A Case of Worsening Stridor in a Neonate

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A Case of Worsening Stridor in a Neonate

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The Case:

A 14-day-old female neonate with a known history of congenital hypothyroidism on supplemental thyroid hormone was admitted with worsening stridor since birth.

- She was noted to have intermittent stridor with agitation soon after birth.
- One week after birth, she developed mild stridor at rest that interfered with feeding and was associated with poor weight gain and intermittent cyanosis.
- She was referred to the inpatient unit by her pediatrician for urgent evaluation.

History:

- The infant was born at 38 weeks via a precipitous vaginal delivery.
- The prenatal history was remarkable for her mother having well-controlled lupus. Prenatal labs, including GBS, were negative and the pregnancy was unremarkable.
- The infant's birth weight was 2500 grams and Apgars were 5 and 7 at 1 and 5 minutes, respectively.
- She developed respiratory distress while in the delivery room and was monitored in the NICU for 4 days. A sepsis evaluation was negative.
- She was noted to have intermittent stridor with agitation while in the NICU. This was not present at rest and did not interfere with feeding and she was discharged home in good condition.
- At one week of age, the state newborn screen was positive for congenital hypothyroidism which was confirmed with formal thyroid testing: TSH was elevated at 52.8 µIU/ml (normal 0.35-5.5 µIU/ml) and free thyroxine was 1.37 ng/dL (normal 0.89-1.76 ng/dL). She was started on levothyroxine, 44 mcg daily, for 6 days before admission.

Physical Exam:

- At presentation, she was afebrile (98.3°F), pulse 150-170 bpm, respirations 23-33 breaths per minute and a blood pressure of 95/45. Her weight was 2540 grams.
- When resting quietly, there was no stridor or increased work of breathing and her oxygen saturation was 100% on ambient air.
- With crying, she developed prominent stridor with chest wall retractions, desaturation to 60% on ambient air with visible perioral cyanosis.
- She was otherwise nondysmorphic with a normal-appearing oropharynx and no palpable thyroid or neck masses.
- The remainder of her physical exam was normal.

Workup and Hospital Course:

- Initial laboratory investigation revealed normal CBC, electrolytes, and liver enzymes.
- Her TSH had normalized (5.05 µIU/ml).
- A noncontrast CT of the neck demonstrated a hyperdense mass at the base of the tongue consistent with an ectopic, lingual thyroid gland that extended into her proximal airway (Figure).
- During the course of her hospitalization, she was provided supplemental oxygen and positioning for feedings that significantly improved her respiratory symptoms and weight gain before discharge to home with close follow-up.

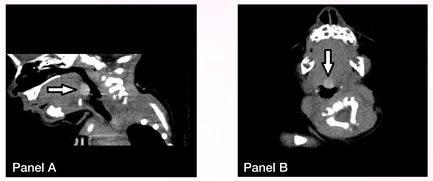

Figure. A noncontrast CT of the neck. Panel A, sagittal view; Panel B, axial view. White arrows denote lingual thyroid.

The Condition: Although 'noisy breathing' is common in neonates due to generally benign conditions, such as laryngotracheomalacia, acute or worsening stridor is a potentially life-threatening condition and warrants immediate assessment. Most cases of neonatal stridor are due to congenital lesions of the oropharynx and/or airway, although acquired conditions, such as infections, are less common at this age. Ectopic thyroid tissue is a recognized cause of airway obstruction in infants and should be considered in any neonate or infant with stridor.[1,2]

Pathophysiology: The thyroid gland begins to develop during the 3rd to 4th week of gestation from tissue located at the base of the tongue. During normal development, the fetal thyroid tissue descends along an anterior and midline tract through the thyroid duct to its final position at the base of the neck. Ectopic thyroid tissue results from a complete or partial failure of descent of the thyroid gland during fetal development. Ectopic thyroid tissue can occur anywhere in the oropharynx along the natural path of thyroid descent, with varying degrees of airway impingement and symptoms.[3]

The lingual area at the base of the tongue is the most common location for ectopic thyroid tissue (80-90%).[3] Estimates of the incidence of a lingual thyroid vary greatly and are reported to be 1:3,000 to 1:600,000 with a strong female predominance.[4,5] Although the presentation of a lingual thyroid can be acute and dramatic, as in our patient, symptoms can be more subtle, such as globus sensation, swallowing dysfunction, chronic cough, or respiratory symptoms. Lingual thyroids can even be asymptomatic and found incidentally in older ages.

Association with Hypothyroidism: Ectopic thyroid tissue and lingual thyroid are often associated with congenital hypothyroidism. In approximately 70-75% of cases, there is no other functional thyroid tissue except the ectopic gland[5], although this may not be sufficient for adequate infant growth and development.

Management: Treatment of a lingual thyroid is made on a case-by-case basis depending on the degree of obstruction and associated symptoms. Treatment of the underlying hypothyroidism can be expected to shrink the ectopic thyroid tissue over time, so adequate thyroxine supplementation and monitoring of TSH may be the only treatment required. Infants and older children with persistent or worsening airway obstruction or associated symptoms may require surgical ablation or excision of the ectopic thyroid tissue.

Conclusion:

A lingual thyroid gland is a rare and potentially life-threatening cause of stridor and should be considered in any neonate or infant with chronic or worsening stridor, particularly in the setting of congenital hypothyroidism.

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