by endothelium. In the past, it was referred to as “lymphangioma” or “cystic hygroma” because the suffix “oma” was used to described neoplasms and malformations. However, because malformations are not neoplasms, words such as “lymphangioma” or “hygroma” are misnomers, and the terminology is being abandoned. It is now termed “lymphatic malformation” based on the currently accepted classification of vascular malformations by Mulliken and Glowacki. It may occur anywhere in the body and frequently involves the cervical region, with extension into the anterior mediastinum. Most lymphatic malformations present early in life. Often asymptomatic, they may present with dyspnea when compressing the airway or other vital mediastinal structures. Depending on the size of

Fig. 15—Thymic cyst in 7-year-old girl who presented with shortness of breath and abnormal chest radiograph. A, Contrast-enhanced axial CT image shows large hypodense cystic mass (asterisk) at level of thoracic inlet in expected location of thymus. Note mass effect on trachea (T). B, Contrast-enhanced coronal CT image better shows craniocaudal extension of mass (asterisk) in lower neck and upper mediastinum and mass effect on airway.

Fig. 16—Lymphatic malformation in 11-year-old boy with shortness of breath and chest pain. Contrast-enhanced axial CT image shows predominantly cystic anterior mediastinal mass with multiple internal septations (arrows) insinuating along mediastinal space and causing mild mass effect.

Fig. 17—Lymphatic malformation in 3-month-old boy with large right neck mass. Coronal T2-weighted image shows large predominately cystic mass with multiple internal septations. Although epicenter of lesion is in right side of neck, there is clear extension into superior mediastinum (arrow), finding commonly seen with these lesions.
the locules, lymphatic malformations may be classified as macrocystic or microcystic. Imaging studies usually reveal a multicystic mass that is smooth and well defined and contains septations and a variable number and size of cysts (Figs. 16 and 17). CT and, more frequently, MRI may show enhancement of internal septa and the cyst wall but no enhancement of central portions (Fig. 16). In the case of previous hemorrhage or infection producing proteinaceous material within the cysts, lymphatic malformations appear as more complex fluid collections instead of simple fluid-filled cystic masses. The primary treatment of lymphatic malformations includes surgical resection or percutaneous sclerotherapy, with the latter more commonly used for the macrocystic type.

**Fatty Lesions**

**Lipoma**—Lipomas are encapsulated masses with a composition identical to subcutaneous fat. They may occur anywhere in the body, including the mediastinum. Because lipomas are soft and pliable, patients are usually asymptomatic and the lesion is found incidentally on a chest radiograph or CT image obtained for another reason. Depending on the size and location of the lipoma, chest radiographs may show a mass that is relatively radiolucent compared with adjacent soft tissues. CT and MRI can confirm the fatty nature of the mass (Fig. 18).

**Thymolipoma**—A thymolipoma is an uncommon benign anterior mediastinal mass that consists of normal thymic tissue interspersed with fat. Because these are pliable fatty masses, patients are usually asymptomatic. A thymolipoma usually appears as a large and sharply marginated mass. Chest radiographs show the low density of the lesion in about one half of cases. CT and MRI can confirm the diagnosis by showing nonenhancing soft tissue interwoven within fat in a somewhat whorled appearance (Fig. 19). No treatment is necessary unless this benign tumor becomes massive and exerts mass effect on adjacent structures.

**Middle Mediastinal Masses**

For practical purposes, middle mediastinal masses in children can be classified into two groups: vascular and nonvascular lesions.

**Vascular Lesions**

**Double aortic arch**—A double aortic arch is a congenital aortic arch anomaly related to the embryologic persistence of both the left and right fourth aortic arches. It accounts for about 55% of symptomatic vascular rings. Because the trachea and esophagus are encircled, patients typically present with airway and esophageal compression, causing inspiratory stridor that worsens with feeding. Patients usually present soon after birth, and there is frequently associated tracheomalacia.
Chest radiography shows prominent soft tissue on either side of the trachea, which can result in an abnormal midline position or slight tracheal deviation away from the dominant arch. The right arch is more commonly higher and more prominent than the left. The lateral view shows anterior and posterior compression of the trachea at the level of the arch. Barium swallow studies show bilateral indentations on the contrast-filled upper esophagus, with focal circumferential narrowing at the expected level of the aortic arch (Fig. 20). On the lateral image, there is an oblique or nearly horizontal sharp indentation on the posterior esophagus. However, fluoroscopic studies rarely obviate cross-sectional imaging to provide the more defined anatomic definition required before surgery (Fig. 21). Each arch gives rise to a carotid and a subclavian artery, resulting in the symmetric “four-artery sign” on CT and MRI at the thoracic inlet. The smaller of the two arches may be partially atretic. Treatment of double aortic arch in symptomatic patients is surgical division performed on the side of the nondominant arch. Despite surgical treatment, approximately 30% of postoperative patients have persistent airway symptoms because of tracheomalacia or persistent extrinsic airway compression.

