Congenital Cystic Neck Masses: Embryology and Imaging Appearances, With Clinicopathological Correlation

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Congenital cystic masses of the neck are uncommon and can present in any age group. Diagnosis of these lesions can be sometimes challenging. Many of these have characteristic locations and imaging findings. The most common of all congenital cystic neck masses is the thyroglossal duct cyst. The other congenital cystic neck masses are branchial cleft cyst, cystic hygroma (lymphangioma), cervical thymic and bronchogenic cysts, and the floor of the mouth lesions including dermoid and epidermoid cysts. In this review, we illustrate the common congenital cystic neck masses including embryology, clinical findings, imaging features, and histopathological findings.

Introduction

Congenital cystic masses of the neck are uncommon but important pediatric neck lesions. Many of these have characteristic locations and imaging findings. The most common congenital cystic neck mass is thyroglossal duct cyst (TGD).[1] The other common lesions are second branchial cleft cyst and cystic hygroma (lymphangioma). The uncommon cystic lesions include first and third branchial cleft cysts, fourth branchial apparatus anomaly, cervical thymic and bronchogenic cysts, and the floor of the mouth lesions including dermoid and epidermoid cysts. Anomalies of the branchial apparatus could also present as sinuses and fistulae in addition to cysts. In this article, we discuss the common congenital cystic neck masses in an orderly fashion, including embryology and pathogenesis, clinical findings, imaging features, and histopathological findings.

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A. Thyroglossal Duct Cyst

Embryology and Pathogenesis

The thyroid gland develops in the region of foramen cecum of the tongue during the third gestational week. The thyroid anlage descends down to reach the thyroid bed anterior to laryngeal cartilages through the TGD by the seventh week of gestation, and TGD begins to involute by 8-10 weeks of gestation.[2] If any segment of the TGD fails to involute, then the persistent secretory activity from the epithelial lining owing to repeated infection or inflammation would give rise to TGD cyst.[1]

Clinical Findings

Most TGD cysts are located either at or below the level of the hyoid bone, with 50% at hyoid bone and 20%-25% in suprahyoid neck, often in the midline. The remaining 25% are located in the infrahyoid neck where they are in the midline or within the strap muscles in a paramidline location (Fig 1).[1] The TGD cyst commonly presents as a painless, enlarging mass in a child or young adult with varying sizes ranging from 0.5-6.0 cm. The duct and cyst characteristically

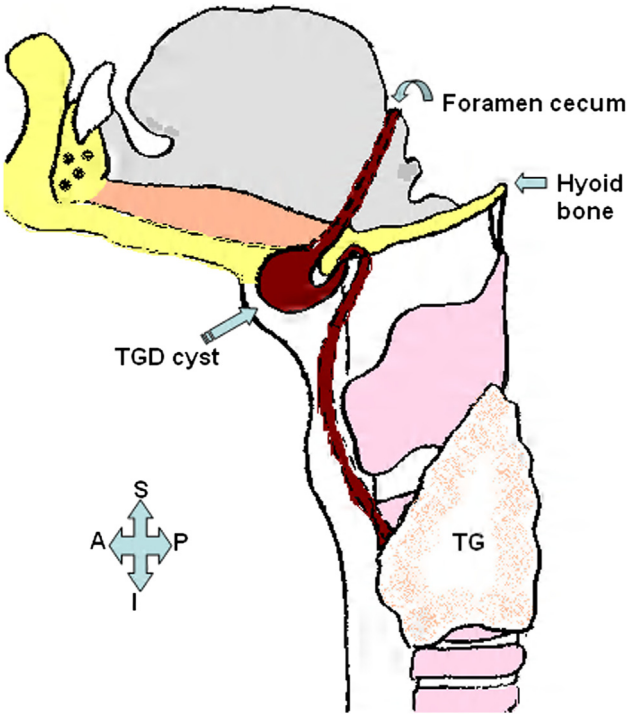

FIG 1. Schematic representation of the thyroglossal duct (TGD) extending from the region of foramen cecum (curved arrow) up to the thyroid gland (TG). Note the close relationship of TGD cyst (broken arrow) with the hyoid bone (solid arrow). (Color version of figure is available online.)

move upward with tongue protrusion. Most of the times the TGD cyst comes to notice owing to repeated infection.[3] Another rare but worrisome complication of the TGD cyst is its association with thyroid carcinoma. Complete excision of the cyst with resection of the central portion of the hyoid bone and a core of tissue following the expected course of the TGD to the foramen cecum (Sistrunk procedure) is the recommended surgical approach. This procedure involves a recurrence rate of 2.6%.[4,5]

Imaging

An uncomplicated TGD cyst has the appearance of a cystic mass either in the midline of the anterior neck at the level of the hyoid bone or within the strap muscles in paramidline location. On ultrasound (US), an anechoic mass with a thin outer wall in close association with the hyoid bone easily establishes the diagnosis of a TGD cyst. However, this "classic" appearance is seen in less than half (42%) of the cases. Other manifestations of these cysts include hypoechoic masses with increased through-transmission, homogeneous or heterogeneous with variable degrees of fine to coarse internal echoes, and may be associated with thick cyst walls (Fig 2A). Fluid levels may be seen owing to settling in of debris. There is no correlation between the sonographic appearance and pathological evidence of infection and inflammation. Heterogeneity seen in TGD cysts on sonograms is more likely due to the proteinaceous content of the fluid secreted from the cyst wall rather than to infection.[6] Preoperative sonographic visualization of normal thyroid tissue is sufficient to exclude a diagnosis of ectopic thyroid tissue and obviates routine thyroid scintigraphy. Pericystic fluid or inflammatory changes have been described in infected TGD cyst.

On computed tomography (CT) scans, a TGD cyst is a benign-appearing cystic neck mass, which may show septations in it. Suprahyoid TGD cyst (Fig 2B and C) may project into the pre-epiglottic space, and infrahyoid TGD cysts are often off-midline embedded in the strap muscles. Contrast administration may demonstrate thin peripheral enhancement. However, in the presence of infection, this enhancement can be significant. If there is associated thyroid carcinoma in the cyst, it can appear as an eccentric mass, which may demonstrate foci of calcifications in it.

On magnetic resonance imaging (MRI), an uncomplicated TGD cyst has low signal intensity on T1-weighted images, sometimes it can be isointense to hyperintense related to proteinaceous contents of the cyst. Most of them are homogeneously hyperintense on T2-weighted images (Fig 2D), which reflect its fluid content. The rim will be nonenhancing unless inflammation is present.[7] In case of infection or hemorrhage, a thick irregular rim may be visualized.

Nuclear scintigraphy is not routinely used. It is recommended only if the sonography fails to identify the normal thyroid tissue.

Histopathology

On histologic examination, the TGD cysts are lined by stratified squamous epithelium or ciliated pseudostratified columnar epithelium and filled with colorless, viscous secretion (Fig 3).[4] Sometimes they may demonstrate mucous glands. Ectopic thyroid tissue along the course of the duct is variably reported in up to 62% of cases.

B. Branchial Cleft Cyst

Embryology and Pathogenesis

Branchial apparatus include 6 arches (mesoderm) interfaced by 4 clefts (ectoderm) and pouches