An Embryological Cause of Primer Hypothyroidism, Lingual Thyroid

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Lingual thyroid is a rare developmental thyroid abnormality usually affecting females. It is often found in the region of the foramen caecum as a lingual thyroid at the base of the tongue in patients whose gland fails to descend. A 16-year-old female patient with severe hypothyroidism applied to our outpatient clinic. She was complaining from growth retardation, primary amenorrhea, weakness, decreases of school performance and short stature. No thyroid tissue was seen at the usual site with ultrasonography. Scintigraphy with 99mTc pertechnetate showed an ectopic functioning thyroid in the lingual area. CT scan showed a mass in the base of lingual region. The thyroid profile implied severe hypothyroidism (free T3:0,142 ng/dl, free T4:0,741 ng/dl, and TSH:100 µU/ml). Patient was accepted as primary hypothyroidism due to ectopic thyroid tissue (lingual thyroid) and substitution treatment with L-thyroxine was started at the dose of 2-3 µgr/kg. Surgical treatment did not consider because of patient was asymptomatic in terms of pressure. As a result we reported a rare cause of primary hypothyroidism due to ectopic thyroid gland (lingual) in a young female patient.

Keywords: Lingual thyroid, juvenile hypothyroidism, ectopic thyroid

Introduction

Ectopic lingual thyroid gland is uncommon developmental abnormality. Abnormality of the embryological development and migration of the thyroid gland can result in ectopic thyroid tissue. Ectopic thyroid tissue may be found anywhere along the embryologic "path of descent" of the thyroid gland between the foramen caecum and the anterior mediastenium. Although uncommon, it is often found in the region of the foramen caecum as a lingual thyroid at the base of the tongue in patients whose gland fails to descend. Of all ectopic thyroids, 90% are found on the lingual dorsum, where they are called lingual thyroid or ectopic lingual thyroid (1). Extra-lingual thyroid tissue is commonly located in the anterior cervical area, in the region of the thyroglossal duct. In addition ectopic thyroid tissue has been described in the submandibular region (2), trachea (3), mediastinum (4), heart (5), lung (6), duodenum (7) and adrenal gland (8).

As the majority of patients with lingual thyroid are asymptomatic, the true incidence is unknown. However obstructive symptoms and hypothyroidism may be observed (9). Ugar-Cankal et al reported that the prevalence of lingual thyroid in Turkish schoolchildren in Ankara was 0,1% (10). The clinical management of lingual thyroid remains controversial. Adjustment of the treatment depends on the presence or absence of symptoms.

We reported a case of primary hypothyroidism due to non-functional ectopic thyroid tissue in the lingual areas in a 16-year-old female patient.

Case Report

A 16 year-old girl applied for our outpatient clinic for growth retardation, primary amenorrhea,
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weakness, decreases of school performance and short stature. Clinical examination revealed dry skin, slow motor functions, and retardation of sexual development. Examination of the neck revealed no palpable thyroid tissue in the normal pretracheal position. Sexual hair and breast development were absent. Body height: 132 cm (below of -3 SD) and body weight: 30 kg, BMI: 17.2 kg/m², below of -3 SD). Bone age was 8 years old, and height age was 10 years old. Growth hormone (GH), insulin like growth factor-1 (IGF-1) and IGF binding protein-3 (IGFBP-3) were found below of normal range according to similar age children (GH:0.74 ng/ml (normal range 0.06-5); IGF-1: 1.81 ng/ml (normal range 237-996); IGFBP3: 2.66 µg/ml (normal range 3.5-10). Insulin tolerance and L-dopa tests were performed for stimulation of growth hormone. GH did not increase during both of stimulation tests. Normal thyroid tissue was not shown in ultrasonography of the thyroid (Figure 1). She had no other medical problems and no prior thyroidal surgery. Radioisotope study using 99mTC pertechnetate showed an ectopic functioning thyroid in the lingual area (figure 2). CT scan showed a large mass in the posterior third of the tongue and its evaluated lingular thyroid gland in the base of lingular region (Figure 3). No thyroid tissue was seen at the usual site in these techniques. She was not complaining from pressure symptoms such as dysphagia, dysphonia, or dyspnea.

