Thyroid Hemiagenesis Presenting As Hypothyroidism – A Rare Case Report

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Abstract

Thyroid hemiagenesis is a very rare abnormality, in which one of the thyroid lobes fails to develop embryologically. Most of the patients are detected incidentally and have an associated thyroid disease. The true prevalence of the thyroid hemiagenesis is still not known but literature reports incidence in 0.02% of population. We report a rare case of hemiagenesis of left lobe of thyroid and isthmus with associated hypothyroidism in a 28 years old female with review of literature.

Keywords: Hypothyroidism, Thyroid hemiagenesis, Ultrasound

Introduction

Thyroid hemiagenesis is a rare congenital developmental abnormality in one lobe of thyroid due to failure in the normal development.¹ First described by Handsfield John in the year 1866. The thyroid gland begins to develop as a median thickening of endoderm on the floor of pharynx between first and second pharyngeal pouches. This area later invaginates to form the median diverticulum which appears later. The diverticulum later grows caudally as a bifurcating tubular duct from which the lateral lobes and isthmus develop.² The cause of unilateral agenesis is not known, but

Correspondence to: Dr. Simran Singh MBBS, Resident, Department of Radiodiagnosis, JSS Medical College, Mysore, Karnataka, India-570004 Email:- <u>dr.simrans@gmail.com</u> it is believed to be arising from the failure of the original anlage to become bilobed and spread out laterally in both halves.¹

Case Report

A 28 year old female patient presented with a history of hypothyroidism and on thyroxin 50 mcg tablets from past 10 years for regular thyroid scan to rule out any focal lesions. No previous histories of any surgery, family history of any thyroid disorder were given.

Ultrasound revealed complete absence of left lobe of thyroid and partial left sided absence of isthmus (Fig. 1) while the right lobe of thyroid was normal and measured $4.1 \times 3.5 \times$ 3.6cms (volume ~25 ml) with normal echogenicity (Fig. 2). No focal nodules, cystic changes or calcifications seen within the right lobe of thyroid.

Discussion

The congenital absence of one lobe of thyroid is rare condition of an unknown etiology. It has been postulated that it occurs due to result of failure of thyroid analage to become bilobed followed by unilateral migration.^{8,11} Most of the cases which are diagnosed showed female predominance with a ratio of 3:1and also predominance of left lobe is far more than the right lobe with a ratio of $4:1.^{10}$ The same is seen in our case where there is complete absence of left lobe along with the isthmus.



Fig. 1: Grey scale image showing complete absence of left lobe of thyroid and partial agenesis of isthmus.

The actual incidence of the hemiagenesis is unknown as most patients worldwide are admitted with a suspicion of other thyroid abnormalities like benign adenoma, multinodular goiter, hyper and hypothyroidism (as in our case) and hemiagenesis is seen as an incidental finding. ¹⁰⁻¹³

Thyroid agenesis should be suspected in all the cases where there is no palpable thyroid tissue felt on one or either side of the neck. Radiological investigations hold the key role in diagnosing this condition. A definitive diagnosis is made by ultrasound and CT, ultrasound examination can easily establish the diagnosis in suspected cases.⁵ Ultrasound examination is far cheaper, easy to perform and above all there are no radiation hazards. Thyroid scintigraphy may also be helpful in differentiating hemiagenesis from a suppressed lobe. It will show total absence of uptake in congenital hemiagenesis, but the activity will reappear in suppressed lobe.³

The lobe of the thyroid which is functioning can be site of pathological changes similarly to a normally developed gland and it may present with spectrum of diseases which include, follicular adenoma, colloid goiter, multinodular goiter, thyroiditis, primary hyperthyroidism and hypothyroidism.⁴⁻⁶ Unilateral absence of superior and inferior thyroid artery, right sided aortic arch is other associated anomalies with hemiagenesis of the thyroid.⁷



Fig. 2: Grey scale ultrasound image showing the normal right lobe of thyroid.

Although our patient had hypothyroidism, the most common disease associated with the remaining thyroid is hyperthyroidism. Hypothyroidism remains an unusual complication of the thyroid hemiagenesis with an uncertain etiology.^{10,14,15} Thyroid hormone deficiency is mainly due to peripheral and central nervous system dysfunctions that makes it even more important to diagnose hypothyroidism.^{12,16}

We conclude that thyroid hemiagenesis and diffuse or focal thyroid disease in the existing thyroid lobe can be demonstrated by ultrasound well and stands as investigation of choice, further associated anomalies are delineated by CT and scintigraphy helps for differentiation between hemiagenesis and suppressed thyroid lobe.

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