



Dual ectopic thyroid – A case report of a very rare disease

Introduction

Ectopic thyroid is a developmental defect of thyroid gland that leads to presence of thyroid tissue at sites other than its normal pretracheal location. It is frequently found along the course of the thyroglossal duct but can also be found at remote distant sites. Lingual thyroid is the most common presentation of ectopic thyroid . It is very rare to have two ectopic foci of thyroid tissue simultaneously and only 19 cases of dual ectopia have been reported in world literature. In 70% of cases of ectopic thyroid the normal thyroid gland is absent. We report a case of 12 year old girl presented with dual ectopic thyroid.

Case report

A twelve year old girl presented with history of a swelling in front of neck in the subhyoid area first noticed about two months back. The swelling gradually increased in size. There was no history of pain, or any pressure symptoms like dyspnoea dysphagia or dysphonia. Birth history ,developmental milestones were normal and menarche was not attained. A detailed general and systemic examination did not reveal any abnormality. Local examination revealed a 2.5 * 2 cm firm swelling in the subhyoid area and 2 * 2 cm midline swelling in the posterior part of tongue. Thyroid gland was not palpable at its normal site. Thyroid function test suggested primary hypothyroidism with elevated TSH , low T3 & T4 . Fine needle aspiration revealed colloid goitre. Ultrasonogram and CECT of the neck revealed absence of normally located thyroid and presence of dual ectopic thyroid – lingual and subhyoid.

Discussion

Ectopic thyroid was first described by Hickman in 1869 in a newborn who was suffocated 16 hours after birth because of a lingual thyroid causing upper airway obstruction. Lingual thyroid is the most common ectopic thyroid accounting for 90% of all cases with a prevalence between 1: 100000 and1:300000 and a clinical incidence between 1:4000 to

1:10000. Other sites of ectopic thyroid are suprahyoid and infrahyoid, lateral aberrant thyroid, substernal goiters, struma ovarii and struma cordis. Ectopic thyroid has also been found in larynx, trachea, oesophagus, pericardium, diaphragm and branchial cysts. Rare cases of ectopic thyroid are described in parathyroid, cervical lymph nodes, submandibular gland, duodenal mesentery, adrenals and carotid bifurcation. Ectopic thyroid occurs more commonly in females and are usually seen during adolescence and pregnancy when the demand for thyroid hormone increases. Up to 70% of patients with lingual thyroid had hypothyroidism and 10% suffer from cretinism. Rest are euthyroid. However hyperthyroidism has been rarely described. Our patient presented during adolescence with hypothyroidism. The thyroid gland is not found in its usual location in 70% of patients with ectopic thyroid. Ectopic thyroid is rare and is often mistaken for thyroglossal duct cyst.

Let's go through the development of thyroid briefly; In the third to fourth weeks of embryonic life, the thyroid gland appears as a midline diverticulum in the pharyngeal wall between the first and second branchial arches. The anterior two thirds of the tongue develops from the tuberculum impar while the posterior one third develops from the hypobranchial eminence and the junction forms the future foramen caecum from which the thyroid gland develops. The thyroid diverticulum becomes bilobed and descends in the neck and fuses with the 2 lateral diverticulae that are derived from the fourth pharyngeal pouch, which contributes to the parafollicular cells. As the developing thyroid advances caudally, it remains attached to pharyngeal wall by the thyroglossal duct, which usually obliterates on the sixth - to eighth weeks of life. The thyroglossal duct descends anterior to the foregut, passing in front of the hyoid bone and larynx and finally localizes in the lower neck anterior to the thyroid cartilage and first few tracheal rings. Ectopic thyroid occurs when there is an arrest or irregularity in such descent, the location of which decides the type of ectopia namely lingual (at the base of the tongue), sublingual (below the tongue), prelaryngeal (subhyoid) or substernal (mediastinal)

Presence of two ectopic foci of thyroid tissue simultaneously is rare and very few such cases of dual thyroid ectopia has been reported in world literature. In an extensive review of literature Sood et al found that the mean age of these patients was 15 years, more common in females with a F:M ratio of 1.25:1. The symptoms varied from asymptomatic to anterior neck swelling with or without altered thyroid status. In almost all of these patients one site of ectopy was at lingual or sublingual region. The second ectopic focus was at subhyoid or suprahyoid level in most cases.

Ectopic thyroid may be asymptomatic or produce symptoms due to its location. Lingual thyroid can cause foreign body sensation in tongue and dysphagia. Large blood vessels present on the surface of lingual thyroid predisposes it for ulceration. All diseases capable of affecting the normal thyroid can affect the ectopic thyroid like adenoma, hyperplasia, inflammation and rarely malignancy. The rate of malignant transformation in ectopic thyroid is no greater than in normally placed thyroid.

A neck ultrasound examination, neck CT scan are valuable modalities for forming a differential diagnosis. US examination plays a role in differentiating cystic and solid masses. On CT scan, thyroid tissue has a characteristically high enhancement due to its great vascularity. FNAC may exclude malignancy or other pathologic changes in heterogeneous tissue. Radionucleotide thyroid scan can detect all the ectopic foci of thyroid in the body.

