Thymopharyngeal Duct Cyst

An Unusual Variant of Cervical Thymic Anomalies

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Background: The thymus develops from the third pharyngeal pouch and descends from the neck into the anterior-superior mediastinum. Thus, it is possible to have thymic remnants in the neck, which most often present as a cervical mass during childhood. One type of cystic thymic remnant is the thymopharyngeal duct cyst, a remnant of one of the paired tracts of embryological thymic descent. Thymopharyngeal duct cysts are rare lesions that can have a similar presentation to more commonly encountered childhood neck masses.

Objectives: To review the embryological development of cervical thymic remnants and to report our experience with the thymopharyngeal duct cyst.

Design: Case series.

Setting: Tertiary care center.

Patients: Two children who presented with asymptomatic neck masses that were caused by cystic remnants of the thymopharyngeal duct.

Results: Both patients underwent preoperative computed tomography, which revealed a multiloculated mass coursing adjacent to the carotid sheath. Surgical treatment was the definitive therapy for both patients, although neither patient had a definitive preoperative diagnosis. In both cases, the mass was approached through an incision anterior to the sternocleidomastoid muscle, and dissection proceeded along the length of the carotid sheath. A fibrous cord extending into the mediastinum was found in both patients. There were no postoperative complications. Histopathologic evaluation revealed the presence of mature thymic elements within the wall of a multiloculated cyst.

Conclusions: Thymopharyngeal duct cysts must be considered in the differential diagnosis of pediatric neck masses. Computed tomography is helpful to delineate the relationship to the carotid sheath. Complete surgical excision is the appropriate therapy in a majority of cases, with minimal morbidity when careful attention is paid to vital structures.


The word thymus derives from the Greek thymos, meaning “soul” or “spirit.” Galen theorized that the function of the thymus was that of a cushion, to protect the mediastinal vessels from the overlying sternum. It was not until the late 1700s that an association was suggested between the development of the lymphatic system and the thymus. It is currently accepted that the thymus serves as a central lymphoid organ that is responsible for maturation of T lymphocytes and induction of self-tolerance.

The thymus is derived primarily from the third pharyngeal pouch in association with the inferior parathyroid glands, with a small contribution from the fourth pharyngeal pouch. Beginning in the sixth gestational week, the right and left portions of the thymic primordium separate from the pharynx and descend into the anterior-superior mediastinum along paired thymopharyngeal stalks. The path of descent involves migration deep to the thyroid gland and sternocleidomastoid muscle along the carotid sheath. In normal development, the inferior portions of the thymopharyngeal stalks enlarge, while the proximal portions form epithelial cords that eventually atrophy.

Cervical remnants of thymic tissue have been found in up to 30% of infants studied at autopsy. When these remnants present clinically as a mass in the neck, it is important to consider the range of variants that exist, based on the embryological path of development. Zarbo et al categorized 7 cervical thymic lesions, distinguishing them by location and by whether the tissue was solid or cystic. Whereas accessory cervical thymus and...
cervical thymic cyst variants represent more than half of all cervical thymic anomalies, persistent thymopharyngeal duct cysts represent only 7% of these fetal remnants. We describe 2 patients with lateral neck masses composed of cystified remnants of the thymopharyngeal duct.

REPORT OF CASES

CASE 1

A 9-year-old boy was referred for evaluation of a mass in the left side of the neck that had been apparent for 4 months. He was asymptomatic and had no comorbidities. On physical examination, a ballotable, nontender mass with indistinct borders was noted in the left side of the neck, extending from the level of the hyoid to the clavicle. The carotid pulse was palpable and transmittable through the mass. A contrast-enhanced computed tomographic scan revealed a somewhat poorly defined, multiloculated cyst in the middle and lower areas of the left side of the neck, extending toward the thymus (Figure 1).

A decision was made to remove the mass surgically. An incision was made along the anterior border of the sternocleidomastoid muscle, extending from mastoid tip to the sternal notch. Superiorly, the mass was encountered adjacent to the internal jugular vein and superficial to the carotid artery and vagus nerve. Blunt dissection was used to elevate the mass from these structures, proceeding in an inferior direction. A dense capsule was identified surrounding the mass as it extended into the mediastinum. As the mass was bluntly retracted into the neck, a fibrous cord was found to be trailing inferiorly toward the thymus. This cord was transected. The patient had an uneventful postoperative course and was discharged home on postoperative day 3.

Gross examination revealed a multiloculated cyst measuring 26 cm in greatest length and 5 cm in greatest diameter. Microscopic examination demonstrated the presence of thymic tissue within the cyst wall, characterized by a lymphoid predominant matrix with numerous Hassall corpuscles (Figure 2).

The patient has been followed up for 18 months, without evidence of recurrence.

CASE 2

A healthy 11-year-old boy presented with a 2-month history of an enlarging left neck mass. On physical examination, the mass was palpable deep to the sternocleidomastoid muscle, extending from mandibular angle to the level of the cricoid cartilage. Although the carotid pulse was palpable through the mass, no bruits were auscultated.

A contrast-enhanced computed tomographic scan revealed a multiloculated cystic mass adjacent to and ramifying within the carotid sheath structures (Figure 3). At the level of the subglottis, the lesion was noted to be splaying the internal jugular vein and the common carotid artery. Based on the findings of imaging, which showed that a multiloculated mass was present throughout multiple fascial layers, the preoperative diagnosis was lymphangioma.

Intraoperatively, the mass was approached through an incision anterior to the sternocleidomastoid muscle. Blunt dissection was used to free the mass from the internal jugular vein between the mandibular angle and the omohyoid muscle. Coursing inferiorly into the mediastinum was a fibrous stalk, which was transected. The patient had an uncomplicated postoperative course and was discharged home on postoperative day 2.

