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ORIGINAL ARTICLE

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When is a mediastinal mass critical in a child? An analysis of 29 patients

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Abstract 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. 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. 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 ($P = 0.033$). 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.

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Introduction

Up to one-third of childhood mediastinal masses have been reported to be discovered incidentally [13]. Clinical presentation is often nonspecific, with a predominance of respiratory symptoms (which mimic common upper respiratory ailments) in 60% of all cases [3]. Death related to the inability to secure an airway in patients with large mediastinal masses is a well-recognised complication that particularly occurs at induction of general anaesthesia [4, 16]. The diagnoses of mediastinal masses are often helped by diagnostic imaging techniques such as plain radiographs, computer tomography (CT), and, occasionally, magnetic resonance imaging (MRI). Despite these recent advances in technology, full histological examination is required to exclude a malignancy, which has been reported in 27–82% of cases in other series [5, 8, 9]. It is therefore crucial to identify the child with a mediastinal mass that could potentially lead to an acute life-threatening catastrophe and to take the necessary precautions in the subsequent diagnosis and management.

Materials and methods

We reviewed the records of 29 consecutive patients who were seen at KK Women's and Children's Hospital between January 1995 and August 2001 with the diagnosis of mediastinal mass. We looked specifically at the characteristics of patients who presented acutely with airway compromise, and we analysed our series for factors associated with acute airway compromise. Demographic data collected for these 29 patients included gender, race, and age at the time of diagnosis.

Clinical presentation was further subclassified into symptoms and signs. In particular, superior vena cava syndrome (SVCS) is defined by symptoms and signs of venous congestion, including plethora, orbital suffusion, upper extremity and facial oedema, prominent venous patterning over the neck and chest, and, rarely, symptoms of cerebral oedema, including dizziness, lethargy, stupor, impaired consciousness, and seizures. Acute airway compromise is defined as the development of respiratory distress with stridor or desaturation or the presence of significant airway narrowing of more than 50% on radiological imaging [16]. These patients were admitted into the intensive care unit for further monitoring before beginning treatment. They were further evaluated for associated factors that may have suggested the development of airway compromise.

Radiological characteristics of the mediastinal mass and other associated thoracic features were documented. The anatomical subdivision of the mediastinum into superior, anterior, middle, and posterior compartments was used to define the location of the mediastinal mass.

The final diagnoses of the masses were based on histological analyses, which were subdivided into neurogenic masses, lymphomas, lymphangiomas, germ cell tumours, enteric cysts, and other histological types.

Statistical analysis was performed using SPSS for Windows version 9.0.0. Associations among factors were examined using Fisher's exact test. Differences were considered significant if the P value was < 0.05.

Results

Demographic data

The median age of the 29 patients at the time of diagnosis was 47 months (range 1–173 months). Seventeen patients were males and 12 were females. Twenty-two (75.9%) were Chinese, four (13.8%) Malay, one (3.4%) Indian, and two (6.9%) Caucasian, which is consistent with the racial distribution of our local population.

Anatomical and histological characteristics

Twelve (41.4%) of the 29 patients studied had masses located in the superior mediastinum. Eight masses each were located in the anterior and posterior mediastinum. Only one of the masses was located in the middle mediastinum. The most common histological group of diagnoses consisted of neurogenic masses (11; 37.9%), which included five neuroblastomas, four ganglioneuromas, one ganglioneuroblastoma, and one peripheral neuroectodermal tumour (PNET). Of the seven (24.1%) patients who were diagnosed with lymphomas, five had non-Hodgkin's lymphoma and two had Hodgkin's lymphoma. Four (13.8%) patients had lymphangiomas, two (6.9%) had germ cell tumours, and one each had oesophageal duplication cyst, acute lymphoblastic leukaemia, Langerhans' cell histiocytosis, extralobar pulmonary sequestration, and matted tuberculous lymphadenopathy. Sixteen (55.2%) of the 29 mediastinal masses were malignant.

Clinical presentation

Seven (24.1%) of the 29 patients were asymptomatic at the time of diagnosis. Respiratory symptoms were the most common ones at presentation (17; 58.6%) (Table 1). These included cough ($n = 13$), dyspnoea ($n = 9$) and wheeze ($n = 2$). The most common physical signs also involved the respiratory system (16; 55.2%); eight patients had decreased air entry, five had stridor, two had tracheal deviation, and one had rhonchi. Palpable lymphadenopathy was present in 10 patients, and four presented with signs of SVCS.

Radiological characteristics

All 29 patients were investigated with chest x-rays and CT scans of the chest. In addition, four patients had MRI to better characterise the nature of their masses. Eleven of the 29 patients had airway compression or displacement by the mediastinal mass. Six had pleural effusion, three had vessel compression or displacement, three had pericardial effusion, three had mediastinal deviation, three had spinal canal invasion, and two had bony erosion.

