# Case 9377
# Congenital hypothyroidism with ectopic lingual thyroid

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**Section:** Head & Neck Imaging
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**Patient:** 50 year(s), female

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## Clinical History

A female patient, 50 years old, non-Portuguese-speaking Eastern Europe immigrant, was referred to our institution because of a low stature (1.37 meters), with foreshortened skull base, short wide face and coarse dry skin. The laboratory analysis showed elevated TSH and low T4 levels with normal T3 levels suggesting primary hypothyroidism with residual hormonal production.

## Imaging Findings

The first examination performed was a neck ultrasound that could not manage to document the existence of the thyroid gland in its expected infrahyoid location. The rest of the neck ultrasound study was apparently unremarkable. Common localisations for ectopic thyroid tissue like the tongue, sublingual and pre-laryngeal spaces were scanned without any relevant finding (Fig. 1). Further radiological investigation of possible ectopic thyroid tissue was followed by a contrast enhanced-CT study which revealed a contrast enhancing well defined round nodule, 18 mm in diameter, lying on the base of the tongue, protruding in the pre-epiglottic space (Fig. 2-4).

Given the patient's clinical context, the nodule was considered to be ectopic thyroid tissue. The

scintigraphic study with iodine-131 was not considered essential for patient management and was not carried out in this patient.

The lesion remained stable in a follow-up CE-CT at 6 months, showing no evidence of growth.

## Discussion

Disturbed morphogenesis of the thyroid gland results in a set of conditions collectively known as thyroid dysgenesis (TD) which present as ectopically located or reduced glandular tissue or in some cases even completely absent. TD is present in 85% of congenital hypothyroidism, a condition that affects one in 3500 newborns worldwide [1].
The ectopic tissue is frequently found along a line following the migration route of the thyroid primordium from the foramen ceacum in the pharyngeal floor to its pre-tracheal final location that can be lingual, sublingual, pre-laryngeal and mediastinal. Interestingly, it has also been described in the lateral cervical area and there are reports in the submandibular region, trachea, heart, lung, duodenum, adrenal gland, parotid gland and gall bladder. The location over the dorsum of the tongue is the most frequent, around 80% of the cases [2].
The existence of ectopic thyroid tissue should always be considered in patients with suspected or proven thyroid gland dysgenesis.

Though extremely rare nowadays in developed countries due to newborn screening of hypothyroidism, cretinism is still a major epidemiological concern in several countries. It is mainly the consequence of inadequate thyroid hormone production by the newborn just after delivery [3]. In some countries it can be endemic, secondary to iodine deficiency [3].
The diagnosis should be suspected in unscreened babies usually as soon as the first month of age showing hypotonia, low or hoarse cry, infrequent bowel movements, exaggerated jaundice, low body temperature, larger anterior fontanel, persistence of a posterior fontanel, an umbilical hernia and macroglossia. If untreated, it results in severe mental impairment, low stature and the recognisable facial and body features of cretinism [4].

Most children born with congenital hypothyroidism and correctly treated with thyroxine grow and develop normally in all aspects [5].
Therapy consists mainly in hormonal replacement with oral levothyroxine (T4) with dosage defined according to the level of endogenous production to suppress the lingual thyroid and reduce its size. Only rarely is surgical excision necessary [5]. Indications for extirpation include failure of suppressive therapy to reduce the size, ulceration, haemorrhage, suspicion of malignancy and even dysphagia, dysphonia with stomatolalia or upper airway obstruction in severe cases [6].

## Final Diagnosis

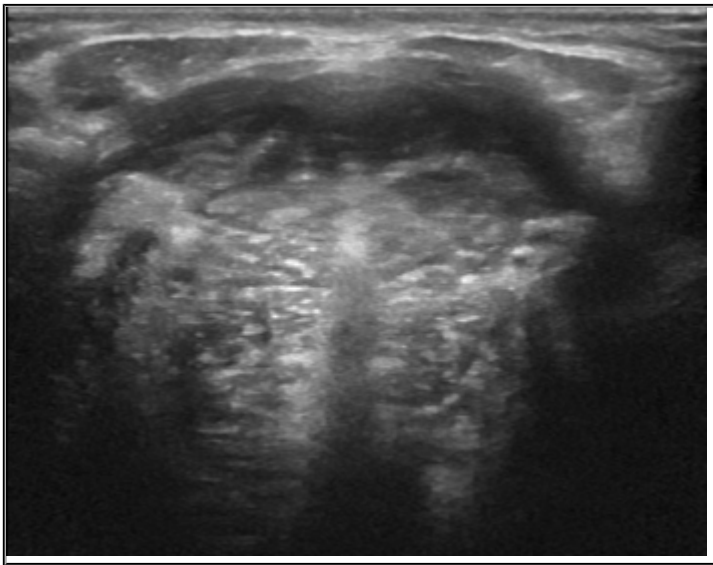
Congenital hypothyroidism with cretinism and ectopic lingual thyroid tissue

