Case Report

Diagnostic evaluation of a case of lingual thyroid ectopia

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ABSTRACT

Thyroid ectopia can occur when the process of thyroid embryogenesis fails. Here, we present the case of a 30-year-old woman with thyroid ectopia that was discovered during magnetic resonance imaging of cervical spine for referred neck pain. Imaging revealed the presence of an encapsulated mass at the base of her tongue. The patient was not symptomatic for any compression of the airways. Diagnosis of ectopic lingual thyroid was confirmed by 99mTc scintigraphy. Incidental diagnosis of thyroid ectopia in asymptomatic adult patients is rare, and it should be considered on diagnostic imaging in case of an anterior midline cervical mass.

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Case report

A 30-year-old woman with no significant medical history presented with long-lasting neck pain. The patient’s family, surgical, and medication history was noncontributory. She had 2 uncomplicated pregnancies in the past, and she was not taking any medication at that time. Her physical examination was unremarkable. She came into the outpatient clinic of our institution for magnetic resonance imaging (MRI) of the cervical spine.

Cervical spine MRI demonstrated soft tissue mass at the base of tongue. On both T1- and T2-weighted sequences, the mass showed heterogeneous signal intensity, and multiple cysts were visualized in its central portion. This tissue did not have any significant mass effect on the surrounding structures or cause airway compression (Fig. 1).

MRI examination of the neck with gadolinium was then performed, confirming the presence of the encapsulated midline mass with small cysts within the tissue, localized on the upper edge of the epiglottis. It appeared isointense if compared to the surrounding musculature on T1-weighted sequences. No signs of compression were evident. The mass demonstrated early enhancement, which persisted into the late contrast phase (Figs. 2-4).

At this time, the diagnosis of a lingual thyroid (LT) was suspected. Laboratory tests showed a free triiodothyronine (FT3) of 3.3 pg/mL (normal range, 2.0-4.2 pg/mL), a free tetraiodothyronine (FT4) of 10.5 pg/dL (normal range, 9.3-16.0 pg/dL), and thyroid-stimulating hormone of 17.848 IU/mL (normal range, 0.37-4.2 IU/mL).

Ultrasound examination of the neck revealed the absence of the thyroid gland in the pretracheal region and no lymphadenopathy of the cervical packages. The patient performed 99mTc scintigraphy. Isotope uptake was observed at the base of tongue in the sublingual region, and the pretracheal region did not show any 99mTc uptake (Fig. 5). The suspected diagnosis of lingual localization of thyroid ectopia was then confirmed.

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Fig. 1 — Cervical spine magnetic resonance imaging in a 30-year-old woman with neck pain. T2-weighted sequence on the sagittal plane showing lingual ectopic thyroid (indicated by arrow).

Fig. 2 — Magnetic resonance imaging T2 FAT SAT sequence on the axial plane showing thyroid tissue at the base of tongue (indicated by arrow).

Fig. 3 — Magnetic resonance imaging THRIVE sequences on the axial plane, showing early vascularization of the ectopic thyroid tissue after gadolinium injection.

Fig. 4 — Magnetic resonance imaging T1-weighted after gadolinium sequence on the axial plane, showing homogeneous signal intensity of the whole tissue after gadolinium.
Discussion

Ectopic localization of the thyroid gland is an embryologic anomaly that occurs in the process of thyroid migration. The thyroid gland is derived from an endodermal thickening in the floor of the primitive pharynx. It arises between the first and second brachial arches. Its development begins at the 24th day of intrauterine life. At first, it is localized at the boundary between the body and the base of the tongue. Around the 7th week of intrauterine life, the thyroid cord forms the thyroglossal duct, and it descends becoming solid tissue, localized at the height of the embryologic laryngeal-tracheal skeleton. The newly formed thyroid tissue follows the descent of the heart and the major vessels, reaching the height of the growing hyoid bone. At this time, the thyroid reaches its final form and position and the thyroglossal duct disappears. Persistence of thyroglossal duct even after birth leads to the formation of thyroglossal cyst. Thyroid activity begins around the 11th week of gestation; from the 20th week, hormonal levels start to increase, and they stabilize to the adult level within the 35th week. Pathogenesis of this condition remains unknown. It has been suggested that maternal antibodies may arrest the descent of thyroid tissue and that the incidence of this disease among family members can be higher if
compared to the general population [1]. In some cases, the variable architecture of the cervical vessels and the different origin of the carotid arteries from the aortic arch can influence thyroid morphogenesis, and then explain the presence of ectopic thyroid tissue [2].

