Multiple Ectopia of the Thyroid Gland: A rare Case Report

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ABSTRACT

Multiple ectopia of the thyroid gland is a term which refers to presence of multiple thyroid tissue in absence or presence of orthotopic thyroid gland. The prevalence of ectopic thyroid is approximately one per 100,000 to 300,000 persons and is reported to occur in one in 4,000 to 8,000 patients with thyroid disease. Multiple ectopia of thyroid is extremely rare entity. We present a case with multiple thyroid ectopia in absence of orthotopic thyroid gland. CT and MRI play an important role in locating ectopic thyroid tissue.

Key words: Computed tomography; magnetic resonance imaging; thyroid dysgenesis; thyroid ectopia; thyroid gland

Introduction

Ectopic thyroid gland is a rare entity which occurs as a developmental aberration which leads to presence of multiple ectopias of the thyroid tissue in the presence or in few cases in absence of orthotopic thyroid gland. The ectopic thyroid sites may be in line with the normal decendence of the gland from the foramen caecum in line with the thyroglossal duct or it may be offline to this path as in our case. We encountered ectopic thyroid tissue at four regions namely across the base of the tongue (posteriorly), the sublingual, the prehyoid and the right submandibular regions.

Case Report

Clinical history

A 25-year-old male patient presented with a chronic painless swelling across the right submandibular region. Ultrasound of the region of interest done at peripheral center suggested neck lesion without mention of the thyroid gland and patient was referred to higher center for further evaluation. Ultrasound evaluation of entire neck region was done at our institute, followed by computed tomography (CT) and magnetic resonance imaging (MRI) scan. The findings were confirmed with fine needle aspiration cytology (FNAC).

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Imaging findings

- Ultrasound examination of the neck revealed a well defined, rounded, heterogeneous, mixed echogenic mass in the right submandibular region [Figure 1]. At the level of the thyroid cartilage, no obvious thyroid gland was seen bilaterally.
- Axial CT revealed well defined hyperattenuating soft tissue mass lesions across the base of the tongue (posteriorly), the sublingual, the prehyoid and the right submandibular regions [Figure 2]. The lesion across the base of the tongue and the sublingual lesion appeared in the midline and were connected with a stalk of similar attenuations best seen on sagittal image [Figure 3]. The prehyoid lesion and the sublingual lesion were also connected through a stalk, which were seen to run through supero right lateral aspect of the hyoid bone. The right submandibular lesion was located adjacent to prehyoid lesion, deep to the platysma and anterior to the sternohyoid and the thyrohyoid muscles. No evidence of any connecting stalk was seen with the other lesions. On post contrast evaluation, all these lesions showed uniform almost homogeneous intense enhancement [Figure 4]. Few small cystic areas were also seen within these lesions. No evidence of other similar lesions were seen. On MRI all these lesions appeared predominantly muscle isointense on the T1 and the T2 weighted sequences [Figure 5]. There was no evidence of bilateral thyroid lobes or isthmus seen in the normal anatomical lower neck location [Figure 6].
- FNAC from the right submandibular lesion revealed thyroid tissue [Figure 7].

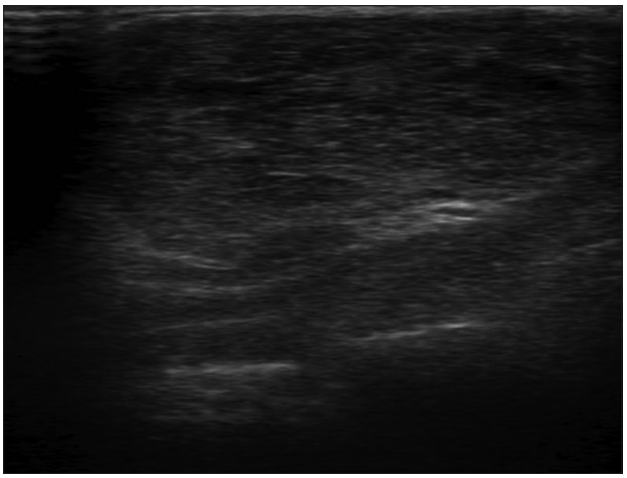

Figure 1: On ultrasound of the neck a well-defined rounded, heterogeneous mixed echogenic mass was seen in the right submandibular region

