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Lingual thyroid and its management

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Lingual thyroid & its management II Edition
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Lingual thyroid is a rare condition. It is seen roughly 1 in 100,000 populations. Managing this condition is filled with historical controversies ranging from leaving it alone to surgical removal of the lesion. Attempt has been made to present in a precise way the management modalities available. All the surgical modalities along with their pluses and minuses are discussed here.

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Preface

Lingual thyroid is a fairly rare ectopic thyroid presentation. Incidence ranges somewhere in the range of 1 in 100,000 normal populations. Commonly lingual thyroid happens to be the only functioning thyroid. Normal thyroid may be absent in the neck of these patients. Common dilemma in managing these patients is whether to operate lingual thyroid or not. Treatment modality invariably depends on the size of the mass. These patients commonly suffer from hypothyroidism as these ectopic tissues are not capable of maintaining normal thyroxin levels in these patients.

This book discusses etiopathogenesis of lingual thyroid with special emphasis on the various management modalities available.
About the Author

Author is a senior faculty in the department of Otolaryngology Stanley Medical College Chennai India, with rich experience in teaching and training undergraduate and post graduate students. He has also created a few online teaching resource sites for the benefit of students of otolaryngology.
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Introduction:

Lingual thyroid is a rare developmental disorder caused due to aberrant embryogenesis during the descent of thyroid gland to the neck. Lingual thyroid is defined as the presence of thyroid tissue in the midline at the tongue base. It can be present anywhere between circumvallate papillae and epiglottis. Lingual thyroid is the most frequent ectopic location of thyroid gland. Prevalence rates of lingual thyroid vary from 1 in 100,000 to 1 in 300,000. In nearly 2/3 of these patients lingual thyroid happens to be the only functioning thyroid as normal thyroid tissue is absent in the neck. Review of literature reveals that only about 400 symptomatic cases have been reported so far. This could well be an understatement and statistical anomaly. This condition is 4 times more common in females than males. Even though this condition is diagnosed clinically radio nucleotide scanning is usually confirmatory in nature. Nearly a third of these patients are hypothyroid since the only functioning lingual thyroid cannot cope up with the normal body demands of thyroxin. These patients need to be identified and supplementation should be started at the earliest. Any delay in thyroxin supplementation could lead to enlargement of lingual thyroid tissue causing problems due to mass effect like dysphagia, bleeding from mouth etc.

Literature suggests that ectopic thyroid tissue can occur anywhere, but commonly it is found in midline of neck in the region of hyoid bone, trachea, oesophagus and rarely even in porta hepatis. Histologically majority of lingual thyroid glands demonstrate normal thyroid tissue. Lingual thyroid can be non-encapsulated and could contain embryonic / mature thyroid follicles. This tissue could extend between the lingual muscle fibers.

In his classic and extensive study Turot reported that lingual thyroid was present commonly in patients with abnormal thyroid function. He put the figure to be around 1 in 500.

Common locations of ectopic thyroid gland include:

1. Between geniohyoid and mylohyoid muscles (sublingual thyroid)
2. Above the hyoid bone (suprahyoid prelaryngeal)
3. Mediastimum
4. Pericardial sac
5. Heart
6. Breast
7. Pharynx
8. Oesophagus
9. Trachea
10. Lung
11. Duodenum

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12. Mesentery of small intestine
13. Adrenal gland

History:
Hippocrates stressed the importance of examination of tongue in diagnosis of diseases. It was Hunt in 1866 first recorded a tumor in the posterior third of tongue.
Hickmann recorded the first case of lingual thyroid in 1869. He reported the death of a 16 hour infant due to asphyxiation caused by goitre at the base of tongue.
Montgomery stressed that for a condition to be branded as lingual thyroid, thyroid follicles should be demonstrated histopathologically in tissues sampled from the lesion. Dore in 1922 collected and analysed 130 cases of lingual thyroid and concluded that majority of them are the only functioning thyroid tissue. He also reported that bleeding from large vessels present over lingual thyroid could be the cause for torrential bleed encountered in these patients.
Bishop practised simple snare removal of lingual thyroid. He suggested snaring lingual thyroid reduced incidence of bleeding. Lahey first classified the various positions the ectopic thyroid tissue could assume.
Montgomery popularised medical management of lingual thyroid by administering Lugol’s iodine to these patients. He was able to demonstrate significant reduction in the size of the mass.
Wapshaw popularized extra oral approach to remove large lingual thyroid masses. It was Thompson who first used diathermy in removing lingual thyroid mass. He was able to demonstrate significant reduction of bleeding during this procedure. Goetsch condemned the use of cautery / radiation in managing lingual thyroid masses.
Lemon and Paschal stressed the importance of neck exploration to ensure normal thyroid tissue is present before proceeding to surgically extirpate lingual thyroid mass. They were also the first to lay down definite indications for surgical removal of lingual thyroid. Lemon after extensive studies concluded that the size of the lingual thyroid mass is an indication for surgical removal of the mass. Other indications suggested by them include:

1. Dysphagia
2. Dysphasia
3. Bleeding from lingual thyroid mass
4. Malignant transformation

Ray and Wapshaw were the first to attempt transplantation of excised lingual thyroid tissue beneath the rectus muscle. Even though their attempt failed, the concept caught the imagination of others who followed them.
Feitelberg was the first person to use radioactive iodine in the management of lingual thyroid mass.
Embryology:
A brief discussion of embryology of thyroid gland will not be out of place as this would ensure better understanding of the pathophysiology involved in the formation of ectopic thyroid gland. Thyroid gland is the first endocrine gland to develop. Its development begins on the 24th day of embryo. Initially thyroid gland appears as proliferation of endodermal tissue in the floor of the pharynx between tuberculum impar and hypobranchial eminence (this area is the later foramen caecum). Cells of thyroid gland descend into the mesoderm above aortic sac into the hypopharyngeal eminence (later pharynx) as cords of cells. During this descent thyroid tissue retains its communication with foramen cecum. This communication is known as thyroglossal duct. This duct disappears as soon as the descent is complete. Thyroid gland descends in front of the hyoid bone and laryngeal cartilages. By 7th week it reaches its final destination in front of trachea. At this time a small median isthmus develops connecting the lobes of thyroid gland. The gland begins to function by the 3rd month when thyroid follicles start to develop. Parafollicular or c cells that secrete calcitonin are developed from ultimobranchial bodies.

Persistence of thyroglossal duct even after birth leads to the formation of thyroglossal cyst. These cysts usually arise from the remnants of thyroglossal duct and can be found anywhere along the migration site of thyroid gland. They are commonly found behind the arch of hyoid bone. Important diagnostic feature is their midline location.

Normal development and migration of thyroid gland needs an intact Tbx1-Fgf8 pathway. This pathway has been identified as the key regulator of development of human thyroid gland. Tbx1 regulates the expression of Fgf8 in the mesoderm, it is postulated that Fgf8 mediates critical Tbx1-dependent interactions between mesodermal cells and endodermal thyrocyte progenitors. Tbx1 is not expressed by thyroid primordium, but is strongly expressed by the surrounding mesoderm. It is also expressed by pharyngeal endoderm lateral to thyroid primordium. Thyroid organogenesis associated with the expression of a set of transcription factor encoding genes. They include Nkx2-1, Foxe1, Pax8 and Hhex1 genes. Expression of these genes in thyroid primordium is also dependent on Tbx1 gene expression.
Figure showing development of thyroid ventral to foramen cecum

Figure showing migration of thyroid gland
It commonly occurs in females. Female: Male ratio is 4:1. Some studies even attribute it to be 7 times more common in women. Even though lingual thyroid may manifest at any age it is commonly seen in patients in whom there is extra demand of thyroxin by the body which causes it to undergo physiological enlargement. It is commonly seen during early childhood and teens.

Symptoms:

Majority of these patients are asymptomatic. They will have no problems other than swelling in the posterior portion of their tongue. Symptoms caused by lingual thyroid include:

1. Dysphagia
2. Dysphonia
3. Bleeding from the mass
4. Sleep apnoea
5. Hypothyroidism
6. Dyspnoea (rarely)

In rare cases lingual thyroid could undergo malignant transformation.

Features seen on examination:

Clinical photograph showing lingual thyroid mass
Table showing symptoms produced by ectopic thyroid tissue according to their location

<table>
<thead>
<tr>
<th>Location</th>
<th>Symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lingual</td>
<td>Dysphagia, bleeding, dyspnoea</td>
</tr>
<tr>
<td>Suprahyoid / Infrahyoid</td>
<td>Midline neck mass</td>
</tr>
<tr>
<td>Thyroglossal duct / cyst</td>
<td>None / Midline neck mass</td>
</tr>
<tr>
<td>Pyramidal lobe</td>
<td>None</td>
</tr>
<tr>
<td>Intratracheal / Intralaryngeal</td>
<td>Stridor</td>
</tr>
<tr>
<td>Intraoesophageal</td>
<td>Dysphagia</td>
</tr>
<tr>
<td>Aortic / pericardium / cardiac</td>
<td>None</td>
</tr>
</tbody>
</table>

Lingual thyroid could be seen as pinkish mucosa covered mass over the posterior third of tongue. On palpation this mass could be felt as solid firm and fixed mass. It would be seen attached to the tongue at the junction of anterior 2/3 and posterior 1/3. This is where approximately foramen cecum is supposed to be present. Attempt should be made to palpate the neck in the region of thyroid to ascertain whether normal thyroid tissue is present in the neck.