The most common site of thyroid ectopia is the lingual region, which is encountered in about 90% of ectopic cases, and it was described for the very first time in 1869 [3]. Other sites of ectopia can be the sublingual region, the hyoid/prelaryngeal area, the lateral regions of the neck, and rarely, the mediastinum [3,4] (Fig. 6). Extremely rare localizations can be pharynx, esophagus, trachea, lung, heart, breast, duodenum, mesentery, and also the adrenal glands [5]. The prevalence rates of LT vary from 1 in 100,000 to 1 in 300,000, with females-to-male ratio ranging from 4:1 to 7:1 [6]. The definition of LT can be used only if thyroid tissue is visualized at the base of the tongue, between the epiglottis and circumvallate papillae, and if scintigraphy demonstrates 131I or 99mTc radiotracer uptake (Table 1).

A normal thyroid is absent in the pretracheal region in up to 80% of the cases of LT. Most symptoms related to an LT are due to oropharyngeal obstruction: dysphonia, dysphagia (from mild to severe), a lump-in-throat sensation, dysnea, and even chronic cough or sleep apneas, but they vary depending on the size of ectopic tissue [7]. Ectopic thyroid gland can encounter all diseases of the normally localized thyroid tissue. Typically, diagnosis occurs during early childhood or puberty adolescence because patients are symptomatic: ectopic tissue is not able to meet physiological requirements, resulting in hypothyroidism in up to 60% of the patients [8,9]. Diagnosis in the adulthood can occur for rapid dimensional increase of overstimulated ectopic tissue during hypermetabolic conditions, such as pregnancy, infections, trauma, and menopause. Our patient had not encountered any discomfort or any hypothyroidism symptoms during her 2 pregnancies.

Some authors have reported that histologic confirmation is needed for a definitive diagnosis of LT [10,11], but it was not performed on our patient, because the ectopic tissue was functioning. On fine-needle aspiration, lingual tissue always resembles normal thyroid follicles. Fine needle aspiration can be considered only if the risk of bleeding and other complications are justified.

In symptomatic patients, levothyroxine suppression therapy can be successful, but for those cases of severe symptoms, a surgical approach is recommended. Unless emergency surgery is needed, suppressive therapy with exogenous thyroid hormone should be tried before surgery to decrease the size of the gland. In case of oropharyngeal severe symptoms, adult patients may refer to radiology centers for a computed tomography (CT) scan. This explains why many case reports in the literature show CT images of incidentally diagnosed ectopic thyroid.

Differential diagnosis of an LT can be metastatic thyroid carcinoma, lingual thyroglossal duct cyst, branchial cleft cysts, dermoid cysts and epidermoid cysts, lingual abscesses, lymphatic malformations, and squamous cell carcinoma of the base of the tongue (Table 2).

Because an increasing number of patients are undergoing routine diagnostic examinations (MRI, ultrasound, and CT), nowadays, this anomaly can represent an incidental diagnosis more frequently than in the past. Thus, thyroid ectopia has been reported more often in the latest literature. Knowledge of the embryologic thyroid typical course is crucial to understand the variant locations along this path. Because lingual localization of thyroid ectopia might remain unknown after puberty if the patient is not symptomatic, it should be considered in the differential diagnosis of enhancing neck lesions in adults.

