

Hemiogenesis of the thyroid gland and T₃ hyperthyroidism

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Summary

A patient with hemiogenesis of the thyroid gland who presented with T₃ toxicosis is described. The diagnosis of thyroidal hemiogenesis was established through the administration of thyroid stimulating hormone and a thyroid scintiscan.

T₃ thyrotoxicosis is a previously unreported clinical presentation of thyroid hemiogenesis.

Introduction

Thyroidal hemiogenesis is a congenital anomaly in which one lobe of the thyroid fails to develop, and was first described by Marshall in 1895. The more recent literature suggests that aplasia of one lobe of the thyroid gland is rare and is diagnosed only because of the concurrence of hyperthyroidism, adenocarcinoma and colloid goitre (Burman, Adler and Wartofsky, 1975; Hamburger and Hamburger, 1970). Although hemiogenesis of the thyroid gland is a benign condition, lack of awareness of its existence may lead to an incorrect diagnosis.

The purpose of this report is to describe a patient with hemiogenesis of the thyroid gland who also presented with T₃ toxicosis. To the authors' knowledge this is the first such report in the literature.

Case report

A 35-year-old woman had been in good health until April 1978, when she began to notice increased nervousness, anxiety, diarrhoea, heat intolerance, palpitation and enlargement of the right lateral region of the neck. She denied weight loss. There was no history of excessive iodine intake or ingestion of medications. There was no family history of thyroid disease.

On physical examination, blood pressure was 160/100 mmHg, pulse rate 116/min. The patient

was anxious, with warm and moist skin and a fine tremor of the hands. There was no lid-lag and proptosis. Examination of the thyroid revealed an enlarged, smooth and easily palpable right lobe. The connecting isthmus was not palpable. No thyroid tissue was palpable where the left lobe should have been. No bruits or venous murmurs were heard. On clinical grounds the patient was thought to be hyperthyroid.

The total serum T₄ measured by radio-immunoassay (RIA) was 126 nmol/l (normal 58-161) (Chopra, 1972), serum T₃ measured by RIA (Maciel *et al.*, 1979) was 5.4 nmol/l (normal 1.10-3.23) and T₃ resin uptake (Braverman, Foster and Mead, 1967) was 35% (normal 25-35%). Thyroxine-binding globulin (TBG) concentration was measured by the method of Roberts and Nikolai (1969) and the result was normal: 23.4 µg/dl (normal 14-28). The ¹³¹I thyroid uptake was 43% in 2 hr (normal 5-12%) and 60% in 24 hr (normal 18-50%). The thyroid scintiscan revealed homogeneous uptake over the right lobe and an absence of radioactivity in the left lobe area (Fig. 1).

A TSH stimulation test (bovine TSH-THY-ROPAR, Armour 10 units was given intramuscularly for 5 consecutive days before the ¹³¹I uptake and scintiscan) revealed a 41 and 50% ¹³¹I uptake in 2 and 24 hr respectively; the scintiscan was similar to the previous one, with an absent left lobe. A T₃ suppression test (100 µg T₃ was given by mouth for 10 consecutive days before the ¹³¹I uptake and scintiscan) demonstrated no suppression (39 and 60% ¹³¹I uptake in 2 and 24 hr respectively). The patient was also submitted to a TRH stimulation test (200 µg TRH was given i.v. in bolus). TSH serum concentrations at 30, 60, 90 and 120 min were less than 1.5 µu./ml.

