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Lateral ectopic thyroid: A case diagnosed preoperatively

Introduction

An ectopic thyroid is a mass of benign thyroid tissue that is located at a site other than the pretracheal area anterior to the second through fourth tracheal rings.[1,2] Ectopic thyroid is uncommon, with a reported prevalence in the range of 1 per 100,000 to 300,000 population; among patients with thyroid disease, the prevalence ranges from 1 in 4,000 to 8,000.[1,2]

Embryology. The thyroid gland is derived from two types of anlage: (1) a large median endodermal anlage that produces most of the thyroid parenchyma and (2) two lateral anlages derived from the fourth pharyngeal pouch that contributes to 1% of the thyroid mass, from where the parafollicular cells are derived.[3-5] Transcription factors TTF-1, Nkx2, Pax-8, FOXE1, and Hhex are involved in the process of organogenesis and migration of the thyroid gland.[3-5]

Around the fourth week of gestation, the thyroid gland descends through the thyroglossal duct as a bilobed diverticulum from an invagination in the foramen cecum from the base of the tongue to the front of the anterior tracheal wall, where it reaches its final position in the seventh week.[3,4] Toward the end of the migration, the lateral thyroid primordium dissociates from the caudal base of the fourth pharyngeal pouch and the ultimobranchial body, which contains parafollicular cells, and migrates medially to join the thyroid gland.[3-5]

Pathophysiology. A failure to descend or an incomplete migration of the medial anlage of the thyroid during embryologic development leads to midline or near-midline ectopias, such as a lingual thyroid or a thyroglossal ectopia.[2-5] In rare cases, an aberrant migration with cell rests deposited laterally during the development of the gland or a failure of the lateral anlage to fuse with the median anlage can result in the development of lateral aberrant thyroid tissue.[3-5] When the process of migration is disturbed, aberrant thyroid tissue may appear in later life; in such cases, it is generally located along the tract of migration.[1,4-8] An ectopic thyroid in the lateral position is located at the submandibular area in 70% of cases; in other cases, it is located near the carotid sheath (table 1).[9]

Table 1. Reported locations of ectopic thyroid[1,2,9,11]

Location	Cases (%)
Midline of the head/neck (along tract of migration)	80 to 95
Lingual thyroid	80 to 90
Thyroglossal duct cyst	5 to 15
Lateral neck (aberrant migration)	1 to 3
Submandibular area	70
Subplatysmally, medial to the carotid sheath	30
Aerodigestive tract (rests left behind)	1

Location	Cases (%)
Thorax or abdomen (overdescended)	<1

Clinical presentation. The clinical presentation varies according to the location and function of the ectopic thyroid. The ectopic tissue is prone to functional insufficiency and, as is the case with lesions in other locations, it might come to attention only after compensatory enlargement, which is otherwise asymptomatic. Hypothyroidism is seen in one-third of patients.[1,2]

Ectopic thyroids in the lateral position are usually slowly growing, nontender, solid masses that are not mobile with swallowing.[9]

Differential diagnosis. Lateral ectopic thyroid should first be differentiated from metastatic thyroid cancer. In fact, in most patients, lateral aberrant thyroid tissue may represent a lymph node with metastatic disease, even in the absence of signs of thyroid carcinoma and especially if biopsy also demonstrates elements of lymphoid tissue that have not been replaced by the tumor.[2,10]

Malignancy has been reported in about 12% of true lateral ectopic thyroids.[2] Other considerations should include submandibular tumors (e.g., pleomorphic adenoma or carcinoma), inflammatory lesions (e.g., Küttner tumor), branchial cleft cysts, lymphangiomas, carotid body tumors, and lymphadenopathy of various etiologies.[2,10]

Diagnostic workup. The diagnosis of ectopic thyroid tissue should be considered in any young woman with an asymptomatic midline or lateral (submandibular) neck mass, since the great majority of cases occur in women (table 2).[1,9-11] The possible presence of ectopic thyroid, which represents the sole source of functional thyroid tissue in 60 to 75% of cases, is the rationale for preoperative imaging and a correct diagnosis.[1,8]

