CHAPTER 54
Mediastinal Masses

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Introduction

Mediastinal masses are a heterogeneous group of lesions that can provide significant diagnostic and management challenges to the paediatric surgeon. The lesions vary from slow-growing congenital cysts to aggressive neoplasms. The symptomatology can be quite varied, and a high index of suspicion needs be maintained to make the diagnosis.

Demographics

Due to the variety of mediastinal masses, it is not possible to determine the true prevalence of these lesions. Each type of lesion, barring mediastinal lymphadenopathy, is quite uncommon. The spectrum of lesions seen on the African continent would include lesions traditionally seen in the developed world, but with infectious causes playing a much larger role in any differential diagnosis. Furthermore, the human immunodeficiency virus (HIV) epidemic has presented myriad presentations not previously seen.

Pathology

Mediastinal masses can be practically classified by the lesion's location within the mediastinum, namely, anterior, middle, and posterior. The location of the mass usually gives a good indication as to the differential diagnosis of the lesion.

The anterior mediastinum extends from the inner aspect of the sternum to the anterior aspect of the trachea, pericardium, and great vessels. Its contents would include the thymus (Figure 54.1), ectopic thyroid or parathyroid, lymph nodes, and connective and adipose tissue. Thymic lesions would include hyperplasia (see Figure 54.1), cysts, thymoma, and thymic carcinoma. Disorders of the lymph nodes would be lymphoma (Figure 54.2), both Hodgkin's and non-Hodgkin's, and more recently an increasing number of patients with tuberculous adenopathy; germ cell tumours, both benign teratomas and malignant seminomas; and lymphatic anomalies, such as lymphatic malformations or lymphangiomas. Uncommon lesions include a retrosternal goitre or ectopic thyroid and malignancies of adipose tissue, a lipoblastoma. A Morgagni diaphragmatic hernia would come to lie in the inferior anterior mediastinum and would thus be considered in the differential of lesions in this location.

The middle mediastinum is situated from the pericardium anteriorly to the prevertebral fascia posteriorly. It contains the major mediastinal viscera, including the oesophagus, trachea, heart, and great vessels. Minor constituents would be the paratracheal spaces and lymphoid tissue. Lesions would thus arise from the aforementioned disorders of the lymphoid tissue (i.e., lymphoma and tuberculosis); congenital anomalies of foregut development (i.e., bronchogenic and enteric, or duplication, cysts); and uncommon lesions related to the heart and pericardium.

The posterior compartment contains the space between the trachea and the spine and the paravetebral sulcus on each side. The contents would include the thoracic spinal ganglia, the sympathetic chain, the proximal part of the intercostal vessels and nerves, lymphatics, and connective tissue. Consequently, lesions in this position are

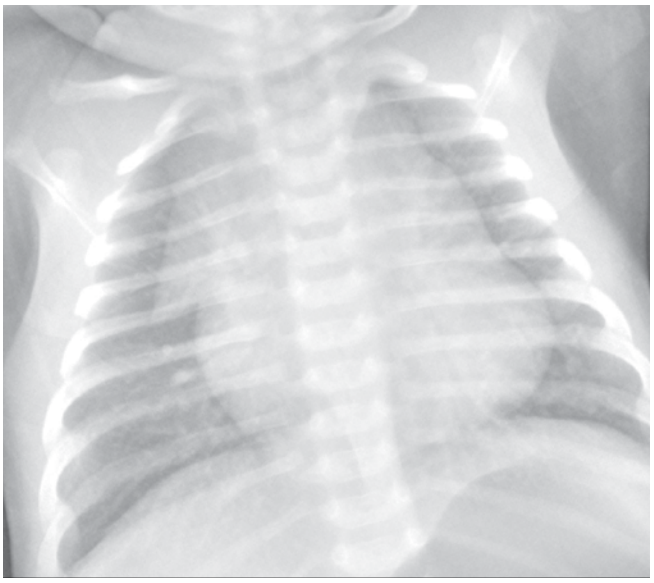

Figure 54.1: Normal thymus in the anterior mediastinum of a neonate.