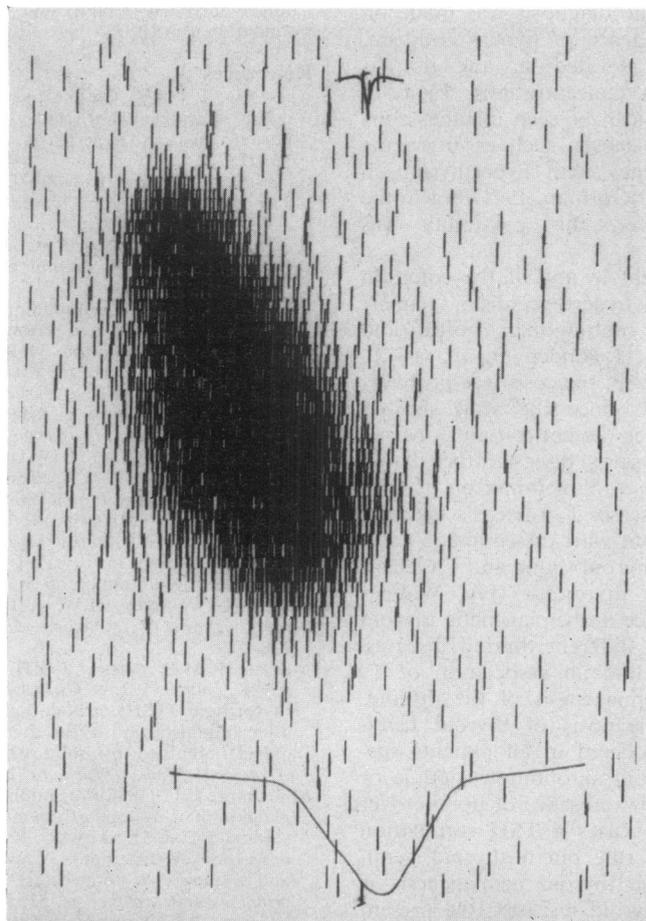


FIG. 1. Thyroid scintiscan revealing homogenous uptake over the right lobe and an absence of radioactivity on the left lobe.

A diagnosis of T_3 thyrotoxicosis was established and therapy with methimazole (30 mg/day) was started. The patient became euthyroid in 45 days and the drug was gradually reduced and finally discontinued after 6 months. The patient is currently in remission.

Discussion

Aplasia of one lobe of the thyroid gland is an unusual condition. Hamburger and Hamburger (1970) found hemiagenesis of the thyroid in 4 of more than 7000 patients with various thyroid disorders. Harada, Nishikawa and Ito (1972) reported 7 cases out of 12 456 surgical thyroid specimens seen over a 13-year period. In all 7 cases the patients were female and 5 of them had classical hyperthyroidism.

As in the present patient, the agenesis seems to occur with a significantly greater frequency on the

left side, as in the 7 patients of Harada *et al.* (1972), 3 of 4 patients found by Hamburger and Hamburger (1970), all 7 patients described by Andreew (1968), 2 of the 3 patients of Burman *et al.* (1975), all 4 patients of Hartemann *et al.* (1976), 3 of 4 patients of Mortimer, Tomlinson and Rosenthal (1981), and all 4 patients of Melnick and Stemkowski (1981).

The actual incidence of thyroid hemiagenesis is unknown, since in most of the cases the diagnosis was 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. Thus, of a total of 30 published cases of this anomaly, 9 occurred in association with hyperthyroidism, 2 in association with benign goitre and 2 in association with carcinoma.

This patient's thyroid studies fulfill the criteria

for T_3 thyrotoxicosis. The diagnosis was made on the basis of clinical evidence of hyperthyroidism, with normal total T_4 , elevated T_3 and normal thyroid-binding globulin concentrations. Elevated T_3 with normal total T_4 can be seen in other conditions than T_3 thyrotoxicosis, such as thyroxine binding globulin deficiency with hyperthyroidism (Wahner, Emstander and Gorman, 1971) but in the present patient, however, this possibility was ruled out.

T_3 toxicosis can present in any of the common forms of conventional hyperthyroidism, namely Graves' disease, toxic multinodular goitre and autonomous adenoma (Hollander *et al.*, 1972). In the present patient the T_3 toxicosis was probably due to Graves' disease, since the scan showed homogeneous uptake over the entire right lobe, an observation suggesting Graves' disease rather than a hot nodule. The response to methimazole is also in favour of Graves' disease. T_3 toxicosis has also been observed in patients with disseminated follicular carcinoma of the thyroid (Sung and Cavalieri, 1973) and Hashimoto's thyroiditis (Ivy, Wahner and Gorman, 1971). Since an hemigenetic thyroid can also be involved in different thyroid diseases, it is not surprising to find an association of T_3 hyperthyroidism and hemigenesis of the thyroid.

In conclusion, the diagnosis of thyroid hemigenesis should be considered in all patients suspected of having a thyroid autonomous nodule or when a thyroid scan shows absence of uptake over an entire lobe. In such cases a TSH stimulation test should be done to rule out a thyroid hemigenesis. Frequently, the thyroid hemigenesis is associated with other thyroid diseases; the present authors demonstrated that T_3 toxicosis can be another of the multiple clinical forms of thyroid hemigenesis.

Acknowledgments

We are grateful to the National Pituitary Agency, Bethesda, Md, U.S.A., and the National Institute for Biological Standards and Control, London, England, for their kindness in supplying the reagents for TSH radio-immunoassay. This study was supported by the National Research

Council of Brazil (CNPq) and São Paulo State Research Foundation (FAPESP).

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