

Lingual Thyroid: A Case Series of Six Lingual Thyroid with Incidence and Different Treatment Modalities

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ABSTRACT

Objectives: Lingual thyroid (LT) is a rare embryological anomaly and originates from failure of the thyroid gland to descend from the foramen cecum to its normal eutopic prelaryngeal site. Lingual thyroid is the most frequent ectopic location of the thyroid gland. Its clinical incidence varies between 1:4000 and 1:10000. The purpose of this case series was to discuss a series of cases of LT gland its incidence, clinical presentation, sex ratio, age group, endocrine status, radiological features and appropriate surgical approaches.

Materials and methods: Six patients of LT were diagnosed who presented to our institution in a period of 6 months out of 225 thyroid cases, out of 14539 outpatient department (OPD) patients. The LT is often asymptomatic but may cause dysphagia, dysphonia with stomatolalia, upper airway obstruction and hemorrhage, often with hypothyroidism. Treatment of a LT depends on the severity of symptoms, the age of the patient, any precipitating factors, such as puberty or pregnancy.

Result: Relevant demographic, clinic-pathological and radiological data were recorded. Four of six were treated conservatively. One underwent complete surgical excision and one pregnant lady in first trimester kept under observation and regular follow-up.

Conclusion: We found female predominance than male with ratio of 5:1. Our study had high prevalence of LT of 1:2400. Thyroid scintigraphy plays the most important role in diagnosing LT, but ultrasonography contributes as well. In cases of symptomatic disease, surgery is the treatment of choice, followed by radioiodine ablation and levothyroxine suppression therapy in more refractory cases.

Keywords: Anatomy, Genetics, Lingual thyroid, Patho, Stomatolalia, Suprahyoid approach, Thyroid scan, Ultrasonography.

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INTRODUCTION

Lingual thyroid (LT) is a rare developmental anomaly originating from aberrant embryogenesis during the passage of the thyroid gland through the neck.¹ The thyroglossal duct is a narrow tube connecting the developing thyroid gland to the tongue and it usually involutes at the 6th or 8th week. The foramen cecum is the opening of the thyroglossal duct in the tongue. The thyroid gland then descends to meet the lateral ultimobranchial bodies; the fusion of these elements leads to the formation of the functional and mature thyroid gland by the third fetal month.²

Lingual thyroid is the most frequent ectopic location of the thyroid gland, although its prevalence varies between 1:100000 and 1:300000 and its clinical incidence is reported to range from 1:4000 to 1:10000.³ Although rare, it is the most common location for ectopic functioning thyroid tissue. Occurs more commonly in females than males and is frequently associated with a lack of a normal cervical thyroid in 70% of cases.⁴

Typically there is an asymptomatic smooth surfaced lump in the midline of the base of the tongue, between sulcus terminalis and epiglottis. Occasionally, a LT may produce dysphagia, cough, pain or rarely airway obstruction.⁵ The diagnosis is made with radioisotope scanning or ultrasonography and it should be suspected whenever a mass is found in the foramen cecum area of the tongue.⁴ The treatment options for lingual thyroid include: levothyroxine suppression therapy, radioactive iodine ablation and lingual thyroidectomy.⁶

MATERIALS AND METHODS

Data were collected at ENT OPD in Dr DY Patil Medical College, Hospital and Research Centre, Pimpri, Pune, from November 2014 to May 2015. In this period of 6 months, we had 6 cases of lingual thyroid out of 14539 outpatient department (OPD) patients.

