Thyroglossal Duct Cyst: A Literature Review

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Abstract: Thyroglossal duct cyst (TDC) develops when the embryonic duct persists after the migration of the thyroid gland, making it possible for it to arise anywhere along the route of the descending thyroid. Approximately 70% of congenital neck masses in children are associated with this cyst. TDC commonly present with an asymptomatic midline neck mass that is painless, movable, soft in consistency and fluctuating, and painful masses can ensue due to infection. Most TDC case are benign with only about 1% case is related to malignancy. Comprehensive history taking and a physical examination are essential for preoperative diagnosis while imaging modalities are useful not only to rule out other differential diagnosis of neck lesion resembling TDC but also to identify a normal thyroid gland in children. Definitive management of TDC is surgery using the relatively safe Sistrunk procedure. This procedure can reduce the recurrence rate of thyroglossal duct cysts.

Keywords: Thyroglossal Duct Cyst, Congenital Neck Mass, Sistrunk Procedure

1. Introduction

Thyroglossal duct cyst develops when the embryonic duct persists after the migration of the thyroid gland, making it possible for TDC to arise along the route of the descending thyroid. TDC is one of the most frequent pathologic conditions encountered in the neck present as midline neck cysts closely associated with the hyoid bone [1], [2]. This congenital abnormality occurs in 7% of the population [3]. Of all congenital neck masses, TDC accounts for 70% and is among the most common cervical abnormality found in children. One study took pediatric patients as samples evaluating the associated mass in the midline of the neck and histopathological results found that 55.9% were TDC and other common findings were dermoid cysts (23.7%), unexplained inflammation and lymph nodes (6.7%), second branchial anomalies (5.1%), and hemangioma (1.7%) [4], [5].

Although TDC is more noticeable in children, it can also be found at any decade of life. These anomalies are observed equally in both sexes, but a slight male predominance had also been reported by several case studies. The highest age predilection is 0 to 20 years (52%), and age up to 5 years is 38%. One of the largest patient reviews of TDC showed that nearly one-third of these lesions occurred in children under 10 years of age. However, nearly one-third of cases were also found in patients in the third decade of life or older [4]. Other researchers reported a predilection for age less than 10 years at 31.5%, in the second decade 20.4%, in the third decade 13.5%, and age over 30 years at 34.6% [6]. One of the studies also reported that the mean age of patients with thyroglossal duct cysts was 13 years. As many as 20% of cases appeared over the age of 16 years. [7]

TDC is mostly found in the midline or near the midline (within 2 cm) of the hyoid bone area. Where the majority of the cysts are found in the infrahyoid, followed by other locations at the level of the hyoid bone and the suprathyroid area. TDC may also be found in less common areas such as the submental, suprasternal, and intralingual [8].

Patients with thyroglossal duct cysts commonly present with an asymptomatic midline neck mass that is painless, movable, soft in consistency and fluctuating, although painful masses can ensue due to infection. Several cases of dyspnea and dysphagia have also been reported [2], [9].

The management of the thyroglossal duct cyst is a surgical procedure that can be done by simple excision or the recommended Sistrunk procedure [10]. There are several complications that can arise if the thyroglossal duct cyst is not treated properly. Recurrent inflammation and infection are the most common complications that accompany the course of this cyst. Infection of the cyst is usually preceded by an upper respiratory tract infection. Another complication is thyroid ectopia, a condition in which the cyst contains ectopic thyroid tissue. This condition is rare, its prevalence is only about 5%. Approximately 1% of all cases of thyroglossal duct cysts are associated with malignancy [4].

Comprehensive history taking and a physical examination are essential; so knowledge of pathogenesis and clinical manifestations is absolutely necessary to differentiate TDC from other neck lesions in order to establish a preoperative diagnosis and appropriate surgical management.

2. Pathogenesis

The thyroglossal duct is a duct lined by transient epithelium serves as a pathway for the descent of the thyroid primordium that happens in the 4th and 7th weeks of gestation. The thyroid primordium arises at the level of the foramen cecum, that is located near the base of the tongue. This primitive thyroid descends, passes through the mesoderm of the tongue and the base area of the oral muscles, and passes anteriorly to the hyoid bone and the laryngeal cartilage. Where at the same period the hyoid bone originating from the second branchial arch is also formed, so that the thyroglossal duct is very closely related to the central part of the hyoid bone [11]. By the 7th week of gestation, after going through the anterior portion of the thyrohyoid membrane, sternothyroid and sternohyoid muscles, thyroid gland reaches its final position on the inferior part of the neck at. As the gland moves caudally, the thyroid primordium is connected to the tongue by a tubular structure known as the thyroglossal duct. This duct normally disappears between the 8th and 10th weeks of gestation and...
the inferior end of this duct becomes the pyramidal lobe of the thyroid gland [12].