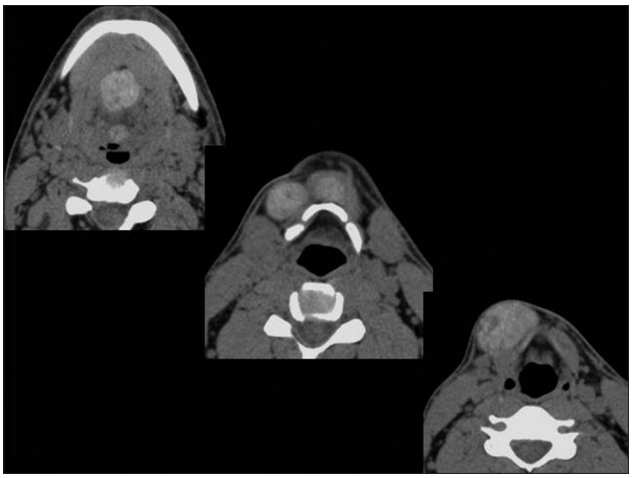

Figure 2: Axial plain computed tomography scan demonstrating well defined rounded hyperattenuating lesions at the base of the tongue, sublingual, prehyoid and right lateral cervical submandibular region

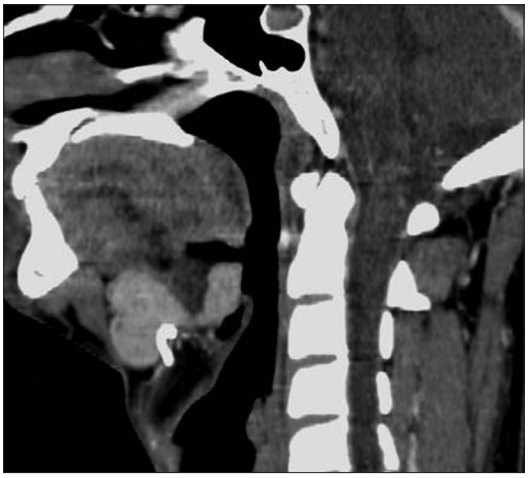

Figure 3: Postcontrast sagittal reconstruction showing a similar attenuation stalk connecting the base of the tongue lesion with the sublingual lesion

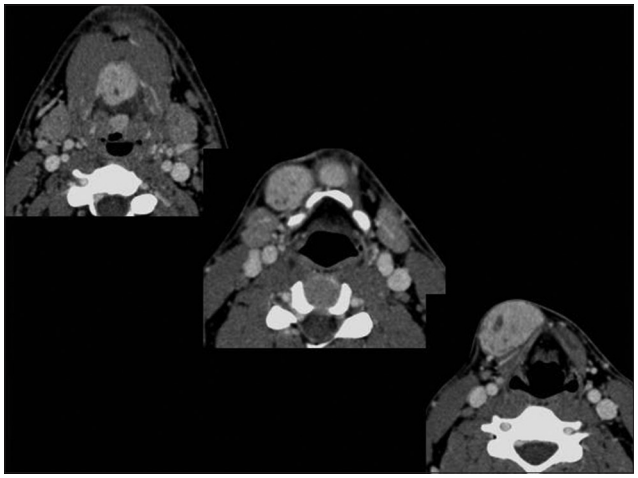

Figure 4: Axial contrast enhanced computed tomography scan at same levels as above demonstrating uniform almost homogeneous brilliant enhancement

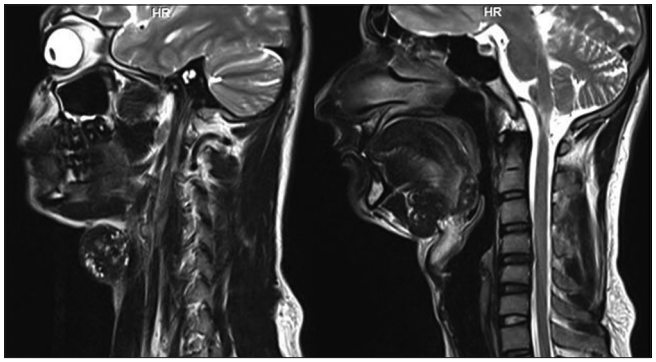

Figure 5: On T2-weighted sagittal magnetic resonance images, the ectopic thyroid glands appears predominantly isointense to the muscle