**Right aortic arch**—Right aortic arch results from the persistence of the right fourth embryologic arch. There are two main types of right aortic arch: right aortic arch with an aberrant left subclavian artery and right aortic arch with mirror image branching. A right aortic arch with an aberrant left subclavian artery is often an incidental finding, with about 95% of patients being asymptomatic. However, the presence of a left ligamentum arteriosum can result in a complete vascular ring. Patients with this condition may present with respiratory distress and dysphagia. Conversely, a right aortic arch with mirror image branching is associated with cyanotic congenital heart disease and usually does not cause respiratory distress. Chest radiographs show an aortic arch located to the right of the trachea deviating the trachea to the left and a right-sided descending aortic line. On the lateral image, there may be anterior bowing of the trachea or an indentation on its posterior aspect. Barium swallow studies can show an oblique filling defect coursing from right-inferior to left-superior, with the lateral image showing a posterior esophageal indentation. Aneurysmal dilatation of the subclavian artery origin may be present in up to 60% of patients with an aberrant left subclavian artery. Known as a diverticulum of Kommerell, it is well visualized on CT or MR angiography.
phy (Fig. 22). CT angiography is superior to MR angiography because it also provides exquisite evaluation of the airway and lungs; however, it is associated with ionizing radiation. An associated constricting left ligamentum arteriosum may cause stridor, requiring surgical division of this structure via a left thoracotomy.

Left aortic arch with aberrant right subclavian artery—The mirror image of a right aortic arch with aberrant left subclavian artery, a left aortic arch with aberrant right subclavian artery usually does not cause airway compression and is an incidental finding. Rarely, esophageal compression produces dysphagia lusoria (an abnormal condition characterized by difficulty swallowing).

A frontal chest radiograph is usually normal but occasionally may show mediastinal widening. On the lateral image, there may be a posterior mass effect on the trachea. Although a barium swallow study may show an oblique indentation on the posterior esophagus coursing from left-inferior to right-superior and suggesting the presence of an aberrant right subclavian artery, cross-sectional imaging studies such as CT and MRI can confirm the presence and show the entire course of an aberrant right subclavian artery (Fig. 23). Symptomatic patients usually require surgery to ligate the right ligamentum arteriosum.
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Pulmonary artery sling—Pulmonary artery sling, also known as anomalous origin of the left pulmonary artery, is a condition in which the left pulmonary artery arises from the right pulmonary artery rather than from the main pulmonary artery. It is the only vascular ring that courses between the trachea and esophagus and is associated with asymmetric lung aeration. Affected patients most commonly present with severe stridor and hypoxia.

A lateral chest radiograph may show a round soft-tissue density between the distal trachea and esophagus. On barium swallow studies, there is usually an anterior indentation on the esophagus. CT and MRI can confirm the diagnosis and provide preoperative delineation of the anatomy (Fig. 24). At surgery, the left pulmonary artery is separated from its anomalous origin and subsequently connected to the main pulmonary artery. If there is concomitant tracheal stenosis or malacia, the trachea also needs to be surgically repaired.

Duplicated superior vena cava—A left SVC results from persistence of the left common cardinal vein. It is usually associated with a normal or slightly smaller right SVC, resulting in a duplicated SVC that is most often an incidental finding but may be associated with congenital heart disease. A left-sided SVC typically drains into the coronary sinus.
On a frontal chest radiograph, a left-sided SVC often manifests as a straight border forming a prominent left paratracheal stripe. The presence of a left-sided SVC is often first detected when a left-sided central catheter or pacer lead is seen entering this vessel. CT and MRI show a tubular structure that originates from the junction of the left internal jugular and subclavian veins and courses along the left side of the mediastinum before draining into the coronary sinus (Fig. 25). No treatment is needed for a left-sided SVC, but note should be made of this anomaly in case the patient subsequently requires catheter placement or thoracic surgery.

**Nonvascular Lesions**

*Congenital foregut duplication cysts*—Intrathoracic foregut duplication cysts are uncommon congenital anomalies that result from developmental malformations of the embryonic foregut. They can be classified into three types: bronchogenic, esophageal duplication, and neuroenteric cysts.

Bronchogenic cysts are caused by abnormal lung budding and development of the ventral foregut during the first trimester. Although more than one half of pediatric patients with
bronchogenic cysts are asymptomatic, some patients may experience dyspnea if the lesion is large enough to exert mass effect on the adjacent airways in the mediastinum (Fig. 26). Bronchogenic cysts can be located anywhere along the tracheoesophageal tree, but they tend to occur near the carina or right paratracheal regions. About 20% are in an intrapulmonary location. The visualization of respiratory epithelium on histology provides the definitive diagnosis of bronchogenic cyst.