There are two theories regarding the etiopathogenesis of thyroglossal duct cysts. Classical theory assumes the formation of a thyroglossal duct cyst is due to failure of obliteration of the thyroglossal duct. If there is any remnant part of the embryological duct, secretions from the epithelium within the duct (due to infection and repeated local inflammation) can cause a cyst-shaped lesion [12]. Otto's theory states that primordial thyroid tissue is formed near the heart which is then followed by the formation of a connecting channel between the thyroid gland and the epithelial tissue on the floor of the mouth. According to this theory, the thyroglossal duct contains two types of epitheliums, namely the epithelium of the oral floor in the superficial part and thyroid epithelium in the inferior part. The area between these two epithelia will rupture by the 6th week of gestation. If the thyroglossal duct is ruptured at a lower level, the thyroid tissue will remain at the cranial end of the thyroglossal duct. Ectopic thyroid tissue or lingual goiter may result from this mechanism. When the thyroglossal duct ruptures at a higher level, non-thyroid epithelial tissue will remain in the area between the base of the tongue and the thyroid gland, so that it can become a site for the formation of an epithelial cyst or thyroglossal duct cyst. These epithelial cysts may also contain thyroid tissue [7], [13].

3. Diagnosis

3.1 Clinical Manifestation

Patients with TDC generally present with an asymptomatic mass in the midline of the neck that is painless, 1-2 cm in size (sometimes up to 10 cm), smooth, well demarcated, and fluctuating. The mass attachment to the hyoid bone will make it move cranially during swallowing or tongue protrusion. As with other congenital cystic malformations, thyroglossal duct cysts can become infected especially after an upper respiratory tract infection. An infected cyst presents with symptoms of an enlarged cyst, erythematous, swelling and pain in the cyst and the surrounding area [2], [9]. Approximately one third of patients present with symptoms of an infected cyst, especially in adults. One fourth of the adults also show draining sinus either spontaneously or due to surgical drainage of abscess. Dyspnea, dysphagia, and dyspnea are rare clinical manifestations, but had been reported in some cases usually in larger cysts that press on the underlying tissues [13], [14].

About 1% of all cases of thyroglossal duct cysts are associated with malignancy [4]. Malignant incidence of TDC was found more frequently in women (68.3%) and in older people with a mean age of 39.5 years. Several studies have stated that the suspicion of a TDC cancer can be seen from the presence of a fixed mass with rapid growth that is accompanied by pain sensation. But it is still quite difficult to identified through clinical findings alone [15].Risk factors that play a role in the occurrence of carcinoma include a history of radiation exposure, a history of thyroid disease, age, tumor size, tumor spread, and histopathological factors [7], [10].

TDC is mostly found in the midline or near the midline (within 2 cm) of the hyoid bone area. Where up to 65% are found in the infrahyoid section, 15% to 50% located parallel to either anterior or posterior of the hyoid bone itself, and about 20% to 25% are positioned in the suprahyoid area. TDC may also be found in the submental (24%), suprasternal (13%), and intralingual (2%) [8].

3.2 Further Evaluation

History and physical examination are very important for TDC diagnosis, but since a variety of different masses can occur in the neck and may be indistinguishable, an imaging study needs to be performed. Investigations in the form of preoperative radiology in cases with suspected TDC are not only to help establish the diagnosis but also to identify a normal thyroid gland. Each radiological examination technique provides advantages and disadvantages in this regard.

Ultrasoundography (USG) is still the most commonly used approach especially in children because of low radiation exposure, non-invasive nature, and relatively low cost compared to other radiological examinations such as computer tomography (CT) and magnetic resonance imaging (MRI) [16]. Ultrasound examination is accurate enough to provide an overview of cystic lesions with a sensitivity level of 75% [7]. Thyroglossal duct cyst on cervical ultrasonography is seen as a thin-walled, well-defined anechoic cyst with posterior acoustic enhancement.In children, USG also plays an important role in differentiating ectopic thyroid tissue from TDC. Where the removal of this ectopic thyroid tissue can cause a condition of postoperative hypothyroidism [4], [16].Fine needle aspiration cytology (FNAC) can be used to establish the differential diagnosis or when a solid lesion is found. On histopathological examination, the cyst was lined by ciliated pseudostratified columnar and/or squamous epithelium. Mucus glands and thyroid follicles are usually seen in adjacent connective tissue [2]. In the case of thyroglossal duct carcinoma, atypical squamous cells or psammoma bodies may be seen on FNAC [17]. In addition to FNAC, CT and MRI examinations can also be used as an adjunct to rule out malignancy and to determine the exact position and size of the TDC if indicated [16], [18]. This includes lesions that are present in unusuallocations, in case of intralaryngeal involvement and/or recurrent TDC remnants[17].

3.3 Differential Diagnosis

There are other neck masses that can closely resemble a thyroglossal duct cyst. The findings from imaging modality of cystic necklesions are not specific thus making it difficult to make differential diagnosis. Paying a close attention to the age predilection, the location of the lesion, and also other association with surrounding structures can give some clues to determine the correct diagnosis [12], [17]. On this section the diagnosis is grouped according to the lesion location in the midline and near or off midline.

