HYPOTHYROIDISM CAUSED BY A NONVISIBLE LINGUAL THYROID

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Abstract: Background. Lingual thyroid is a developmental anomaly caused by failure of migration of the thyroid gland to its normal position in the neck. This anomaly is usually manifested in childhood as hypothyroidism or local symptoms.

Methods. We present a case of a 32-year-old patient in whom lingual thyroid was diagnosed after complaints of a chronic cough. Thyroid-stimulating hormone levels were significantly elevated. Sonograms of the neck revealed atrophy of both thyroid lobes, with nonhomogeneous consistency and no lymphadenopathy. Technetium-99m scan revealed uptake at the tongue base, with no uptake at the neck or other locations.

Results. L-thyroxine treatment was begun, with progressive decrement in thyroid-stimulating hormone levels and cessation of the cough.

Conclusion. This case is unique in both the advanced age of the patient and the unusual clinical presentation.

Keywords: lingual thyroid; hypothyroidism

Lingual thyroid is a rare developmental anomaly defined as the presence of thyroid tissue in the midline of the base of the tongue between the circumvallate papillae and the epiglottis. Its clinical incidence varies between 1/3000 and 1/10,000, yet the tongue is the most frequent ectopic location of the thyroid. The age of presentation ranges from birth to 74 years, although most cases are diagnosed in childhood. We describe a 32-year-old military aviator who was diagnosed with lingual thyroid because of a complaint of chronic cough. This case is unique in both the relatively late age of the patient at the time of diagnosis and the unusual clinical presentation. The fact that the patient was a military aviator involved in everyday strenuous activity is also unique, because previous reports have demonstrated a relationship between stress and local symptoms, whereas in our patient no apparent relationship was demonstrated.

CASE REPORT

A 32-year-old F-16 pilot was seen by his primary care physician with a 3-month history of nonproductive cough. His medical history was unremarkable, and he was taking no medications. He had no additional complaints, but when specifically asked, he recalled gaining 4 kg in the preceding 4 months. The physical examination was unrevealing, including the ear, nose, and
throat (ENT) examination. Routine laboratory tests revealed a mild normocytic, normochromic anemia (hemoglobin, 12.7 g/dL; mean corpuscular volume [MCV], 86 fL; mean corpuscular hemoglobin [MCH], 26.2 pg/cell) and mildly elevated cholesterol (207 mg/dL). Similar hemoglobin values were observed in the past, and his ferritin values were within normal limits. Thyroid function tests revealed significantly elevated thyroid-stimulating hormone (TSH) levels with free serum triiodothyronine (FT₃) and free serum thyroxine (FT₄) at the lower limit of normal values. Follow-up thyroid function tests revealed elevated TSH and low FT₄, with FT₃ being at the lower limit of normal values. He was restricted from continued flying duty pending the completion of work-up of his thyroid function abnormality.

His chest X-ray was found to be normal. An ultrasound examination of the neck revealed atrophy of both thyroid lobes with nonhomogenous consistency and no lymphadenopathy. A technetium-99m scan revealed uptake at the tongue base with no uptake at the neck or at other locations. A diagnosis of lingual thyroid was made, and L-thyroxine treatment was begun with a progressive decrement in TSH values and disappearance of the cough.

DISCUSSION

Lingual thyroid is the term applied to a mass of ectopic thyroid tissue located at the tongue base at the midline, usually between the circumvallate papillae and the epiglottis, in the area of the foramen cecum. It is believed to result from defective descent of the thyroid anlage from its embryonic position at the base of the tongue to its normal pretracheal location, which usually occurs on weeks 3 to 7 of embryonic development. Defects in migration may lead to locations of the thyroid gland in other midline structures such as the trachea, the esophagus, or near the hyoid bone. Hickman¹ reported the first case of lingual thyroid in 1869, and 400 cases have been described in the literature to date. This is the most common benign mass found at the base of the tongue and in other midline structures such as the trachea, the esophagus, or near the hyoid bone. Postmortem studies indicate that up to 10% of people have thyroid tissue remnants near the base of their tongue,²,³ although clinically apparent lingual thyroid is an unusual condition, with a few hundred cases reported in the literature. The estimated frequency of clinically significant lingual thyroid varies between 1/3000 and 1/10,000.⁴ In 70% of these cases, the lingual thyroid is the only functional thyroid tissue.⁵ Hypothyroidism is observed in an estimated 33% of the cases and is commonly precipitated by increased physiologic demands.⁶ This entity is much more prevalent in females, with a male-to-female ratio of 1:3 to 1:8.⁷

The pathogenesis of this condition remains unknown. It is postulated that maternal antithyroid antibodies may arrest the gland’s descent and predispose the patient to poor thyroid function later in life.⁸ The incidence of thyroid disease among family members of patients with lingual thyroid is higher than among the population at large.⁹

The symptoms of lingual thyroid are variable and depend on the patient’s age. Infants and young children are usually diagnosed on routine screening and often have failure to thrive and mental retardation. Older children, adolescents, and adults are diagnosed after the onset of obstructive symptoms, such as dysphagia, dysphonia, dyspnea, a cough worse on lying down, or local hemorrhage. Glandular hypertrophy occurs in these populations as a response to the rise in TSH levels that is generated by the increase in metabolic demand for thyroid hormone that occurs in situations such as puberty, pregnancy, trauma, infection, or menopause.¹⁰

