

East African Medical Journal Vol. 87 No. 7 July 2010

LINGUAL THYROID: CASE REEPORT

R. J. Sonigra, MBBS (New Delhi), MMed (ENT Surg.) Consultant ENT Surgeon, Kenyatta National Hospital, P.O. Box 19303-00202, Nairobi, Kenya and B. A. Ongulo, MBChB, MMed (ENT Surg.), Consultant ENT Surgeon, Armed Forces Memorial Hospital, P.O. Box 13419-00100, Nairobi, Kenya

Request for reprints to: Dr. R. J. Sonigra, Kenyatta National Hospital, P.O. Box 19303-00202, Nairobi, Kenya

LINGUAL THYROID: CASE REPORT

R. J. SONIGRA and B. A. ONGULO

SUMMARY

A sixteen year old male presented with progressive dysphagia, dysphonia and haemoptysis over eight months. Radionuclide studies and computed tomographic scans confirmed an only functional thyroid gland at the base of tongue which was excised wholly via mandibular split transoral route and patient put on thyroxin replacement therapy.

INTRODUCTION

The thyroid gland develops quite early during embryogenesis starting from approximately 24 days of foetal life from the median endodermal thickening in the floor of the primordial pharynx and descends from the foramen caecum passing ventral to the developing hyoid bone to be located anteriorly to the developing second and third tracheal rings (1). When not located here, the thyroid gland is characterised as ectopic (1-2). Ectopic thyroid tissue is an uncommon congenital aberration which occurs in approximately 1 in 100,000 people and there is a marked female preponderance (2). A lingual thyroid gland results from failure of the thyroglossal duct to migrate from foramen caecum to its final prelaryngeal position during foetal life.

CASE REPORT

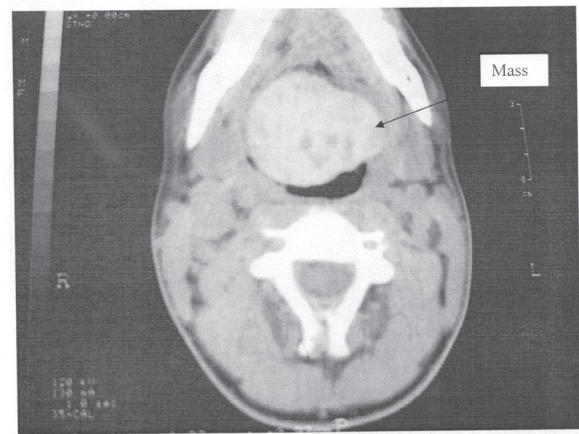
A sixteen year old male presented with eight month history of dysphagia, dysphonia, haemoptysis and what was said to be recurrent sore throat not responding to antibiotics. Lately he had started having obstructive sleep apnoea. There was nothing significant in his past medical history.

On examination he had low weight, muffled speech and in the oral cavity there was a smooth pink mass in the posterior third of tongue with a small surface ulceration. There were no signs of hypo or hyperthyroidism and no palpable thyroid in the neck. Thyroid hormones were within normal ranges and technetium (^{99m}Tc) scanning showed a large area of increased uptake of 6.5 % (normal range 0.3 to 1.5%)

in the region of the midline base of the tongue but no radiotracer uptake in the normal region of the thyroid gland. A computerised (CT) scan showed a well delineated hyperdense mass in the region of the base of tongue which was encroaching on the aero digestive track (Figure 1).

Figure 1

Hyperdense mass at the base of tongue encroaching on the aerodigestive track



The patient was prescribed thyroid hormone to suppress hypertrophy of the thyroid gland but he developed hyperthyroidism after one month hence therapy was stopped and surgery recommended after endocrinology review. Surgery was performed after the T4 and T3 levels had normalised and a day after embolisation of the major artery. Via mid labial and mandibular split (Figure 2) after tracheostomy and the tongue mobilised

anteriorly, a well encapsulated spherical mass (Figure 3) was removed from the posterior third of the tongue which on histology showed features of colloid goiter with chronic sub-acute inflammation. Post operation the patient did well, tracheostomy was reversed and patient discharged home one week post operation. He has been on close follow up for two years now and is currently prescribed thyroxin 150 mcg daily and has remained euthyroid with no complaints.