The thyroid profile showed a free T3 value of 0.142 ng/dl (normal range: 0.182-0.462 ng/dl), free T4: 0.741 ng/dl (normal range: 0.932-1.710 ng/dl), and a thyroid-stimulating hormone (TSH): 100 µIU/ml (normal range, 0.270 to 4.2 µIU/ml) and thyroglobulin were 24.05 ng/mL (normal range: 1.4-78) respectively. Anti-thyroglobulin antibody and thyroid peroxidase antibody were negative. With these findings, the patient was accepted as primary hypothyroidism due to ectopic thyroid tissue (lingual thyroid), and substitution treatment with L-thyroxine was started at the dose of 2-3 µgr/kg. We didn’t consider surgical treatment as the patient was asymptomatic. We decided to follow-up monthly interval under the suppressive dose of L-thyroxine. When the patient become euthyroid we will perform a new growth hormone stimulation test to confirm growth hormone deficiency.

Figure 1. Thyroid ultrasonography of patient.

Figure 2. Scintigraphy with 99mTC pertechnetate showed an ectopic functioning thyroid in the lingual area.

Figure 3. CT scan showed a mass in the base of lingular region.
Discussion

Lingual thyroid is the result of a defective migration of the thyroid anlage occurring between the 3rd and 7th week of embryologic development (11,13). Whereas mutations in the transcription factor-2 (TTF-2) and PAX8 and in the TSH receptor genes have been reported in a minority of patients with thyroid dysgenesis (11), but etiopathogenesis of disease did not clearly understand majority of case with ectopic thyroid.

The lingual thyroid is four-seven times more common in females than in males (1,15). Generally, lingular thyroid can be seen like noduler mass in the base of the tongue, usually less than a centimeter in size but sometimes reaching more than 4x8 cm. in size (1). The surface of the lesion is usually smooth and vascularity (12). Larger lesions can interfere with swallowing and breathing, but most patients are unaware of the mass at the time of diagnosis (16). Patients with lingual thyroid usually have signs of dyspnea, dispagia, bleeding sensation of foreign body in the throat or may mimic a lingual tumor and upper airway obstruction (16). But our patient was not complaining any pressure symptom.

Clinically, the majority of patients with lingual thyroid is asymptomatic (9), but, in the case of a normal functioning thyroid gland with presenting as an oropharyngeal mass has been reported in the literature (14). However, up to 70% of patients with lingual thyroid have hypothyroidism and 10% suffer from cretinism. In 75% of these cases, the lingual thyroid is the only thyroid tissue in the body (17). Isotopic studies indicate that approximately 30% of individuals have functioning thyroid tissue along the thyroglossal duct; in 70% of those with ectopic contained in the tongue (11). Moreover, ectopic lingual thyroid tissue may undergo neoplastic transformation and may develop papillary thyroid carcinoma (3,4).

If the patient is asymptomatic, the use of suppressive therapy with exogenous thyroid hormone is the mainstay of the treatment. The goal of therapy is to suppress TSH and thereby remove the stimulus for gland enlargement. This suppressive therapy is mandatory for symptomatic patients and those asymptomatic patients with elevated TSH levels. In asymptomatic euthyroid patients it prevents the hypothyroid state that eventually develops in most cases and leads to hypertrophy of the gland. The surgical management of lingual thyroid depends on the severity of present symptoms. Surgical intervention is necessary for selected patients who become symptomatic or have worsening of their initial symptoms while on suppressive therapy. Surgical treatment was not considered, because our patient has not got any pressure symptoms.

Thyroid ablation with radioactive iodine can also be used as an alternative treatment to surgical excision for lingual thyroid, but no treatment should be attempted until an 131 iodine radioisotope scan has determined that there is adequate thyroid tissue in the neck. It has been found to result in unpredictable thyroid shrinkage and is reserved for patients who are deemed unfit for surgery or who refuse surgical intervention (18). As a result we described a lingual thyroid gland in a 16-year-old female patient with severe hypothyroidism.

References