Surgical procedures in providing diagnosis

Histological diagnoses were made after surgical biopsies in 26 of the 29 patients, of which 17 (65.4%) were biopsies of the mediastinal mass and nine (34.6%) were specimens obtained from extrathoracic sites. In the remaining three patients, diagnoses were made from thoracocentesis in two patients and bone marrow biopsy in one. Twelve (46.2%) of the 26 patients who underwent mediastinal surgery had complete excision of the mediastinal masses, whereas biopsies were obtained from the remaining 14. There were no surgical mortalities.

Acute airway compromise

Eight (27.6%) of the 29 patients presented with acute airway compromise. The median age at presentation was 110 months (range 7–173 months). Five (62.5%) were males, and all were Chinese. We evaluated the relationship of age, gender, and race with the tendency for airway compromise, and none of the associations tested reached statistical significance ($P = 0.092$, $P = 1.000$, $P = 3.515$ respectively).

Table 1 Clinical presentations of children with mediastinal masses

	Absence of airway compromise ($n=21$) *N* (%)	Presence of airway compromise ($n=8$) *N* (%)	*P* value
Asymptomatic	7 (33.3)	0	0.142
Systemic[a]	7 (33.3)	6 (75.0)	0.092
Respiratory	9 (42.9)	8 (100)	0.012
Wheeze	1 (4.8)	1 (12.5)	0.483
Cough	7 (33.3)	6 (75.0)	0.092
Dyspnoea	5 (23.8)	4 (50.0)	0.209
Chest pain	4 (19.0)	0	0.552
Dysphagia	0	1 (12.5)	0.276
Superior vena cava syndrome	0	3 (37.5)	0.015
Other symptoms[b]	1 (4.8)	0	1.000
Normal	4 (19.0)	0	0.552
Respiratory	8 (38.0)	8 (100)	0.003
Rhonchi	0	1 (12.5)	0.276
Tracheal deviation	1 (4.8)	1 (12.5)	0.483
Decreased air entry	5 (23.8)	3 (37.5)	0.646
Stridor	0	5 (62.5)	<0.001
Superior vena cava syndrome	0	4 (50.0)	0.003
Lymphadenopathy	7 (33.3)	3 (37.5)	1.000
Other signs[c]	7 (33.3)	2 (25.0)	1.000

[a]Systemic symptoms included fever, loss of weight, and loss of appetite
[b]Bilateral lower limb weakness secondary to cord compression
[c]Other signs included neck masses, swelling over chest wall, hepatosplenomegaly, generalised maculopapular rash, and neurological signs

Five (62.5%) of the masses in these eight patients were located in the anterior mediastinum, and the remaining three were in the superior mediastinum. The location of the mass in the anterior mediastinum was associated with a significant risk of acute airway compromise ($P=0.019$). Four patients with non-Hodgkin's lymphomas and one with acute lymphoblastic leukemia had anterior mediastinal masses. The superior mediastinal masses included a Hodgkin's lymphoma, an oesophageal duplication cyst, and a cervicothoracic lymphangioma. Even though a malignant histology was not significantly associated with acute airway compromise ($P=0.238$), the diagnosis of lymphoma was significantly associated with the presence of acute airway compromise ($P=0.008$), whilst neurogenic tumours were associated with the absence of airway compromise ($P=0.012$).

As expected, all eight patients who developed acute airway compromise presented with respiratory symptoms and signs. We evaluated the relationship of symptoms of wheeze, cough, and dyspnoea with acute airway compromise and found no statistical significance. Not surprisingly, as reflected in the definition of acute airway compromise, all five patients with stridor also had acute airway compromise ($P<0.001$). However, rhonchi, tracheal deviation, and decreased air entry were not significantly associated with acute airway compromise. Symptoms and signs suggestive of SVCS were found to be significantly associated with the development of acute airway compromise ($P=0.015$ and 0.003, respectively).

Seven (87.5%) patients with acute airway compromise presented with airway compression or displacement on radiological imaging ($P<0.001$). Other significant radiological findings that were associated with acute airway compromise included pleural effusion, vessel compression or displacement, and pericardial effusion ($P=0.033$, $P=0.015$, and $P=0.015$, respectively) (Fig. 1).

Discussion

Childhood mediastinal masses pose difficult diagnostic and therapeutic challenges. The development of masses in a uniquely limited space within the small thoracic cavity in children renders the masses more likely to cause acute symptoms by compression as compared with masses in adults [6, 14]. Most of these masses occur near the tracheobronchial tree and major blood vessels, a position that may potentiate life-threatening airway

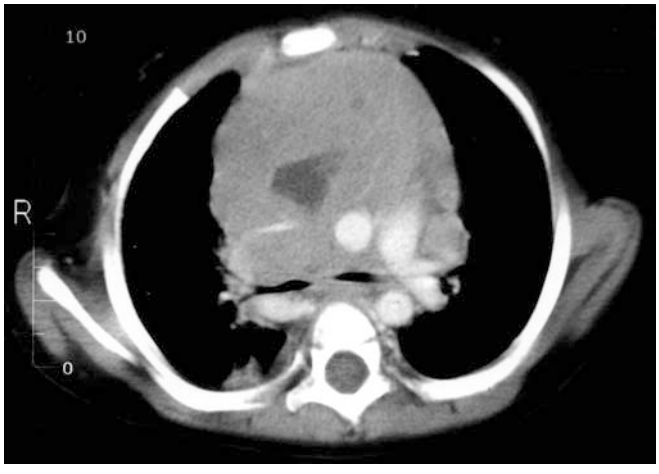

Fig. 1 CT scan showing anterior mediastinal mass (non-Hodgkin's lymphoma) causing superior vena caval and bilateral bronchial compressions

obstruction and/or cardiovascular compromise either by growth of the mass or during the process of general anaesthesia (GA). Our review of 29 children who presented with mediastinal masses determined the pattern of presentation in our community and added further predictive factors associated with the development of cardiorespiratory compromise.

