Thyroid Hemiagenesis: Report of Two Cases

Tiroid Hemiagenezis: İki Olgu Sunumu

Tülin ÖZTÜRK,[a]
Muammer AKYOL,[a]
Alperen KAYALI,[a]
Gülen BURAKGAZİ[a]

[a]Clinic of Radiology,
Elazığ Training and Research Hospital,
Elazığ

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Yazışma Adresi/*Correspondence:*
Tülin ÖZTÜRK
Elazığ Training and Research Hospital,
Clinic of Radiology, Elazığ,
TÜRKİYE/TURKEY
drtulin58@gmail.com

ABSTRACT Thyroid hemiagenesis (TH) is a very rare congenital anomaly of the thyroid gland with absence of a lobe. It is usually diagnosed incidentally, because does not cause clinical symptoms. The etiology of TH is not clear. The accurate prevalence of TH is not known because the patiens are usually asymptomatic. Several studies showed that the prevalence of the TH in normal population is 0.05-0.2%. The diagnosis of TH can be easily made by ultrasound (US) in most patients. In this report we present two patients who diagnosed by US with left thyroid lobe agenesis.

Key Words: Thyroid gland; ultrasonography; thyroid dysgenesis

ÖZET Tiroid hemiagenezis (TH) tiroid bezinin bir lobunun yokluğu ile karakterize çok nadir doğumsal bir anomalisidir. Klinik semptomlara neden olmadığı için genellikle tesadüfi olarak tanı konulmaktadır. TH etyolojisi tam olarak açık değildir. TH'li hastalar genellikle asemptomatik olduğundan tam prevalansı bilinmemekle beraber yapılan birkaç çalışmada normal popülasyondaki sıklığı %0,05-0,2 olarak gösterilmiştir. TH tanısı pek çok hastada ultrason (US) ile kolayca konabilir. Bu yazıda sol tiroid lobu agenezisi olan ve US ile tanı konulan iki hasta sunulmuştur.

Anahtar Kelimeler: Tiroid bezi; ultrasonografi; tiroid disgenezisi

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Hemiagenesis of the thyroid gland (TH) is a very rare congenital anomaly defined by absence of one lobe. TH was described in 1866 by Handsfield-Jones.[1] Absence of one thyroid lobe usually does not cause clinical symptoms. Therefore its prevalence is unknown. But in the literature there are some prevalence studies in normal children. In these studies, the prevalence of TH was found 0.05% and 0.2%.[1,2] The etiology of TH is uncertain. There are two proposed theories related with the etiology: a descent defect from the floor of the primitive pharynx to trachea or failure of the original anlage to become bilobed and spread out laterally to both sides.[3,4] There may be a genetic component to the etiology as this rare condition has been documented in monozygotic twins.[5]

CASE REPORT

A 38 year-old woman and 11 year-old girl were referred for ultrasound (US) examination of thyroid gland. The right lobe and isthmus dimensions and

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parenchyma were normal in both patients (18x16x35 mm and 12x10x17 mm in anterior-posterior, transverse and craniocaudal planes, respectively). US examination showed no thyroid tissue on the left side of both patients (Figure 1). A diagnosis of thyroid hemiagenesis was made on the basis of US findings. The patients were complaining excessive sweating and growth retardation, respectively. The patients had no history of thyroid disease. The patients did not have any abnormalities in the neck examination. Serum concentration of TSH, free T3 and free T4 were in normal limits.

DISCUSSION

Thyroid agenesis may be complete, unilateral or isthmic. Congenital hemiagenesis is a rare developmental anomaly of the thyroid gland and usually does not cause clinical symptoms. It is usually diagnosed incidentally. Embryological development of the thyroid gland begins from an invagination of the endoderm in the primitive pharynx. The thyroid is then displaced inferiorly until it comes to its final location anterior to the trachea by the seventh week.[6] It is thought to result from failure of the cells to migrate laterally resulting in agenesis of a part of the thyroid.[7] It is unclear whether defect of the lobulation process is due to the interference of environmental factors or to some genetic factors.

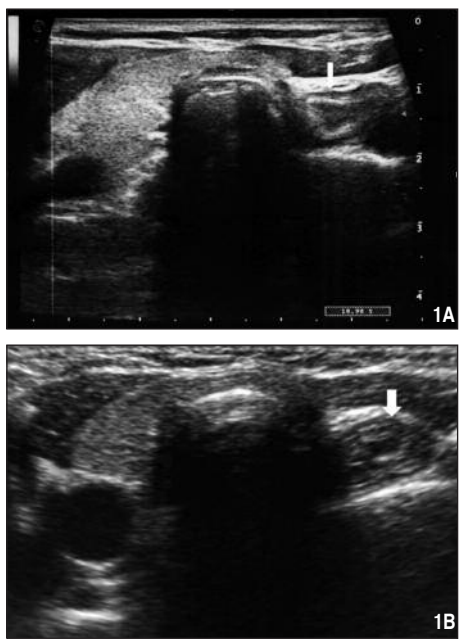

FIGURE 1: US shows right lobe (long arrow) of thyroid gland, esophagus (short arrow) in 38 year-old (Figure 1A) and US shows right lobe (long arrow) of thyroid gland, esophagus (short arrow) in 11 year-old (Figure 1B) patients but left lobes are absent.

It is known that the large majority of thyroid hemiagenesis is seen in the left lobe. In many normal subjects the thyroid is asymmetric, with the right lobe larger in size in respect to the left lobe. Thyroid hemiagenesis, therefore, could be considered an exaggeration of this difference producing an extreme asymmetry of the thyroid gland.[8]

TH has been reported three times more common in females as compared to males.[9] It may indicate a possible sex-related background. Most commonly, investigators have found absence of the left lobe versus the right with a concurrent absent thyroid isthmus in 50% of patients.[10,11] Coexisting thyroid disorders in the remaining lobe are a common occurrence with hemiagenesis. Any type of thyroid disorders might have been found in conjuction with TH; including hyperthyroidism, hypothyroidism, multinodular goiter, chronic thyroiditis and thyroid carcinoma.[5,12]

The diagnosis of TH can be made by US in most patients. US can demonstrate an absent lobe and is also helpful in follow up of euthyroid patients. It is cost effective, can be performed easily, and does not expose patients to radiation. However, other diagnostic methods can be used to diagnose thyroid hemiagenesis such as magnetic resonance imaging (MRI), computerized tomography (CT) and thyroid scintigraphy using radioisotopes. However, these techniques can be expensive, time consuming and there may be radiation exposure. Scintigraphy can be combined with US to locate ectopic thyroid tissue.[1,13]

In conclusion, TH is a very rare congenital anomaly of unknown etiology favoring females and absence of the left lobe. The remaining tissue may be affected by the whole spectrum of thyroid diseases. In diagnostic procedures, US is the most cost-effective way of diagnosis.

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