

Dual ectopic thyroid: report of a case

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Abstract

Dual ectopic thyroid is very rare. We report a case of dual ectopic thyroid in the lingual and infrahyoid areas in a 20-year-old female patient with no thyroid gland in its normal anatomical location. On physical examination, there was a 7 × 5 cm anterior midline neck swelling just below the hyoid bone and a 2 × 2 cm mass in the base of the tongue. Triiodothyronine (T₃), thyroxine (T₄), and thyroid-stimulating hormone (TSH) levels were normal. A thyroid scan with technetium-99m sodium pertechnetate confirmed dual ectopic thyroid with no iodine uptake in the normal anatomical location of the thyroid gland. The infrahyoid ectopic thyroid was surgically removed for cosmetic reasons, and the lingual thyroid, which was symptomatic, was left untouched. The importance of thyroid scanning in the evaluation of anterior midline neck swellings and treatment options are discussed.

Key words: Thyroid Gland, Abnormalities

Introduction

The thyroid is the first endocrine gland to appear in embryonic development. It begins to develop about 24 days after fertilization from a median endodermal thickening in the floor of the primitive hypopharynx. This thickening soon forms a downgrowth or outpouching known as the thyroid diverticulum. As the embryo and tongue grow, the developing thyroid gland descends in the neck, passing ventral to the developing hyoid bone and laryngeal cartilages.¹ For a short time, the developing thyroid gland is connected to the tongue by a narrow tube, the thyroglossal duct, which normally involutes and atrophies by the sixth or eighth week. The opening of this duct in the tongue is called the foramen caecum. During this time, the descending thyroid component, also known as the medial anlage, meets and fuses with the other primitive thyroid tissue, the right and left ultimobranchial bodies, which are derivatives of the fourth branchial pouches. The medial and lateral components fuse and begin to function as mature thyroid by the third fetal month.²

Ectopic thyroid tissue is an uncommon entity resulting from abnormal migration of the gland. Its prevalence is approximately 1 per 100 000 to 300 000 persons and is reported to occur in 1 in 4 000 to 8 000 patients who have thyroid disease.³ Dual thyroid ectopy is very rare; our MEDLINE review to June 2002 revealed only eight previously reported cases. Anatomically, ectopic thyroid can be classified as lingual (just below the foramen caecum), sublingual (between the geniohyoid and mylohyoid muscles), or prelaryngeal (just above or below the level of the hyoid bone).^{3,4} Other rare sites of occurrence of ectopic thyroid are the mediastinum, precardial sac, trachea, oesophagus, pharynx, lung, heart, breast, duodenum, and mesentery of the small intestine.^{3–7}

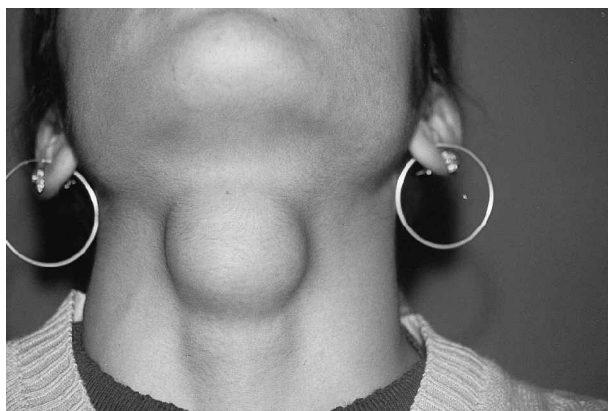


FIG. 1
Midline anterior neck swelling.

Case report

A 20-year-old female patient presented with a history of swelling in the anterior neck for 12 years. The swelling had enlarged gradually during these years, reaching its current size. The patient did not complain of any pain or pressure symptoms, her major complaint being the unpleasant appearance of the swelling in the neck.

Her past history and family history were negative. Examination of the neck revealed a soft, mobile, non-tender midline swelling at the level of the hyoid. Its dimensions were 7 × 5 cm (Figure 1). The swelling showed movement upon swallowing. On indirect laryngoscopic examination, a 2 × 2 cm mass in the tongue base was observed (Figure 2). The mass was soft in consistency and had a normal appearing mucosa overlying it. The thyroid gland was not palpable in its normal anatomical position. A thyroid scan with technetium Tc-99m sodium showed