Even though our centre manages patients from birth to 18 years of age, we realised that a significant number of adolescents are managed by the adult oncologists. Our series captured the younger patients of the cohort with a median age of 47 months at diagnosis. This translated into a peculiar histological distribution, with more neurogenic masses (37.9%), which are more common in younger children [2, 10, 15, 17], than lymphomas (24.1%), which are known to occur more frequently in adolescents [13].

Clinical presentation was often nonspecific or incidental; 24.1% of patients were asymptomatic at the time of diagnosis, which is consistent with previous published reports [13, 15]. Half of the patients had nonspecific presentations that often mimicked common upper respiratory ailments or viral illnesses, none of which would have signaled a potential life-threatening catastrophe. Signs of acute airway compromise were present in 27.6% and were managed in the intensive care unit. All of the mediastinal masses were detected by plain chest radiographs.

Identification of risk factors for cardiorespiratory collapse at pre-anaesthesia or at induction of GA had been a major challenge. In other series [3, 4, 11, 13], deaths had occurred in patients who could not be intubated due to extreme airway compression by the tumour's direct mass effect. Previous authors have identified several factors as unsafe for GA: stridor, orthopnea [3], CT findings of more than a 50% decrease in the cross-sectional area of the trachea, and a peak expiratory flow rate $< 50\%$ of predicted value [16]. Of our patients, 27.6% were identified as having the above. In addition, other symptoms previously reported include progressive dyspnoea and a history of recurrent chest infections [1], which may be associated with complete airway obstruction at induction of anaesthesia. We evaluated our patients for any additional factors or clues that may have alerted us to the potential risk.

The risks of anaesthesia in patients with anteriosuperior mediastinal lesions are well recognised [3, 7, 12]. In our review, we identified anterior mediastinal masses to be significantly associated with the development of a critical life-threatening situation. Besides their proximity, which would allow mass effect on both the airways and major blood vessels, their anterior location would also allow for direct gravitational compression of the airways at paralysis during induction of GA in a supine position.

Symptoms and signs of SVCS and radiological evidence of vessel compression—in particular, the superior vena cava (SVC)—were other significant factors associated with the development of a life-threatening situation. SVCS was the only nonrespiratory presentation that was significant in our analysis. All four children with SVCS developed acute airway compromise. This was not surprising when we consider the proximity of the SVC and the tracheobronchial tree in the superoanterior mediastinum. A notion is that if a child can develop SVCS from a mediastinal mass, his airway would also be at risk.

Radiological evidence of pleural and pericardial effusions was significantly associated with acute airway compromise. Besides airway obstruction, mediastinal masses caused SVC compression, pulmonary artery compression, and/or myocardium compression, which led to decreased right ventricular output and pericardial and pleural effusions. Though these effusions worsen the severity of cardiorespiratory distress, they may be useful sources for fluid cytological examination.

The close association of SVCS and acute airway compromise in childhood mediastinal masses suggests that they should be dealt with as a single complex syndrome, which we would like to term "critical mediastinal mass syndrome". In this syndrome, the mediastinal mass not only threatens the airways, but it also causes SVCS, leading to right ventricular output reduction and right heart failure and, sometimes, pulmonary artery and myocardium compression. Under GA, a fixed low cardiac output associated with SVCS is further diminished by GA-associated myocardial depression and the supine position of the patient, which may lead to a catastrophe.

With improved understanding, alternative strategies of extrathoracic biopsies under local anaesthesia (e.g. bone marrow and extrathoracic lymph node biopsies, pleurocentesis, and pericardiocentesis) should be considered whenever feasible. When faced with a probable malignancy, and if alternative strategies are not feasible, physicians should give empirical treatment with steroids, chemotherapy, and radiotherapy to restore the airway. GA in a compromised patient is reserved for carefully selected patients, such as those with benign lesions, and should be provided by an experienced anaesthetic and surgical team familiar with managing such patients. In such situations, anaesthetic precautions, including a sitting position induction, avoidance of neuromuscular blocking agents, turning of the patient to a semiprone position, direct laryngoscopy, rigid bronchoscopy, and cardiopulmonary bypass, should be available.

Conclusion

Mediastinal masses are often difficult lesions to diagnose and manage in children and have a high incidence of asymptomatic presentation. They often result in a critical mediastinal mass syndrome, which may lead to cardiorespiratory compromise. A high index of suspicion based on likely signs and symptoms aided by radiological imaging may help identify patients at risk and prompt the involvement of an experienced multidisciplinary team.

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