Investigation:
Ultrasound neck:
In all patients with lingual thyroid the presence of normal thyroid in the neck should be ascertained. This can easily be done by performing ultrasound examination of neck. It will reveal the presence or absence of normal thyroid gland in the neck.

Picture showing ultrasound neck with absence of thyroid gland in the neck

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Doppler images reveal peripheral blood vessels and low resistance arterial blood flow. Trans oral ultrasound reveals mass lesion in the posterior third of tongue. It also reveals areas of tissue necrosis if present.

X-ray soft tissue neck lateral view:

This will just reveal the presence of soft tissue shadow in the region of the tongue. It will also demonstrate the lower extent of the mass.

X-ray soft tissue neck lateral view showing a globular soft tissue mass in the region of tongue above the level of hyoid bone

CT scan:

This will help in accurately assessing the extent of lesion. If contrast is used it would give valuable input regarding its vascularity. CT scan of neck will also categorically reveal the presence or absence of normal thyroid tissue in the neck.
CT scan axial cut taken at the level of lower border of mandible clearly shows soft tissue mass occupying the posterior portion of tongue.
CT scan neck axial view with contrast shows absence of thyroid gland in the neck. The internal jugular vein and carotid artery could be seen as enhancing masses. Jugular vein of one side appears to be predominantly enlarged.

Technitium 99 scan is virtually diagnostic. It will clearly reveal the radioactive isotope uptake by the thyroid tissue present on the tongue. It will also clearly demonstrate the presence or absence of thyroid tissue in the neck region. These images are obtained in either dynamic or static mode 20 minutes after intravenous injection of 74-111MBq of Technitium 99 pertechnetate. Its molecular weight is comparable to that of iodine and is transported actively into the thyroid tissue via the sodium iodide symporter system.
Figure showing Technitium 99 scan. It clearly shows increased uptake in the region of the tongue (due to lingual thyroid tissue) and absence of uptake in the neck region due to absence of normal thyroid tissue in this area. It also helps in location of ectopic glands.  

Role of radio active iodine uptake studies:

This helps in ascertaining the functional status of the thyroid gland. It also helps in ascertaining the viability of the transplanted ectopic thyroid gland 100 days after the surgical procedure.

Both I 131 and I 123 can be used for this purpose. I 123 have a favourable dosimetry for imaging. Since it is produced in a cyclotron it is rather expensive. Whereas I 131 is reactor produced and is reasonable cheap. It is also freely available. It has poor imaging characteristics and emits beta radiation. Its half life is about 8 – 10 days as compared to 12 hours of I 123. Hence I 123 is preferred for functioning radioactive imaging purposes.

Radioactive iodine is usually administered in small doses orally and uptake is measured at different intervals i.e. 2 hrs, 4 hrs, 24hrs and 48 hrs.

Estimation of serum T3 T4 and TSH levels:

This will help in assessing the functional status of the ectopic gland. Invariably
majority of these patients are euthyroid. If TSH levels are raised then suppression can be attempted using regular doses of oral thyroxine.

Management:

Conservative: If the lingual thyroid is the only functioning thyroid suppression therapy using regular oral doses of thyroxin can be attempted. This is more so in patients whose normal physiological requirement of thyroxin 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.

Surgical management:

Indications for surgery:

1. If the mass produces obstructive symptoms
2. If the mass produces bleeding
3. If the mass demonstrates sudden increase in size
4. If malignancy is suspected

FNAC is not advised as it would cause unnecessary bleeding. Similarly instead of biopsying the lesion total excision is preferred.

Methods of excision:

Transoral method of excision:

This method of excision is preferred for small lingual thyroid masses. It is ideally suited for lesions which are above the level of hyoid bone. Clinically if the posterior border of the swelling is seen on depressing the tongue with a tongue depressor then one can safely go ahead and remove the mass transorally.

Transoral removal is assisted by:

1. Cautery
2. Coablation
3. Debrider
4. Laser

Surgery is usually performed under general anesthesia induced via nasotracheal intubation. This is the preferred intubation modality in these patients as it would avoid troublesome bleeding following intubation trauma.

Patient is placed in Rose position. Boyles Davis mouth gag is used to hold the mouth
open. Throat is packed tightly using ribbon gauze to avoid spillage into larynx. The mass is held with a tenaculum forceps and is pulled anteriorly. The anterior border is incised using diathermy cautery / coblator /laser. The tumor is gently dissected and stripped away from the lingual tissue. Perfect hemostasis is secured by coagulating the bleeding points seen in the base of the tumor.

Debrider blade can be used to shave off the tumor from the tongue base. Bleeding points seen in the base can be cauterized using bipolar cautery.

