

## Lingual Thyroid: A Case Report and Literature Review

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### Abstract

Thyroid ectopia is a dysgenesis of thyroid gland and Lingual position represents the most frequent ectopic location accounting up to 90% of ectopic cases. Hypothyroidism is commonly present because of absence of a normal thyroid gland in most instances. Primary hypothyroidism in juvenile population generally leads to retardation of linear growth and delay or even arrested puberty. We present a 20 years old female with typical and profound presentation of hypothyroidism due to lingual thyroid.

**Key Words:** Ectopic thyroid, Hypothyroidism, Lingual thyroid, Thyroid dysgenesis.

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## 1- INTRODUCTION

Thyroid hormone is necessary for growth and neurologic development in childhood. The thyroid begins to take shape at 7 weeks' gestation, and thyroid hormone (T4, thyroxine) is produced starting at 12 weeks' gestation. Thyroid dysfunction in the neonate, infant, or child has a significant impact on development (1). Hypothyroidism is among the most common endocrine disorders in children, and those affected present with a short stature and delayed puberty (2). The etiology in younger children is usually congenital hypothyroidism (3). Clinical manifestations include a decline in linear growth, fatigue, constipation, cold intolerance, poor school performance, weight gain, irregular menstrual periods, and somnolence (1). Based on strong clinical evidence, congenital hypothyroidism is the most treatable preventable cause of potential intellectual disability (1, 4).

The prognosis for recovering lost linear growth depends on the duration of the hypothyroidism as well as the age at which treatment is started (1). Thyroid is the first endocrine structure to develop during fetal life. Morphogenetic errors during the development period result in the most developmental thyroid abnormalities (5). Thyroid dysgenesis accounts for 80% to 90% cases of permanent primary congenital hypothyroidism. Out of these, nearly 60% cases are due to thyroid ectopia (6). Location of ectopic thyroid tissue can be anywhere from the base of the tongue till diaphragm. Lingual thyroid is the most common (up to 80%) form of ectopia (7). Hypothyroidism is commonly present because of the absence of a normal thyroid gland in most instances (5). Clinical features vary from asymptomatic (8) to dysphagia, dysphonia (9). Diagnosis of lingual thyroid (LT) includes local examination of the tongue base associated with the absence of normally located

gland, and imaging examinations. Imaging studies consist in neck ultrasound, neck CT-scan, neck-MRI (magnetic resonance imaging) and scintigraphy using Tc 99m, I-131, I-123 (10).

## 2- CASE REPORT

A 20 year-old female was referred to pediatric department of Imam Reza hospital, Mashhad, Iran. She presented with short stature, arrested puberty (absence of thelarche and menarche), and developmental delay, otherwise, clinical history was unremarkable.

Physical examination revealed stable vital sign with temperature 37° C, Heart rate 94/min, respiratory rate 24/min, and blood pressure 100/68 mm Hg. Her weight was 21 kg (< 3th percentile, Z score: -14.589, height 100 cm (< 3th percentile, Z score: -9.842) and body mass index (BMI) 21.8 (50th percentile).

She had coarse facial features with open anterior fontanel, remnant milk teeth, short neck, coarse, brittle and scanty hair (**Figure.1a**), no palpable thyroid gland, absent axillary and pubic hair and no breast enlargement, cold extremities, and no dysmorphic features. Abdominal examination revealed hepatomegaly 5cm below the costal margin. Cardiac and chest examination was normal. Neurological examination was normal except developmental delay. Initial laboratory investigations are summarized in **Table.1**.

Thyroid ultrasound (U/S) had normal report and Abdominal U/S showed hepatomegaly. While left wrist and hand X-ray revealed delayed bone age, estimated about 2.5 years old (**Figure.1b**).