Esophageal duplication cysts result from abnormal development of the posterior division of the embryonic foregut (Fig. 27). Dysphagia is the most common presenting symptom. Although esophageal duplication cysts are typically located adjacent to the upper third of the esophageal wall, they may be found within the lung parenchyma if they...
become detached from the esophagus during development and subsequently migrate with a lung bud. The histologic visualization of a submucosa or muscular layer of the gastrointestinal tract within the lesion provides the definitive diagnosis of esophageal duplication cyst.

Neurenteric cysts result from failure of separation of the gastrointestinal tract from the primitive neural crest during early embryonic life. Most neurenteric cysts are found in the posterior mediastinum (Fig. 28) where they can extend into or communicate with the spinal canal and be associated with congenital osseous defects of the spine. Histologically, both neural elements and gastrointestinal epithelium are typically seen. Affected patients often present with pain, which usually leads to early imaging evaluation and subsequent diagnosis.

On imaging studies, all three types of foregut duplication cysts usually appear as solitary well-circumscribed round or oval cystic masses (Figs. 26–28). About one half of them show simple fluid attenuation on CT. However, foregut duplication cysts commonly contain proteinaceous material, which may have an attenuation value higher than water. There is no substantial internal contrast enhancement within a foregut duplication cyst unless there is superimposed infection. Complete surgical resection is the usual treatment of all foregut duplication cysts, particularly in symptomatic pediatric patients.

Lymphadenopathy—Lymphadenopathy in the middle mediastinal compartment in children is most commonly due to either neoplastic or infectious processes. Middle mediastinal lymph node masses are often malignant, reflecting either a primary neoplasm, such as lymphoma (which also can extend from the anterior mediastinum) or metastases from a distant tumor (Fig. 29). In children, metastatic lymphadenopathy often results from primary tumors in the abdomen and pelvis, such as Wilms tumor, testicular neoplasms, and various sarcomas (Fig. 30). Mediastinal metastatic lymphadenopathy usually appears on CT and MRI as homogeneous or heterogeneous soft-tissue masses that can represent a conglomeration of nodes. Determining the primary tumor responsible for metastatic mediastinal lymphadenopathy is often difficult. The presence of calcification suggests treated lymphoma or osteosarcoma metastases in pediatric patients. As with other mediastinal masses, patients with metastatic mediastinal lymphadenopathy may be asymptomatic or present with a variety of symptoms (such as dyspnea or chest pain) that depend on the extent of the lymphadenopathy and its mass effect on vital adjacent mediastinal structures.

The most common infectious causes of middle mediastinal lymphadenopathy in children are tuberculosis and histoplasmosis (Fig. 31). Affected patients generally also have pulmonary parenchymal abnormalities on imaging studies that aid in reaching a correct diagnosis. However, isolated lymph node enlargement may occur. Treatment consists of appropriate therapy of the underlying infection.

**Posterior Mediastinal Masses**

**Sympathetic Ganglion Tumors**

**Neuroblastoma**—About 90% of posterior mediastinal tumors are neurogenic, derived from the sympathetic chains that are located
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along the thoracic vertebral bodies. In pediatric patients, the vast majority of these neurogenic tumors are neuroblastomas; the rest are ganglioneuroblastoma or ganglioneuroma.

Neuroblastoma is a malignant tumor of primitive neural crest cells. Although most develop in the adrenal gland, neuroblastomas can arise anywhere along the sympathetic chain, particularly within the posterior mediastinum (about 20% of neuroblastomas in pediatric patients). The typical clinical presentation includes fever, irritability, weight loss, and anemia. Cord compression by the tumor may cause paraplegia, extremity weakness, and altered bowel or bladder function.

Chest radiographs often show soft-tissue opacity in the paravertebral region, sometimes associated with rib or vertebral erosion or destruction or widening of intercostal spaces. Calcification within the tumor is common, reportedly occurring in up to 30% of cases. Bone metastases from neuroblastoma are also common and usually appear as lytic and permeative. CT can delineate the margins of the tumor and internal calcification as well as local extension and distant metastases (Fig. 32). Because it is an invasive tumor, neuroblastoma tends to surround and

Fig. 31—Tuberculosis in 12-year-old boy with cough and low-grade fever who underwent chest radiography for positive purified protein derivative test.
A, Frontal chest radiograph shows slightly prominent right paratracheal stripe and dense pulmonary nodule (arrow) in right upper lobe.
B, Contrast-enhanced sagittal CT image shows lymphadenopathy with coarse calcifications (arrows) in middle mediastinum.