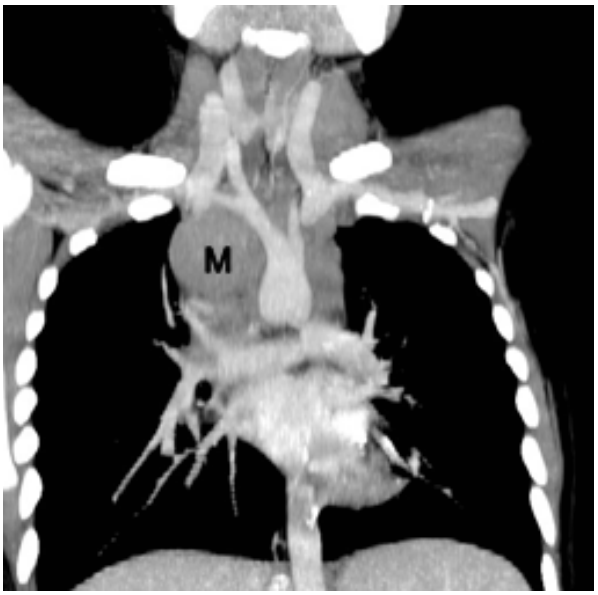

Figure 54.2: Anterior mediastinal mass (M) lymphoma.

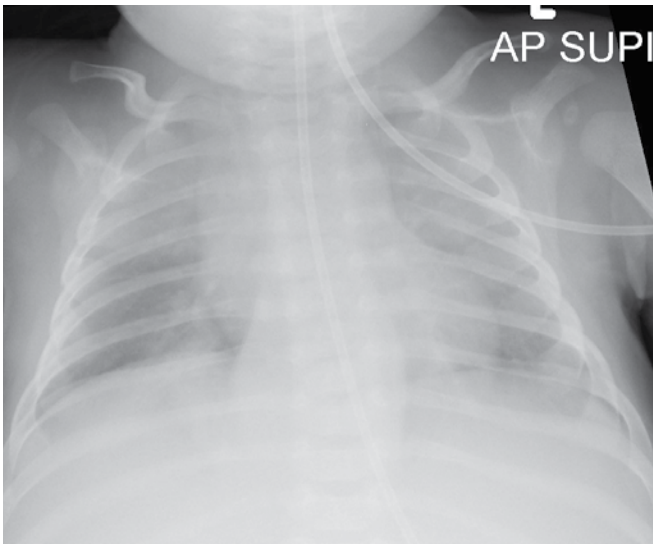

Figure 54.3: Posterior mediastinal mass (neuroblastoma).

typically neurogenic in origin—namely, neuroblastoma (Figure 54.3), ganglioneuroma, neurofibroma, neurilemoma, paeochromocyctoma, and neuroenteric cysts. Other, less common lesions would include a primitive neuroectodermal tumour (PNET) and hamartomas. Lastly, as for the mediastinal compartment, occasionally cystic lesions in the inferior posterior compartment may represent a sliding or paraoesophageal hiatus hernia.

A further mechanism for classifying mediastinal masses are as cystic and solid. Cysts are usually congenital in nature. Cystic lesions would include bronchogenic, duplications, neuro-enteric dermoids, lymphatic, and pericardial cysts. Solid masses are usually neoplasms, which may be benign or malignant and are well detailed in the preceding section.

Clinical Presentation

The location and the age of the patient are often the most useful factors in making a diagnosis. As discussed in the previous section, certain lesions have characteristic sites of occurrence.

Mediastinal lesions are often asymptomatic; many of them are found on routine chest radiology (see Figures 54.1 and 54.3). Symptoms usually occur secondary to a mass effect of the lesion on adjacent structures in the mediastinum. The symptoms, when present, are usually a cough, respiratory distress, wheeze, stridor, or dysphagia. In malignant mediastinal lesions, or occasionally in tuberculous lymphadenopathy, children can present with a superior vena caval syndrome, representing both vascular and airway compression. Depending on the rate of growth of the lesions, the presentation may be insidious; in malignant lesions, however, it may occur over a few weeks, or in lesions complicated by superadded infection or haemorrhage, it may present as a life-threatening emergency over days or even hours.

Posterior mediastinal lesions will usually present with pain secondary to bony erosions or neuralgia. Occasionally, with neurogenic tumours, patients may present with loss of power to the lower limbs and paraplegia secondary to a dumbbell lesion and cord compression. Neurological symptoms can be found with neuro-enteric cysts, but usually take the form of recurrent meningitis and only rarely paraplegia.

Systemic symptoms of loss of weight and night sweats are usually the initial symptoms for both tuberculosis (TB) and lymphoma. Differentiation between these two diseases in the African setting can often be difficult, especially in children coinfected with HIV. Both TB and lymphoma have an atypical and more aggressive course. In younger children, TB is the most common of the mediastinal lesions causing compressive symptoms of the airways. This is secondary to subcarinal and peribronchial nodes.

In the clinical examination, it is important to pay attention to a systemic examination. Features of weight loss, lympahedenopathy, visceromegally, or skin lesions will all contribute to making the clinical diagnosis. A systematic approach to examination is important and can also provide a valuable alternative for a tissue diagnosis.

Investigations

Diagnostic studies will be directed by the type and severity of the symptoms and location of the mass.

