CASE REPORT

Hashimoto's thyroiditis: an accidental discovery of a lingual thyroid in a 7-year-old child

Michela Mariani, Alessandra Palpacelli, Annalisa Mussoni, Aurora Natalia Rossodivita

Department of Paediatrics,
Catholic University, Rome, Italy

Correspondence to
Dr Aurora Natalia Rossodivita,
arossodivita@libero.it

SUMMARY

An ectopic thyroid located at the base of the tongue is a rare entity, resulting from early developmental defects of thyroid gland embryogenesis during its descent from the foramen caecum to its normal eutopic pretracheal site. This condition is the main cause of congenital primary hypothyroidism, even though signs and symptoms of thyroid dysfunction may also appear later in childhood. Lingual thyroid may sometimes present with symptoms of respiratory obstruction or feeding difficulties. We illustrate the case of a 7-year-old girl with subclinical hypothyroidism due to Hashimoto's thyroiditis arising in a lingual thyroid. She had never suffered from upper airway obstructive symptoms, but did refer a 3-month history of cough. Rapid normalisation of thyroid-stimulating hormone levels and termination of the cough were attained when the L-thyroxine treatment started. After 6 months a significant reduction of lingual thyroid size was also noted. The diagnostic procedures and therapeutic options in childhood are discussed.

BACKGROUND

Hashimoto's thyroiditis occurring in ectopic tissue is a very rare event in childhood. Moreover, it is hard to diagnose when the migration abnormality is unknown, as in our patient.

This case emphasises that ectopic tissue may retain its normal function until a pathological process develops.

Ectopic lingual thyroid should be taken into account during the differential diagnosis of all lingual and pharyngeal masses, to avoid its accidental excision.

CASE PRESENTATION

A 7-year-old girl came to our attention due to the presence of a pink spherical mass at the base of her tongue. She reported a 3-month history of sore throat and cough. Her otorhinolaryngologist suspected the mass to be an ectopic lingual thyroid.

The patient had a family history of thyroid diseases, but no cases of congenital hypothyroidism had been recorded. The patient's physical and psychomotor development was completely normal.

A thorough head, neck, chest, cardiovascular, respiratory and lymph node examination did not show anything abnormal. Her thyroid gland was not palpable in its normal pretracheal position.

INVESTIGATIONS

The neck ultrasound revealed an atrophic orthotopic gland.

A Tc-99m pertechnetate scan yielded marked isotope accumulation at the base of her tongue and no uptake at the usual thyroid location (figure 1).

Elevated thyroid-stimulating hormone levels (TSH=36.38 μ IU/mL, range 0.27–4.2) with normal free serum tri-iodothyronine=5.64 pmol/L, range 3.10–6.80) and free serum thyroxine=13 pmol/L, range 12–22) values were found. In addition, high titres of antithyroglobulin=1508 IU/mL) and TPOAb=390.8 IU/mL) were detected. Biochemical and lipid parameters were in the normal range. Her bone age was appropriate for her chronological age.

All these data pointed to a subclinical hypothyroidism due to Hashimoto's thyroiditis in the lingual thyroid.

TREATMENT

The levo-thyroxine (L-T4) therapy was started at a dose of 1 μ g/kg/day and gradually titrated up to 2.5 μ g/kg/day until TSH normalisation was achieved.

OUTCOME AND FOLLOW-UP

Cough and sore throat disappeared after a few weeks of treatment.

Six months later the lingual thyroid size had significantly decreased. The patient never reported therapy side effects during the follow-up period (5 years). Her growth (73rd centile for height and 50th centile for weight) and bone age proceeded steadily. Her pubertal development started at the age of 9 years and 8 months and has not yet completed.

Presently the 12-year-old girl is in good health. She is still being treated with L-T4 at a dosage of 100 μ g/day.

