A Case of Thyroid Hemiagenesis: An Exceptional Case

Tiroid Hemiagenezi Vakası: Sıra Dışı Bir Vaka

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Abstract

Thyroid hemiagenesis is a rare congenital anomaly in which one thyroid lobe fails to develop. We recently observed a case of thyroid hemiagenesis accompanied by parathyroid adenoma and papillary thyroid microcarcinoma in a 51-years-old woman. The patient’s serum calcium level was 10.9 mg/dL and that of intact parathyroid hormone was 218 pg/mL. Although she had a history of thyroid hemiagenesis, the patient was in a euthyroid state without thyroxine replacement. Thyroid ultrasonography detected no right lobe and four nodules of varying sizes in the left thyroid lobe. Tc-99m scintigraphy also demonstrated the absence of the right thyroid lobe. In addition, MIBI-parathyroid scintigraphy showed a parathyroid adenoma at the lower pole of the right cervical region. Based on the diagnosis, parathyroid adenoma excision and thyroidectomy were performed. Postoperative pathological diagnosis revealed parathyroid adenoma and papillary thyroid microcarcinoma in the form of a 0.4 cm sized tumor in the left thyroid lobe. The coexistence of thyroid hemiagenesis, primary hyperparathyroidism, and papillary thyroid microcarcinoma is truly exceptional and has never been reported in the literature before.

Keywords: Thyroid hemiagenesis; primary hyperparathyroidism; thyroid papillary microcarcinoma

Case Report

A 51-year-old woman with a history of Colles fracture presented for the control of bone health. Laboratory examination revealed elevated calcium level of 10.9 mg/dL, elevated intact parathyroid hormone (PTH) level of 218 pg/mL, and 24 h urinary calcium excretion at 580 mg/day (Table 1). She had a T score of -2.5 at the L1-L4 level of bone mineral densitometry. The patient had a medical history of thyroid hemiagenesis but was...
in a euthyroid state without thyroxine replacement. After primary hyperparathyroidism, she was diagnosed on the basis of the laboratory tests, and we performed several imaging studies. Thyroid ultrasonography detected no right lobe, four nodules of sizes 1.5x0.9 cm, 1.3x0.7 cm, 0.4x0.2 cm and 0.5x0.3 cm in the left thyroid lobe, and no signs of parathyroid adenoma. Tc-99m scintigraphy also demonstrated the absence of the right thyroid lobe (Figure 1), while MIBI-parathyroid scintigraphy showed parathyroid adenoma at the lower pole of the right cervical region (Figure 2). After diagnosing primary hyperparathyroidism and thyroid hemiagenesis with multiple nodules, parathyroid adenoma excision and thyroidectomy were performed. The postoperative pathological diagnosis revealed a 1.3 cm sized parathyroid adenoma, a 0.4 cm sized papillary thyroid microcarcinoma in the left thyroid lobe, and no lymph node metastasis. On the day after surgery, PTH and serum calcium levels were normalized. The patient’s postoperative course was overall favorable and she was discharged on the second day after surgery.

**Discussion**

Thyroid hemiagenesis is a rare congenital anomaly in which one thyroid lobe fails to develop embryonically. The incidence rate varies across reports but is estimated to be 0.01%, with three times as many women are affected as men, and the left lobe is affected four times more than the right lobe (1). Ruchala et al. studied 40 cases of thyroid hemiagenesis (2), and observed left-sided hemiagenesis in 87.5% of the patients. Our patient had right lobe hemiagenesis, which is considered to be relatively rare. The remaining thyroid lobe can compensate functionally for the missing lobe, resulting in clinically euthyroid status. Patients with thyroid hemiagenesis have significantly higher levels of the thyroid growth hormone thyrotropin (TSH) and free triiodothyronine (FT3) when compared to people with normally developed thyroid. Increased FT3 levels may be explained by the increased peripheral conversion of thyroxine (T4) to T3. Since the changes in the hypothalamic-pituitary-thyroid axis in thyroid hemiagenesis patients results in

