

Graves' Thyrotoxicosis and Moyamoya Disease

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ABSTRACT: Cerebrovascular accidents due to Moyamoya disease, a disorder characterized by arterial stenosis at the base of the brain accompanied by typical net-like collateral vessels, occurred in two young Japanese women with Graves' disease when they were in thyrotoxicosis. In one patient, a second attack of cerebral infarction occurred with the recurrence of thyrotoxicosis. Association of Moyamoya disease and Graves' thyrotoxicosis is rare and the pathogenetic relationship is discussed.

RÉSUMÉ: Thyrotoxicose de Graves et maladie de Moyamoya. Deux jeunes femmes japonaises, atteintes de la maladie de Graves, ont subi un accident cérébrovasculaire dû à la maladie de Moyamoya, une affection caractérisée par une sténose artérielle à la base du cerveau accompagnée par des vaisseaux collatéraux disposés typiquement en filet, alors qu'elles étaient en thyrotoxicose. Chez une patiente, un deuxième épisode d'infarctus cérébral est survenu lors d'une récurrence de la thyrotoxicose. L'association de la maladie de Moyamoya et d'une thyrotoxicose de Graves est rare et nous discutons de la relation pathogénique entre ces deux maladies.

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Moyamoya disease is a cerebrovascular disorder characterized by bilateral stenosis or occlusion of the distal segments of the internal carotid arteries accompanied by typical net-like collateral vessels.¹ The pathogenesis of Moyamoya disease is unknown, but hyperactivity of cervical sympathetic nerves may contribute to the stenosis of cerebral arteries.²

Many symptoms of thyrotoxicosis are similar to those of hyperactivity of the sympathetic nervous system, and thyroid hormones are thought to enhance the effects of sympathetic nervous activity.³ We report here two cases of Moyamoya disease associated with thyrotoxicosis due to Graves' disease.

Case Reports

Patient 1

The patient was a 26-year-old Japanese housewife. When she was 21 years old, the patient developed severe headache, muscle weakness and sensory disturbance in the right upper limb, cramp in the right side of her face and walking disturbance. A few months later, she was diagnosed at a hospital as having hyperthyroidism. The hemiparesis was improved along with the normalization of the thyrotoxicosis by an antithyroid drug. Five years later, after the patient had stopped medication, hyperthyroidism developed again in association with high grade fever (38-39°C), right sided headache, muscle weakness and numbness

in the left upper limb. She was admitted to Ito Hospital (Thyroid clinic). On admission, laboratory data of her thyroid function showed total triiodothyronine (T3) of 7.7 nmol/L, free thyroxine (FT4) of >66 pmol/L, thyroid stimulating hormone (TSH) of <1.0 mU/L, and she was diagnosed as having Graves' disease. Neurological examination revealed left hemiparesis and speech disturbance. Brain CT revealed findings that resembled A-V malformation and a low density area in the right fronto-parietal region (Figure 1a). One month later, she was admitted to Showa University Hospital for further examination. Cerebral angiography showed the typical net-like vessels at the base of the brain that led to a diagnosis of Moyamoya disease (Figure 1b). Left hemiparesis and speech disturbance were alleviated along with the normalization of thyrotoxicosis by antithyroid drug therapy.

Patient 2

The patient was a 22-year-old Japanese woman. When she was 17 years old, she developed swelling of the neck, exophthalmos, hyperhidrosis and body weight loss. At the age of 22, she suffered from left hemiparesis and motor aphasia, and was admitted to a hospital. Brain CT showed a low density area in the right temporo-parietal region. Diagnosis of Moyamoya disease was made after cerebral angiography revealed a network of vessels at the base of the brain. Laboratory data of thyroid function were as follows: T3 of 5.5 nmol/L, T4 of 265 nmol/L, TSH of 1.0 mU/L. She was started on an antithyroid drug, but stopped taking the medicine some months later. Five years later, thyrotoxicosis recurred and she was started again on antithyroid drug, but her thyroid function remained high. She was further examined at the Showa

