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Thyroid hemiagenesis associated with multinodular goiter and Hashimoto's thyroiditis

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SUMMARY: **Thyroid hemiagenesis associated with multinodular goiter and Hashimoto's thyroiditis.**

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Thyroid hemiagenesis is a rare congenital abnormality in which one of the thyroid lobes is not developed. It can be associated with various thyroid diseases, such as Grave's disease, nodular goiter and thyroid neoplasm, rarely with hyperparathyroidism. We report a case of a 50-year old woman with left thyroid lobe agenesis diagnosed by ultrasonography and scintigraphy. Right thyroidectomy was performed and the histopathological examination showed diffuse hyperplasia, multinodular goiter and Hashimoto's thyroiditis. To our knowledge, this is the first description of multinodular goiter and Hashimoto's thyroiditis in a patient with thyroid hemiagenesis.

KEY WORDS: Thyroid hemiagenesis - Multinodular goiter - Hashimoto's thyroiditis.

Introduction

Thyroid hemiagenesis characterized by the total absence of one lobe is a very rare congenital anomaly of the thyroid gland (1). The true prevalence of this malformation is not known. However it is estimated to be about 0.02- 0.05% and it is generally detected incidentally (2-3). Etiology of thyroid hemiagenesis is still unclear and it can result from either an abnormal descent or an agenesis of one lobe (2-4).

Left lobe involvement is more frequent and these patients have a higher incidence of associated functional, morphological and autoimmune thyroid disorders (4).

We report a case of a female patient went to our observation for an aspecific symptomatology in which an incidental left thyroid lobe agenesis was found.

Case report

A 50 years old female patient came to our observation for palpitation, insomnia, fatigue and a swelling in the neck that had been gradually growing in the last months.

On physical examination, she had a rate of 75 pulse/minute, blood pressure of 110/60 mmHg and a palpable right thyroid lobe.

The patient was being treated with thyroxine at dose of 50 μg/day and at the laboratory tests, the biochemical thyroid profile was as follows: *FT3* 5.68 (range 3.32- 7.45 pmol/L); *FT4* 10.7 (range 7.9- 14.4 pmol/L); *TSH* 0.29 (range 0.34- 4.4 μUI/mL); *Thyroglobulin* 0.78 (<50 ng/mL); *TgAb* 15 (<4 UI/mL); *TPOAb* 62 (<9 UI/mL); *Parathormone* 77.5 (range 12-88 pg/mL); *Total Calcium level* 9.1 (range 8.4-10.2 mg/dL).

On direct chest X-ray, trachea was shifted to the left.

On thyroid ultrasonography left lobe and isthmus could not be visualized. Right lobe measured 40 x 31 mm with an heterogeneous parenchyma caused by multiple circumscribed hypoechoic solid nodules with partially cystic degeneration areas.

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No lymphadenopathy on either side of the neck was found.

Tc-99m scintigraphy showed one hypercaptant nodule in the right lobe. No ectopic thyroid tissue was revealed.

Fine needle aspiration was performed on the nodule of the right thyroid lobe. Cytological examination showed the presence of normal thyrocytes compatible with a benign nodule.

A right thyroidectomy was performed and the agenesis of the left lobe and isthmus was confirmed intraoperatively (Figures 1, 2) while left parathyroids were found. The right parathyroid glands and the inferior laryngeal nerve were in their normal place.

Hystopathological findings showed diffuse hyperplasia and multinodular goiter in patient with thyroid hemiagenesis and Hashimoto's thyroiditis.

No complication were observed and the patient was discharged on the second postoperative day.

Discussion

Thyroid hemiagenesis is a rare congenital abnormality characterized by the lack of development of one of the thyroid lobes (3). First case was described by Handfield-Jones in 1866 (5-6).

The prevalence has been reported between 0.025 and 0.05% in general population and between 0.16 and 0.25% in patients with thyroid disorders (1-2). For unknown reasons, agenesis concerns the left thyroid lobe in 60-80% of the cases, and women are more frequently affected than men (3:1) (7), like in our case.

Embryological development of the thyroid gland begins from an invagination of the endoderm in the primitive pharynx. This invagination grows ventrally while remaining attached to the pharyngeal floor. The thyroid rudiment migrates to its correct anatomical position, anterior to the pharynx and only there it begins to grow laterally to create the bilobed thyroid gland (8). Accessory thyroid gland tissue generally arises from remnants of the thyroglossal duct, including an ectopic lingual thyroid thus, embriologically, the most common congenital abnormalities of the thyroid include a lingual thyroid and a thyreoglossal cyst (9).

Congenital thyroid anomalies may be caused either by abnormal descent of the gland or by incomplete genesis of a lobe. However the etiology of the hemiagenesis still remains unclear (5-8). A high incidence of the condition in monozygotic twins or in