Skeletal survey X-ray showed decrease in bone density and epiphyseal dysgenesis in the proximal epiphyses of femoral bone and in the head of humerus (**Figure.1c**). Echocardiograph showed mild right ventricular hypertrophy (RVH), and Left ventricular hypertrophy (LVH), moderate

tricuspid regurgitation (TR) and pulmonary Hypertension (PH). Thyroid scan showed the thyroid gland in the posterior aspect of the tongue (**Figure.1d**). Based on physical findings, laboratory results, including high levels of thyroid stimulating hormone (TSH) with low

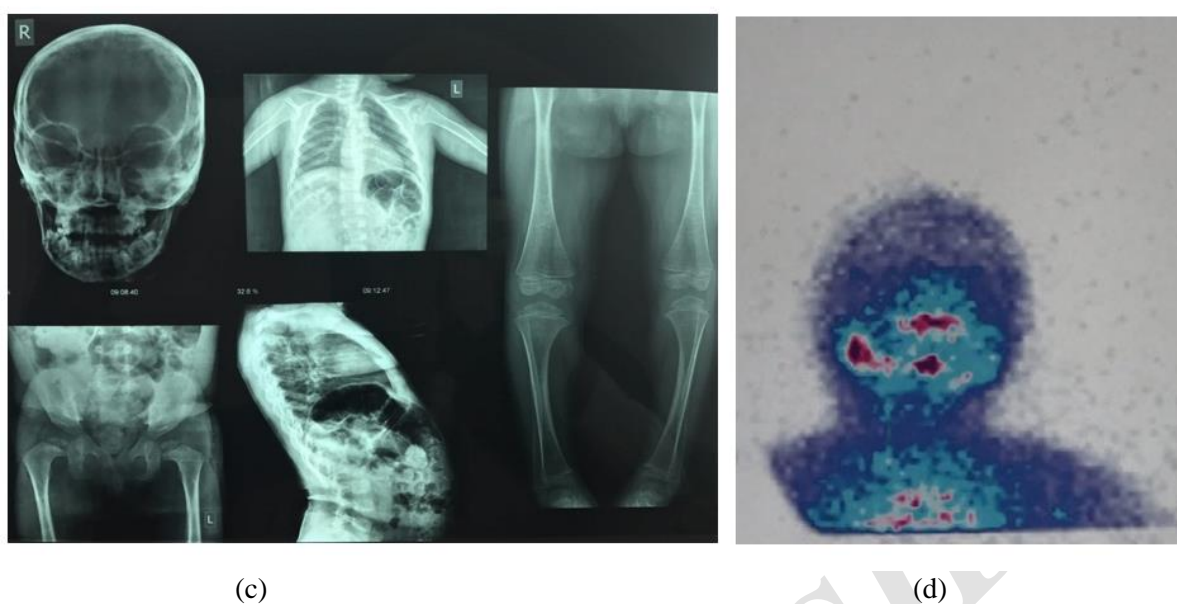
levels of T3 and T4, and radiological investigations, the diagnosis of hypothyroidism due to lingual thyroid was confirmed. Levothyroxine was started at 50 $\mu$ g/day and the patient was discharged from the hospital.



(a)



(b)



**Fig.1:** Clinical signs and imaging findings of the patient. (a) The coarse facial features. (b) Wrist X Ray and delay bone age. (c) Whole body study showing decrease in bone density and epiphyseal dysgenesis of long bones. (d) Thyroid CT-scan showing thyroid gland in posterior aspect of the tongue.

**Table-1:** Laboratory results of the patient

Parameter	Result	Normal value
Complete blood count	Normal	
Renal functions	Normal	
Thyroid stimulating hormone (TSH)	836 mU/L	0.30–5.1 mIU/L
T3	0.4 ng/mL	0.55–2 ng/mL
T4	0.2 µg/dl	5.4–12.6 µg/dl
T3 Uptake	0 %	25-38 %
25 Hydroxy Vit D3	18 ng/mL	Vit D deficiency 10-20 ng/mL Optimal vit D level 30-50 ng/mL

