

CASE REPORT

Dual Ectopia with Absence of Orthotopic Thyroid Gland

Kuberan Krishnan,** Vijayalakshmi Ramanathan,* Chandrasekar,* Sivakumar Ramasamy*

ABSTRACT

Ectopic thyroid gland is a rare developmental anomaly of thyroid embryogenesis. Dual thyroid ectopia is even more rare¹. Among the dual thyroid ectopia the simultaneous occurrence of lingual thyroid and thyroglossal cyst in the absence orthotopic thyroid gland is even more rare entity. Only three cases have been reported so far. We present a case of dual thyroid ectopia in a 28 year old lady who presented with anterior neck mass, swelling in the posterior one third of tongue with subclinical hypothyroidism.

KEY WORDS

Ectopic thyroid, dual ectopia, orthotopic thyroid, lingual thyroid, thyroglossal cyst.

INTRODUCTION

During third week of intra uterine life thyroid develops as a median thyroid diverticulum in the pharyngeal wall between the first and second pharyngeal pouch. This thyroid diverticulum fuses with two lateral diverticulum derived from neural crest portion of the fourth pharyngeal pouch which contributes to the parafollicular C cells. This thyroid diverticulum as a thyroglossal duct descends in front of the hyoid bone to reach its final position in the neck in front of thyroid cartilage and few tracheal rings. This tract usually disappear by 8th to 10th week if intrauterine life. Ectopic thyroid occurs as a result of failure to descent or persistence of the duct. Site of the arrest determines the type of ectopia namely lingual (tongue), sublingual (below the tongue), prelaryngeal (subhyoid), substernal.

CASE REPORT

A 28 year old female patient was admitted with complaints of swelling in the front of the neck for past 2 years, gradually increasing in size. On examination swelling was 3x3 cm, firm in consistency, with well defined margins which moved on deglutination and on protrusion of tongue (**Fig. 1**). Ultrasound of neck shows cystic lesion in the subhyoid region with absence of normal thyroid gland. Rigid laryngoscope showed 2x2 cm soft pink swelling in the posterior 1/3rd of the tongue. Thyroid function test showed normal T3, T4 with mildly elevated TSH level. Fine needle aspiration cytology from the subhyoid



Fig. 1 : Clinical photograph of patient with midline neck swelling

mass showed feature of colloid goitre with cystic degeneration. The technetium 99 thyroid scan showed absence of normal thyroid, uptake in the subhyoid region and lingual mass (**Fig. 2**). All of these features suggestive of dual thyroid ectopia.

Initially patient was treated with 50 mcg Levothyroxine and after 6 months of followup her TSH came to normal, but size of the swelling did not decrease. The patient was keen for surgical intervention due to cosmetic reasons. The patient was explained about the surgical procedure and after informed consent underwent Sistrunk operation with Excision of lingual thyroid (**Fig. 3**).

** Professor

* Assistant Professor / Senior Resident,
Govt. Stanley Medical College, Chennai, India.

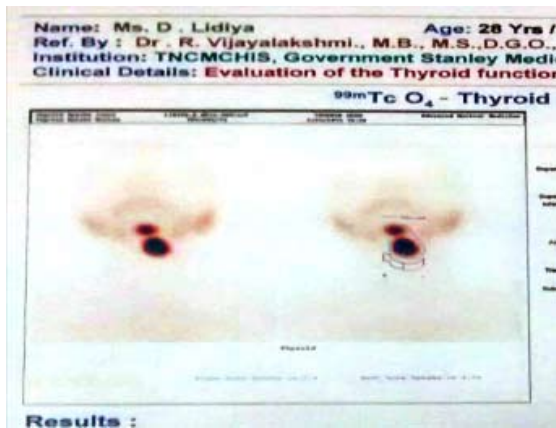


Fig. 2 : Technetium 99 scan shows uptake in the subhyoid region and in the lingual mass



Fig.3 Postoperative specimen

and an ectopic thyroid is suspected or if a normally placed thyroid cannot be located. This is important in determining the size, locations and activity of ectopic thyroid. The use of technetium 99 is preferable to iodine 131 owing to its lower radiation dose.

In asymptomatic and euthyroid patient 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

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. The male to female ratio is approximately 1:4.3. Since Hickman 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. According to its location, it can be classified as follows: infra-hyoidal (61%), supra-hyoidal (24%), supra-sternal (13%), and intra-lingual region (2%).⁵ Preoperative evaluation of TGDC includes thyroid scan, thyroid function test, US and CT. The thyroid scan plays a role in the detection of other ectopic and functioning thyroid tissue.⁴ The differential diagnosis of TGDC contains epidermoid, dermoid, lipoma and teratoma, the radiologic findings are the most distinctive points.⁶ On the other hand, it is located inside the strap muscle, but epidermoid and dermoid exist outside the muscle.⁶ The incidence of malignancy in TGDC is about 1%, mostly papillary adenocarcinoma (80%), papillary/follicular mixed (7%) and then squamous type (5%).⁷ The medullary carcinoma has not been reported.

Both ultrasound 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. Radionuclide study however, is indicated if the lesions are solid

gland, symptomatic goiters or for cosmetic reasons. Kansal *et-al*⁷ 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.

REFERENCES

1. Sadler TW. Langman's Medical Embryology. Baltimore (MD): *Williams & Wilkins*; 1990. p.312-313.
2. Krishnamurthy GT, Bland WH. Lingual thyroid associated with Zenker's and vallicular diverticula. *Arch Otolaryngol* 1972; 96:171-175.
3. Yeung VT, Loong EP, Cockram CS. Cretinism and lingual thyroid presenting in an adult. *Postgrad Med J* 1987; 63:881-883.
4. McCoul ED, Vries JD. Concurrent lingual thyroid and undescended thyroglossal duct thyroid without orthotopic thyroid gland. *Laryngoscope* 2009; 119:1937-40.
5. Plaza CPR, Lopez MED, Carrasco CEG, Meseguer LM, Perucho AF. Management of well-differentiated thyroglossal remnant thyroid carcinoma : time to close the debate? Report of five new cases and proposal of definitive algorithm for treatment. *Annals of Surgical Oncology* 2006; 13(5):745-52.
6. Huoh JC, Durr ML, Meyer AK, Rosbe KW. Comparison of imaging modalities in pediatric thyroglossal duct cysts. *Laryngoscope* 2012; 122(6):1405-8.
7. Kansal P, Sakati N, Rifai A, Woodhouse N. The lingual thyroid: diagnosis and treatment. *Arch Intern Med* 1987; 147:2046-2048.