A. Midline Lesion

In addition to TDC, masses that can be found in the midline area are ranulas and dermoid cysts [17].
a) Ranula
Also called sublingual gland mucocele or mucous retention cyst, is a fluid collection or cyst that forms in the mouth under the tongue due to obstruction of the sublingual or the minor salivary glands. There are simple ranula and diving ranula which was recorded in the age range of 3 – 61 years. Simple ranula is restricted within the buccal floor to the level of mylohyoid muscle while diving ranula (pseudocyst) practically originates from successive rupture of simple ranula which then expands to posterior part of mylohyoid muscle [17].

b) Dermoid cyst
Dermoid cyst as neck cyst originates from dermal portion of the first and second branchial arches [12]. Due to this nature, dermoid cyst presents frequently in midline, typically from buccal floor and superficially within the subcutaneous tissues of the anterior neck, which is in contrast to TDC that is closely associated with the hyoid bone. Other than their predilections, dermoid cyst can also arise in other parts of the body such as the orbital and nasal area. The midline cyst might be presents with a tuft of hair. The size of dermoid cyst is smaller with rounded shape that does not move with deglutition. It is present at birth and usually found in children under the age of 5 years [2], [12], [17].

B. Off Midline Lesion
The differential diagnosis based on its off-midline location are as such:

a) Branchial cleft cyst
This is the second most frequent congenital neck masses in the pediatric population. It arises from incomplete obliteration of any branchial tract resulting in cysts, sinuses, or fistula. First branchial cleft cyst predilections are in peri-parotid or submandibular area. This branchial cleft can manifest as parotitis or recurrent abscess at the angle of the mandible or submandibular area. Approximately 80 to 95% of branchial cysts is originated from second branchial cleft. It occurs in the lateral aspect of the neck anterior to the sternocleidomastoid muscle. Cyst presents as a fluctuant neck mass with or without pain sensation. The third and fourth branchial cleft anomalies are excessively rare and usually present as sinus tract rather than a cyst or fistula. They mostly appear on the left side of the lower anterior neck and can be closely associated with thyroid gland [2]. Although the branchial cleft lesion is congenital, it can also present in later stage of life [17].

b) Lymphatic malformation
Sequestration of embryonic lymphatic channels can give a rise to lymphatic malformations. This lesion reported no sex predilection and commonly seen in children of 2 years which is the age of greatest lymphatic growth. Lymphatic malformation can be associated with chromosomal defects, such as Turner’s syndrome, Trisomy 13, 18 and 21 as well as Noonan syndrome. Patients mostly asymptomatic. The lesion commonly presents in the posterior triangle or in the submandibular space as a soft, painless and compressible mass in the neck. The lesion may become painful or enlarge rapidly from acute spontaneous hemorrhage or infection [2], [12], [17].

c) Thymic cyst
This is a rare cystic lesion derived from the persistent thyrompharyngeal duct. Most commonly present during the first decade of life, age 2 to 13 years old, with a slight predilection in males. They are most commonly found in the left neck, anterior to the sternocleidomastoid muscle, with or without extension into the superior mediastinum. Patients can be asymptomatic or can present with upper respiratory infection, hoarseness, wheezing, coughing, dysphagia, and even respiratory distress [12], [17].

4. Management
There are several complications that can arise if TDC is not treated properly. The definitive management of the thyroglossal duct cyst is a surgical procedure that can be done by simple excision or by the widely accepted Sistrunk procedure which involves excision of the entire cyst and thyroglossal duct channel to the foramen cecum at the base of the tongue. The close association between the hyoid bone and the tract requires removal of the midportion of the bone. Sistrunk procedure still remains as the gold standard for surgical management in TDC [10], [19].

Complications due to surgery are quite rare (at the rate of 8%) and most of them are minor with minimal morbidities such as local infection, seroma, hematoma, and wound dehiscence [1]. Recurrence rates may vary in the literature, and the recurrence of a TDC after complete excision with the Sistrunk procedure is reported to be 2.6 – 5% compared to a higher percentage of 37 – 70% resulting from simple excision [1], [10]. Incomplete surgical removal due to misdiagnosis or mismanagement might lead to a greater risk of recurrence [16]. Indications of excision in TDC include complaints of growing cyst size, cosmetics, previous history of infected mass, and possible malignancy [4].

Another study reported a no-scar transoral excision approach for managing TDC in consideration of the external operational scar that is inevitably left by the Sistrunk procedure. The exclusion criteria were the presence or suspicion of abscess or inflammation preoperatively, or prior head and neck malignancy, or history of radiation therapy. This procedure promotes a good cosmetic outcome but still has no comprehensive data regarding the recurrence rate and complications, considering that it has only recently been introduced [19].

In association with malignancy, some experts stated that the Sistrunk procedure alone is sufficient as curative therapy in most cases. However, looking at the high incidence of papillary carcinoma or mixed type carcinoma in the thyroid gland, a total thyroidectomy procedure should be performed in cases with TDC malignancy [20].

5. Summary
Thyroglossal neck cyst is one the most common pathologic conditions present in the midline neck. Patients with TDC generally present with an asymptomatic mass that is painless, smooth, well demarcated, and fluctuating that moves with deglutition and tongue protrusion. Meticulous
References