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<th>Table 1. Preoperative laboratory values of the case.</th>
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<td><strong>Preoperative values</strong></td>
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<tr>
<td>Serum Ca (mg/dL)</td>
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<td>Serum P (mg/dL)</td>
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<td>24 h urinary Ca (mg/day)</td>
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<td>PTH (pg/mL)</td>
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<td>FT4 (ng/dL)</td>
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<td>TSH (µIU/mL)</td>
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PTH: Parathyroid hormone; TSH: Thyroid stimulating hormone.
higher TSH thresholds, the higher TSH levels observed are usually within normal ranges (2). In addition, patients with hemiagenesis can display various thyroid dysfunctions like euthyroidism, hyperthyroidism, and hypothyroidism (1). One study reported thyroid hemiagenesis accompanied by Graves’ disease in 40 patients (3).

The available literature reports approximately 350 cases with the diagnosis of thyroid hemiagenesis (3), and 56% (195/350) of the cases were associated with other thyroid disorders including euthyroid multinodular goiter, Hashimoto thyroiditis, congenital hypothyroidism, differentiated thyroid cancer, and thyroglossal duct cysts. Hypothyroidism and/or Hashimoto thyroiditis are the most common thyroid disorders accompanying hemiagenesis (3).

Co-existence of hemiagenesis and papillary thyroid carcinoma is rare (4,5), with only eight cases of accompanying differentiated thyroid carcinoma and one case of medullary thyroid carcinoma reported so far (6). Elevated levels of TSH may lead to diffuse or nodular goiter and is associated with an increased risk of neoplastic transformation (7). In a study by Haymart et al. (8), higher TSH levels were significantly correlated with an increased risk of thyroid malignancies, more likely via TSH receptor stimulation. Therefore, individuals with thyroid hemiagenesis that have both the high prevalence of thyroid nodular diseases and increased baseline TSH levels are at a higher risk of developing thyroid neoplasms. Therefore, thyroid hemiagenesis patients should be followed-up closely for monitoring TSH levels and the possibility of a neoplastic disease.

The association of thyroid hemiagenesis and primary hyperparathyroidism is very rare. Ferrari et al. (9) reported that only nine cases of thyroid hemiagenesis associated with primary hyperparathyroidism are known so far, and in six of those, parathyroid adenoma was present on the same side as thyroid hemiagenesis. In those ipsilateral cases, adenomas were also observed in the lower parathyroid glands. In our case, parathyroid adenoma was present in the lower parathyroid gland of the thyroid hemiagenesis side.

Although the thyroid and parathyroid glands originate from the pharyngeal endoderm, they develop into different structures embryonically. The thyroid diverticulum becomes lobulated and joins the two lateral anlagen (30% of the gland weight) originating from the fourth and fifth pharyngeal pouches during the descent. Parathyroid glands result from dorsal epithelium of the third and fourth pharyngeal pouches. The inferior parathyroid gland emerging from the third pharyngeal pouch migrates caudally to lie on the dorsal surface of the inferior aspect of the thyroid glands. The superior parathyroid gland emerging from the fourth pharyngeal pouch migrates before it lies on the dorsal and superior aspect of thyroid gland.

**Conclusion**

The coexistence of thyroid hemiagenesis, primary hyperparathyroidism and thyroid papillary microcancer is truly exceptional, and to the best of our knowledge, has never been reported before in medical literature.

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**Author Contributions**

Concept: Banu Sarer Yürekli; Design: Banu Sarer Yürekli, Nilüfer Özdemir Kutbay; Data Collection or Processing: Özer Makay; Analysis or Interpretation: Gökan İçöz, Gökhan Özgen; Literature Search: Nilüfer Özdemir Kutbay, Mehmet Erdoğan; Writing: Banu Sarer Yürekli.

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