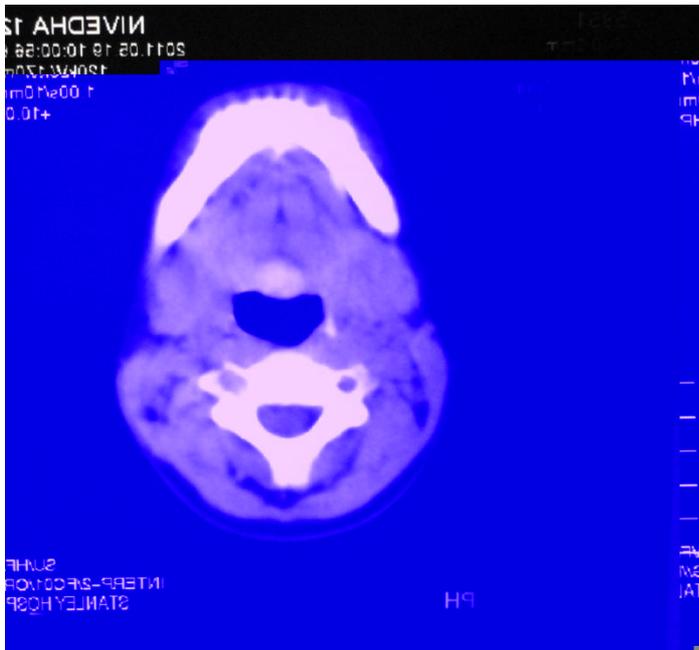
Asymptomatic and euthyroid patients do not require any treatment but they should be followed up and looked for any complications. Patients with raised TSH with swelling should be put on replacement therapy with thyroid hormone which can produce a slow reduction of the mass. When medical treatment fails or there are obstructive symptoms or haemorrhage or suspicion of malignancy then surgery should be considered. However, 70-90% of patients have no functioning thyroid tissue besides the ectopic thyroid, so extreme caution must be exercised; removal of the only functioning thyroid tissue will result in permanent hypothyroidism.

Our patient was put on 0.1 mg of levothyroxine once daily and on regular follow up for last 2 months. She attained menarche after 1 month of the thyroxine supplementation therapy.

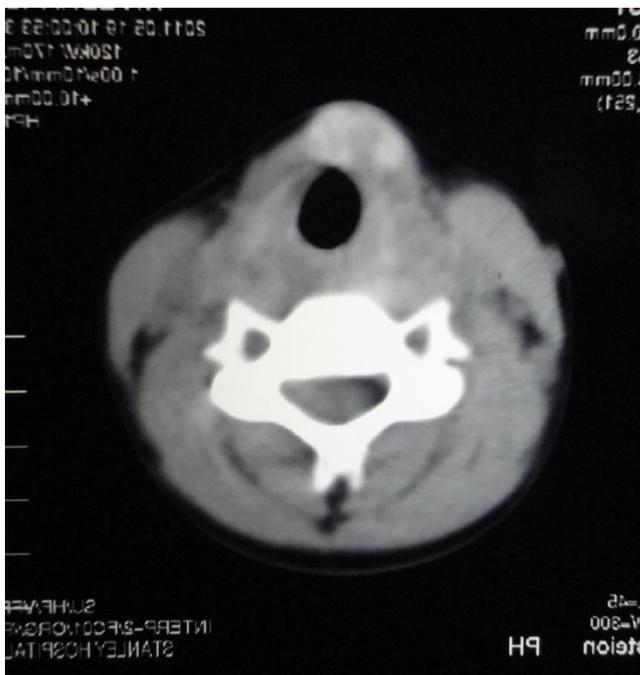
Conclusion

Ectopic thyroid should be considered as a differential diagnosis in a case of midline neck swelling since it is often misdiagnosed as thyroglossal cyst. The possibility of another ectopic focus should be kept in mind when an ectopic thyroid is encountered. Unnecessary surgery must be avoided in case of asymptomatic ectopic thyroid as it may be the only functioning thyroid in the body in most of the cases.

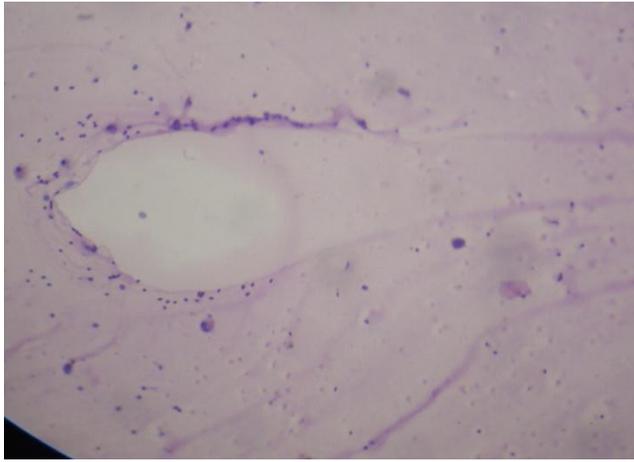




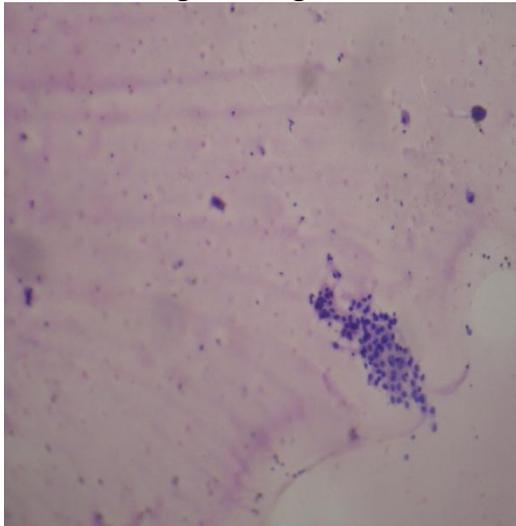
CT picture showing lingual thyroid



CT picture showing subhyoid ectopic thyroid



FNAC showing colloid goitre



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