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Case Report

Ectopic thymus as a cause of Horner's syndrome

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ABSTRACT

Ectopy of the thymus is a rare anomaly arising during fetal development, where the thymus does not make a complete decent into the thoracic cavity where it should involute in adolescence. The most common complications of an ectopic thymus include tracheal or esophageal compression presenting in childhood. This is a report of a single case of ectopic cervical thymus identified in a 2-month-old infant presenting with Horner's syndrome. Thymic ectopy should be on the differential when performing a radiologic evaluation of a neck mass when imaging characteristics are similar to thymic tissue.

Introduction

The thymus is the primary lymphoid organ in infancy and early childhood and facilitates the differentiation and maturation of T lymphocytes [1]. The thymus arises from the third pharyngeal pouch, eventually forming thymic primordial that descends into the anterosuperior mediastinum. As children age into adolescence, the thymus involutes under the influence of surges in circulating sex hormones [1].

Ectopy of the thymus most commonly occurs in the first trimester of pregnancy during organogenesis and migration, and thus ectopic thymic tissue can be found anywhere from the angle of the mandible to superior mediastinum [2]. Abnormalities are often associated with immunodeficiency diseases such as DiGeorge syndrome, autoimmune diseases such as myasthenia gravis, and malignancies associated with paraneoplastic syndromes [1]. Symptomatic ectopic cervical thymus commonly presents in childhood secondary to tracheal or esophageal compression.

The prevalence of ectopic thymic tissue in children is very low, with reports ranging from 0.99% to 1.8%, while scant reports exist on published cases of presentation in infancy [1,3,4]. The following case is that of an infant presenting with Horner's syndrome due to the compressive effects of a neck mass that was found to be ectopic thymic tissue.

Case description

A 3-week-old girl initially presented to an outpatient pediatrician with unequal pupil size. Her parents first noticed the anisocoria at 6 days of age, reporting that the right pupil was

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larger than the left. At that time, the parents denied any observable ptosis or other neurological symptoms, nor were any further remarkable findings noted in the physical exam. She was born at full-term and was otherwise healthy in the first week of life. The pregnancy was uncomplicated and the labor atraumatic.

Given the patient's age, additional testing was deferred and close clinical follow up was planned. At 2 months of age, the parents reported by phone symptoms consistent with left upper lid ptosis. The patient was brought in for urgent evaluation where she was found to have persistent unchanged anisocoria and left lower eyelid reverse ptosis. She underwent an apraclonidine test in clinic, which was positive in the left eye given reversal of anisocoria and periodic heart rate decrease throughout the exam. The clinical history, course, and exam findings suggested Horner's syndrome and further imaging was ordered to investigate potential etiologies. Otherwise, serum and urine studies were unremarkable.

Magnetic resonance imaging (MRI) of the chest, abdomen, and pelvis were performed, but did not reveal any abnormalities. Of note, normal thymic tissue was seen in the chest (Fig.1). The patient then underwent a neck MRI, which demonstrated a 12 × 20 × 19 mm soft tissue mass within the left submandibular space of the neck at the level of the angle of the mandible, situated just anterior to the left common and internal carotid arteries, compressing the carotid space (Fig. 2). The neck mass was smooth, well circumscribed, lobulated, had very homogenous signal on T1 and T2 sequences and enhanced similar to the adjacent salivary gland. The mass had similar signal characteristics to the normal thymic tissue within the chest (Fig. 1).

The patient then underwent operation to remove mass pressing on the left sympathetic chain. Review of the surgical specimen by pathology demonstrated ectopic thymus.

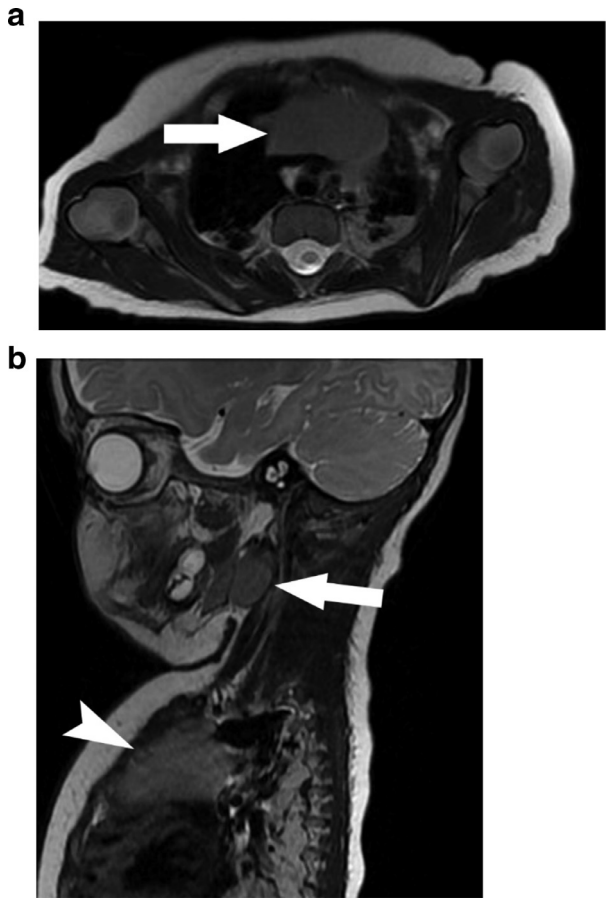

Fig. 1 – Thymic tissue in the mediastinum: (a) Normal homogenous thymic tissue is seen in the anterior superior mediastinum on axial T2 images (arrow). (b) The neck mass (arrow) and the thymus (arrowhead) can both be seen on the sagittal T2 series with similar signal characteristics. Posterior displacement and compression flow void of the internal carotid at the tip of the arrow is seen.

