Thymopharyngeal Duct Cyst: A Case Presentation and Literature Review

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Abstract

The differential diagnosis of a lateral cystic neck mass includes branchial cleft cysts and lymphangiomas. Cervical thymic cysts are rare, and thymopharyngeal duct cysts, also known as thymic thymopharyngeal duct cysts, are uncommon. The surgeon must be prepared to dissect along the carotid sheath and into the mediastinum. For a suspected thymopharyngeal duct cyst, cervical laryngoscopy identifies the fistula tract often before the pre-operative diagnosis. The thymic cyst can present with mediastinal extension in up to 50% of cases. Direct laryngoscopy identifies the fistula tract extending into the anterior mediastinum. Transillumination is a non-specific finding. Cyst expansion with a Valsalva maneuver suggests pharyngeal continuity.

Case Report

A 6-year-old boy developed a lateral left-sided cystic neck mass after an antecedent upper respiratory tract infection. He was febrile at presentation without leukocytosis. An ultrasound profiled a complex cystic structure measuring 5x3x5 cm (Image 1). Subsequent aspiration yielded only polymorphonuclear leukocytes, but no organisms. The CT scan uncovered a heterogeneous multicollated cyst obscuring the left thyroid lobe and causing right-sided tracheal deviation (Image 3). Of note, there were inflammatory changes surrounding the mass that extended superiorly to the left piriform sinus. The left thyroid lobe was not palpable. A subsequent ultrasound profiled a complex cystic structure measuring 5x3x5 cm (Image 1).

Discussion

Lateral cystic neck masses were first described in 1785 by Hunczovsky. Over the past two centuries, a wealth of names and theories about this entity have been described by well recognized medical figures including Langenbeck, Luschka, and Virchow. Currently, the differential of a lateral cystic neck mass includes lymphangiomas, branchial cleft cysts, thyromimthic cysts, and thymopharyngeal duct cysts. Despite the diverse nomenclature, this differential has engendered over time, the thymic cyst can present with mediastinal extension in up to 50% of cases. Transillumination is a non-specific finding. Cyst expansion with a Valsalva maneuver suggests pharyngeal continuity.

By definition, a thymic cyst that involves the neck from the mandible to the clavicle is a thymopharyngeal duct cyst; it traverses the aforementioned anatomic structures. The presence of a fistula tract to the piriform sinus is a feature of some of these cysts, but is not a criterion for diagnosis. Three major imaging modalities are used to aid diagnosis. Ultrasound has been reported as useful in distinguishing cystic versus solid structure, assessing proximity to the carotid sheath, and in some instances, identifying thymic tissue specifically by echo patterns. CT gives important information to distinguish thymic cysts from other congenital anomalies such as lymphangiomas and branchial cleft cysts based on specific anatomic location and appearance. CT also provides information regarding proximity to vital structures that optimize operative planning. A contrast study shows a homogenous hypodense mass with minimal rim enhancement when imaging congenital thymic cysts. Air fluid levels within the cyst can indicate foreshadowed communication. The MRI shows low T1 signal and high T2 signal as well as superior soft tissue definition. This study is particularly useful in imaging a fistula tract connecting to the pharynx as well as determining the relationship between the cyst and mediastinal thymus, an area in which the CT falls short. It is crucial to radiographically determine the presence of normal thymic tissue to ensure immunocompetence. Theoretically, a barium esophagram can confirm the presence of a piriform fistula, but little data is available to support its efficacy.

Definitive surgical excision of the cyst is the treatment of choice once the diagnosis is confirmed. If further analysis reveals the absence of mediastinal thymic tissue, surgery can be delayed until immunologic development is complete. Once the presence of normal thymic tissue is confirmed, a transverse cervical neck incision is the most common approach, although vertical approaches parallelizing the anterior sternocleidomastoid border have been reported. For a suspected thymopharyngeal duct cyst, the surgeon must be prepared to dissect along the carotid sheath and into the mediastinum. A connection between the cyst and pharynx is often unknown pre-operatively despite appropriate imaging and laryngoscopy; therefore it is imperative intraoperatively to meticulously dissect out any fistula tracts as failure to do so can result in cyst recurrence. Once resected, histologic analysis is critical to confirm the suspected diagnosis.

The majority of thymopharyngeal duct cysts have a multicollated structure. The cyst contents are highly variable ranging from clear to dark brown turbid fluid. The white flecks commonly found in the fluid represent cholesterol crystals. The capsule is composed of dense fibrous tissue containing epithelium of cuboidal, columnar, or squamous varieties and frequently contains cholesterol granulomas. The pathognomonic histologic finding is the presence of Hassall’s corpuscles within the cyst wall. These are described as conglomerates of “keratinizing epithelial cells originating in the thymic medulla.” This feature differentiates thymic cysts from branchial cleft cysts and thymomas, which can become cystic in up to 40% of cases. This particular histologic finding is paramount in establishing an accurate diagnosis.

References


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