Thyroid Hemiagenesis with Hashimoto’s Disease: A Case Report and Review of the Literature

Seval ERHAMAMC1, Mehmet REYHAN1, Enver AVCI2

Received 20th July 2013, Accepted 13th August 2013

Abstract: Thyroid hemiagenesis is a rare abnormality in which one thyroid lobe with or without isthmus fails to develop. It is usually found incidentally in patients who are evaluated for some other disease. Thyroid scintigraphy with Tc-99m pertechnetate and ultrasonography play an important role in diagnosis. Here, we report a case of 77-year-old man with Hashimoto’s Disease associated with hemiagenesis of the left lobe. The purpose of this presentation is to review the literature and discuss the scintigraphic finding on thyroid hemiagenesis.

Keywords: Thyroid hemiagenesis, Tc-99m pertechnetate, Scintigraphy, Hashimoto’s Disease.

1. Introduction

Thyroid hemiagenesis is a rare embryological condition, which was first reported by Handfield-Jones in 1866 [1]. The true prevalence of this congenital abnormality is not known because the absence of one thyroid lobe usually does not cause clinical symptoms by itself. However, several studies show that the prevalence of this abnormality was estimated as 0.05-0.2 % in literature [2-5]. Thyroid hemiagenesis is an asymptomatic condition, which is usually detected incidentally. However, all forms of functional status can be observed in patients with hemiagenesis, including euthyroidism, hyperthyroidism and hypothyroidism [6-8]. Hyperthyroidism was reported to be the major reason for diagnosis. Most of the patients with thyroidal hemiagenesis had several thyroid disorders [3-13]. Thyroid scintigraphy and ultrasonography play an important role in diagnosis. Here, we report a case of 77-year-old man with Hashimoto’s Disease, associated with hemiagenesis of the left lobe.

2. Case Report

A 77-year-old man patient was admitted to the endocrinology clinic with the complaint of palpitation and sweating. On physical examination, the left lobe was non-palpable. He had no history of operation on thyroid gland. The thyroid function tests were as follows: free triiodothyronine (fT3): 2.84 pg/mL (normal range: 1.71-3.71), free thyroxine (fT4): 1.46 ng/dL (normal range: 0.7-1.48), thyroid stimulating hormone (TSH): 0.0134 µU/mL (normal range: 0.36-4.94), antithyroidperoxidase (anti-TPO): 868 IU/mL (normal range: 0-35) and antithyroglobulin antibodies (anti-TG): 183.23 IU/mL (normal range: 0-40). It was concluded that the patient had subclinic hyperthyroidism. On ultrasonography (USG), right thyroid lobe was measured 48x19x17 mm and istmus was 7mm, and the left lobe was absent. Ultrasonographic findings were consistent with left thyroid lobe agenesis. To rule out the possible associated diseases, thyroid scintigraphy examination was requested. Thyroid scan with Tc99m pertechnetate confirmed the absence of the left thyroid lobe, with homogeneous uptake in the slightly enlarged right lobe and istmus (Fig 1). We noted the characteristic hockey stick sign found on scan. The case was diagnosed as left lobe hemiagenesis of thyroid. Due to the mild symptoms and the subclinic hyperthyroidism, and in view of the possibility of developing hypothyroidism or hyperthyroidism in the future, follow-up with thyroid function tests was advised.

Figure 1. Thyroid scan with Tc99m pertechnetate revealed absence of the left thyroid lobe and homogeneous uptake in the slightly enlarged right lobe and istmus.

One year later, the thyroid function tests were as follows; fT3: 2.60 pg/mL, fT4: 1.18 ng/dL, TSH: 3.97 µU/mL, anti-TPO: 1000 IU/mL and anti-TG: 250.78 IU/mL. The laboratory analysis

1 Department of Nuclear Medicine, Faculty of Medicine, Baskent University, Konya, Turkey
2 Division of Internal Medicine, Faculty of Medicine, Baskent University, Konya, Turkey
* Corresponding Author: Email: sevaler@yahoo.com
showed that fT3, fT4 and TSH were within normal limits, specific antibodies were elevated. The patient was finally diagnosed as having Hashimoto’s Disease, and was followed without therapy.