DISCUSSION

Thyroid dysgenesis is the most common cause (85% of cases) of permanent primary congenital hypothyroidism. Ectopic thyroid tissue accounts for approximately two-thirds of thyroid dysgenesis, followed by gland aplasia or hypoplasia.¹

Thyroid development begins during the third week of intrauterine life as a median endodermal thickening in the floor of the primitive hypopharynx. Gland descent usually occurs between the third and seventh weeks of embryological development.²

Ectopic tissue may occur in the following four locations based on its natural migration from the base of the tongue to its final position anterior to the trachea: lingual, sublingual, prelaryngeal and substernal (in the mediastinum). Lingual thyroid, located at the base of the tongue between the circumvallate papillae and the epiglottis, is the most common site for ectopic thyroid tissue (90%).

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Figure 1 Scintigraphy with Tc-99m: radionuclide uptake focus at the base of the tongue. No thyroid tissue is seen in normal thyroid bed.

Sublingual types are less frequently encountered. Very few cases of ectopy in more distant sites (heart, hepatic portal, ovaries, adrenal glands, etc.) are reported in medical literature.³

The pathogenesis of this condition remains unknown. It has been suggested that maternal antithyroid antibodies may disturb the gland's descent and predispose the child to the development of dysthyroidism even at an older age.⁴ Generally the ectopic thyroid is sporadic, but recent studies have highlighted the possibility of a genetic component in this disorder. Familiarity has been demonstrated in 2% of cases but the specific genes responsible have not yet been identified.

The first report on lingual thyroid was published in 1869 by Hickman⁵, describing the case of a 16-hour-old girl infant who died of suffocation due to an obstructive mass in the oropharynx.

The true incidence is unknown, probably ranging from 1:3000 to 1:10 000, with a male-to-female ratio of 1:3–1:8.⁶

In most cases, patients are asymptomatic and the finding of a lingual thyroid may be incidental. Clinical presentation depends on the patient's age, the size and the location of the mass, and on the presence of thyroid dysfunction. The most common symptoms are related to the compression caused by the mass and include dysphagia, dysphonia, foreign body sensation, stomatolalia, cough, sleep apnoea and bleeding. Stridor is more common in newborns.⁷

The ectopic thyroid may become symptomatic at any age but is more common during childhood, puberty, pregnancy and menopause, when the higher metabolic demands cause an increase in TSH levels with consequent glandular hypertrophy.

In 70% of cases lingual thyroid is the only functional tissue and an overt hypothyroidism is observed in 33% of these patients.^{4–7} Hyperthyroidism related to lingual thyroid is on the other hand a rare event, and only a few cases have been reported in medical literature, mostly in adults.⁸

Primary malignant transformation may also occur in ectopic thyroid tissue, regardless of location. The frequency of carcinoma in lingual thyroid is estimated to be approximately 1 in 100 cases with a female-to-male ratio ranging from 3:1 to 8:1. The majority of these tumours are of the follicular type, while papillary carcinomas are found in 23% of cases. This contrasts with normal thyroid gland neoplasms, whose predominant types are papillary.^{4–9–10}

The initial evaluation of lingual thyroid includes a careful and thorough head and neck examination. Radiological imaging studies (colour Doppler ultrasound, CT, MRI) are also useful in determining the location and extension of ectopic tissue. Scintigraphy using I-123 (whenever available) or Tc-99m is the most important diagnostic tool used to confirm the site of

ectopic thyroid tissue and the presence or absence of the thyroid in its normal position.³

Tissue biopsy for histology or fine needle aspiration cytology (FNAC) is especially important when malignancy is suspected. FNAC is one of the most accurate diagnostic methods for the detection and differential diagnosis of neck masses, and gives a correct diagnosis in 95–97% of cases.⁴

Owing to the rarity of ectopic thyroid, there is still no consensus about the optimal therapeutic strategy. The choice of treatment is influenced by several factors: the size of the residual gland, the nature of the symptoms, the patient's age, the thyroid function and the presence of complications (ulceration, haemorrhage, cystic degeneration or cancer).¹¹ No treatment is required when the mass is small and asymptomatic, thus needing only a careful follow-up. In case of obstructive symptoms or in the presence of subclinical hypothyroidism, suppressive thyroxine therapy is the best option to reduce gland size. Some authors, however, recommend this therapy also in euthyroid and asymptomatic patients, especially in childhood to prevent hypothyroidism and the progressive growth of the ectopic mass, as well as to reduce the risk of malignancy.¹²