Radiology

The initial radiological work-up will be the anteroposterior (AP) and lateral chest x-rays. An enormous amount of information can be gained from this affordable and widely available investigation (see Figures 54.1 and 54.3). Most important, one needs to assess the position of the mass, especially on the lateral film, which would place it in either the anterior, middle, or posterior compartments. The airways can be well assessed by looking for compression and deviation of the trachea and major bronchi.

In instances where dysphagia is a predominant symptom, a contrast swallow would be helpful to identify the location and extent of the oesophageal compression. In instances of foregut duplication, this will communicate with the normal oesophagus in 20% of the cases.

The mainstay of investigation will be the computed tomography (CT) scan, which provides an excellent outline of the mediastinum and major airways (see Figure 54.2). It gives a precise relationship of the mass to the airway, oesophagus, and major vascular structures. In instances where one suspects a foregut duplication, imaging should continue into the upper abdomen, as these lesions may extend below the diaphragm. CT scans should be kept to an absolute minimum amidst concerns of radiation-induced neoplasia.

In children who present with neurological symptoms, magnetic resonance imaging (MRI), where available, should strongly be considered, as—especially with dumbbell lesions—cord compression may be present.

Occasionally, radioisotope studies may be required, notably a metiodobenzylgaunidine (MIBG) scan, which is both specific and sensitive for neuroblastoma or phaechromocytoma.

The goal of the radiological work-up is to aid diagnosis and to help define the optimal surgical approach.

Laboratory Testing

Specific laboratory tests can aid in the diagnosis of mediastinal masses. Serum lactate dehdrogenase (LDH) is a sensitive but nonspecific marker of lymphoma and neuroblastoma. Homovanillic acid (HVA) or vanillulmandelic acid (VMA) are both markers of neurogenic tumours, and these tests should be done in posterior mediastinal lesions.

Finally, TB testing, either in the form of skin antigenicity (Mantoux or PPD—purified protein derivative) or white cell interferon-gamma testing (ELISpot or QuantiFERON®-TB Gold) should be performed. This testing must be correlated with the clinical picture—either induced sputum or gastric washings, depending on the age of the child. One note of caution: if TB is diagnosed, it can coexist with lymphoma, and hence failure to respond or progression on TB treatment should alert one to an alternative diagnosis.

Histology

Ultimately, treatment of solid mediastinal masses rests on the histological diagnosis. This is usually most pertinent to masses in the anterior mediastinum, where lymphoma is suspected. In these cases, peripheral nodes may provide the answer and avoid entrance of the thoracic cavity. In cases of smaller masses or cysts, excision biopsy can be done.

Treatment

The treatment for mediastinal masses ranges from curative excision to medical management, depending on the cause. In general, however, apart from lymphoma and TB, most lesions will require excision.

Airway Management

A compromised airway is often the reason for emergent presentation of these lesions. Airway compromise secondary to mediastinal lesions can be particularly difficult to manage, as the area of compression may often be at a carinal level and hence not alleviated by intubation or a tracheostomy. At presentation, patients may be unable to lie flat, and if given any sedation or anaesthetic will completely lose their airway.

The best form of management would be avoidance of any sedation. Obtaining tissue under local anaesthetic from other sites is preferable. If this is not possible, then careful liaison with anaesthetic services and use of a CT scan best identify the degree and location of the compression. A rigid bronchoscope is a necessity, as it may be the only option to re-establish an airway distal to the carina. Full intensive-care facilities must be available. Discussion with oncology services should be undertaken to balance the risk of biopsy, or empirical initial therapy to alleviate some of the airway compromise, with the disadvantage of losing valuable histological information.

Definitive Management

Surgical involvement is twofold: first, obtaining tissue for histology in unresectable lesions; and second, for excision of cysts and masses.

Careful attention needs to be paid to the access incision that is to be made. Anterior mediastinal lesions are best performed through a sternotomy, and middle and posterior lesions via a posterior lateral thoracotomy. Lesions that have a spinal component should either have a combined procedure, or alternatively have the spinal component done first, as failure to do so could result in paraplegia. In rare cases, thoracic foregut duplication cysts may transverse the diaphragm and end in the abdomen, requiring a combined abdominal and thoracic approach.

Occasionally, surgical intervention will be required for node decompression in cases of TB.

Thorascopic surgery offers an excellent diagnostic and therapeutic tool when available, but expertise and equipment may make this a limited option in most African settings.

Postoperative Complications

Postoperative complications are usually secondary to inadequate analgesia, resulting in pulmonary atelectasis. Incompletely excised lesions may recur, especially lymphatic malformations and malignant lesions.

