# Right thyroid hemiagenesis with multinodular goitre on the left lobe in female patient

## Bayan bir hastada sol lobta multinodüler guatr ile birlikte görülen sağ tiroid lobu hemiagenezisi

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## Abstract

Thyroidal hemiagenesis is a congenital anomaly, 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 this anomaly is not known yet. In this study, we report a 40 - year-old female patient with a multinoduler goitre on the left lobe, associated with hemiagenesis of the right lobe.

**Key words:** Thyroid, hemiagenesis, multinodular goitre

## Özet

Tiroid hemiagenezisi, tiroid glandının bir lobunun embriyoneal gelişim bozukluğu nedeniyle gelişememesi sonucu meydana gelen konjenital bir hastalıktır. Hastaların çoğu tesadüfen tanı alır ve diğer tiroid hastalıklarına sahiptirler. Bu nedenle bu anomalinin gerçek prevalansı tam olarak bilinmemektedir. Biz bu olgumuzda tiroid sağ lobunda görülen ve sol lobta multinodüler guatrın eşlik ettiği bir vakayı sunmayı amaçladık.

**Anahtar kelimeler:** Tiroid, hemiagenezis, multinodüler guatr

### Introduction

The absence of left or right lobe of the thyroid gland is named thyroid hemiagenesis. Thyroid hemiagenesis, was first reported in 1866 by Handsfield-Jones[1], is a very rare congenital anomaly of the thyroid gland[2,3]. Most of the patients are detected incidentally and have an associated thyroid disease[4]. The true frequency of thyroid hemiagenesis is unknown because the absence of one thyroid lobe usually does not cause clinical symptoms by itself. It is more common in women than men[4,5]. Hemiagenesis of the left lobe is more often than the right[3-5]. In this report, we present a 40 - year-old female patient with a multinoduler goitre in the left lobe, associated with hemiagenesis of the right lobe.

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### Case Report

A 40 - year-old female patient admitted to the outpatient clinic with a gradual onset swelling on her left side of the neck for 2 years. Physical examination of the thyroid gland revealed easily palpable, smooth and enlarged left lobe. However the right lobe could not be palpated. There was no known history of thyroid operation or neck dissection. The rest of the physical examination was unremarkable. Her thyroid function test was normal [free $T_3$: 3.64 pg/ml (normal range 2.30-4.20), free $T_4$: 1.11 ng/dl (normal range: 0.88-1.72) and TSH: 0.85 mIU/L (normal range: 0.57-5.6)], but antithyroperoxidase (Anti-TPO) and antithyroglobulin (Anti-TG) ranges were elevated, 221 U/ml (normal range:0-60) 89

U/ml (normal range:0-60), respectively. Ultrasound examination showed no thyroid tissue on the right side while the left lobe of thyroid was measured as 2.7x2.6 x6.2 cm. The thickness of isthmus was measured 1.4 mm. There was a large left lobe including 8 nodules. The solid dominant nodule located in the upper pole of the left lobe and was measured as 26×20 mm. The some nodules were including calcification and cystic degeneration areas. Ultrasound-guided fine needle aspiration biopsy was performed two times but was reported as non-diagnostic. Based on clinical and ultrasonographic findings thyroidectomy was planed. We performed left thyroid lobectomy and isthmusectomy. During the operation, we explored the right lobe but we did not find it ***(figure 1)***. In this way intraoperatively, right thyroid hemiagenesis was confirmed. Pathologic examination of the left lobe was reported as a hashimoto thyroiditis. The patient was discharged from hospital on the second postoperative day.