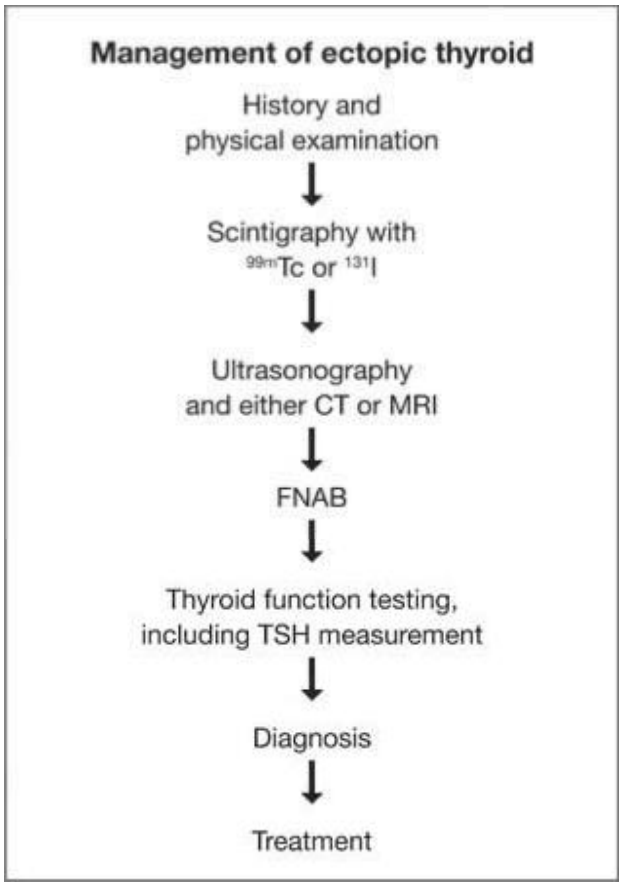
Table. 2. Reported clinical and pathologic characteristics of lateral ectopic thyroid[1,9-11,30,38]

Characteristic	Cases (%)
Age, yr	
≤19	3.5
20 to 45	50.0
≥46	46.5
Sex	
Female	79
Male	21
Symptoms	
No	60
Yes	40

Characteristic	Cases (%)
Site	
Submandibular area	70
Other	30
Pathology	
Benign	76
Malignant	12
Unknown	12

When findings on an adequate history and physical examination have raised a clinical suspicion of ectopic thyroid presenting as a neck mass, the diagnostic workup should include scintigraphy with either technetium-99m (^{99m}Tc) or iodine -131 (^{131}I), ultrasonography and either computed tomography (CT) or magnetic resonance imaging (MRI), fine-needle aspiration biopsy (FNAB), and thyroid hormone testing, including measurement of the thyroid-stimulating hormone (TSH) level (figure 1).[1,9]

Figure 1. Algorithm shows a stepwise approach to the management of ectopic thyroid.



CT with contrast should be obtained after scintigraphy is performed; otherwise, the content of the CT contrast material might provoke hyperthyreosis and possibly result in an inaccurate scintigraphy result. A single FNAB may not be diagnostic, and it cannot completely rule out a malignancy, especially a follicular carcinoma. Clinical photographic documentation is also advised.

In surgical cases, frozen-section analysis should be performed intraoperatively. It is important to rule out a well-differentiated metastasis that has totally replaced a lymph node in cases in which the primary thyroid carcinoma is small or even microscopic. Frozen-section evaluation can also yield the histologic diagnosis and help prevent the removal of benign ectopic tissue, which represents the patient's principal functioning thyroid.[2,12]

Treatment. Most of the reported cases of true lateral aberrant thyroid tissue were diagnosed after surgical excision for an enlarging mass of unknown etiology and indeterminate FNAB results.[1,2,10] Hormonal suppression can decrease the size of an ectopic submandibular goiter, and therefore it should be attempted prior to surgery.[1]

Surgical excision has the advantage of ruling out a diagnosis of metastatic thyroid cancer by allowing for pathologic analysis of the mass.[2] However, unless carcinoma is suspected or diagnosed, ectopic submandibular thyroid tissue should not be excised because of the inherent risk of hypothyroidism.[2] No treatment is required for a patient who has an asymptomatic ectopic thyroid with normal thyroid function and cytology.[1]

To the best of our knowledge, fewer than 40 cases of lateral ectopic thyroid have been reported in the literature.[6-40] In this article, we report a new case.