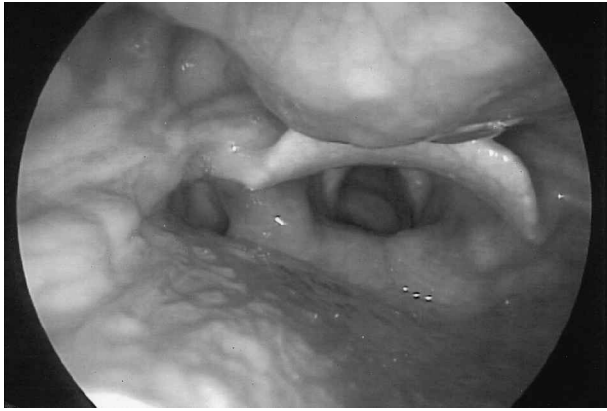


FIG. 2

Laryngoscopic view of the mass in the tongue base.

uptake in the tongue base and in the area of the anterior midline neck swelling (Figure 3). There was no uptake in the normal anatomical location of the thyroid gland. The T_3 level was 1.9 ng/ml (normal, 0.8–2.0 ng/ml), the T_4 level was 7.2 $\mu\text{g/dl}$ (normal, 5.0–14.0 $\mu\text{g/dl}$), the free T_4 level was 13 pmol/L (normal 12–22 pmol/L), and the TSH level was 3.48 mIU/L (normal 0.27–4.20). Based on these findings, the diagnosis of dual ectopic thyroid was made. The patient was observed for six months in the expectation of a reduction in the size of the cervical mass with thyroxine therapy. Since the mass did not show a significant reduction in size, surgery was planned for this cosmetic disturbance. During the operation, the mass was removed through a horizontal skin incision just below the level of the hyoid bone. The ectopic infrahyoid thyroid tissue was removed along with its capsule (Figure 4). The post-operative course was uneventful and the patient was discharged on the second day. The patient has been receiving a maintenance dose of thyroxine (100 μg daily) both before and since the operation. At the six months follow-up, the T_4 level was 9.3 $\mu\text{g/dl}$ (normal, 5.0–14.0 $\mu\text{g/dl}$) and the TSH level was 5.60 mIU/L (normal 0.27–4.20 mIU/L).

Discussion

Ectopic thyroid was first described by Hickman in 1869. It is seen more commonly in females; the female to male ratio being 4:1. Ectopic thyroid can occur at any age, but patients are usually seen during adolescence or pregnancy when the demands for thyroid hormones increase.⁷ The

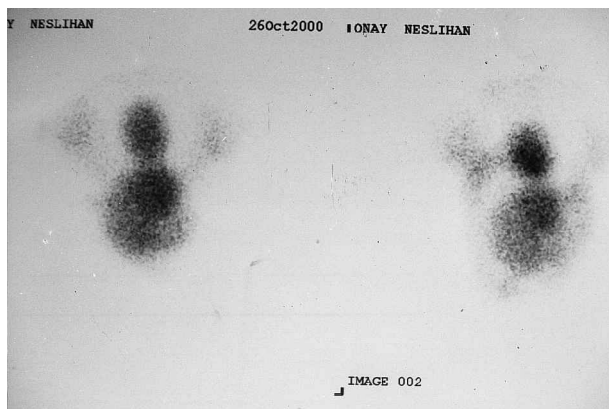


FIG. 3

Thyroid gland scan demonstrating uptake in the tongue base and anterior midline neck swelling.

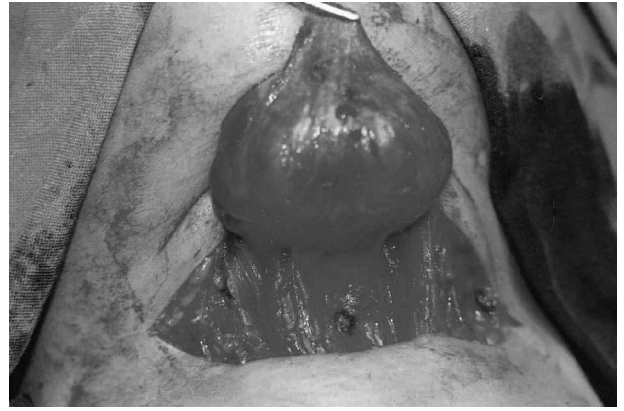


FIG. 4

Intra-operative view of the mass.

organism's demand for thyroid hormones elevates the circulating TSH levels causing ectopic thyroid tissue to increase in size. This in turn is clinically seen as an enlarging neck mass or with pressure symptoms such as dysphagia, dyspnoea, or dysphonia. Occasionally, a lingual thyroid may be an accidental finding complicating intubation, especially since mild trauma can cause profuse bleeding.⁸ However, most ectopic thyroids are asymptomatic and usually no therapy is needed. In our case, the infrahyoid mass was excised for cosmetic reasons, whereas the lingual mass was managed conservatively due to the fact that it was asymptomatic.

