Thyroid Hemiagenesis Associated with Mixed Hyperactive Nodule: A Case Report

Mikst Hiperaktif Nodülle Birlikte Olan Tiroid Hemiagenesi: Bir Olgu Sunumu

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Abstract
Failure of embryologic development of one lobe of the thyroid gland is a rare congenital anomaly. This condition is usually diagnosed as a coincidental finding when the patient is being evaluated for a thyroid nodule, which develops as a compensatory hypertrophy of the other side that is present. A variety of pathological conditions may occur in the remaining thyroid tissue. For instance, thyroid adenoma, thyroid carcinoma, subacute thyroiditis, colloid nodule, Graves’ disease, simple goiter, or Hashimoto’s thyroiditis may be associated with this rare anomaly. However, thyroid hemiagenesis complicated with mixed (cystic-solid) hyperactive nodule is very rare. We herein report a 24-year old-female presented as mixed hyperactive nodule together with right lobe hemiagenesis of the thyroid gland. Turk J Em 2007; 11: 121-3

Key words: Thyroid hemiagenesis, hyperactive nodule

Özet
 Anahtar kelimeler: Tiroid hemiagenesi, hiperaktif nodül

Introduction
Thyroidal hemiagenesis is defined as congenital absence of one thyroid lobe. This rare condition was first described by Handfield and Jones in 1886 (1). This may result in dysfunction or hyperthyroidy of the remaining lobe. Moreover, thyroid adenoma, thyroid carcinoma, subacute thyroiditis, colloid nodule, Graves’ disease, simple goiter, or Hashimoto’s thyroiditis may be associat-ed with hemiagenesis of the thyroid (2-8). However, thyroid hemiagenesis complicated with mixed (cystic-solid) hyperactive nodule is exceptional. We herein report a 24-year old-female presented as mixed (cystic-solid) hyperactive nodule together with right lobe hemiagenesis of the thyroid gland.

Case Report
A 24-year-old female was admitted to our clinic with the complaint of cervical mass. She has had cervical mass for one year. Se reported no previous thyroid surgery. She reported no signifi-cant family history of a thyroid disorder. A general physical examination was free of any pathological finding. Local examination of the neck region revealed an enlarged and palpable left lobe of the thyroid gland, which was firm and not tender. The right lobe could not be palpated. No ocular signs were detected. Laboratory evaluation revealed normal serum biochemistry and abnormal thyroid functions tests (serum free T3, freeT4 and thyroid stimulating hormone (TSH) levels were 6.80 (1.8-4.6) pg/ml, 2.37 (0.9-1.7) ng/ml, and 0.01 (0.2-4.2) mU/ml, respectively). Technetium thyroid scan showed homogenous uptake of the radioisotope only in the left lobe (Figure 1). The antithyroid peroxidase antibody, antithyroglobulin antibody and TSH receptor antibody levels were within normal limits. Thyroid ultrasonogra-phy showed the absence of the right lobe in the thyroid bed and cystic–hypoechoic mass in the left thyroid lobe (Figure 2). Cytologic analysis of the fine needle aspiration material from the left-sided
mixed hyperactive nodule revealed benign colloidal nodule. Accordingly, right lobe hemiagenesis associated with cystic hyperactive colloid nodule in the left lobe was diagnosed.

**Discussion**

The absence of one thyroid lobe or both one thyroid lobe and the isthmus are rare conditions. The reported cases are few in number in the literature. A variety of physiologic and pathological conditions may develop in the remaining thyroid tissue including compensatory hypertrophy, thyroid adenoma, thyroid carcinoma, subacute thyroiditis, colloid nodule, Graves’ disease, simple goiter, or Hashimoto’s thyroiditis [2-8]. Thyroid hemiagenesis is usually discovered incidentally during the evaluation of those unrelated physiologic or pathologic conditions in the gland, and its real prevalence in the general population is unknown [9]. However, thyroid hemiagenesis complicated with cystic hyperactive nodule is very rare. In a medline search by using PubMed, we could find only one report of thyroid hemiagenesis and cystic thyroid nodule [10]. In this paper, we reported a new case of thyroid hemiagenesis together with cystic-solid hyperactive nodule.

![Figure 1](image1) Tc-99m pertechnetate scan shows the supression of the right lobe of the thyroid gland, and shows a hyperactive nodule in the left lobe. And sub-mandibular gland activity at the superior.

![Figure 2](image2) Thyroid sonogram through the thyroid gland shows a normal echo pattern and a cystic-hypoeoic nodule (14X10 mm,mixed, no calcification) from the left lobe of the thyroid gland and absent right lobe.

Women are likely than men to have this developmental anomaly, and a female to male ratio of 3:1 has been reported. Thyroid diseases are mainly seen in women and as most cases of thyroid hemiagenesis were diagnosed coincidentally with any of the above-mentioned conditions in the gland, this could be reason for female predominance [11]. Similarly, our patient was a young female who was admitted to our clinic with the complaint of a mass lesion in the remaining thyroid lobe. On the other hand, although agenesis of the left lobe is four times more common than the right [12-14], our patient had right thyroid lobe hemiagenesis.

If the radionuclide scintigraphy shows an absolute absence of tracer uptake in an area corresponding to one lobe of the thyroid gland, and the other lobe and isthmic portion shows normal and homogenous tracer uptake, the diagnosis of hemiagenesis should be considered. However, certain pathological conditions such as toxic adenoma, primary or secondary neoplasms, infiltrative diseases, and unilateral inflammations should be considered in the differential diagnosis. When those pathologic states affect an entire lobe of the thyroid gland, a functional hemiagenesis may appear on scintiscan. The thyroid scintiscan could not differentiate between functional and real hemiagenesis [15]. To overcome this problem technetium angiography, thallium scintigraphy, TSH or TRH tests are suggested. Even by using these tests, scintigraphy could fail to detect the other thyroid lobe. On the other hand, TSH stimulation scintigraphy is obsolete and should not be used for this purpose. The preferred method to demonstrate the absence versus suppression of a thyroid lobe is ultrasonography. It can be performed easily in many centers, is cost effective, has no radiation exposure risk, and is the most valuable diagnostic tool for detecting thyroid hemiagenesis [9]. Likewise, although technetium thyroid scan showed uptake of the radioisotope only in the left lobe, the diagnosis of hemiagenesis was confirmed by ultrasonography in our patient.

Three thyroid transcriptions factors, TTF1, TTF2, and pax-8 have been discovered. They are expressed not only in mature thyroid cells but also in their precursors. In TTF2 knock-out mice it was found that in embryos homozygous for the null allele the thyroid bud forms but does not migrate from the floor of the pharynx. There may be a specific gene regulated by TTF2 and a potential candidate in thyroid development defects. Newly identified sonic hedgehog (Shh) may cause hemiagenesis and ectopic development of the thyroid in mice [16]. Therefore, the pathogenesis of thyroid hemiagenesis may include genetic defects.

In conclusion, thyroid hemiagenesis could be associated with cystic hyperactive nodule. This anomaly should be considered in the differential diagnosis of thyroidal cystic lesions especially when physical examination shows no apparent thyroid tissue on one side of the neck. Especially, Thyroid ultrasonography should be performed in such cases to reach a definite diagnosis since thyroid scintiscan could not differentiate between anatomic hemiagenesis and functional hemiagenesis secondary to neoplastic or infiltrative disorders. Patients who undergo thyroid surgery for hyperactive nodule may be develop hypothyroidism due to hemiagenesis.
References