Discussion

Horner's syndrome is a neurologic oculo-sympathetic paresis characterized by the classic triad of miosis, ptosis and anhidrosis. In pediatric populations, the addition of enophthalmos ipsilateral to the lesion secondary to an interruption of ocular sympathetic innervation is frequently present. Lesions can arise anywhere along the sympathetic pathway between the hypothalamus through the level of T2 spine [5]. In adults, the etiology and presentation of Horner's syndrome can be broken down by subtype based on location; first order neuron central lesions arising from the hypothalamus, brain stem and cervicothoracic spinal cord; second order preganglionic lesions arising from the pulmonary apices or thyroid; and third order post ganglion lesions arising from skull base, cavernous sinus, internal carotid artery, or superior cervical ganglion [5]. In children, etiologies are less established based on location given the rarity of presentation, and as such are often classified as either congenital (defined as diagnosed within 4 weeks after birth) or acquired. The most common congenital and acquired cause of Horner's syndrome in the pediatric population is neuroblastoma, followed by central astrocytoma and rhabdomyosarcoma [6]. The remaining causes are often iatrogenic or idiopathic from birth related trauma or surgery, congenital infections or vascular disease and venous malformations that exert a compressive effect on neurons that innervate the sympathetic chain. Imaging, such as MRI, can be a powerful, noninvasive tool in identifying the location of a lesion and differentiating etiologies of Horner's syndrome due to pediatric neck masses. Neoplastic lesions such as neuroblastoma are more likely to appear as nonhomogeneous and cystic enhancing masses while birth related trauma to the brachial plexus might exhibit nerve root edema, enhancement of the nerve roots, or pseudo-meningoceles. Vascular malformations would demonstrate flow voids and be much more heterogenous on conventional MRI.

In the case above, Horner's syndrome resulted from the compressive effects of an ectopic cervical thymic neck mass on the sympathetic pathway. This presentation is exceedingly rare, with only 2 reported cases in the literature [2,7,8]. Ectopic cervical thymic tissue can be found anywhere from the angle of the mandible to superior mediastinum, and if adjacent to the sympathetic chain ganglion, can lead to a Horner's

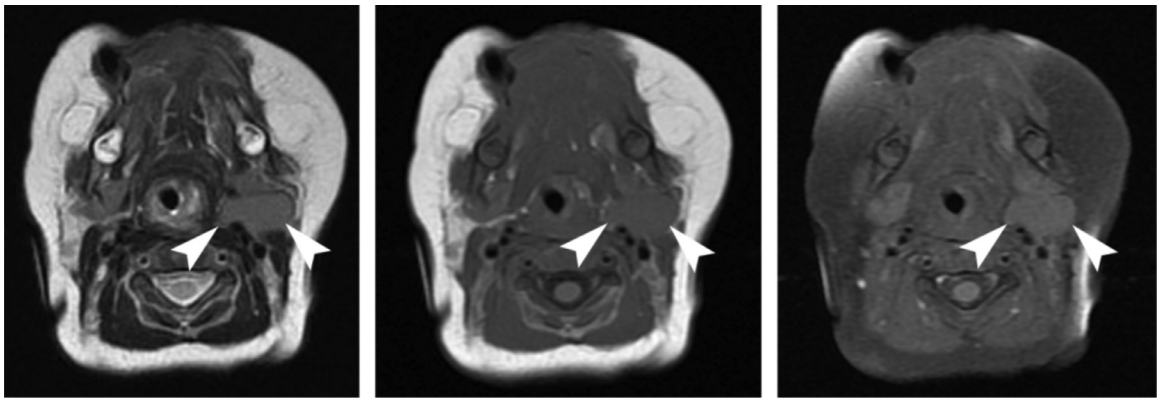

Fig. 2 – (a) (left): Axial T2 weighted MRI of the neck demonstrates a homogenous well demarcated mass with isointense signal to the thymic tissue seen on Figure 1 immediately anterior to the left internal carotid artery (arrowheads). No flow voids are seen in the mass. (b) (center): Axial T1 weighted MRI of the neck shows the mass is slightly hyperintense to muscle on T1 (arrowheads). (c) (right): Postgadolinium contrast T1 fat saturated image shows the mass enhances similar to the adjacent submandibular gland.

syndrome [9]. On MRI, both normal and ectopic thymic tissue will appear homogeneous on T1 and T2 sequences. On T1 and T2 weighted imaging, the signal intensity will be greater than that of muscle and should lack septations, calcifications, cysts [10].

Conclusion

Although extremely rare, an ectopic cervical thymus should be included in a comprehensive differential alongside more common causes of a pediatric neck mass causing Horner's syndrome when the signal characteristics of the mass are similar to thymic tissue. MRI is the most accurate noninvasive method for diagnosis of pediatric neck mass, with location to adjacent structures and imaging characteristics aiding in the differentiation between neoplastic, congenital, and idiopathic etiologies.

Integrity of research and reporting

This article does not contain any studies with human participants or animals performed by any of the authors. Therefore, this article is exempt from IRB review.

For this type of study, informed consent is not required.

Consent to publish was obtained by patient for case report.

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