

## Case report

### **Functional lingual thyroid in an adolescent female: a case report and brief literature review**

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#### Abstract

**Background:** Lingual thyroid is a rare clinical entity that represents faulty migration of normal thyroid gland. It commonly presents as a benign mass found at the junction of the anterior two-thirds and posterior one-third of the tongue. Although usually asymptomatic, glandular hypertrophy can cause dysphonia, dysphagia, bleeding, or stridor at any time from infancy through adulthood. We report a case that we encountered, discuss the diagnosis and its management, and review the literature. An otherwise asymptomatic 14 year-old girl presented with a posterior tongue mass that had been present since childhood but was never investigated. She was clinically and biochemically euthyroid, with normal thyroid function tests. Physical examination revealed a smooth, globular mass occupying the whole tongue base and valleculae. The epiglottis was slightly displaced posteriorly but the laryngeal inlet was patent. A <sup>99m</sup>Tc-radioisotope scan showed accumulation of tracer in the tongue base and no uptake in the neck. MRI revealed a 1.8-cm diameter soft tissue mass in the posterior part of the tongue. To date she has not required suppressive therapy or surgical intervention. **Conclusions:** Treatment of lingual thyroid depends on patient symptoms. Most importantly, patients should be followed at regular intervals and educated on the possibility of developing complications.

**Key words:** Lingual thyroid, ectopic thyroid, thyroid gland.

#### Introduction

Lingual thyroid is a rare anomaly defined as presence of thyroid tissue in the midline at the base of the tongue anywhere between the circumvallate papillae and the epiglottis. The true incidence of lingual thyroid is unknown and is estimated at 1 in 100,000<sup>1</sup>. There is no cervical thyroid in 70% of patients<sup>2</sup> and the lingual thyroid represents the only functional thyroid tissue. Only 0.08% had a thyroid gland in the normal position [3]. Ectopic thyroid is reportedly more common among females, who form 61-88% of cases<sup>3,4</sup>. Mean age of presentation is 14 years. It usually presents as a smooth mass in the midline at the base of the tongue and may cause dysphagia, dysphonia, choking, cough, pain, or airway compromise<sup>5</sup>. The diagnosis of lingual thyroid is usually made clinically and radionuclide scanning is used to confirm the diagnosis. In this report, a case of lingual thyroid in 14-year-old girl is presented and the choices of treatment are discussed.

#### Case report

A 14-year-old girl presented to the ENT head and neck clinic with an oral mass in the posterior part of the tongue. It had been present since childhood but had never been investigated. She had no dysphagia, shortness of breath, dysphonia, loss of appetite, or weight loss. Clinical examination revealed an oropharyngeal mass at the posterior part of tongue, partly abutting the epiglottis. On indirect laryngoscopy and flexible nasopharyngolaryngoscopy, a hemispherical swelling was seen (Figure 1). The mass was smooth and pink, and showed no evidence of ulceration. There was no palpable thyroid gland in the normal pretracheal position. Thyroid function tests revealed normal thyroxine (T<sub>4</sub> = 17.13 mmol/l) and thyroid-stimulating hormone (TSH = 5.33 mmol/l). Radioactive thyroid scanning (<sup>99m</sup>Tc scan) revealed foci of isotope uptake in the tongue base and no uptake in the neck. Magnetic resonance imaging (MRI) revealed a 1.8-cm soft tissue nodule at the

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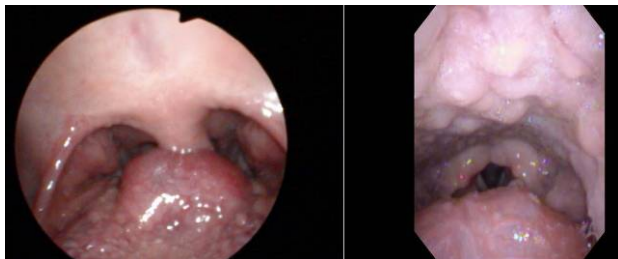
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posterior aspect of the tongue displacing the epiglottis posteriorly (Figure 2). A diagnosis of lingual thyroid was made, the patient was counseled regarding the anomaly, and was advised to return for scheduled follow-up. Repeat thyroid function tests 6 months later were normal. The swelling was unchanged with no new symptoms. To date, she has not required suppressive therapy or surgical intervention.



**Figure 1:** Lingual thyroid, transoral view



**Figure 2:** MRI T1-weighted sagittal scan shows a mass in the posterior one third of the tongue protruding into the vallecula and oropharynx

## **Discussion**

In normal embryological development, thyroid primordium develops at foramen caecum and descends through the tongue tissue, passing anterior to hyoid bone to lie in the pretracheal region. Lingual thyroid is believed to result from failure of the embryonic gland anlage to descend from its position posterior to the tuberculum impar to the normal position in the anterior neck, and can occur anywhere along this tract. The most common site is lingual with other sites being sublingual, suprahyoid, and infrahyoid.