Advantages of transoral approach:

1. Easy to perform
2. Neck incision is avoided
3. Patient's recovery is rapid
4. Complications are minimal

Transmandibular translingual approach:

This approach is very useful in removing very large lingual thyroid masses.

Procedure:

Preliminary tracheostomy is performed under local anesthesia. General anesthesia is introduced via tracheostome. This protects and takes control of the airway in an efficient manner. An incision over the mucoperiosteum of the buccogingival sulcus is performed over the interior region of mandible and the bone over the mental area is exposed. A midline vertical osteotomy of the mandible is performed. The tongue is sectioned sagittally in the midline up to the floor of the mouth till the tongue base is reached. The lingual thyroid mass is excised in toto. The wound is closed in layers. The mandible is immobilized by wiring and dental arch bar.
Figure showing the transmandibular approach

Figure showing wound closure

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Advantages:

1. Excellent visualization
2. No need for ligating lingual vessels
3. Important structures are spared i.e lingual nerve, hypoglossal nerve, and submandibular salivary gland

Lateral pharyngotomy approach:

This approach is preferred if transpositioning of lingual thyroid is planned. Anaesthesia is induced via nasotracheal intubation. Patient is positioned in such a way that the neck is slightly extended. An oblique curved incision is made about 8 cms long in the left lateral portion of upper neck just anterior to sternomastoid muscle. The dissection is proceeded in the subplatysmal plane. The following structures are identified:

1. Carotid bifurcation
2. Lingual artery
3. Superior thyroid artery
4. Hypoglossal nerve

Using the finger guide passing through the oral cavity to the left lateral pharynx at the level of base of tongue a lateral transverse pharyngotomy of 3-4 cms is made inferior to the hypoglossal nerve and above the hyoid bone. Through this pharyngotomy opening the posterior 1/3 of tongue, epiglottis and lingual thyroid mass could be identified. The gland is dissected out of the tongue. The right side of the mass is totally freed of the tongue. The mass is mobilised by an encircling incision over the tongue. A small attachment to the left side of tongue base is retained. This will ensure adequate vascularity to the mass after transposition. The mass is delivered via the pharyngotomy opening and is implanted in the left side of the neck with its attachment to the left tongue base remaining intact. The wound is closed in layers.

Advantage:

The most important advantage of this approach is that it ensures tension free transposition of lingual thyroid to the left side of neck. After transposition the gland can easily be examined on the left lateral neck of the patient.

Suprahyoid midline approach:

This approach is preferred in removing large lingual thyroid mass even if it extends to a level below that of hyoid bone.
Procedure:

This surgery is performed under general anesthesia administered via nasotracheal intubation. This intubation modality prevents intubation injury to lingual thyroid mass.

Infiltration:

The surgical area in the neck is liberally infiltrated using tumescent fluid.

Tumescent fluid is prepared using:

1. one litre of ringer lactate solution
2. 40 ml of 2% xylocaine
3. 1ml of 1 in 1000 adrenaline
4. 20 ml of 8.4% soda bicarb

Advantages of using tumescent fluid infiltration:

1. Breaks open tissue planes facilitating easy dissection i.e Hydro dissection
2. Reduces bleeding due to vasoconstrictive effect of adrenaline
3. Facilitates uniform heat dissipation when diathermy is used during surgical procedure
4. Prevents development of local tissue level acidosis

Figure showing infiltration being given
Incision:

Transverse skin crease incision is made at the level of hyoid bone. Skin, subcutaneous tissue and cervical fascia are elevated in the subplatysmal plane. Sticking on to the subplatysmal plane helps in preserving the cervical branches of facial nerve. Dissection in this plane is continued and the flap is raised above the level of hyoid bone.

Incision being widened using cutting diathermy
Hyoid bone visualized

Supra hyoid dissection:

In this stage the muscles attached to the hyoid bone are cut and dissected subperiosteally.

Figure showing hyoid bone being skeletonized using a cutting diathermy
Figure showing suprahyoid subperichondrial dissection being performed

The suprahyoid 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 full. The wound should be meticulously closed in layers. Ryle’s tube should be inserted to facilitate early feeding. Ideally the Ryles tube should be left in place at least for 3 days.
Figure showing lingual thyroid being delivered into the neck

Lingual thyroid attached to the base of tongue
After surgery all these patients should be started on oral supplemental doses of thyroxin.

If you are wondering about the status of parathyroids, you need not worry as they will be in their normal position i.e. neck because embryologically their developmental process is different.

Radiofrequency ablation: This has been successfully used in managing lingual thyroid masses. Bleeding is minimal and complete removal is possible with minimal morbidity and tissue damage. Lingual oedema is also minimal in these patients.
References:
