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Cervical thymic cysts

Hongzhao Ji, MD, Eric Gantwerker, MD, MS

From the Department of Otolaryngology, Head and Neck Surgery, University of Texas Southwestern Medical Center, Dallas, Texas

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Thymic cysts are rarely occurring benign neck masses, most frequently seen in children. They are remnants of the third and fourth branchial pouches. They may occur at any point along the thymic tract, though they have a predilection for the left side of the neck, often deep in the neck. Patients often present with compressive symptoms. Treatment is surgical excision. Care must be taken, however, to confirm the presence of thoracic thymic tissue before excision of this neck lesion. On histology, they are defined by the presence of thymic parenchyma and Hassall corpuscles in the cyst wall.

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Introduction

The thymus is a lymphatic organ, normally situated in the thorax. The name of the organ is believed to originate from its appearance, which resembles the thyme plant. Other possible etymologies of the name come from the ancient Greek word for “smoke” or “soul”, as the thymus was believed to be the seat of the soul owing to its proximity to the heart and otherwise unknown function at that time. This also led to the Latin name “animella”, the small soul, used in culinary tradition to describe the thymic tissues.¹

In antiquity, it was known to diminish with age, but for most of history was thought of as a cushioning structure for other thoracic structures. Its nature as a lymphatic organ was first published in 1774. The first known surgeries to remove enlarged thymus for symptomatic hypertrophy occurred in the late 1800s. The functions of the thymus in the immune system and its role in the maturation of T cells were not discovered until the last decades of the 20th century.¹

Embryology

The thymus epithelium derives from the ectoderm and endoderm of the third and fourth pharyngeal pouches. The mesenchyme develops from the neural crest cells associated with these pouches. These neural crest cells arise at stage 10 of human embryologic development (23 days).¹

The third branchial pouch gives rise to the inferior parathyroid gland and the thymus; the former travel inferiorly with the rest of the third branchial pouch whereas the thymic buds separate from their ipsilateral third branchial pouches at approximately the end of the fourth week (Figure 1). These 2 buds, then travel inferiorly and are joined to each other in the aortic sack by connective tissue only between the fourth and seventh weeks. The remaining connection with the third branchial pouch is then lost, but a cellular cord may persist.² With second branchial cleft cysts, the tract usually persists between the internal and external carotids, whereas thymic tracts usually travel between the carotid and the vagus nerve.¹

The cells of the thymus then form a densely packed epithelial mass, which becomes a loose epithelial reticulum. Lymphoid stem cells then migrate from bone marrow to fill this reticulum during the third month of gestation. Lymphocytes and dendritic cells appear, followed by vascular mesenchyme and vagal nerves.² The medulla and cortex differentiate when the embryo is at approximately 40 mm in length. Hassall corpuscles, whorl-like clusters of

Address reprint requests and correspondence: Eric Gantwerker, MD, MS, Department of Otolaryngology—Head and Neck Surgery, University of Texas Southwestern Medical Center, 2350 N. Stemmons Freeway, F6.218, Dallas, TX 75207.

E-mail address: Eric.Gantwerker@UTSouthwestern.edu

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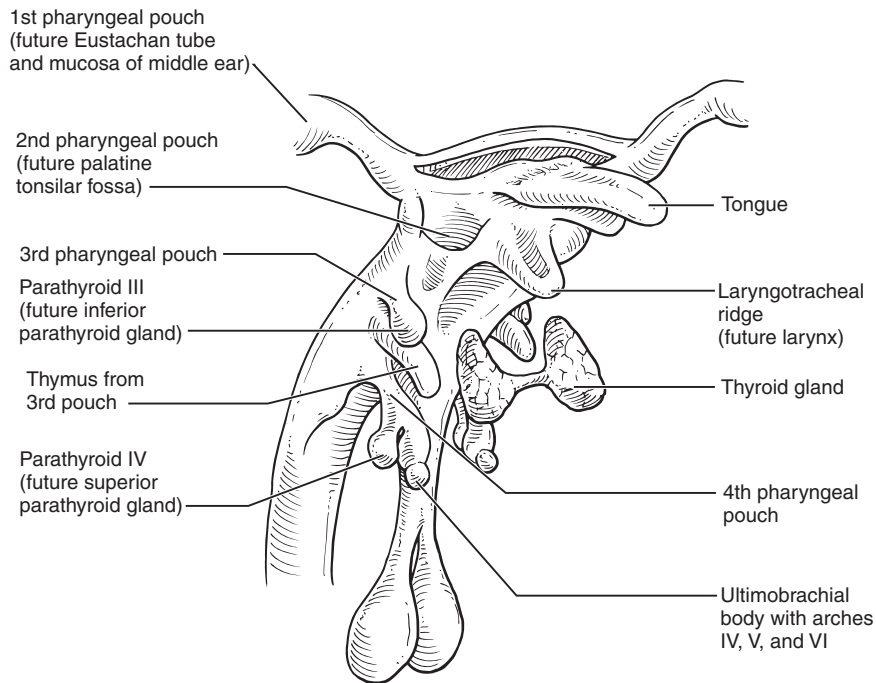