Three out of 6 patients presented with complaints of dysphagia. One case presented with stomatolalia. One case presented with blood stained sputum. To our surprise one male patient came with complaints of earache while LT was an accidental finding on throat examination, while he was asymptomatic (Table 1). The cases which we found are in age group of 10 to 35 years,

Table 1: Presentation and management of LT cases

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Gender	Female	Female	Male	Female	Female	Female
Age	12	25	30	16	19	22
Presentation	Stomatolalia	dysphagia and dyspnea	Asymptomatic (incidental finding)	Dysphagia	Blood stained sputum	Dysphagia
Endocrine status	Hypothyroidism	Hypothyroidism with amenorrhea	Hypothyroidism	Euthyroid	Hypothyroidism	Hypothyroidism
USG Neck	Absence of normal thyroid	Absence of normal thyroid	Absence of normal thyroid	Absence of normal thyroid	Absence of normal thyroid	Absence of normal thyroid
Thyroid scan	Lingual thyroid	Lingual thyroid	Lingual thyroid	Lingual thyroid	Lingual thyroid	Lingual thyroid
Treatment	Conservative	Conservative	Conservative	Surgical excision	Conservative	Conservative

in whom there is extra demand of thyroid hormone by the body which causes physiological enlargement of the gland.

In our data, we found female predominance of lingual thyroid with female to male ratio of 5:1. All females were between 10 and 35 years in whom there is extra demand of thyroxine by the body which causes it to undergo physiological enlargement (Graph 1).

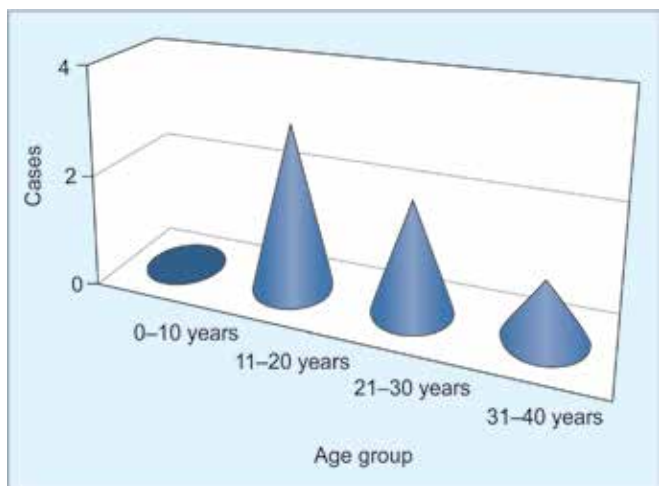
On examination, mass was visible which was single or bilobed and pinkish to red in color, ovoid in the posterior 1/3rd of tongue just posterior to the circumvallate papillae, over foramen cecum (Fig. 1). On palpation, the mass was single or bilobed, soft to firm in consistency with well-defined margins, nonmobile, nontender and did not bleed on touch. Examination of the neck revealed no palpable thyroid gland in the pretracheal position and no cervical lymphadenopathy. Patients were further investigated. Video laryngoscopy was done in all patients to look out the extent of the mass from base of tongue to vallecula. Biopsy of the mass was not done as chances of bleeding are more.

Ultrasonography of neck shows absence of thyroid gland in its normal anatomical position with the hypo-echoic mass lesion in posterior one third of the tongue suggestive of LT. Thyroid function test was suggestive of hypothyroidism in all patients. Thyroid scan showed

an area of radiotracer concentration noted in the midline, corresponding to the swelling at the base of the tongue (Fig. 2). Ectopic functioning thyroid tissue-LT. No tracer concentration was noted in the region of thyroid bed: absence of functioning thyroid gland in the thyroid bed.

Computed tomography neck was suggestive of LT. Magnetic resonance imaging (MRI) neck showed a fairly well-defined mixed signal intensity mass seen at the posterior aspect of the tongue which showed heterogeneous post contrast enhancement and narrowing of the oropharynx suggestive of LT.