Case report

A 34-year-old woman was referred to our ENT department for evaluation of an asymptomatic left cervical mass that had been gradually increasing in size over the previous year. She denied any episodes of pain or tenderness over the mass,

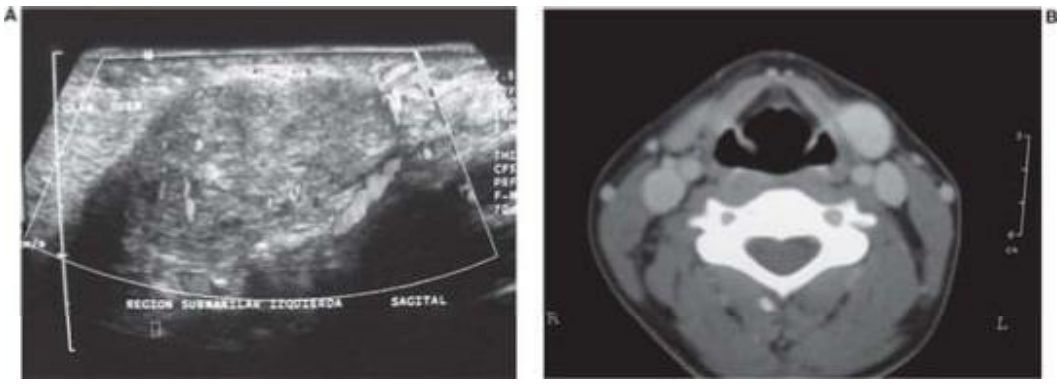
and she had no fever, swallowing difficulty, or voice changes. She also denied recent weight changes or any other symptoms related to hypothyroidism.

On physical examination, the patient had a firm, mobile, nontender, 3 x 2-cm mass in the left submandibular area (figure 2). On bimanual palpation, the mass was independent of the submandibular gland, and it did not move with swallowing. No associated lymphadenopathies were palpated. The thyroid gland was not palpable in its normal position. Findings on examination of the tongue base and a fiberoptic examination of the pharynx and larynx were normal.

Figure 2. At presentation, the mass in the lateral neck measures 3 x 2 cm.

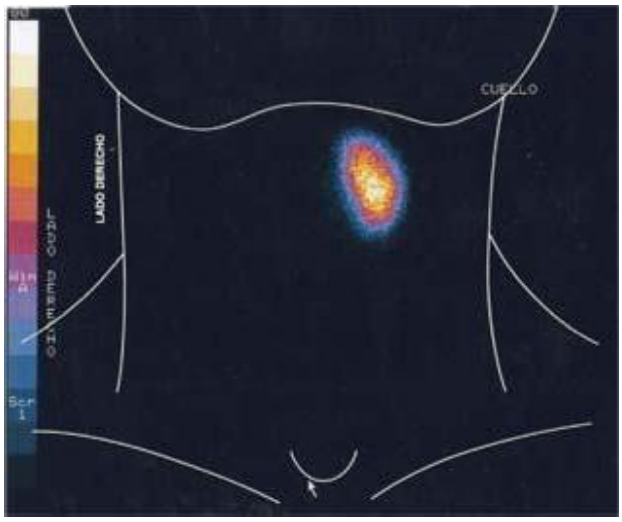
Prior to referral, the patient had undergone ultrasonography and contrast-enhanced CT of the head and neck. The ultrasonography demonstrated a solid, moderately vascular mass adjacent to the submandibular gland (figure 3, A), while the CT showed a solid, enhancing, 2-cm submandibular mass without calcifications (figure 3, B). The left submandibular gland was pushed posteriorly, and an absence of an orthotopic thyroid gland was observed.

Figure 3. Previously obtained ultrasonography (*A*) and contrast-enhanced CT (*B*) show the solid mass.



