# Large Cervicothoracic Thymic Cyst Causing Prominent Airway Deviation in a 3-Day-Old Neonate

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#### **ABSTRACT**

Cervicothoracic mass in the pediatric population is uncommon and has a broad differential diagnosis. Frequently, masses in the cervical region present with airway compromise, particularly in younger patients. We present a case of an extremely large cervicothoracic mass causing airway obstruction in a 3-day-old, otherwise healthy male infant. Following awake intubation for airway protection, a  $4.5~\rm cm \times 2.5~\rm cm \times 1.5~\rm cm$  thymic cyst was removed. This case illustrates the wide differential diagnosis of cervicothoracic masses and shows the difficulty of preoperative diagnosis, especially in the case of thymic cysts with extension into the cervical space.

### **CASE**

Our patient was a 3-day-old male infant born to a 17-year-old G1P1 (Gravida 1, Para 1) female. Pregnancy was uncomplicated with routine prenatal care. Prenatal labs were notable for negative Group B streptococcus, hepatitis B, rubella, and a nonreactive VDRL (Venereal Disease Research Laboratory). The patient was born by vaginal delivery at 39 and 5/7 weeks gestation and had APGARS of 9 and 9 at 1 and 5 minutes, respectively. Initial course in the newborn nursery was unremarkable, with no signs of respiratory distress or feeding difficulty. Patient appeared well, without any outward signs of neck mass. He was discharged to home after 2 days.

Over the following 24 hours, the patient developed difficulty feeding with increasing lethargy and intermittent apnea at home and was brought to the local emergency department for evaluation. On exam, he was found to have a large, soft neck mass with biphasic stridor. A computed tomography (CT)

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scan, performed without sedation, noted a cystic lesion extending posteriorly from the anterior mediastinum to the prevertebral space and superiorly into the hypopharyngeal region, causing prominent rightward and posterior laryngeal and tracheal deviation. The mass was noted to contain an air-fluid level with homogenous fluid on CT imaging (Figure 1). Given this finding, he was transported to the University of Wisconsin Hospital &

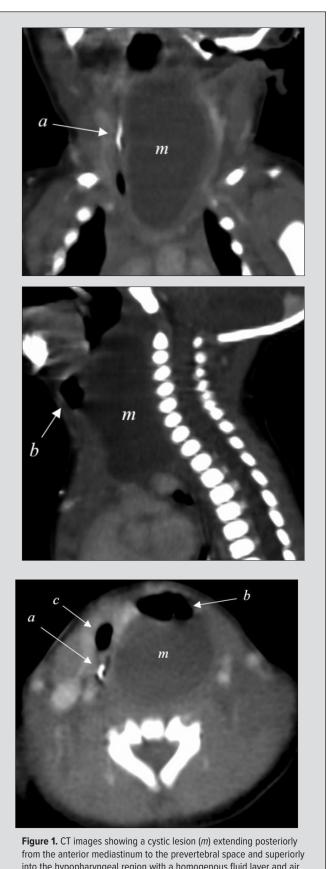
Clinics for evaluation by pediatric otolaryngology.

On arrival, he was on nasal cannula oxygen but continued to have significant respiratory distress and intermittent desaturations, which were more prominent with agitation. Following evaluation by pediatric otolaryngology, he was taken to the operating room for mass resection. Airway control was obtained through awake intubation using rigid bronchoscopy after failed direct laryngoscopy.

During resection, the lesion was noted to arise from the hypopharynx in the area of the left pyriform sinus and contained foul smelling, purulent fluid (Figure 2). Tissue pathology revealed a thymic cyst with components of thyroid and parathyroid tissue. The fluid grew multiple organisms, including S viridans, H parainfluenzae, and S anginosus, consistent with oral flora. Given this finding, we hypothesized that the mass, which may have had a connection to the oral cavity, enlarged secondary to the introduction of food. Post-operatively, the patient developed hypocalcaemia, but was responsive to vitamin D and calcium supplementation. The hypocalcaemia was determined to be secondary to parathyroid tissue removal and the stress of surgery in a newborn. Specifically, evaluation for 22q.11 deletion by fluorescent in situ hybridization (FISH) was negative. The patient recovered from his surgery without complications and was discharged from the hospital at 13 days old.

#### **DISCUSSION**

Differential diagnosis of cervicothoracic masses can be classi-



into the hypopharyngeal region with a homogenous fluid layer and air pocket (b) causing prominent esophageal (a) and tracheal (c) deviation.