## Differential Diagnosis List

Squamous cell carcinoma, Haemangioma

## Figures

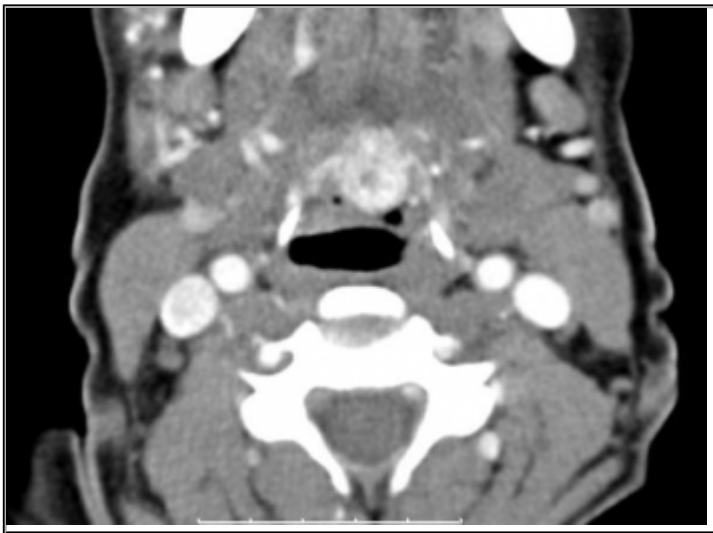
**Figure 1 Tongue**



No ectopic thyroid tissue was found in the neck during the ultrasound. An apparently normal tongue can be seen.

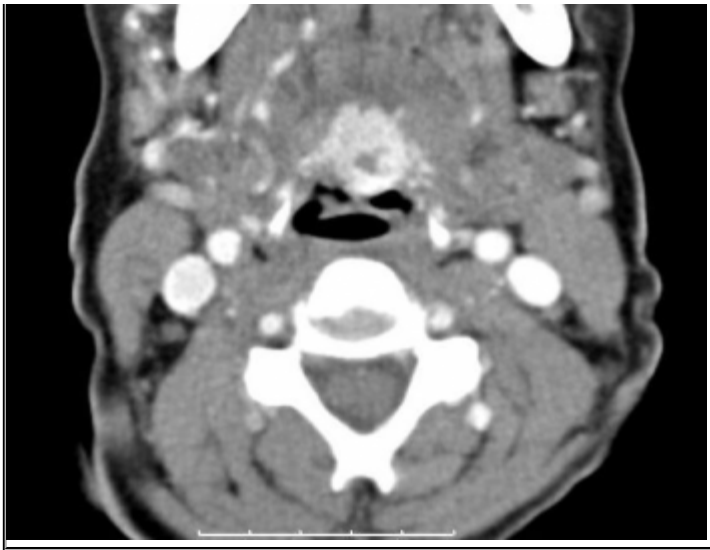
Area of Interest: Head and neck;

**Figure 2 Ectopic thyroid tissue**



CE-CT axial view: Slightly heterogeneously enhancing 2 cm nodule lying over the base of the tongue corresponding to ectopic thyroid tissue.

