

Dual thyroid ectopia

Mohammed H. Al-Akeely, MBBS, ABS.

ABSTRACT

Ectopic thyroid gland is a rare embryological fault of thyroid development. Dual ectopic thyroid is even more rare and only cases have been reported in the literature. The author presents a case of dual ectopic thyroid in a 16-year-old boy with anterior neck mass, which is gradually growing in size particularly in the last 2 years. The initial diagnosis was thyroglossal duct cyst. Thyroid function test revealed elevated thyroid-stimulating hormone. Ultrasound of the neck did not show thyroid gland in its normal pretracheal position. Thyroid scan (Technetium 99) revealed the diagnosis of dual thyroid ectopia (lingual and subhyoid).

Saudi Med J 2003; Vol. 24 (9): 1021-10

In the third to fourth weeks of embryonic life, the thyroid gland appears as a midline diverticulum in the pharyngeal wall between the first and second branchial arches.¹ The anterior two thirds of the tongue develops from the tuberculum impar while the posterior one third develops from the hypobranchial eminence and the junction forms the future foramen caecum at which the thyroid gland develops.¹ The thyroid diverticulum becomes bilobed and descends in the neck and fuses with the 2 lateral diverticulae that are derived from the fourth pharyngeal pouch, which contributes to the parafollicular cells. As the developing thyroid advances caudally, it remains attached to pharyngeal wall by the thyroglossal duct, which usually obliterates on the sixth to eighth weeks of life. The thyroglossal duct descends anterior to the foregut, passing in front of the hyoid bone and larynx and finally localizes in the lower neck anterior to the thyroid cartilage and first few tracheal rings.¹ Ectopic thyroid occurs when there is an arrest or irregularity in such descent, the location of which decides the type of ectopia namely lingual (at the base of the tongue), sublingual (below the tongue), prelaryngeal (subhyoid) or substernal (mediastinal).² The author reports on a 16-year-old boy with dual thyroid ectopia (lingual and subhyoid) and reviews the literature.

Case Report. A 16-year-old boy was admitted electively with a 3 x 3 cm firm swelling in the subhyoid region, which was present for 6 years but has grown larger in the past 2 years (**Figure 1**). He had no respiratory symptoms (dyspnea, dysphagia, dysphonia). His thyroid function test revealed normal triiodothyronine (T₃) and thyroxine (T₄) but elevated thyroid-stimulating hormone (TSH) 9 mmol (NR 0.27-4.2 mmol). Ultrasound of the neck revealed absence of a normally located thyroid gland and a solid subhyoid mass. The fine needle aspiration of this subhyoid mass revealed colloid goiter. The technetium 99 thyroid scan showed absent thyroid gland from its normal pretracheal location, uptake in the subhyoid mass, lingual mass, all suggestive of dual thyroid ectopia (**Figure 2**). The patient was treated conservatively with Eltroxin 100 µg daily. He was followed up with clinical examination and thyroid function tests. Six months later, the swelling was smaller (1x1 cm), T₃, T₄ were normal and TSH had returned to normal (3.5mmol).

Discussion. The prevalence of ectopic thyroid gland 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.³ The male to female ratio

From the Department of Surgery, College of Medicine, Riyadh Medical Complex, Riyadh, Kingdom of Saudi Arabia.

Received 30th April 2003. Accepted for publication in final form 16th June 2003.

Address correspondence and reprint request to: Dr. Mohammed H. Al-Akeely, Consultant Surgeon, Department of Surgery (37), College of Medicine, PO Box 2925, Riyadh 11461, Kingdom of Saudi Arabia. Tel. +966 (1) 4671575/55485810. Fax. +966 (1) 4116772.

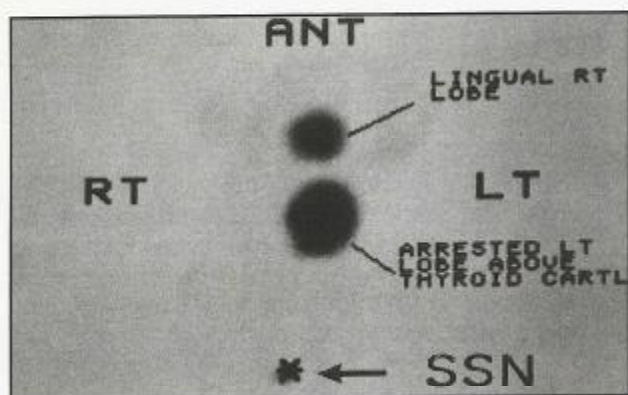


Figure 1 - Thyroid scan (tech99) showing the ectopic lingual (upper), subhyoid (lower) and absent normally located thyroid. SSN - suprasternal notch, ANT - anterior, RT - right thyroid, LT - left thyroid