Fig. 32—Neuroblastoma in 1-year-old boy who presented with opsoclonus-myoclonus syndrome. Chest radiographs showed posterior mediastinal mass.
A, Contrast-enhanced axial CT image shows well-defined oval soft-tissue mass (asterisk) with internal calcifications located in posterior mediastinum.
B, Contrast-enhanced coronal CT image shows large soft-tissue mass (asterisk) with calcifications extending from upper thoracic spine to right hilum.
encase blood vessels and to spread through the neural foramina into the spinal canal. MRI is the best imaging modality for showing local spread of tumor and for detecting tumor extension into the spinal canal, which influences surgical planning and management. On MRI, neuroblastomas show high signal on T2-weighted images and low signal on T1-weighted images. They tend to enhance early, reflecting the high vascularity of the tumor. Metaiodobenzylguanidine (MIBG) scintigraphy is a highly sensitive imaging modality for determining the extent of disease, with avid uptake of the radionuclide related to catecholamine production by the tumor.

The current treatment of choice for neuroblastoma in pediatric patients is surgical resection, with adjuvant chemotherapy and radiation therapy for advanced disease. The prognosis varies depending on the location and extent of the tumor. Children with thoracic neuroblastoma have a more favorable prognosis than those with abdominal tumors.

Ganglioneuroblastoma and ganglioneuroma—Ganglioneuroma and ganglioneuroblastoma are less aggressive tumors than neuroblastomas. These tumors of primitive neural crest cells also often arise in the posterior mediastinum. Ganglioneuroma is currently considered to be a benign neoplasm, whereas ganglioneuroblastoma has the potential for distant metastases and thus is considered malignant. Radiologically, they are difficult to differentiate from neuroblastomas because of similar imaging characteristics on all imaging modalities (Figs. 33 and 34). However, ganglioneuroblastoma and ganglioneuroma may have a more fusiform configuration than neuroblastoma. Age is also a diagnostic clue: ganglioneuroblastomas and ganglioneuromas tend to occur in older children, whereas neuroblastomas are seen during the first few years of life. Histopathologic analysis is required for a definitive diagnosis.
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Fig. 34—Ganglioneuroblastoma in 5-year-old boy with chest pain and limping.
A, Coronal T1-weighted image shows large relatively homogeneously enhancing paraspinal soft-tissue mass (asterisk).
B, Contrast-enhanced axial T1-weighted image of large paraspinal mass (asterisk) shows extension (arrow) into spinal canal, causing mass effect on thecal sac (supporting neural origin of mass).

Fig. 35—Neurofibroma in 12-year-old boy with known neurofibromatosis type 1.
A, Frontal chest radiograph shows mild widening of right and left paratracheal stripes (arrows).
B, Contrast-enhanced axial CT image shows multiple lobulated soft-tissue masses in paraspinal regions, following intercostals spaces, and near aortic arch, consistent with neurofibromas (arrows). Note extension along neural foramen on right.

Nerve Sheath Tumors

Although most posterior mediastinal masses in the pediatric population are sympathetic ganglion tumors, some reflect benign (schwannoma, neurofibroma) (Fig. 35) or malignant peripheral nerve sheath tumor. Malignant peripheral nerve sheath tumors are highly cellular pleomorphic spindle cell sarcomas of nerve sheath origin. In pediatric patients, these tumors most commonly arise from a preexisting plexiform neurofibroma, as in patients with neurofibromatosis type 1. Less commonly, a malignant peripheral nerve sheath tumor arises de novo or from a preexisting schwannoma.

A malignant peripheral nerve sheath tumor often appears as a round or oblong posterior mediastinal mass that is associated with a widened neural foramen and follows the course of the involved nerve. On CT, these tumors usually show decreased attenuation because of their lipid contents or cystic degeneration and have variable enhancement (Fig. 36). Dumbbell-shaped extension into the spinal canal can be seen on both CT and MRI. Features suggestive of a malignant peripheral nerve sheath tumor, whether arising de novo or from degeneration of a preexisting...
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mass, include rapid growth, local invasion, osseous destruction, and pleural effusion. Surgical removal for symptomatic or malignant lesions is the current treatment of choice.

Conclusion

Mediastinal masses in children may represent congenital anomalies, infections, and benign and malignant neoplasms. Pseudo-masses can pose diagnostic challenges to the unwary eye. In both asymptomatic and symptomatic children with mediastinal masses, imaging evaluation plays a paramount role by providing precise information regarding location, appearance, size, and relationship to the adjacent mediastinal structures as well as detecting metastases. Knowledge of the practical diagnostic imaging approach based on the three mediastinal compartments and the characteristic imaging appearances can lead to a correct diagnosis and subsequent optimal management.

Selected Reading


Fig. 36—Malignant peripheral nerve sheath tumor in 20-year-old woman with known neurofibromatosis type 1. Contrast-enhanced axial CT image shows multiple round heterogeneously enhancing masses (asterisks) located in posterior mediastinum and on pleural surfaces along course of intercostal nerve.