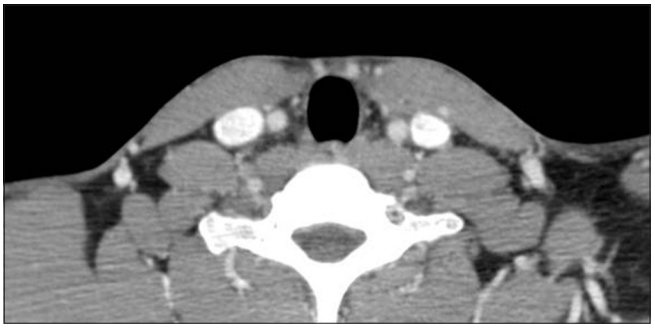

Figure 6: Axial computed tomography sections show absence of thyroid lobes at their anatomical location

Final diagnosis

Multiple thyroid ectopia with the absence of thyroid tissue in its normal anatomical location.

Differential diagnosis

- Thyroid cancer metastasis
- Midline masses like thyroglossal cysts.

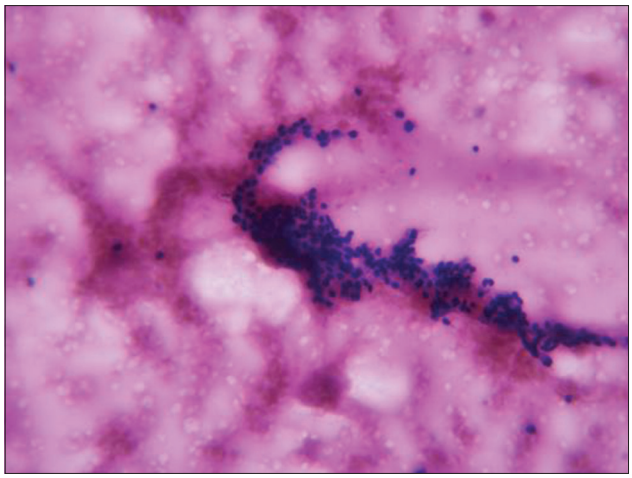

Figure 7: Smears show few clusters of thyroid follicular cells. Background shows plenty of foamy and hemosiderin laden macrophages

Discussion

Ectopia of the thyroid gland is a rare entity, and it may not be clinically detected until adulthood. Only a few cases of thyroid ectopia are reported in the literature.[1,2] Association of multiple ectopia without the presence of the normal orthotopic thyroid gland is very rare.[3] Some of the anomalies are detected as incidental findings in asymptomatic patients. The developmental abnormalities of the thyroid gland can be divided into three major groups agenesis, dysgenesis and abnormalities due to the persistence of the thyroglossal duct.[4]

The thyroid gland is the first endocrine gland to be developed in a fetus by 4th week.[5] During embryogenesis, the endodermal diverticulum from the median plate of the floor of the pharyngeal gut descends from the midline from the foramen cecum to the final location of the gland. During migration, a narrow tract called the thyroglossal duct is created, which later atrophies. Failure of migration leads to ectopia in the midline. In rare cases when the cells of the lateral anlage do not join the median, a lateral ectopic thyroid gland is formed. In such cases, the location is usually submandibular region.[6]

The most common location for ectopic thyroid is the lingual thyroid, just posterior to foramen caecum, The extra-lingual sites include sublingual, perihyoid, intratracheal, laryngo-tracheal, intraesophageal, mediastinal, or cardiac or even abdomen.[4,7]

The mean age of presentation may vary depending upon the symptoms. In asymptomatic cases, the finding is incidental. In symptomatic cases they may present at any age from birth to 80 years.[6] The most common symptoms are related to the growth of the lingual thyroid presenting as dysphagia, dysphonia, foreign body sensation, cough, snoring or sleep apnea.[8] Patients with dual ectopia usually present with neck swelling.[9]