| Table 1 – Summary table. |
|--------------------------|
| **Etiology** | **Unknown** |
| Prevalence | Overall lingual ectopia prevalence varies between 1:100,000 to 1:300,000. Average prevalence among patients with thyroid disease: 1 in 4000-8000. Lingual ectopia: 90% of cases. |
| Gender ratio | Female gender predilection (female-to-male ratio ranging from 4:1 to 7:1). |
| Age predilection | Age at the time of diagnosis can vary. Diagnosis is more frequent during childhood. |
| Risk factors | Unknown. |
| Treatment | Medical treatment with hormone-suppressing or hormone-stimulating therapy. Severe cases may require surgical approach. |
| Prognosis | Prognosis is variable. |
|                  | X-ray | US                          | CT                              | MRI                              | Nuclear medicine imaging |
|------------------|-------|-----------------------------|---------------------------------|----------------------------------|--------------------------|
| Lingual thyroid  | Nondetectable | No thyroid is visualized. No associated lymphadenopathy. | Multiloculated encapsulated mass, on the midline. Contrast enhancement: present. | T1 and T2 variable signal intensity. Multiple cysts inside encapsulated mass. T1 + Gd: heterogeneous enhancement. | SCINTI: at the level of the posterior third of the tongue, midline, functioning. |
| Metastatic thyroid carcinoma (only with X-ray of the neck) | Nondetectable | Hypoechoic lesions; irregular margins. Concurrent cervical lymphadenopathy. | Variable presentation. Non–well-defined mass. Possible great extension. Heterogeneous enhancement. | T1 and T2 variable signal intensity. T1 + Gd: heterogeneous enhancement. | PET: diffuse spreading. Functioning. |
| Lingual thyroglossal duct cyst | Nondetectable | Fluctuant mass, with thin walls and anechoic central component. No vascularization (unless infected). If complicated: internal debris and heterogeneous echogenicity. US–imperceptible walls. Variable echogenicity. | Thin walled, smooth, homogeneous mass. Possible rim enhancement. Sternalcleidomastoid muscle is typically displaced posteriorly or posterior-laterally. | T1: variable signal intensity: low signal if low protein content. T2: typically high signal intensity. T1 + Gd: no enhancement in uncomplicated lesions. Possible rim enhancement. | SCINTI: nonfunctioning, normal thyroid present. |
| Branchial cleft cysts | Nondetectable | Sharply demarcated mass, typical posterior acoustic enhancement. US–imperceptible walls. Variable echogenicity. | Nodular, well-defined mass with thin walls and typical central fluid density. At the level of carotid bifurcation. Enhancing if complicated. | T1: variable signal (high signal intensity if high protein content) T2: usually high signal T1 + Gd: no enhancement in uncomplicated lesions. | Nondetectable if not complicated, usually nonfunctioning. |
| Dermoid and epidermoid cysts | Nondetectable | Well-defined anechoic homogeneous mass on the midline. Heterogeneous appearance, due to the presence of echogenic fat, osseous, or dental elements. | Low-density, well-circumscribed mass. Fat, mixed-density fluid, and calcification (<50%) may also be seen. Nonenhancing if uncomplicated. | T1 hypointense well-defined mass. T2 hyperintense heterogeneous mass. T1 + Gd nonenhancing if not complicated. | |
| Lingual abscesses | Nondetectable | Complicated mass, with internal debris and heterogeneous echogenicity. Vascularized. | Heterogeneous density. Rim enhancement and heterogeneous enhancement. Lymphadenopathy. | T1 and T2 variable signal intensity. T1 + Gd: heterogeneous and remarkable enhancement. | PET: functioning. |
| Squamous cell carcinoma of the base of the tongue | Nondetectable | Not gold standard examination | Heterogeneous and enhancing mass located at the base of the tongue. It can be lateral or on the midline. Associated lymphadenopathy. | T1 and T2 variable signal intensity of the mass, typically bulky, lobulated. T1 + Gd: heterogeneous and remarkable enhancement. | PET: functioning. |

CT, computed tomography; MRI, magnetic resonance imaging; PET, positron emission tomography; US, ultrasound.
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