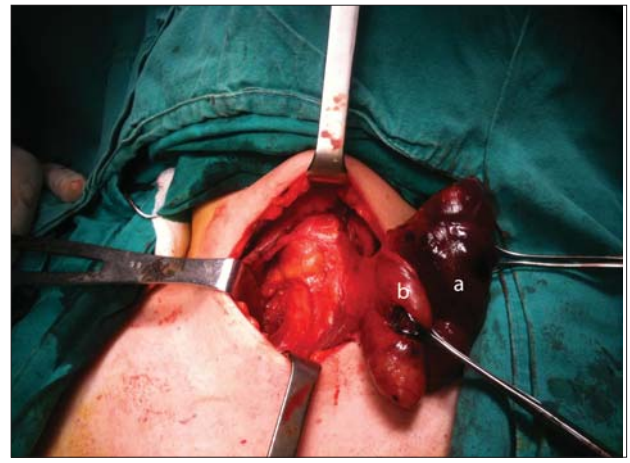


**Figure 1.** During the operation, right thyroid lobe was not found. a: left thyroid lobe, b: isthmus

## Discussion

Thyroid hemiagenesis (THA) is a rare congenital abnormality that is characterized by the absence of one lobe of the thyroid gland with or without the absence of the isthmus[6]. It was first described in 1866 by Handsfield-Jones[1]. The actual incidence of THA is unknown, since in most of the cases the diagnosis is made coincidentally, usually in patients submitted to thyroid scan or thyroid surgery because of the suspicion of other thyroid abnormalities. This could explain the high frequency of the association of hemiagenesis with other thyroid abnormalities[7]. In the literature its prevalence rate has been reported between 0.05% and 0.2% in the normal population[2-4].

The cause of the abnormal development of the thyroid that leads to THA is not known. It is thought to result from failure of the cells to migrate laterally resulting in agenesis of a part of the thyroid[8]. Recently, several genes have been found to be involved in thyroid morphogenesis and descent, but these have not been studied in hemiagenesis[9,10].

The prevalence of this developmental defect is greater in females[2,4,5]. The preponderance of women with THA may only be a result of better detectability due to the fact that thyroid disorders occur more often in the female sex[5]. In contrast to our case, the left lobe of the thyroid gland is involved in most of the cases of thyroid hemiagenesis and the isthmus may also be absent in up to half of them[2,5,11].

The clinical presentation of thyroid hemiagenesis is highly variable. Although person with THA may have a normal thyroid lobe with euthyroidism, both hypothyroidism and hyperthyroidism are known to occur[4,12-16]. Our case was clinically euthyroid. TSH is observed to be elevated in person with thyroid hemiagenesis when compared with normal person, presumed to be due to overstimulation of the normal lobe and it is suggested that this may not truly represent subclinical hypothyroidism. Furthermore, a higher incidence of associated functional, morphological, and autoimmune thyroid disorders in patients with THA is observed when compared to subjects with bilobate thyroid[3,4,6]. In most of the clinical reports on THA, an association with other thyroid disorders was found such as hyperthyroidism, multinodular goitre, hypothyroidism, benign adenoma and Graves' disease and acute and subacute thyroiditis[4,5,7,11-16]. Because elevated TSH, as a thyroid growth-promoting factor, may lead to diffuse or nodular goiter and is connected with an increased risk of neoplastic transformation. Ruchala M, et al.[5] showed that the frequency of thyroid abnormalities in patients with THA varies with age, which is probably due to the longer exposure of the hemiagenetic gland to TSH overstimulation in older patients. In our case, histopathologic examination was reported as hashimoto thyroiditis5.

Most of the cases with THA are clinically asymptomatic becasue the absence of one thyroid lobe does not usually cause clinical symptoms[4]. They are discovered when patients present with a lesion in the functioning lobe or are diagnosed incidentally. Clinical examination has a limited diagnostic value, but tracheal rings may be easily palpable in patients with absent isthmus[6].

Ultrasound is an indispensable tool in the workup and diagnosis of thyroid disease. It is very cheap, easy to perform and above all there are no radiation hazards. Ultrasound-guided fine needle aspiration biopsy (UG-FNAB) should be performed in all patients who have nodules in the their remaining thyroid lobe[4,5,14,16]. In our case, we performed UG-FNAB two times but the histopathologic examination did not have diagnostic value.

Thyroid scintigraphy may also be helpful in differentiating hemiagenesis from a suppressed lobe. However there are several clinical conditions mimicking THA in scintigraphic evaluation. Autonomously functioning nodules with suppressed normal thyroid tissue, primary or secondary neoplasms, infiltrative diseases such as amyloidosis and unilateral inflammations of one lobe can mimic thyroid hemiagenesis[6,7,14].

In conclusion, THA occur more often in the left lobe of the thyroid gland but It should be keep in mind that the condition can be found in the right lobe. Surgery should be performed if the remaining lobe contains radiologic or histopathologic findings or clinical suspicion of malignancy.

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