Severe hyperparathyroidism in patient with right thyroid hemiagenesis

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Abstract
Thyroid hemiagenesis is an infrequent congenital disorder which is rarely associated with hyperparathyroidism. We present a case of an adult woman who presented with hyperparathyroidism and ipsilateral thyroid hemiagenesis. Parathyroid adenoma was excised by minimal invasive parathyroidectomy.

Keywords: Right thyroid hemiagenesis, Parathyroid adenoma, Primary hyperparathyroidism.

Introduction
The thyroid gland consists of two lobes connected by an isthmus. Thyroid hemiagenesis is an infrequent congenital disorder. The incidence of thyroid hemiagenesis is not known exactly. Most of the reported cases have been related with thyroid diseases such as Graves diseases and nodular goiter.1-3 Rarely, thyroid hemiagenesis is associated with hyperparathyroidism.4-8 We present a case of hyperparathyroidism and ipsilateral thyroid hemiagenesis.

Case Report
A 27-year-old woman was admitted in January 2015 to the Endocrinology Department complaining of nausea, vomiting, abdominal pain and fatigue. Physical examination was unremarkable other than a mild abdominal tenderness. Laboratory tests revealed high serum and urine calcium (18.7 mg/dL and 1434 mg/24h, vs. normal value of 8.6 - 10.5 mg/dL and 0-250 mg/24h, respectively), and intact parathyroid hormone (PTH) (1542pg/mL vs. normal value of 16-87 pg/mL) levels, low phosphorus (1.5 mg/dL vs. normal value of 2.5 - 4.5 mg/dL) and vitamin-D levels (3.92 ng/dL vs. normal value of > 30 ng/dL), and normal thyroid stimulating hormone (TSH) levels (1.82 uIU/mL vs. normal value of 0.270-4.20 uIU/mL). We diagnosed primary hyperparathyroidism, and performed other tests. Cervical ultrasound revealed left thyroid lobe, but the absence of right thyroid lobe,

Figure-1: Cervical ultrasound revealed left thyroid lobe, but the absence of right thyroid lobe and a hypoechoic lesion compatible with a parathyroid adenoma on the right side.

Figure-2: Thyroid scintigraphy revealed the absence of the thyroid right lobe.
thyroid left lobe measuring 18x20x54 mm in size, and a hypoechoic lesion measuring 20x11 mm in size were compatible with a parathyroid adenoma on the right side (Figure-1). Technetium-99m (Tc-99m) scintigraphy revealed a parathyroid adenoma on the right side. Thyroid scintigraphy showed the absence of right thyroid lobe (Figure-2). The patient was evaluated for multiple endocrine neoplasia (MEN) 1 and 2 syndromes because she was at a young age. However, we did not find an association with the syndromes.

We performed saline and furosemide infusion. The patient’s serum calcium levels fell to 11.9 mg/dL. Parathyroid adenoma was excised by minimal invasive parathyroidectomy. Postoperative PTH and calcium levels fell to 76 pg/mL and 8.3 mg/dL, respectively. The patient was discharged the next day. The pathology of specimen was consistent with parathyroid adenoma.

Discussion
Thyroid hemiagenesis has been reported in literature with Hashimoto's thyroiditis, hyperparathyroidism due to parathyroid adenoma or hyperplasia, thyroglossal duct cyst, and thyroid neoplasms. However, thyroid hemiagenesis have been very rarely reported with hyperparathyroidism. Our patient had severe hypercalcemia symptoms with the highest calcium and PTH levels in patients with thyroid hemiagenesis. Data regarding the prevalence of thyroid hemiagenesis in adult population is not substantial. However, it is estimated to be 0.002-0.05% in the paediatric population. The absence of one thyroid lobe is usually asymptomatic. Therefore, these patients are usually detected during neck imaging for contralateral thyroid diseases or other neck pathology. Our patient was also detected during parathyroid imaging. Other thyroid lobe is usually compensatory hypertrophic due to thyroid gland overstimulation by TSH. Also, our patient's left thyroid lobe was mild hypertrophic.

The cause of the thyroid hemiagenesis is unknown. The majority of it is due to the absence of the left lobe. Many normal people have asymmetric, larger in size thyroid right lobe than left lobe. Therefore the cause of the lack of left lobe predominance could be some imbalance in the development of the bilobed gland during embryogenesis. However, that was unlikely for our patient who lacked the right thyroid lobe.

The thyroid hemiagenesis can be seen among members of the same family. It is suggested that genetic factors could affect this malformation. However, the molecular mechanism of thyroid hemiagenesis is largely unknown. We evaluated patient’s relatives by cervical ultrasound. However, we could not detect another thyroid hemiagenesis.

Conclusion
Thyroid hemiagenesis is infrequently encountered in clinical practice. Therefore it may be overlooked during routine cervical ultrasound. Many patients' thyroid hemiagenesis can only be found intraoperatively. However, Tc-99m scintigraphy can demonstrate thyroid hemiagenesis in early images in patients with hyperparathyroidism. However, the malformation may be diagnosed during ultrasound imaging if there is a clinical suspicion.

References