In terms of function, most of the patients with the lingual thyroid are hypothyroid. Fifty percent of patients with dual ectopia are euthyroid. In our case also the patient was euthyroid, and it presented with swelling. However, patients with ectopic thyroid with normal thyroid function can become hypothyroid during puberty, menstruation, pregnancy, infections, trauma and surgery. This hypothyroid status can inturn trigger enlargement of ectopic thyroid tissue.[10]

For diagnosing ectopic thyroid, ultrasound, CT scan and MRI are useful. They also help in detecting extension and location of the ectopic thyroid gland.[6] Ultrasound is inexpensive and readily available modality. Some of the ultrasound features help to distinguish benign and malignant nodules.[6,11] CT can be used to scan the entire neck from the nasopharyngeal region to the mediastinum. On plain CT thyroid tissue shows higher attenuations than surrounding soft tissues. On contrast administration it shows intense enhancement. In our case CT was useful in delineating the extra lesions that were not picked up on ultrasound, namely the lesions at the base of the tongue, the sublingual and the prehyoid lesions. The main disadvantage of CT is that after the administration of iodinated contrast media, iodine based nuclear medicine agents cannot be performed until 6 weeks. A typical CT study uses about 100 cc of intravenous contrast material, which translates to about 30 g of iodine. This transient increase in iodine can compete with ^{131}I and interfere with subsequent management, such as whole-body scans and treatment with radioactive iodine. Thus, avoidance of iodinated contrast material is particularly important as it can lessen the effectiveness of radioactive therapy.[12] MRI is nonradiation based technique which also gives information about the extent and location of the lesion. The ectopic thyroid tissue mostly appears as rounded mass with signal intensity similar to muscle on the T1-weighted and the T2-weighted sequences. In our case also the lesions are isointense to muscle on the T1-weighted and T2-weighted sequence. However, on convention MRI in fat suppressed sequences, susceptibility artifacts may cause imaging problem. This is overcome by eight channel high resolution MRI scanners.[13] Sometimes visualization of the ectopic thyroid gland may be missed because some of the lesion appear muscle isointense and hence difficult to delineate. Scintigraphy using Tech - 99 m, I-131r I-123 is the most important diagnostic tool to detect ectopic thyroid tissue. CT and MRI are useful in cases when the radiotracer uptake by normal thyroid gland masks the uptake of ectopic thyroid tissue especially in the midline.[6] FNAC is however, confirmatory in the diagnosis of ectopic thyroid.

Primary thyroid cancers arising from ectopic thyroid are uncommon.[6] Management is based on the size and local symptoms. For completely asymptomatic and euthyroid patients regular follow-up is recommended in order to look for mass enlargement or development of complications. In our case, the patient was conservatively managed. For mildly symptomatic and hypothyroid states, levothyroxine replacement therapy may be effective leading to considerable mass reduction.[6] When medical treatment fails or there are obstructive symptoms or occurrence of any complications like hemorrhage or suspicion of malignancy, then surgery should be considered.

Conclusion

Developmental defects that occur at an early stage of embryogenesis generate ectopic thyroid tissue, which may be seen anywhere along the gland's embryological pathway as well as distant areas. Ultrasound, CT and magnetic resonance scanning are among the diagnostic modalities though thyroid scintigraphy plays an important role in diagnosis. In majority of cases, patients are asymptomatic, but when symptoms arise due to size and location of the lesion or its malignant transformation, then surgery needs to be considered. Radioiodine ablation also plays a significant role in cases of recurrence.

Relevance

Ectopic thyroid gland presenting as lateral neck swelling should not be operated without checking for the presence of the thyroid gland in its normal anatomical location because this might be the only functional thyroid tissue. Ultrasound of the neck masses, in general, should involve detailed scan of the entire neck region and not a just region of interest. Hence our case implies the significance of the detailed neck ultrasound which was carried out at our institute where absence of thyroid gland was seen in normal anatomical location and contrast-enhanced CT was done to look for the number and sites of ectopic thyroid tissue.

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