Case study

A Rare case report of ectopic lingual thyroid with subclinical hypothyroidism

ABSTRACT

Ectopic thyroid gland located in the midline of the tongue due to the embryonic failure to descend to the anterior neck is an uncommon clinical entity, known as lingual thyroid (LT).
Patients with LT tissue can experience a range of clinical manifestations, depending upon the extent of the obstruction due to an enlarged thyroid or symptoms related to thyroid dysfunction.

Herein, we report a 38-year-old female who presented with dysphonia associated with foreign body sensation in the throat for three months.

Based on clinical examination, assessment of thyroid function and radiological findings, the patient was diagnosed as LT with subclinical hypothyroidism

Conservative approach with thyroid hormone suppression resulted in size reduction of ectopic thyroid gland and a complete resolution of dysphonia.

Keywords: Ectopic; nodule; lingual thyroid; conservative

1. INTRODUCTION

The thyroid gland is typically located in the anterior neck and spans the C5-T1 vertebrae [1].

Ectopic thyroid tissue is caused by failure in descent of thyroid gland from the foramen cecum to its normal location during embryogenesis and when is situated at the tongue base, it is known as lingual thyroid (LT).

The estimated incidence of ectopic thyroid is one case per 100,000 people [2,3],which is even more frequent among individuals with thyroid disorders ,with a prevalence rate ranging from 1 /4000 to 1/8000 cases [4].

The lingual thyroid (LT) is uncommonly observed in clinical practice and it affects 1/100000 to 1/300000 people [5].

Nevertheless, it represents the most common site of all ectopic thyroids (> 90 %) [3].

Lingual thyroid is more prevalent in female (70-80%) [2,5].

Patients with lingual thyroid are commonly asymptomatic [6), until the ectopic gland begins to increase in size leading to dysphonia ,dysphagia ,dyspnea or bleeding and may be associated with thyroid disorders [3].

Meticulous physical examination, hormonal profile and radiological tools are imperative to success the management of lingual thyroid [7,8].

In the present case, we report a rare case of lingual thyroid (LT) associated with subclinical hypothyroidism.

2. CASE REPORT

A 38-year-old non-smoker female presented to our department with a history of dysphonia for several months, with a foreign body sensation, especially during swallowing. She additionally reported a long-standing infertility for five years. There was no family history of thyroid disorders or autoimmune diseases.

On admission, her vital signs were within normal range.

Throat examination revealed a well-defined nodular lesion in the midline of the tongue base measuring 2 cm without sings of hemorrhage (Fig. 1).

Fig. 1. Throat examination revealing a 2-cm nodule at the base of the tongue

Neck examination demonstrated absence of thyroid gland on palpation.

Neck ultrasonography (USG) did not detect thyroid tissue in its usual site. There were no enlarged cervical lymph nodes (Fig. 2).

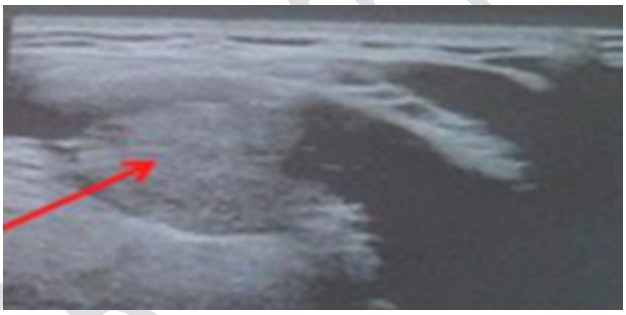

Fig. 2. Ultrasonography (USG) of the neck showing absence of thyroid tissue

Contrast-enhanced computed tomography (CT) scan showed a well-defined hyperdense homogenous enhancing mass measuring 19 x 15 x 19 mm extending from the base of the patient's tongue to her epiglottis (Fig. 3). There was no thyroid gland seen in the orthotopic site.