Figure 2

Mid labial and mandible split intra-operation

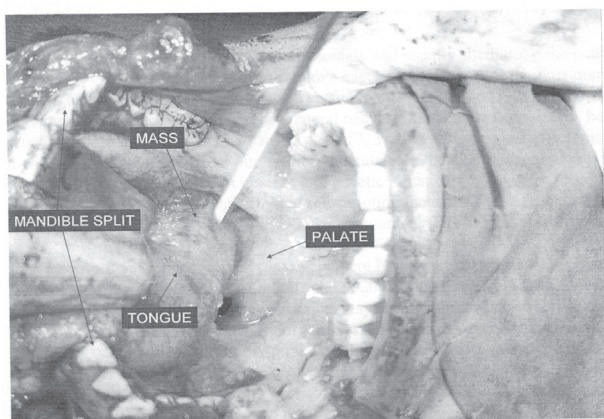


Figure 3

Spherical specimen excised. Histology showed features of colloid goiter with chronic sub-acute inflammation



DISCUSSION

Ectopic thyroid (ET) is a rare congenital anomaly and usually sited along the path of migration of the thyroid anlage from foramen caecum to the anterior mediastinum. Lingual thyroid is the most common

and accounts for over 90% of ET (1,3). Other rare sites include the mediastinum, trachea, lateral neck, submandibular area, aorta, heart, oesophagus, uterus, abdomen and parotid gland (2, 3). The pathogenesis of ectopic lingual thyroid (ELT) remains unclear but has been postulated to be due to maternal anti-thyroid immunoglobulins which impair descent and genetic mutations in the paired box transcription factor (PAX-8) and thyroid transcription factor-1 (TTF-1) required in the survival and proliferation of the follicular cell precursors. TTF-2 is necessary for the correct downward migration (3,4).

ELT has two main clinical manifestations that are based on the patient's age at onset of symptoms. One group consists of infants and young children whose lingual thyroid is detected on routine screening. These patients may suffer from failure to thrive and mental retardation (4) or even severe respiratory distress (5). The second group presents as in the case of our patient during or before puberty with a feeling of foreign body sensation and symptoms related to oropharyngeal obstruction which include, dysphagia, dysphonia, dyspnoea (4,6) and rarely haemorrhage (2). Our patient also had low weight for age. Thyroid gland hypertrophy occurs in response to the elevation in TSH levels generated by the increased metabolic demand for thyroid hormone during puberty. A similar effect is seen in other states of metabolic stress, such as pregnancy. Trauma, infection and menopause (4), this may partly explain why it is common in more females even though our patient was a male. About 10% of these patients present with hypothyroidism in spite of hyperplasia of the gland but the majority are euthyroid as in the case of our patient (6).

A thorough head and neck examination is mandatory with attention paid to visualising the oropharynx since lingual thyroid gland typically appears as a raised mass posterior to the circumvallate papillae (4). Endoscopic examination of the aerodigestive track is necessary to determine gland size and airway patency and also to inspect the larynx for other possible anomalies, such as haemangioma, vallecular cysts and hypertrophied lingual tonsils (2), however biopsy is not advisable because of possible haemorrhage. The diagnostic aids which can be used in evaluating a midline mass are radionuclide thyroid scanning, ultrasound (US) and computed tomography (CT) scanning (7). US should be done for the first screening in all cases since it is non-invasive and does not need any prior patient preparation but it is at times difficult to make a satisfactory examination especially in neonates and infants (8) moreover care should be taken to avoid the pitfalls where altered configuration of the infrahyoid muscles produce a "pseudothyroid" gland appearance in cross section

which may lead to inadvertent excision of the only functioning but ectopic thyroid gland leading to hypothyroidism (7). Thyroid scintigraphy is the most definitive non-invasive technique for establishing the diagnosis of ET and in most cases it can identify all the sites of ectopic thyroid tissue obviating the need for biopsy (9). CT scan plays an adjunctive role in defining the anatomical relationships of the gland especially when surgery is contemplated and the characteristic high attenuation due to intrinsically high iodine content prompts a diagnosis of ET (9), however, in infants or younger children CT scan has been shown to be more useful in identifying or localising thyroid tissue than US or scintigraphy (8). Magnetic resonance imaging (MRI) helps in better evaluation of the extent of the lesion and extent of oro- and hypopharyngeal involvement in a sagittal plane (4,8). Fine needle aspiration cytology may exclude malignancy and help elucidate the cause of ET glandular enlargement and inhomogeneity (9).

The management of ELT can either be medical or surgical depending on the presentation. The use of suppressive therapy with exogenous thyroid hormone is the mainstay of medical management. The goal of therapy is to suppress TSH and thereby remove the stimulus for gland enlargement. This suppressive therapy is mandatory for symptomatic patients and those asymptomatic patients with elevated TSH levels. In asymptomatic euthyroid patients it prevents the hypothyroid state that eventually develops in most cases and leads to hypertrophy of the gland. Clinical examination can then be performed and thyroid function monitored at regular intervals, ideally every three months or less. This regimen provides control of gland size and resultant symptoms in most patients. Frequent follow-up and examination may be necessary during periods of metabolic stress such as puberty or pregnancy (4, 10).