Prognosis and Outcome

Generally, the prognosis for mediastinal masses is excellent. Mediastinal cysts that are excised offer complete cysts. Effective medical therapy for tuberculosis can prevent damage to the bronchi and lungs. Malignant lesions would depend on the histology, but most of the lesions found in the mediastinum are responsive to chemotherapy.

Evidence-Based Research

The wide spectrum of disorders that make up mediastinal masses do not lend themselves to comparative trials. Each of the neoplastic lesions (i.e., lymphoma, germ cell tumours, and neuroblastoma) has been extensively studied with respect to multimodal therapy, but these are not specific for mediastinal masses.

The largest case series is that of Grosfeld et al. (see Suggested Reading). Most subsequent series focus on thorascopic approaches to these lesions. Table 54.1 presents an analysis of mediastinal masses in 29 children. Table 54.2 discusses a study of airway obstruction and management in mediastinal tumours.

Table 54.1 Evidence-based research.

Title	When is a mediastinal mass critical in a child? An analysis of 29 patients
Authors	Lam JC, Chui CH, Jacobsen AS, Tan AM, Joseph VT
Institution	Department of Paediatric Surgery, KK Women's and Children's Hospital, Singapore
Reference	Pediatr Surg Int 2004; 20(3):180–184
Problem	The aims of this study were to determine the pattern of presentation of childhood mediastinal masses in our community and to identify factors associated with the development of acute airway compromise.
Intervention	The authors retrospectively reviewed the records of 29 consecutive patients with mediastinal masses managed at their institution between January 1995 and December 2001. Demographic data, mass characteristics, clinical presentation, and surgical procedures were recorded.
Comparison/ control (quality of evidence)	Seven patients (24.1%) were asymptomatic at presentation. Eight (27.6%) were classified as having acute airway compromise at presentation. Respiratory symptoms and signs were the most common mode of presentation (58.6% and 55.2%, respectively). The most common histological diagnosis was neurogenic mass (37.9%), followed by lymphoma (24.1%). Most masses were located in the superior mediastinum (41.1%). Factors associated with the development of acute airway compromise were (1) anterior location of the mediastinal mass (P=0.019); (2) histological diagnosis of lymphoma (P = 0.008); (3) symptoms and signs of superior vena cava syndrome (P = 0.015 and 0.003, respectively); (4) radiological evidence of vessel compression or displacement (P = 0.015); (5) pericardial effusion (P=0.015); and (6) pleural effusion (P = 0.033).
Outcome/ effect	Clinical presentation of childhood mediastinal masses is often nonspecific or incidental. Yet they have the propensity of developing acute airway compromise, which is closely associated with superior vena cava obstruction. Such patients should be managed as a complex cardiorespiratory syndrome, termed "critical mediastinal mass syndrome", by an experienced multidisciplinary team.

Table 54.2 Evidence-based research.

Title	Mediastinal tumors-airway obstruction and management
Authors	Robie DK, Gursov MH, Pokorny WJ
Institution	Cora and Webb Manning Department of Surgery, Baylor College of Medicine, Houston, Texas, USA
Reference	Semin Pediatr Surg 1994; 3(4):259–266
Historical significance/ comments	Large mediastinal masses can cause compression of surrounding mediastinal structures. Patients may have symptoms of airway obstruction or cardiovascular compromise. The additive effects of anaesthetics, paralysis, and positioning during biopsy can lead to acute airway obstruction and death. In some cases, tissue diagnosis can be achieved and treatment initiated without general anaesthesia. When general anaesthesia is necessary, specific measures should be taken to avoid disaster or immediately alleviate obstruction should it occur. Some patients at greatest risk will require pretreatment of the mass before tissue diagnosis. This article reviews these issues and provides a useful algorithm for managing patients with mediastinal masses.

Key Summary Points

1. Mediastinal masses are presented by their anatomical location into anterior, middle, and posterior mediastina.
2. The location usually indicates the differential diagnosis.
3. Chest x-ray and CT scan are the most useful imaging modalities for mediastinal masses.
4. Most lesions require excision (except infective causes and lymphoma).
5. Airway management is paramount.

Suggested Reading

Engum SA. Minimal access thoracic surgery in the pediatric population. Semin Pediatr Surg 2007; 16:14–26.

Grosfeld JL, Skinner MA, et al. Mediastinal tumors in children: experience with 196 cases. Ann Surg Oncol 1994; 1:121–127.

Hammer GB. Anaesthetic management for the child with a mediastinal mass. Paediatr Anaesth 2004; 14:95–97.

Jaggers J, Balsara K. Mediastinal masses in children. Semin Thorac Cardiovasc Surg 2004; 16:201–208.

Williams HJ, Alton HM. Imaging of paediatric mediastinal abnormalities. Paediatr Respir Rev 2003; 4: 55–66.