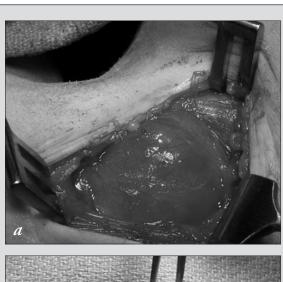






Figure 2. Imaging demonstrating the position of the lesion at dissection (a), decompression of cyst with yellow fluid noted (b, arrow), and size of mass after resection (c).

fied into 4 broad categories: congenital lesions, inflammatory lesions, tumors - benign or malignant, and traumatic lesions. Congenital lesions include lymphatic malformations, hemangioma, thymic cysts, and vascular abnormalities. Inflammatory lesions include inflammatory adenopathy reactive to tuberculosis, mononucleosis, tularemia, cat-scratch fever, or other upper respiratory illness. In addition, inflammatory lesions may include abscess formations from tuberculosis or other causes. Benign tumors include lipoma, lipoblastoma, aggressive fibromatosis, and nerve sheath lesions. Malignant tumors may include lymphoma, thyroid carcinoma, neuroblastoma, and chest wall tumors such as rhabdomyosarcoma and Ewing sarcoma. Traumatic lesions may include esophageal foreign body, granuloma, and cervicothoracic hematoma. Within the pediatric population, congenital lesions are most frequent, with lymphatic malformations being most common.1

The differential for thymic enlargement, in contrast, is associated with 4 causes: cysts, hyperplasia, hemorrhage, or neoplasm.<sup>2</sup> The differential diagnosis when presented with a fluid-filled cyst in the cervical region includes thymic cysts, brachial cleft cysts, thyroglossal duct cysts, dermoid cysts, thymic tumors, cystic hygromas, teratomas, and abscesses.<sup>1,3</sup>

Congenital thymic cysts compose <1% of pediatric cervicothoracic masses and are essentially benign, with 80%-90% presenting as asymptomatic, slow growing masses.<sup>2,4,5</sup> Presentation with symptoms often occurs between 5 and 7 years of age,<sup>5</sup> with 6% to 10% having airway complaints, vocal cord paralysis, or pain. Thymic cysts develop from the persistence of thymic tissue during the degeneration of the thymopharyngeal duct in the third and fourth pharyngeal pouch.<sup>3,5</sup> Consequently, the cyst will occasionally connect to the pyriform sinus by an internal sinus, which provides access for bacterial organisms, as happened with our patient. Most lesions are soft, nontender, and lie on the anterior border of the sternomastoid muscle opposite the thyroid and found on the left side of the neck. Approximately 39% to 50% of cervical thymic cysts will extend into the thoracic space.<sup>3,5</sup>

On CT imaging, these cysts are well marginated, with attenuation close to that of water. In comparison, on MRI, the T1-weighted lesions are of low signal intensity, and T2-weighted lesions are of intermediate or high signal intensity. Histologically, the thymic cysts are commonly lined by a flattened cuboidal epithelium and Hassall's corpuscles. Management of thymic cysts is surgical, with excision of the lesion.

There have been 4 reported cases of cervicothoracic thymic cysts within the pediatric literature.<sup>2-4,6</sup> Two of the 4 cases presented with airway complications (1 patient was 6 years of age and the other 20 months). The 6-year-old patient presented

with difficulty breathing but not frank respiratory failure, most likely due to a larger cervical space. In comparison, the 20-month-old with a smaller airway presented with both apnea and respiratory failure. Both patients were intubated prior to resection of the mass, although the airway control strategy was not commented on within the case reports. In our patient, who clinically was in impending respiratory failure, an awake intubation strategy was used to allow for spontaneous respiration during intubation. This strategy lessened the impact of mass effect on the airway during intubation. Had anesthetics been used, the airway may have been compromised due to decrease in muscle tone and sedation.

Even though the final diagnosis in our patient was determined by pathology, he had features suggestive of a thymic cyst in the homogenous appearance of the fluid with an air fluid level. Imaging may be beneficial, and either CT or MRI modalities are recommended, following initial evaluation by plain neck film.<sup>1,5</sup> However, despite imaging, preoperative diagnosis is quite difficult and lesions almost always require histological analysis for confirmation.<sup>5</sup>

#### CONCLUSION

The differential diagnosis for cervicothoracic lesions is broad. In addition to diagnosis, clinicians must carefully regard the airway impact of such masses. Clinical features and radiographic findings are helpful, but pathology is ultimately required for diagnosis.

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