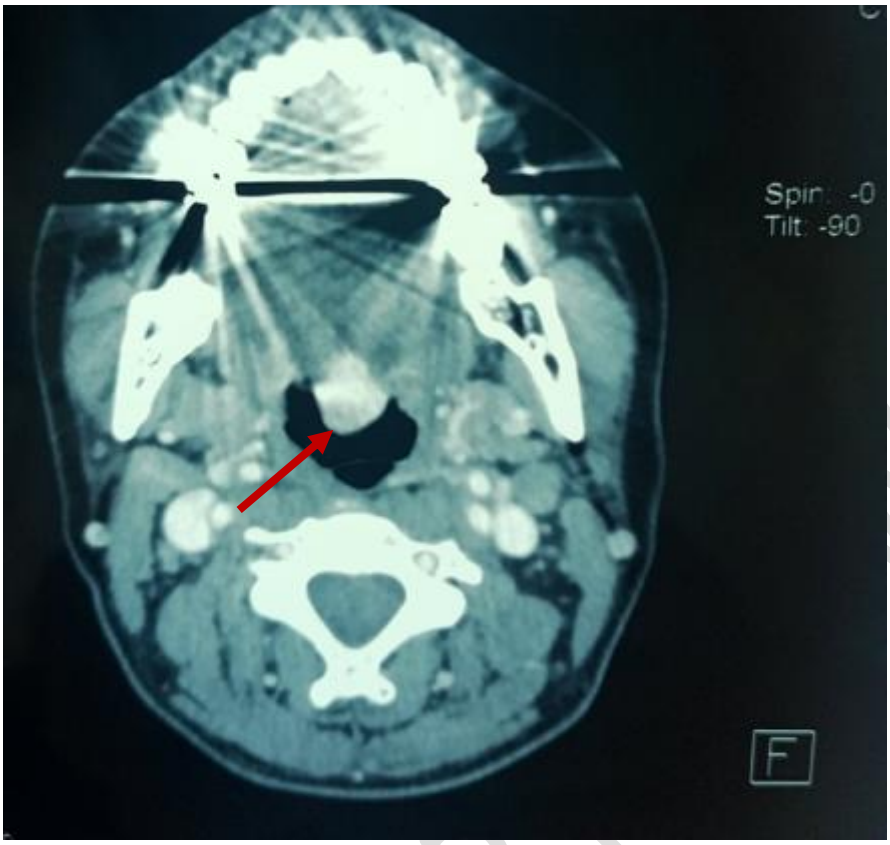

Fig. 3. Contrast-enhanced computed tomography (CT) scan of neck showing a well-defined homogenous enhancing nodule in the midline of the base of the tongue measuring 19 x 15 x 19 mm

Thyroid scintigraphy with technetium (99mTc) pertechnetate confirmed an increased uptake on the superior lingual region and the absence of thyroid tissue in the usual location.

Thyroid function tests showed normal FT4 :13,8 pmol/l (reference range :9-22 pmol/l) and moderately elevated TSH :5,8 mUI/l (reference range :0,27-4,2 mUI/l).

Screening for thyroid peroxidase (TPO) antibodies and thyroglobulin-antibodies was negative.

Based on the physical examination, thyroid function evaluation and imaging findings, the diagnosis of ectopic lingual thyroid with subclinical hypothyroidism was made.

Conservative management with hormone suppression was considered and the patient was started on 50 µg of levothyroxine, given daily.

Hormone suppression therapy resulted in a complete clinical resolution by 3 months with a remarkable decrease of the lingual gland. TSH was 1,3 mUI/l.

The patient is now on a regular follow-up since four years and continues on levothyroxine 75 µg daily.

3. DISCUSSION

Embryologically, the thyroid gland is the first of the body's endocrine glands to develop during prenatal development, on approximately the 24th day of gestation [1].

It descends to end up at the pretracheal space inferior to the thyroid cartilage [8].

The failure of this embryonic migration to the orthotopic position leads to an ectopic thyroid gland [4].