The surgical management of lingual thyroid depends on the severity of the symptoms. Surgery becomes crucial under condition such as severe or repeated haemorrhage, gland enlargement with dysphagia that prevents adequate oral intake, and significant airway compromise or dysphonia as was the case in our patient. Preoperative tracheostomy may be necessary to secure the airway. Suprahyoid, transhyoid and transoral approaches have been described to be excellent (4). We achieved good exposure via transoral route with mandibular split and managed total excision of the lingual thyroid. The use of CO₂ laser for endoscopic transoral removal of lingual thyroid is associated with less morbidity and short hospitalisation and has the advantage of being precise in cutting and haemostasis (10) but does not

achieve total excision of the gland as advocated by Ward *et al* (11). However, total removal of the gland together with a cuff of muscle may affect swallowing and speech, this may be unnecessary since the main indication for surgery is to relieve obstruction (10).

Ablation with radioactive iodine (11,13) can also be used as an alternative to surgical excision of a symptomatic gland, after which the patient should be placed on thyroid hormone replacement for life. It is rarely used because of concerns regarding radiation induced tumours since the dose required is high because the thyroid tissue is under-active and it has also been found to result in unpredictable thyroid shrinkage. It is therefore reserved for patients who are deemed unfit for surgery or who decline surgical intervention (4,10).

Euthyroid patients may be considered for auto transplantation of thyroid tissue (2). The excised thyroid gland can be placed into the anterior rectus sheath or under the strap muscles. Exogenous hormone should not be given immediately postoperatively to prevent suppression of the graft. Despite auto transplantation, 70% of patients will require exogenous thyroid hormone replacement (12).

In conclusion, ectopic lingual thyroid is a rare clinical entity which presents a challenge in management. When the presentation is due to obstruction of the aerodigestive pathway, surgical intervention is likely to be required and airway management has to be considered. Females are affected more probably because they experience more stresses which require increased thyroxin. The work up of these patients should include localisation of other thyroid tissue and in cases where there is no other thyroid tissue, long-term thyroid hormone replacement is necessary.

REFERENCES

1. Keith, L.M. and Persaud, T.V.N. Developing human-clinically oriented embryology. 7th edition. pp 215-217.
2. Williams, J.D., Sclafani, A.P. and Slupchinskij, O. Evaluation and management of lingual thyroid gland. *Ann. Otol. Rhinol. Laryngol.* 1996; **105**: 312-316.
3. Hazel, E. Bowen-Wright. and Jonklaas J. Ectopic intratracheal thyroid: An illustrative case report and literature review. *Thyroid.* 2005; **15**: 478-484.
4. Kumar, V., Nagendhar, Y., Prakash, B., *et al.* Lingual thyroid gland: Clinical evaluation and management. *Indian J. Pediatr.* 2004; **71**: 1143-1143.
5. Chan, F.L., Low, L.C., Yeung, H.W. and Saing, H. Case report: lingual thyroid, a cause of neonatal stridor. *Brit. J. Radiol.* 1993; **66**: 462-464.

6. Yang, Y.S. and Hong, K.H. Case of thyroid hemiagenesis and ectopic lingual thyroid presenting as goiter. *J. Laryngol. Otol.* 2008; **17**: 1-4.
7. Mussak, E.N. and Kacker, A. Surgical and medical management of midline ectopic thyroid. *Otolaryngol Head Neck. Surg.* 2007; **136**: 870-872.
8. Kobayashi, H., Tashita, H. and Hara, H. Utility of computed tomography in identifying an ectopic thyroid in infants and pre-school children. *Endoc. J.* 2005; **52**: 189-192.
9. Bayat, M.R., Vawda, F. and Campbell, H. Dual ectopic thyroid. *Clinical Rad.* 2005; **60**: 821-825.
10. Hafidh, M.A. Sheahan, M.B. and Khan, N.A. Role of CO₂ laser in the management of obstructive ectopic lingual thyroids. *J. Laryngol. Otol.* 2004; **118**: 807-809.
11. Ward, G.E., Cantrell, J.R. and Allan, W.B. The surgical treatment of lingual thyroid. *Ann. Surg.* 1954; **139**: 536-532.
12. Reza, R. Yoon, M.J. and Connolly, L.P. Lingual thyroid in children: A rare clinical entity. *Laryngoscope.* 2008; **118**: 1174 – 1179.