Congenital anomalies are rare among thyroid disorders. Thyroid ectopy occurs 1 in 4 000 to 8 000 patients with thyroid disease. Concurrent thyroid ectopy of the lingual and infrahyoid region is a much rarer entity that has been reported in only eight patients previously. Ninety per cent of all ectopic thyroids are found in the region of the foramen caecum, as a lingual thyroid. In approximately 75 per cent of the patients, this is the only functioning thyroid tissue in the body.⁴ The management of ectopic thyroid disease must be carefully planned, keeping in mind the risk of permanent hypothyroidism.

The diagnosis of an ectopic thyroid requires a high index of clinical suspicion. In addition to a detailed physical examination, the most important diagnostic tool is the thyroid scan. This provides an estimation on the size, the degree of activity of the ectopic thyroid, and also ascertains the presence or absence of normal cervical thyroid tissue. The differential diagnosis should include different conditions such as epidermal cysts, lymphadenopathy, lymphangioma, lipomas, sebaceous cysts, fibroma, minor salivary gland tumours, midline branchial cysts, and most importantly, thyroglossal duct cysts.^{3,8} In all of these conditions, the neck should be carefully examined for the presence of a thyroid gland in its usual anatomical location. The value of thyroid scan in differentiating thyroid ectopy from these diseases cannot be over-estimated. Ultrasonography, computerized tomography (CT), and magnetic resonance imaging (MRI) can be of help in determining the location and dimensions of the ectopic thyroid gland.

Hypothyroidism has been reported to occur in 33 per cent of patients with ectopic thyroids.³ Although the chemical structure of the thyroid hormones that are synthesized in the ectopic thyroid glands is normal, the amount that is produced may not be sufficient for increased physiological demands such as seen during puberty, pregnancy, infection, trauma, and surgery. The thyroid hormone profile of our patient was within normal

ranges. However, the TSH level was close to its physiological peak, showing the limited potential of ectopic thyroid tissue in producing thyroid hormones.

- **Dual ectopic thyroid tissue is very rare**
- **This case reports ectopic thyroid tissue in the lingual and infrahyoid areas with no thyroid tissue in the normal anatomical location**
- **This was confirmed by technetium-99m scanning**
- **The infrahyoid ectopic thyroid was surgically removed for cosmetic reasons**
- **The importance of thyroid scanning in cases of anterior midline neck swellings is stressed**

The management of patients with ectopic thyroid tissue is dependent upon several factors, including the size of the lesion, the presence of local symptoms, the patient's age, the presence of the thyroid gland, and the presence of complicating factors such as cosmetic disturbance, ulceration, haemorrhage, cystic degeneration, or malignancy.⁹ Observation is all that is needed for asymptomatic patients. Suppressive treatment with thyroxine can be used to reduce the size of the gland in patients with pressure symptoms or cosmetic problems. The major drawback of this treatment method is that the reduction in the size of the ectopic tissue may be rather slow.⁹ Ablation of the ectopic thyroid tissue with radioactive iodine is an alternative approach. Radioactive iodine is reserved for older symptomatic patients, in whom surgery is not considered appropriate, but it is contra-indicated in children and young adults for whom the systemic doses required have potentially damaging effects on the gonads and other organs.⁴ There is also the possibility of malignant transformation in normal functioning thyroid tissue.⁸ Surgical removal of the mass can be performed in cases with marked swelling causing pressure symptoms or cosmetic problems, as well as thyrotoxicosis, and in cases of malignancy in the ectopic gland.⁷ When surgical or radioactive iodine treatment is chosen as the preferred method for an apparent ectopic thyroid, the possibility that it may be the only functioning thyroid tissue must be considered. In such cases, hypothyroidism inevitably ensues; hormone supplementation or transplantation of the excised thyroid tissue into the cervical, thoracic, and abdominal muscle layer have been attempted as therapeutic modalities.^{4,8} In our case, our chosen treatment method for the solution of the cosmetic problem of our patient was surgical excision, due to the fact that the patient's response to thyroxine therapy was insufficient. The lingual ectopic thyroid was left untouched for two reasons: The first is that

it was asymptomatic and the second reason is that it would be the only thyroid tissue left after surgery. Since the surgical procedure, the patient has been on a low dose of thyroxine therapy, both to suppress the ectopic lingual tissue from increasing in size and to uphold the insufficient amount of thyroid hormones produced by the ectopic lingual thyroid.

In conclusion, although rare, ectopic thyroid must be kept in mind in the management of midline neck masses. Thyroid scanning should be performed in all these cases before treatment to avoid permanent hypothyroidism. The treatment approach should be tailored according to the patient's age, the status of the thyroid gland, and the presence of complicating factors such as ulceration, haemorrhage, cosmetic disturbance, cystic degeneration, or malignancy.

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Dr T. Ulug takes responsibility for the integrity of the content of the paper.

Competing interests: None declared