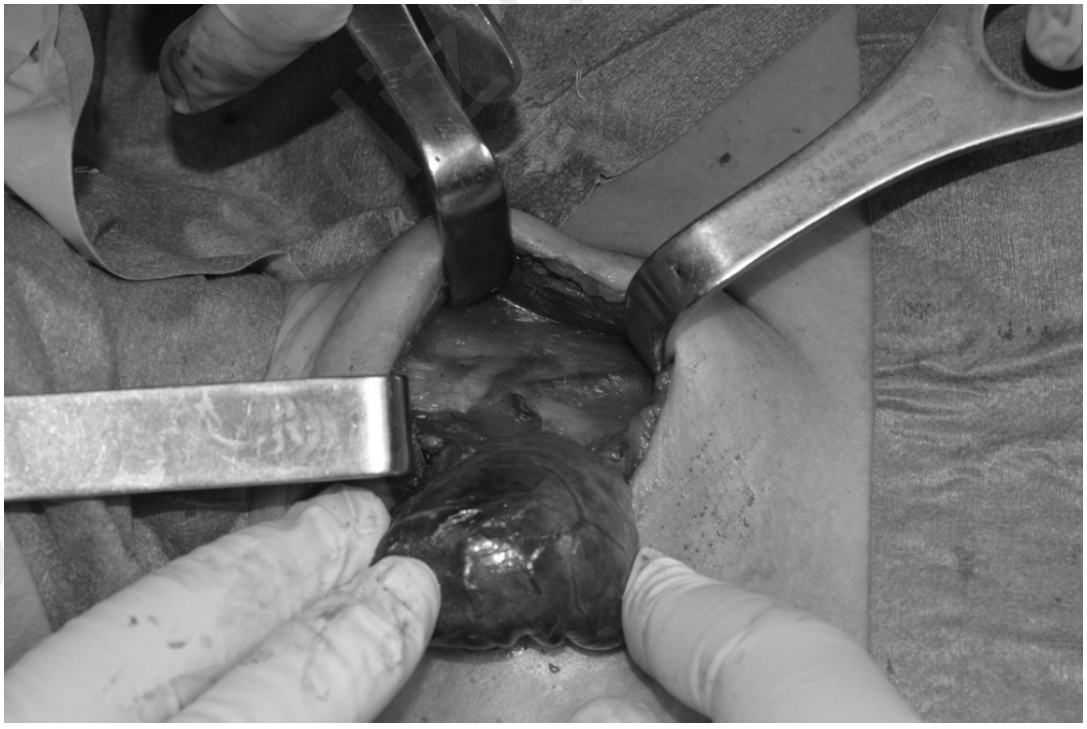

Figure 1 - Intraoperative picture.

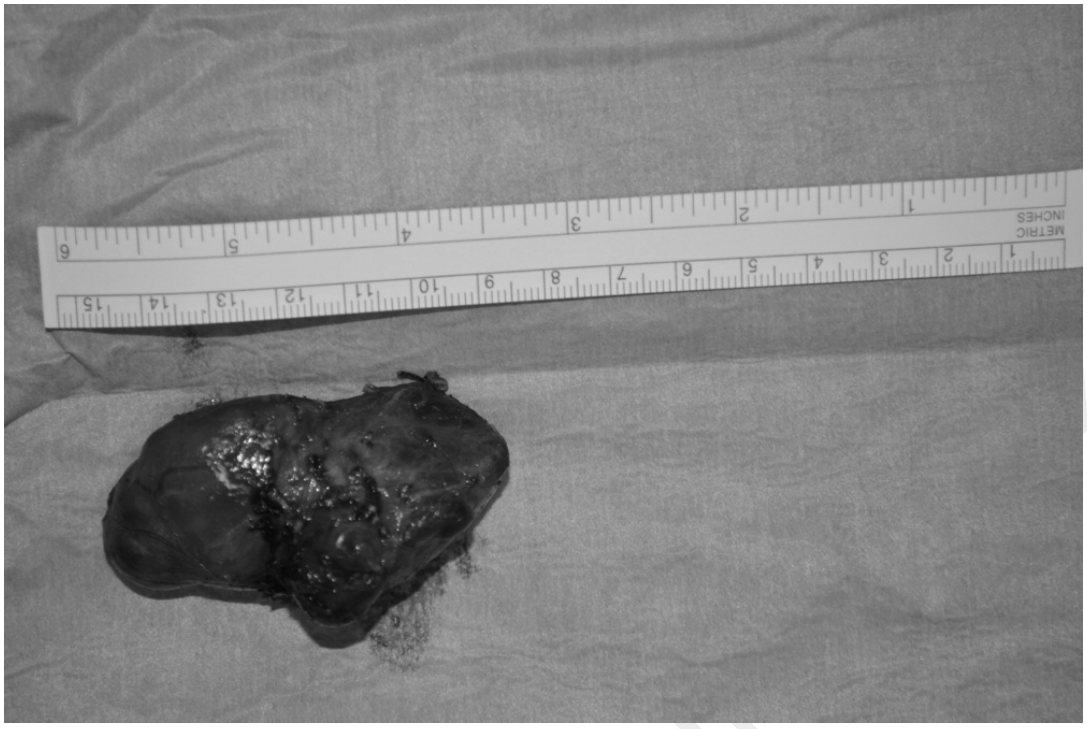

Figure 2 - Excised thyroid right lobe.

association with other thyroid malformations within the family may indicate a possible role of genetic factors. Several genes have been implicated in thyroid morphogenesis and descent such as PAX-8, TTF-1 and TTF-2 (2-10). PAX-8 partecipates in regulation of embryogenesis of the thyroid gland as well as mullerian and renal/ upper urinary tracts (11). Most of the reported cases of thyroid hemiagenesis are associated with thyroid diseases such as Grave's disease, nodular goiter and thyroid neoplasm, rarely with hyperparathyroidism (3-12).

Conclusions

In conclusion, we reported a case of thyroid hemiagenesis associated with a multinodular pretoxic goiter and Hashimoto thyroiditis. This is the first description of such an association. In this case, the excision of the remaining lobe provided to be the appropriate treatment.

Conflict of interests

The Authors declare that there is no conflict of interests regarding the publication of this article.

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