The initial evaluation of a lingual thyroid involves a thorough head and neck examination. The mass appears light pink to bright red or blue, and its surface can be smooth or irregular. Flexible or rigid endoscopy of the upper airway is performed to determine the size of the thyroid and airway patency. Laboratory evaluation should include thyroid function tests, which frequently demonstrate normal to low levels of T₃ and T₄ and low levels of TSH and thyroglobulin. Hyperthyroidism, although rare, has been reported.¹¹ A technetium scan should be used to confirm the diagnosis. These scans frequently show radionuclide activity at the level of the mouth and no apparent activity in the normal position in the neck. CT provides an accurate method of determining the gland size, and it can be performed without contrast, because the thyroid tends to accumulate iodine.¹² MRI is considered the method of choice to study lingual thyroids in a noninvasive way because of its multiplanar capabilities and its high degree of soft tissue resolution, which result in better delineation of the lesion.¹³ Ultrasound is less dependable and is used to follow an enlarging component of
a thyroid gland without the need for additional ionizing radiation.

Malignant degeneration in lingual thyroid seems to be the same as for orthotopic thyroid tissue. Lingual thyroid carcinoma has been described in patients from the age of 12 to the age of 86, with the mean age of presentation in the third decade. No risk factors have been described in the literature, but it is assumed that they are the same as those for malignancy of a cervical thyroid (x-ray exposure of the head, neck, or mediastinum in infancy or childhood). There are no clinical findings that can differentiate lingual thyroid carcinoma from a symptomatic lingual thyroid. Therefore, it is recommended that biopsy or aspiration be taken in every case of lingual thyroid to rule out thyroid carcinoma. The natural history of lingual thyroid carcinoma is unknown because of the rarity of this condition, yet most cases described in the literature did not extend beyond the tongue.

Treatment of lingual thyroid is controversial. All patients should receive lifelong thyroxine suppression, even those with small and asymptomatic lingual thyroids, because this will prevent the gland's enlargement, diminish the risk of malignancy, and prevent hypothyroidism. The ectopic thyroid tissue can be ablated by a therapeutic dose of $^{131}$I. This modality is safe and effective and should be included in the algorithm when formulating a treatment plan for the symptomatic patient. Surgery is reserved for those whose initial symptoms worsen while on suppressive therapy. Surgery becomes crucial in cases of recurrent hemorrhage, severe dysphonia or dysphagia, or significant airway compromise. CT and MRI are helpful in determining the best approach for total gland excision. Complete excision of the gland is difficult, and in most cases, there are remnants left at the base of the tongue, which have the potential to enlarge in the future unless suppressed with thyroid replacement therapy.

Our case is unique in several aspects. The relatively late age of presentation of our patient is surprising, because most cases of lingual thyroid are diagnosed at times of increased metabolic demands. Our patient was a military aviator, who was certainly exposed to increased metabolic demands, whether while being a student aviator or while performing certain military missions. Previous reports linked stress and development of symptoms in patients with lingual thyroid, and there are descriptions of symptomatic cases linked to enrollment and training in the army. Our patient completed a difficult army training program with no apparent symptoms. No apparent cause of increased metabolic demands was identified before his clinical presentation, and we do not have a good explanation for the timing of his symptoms. The patient did gain 4 kg in weight in the months before the diagnosis. This weight gain may have increased his metabolic demands and caused his lingual thyroid to become symptomatic, but they may also have been a result of his hypothyroidism.

The presenting symptom of prolonged cough in our patient is also unique. Most cases described in the literature of adults with symptomatic lingual thyroid manifested as dysphagia, dysphonia, a foreign body sensation in the throat, bleeding, or intermittent respiratory obstruction. A cough, which worsens on lying down, has been described but is rare. In our patient, no other cause for the cough was identified, and the cough improved on start of treatment; therefore, we assume that the cough was related to the lingual thyroid, although the lingual thyroid was not visible on the ENT examination. The laboratory evidence of hypothyroidism and the technetium scan eventually led to the diagnosis, despite a normal ENT examination. We recommend that thyroid function tests be included in the laboratory evaluation of patients with complaints of the upper airway, including prolonged cough. Those with impaired thyroid function tests should undergo a thorough evaluation for a lingual thyroid by an otolaryngologist. Even in cases of a normal ENT examination, a technetium scan should be performed, because clinically significant lingual thyroid may be difficult to diagnose solely on clinical grounds. Should a lingual thyroid be found on a radionuclide scan, we believe that treatment with levothyroxine should be started to suppress the thyroid gland. In most cases, this treatment will cause decrement of the gland and resolution of symptoms. If symptoms fail to resolve or the gland fails to decrease in size, a biopsy should be taken to rule out a malignancy.

**CONCLUSIONS**

Lingual thyroid is a rare entity that has the potential to present at any age. The clinical presentation in adults is variable, but most patients have symptoms that result from local irritation. The combination of such complaints and disturbances in thyroid function tests should prompt the physician to search for a lingual thyroid by
use of a thorough clinical examination and a radionuclide scan.

REFERENCES