

A Case of Cervico-Mediastinal Thymic Cyst Causing Tracheal Compression in a Child

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INTRODUCTION

Stridor, a high-pitched monophasic sound indicative of airway obstruction, is a common presenting symptom in pediatric emergency department (ED).^{1,2} Although underlying cause of stridor is infectious in most cases, such as croup or bronchiolitis, clinicians should maintain a broad differential diagnoses when dealing with a child with stridor. We are presenting a unique case of cervico-mediastinal thymic cyst presenting as stridor and ultimately leading to acute hypoxic respiratory failure.

CASE REPORT

A previously healthy 18-month-old boy presented to an emergency department (ED) with fever, tachypnea, inspiratory stridor, and brief period of apnea with cyanosis. Oxygen saturation improved with adequate airway positioning and administration of oxygen via nasal cannula. He was given racemic epinephrine for continued stridor in addition to combination therapy with ipratropium bromide and albuterol. He then was transferred to a pediatric ED for a higher level of care.

In the pediatric ED, he was noted to have dyspnea and stridor with sudden desaturation, along with seizure-like activity, including clenching of his jaws. He was given lorazepam for seizure-like activity. He continued to receive supplemental oxygen via placement of oral airway for airway protection. He required intubation by direct laryngoscopy following an episode of emesis associated with desaturation. He was admitted to the pediatric intensive care unit (PICU), where septic workup was completed with chest x-ray (CXR) showing a right patchy opacity, negative blood cultures, and negative respiratory viral panel. He was started on intravenous (IV) dexamethasone and IV ceftriaxone. A diagnosis of febrile seizures was made after an electroencephalogram showed no evidence of focal or epileptiform abnormalities. A repeat CXR showed diminished lung volumes with a consolidative medial right upper lobe opacity. Heart size and central vascularity were within normal limits but a prominent thymic shadow was seen (Figure 1).



Figure 1. CXR with diminished lung volumes and a consolidative medial right upper lobe opacity. Heart size and central vascularity within normal limits. A prominent thymic shadow is evident.

He was extubated after three days on mechanical ventilation. Immediately after extubation, he had severe desaturations which improved with jaw thrust and bag and mask ventilation. He was supported on continuous positive airway pressure (CPAP), and an otolaryngologist was consulted. A bedside flexible laryngoscopy showed no evidence of upper airway obstruction. Immediately following the procedure, he had another decompensation requiring re-application of CPAP. He received a high-dose methylprednisolone burst over seven days and was weaned off of CPAP. He eventually was discharged home with a one-week prescription of dexamethasone and antibiotics along with a referral for a primary care physician (PCP) follow-up.

Due to persistent dyspnea and stridor, the patient was continued on oral steroids at home by his PCP and again was seen by an otolaryngologist who recommended further evaluation. He underwent microlaryngoscopy and rigid bronchoscopy six weeks after admission to the PICU, where he was found to have severe distal tracheal obstruction. A computed tomography (CT) scan of chest with angiogram revealed a large thymic cyst compressing his innominate artery and distal airway (Figure 2). He underwent an excision of cervicome-diastinal cyst and was extubated immediately after surgery without having desaturation episodes. He was weaned to room air and was discharged home on post-operative day two.

DISCUSSION

By the sixth week of gestation, the thymus can be recognized as a separate paired organ.³ Embryologically, it is derived from the third and fourth pharyngeal pouch. On the onset of the eighth week of gestation, thymic anlage develops into the thymopharyngeal duct. This duct runs from the mandibular angle to the anterior superior mediastinum, the ultimate position of the thymus. The upper end of the thymus regresses and gradually disappears. The remnant of endodermal epithelium undergoes regression and forms the Hassall's corpuscles.

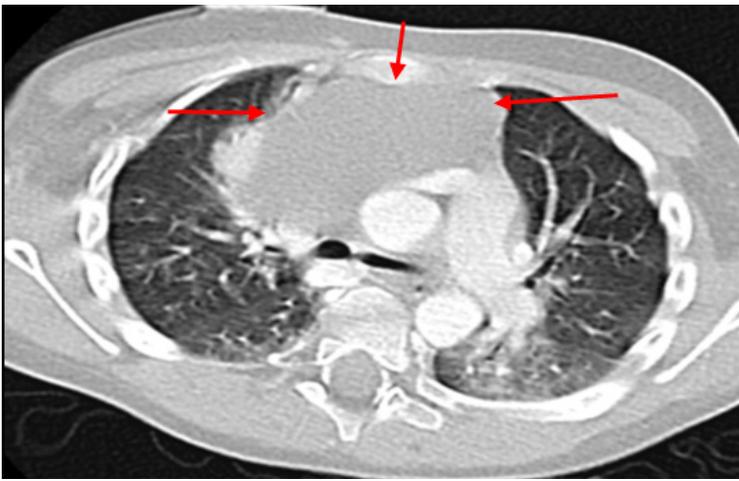


Figure 2. CT scan of chest with angiogram with a large thymic cyst compressing innominate artery and distal airway.

Two main theories for the origin of cervical thymic cyst have been suggested.⁴ First, a unilocular cyst is more common which is thought to originate from the persistence of thymopharyngeal duct. Second, a multilocular cyst is attributed to the degeneration of thymic Hassal's corpuscle.

Cervico-mediastinal thymic cysts (CTC) are increasingly rare with only 100 cases reported in the literature.⁵ Most of the time, it is asymptomatic, making diagnosis difficult; thus a large number of cases are discovered incidentally. CTCs have strong male preponderance, more predilection toward the left side and present themselves during the first decade of life as a painless slow growing mass in the areas located between the angles of the mandible to sternum. In nearly half of the cases, the CTC may reach up to and make contact with the mediastinum.⁶ If symptomatic, patients usually present to the outpatient department with complaints of stridor, dysphonia, or dysphagia, as was in our case.⁵

The differential diagnosis of CTCs in the pediatric population is extensive, ranging from common congenital causes, such as thyroglossal duct cyst, branchial cyst to benign tumors (dermoid cysts, epidermoid cysts), malignant tumors (lymphoproliferative, soft tissue sarcoma and other metastatic lesions), and tumors arising from thyroid and parathyroid.⁷ Imaging studies, mainly CT scan and magnetic resonance imaging (MRI), along with surgical findings and histopathological correlation play an important role in diagnosing a thymic cyst.⁶

The treatment of choice is surgical excision and no recurrences have been reported after complete resection.⁸ However, it is imperative that the existence of a mediastinal thymus be confirmed with MRI or fine needle aspiration cytology prior to surgery, because thymectomy during childhood can cause severe impairment of immune status later in life.⁹ Thymic carcinoma and myasthenia gravis are some of the rare complications of surgery which should be taken into account.

CTC, albeit a rare cause of pediatric neck mass, should not be overlooked as a diagnosis when a child presents with asymptomatic cervical neck mass or persistent stridor with hypoxic respiratory failure despite adequate ventilatory support. Ultrasound, MRI, and CT scan along with histopathological examination are the best

modalities to create a definitive diagnosis. CTCs have excellent prognosis and can be treated with surgical excision with minimal chance of recurrence.

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