Figure 1 Embryologic anatomy showing development at 6-7 weeks. Thymic buds have separated from the rest of the third branchial pouch structures and will travel inferiorly.

cytoreticulum, then appear. T-lineage lymphoblasts comprise more than 95% of cells by the 10th week. The thymus is fully developed by the 17th week.

In 1938, 5 possible mechanisms of thymic cyst formation were described. The 2 most accepted theories are (1) thymic cysts develop from congenital persistence of the thymo-pharyngeal ducts and (2) cysts arise from Hassall corpuscle degeneration in ectopic rests of the thymus. During development, there is the potential for thymic tissue to be left at any point along the path of embryological migration from the neck to the thorax.³

Anatomy

The thymus initially increases in size over the first year of life, with a mean weight of approximately 25 g at 1 year of age. This weight is maintained for the first 6 years of life, at which time the gland begins to involute. This may be triggered by elevated levels of sex hormones, as evidence by the fact that castration, either physical or chemical, has been associated with increased size and activity of the thymus. The thymic lymphatic tissue decreases in size annually by about 5% per year until the 10th year of life. After this point, lymphocytes begin to infiltrate the thymic medulla until the 25 years of age. These adipocytes then begin to infiltrate the cortex until the 40th year of life. During this time, thymic involution continues at a rate of approximately 5% per year until after the fourth decade of life when involution slows down and most of the thymus is fatty tissue. Islands of active thymic tissue, however, do persist into old age.

The normal thymus exists in the anterior thorax, separated into a right and left, connected by only connective tissue. The 2 sides of the thymus descend separately and can both leave persistent cellular tracts. Ectopic areas of thymus

usually exist along the neck, from the angle of the mandible superiorly to the mediastinum inferiorly (Figure 2). Residual thymic tracts tend to be associated with the pyriform sinus, unlike second branchial cleft cysts, which tend to track into the tonsillar fossae.

Most of the thymic cysts reported are multilobular; however, unilobular cysts are also reported.⁴ Thymic cysts occur on the left side more frequently than the right with several authors reporting 2-3:1 left sided predominance.^{5,6} Also reported is a cyst in the midline or pharynx in 5%-7% of patients. The airway can often be affected, but it is typically only minimally displaced by the mass. Symptomatic mass effect on the airway is largely limited to infants.

Thymic cysts are usually deep to the platysma muscle and often even deep to the sternocleidomastoid (SCM) muscle. They are frequently associated with, though outside, the carotid sheath, usually from the posterior or medial aspect, and can often extend into the retropharyngeal space.^{7,8}

Some authors have reported unusual anatomical associations and presentations of thymic cysts. These include involvement of the parotid, connection to a thyroglossal duct cyst, sinus tracts that extend to the skin in the neck, masses that increase with valsalva, masses on the anterior border of the SCM, connection to the mediastinal thymus, and presentation as a mass associated with parathyroid and even cartilage.^{9,10}

Histology

Histology is currently the only way to definitively diagnose thymic cysts. The typical histology of a thymic cyst shows cholesterol clefts surrounded by histiocytic giant cells, thymic cyst wall and, most importantly, Hassall corpuscles.

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