Although it represents a rare clinical entity, lingual thyroid is the most common ectopic location of the thyroid gland followed by sublingual and prelaryngeal thyroid. Ectopic thyroid tissue may be also located in other rare sites such as the cardiac muscle, esophagus, duodenum, mesentery of the small intestine, lung, breast ,ovary ,uterus , pancreas and adrenal gland [2].

Hickmann reported the first case of lingual thyroid in 1869 [7].

The estimated incidence of ectopic thyroid is one case per 100,000 people [2,3].

Females account 60-80 % of all cases .The average age of presentation is nearly 40 years old [3].

Although the pathogenesis of this embryological anomaly is unknown, genetic studies have demonstrated that mutations of genes encoding the transcription factors Foxe1 (TITF-2), TITF-1 (Nkx2-1) and PAX-8 may be implicated in aberrant migration of the thyroid gland [9,10].

Furthermore, some authors reported that maternal antithyroid immunoglobulins could be involved in failure of the gland to descend to its anatomical position, during early embryogenesis [5,6].

Most patients with lingual thyroid are asymptomatic, as some cases are discovered fortuitously. Symptoms are often related to mass effect of the ectopic gland as well as dysthyrodism. Most of clinical manifestations are caused by oropharyngeal obstruction, and may include dysphonia, dysphagia, dyspnea, foreign body sensation in the throat or bleeding [11,12].

In the reported case, dysphonia was the chief complaint that stimulated the search for diagnosis.

Palpation of the neck is a crucial part of physical examination to assess the thyroid gland in its normal location [11].

On oral cavity examination, lingual thyroid (LT) typically presents as a midline nodule in the base of the tongue [12].

Additionnaly,LT is more frequent among patients with thyroid disorders [11].

Ectopic thyroid gland may become goitrous and hypothyroidism may occur in 33 % of cases.

Then, LT has rarely malignant potential with an incidence rate of 1 in 300 cases [13].

Most common tumors are papillary carcinomas. Nevertheless, medullary, follicular and papillary Hurthle cell carcinomas have also been reported [14].

Once the LT is suspected clinically, imaging allows the physician to confirm the diagnosis, assess the extent, and identify complications.

Ultrasonography (USG) is a noninvasive routine tool demonstrating the absence of the orthotopic located gland in most cases and revealing the ectopic thyroid tissue [5].

Color Doppler may help to determine the vascularity of the nodule.

Computed tomography (CT) and magnetic resonance imaging (MRI) are extremely valuable tools to identify the location of the ectopic thyroid gland, detect calcifications and recognize its relation to the surrounding structures. On CT, ectopic thyroid tissue presents the same characteristics of orthotopic thyroid gland, a midline well-circumscribed homogenous nodule with a mildly increased attenuation (70 HU ± 10) compared to adjacent muscles, as a result of its iodine content [11,15].

Radioisotopic exams (99m Tc: technetium-99m pertechnetate and/or $^{131-}$Iodine) may be performed to localize ectopic thyroid tissue and detect an orthotopic thyroid gland [8].

Fine-needle aspiration cytology (FNAC) is helpful to attest the ectopic thyroid tissue and to exclude malignancy [3].

Thyroid function tests should further be carried out to screen for thyroid disorders.

Management of the ectopic LT is mainly dependent on the severity of symptoms, thyroid function and complications.

For asymptomatic euthyroid patients, observation is recommended and a regular follow-up is essential [2]. Patients who develop subclinical or overt hypothyroidism with symptoms related to mass effect on regional structures are treated conservatively with suppressive therapy with levothyroxine in order to reduce the size of the ectopic gland ,such as the one described in the present report [16].

Surgical management is justified in cases of lingual thyroid uncontrollable with thyroid hormone supplement and patients with serious airway obstruction, ulceration, hemorrhage and malignancy [17]. Generally, a transcervical or transmandibular approach is performed [18,19].

Then, surgical excision of the only functioning thyroid tissue requires lifelong thyroid hormone replacement.

4. CONCLUSION

Lingual thyroid (LT) is a rare condition in clinical practice. Its approach should be multidisciplinary, taking into account the patient's hormonal status as well as the clinical condition of the lesion. Conservative strategy represents a legitimate approach for many cases such as mild symptoms and subclinical hypothyroidism.

CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Rosen RD, Bordoni B. Embryology, Parathyroid. 2021; Disponible sur: http://europepmc.org/abstract/MED/32119467

2. Santangelo G, Pellino G, De Falco N, Colella G, D'Amato S, Maglione MG, et al. Prevalence, diagnosis and management of ectopic thyroid glands. Int J Surg Lond Engl. avr 2016;28 Suppl 1:S1-6.

3. Cruz-Dardíz N, Rivera-Santana N, Torres-Torres M, Cintrón-Colón H, Lajud S, Solá-Sánchez E, et al. Lingual thyroid gland: it's time for awareness. Endocrinol Diabetes Metab Case Rep. 29 mai 2020;2020:EDM200026.

4. Ibrahim NA, Fadeyibi IO. Ectopic thyroid: etiology, pathology and management. Horm Athens Greece. déc 2011;10(4):261-9.

5. Koc G, Taskaldiran I, Aslan Felek S, Saltabas MA, Omma T, Akbulut A, et al. ECTOPIC LINGUAL THYROID PRESENTING WITH MASSIVE HEMATEMESIS. Acta Endocrinol Buchar Rom 2005. juin 2019;15(2):244-6.

6. Rao K. A Case of Lingual Thyroid. 2020;1(1):3.

7. Kaushal D, Goyal A, Shakrawal N. Ectopic lingual thyroid: an entity not to be missed! 22 oct 2019;4.

8. Kumar SS, Kumar DMS, Thirunavukuarasu R. Lingual thyroid-conservative management or surgery? A case report. Indian J Surg. juin 2013;75(Suppl 1):118-9.

9. Gillam MP, Kopp P. Genetic regulation of thyroid development. Curr Opin Pediatr. août 2001;13(4):358-63.

10. Cortés JMR, Zerón HM. Genetics of Thyroid Disorders. Folia Med (Plovdiv). 1 juin 2019;61(2):172-9.

11. Ramanathan R, Veerapandian J, S. S. Lingual thyroid with hypothyroidism in a child. Int J Contemp Pediatr. 27 juin 2019;6:1747.

12. Castro PH de S, Volpato LER, Tramujas J, Borges AH. Ectopic Thyroid at the Base of the Tongue of a Young Patient. Case Rep Dent. 2016;2016:9174970.

13. Stokes W, Interval E, Patel R. Lingual Thyroid Carcinoma: A Case Report and Review of Surgical Approaches in the Literature. Ann Otol Rhinol Laryngol. 1 juill 2018;127(7):475-80.

14. Lukáš J, Drábek J, Lukáš D, Zemanová I, Rulseh A. Ectopic thyroid with benign and malignant findings: A case series. Int J Surg Case Rep. 1 janv 2020;66:33-8.

15. Abdel Aal M, Scheer F, Andresen R. Ectopic Mediastinal Thyroid Tissue With a Normally Located Thyroid Gland. Iran J Radiol. 1 janv 2015;12(1):e7054.

16. Sigua-Rodriguez EA, Rangel Goulart D, Asprino L, de Moraes Manzano AC. Conservative Management for Lingual Thyroid Ectopic. Case Rep Otolaryngol. 15 févr 2015;2015:e265207.

17. Adelchi C, Mara P, Melissa L, De Stefano A, Cesare M. Ectopic thyroid tissue in the head and neck: a case series. BMC Res Notes. 6 nov 2014;7(1):790.

18. D'Andréa G, Vairel B, Vandersteen C, Chabrillac E, Vergez S, Bonnecaze GD. Is Transoral Robotic Surgery the Best Surgical Treatment for Lingual Thyroid? A Case-Report and Literature Review. Ann Otol Rhinol Laryngol. 1 janv 2022;131(1):39-51.

19. Thapa S, Khanal P. Lingual Thyroid with Subclinical Hypothyroidism in a Young Female. Case Rep Endocrinol. 30 janv 2021;2021:e6693477.