Gross examination of the specimen revealed a multiloculated cystic mass, measuring 7.0 × 1.2 × 1.5 cm. Microscopic examination revealed a cyst wall lined by squamous epithelium. Hassall corpuscles, cholesterol clefts, and lymphoid aggregates were found within the cyst wall.

The patient has been followed up for 4 months, without evidence of recurrence.

Figure 1. A, Contrast-enhanced computed tomographic scan demonstrating a poorly defined cystic mass (arrow) splaying the internal jugular vein and common carotid artery. B, The mass (arrow) is noted to be descending into the superior mediastinum toward the thymus gland.
The mechanisms that account for a retained cervical thymus include arrest in migration, failure of involution, or sequestration of thymic tissue during descent. Any of these processes results in cervical rests of solid thymic tissue, which will generally be asymptomatic. Cervical thymic cysts are more important clinically, as they will have a similar clinical appearance to other congenital neck masses and may demonstrate progressive growth. The transition from solid to cystic thymic tissue has been hypothesized to occur as a result of cystic degeneration of Hassall corpuscles or cystic change in persistent remnants of the thyropharyngeal duct. A classification system for cystic thymic remnants has been proposed, differentiating these lesions based on structures of origin. True cysts are those lesions that originate in maldevelopments of the thyropharyngeal duct and may span the entire length of the neck with or without a patent fistula tract to the pyriform sinus. Mixed cysts contain elements of second branchial cleft and third pharyngeal pouch origin and are considered thymic fistulas. Those lesions derived from branchial cleft remnants, with incidental thymic tissue present, are referred to as false thymic cysts.

Cervical thymic cysts are rare in comparison to other congenital neck masses, such as thyroglossal duct cysts, lymphangiomas, and branchial cleft cysts. Nicollas et al, in their retrospective review of 191 congenital neck masses, found that only 2% were thymic cyst malformations. All cervical thymic cysts are characterized as congenital lesions; accordingly, the majority of patients present in the first decade of life. Males are more commonly affected, and there is a higher incidence of left-sided lesions. Lesions are most often asymptomatic, although

Figure 2. A, On low-power magnification, mature thymic elements are depicted within the cyst wall. B, At higher magnification, it is easy to appreciate the presence of Hassall corpuscles (arrow); also note cystification within a Hassall corpuscle (arrowhead) (hematoxylin-eosin, original magnification ×40).

Figure 3. A, Contrast-enhanced computed tomographic scan demonstrating a cystic mass (arrow) at the level of the mandibular angle. B, More inferiorly in the neck, the mass (arrow) is well defined and ramifies within the carotid sheath structures.
there have been reports of associated hoarseness, dysphagia, and stridor, especially in neonates. On physical examination, thymic cysts are usually palpable deep to the sternocleidomastoid muscle. Because of the intimate relationship between these lesions and the carotid sheath, the carotid pulse will often be palpable through palpation of the mass. By definition, when these masses span the length of the neck from mandibular angle to clavicle, they are termed thymopharyngeal duct cysts. It may be difficult to completely palpate the inferior aspect of these masses, because in the 50% of cases that maintain a connection with the mediastinal thymus, there is an atrophic fibrous cord attachment. Occasionally, a thymopharyngeal duct cyst will have a fistula tract to the pyriform sinus, in which case a barium esophagram may be of diagnostic assistance.

Contrast-enhanced computed tomographic scans are important for distinguishing thymic cysts from other congenital neck masses, such as second branchial cleft cysts or lymphangiomas. Second branchial cleft cysts occur superficial and lateral to the internal jugular vein and common carotid artery, while lymphangiomas occur most often in the posterior triangle of the neck, often violating normal fascial planes. It is only thymic cysts that maintain their relationship with the carotid sheath, based on the embryological path of migration. Magnetic resonance imaging has been used to confirm an association between the cervical neck mass and the mediastinal thymus because of its ability for greater soft tissue delineation. It also may assist in determining whether the cervical mass represents the only source of thymic tissue. Complete excision of a cervical thymic mass should be deferred in a young child with no evidence of mediastinal thymic tissue. Immune dysfunction after total thymectomy in children has been reported.

Although not always clinically indicated, fine-needle aspiration may assist in proper management. If the findings of computed tomography support a diagnosis of thymic cyst, the results of fine-needle aspiration can confirm the presence of thymic tissue and thus promote further investigation as to the presence of a mediastinal thymus prior to surgical therapy. Complete surgical excision of the mass is the recommended therapy in most cases. When undertaking surgery for suspected thymopharyngeal duct cysts, one must be prepared for dissection along or within the carotid sheath into the superior mediastinum. The presence of an atrophic fibrous cord can be a clue as to the inferior extent of dissection. As previously mentioned, if workup reveals the absence of a mediastinal thymus, one may consider excisional biopsy or deferring surgery until the patient has completed immune system development.

Thymic cysts are most often multiloculated lesions with a smooth, fibrous capsule. Unlike other congenital neck masses, the cyst has an epithelial lining of the columnar, cuboidal, or squamous variety. Within the cyst wall, it is common to find lymphoid aggregates, granulomas, and cholesterol clefts. The pathognomonic histological finding in thymic lesions is the presence of Hassall corpuscles, which are formed from swirls of keratinizing epithelial cells originating in the thymic medulla.

Prognosis is excellent after complete surgical excision, with minimal morbidity. Of the cases of thymic cysts described in children, to our knowledge there have been no recurrences reported in the literature.

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