### 3- DISCUSSION

Traditionally, long-standing primary hypothyroidism leads to both pubertal and growth delay (11-13). Infants who born from areas with no screening program, severe cases manifest in the 1st few weeks of life, but in cases of milder deficiency, manifestations may be delayed for months (14). The prevalence of congenital hypothyroidism based on nationwide programs for neonatal screening was

initially estimated 1 in 4,000 infants worldwide (15). Over the last 2 decades, the prevalence has dropped to 1 in 2,000, likely the result of detection of milder cases of hypothyroidism (14). If congenital hypothyroidism goes undetected and untreated, Retardation of physical and mental development becomes greater during the following months (16-19). The child's growth will be stunted, the extremities are short, and the head size is normal or even increased. The anterior

fontanel is large and the posterior fontanel may remain open. Development is usually delayed (17). There are few reports of long standing primary hypothyroidism, not detected till adolescence. The first report of severe long standing primary hypothyroidism in combination with delay bone age and precocious Puberty, described as Van Wyk-Grumbach syndrome in 1960 which found in girls mostly. In 2012, a boy with Van Wyk-Grumbach syndrome was reported at 12 years old (12).

Some forms of thyroid dysgenesis (aplasia, hypoplasia, or an ectopic gland) are the most common cause of permanent congenital hypothyroidism, accounting for 80-85% of cases; the cause of thyroid dysgenesis is unknown in most cases. It occurs sporadically, but familial cases occasionally have been reported (17). Thyroid ectopia is a dysgenesis of thyroid gland (5) and a rare cause of hypothyroidism, occurring in approximately 1 in 200,000 individuals, though this may be a conservative estimate due to the often asymptomatic nature of this anomaly. Previous studies reported that 33% to 62% of all patients with ectopic thyroid showed hypothyroidism with increased levels of TSH (21-23).

Lingual thyroid is a rare developmental disorder due to the aberrant embryogenesis (10, 23), but the most common form of thyroid ectopia (7), accounting up to 90% of ectopic cases (10). Imaging techniques such as thyroid scan with Tc-99m or iodine-123 or iodine-131, CT and magnetic resonance imaging are frequently used for the exploration of a lingual thyroid, topographic diagnosis, and confirm the presence or absence of orthotopic thyroid gland. In 1869, Hickman reported the first case with lingual thyroid on a newborn baby (24). Hunt in 1865 reported a similar case of a lingual thyroid in a young woman (tumor on the base of the tongue), although not

proved (25). Ectopic lingual thyroid is commonly detected during periods of increased demand for thyroid hormones, for example adolescence and pregnancy (25). Our case came to us at the 20th year of age with severe clinical manifestation of hypothyroidism like short stature, no sign of puberty and delay bone age, like noted by Hunt or Hickman. As we found in literature, the majority of reported cases with lingual thyroid presented with local symptoms like dysphagia, dysphonia, neck swelling, stridor and respiratory obstruction reported by Bode Rovena (10) and Madhuri Patil (5), and foreign body sensation in a 12 years old girl presented by Kumar (22), or a 33 year-old man with chronic cough that reported by Grossman (26); but in our case there was nothing similar to other previous cases.

Lilley and Lomenick reported a 6 year -old girl with growth deceleration due to iatrogenic hypothyroidism after thyroglossal duct cyst excision (3). Also, a report of 10 year- old female with poor development and missed newborn screening test by Asami et al., showed ectopic thyroid with delay rise in blood TSH concentration (27); also, in a retrospective evaluation of children with primary congenital hypothyroidism by Devi et al., 18.7%, identified to have ectopic thyroid glands, and the most common presenting symptoms were consist of poor weight gain and developmental delay and lingual thyroid as the most common location (28).

Our case is different in several aspects, like the age of diagnosis (mean age of  $64\pm 47$  months) (20 years old in our patient) and the mode of diagnosis (newborn screening). We have no document from screening test or additional thyroid function assessment in our patient, because of no referral to the physician from birth to now. Diagnosis was confirmed by laboratory and technetium scan evidences. Our patient was treated for

hypothyroidism. In some reports treatment will cause decrement of the gland and resolution of symptoms (26).

#### 4- CONCLUSION

Newborn screening has a major role in detection of primary hypothyroidism. So, making easy the access to the health care facilities and education should be the first and most important goal toward proper treatment and prevention of hypothyroidism. Lingual thyroid is a rare entity that has the potential to present at any age. The combination of such complaints and disturbances in thyroid function tests should prompt the physician to search for a lingual thyroid by use of a thorough clinical examination and a radionuclide scan.

**5- CONFLICT OF INTEREST:** None.

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