Surgery is indicated in case of severe obstructive symptoms, ineffectiveness of medical therapy and in presence of complications. Several surgical techniques have been described, such as the transoral, trans-hyoid, supra-hyoid or lateral pharyngotomy. The first technique is preferred in case of small lesions because it reduces the risk of damaging other close anatomical structures, such as the lingual nerve. The latter approach is preferred in case of large lesions to ensure better control of bleeding.⁴

The radio ablation with I-131 is a viable alternative to surgery when anatomical causes or clinical status contraindicate it. Higher doses of radioiodine than those used to ablate orthotopic thyroid tissue may, however, be required for the reduction of ectopic tissue. For this reason, this approach should be avoided in children and adolescents to prevent the potentially deleterious effects on surrounding tissues and on other organs.^{13–14}

In both treatments, hormone replacement therapy is later required. Regardless of the treatment choice, a careful clinical and laboratory follow-up is recommended to monitor the possible occurrence of complications.

When analysing our case some peculiar aspects can be found. The patient ought to have been euthyroid for a long time as her ectopic tissue could have retained its normal function and allowed her a normal growth and psychomotor development. It is in fact highly probable that the hyperthyreotropinaemia was not present at birth, thus escaping the neonatal screening tests.

Some studies report that the function of ectopic thyroid is long preserved in childhood.^{15–17} McGirr¹⁸ and Hutchison presume that the amount of residual thyroid tissue may mostly influence the age of thyroid insufficiency onset. Stoppa-Vaucher S. *et al*¹⁹ also suggest that the hypothyroidism in patients with ectopic gland is due to a smaller amount of tissue, with a limitation to TSH-induced growth that is possibly a consequence of the embryogenesis defect.

Hashimoto's thyroiditis arising in ectopic tissue is a rare event in childhood. It is hard to diagnose when the migration abnormality is unknown.

The persistent cough and sore throat, and the presenting respiratory obstructive symptoms in our patient, were probably related to the TSH-induced growth of the lingual thyroid as a result of the autoimmune process.

As far as we know, only two similar cases have been reported in children,^{20–21} both characterised by the appearance of a middle subhyoid neck swelling, mimicking a thyroglossal duct cyst. In these cases surgical excision was necessary due to the persistent compressive symptoms, and histopathology unexpectedly revealed chronic lymphocytic thyroiditis. In our patient the choice of a medical treatment has been successful, in relieving her symptoms as well as in avoiding an invasive approach.

In conclusion, lingual thyroid is a rare clinical entity that can occur at any age. It should always be taken into consideration in the differential diagnosis of all lingual and pharyngeal masses, even if they are asymptomatic. Lingual thyroid must be searched for particularly in the presence of oropharyngeal chronic irritative symptoms such as the persistent obstruction of the upper airways, unexplained cough, dysphagia and/or dysphonia. A careful and thorough neck and oral cavity examination is the first step in the diagnostic work-up of this condition. In addition, imaging tools and thyroid function tests can lead the clinician to the diagnosis of ectopic thyroid. Finally, a cautious approach is recommended before removing the only functional tissue.

Learning points

- ▶ Ectopic lingual thyroid is the most common cause of congenital hypothyroidism.
- ▶ Despite its rarity, the presence of ectopic thyroid should be considered in the differential diagnosis of all tongue, neck and trachea masses.
- ▶ The ectopic tissue may be functional and affected by the same pathological processes as the orthotopic thyroid gland.
- ▶ Scintigraphy with I-123 or Tc-99m is the gold standard for the detection of ectopic thyroid tissue. However, colour Doppler ultrasound is the most accessible and cheapest tool for a frontline assessment of neck ectopic glands.
- ▶ The therapeutic approach is still controversial and depends on the clinical presentation and the patient's age. A multidisciplinary team should therefore be ensured in the management of such cases.

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