3. Discussion

Thyroid hemiagenesis is one of the rare congenital anomalies [1]. It is more common in females than males (75% versus 25%). The absence of the left lobe was detected in 80% of cases, and agenesis of the isthmus was seen in 50% of cases [5-7]. In accordance with the literature, left lobe was absent in our case.

Although exact pathogenesis of thyroid hemiagenesis is unknown, some genetic alterations in transcriptional control of thyroid development and in the control of migration of the median thyroid bud during embryogenesis have been questioned. Some familial cases have been reported, including the one associated with heterozygous mutation in the Pax8 gene [14-15].

The absence of one thyroid lobe usually does not cause clinical symptoms unless there is a thyroid hormone imbalance and possible thyroid disease of the other developmentally normal thyroid lobe [5-10]. Compensatory hypertrophy of the residual thyroid lobe was reported and is probably due to thyroid tissue overstimulation by TSH [10]. Because of the high risk of goiter or hypothyroidism, systematic follow-up of all identified cases of thyroid hemiagenesis was recommended [10]. Thyroid hemiagenesis is sometimes incidentally diagnosed during routine work-up for suspected thyroid disorders [4]. The present case was evaluated for thyroid disease because of the clinical suspicion.

Thyroid hemiagenesis is frequently associated with other thyroidal disease in the remaining thyroid tissue. Some of these are benign adenoma, multinodular goiter, chronic thyroiditis, Graves’ disease, carcinoma, toxic adenoma [11-13]. Therefore, these patients need careful follow up. In addition, extrathyroidal lesions, such as parathyroid adenoma or hyperplasia, cervical thyreoid cysts, ectopic sublingual thyroid gland and thyroglossal duct cyst have been reported [16-19].

Thyroid hemiagenesis can be diagnosed in any patient in whom, on physical examination, no thyroid tissue can be palpated on one side. However, the role of the physical examination is limited. Imaging modalities play an important role in diagnosis. The scintigraphic examination is useful in the demonstration of absent thyroid tissue and probable ectopic thyroid tissue. Scintigraphic imaging is reported to show the characteristic cockeye stick sign in patients with thyroid hemiagenesis, as the present case [7]. However, there are several clinical conditions mimicking thyroid hemiagenesis. Other causes for nonvisualization of one thyroid lobe include a contralateral autonomous solitary thyroid nodule that is suppressing normal extranodular tissue; focal or unilateral subacute thyroiditis; primary or metastatic carcinoma; postinflammatory atrophy of thyroid tissue as a result of Hashimoto’s disease; infiltrative diseases. In these clinical conditions functional hemiagenesis based on thyroid scan alone may be misdiagnosed as congenital hemiagenesis. Therefore, thyroid scans should be confirmed by other methods revealing the morphology of the thyroid. Ultrasonographic examination, clinical and laboratory findings, and fine-needle biopsy may be helpful in the differential diagnosis. Ultrasonography is widely available. It is cost effective, can be performed easily, and does not expose patients to radiation. Computed tomography and magnetic resonance imaging can be useful to distinguish between unilateral thyroid disease and true hemiagenesis. Both of the techniques are expensive and time consuming. In addition, diagnostic performance of these investigations is comparable with USG for the thyroid. If nodular thyroidal disease is associated, fine-needle biopsy should be performed in order to rule out primary and secondary malignancies.

We have described a case of thyroid left-lobe hemiagenesis with Hashimoto’s Disease in the right lobe. Laboratory analysis initially showed subclinical thyroid hyperfunction and the presence of thyroid stimulating antibodies. Ultrasonography and thyroid scan confirmed the left lobe agenesis.

In conclusion, patients with thyroid hemiagenesis need careful follow-up, because of the possible development of thyroid disease. The combined use of scintigraphy and ultrasonography can prevent the misdiagnosis.

References

[15] Castanet M, Leenhardt L, Leger J, Simon-Carre A, Lyonne S, Pelet A et al. Thyroid hemiagenesis is a rare variant of thyroid dysgenesis with a familial component but without