A male patient of LT (Fig. 3) was asymptomatic and was in hypothyroid state, managed conservatively on tablet thyroxine and routine follow-up. Other female patient complained of blood stained sputum 5 to 6 episodes since 6 months. She was investigated and was diagnosed as LT without any respiratory cause. She was in hypothyroid state and was started on tablet thyroxine and routine follow-up. One 12 years old female child's parents complained of stomatolalia and was diagnosed LT after routine hematological and radiological investigations. She was in hypothyroid state so was started on conservative treatment of thyroxine. Two out of 5 female patient of LT complained of dysphagia and dyspnea. One was in physiological hypothyroid state and were treated conservatively. Other female complained of



Graph 1: Incidence of lingual thyroid in age group



Fig. 1: Lingual thyroid on videolaryngoscopy

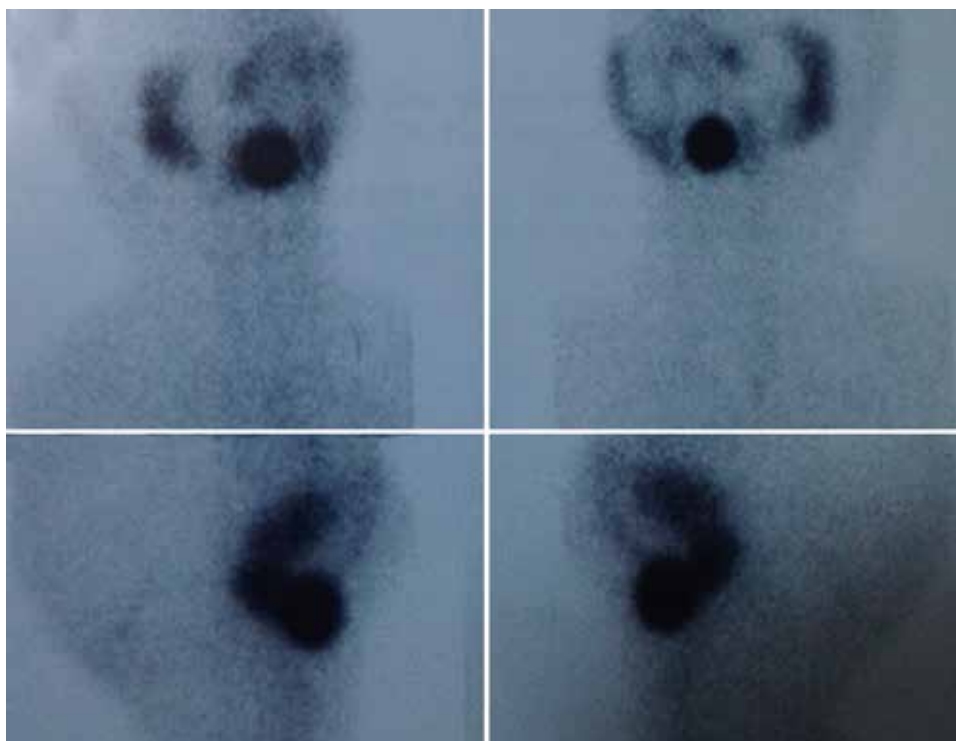


Fig. 2: Radioactive iodine uptake and scan test for lingual thyroid

2 months of amenorrhea. She was advised by endocrinologist to undergo surgery in second trimester and was given tablet thyroxine 25 µg and was kept under observation.

If the LT is the only functioning thyroid, suppression therapy using regular oral doses of thyroxine can be attempted. This is more so in patients whose normal physiological requirement of thyroxine is raised as during periods of active growth, menarche, pregnancy, etc. This suppression therapy will help in preventing abnormal physiological enlargement of the ectopic thyroid tissue.

A 16 years old female patient complained of dysphagia. She was already diagnosed of hypothyroidism and was on treatment for the same for 1 year. Her repeat thyroid function test was euthyroid state. She was

investigated further. Her video laryngoscopy showed mass over posterior 1/3rd of tongue and extending till median aryepiglottic fold. On palpation, there was absence of normal thyroid tissue in its normal position. Further hematological and radiological investigations were done which all suggestive of LT.