Figure 2 - Subhyoid thyroid. The pen is pointing to the thyroid cartilage

approximately 1:4.3 Since Hickman⁴ described the first case of ectopic thyroid gland in 1869, sporadic case reports of thyroid ectopia continued to appear in literature. Ectopic thyroid may present at any age but commonly appear at periods of physiological stress like pregnancy and puberty. These usually present with an enlarging anterior neck mass with or without pressure symptoms of dysphagia, dyspnea and dysphonia. Such enlargement is due to hyperplasia of the ectopic thyroid tissue owing to increased stimulation by the elevated TSH in response to increased body demand of thyroxin in physiological stress. Ectopic thyroid is rare and is often mistaken for thyroglossal duct cyst as in the present case. In addition to thyroglossal duct cyst, the other differential diagnosis of masses in the anterior neck and base of the tongue that may be confused with ectopic thyroid includes lymphadenopathy, epidermoid cyst, lipoma, sebaceous cyst, lymphangioma and rarely branchial cyst. Among the different types of thyroid ectopia, the sublingual ectopia is the most common, accounting for 90% of the cases followed by the high cervical (subhyoid), and in 70% of them it is the only thyroid tissue in the body.⁵ The first sublingual ectopic thyroid was reported by Hickman⁴ in a newborn who died of suffocation due to laryngeal pressure. The subhyoid ectopic thyroid glands on the other hand, were diagnosed in the past after excision of what clinically diagnosed as thyroglossal duct cysts.⁶ In patients who presents with anterior neck or base of tongue mass the presence of normally, located thyroid should be confirmed by physical examination and radiological investigations. Pinczower et al⁷ suggested that physical examination of pediatric normal thyroid gland is unreliable. If the normal thyroid cannot be palpated or if ectopic thyroid is suspected then ultrasonography (U/S), computed tomography (CT) or radionucleotide scanning of the neck and oropharynx should be considered. Both U/S and CT are non-invasive and appropriate initial investigations, which may help to distinguish solid and cystic masses and delineate their relations to adjacent

structures. Radionucleotide study however, is indicated if the lesions are solid and an ectopic thyroid suspected or if a normally placed thyroid cannot be located. This is important in determining the sites, locations and activity of ectopic thyroid. The use of technetium 99 is preferable to iodine 131 owing to lower radiation dose. Jennifer et al⁸ studied 30 children with proven thyroglossal duct cysts and found that U/S is reliable in confirming the presence of normally placed thyroid gland in patients with thyroglossal duct cyst and therefore, thyroid scan is not necessary in the routine evaluation of patients with thyroglossal duct cyst. Hypothyroidism has been reported in up to 33% of patients of ectopic thyroid gland. The thyroid hormone levels are normal in the chemical structure but at a lower level than normal, which may result in subclinical hypothyroidism which becomes manifest clinically during periods of stress for example, puberty with subsequent development of goiter in patients with ectopic thyroid.⁹

A Medline review of literature to February 2003 revealed 8 cases of dual thyroid ectopic. Hazarika et al¹⁰ reported 2 cases of dual thyroid ectopy, one in a 32-year-old man with a lingual and subhyoid swelling and the other in 18-year-old man with sublingual and subhyoid swellings. Misaki et al¹¹ reported a mother whose son had lingual and subhyoid ectopic thyroid. Rosen and Walfish¹² described a 12-year-old boy with lingual and subhyoid ectopia. Kumar et al¹³ reported lingual and subhyoid ectopia in 14-year-old boy. Ba et al¹⁴ has recently reported a lingual and subhyoid ectopy in a 15-year-old girl. In all these 7 cases, there were no normal thyroids in the pretracheal location. However, Kuehn et al¹⁵ has reported the only dual thyroid ectopia (lingual and subhyoid) in a patient who had normally placed thyroid gland. The pretracheal and the subhyoid glands were resected for thyrotoxicosis which recurred after surgery, and thyroid scan subsequently revealed a thyrotoxic lingual ectopic thyroid as well. In asymptomatic and euthyroid children

