Retropharyngeal Thymus and Parathyroid Gland: A Case Report

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ABSTRACT

Ectopic cervical thymus is a rare disorder. It occurs when thymic tissue arrests in its descent from the third pharyngeal pouch in the embryo to the mediastinum in the fetus. Most often it results in an asymptomatic neck mass somewhere along its pathway of descent, which is traditionally described as from the piriform sinus to the upper mediastinum. Rarely it can present with airway compromise, particularly in the neonatal population. This is a case of a retropharyngeal mass in a neonate causing upper airway obstruction. While clinically this mass behaved like a venolymphatic malformation, it ultimately proved to be aberrant ectopic thymus with an associated parathyroid gland. This represents the second known case of retropharyngeal thymus. It is proposed that the thymus and parathyroid glands have their embryologic origin more cephalad and posterior than previously described.

INTRODUCTION

Cervical ectopic thymus is an uncommon clinical problem. Of the approximately 100 total cases reported in the literature, ten percent occurred in infants.1 The typical presentation of ectopic cervical thymus is an asymptomatic neck mass.2 There is a single previous case report in the literature of cervical ectopic thymus which occurred in the retropharynx and caused airway compromise.3 Upper airway obstruction occurring in an infant requires urgent evaluation and treatment. While the differential diagnosis includes many more common etiologies, such as infection, trauma, neoplasm, and congenital abnormalities, retropharyngeal thymus is a potential cause of upper airway obstruction in a neonate.

METHODS AND MATERIALS

This is a case report of a neonate with retropharyngeal thymus causing dynamic upper airway obstruction and review of the literature.

RESULTS

A three-day-old male born at term presented with intermittent choking, coughing, desaturations, and cyanosis with oral feeds. On flexible fiberoptic nasopharyngoscopy a nasopharyngeal mass was identified which was highly dynamic with respirations. Magnetic resonance imaging (MRI) of the neck demonstrated a high retropharyngeal soft tissue abnormality with localized mass effect possibly representing an atypical venolymphatic malformation (Figures 1 and 2). The patient required a tracheotomy for a stable airway. After watchful waiting with repeat imaging and endoscopy, the mass was ultimately biopsied and found to be ectopic thymus with an associated histologically normal appearing parathyroid gland.

DISCUSSION

Embryologically, the thymus is derived primarily from the third pharyngeal pouch, with a small contribution from the fourth pharyngeal pouch. The dorsal portion of the third pharyngeal pouch gives rise to thymus, and the ventral portion gives rise to the inferior parathyroid gland. The thymus then descends from the piriform sinus, through the thyrohyoid membrane, and continues anteroinferiorly along the carotid sheath until the two lateral portions fuse in the midline and descend into the upper mediastinum.4 Aberrant ectopic thymus may occur anywhere along this pathway when arrest of descent occurs.

There have been only approximately 100 cases of solid cervical ectopic thymus reported in the literature, with less than ten of these occurring in infants.1 Most commonly, these cases present as an asymptomatic neck mass. The single prior case of retropharyngeal ectopic thymus that has been published also occurred in an infant with airway compromise.3 The present case is unique in that a normal appearing parathyroid gland was also present.

CONCLUSIONS

This case demonstrates the importance of considering ectopic thymus in the differential diagnosis for congenital neck mass, even in cases not typically associated with ectopic thymus. While the embryological path of descent of the thymus gland is classically described as beginning at the angle of the mandible and ending in the mediastinum, the present case is the second reported of ectopic thymus in the retropharyngeal space. Further, an ectopic parathyroid gland was intimately associated with the ectopic thymus in the present case, a finding not present in the prior case report. Given that both the thymus and the inferior parathyroid are third pharyngeal pouch derivatives, it stands to reason that both begin their descent more cephalad and posterior than previously thought.

REFERENCES