Although lingual thyroid may present with dysphagia, dysphonia, choking<sup>5</sup>, or obstructive

sleep apnea<sup>6</sup> depending on its size, most patients are unaware of the mass at the time of diagnosis<sup>7</sup>. It has two modes of presentation based on patient's age and onset of symptoms<sup>8,9</sup>. One group of patients consists of infants and young children who suffer from failure to thrive and mental retardation, in whom lingual thyroid is detected on routine examination. The second group consists of peripubertal patients with symptoms of dysphagia and oropharyngeal obstruction. Higher TSH levels that are present in response to the increased demand for thyroid hormone during puberty stimulate the gland to hypertrophy. A similar response is seen in conditions of metabolic stress such as pregnancy, infections, trauma, and menopause<sup>8</sup>.

The ectopic thyroid is usually not adequate to maintain a euthyroid state, and 60-80% of patients will have hypothyroidism and 10% suffer from cretinism<sup>1,3,4</sup>. Therefore, thyroid function tests should be done in all patients with lingual thyroid even in the absence of symptoms. Hypothyroidism or subclinical hypothyroidism, which may result in hypertrophy of the gland, will benefit from replacement therapy using levothyroxine. Levothyroxine therapy corrects hypothyroidism, and by suppressing TSH, eventually reduces the ectopic thyroid gland size<sup>7</sup>. However, the rate of size reduction is very slow; hence, dramatic results should not be expected<sup>10</sup>.

Radionuclide scanning using <sup>99m</sup>Tc-pertechnetate (technetium scanning) should be performed to confirm the clinical diagnosis and avoid unnecessary biopsy. It typically shows radionuclide activity at the level of the mouth and no apparent activity in the neck. Most lingual thyroid glands contain histologically normal tissue and histological confirmation is not necessary unless iodine or CT scanning is negative, or malignancy is suspected<sup>11</sup>. The presence of normal functioning thyroid gland in the neck is helpful in management especially if surgical intervention is required. Total excision of lingual thyroid gland may be possible without requirement for replacement.

The role of MRI and computed tomography (CT) is similar, depending on the service available. Both CT and MRI are useful adjuncts in demonstrating airway patency, gland size, and in planning surgical intervention. There are reported cases of mixed lingual and sublingual ectopic thyroid gland. In our case, MRI revealed a single 1.8-cm soft tissue

nodule at the posterior aspect of the tongue displacing the epiglottis posteriorly.

Due to the rarity of this condition, treatment options for lingual thyroid are controversial, and there is little consensus in the literature regarding the ideal treatment. The management of lingual thyroid depends on its size, the presence and severity of local symptoms, age of the patient, status of the thyroid gland, and presence of complicating factors such as ulceration, hemorrhage, or malignancy<sup>12</sup>. When obstructive symptoms are absent and the patient is euthyroid, no treatment is required. However, the patient should be followed at regular intervals and educated on the possibility of developing complications<sup>13</sup>.

Suppressive therapy with exogenous thyroid hormone is the mainstay of medical management for the majority of patients. This therapy suppresses TSH secretion and removes the stimulus for gland enlargement. The best guide for initiating treatment is the presence of symptoms<sup>8</sup>. In patients with mild symptoms or those who are asymptomatic with elevated TSH, long-term exogenous thyroid hormone may be successful. Kalan et al. suggested suppression in asymptomatic euthyroid patients to prevent hypothyroidism, glandular hypertrophy, and diminish the risk of malignancy<sup>14</sup>. Taibah et al. also advocated lifelong treatment with L-thyroxine of those who have not yet developed hypothyroidism or obstructive symptoms<sup>15</sup>.

Surgical excision is necessary for selected patients who become symptomatic or whose symptoms do not respond to hormonal treatment. Surgery is considered crucial in severe or repeated hemorrhage and gland enlargement with dysphagia, significant airway compromise, or dysphonia<sup>7</sup>. Several approaches have been described including transoral and midline or lateral pharyngotomy through the

neck, all of which have potential for significant morbidity. The decision to use one versus the other depends on surgeon preference and the technology available. The new technologies available include CO<sub>2</sub> laser and radiofrequency ablation. Ward et al. suggested complete excision of ectopic thyroid gland to avoid recurrence of hypertrophy and airway obstruction<sup>16</sup>. However, even in the best hands, total excision of an ectopic thyroid is difficult, regardless of the surgical approach. Remnants of thyroid tissue following an attempted excision are usually located deep in the tongue substance and may be less likely to produce obstructive symptoms should they hypertrophy again<sup>14</sup>. Moreover, patients invariably require thyroid hormone replacement for hypothyroidism, which prevents possible regrowth of the gland. Hafidh et al. demonstrated the success of this approach in a series of patients, as evidenced by absence of recurrence during 3 years of follow-up<sup>17</sup>. Postoperative hypothyroidism can be avoided by auto-transplantation of the excised tissue, usually into the anterior abdominal wall or under the strap muscles<sup>2,18</sup>.

Radioactive iodine-131 thyroid ablation has been used as an alternative to surgical excision of a symptomatic gland. It has been proven successful in reducing the gland size and is reserved for patients who are deemed unfit for anesthesia or refuse surgery<sup>19</sup>.

### **Conclusions**

In our case, no treatment was required because the lingual thyroid gland was small, did not cause any symptoms, and the patient was euthyroid. Follow up is essential to detect changes in thyroid function or malignant transformation. Lingual thyroid should always be considered in the differential diagnosis of tongue base swelling.

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