with ectopic thyroid gland, no treatment is required and long-term follow up is necessary. However, thyroxin supplement is indicated in patients with hypoactive ectopic thyroid gland, symptomatic goiters or for cosmetic reasons. Kansal et al¹⁶ has suggested that all patients with ectopic thyroid glands should have lifelong suppression doses of thyroxin to prevent hypothyroidism and hence goiter formation. Hyperthyroidism has rarely been reported in association with thyroxin prophylaxis.¹⁶ Surgical excision of ectopic thyroid is seldom necessary, but has to be considered in life threatening airway obstruction, malignancy and thyrotoxicosis. Cosmetic desires are also an indication for surgery particularly if a period of hormone treatment has failed.

In conclusion, ectopic thyroid gland should be ruled out before surgical excision of any midline anterior neck mass. The possibility of dual thyroid ectopia has to be kept in mind when a thyroid ectopia is encountered.

Acknowledgment. I would like to thank Dr. M. K. Alam, Prof. and Consultant Surgeon, for his expert advice and opinion, and Ms. Reena Sebastian, Secretary, Postgraduate Department, Riyadh Medical Center, Riyadh, Kingdom of Saudi Arabia for typing the manuscript.

References

- Sadler TW. Langman's Medical Embryology. Baltimore (MD): Williams & Wilkins; 1990. p. 312-313.
- Krishnamurthy GT, Blahd WH. Lingual thyroid associated with Zenker's and vallecular diverticula. *Arch Otolaryngol* 1972; 96: 171-175.
- Yeung VT, Loong EP, Cockram CS. Cretinism and lingual thyroid presenting in an adult. *Postgrad Med J* 1987; 63: 881-883.
- Hickman W. Congenital tumour of the base of the tongue pressing down the epiglottis on the larynx and causing death by suffocation sixteen hours after birth. *Trans Pathol Soc Lond* 1869; 20: 160-161.
- Pintar JE, Allerand CDT. Normal development of the hypothalamic-pituitary-thyroid axis. In: Braverman LE, Utiger RD, editors. Werner and Ingbar's: The Thyroid. 6th ed. Philadelphia (PA): JB Lippincott; 1991. p. 11-14.
- Strickland AL, Macfie JA, Van Wyk JJ, French FS. Ectopic thyroid glands simulating thyroglossal duct cyst: hypothyroidism following surgical excision. *JAMA* 1969; 221: 307-310.
- Pinczower E, Crockett DM, Atkinson JB, Kun S. Preoperative thyroid scanning in presumed thyroglossal duct cyst. *Arch Otolaryngol Head Neck Surg* 1992; 118: 985-988.
- Lim-Dunham JE, Feinstein KA, Yousefzadeh DK, Ben-Ami S. Sonographic demonstration of normal thyroid gland excluding ectopic thyroid in patients with thyroglossal duct cyst. *AJNR* 1995; 164: 1489-1491.
- Leung AK. Ectopic thyroid gland and thyroxine-binding globulin excess. *Acta Paediatr Scand* 1986; 75: 872-874.
- Hazarika P, Siddiqui SA, Pujary K, Shah P, Nayak I, Balakrishnan R. Dual ectopic thyroid: A report of two cases. *Laryngol Otol* 1998; 112: 393-395.
- Misaki T, Koh T, Shimbo S, Kasagi K, Konishi J. Dual thyroid ectopy in mother and son. *Thyroid* 1992; 2: 325-327.
- Rosen RB, Walfish PG. The subhyoid ectopic median thyroid. *Can Med Assoc J* 1967; 96: 544-549.
- Kumar R, Khullar S, Gupta R, Marwah A, Dhillon MA. Dual thyroid ectopy and review of literature. *Clin Nucl Med* 2000; 25: 253-254.
- Baik SH, Choi JH, Lee HM. Dual ectopic thyroid. *Eur Arch Otorhinolaryngol* 2002; 259: 105-107.
- Kuehn PG, Newell RC, Reed JF. Exophthalmos in a woman with lingual subhyoid, and lateral lobe thyroid glands. *N Engl J Med* 1966; 274: 652-654.
- Kansal P, Sakati N, Rifai A, Woodhouse N. The lingual thyroid: diagnosis and treatment. *Arch Intern Med* 1987; 147: 2046-2048.