Given the clinical picture and imaging results, a presumptive diagnosis of ectopic submandibular thyroid was considered. On ^{99m}Tc scintigraphy, an ectopic concentration of the tracer was seen in the left submandibular area without uptake in the pretracheal thyroid area (figure 4). (As mentioned, it would have been better to obtain scintigraphy prior to CT for diagnostic purposes, but the CT had already been performed elsewhere before the patient was referred to us.)

Figure 4. On ^{99m}Tc scintigraphy, uptake of the tracer is seen only in the left submandibular area.



Thyroid function testing revealed that the patient's serum TSH level was slightly elevated, which suggested subclinical hypothyroidism. FNAB analysis revealed benign thyroid cells.

The patient was treated with levothyroxine supplementation, which resulted in shrinkage of her mass and normalization of her serum TSH level. Her triiodothyronine (T3) and thyroxine (T4) levels remained within normal ranges.

During follow-up, a repeat FNAB identified normal follicular cells. At 3 years from presentation, the patient remained asymptomatic and euthyroid under thyroid hormone replacement therapy.

Discussion

Ectopic thyroids are located along the embryologic descent path as either a lingual thyroid (80 to 90% of cases) or a thyroglossal duct cyst (5 to 15%).[1,2] Only 1 to 3% of all ectopic thyroids are located in the lateral neck.[9,11] Reported cases have occurred in patients aged 12 to 81 years, with almost all occurring in adults (table 2).[9,11] As mentioned, the vast majority of reported cases (79%) have occurred in women, as did our case.[9,11]

The clinical presentation of our patient-with a slowly enlarging asymptomatic mass-was similar to those of previously reported cases.[9,11] Our patient's case was also typical in that her lateral ectopic thyroid was located in the submandibular area (70% of cases); a smaller proportion of these lesions are located subplatysmally, medial to the carotid sheath.[9,11] Finally, our case was similar to most others (59%) in that our patient did not have a thyroid gland in the orthotopic position.[9,11] It is noteworthy that in 65% of cases of lateral ectopic thyroid, there was another site of thyroid tissue; in 63% of those cases, the other site was the normal orthotopic pretracheal position, and in 37%, it was elsewhere, usually as a coincident lingual ectopic gland or as a bilateral submandibular ectopic thyroid.[9,11] In cases of ectopic lingual thyroid, less than 25% of cases include a second thyroid tissue site.[9,11]

It is important to stress the importance of clinical suspicion and a diagnostic workup-including scintigraphy, ultrasonography, either CT or MRI, FNAB, and thyroid function testing-for masses that appear laterally in the anterior neck. Most of the reported cases of lateral ectopic thyroid (76%) were diagnosed after they had been surgically removed.[9,11] In these cases, the patients lost their only functional thyroid tissue, which put them at risk for hypothyroidism. Fortunately for our patient, she was diagnosed without surgical resection.

The decision regarding whether to excise lateral ectopic thyroid tissue should be made judiciously and based on many factors: the patient's symptoms, the degree of cosmetic deformity, the presence or absence of other thyroid tissue, pathologic findings on FNAB, the results of other diagnostic tests, and the results of drug therapy.[1]

Some 76% of cases of true lateral ectopic thyroid tissue were reported as benign on cytopathologic or histopathologic examination.[9,11] Of the remainder, 12% of cases were malignant and 12% were undetermined.[30,38] Roughly half of the malignancies were follicular carcinomas, and half were papillary carcinomas.[1,2] It has been reported that malignancy is not more common in ectopic thyroid tissue than it is in orthotopic thyroid tissue (5% of cases).[1,2,40] While this may be true for the most common type of ectopic thyroid (i.e., those at the lingual position), in cases of lateral and thyroglossal ectopic thyroid, malignancy has been reported in 12 and 20% of cases, respectively.[9,11]

In cases of laterally located thyroid tissue, a metastasis of occult thyroid carcinoma should be considered, especially if the thyroid tissue is within a lymph node or located lateral to the carotid sheath.[9,11]

From the Department of Otolaryngology-Head and Neck Surgery, Hospital General Dr. Manuel Gea González, Distrito Federal, México. Corresponding author: Héctor Prado Calleros, Durango #49-801, Colonia Roma, 06700 Distrito Federal, México. Email: hmpradoc@hotmail.com

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