Suprahyoid approach for this patient was decided. Preliminary tracheostomy was performed under local anesthesia. General anesthesia is introduced via tracheostome. This protects and takes control of the airway in an efficient manner. Transverse skin crease incision is made at the level of hyoid bone. Skin, subcutaneous tissue and cervical fascia are elevated in the subplatysmal plane (Fig. 4) Dissection in this plane is continued and the flap



Fig. 3: Videolaryngoscopy showing lingual thyroid



Fig. 4: Intraoperative picture of suprahyoid approach for lingual thyroid excision



Fig. 5: Excised lingual thyroid specimen sent for HPE

is raised above the level of hyoid bone. In this stage, the muscles attached to the hyoid bone are cut and dissected subperiosteally. The supra hyoid muscles are split and the oral cavity is entered. Using a finger guide inside the oral cavity the mass is pushed downwards and delivered via the suprahyoid neck incision. The mass is removed *in toto* (Fig. 5) and was sent for histopathological examination. The wound was meticulously closed in layers. Ryles tube feeding was started. After surgery, the patient was started on oral supplemental doses of thyroxine.

DISCUSSION

Hickmann recorded the first case of LT in 1869. Montgomery stressed that for a condition to be branded as LT, thyroid follicles should be demonstrated histopathologically in tissues sampled from the lesion.⁷ Any functioning thyroid tissue found outside the normal thyroid location is termed ectopic thyroid tissue.⁸

Ectopic thyroid tissue can also occur between the geniohyoid and mylohyoid muscles (sublingual thyroid), above the hyoid bone (prelaryngeal thyroid) and in other rare sites, such as the mediastinum, precardial sac, heart, breast, pharynx, esophagus, trachea, lung, duodenum and mesentery of the small intestine, adrenal gland.⁹⁻¹³

Lingual thyroid is the result of failure of descent of the thyroid anlage from the foramen cecum of the tongue. The reasons for the failure of descent are unknown.¹⁴ Genetic research has shown that the gene transcription factors TITF-1 (Nkx2-1), Foxe1 (TITF-2) and PAX-8 are essential for thyroid morphogenesis and differentiation. Mutation in these genes may be involved in abnormal migration of the thyroid.¹⁵ Lingual thyroid is found in 10% of general population. Not all LT tissue is functional and function tends to decline with age.⁵ The majority of patients with ectopic thyroid are asymptomatic, while some cases are detected incidentally. Symptoms are usually related to size and location of the ectopic

gland as well as endocrine dysfunction. Clinical presentation are varied, most of them related to oropharyngeal obstruction, and may include dysphagia (mild or severe), dyspnea and dysphonia, fullness in throat, stomatolalia, bleeding, sleep apnea. Stridor is most common in neonates. About 33% of the patients show hypothyroidism findings.¹⁴ Lingual thyroid usually presents itself as a midline, nodular mass in the base of tongue.¹⁶ Palpation of the neck is extremely essential, in order to check the presence or the absence of the thyroid gland in its normal position.

Ectopic thyroid may become goitrous and may also be associated with clinically evident thyroid dysfunction, which could be either hypofunction or hyperfunction.¹⁷ Rarely, benign and malignant neoplastic changes can occur in ectopic thyroid tissue. Thyroiditis occurring in ectopic thyroid tissue has also been reported.¹⁸ Ectopic thyroid is commonly detected during periods of increased demand for thyroid hormones, e.g. puberty and pregnancy. Increased levels of thyrotropin at these periods causes enlargement of the ectopic thyroid tissue, thereby making it clinically detectable as a mass, or following pressure symptoms. During pregnancy, thyroid gland size increases by an average of 30% in borderline iodine deficient regions.¹⁹ It has been speculated that epidermal growth factor could also stimulate thyroid growth (n). Ectopic thyroid is the most common cause of congenital hypothyroidism in infants. In addition hypothyroidism occurs in about 33% of patients with thyroid ectopy.²⁰ While some amount of normal thyroid hormone is secreted by the ectopic gland, this may not be sufficient for higher physiological demands during puberty, pregnancy, infections and trauma.²¹

Frequency of carcinoma in LT is estimated to be approximately 1 in 100 cases with a female to male ratio ranging from 3:1 to 8:1. Most tumors are papillary carcinomas. However, follicular, mixed follicular, and papillary Hurthle cell and medullary carcinomas have also been described.²²