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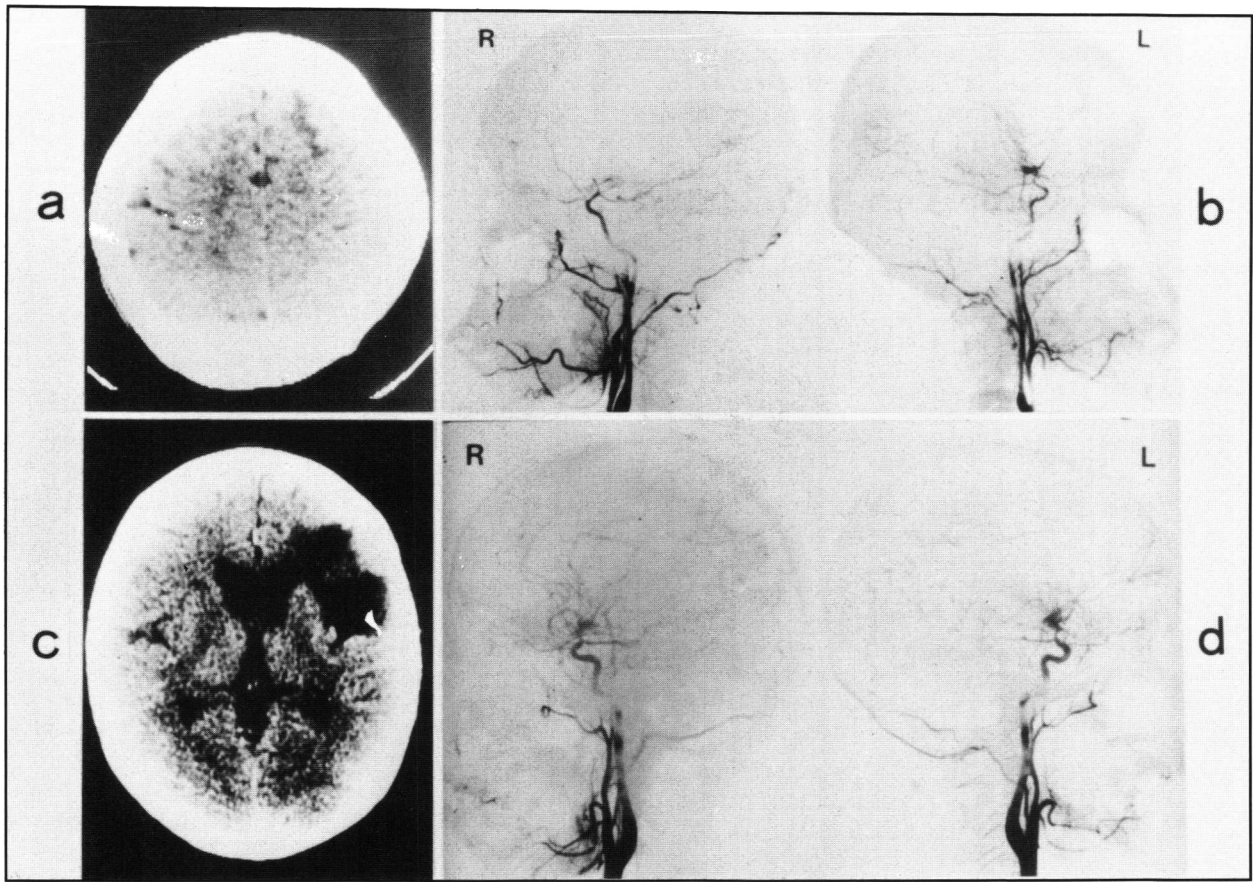


Figure 1 — Brain computed tomogram (CT) of Patient 1 shows a low density area (upper left (a)). Brain CT of Patient 2 shows a A-V malformation like appearance (lower left (c)). Cerebral angiograms of Patient 1(b) and Patient 2(d) reveal moyamoya net-like collateral vessels (right figures).

University Hospital. She had exophthalmos, swelling of the tonsils and a large goiter when she was admitted. A diagnosis of Graves' disease was made because of the thyrotoxicosis and high titer of TSH receptor antibodies. Brain CT showed low density areas in the right frontal, temporal and parietal lobes (Figure 1c). Cerebral angiography showed narrowing of both internal carotid arteries, and net-like vessels at the base of the brain (Figure 1d), which confirmed a diagnosis of Moyamoya disease.

DISCUSSION

Moyamoya disease was first reported in Japan in 1963,⁴ and as many as 2,000 cases have since been identified.⁵ To our knowledge, coexistence of Moyamoya disease and thyrotoxicosis has never been reported in any English language journal. However, coexistence of these two diseases may not be extremely rare, since 3 cases of thyrotoxicosis or high serum level of thyroid hormones with cerebrovascular Moyamoya phenomenon were reported at the annual meeting of the Japanese Neurological Association.⁶

Although relations between Graves' disease and cerebrovascular accident in Moyamoya disease are obscure, coexistence of both diseases is noteworthy. First, these cerebrovascular accidents occurred when the patients were thyrotoxic. It has been known that cerebral blood flow increases in thyrotoxicosis.⁷ Alterations of cerebral hemodynamism in thyrotoxicosis may have been the trigger of the vascular attack in these patients.

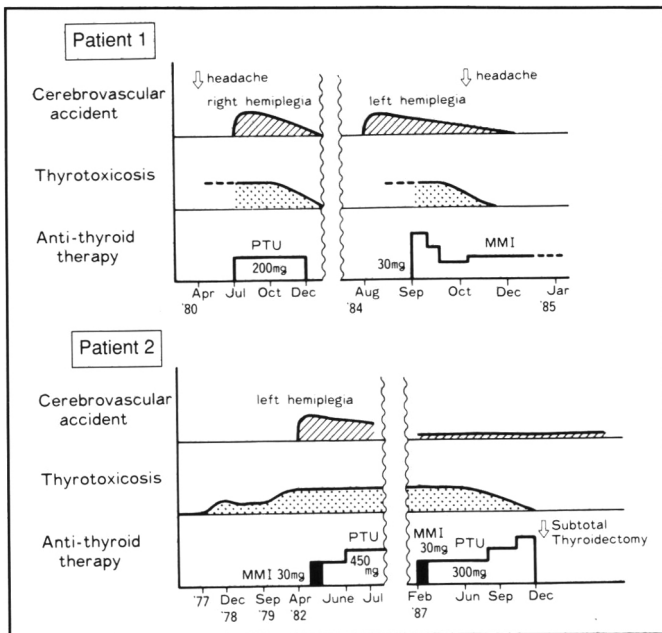


Figure 2 — Relation between cerebrovascular accidents and thyroid status of the patients.

Furthermore, thyrotoxicosis may involve the formation of abnormal cerebral vessels. Suzuki and his coworkers proposed a hypothesis that regional sympathetic nervous stimulation may contribute to the pathological changes of the carotid arteries.² Sympathetic nervous tone itself is not accentuated in thyrotoxicosis, but thyroid hormones are thought to increase sensitivity to the sympathetic nervous system. Thyrotoxicosis might have accelerated the formation of abnormal cerebral vessels. Further investigation is needed.

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