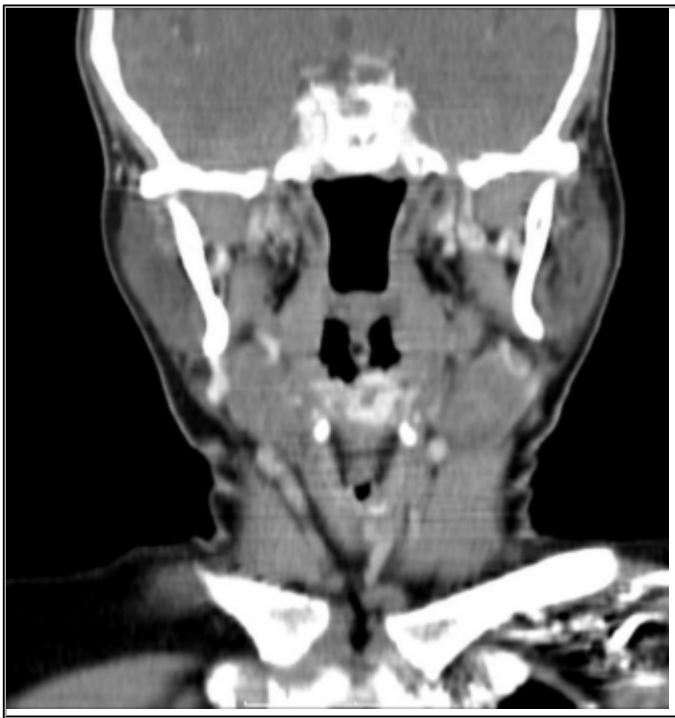
Area of Interest: Head and neck;



CE-CT axial view: Slightly heterogeneously enhancing 2 cm nodule lying over the base of the tongue corresponding to ectopic thyroid tissue.

Area of Interest: Head and neck;

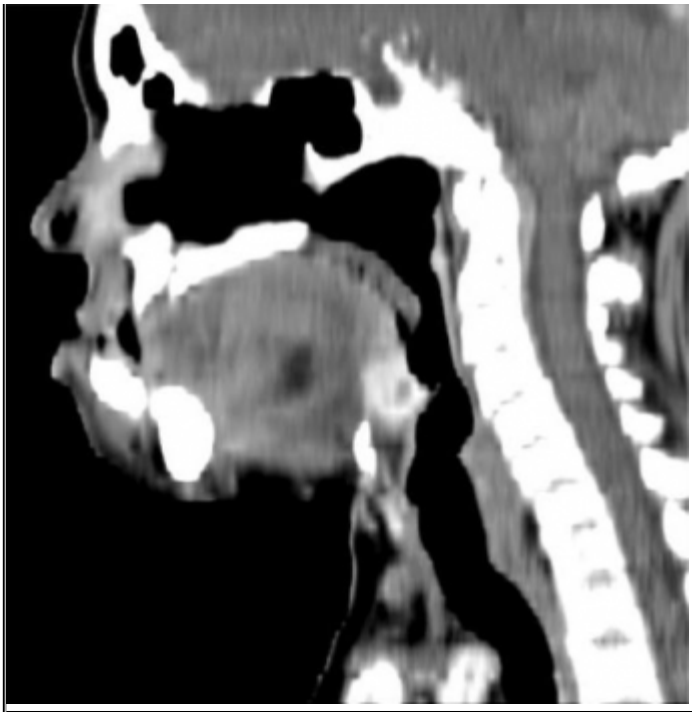
**Figure 3 Ectopic thyroid tissue**



CE-CT coronal view: This reconstruction helps to better understand the anatomic relations of the enhancing ectopic thyroid and the base of the tongue.

Area of Interest: Head and neck;

**Figure 4 Ectopic thyroid tissue**



CE-CT sagital view: The ectopic thyroid can be located between the base of the tongue and the epiglottis.

Area of Interest: Head and neck;

## MeSH

**Cretinism** [C19.874.482.281]
A condition due to congenital lack of thyroid hormone, marked by arrested physical and mental development, dystrophy of the bones and soft parts, and lowered basal metabolism. It is the congenital form of thyroid deficiency, while MYXEDEMA is the acquired form. (From Dorland, 27th ed)

**Hypothyroidism** [C19.874.482]
The clinical syndrome that results from decreased secretion of thyroid hormone from the thyroid gland. It leads to a slowing of metabolic processes and in its most severe form to the accumulation of mucopolysaccharides in the skin, causing a nonpitting edema termed myxedema. Cretinism is the congenital form leading to abnormalities of intellectual and physical development. (Bennett, et al., Cecil Textbook of Medicine, 20th ed)

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