A Rare Case of Lingual Thyroid with Hypothyroidism

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Abstract: Lingual thyroid gland is a rare clinical entity and occurs due to the failure of the thyroid gland to descend to its normal cervical location during embryogenesis. This ectopic thyroid gland located at the base of the tongue may present with symptoms like dysphagia, dysphonia, upper airway obstruction or haemorrhage and maybe associated with thyroid dysfunction. We are presenting here the case of a 16 year-old girl who reported to our hospital with complaints of foreign body sensation in the throat & dysphagia. The CT scan revealed an SOL base of tongue. An emergency surgery was conducted due to sudden appearance of dyspnoea & increase in dysphagia. A post operative Thyroid scan & Biopsy confirmed the diagnosis of Lingual thyroid. Thyroid functions showed Hypothyroidism. The TSH level was 98.1 µU/ml and T3 & T4 levels were 0.1 ng/ml & 2.3 µg/dl respectively. Conclusion: Ectopic Thyroid is a rare anomaly with Lingual thyroid accounting for majority of cases. Dysphagia and dysphonia are common presenting symptoms and majority of cases with thyroid dysfunction have hypothyroidism. Pathogenesis of this ectopic is unknown. Genetic factors have been associated with thyroid gland morphogenesis & differentiation but so far no mutation in known genes has been associated with human thyroid ectopy.

Keywords: lingual thyroid, ectopic thyroid, dysphagia, dysphonia

1. Introduction

The thyroid gland is one of the largest endocrine glands in the body, it lies approximately at the same level as the cricoid cartilage(1,2). Any functioning thyroid tissue found outside of the normal thyroid location is termed Ectopic thyroid tissue(3). Ectopic thyroid tissue has been found from the tongue to the diaphragm. Ninety percent of the reported cases of ectopic thyroid are found in the base of the tongue(4). Lingual thyroid is a rare established embryological anomaly and originates from failure of the thyroid gland to descend from the foramen caecum to its normal eutopic pre-laryngeal site. It generally originates from epithelial tissue of non-obliterated thyroglossal duct(5). The ectopic gland located at the base of the tongue is often asymptomatic but may cause local symptoms such as dysphagia, dysphonia with stomatolalia, upper airway obstruction and haemorrhage, often with hypothyroidism at any time from infancy through adulthood. Symptomatic lingual thyroid tissue is unusual with approximately 400 previously reported cases(6). Prevalence rates of LT vary from 1 in 100,000 to 1 in 300,000 (7). Sixty five to eighty percent of cases occur in females(8).

2. Case Report

We present the case of a 16 year-old girl who reported to the surgical outpatient clinic of our hospital with complaints of foreign body sensation in the throat & mild dysphagia of many years duration. Her past medical history was insignificant. Examination revealed a 3 cm×4 cm midline, smooth, rubbery and reddish mass at the base of the tongue, with prominent telangiectasias. (Figs.1). Neck examination revealed neither palpable thyroid gland nor any other palpable masses. The patient did not give any history of past or present thyroid disease. Possibility of lingual thyroid, Dermoid cyst, & minor salivary tumor were considered and she was advised to undergo various investigations. However before all investigations could be completed the patient reported again with severe complaints of dyspnoea, increasing dysphagia and increasing size of mass.

The CT scan revealed an SOL at base of tongue. Due to the deteriorating condition of the patient she was immediately taken up for surgery transorally after difficult endotracheal intubation. The swelling was very vascular & because of excessive bleeding, ligation of external carotid was considered at one stage. However it was not needed after achievement of haemostasis. The post-operative period was uneventful.

A thyroid scan conducted post-operatively revealed no evidence of any functioning thyroid tissue in the neck. An ultrasound of the neck confirmed that there was no evidence of lobes of thyroid in the neck. Histopathological examination of the excised mass revealed a follicular adenoma(embryonal type) of thyroid tissue. All the above investigations confirmed a final diagnosis of lingual thyroid (LT). Thyroid function tests showed that the patient had hypothyroidism with elevated TSH levels of 98.2 µIU/ml and low T3, T4 levels of 0.1 ng/ml & 2.3 µg/dl respectively. Substitutive Thyroid hormone therapy was started in order to maintain a euthyroid state. The patient was followed up till 3 months after surgery and was doing well with replacement thyroid hormone.

3. Discussion

Ectopic thyroid is a rare embryological aberration which can occur in any moment of the migration of the thyroid resulting in lingual (at tongue base), sublingual (below the tongue), prelaryngeal (in front of the larynx), and substernal.
(in the mediastinum) ectopy\(^9\). Lingual thyroid is the result of failure of descent of the thyroid anlage from the foramen cecum of the tongue.