Ectopic thyroid glands in infants and young children who suffer from failure to thrive and mental retardation is often detected during routine screening and work-up for hypothyroidism.²³

Diagnosis depends on finding thyroid tissue at the base of the tongue with the absence of normally located gland. Only occasionally do patients have thyroid tissue both at the tongue base and elsewhere in the neck. Imaging studies as ultrasound scan, CT scan and technetium-99 m (Tc99m) thyroid scan would be of great value establishing the diagnosis.⁸ Ultrasound is only use in demonstrating absent thyroid tissue in the normal location, which is the case in the majority of cases.²⁴ The sensitivity of detection of an ectopic thyroid is enhanced by the use of color

Doppler technique by demonstrating peripheral or internal color flow signals that are reflective of hyper-vascularity.²⁵ Radionuclide thyroid imaging employing Tc99m pertechnetate, iodine-131 or iodine 123 is useful in the evaluation for ectopic thyroid. Thyroid tissue takes up the radioisotope and this helps in localizing the ectopic thyroid and at the same time in determining the presence of ectopic thyroid gland. This crucial to know before surgical removal of the ectopic tissue since in more than half of patients with thyroid ectopy, no other functioning thyroid tissue exists.²⁶

Chest radiography may also be useful in revealing some cases of intrathoracic goiter.²² Computed tomography and MRI are valuable tools in identifying the site of ectopy, especially when it is distant from the descending pathway of thyroid. Magnetic resonance imaging is particularly useful in LT when there is difficulty in differentiating thyroid tissue from tongue muscle.²⁷

Asymptomatic euthyroid patients with ectopic thyroid do not usually require therapy but are kept under observation. In patients with lingual, sublingual, lateral neck or intratracheal thyroid with hypothyroidism, suppressive therapy is administered using exogenous thyroid hormone. This suppresses the TSH level and causes reduction in the size of the gland. Euthyroid patients with mild obstructive symptoms can also benefit from suppressive therapy.²³ Surgical intervention is indicated when severe obstructive symptoms, bleeding, ulceration, cystic degeneration and malignancy occur.²⁸ Levothyroxine therapy should be initiated after surgical excision as the LT is the only functioning thyroid tissue found in 70% of these patients.¹⁶

Excision of a LT is usually carried out by transoral route. It is best for small lesions because of the limited exposure it provides. In bigger lesions, exposure can be improved by adopting the midline mandibulectomy and tongue splitting technique. Large swellings are often removed surgically through lateral pharyngectomy, suprahyoid or transhyoid approaches. The transposition technique of thyroid tissue into the anterior rectus sheath or under the strap muscles enables satisfactory long-term postoperative thyroid function, while 70% of patients will require exogenous thyroid hormone replacement following autotransplantation.²³ Parathyroids, will be in their normal position, i.e. neck because embryologically their developmental process is different.

Radioactive iodine 131 therapy is an alternative to surgical ablation. It is indicated in patients who are not fit for surgery, in those who refused operation and where surgical resection is not feasible due to anatomical difficulties.²³ It is contraindicated in pregnant women and avoided in younger pediatric patients. Other disad-

vantages include slow response to medication, fibrosis, radiation induced tracheitis and dependence on life-long thyroid hormone replacement.^{23,29}

CONCLUSION

Ectopic thyroid remains a rare developmental anomaly. The majority are asymptomatic. Symptoms may arise following enlargement of the gland during periods of stress. Patients may present insidiously or as an emergency. Dysphagia and dysphonia are common presenting symptoms. Thorough head and neck examination with special attention to base of tongue is essential. Radionuclide thyroid imaging, ultrasonography, CT scan, MRI, biopsy and thyroid function tests are diagnostic tools. Treatment could be conservative with substitutive levothyroxine treatment in patients with mild symptoms, while transoral surgery is recommended in cases with airway obstruction. Transoral approach provides good exposure and is less traumatic for the patient, with better postoperative recovery.

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