The reasons for the failure of descent are unknown\(^{10}\). Although the pathogenesis of lingual thyroid is unclear, some authors have postulated that maternal antithyroid immunoglobulins may impair gland descent during early fetal life\(^{20}\). The molecular mechanisms involved in thyroid dysgenesis are not fully known but studies have shown that mutations in regulatory genes expressed in the developing thyroid could be responsible\(^{11}\). Genetic research have shown that the gene transcription factors TITF-1(Nkx2-1), Foxe 1(TITF-2) and PAX-8 are essential for thyroid morphogenesis and differentiation. Mutation in these genes may be involved in abnormal migration of the thyroid\(^{11,12}\). In humans more than 50% of thyroid dysgenesis cases are associated with an ectopic thyroid. However no mutation in known genes has so far been associated with the human ectopic thyroid\(^{11}\).

Ectopic thyroid is seen at any age but more commonly during childhood, adolescence and around menopause\(^{10}\). This probably occurs when demands for thyroid hormones increase, causing the increase in circulating TSH levels with growth of the ectopic thyroid tissue\(^{13,14}\). Similar response is also encountered during other metabolic stress conditions like pregnancy, infections, trauma etc\(^{11}\).

About 33% of all patients with ectopic thyroid show hypothyroidism findings\(^{10}\). This was also the case in our patient. Most ectopic thyroids are asymptomatic and no therapy is necessary. Symptoms are related to the growth of the thyroid tissue, causing dysphagia, dysphonia with stomatolalia, bleeding or dyspnœa\(^{13,16,17}\).

Clinically, lingual thyroid presents as a midline mass at the base of the tongue, pink and firm. Palpation of the neck is extremely essential, in order to check the presence or the absence of the thyroid gland in its normal position. Diagnosis depends on finding thyroid tissue at the base of the tongue with the absence of normally located gland. Imaging studies as ultrasound scan, C.T scan and Technetium (Tc99m) thyroid scan are of great value in establishing the diagnosis\(^{15}\). Thyroid scan can also reveal whether there are other sites of thyroid tissue. In approximately 75% of patients the ectopic tissue is the only functioning thyroid tissue in the body\(^{19}\). Therefore, it is important to observe the patient at follow-up, being aware of the risk of post-operative hypothyroidism.

Differential diagnosis includes lymphangioma, minor salivary gland tumours, midline branchial cysts, thyroglossal duct cysts, epidermal and sebaceous cysts, angiomata, adenoma, fibroma and lipoma\(^{19,20}\). Management of lingual thyroid is still controversial. No treatment is required when the lingual thyroid is asymptomatic and the patient is in an euthyroid state; the patient has to be followed to be aware of development of complications. Malignant transformation can occur though the incidence is only 1%\(^{21}\) and the rate of malignant transformation in ectopic thyroid tissue is no greater than in the normally placed thyroid gland\(^{22}\). Follicular carcinoma of the lingual thyroid is the commonest histopathological subtype\(^{23}\). Some Authors consider complete surgical removal of the gland as appropriate treatment\(^{24,25}\). For patients with no, or only mild, clinical symptoms and elevated TSH concentration, substitutive therapy with thyroid hormone may be successful, producing a slow reduction of the mass. Ablative radioiodine therapy is an alternative approach recommended in older patients or patients who are deemed unfit for surgery. This treatment should be avoided in children and young adults since the systemic doses required have potentially damaging effects on the gonads or other organs\(^{26}\). Moreover, the thyroid tissue is often hypoactive and the dose of radioiodine required is generally high\(^{27}\). When medical therapy fails, and in symptomatic or complicated cases, surgery is the treatment of choice. Levothyroxine therapy should be initiated after surgical excision as the lingual thyroid is the only functioning thyroid tissue found in 70% of these patients\(^{28}\).

4. Conclusions

Lingual thyroid is a rare developmental anomaly representing faulty migration of normal thyroid gland. The exact pathogenesis of this ectopic is not known. The incidence is higher in females. Dysphagia is a common presenting symptom. Thorough head and neck examination with special attention to base of tongue is essential. Investigations include thyroid function tests, neck ultrasound scan, Technetium scanning and C.T scan. The treatment is still controversial on account of the rarity of the condition. Treatment could be conservative with substitutive hormone treatment in patients with mild symptoms, while surgery is recommended in cases when complications such as dysphagia or dyspnœa occur. Evaluation of thyroid function is recommended before and after surgery due to the risk of post-operative hypothyroidism. Substitutive hormone treatment is often needed. Follow-up is recommended to monitor possible recurrence or complications and to monitor the thyroid hormone status.

More research is required to determine cause of thyroid ectopy. Detection of the gene responsible for this condition in humans will assist early or prenatal diagnosis and better management of the disease.

Reference


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Figure 1: Thyroid Tissue at Root of Tongue