

Case Report

Neurologic Improvement without Angiographic Improvement after Antithyroid Therapy in a Patient with Moyamoya Syndrome

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Moyamoya disease with special complications, including Graves' disease, is called as moyamoya syndrome. A 22-year-old Japanese woman had left middle cerebral artery (MCA) territory infarction complicated with Graves' disease. She had right-sided hemiparesis that deteriorated on day 8 with the infarct growth and thyrotoxicosis. On angiogram, the left MCA was occluded at the origin without moyamoya vessels. Positron emission tomography (PET) revealed misery-perfusion phenomenon in the left MCA territory. After initiation of the antithyroid therapy, her hemiparesis became milder. Seventeen months later, her thyroid function was normalized and net-like collateral moyamoya vessels proliferated in the left MCA territory. Misery-perfusion phenomenon persisted on PET. This report is unique in the point of neurologic recovery of the moyamoya patient right after initiation of anti-thyroid medication without radiological improvement. **Key Words:** Moyamoya syndrome—Graves' disease—antithyroid therapy.

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Case Report

A 22-year-old Japanese woman was admitted to our hospital because of sudden right-sided motor weakness. She was obese, exophthalmic, and the thyroid lobes were enlarged with hyperthyroidism (free tri-iodothyronine 12.6 pg/mL, free thyroxine 6.7 ng/dL, thyroid-stimulating

hormone <.01 µIU/mL, thyroid-stimulating hormone receptor antibody 15.8 IU/L thyroid-stimulating antibody (TS-Ab) 885%). She had aphasia, left conjugate deviation, and right-sided hemiparesis and sensory loss. In the prone position, she could not lift the right arm and could lift the right leg slightly. Brain MRI showed a fresh infarct in the left middle cerebral arterial (MCA) territory (Fig 1, A). On angiography, the left MCA was occluded at the origin, and the right anterior cerebral artery (ACA) was stenotic (Fig 1, B,C). Positron emission tomography revealed the misery-perfusion phenomenon in the left MCA territory. On a thyroid scintigram, a diffuse goiter and high uptake of technetium-99m were seen (Fig 1, D). She was diagnosed as having an ischemic stroke and Graves' disease.

Intravenous injection of edaravone and heparin was initiated. On day 8, she developed complete paralysis of the right leg, with infarct growth. On day 23, she developed thyrotoxicosis (free tri-iodothyronine 30.2 pg/mL, free thyroxine > 7.7 ng/dL). Oral thiamazole was started immediately, and the dosage was increased on day 42. She

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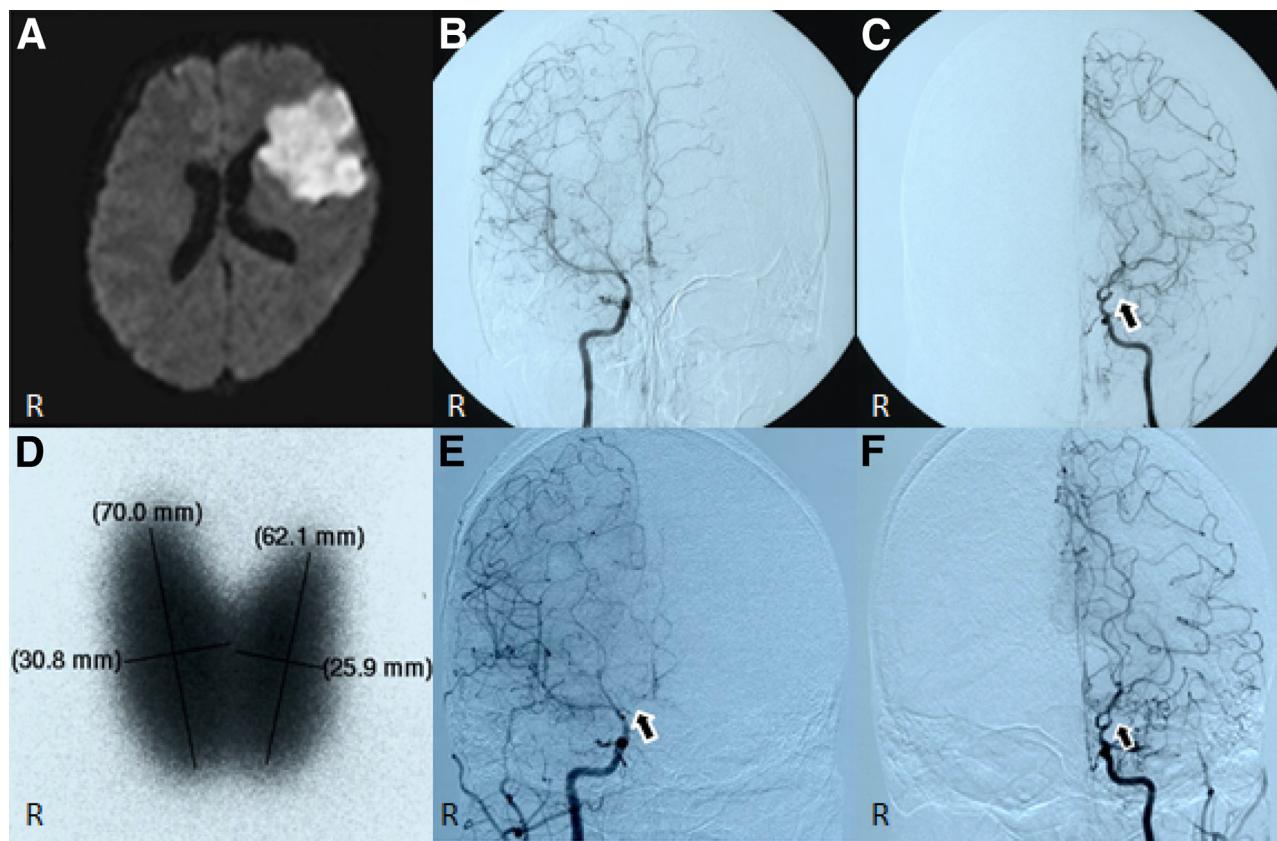


Figure 1. (A) Brain magnetic resonance imaging diffusion-weighted image on admission showing a fresh infarct in the left middle cerebral artery (MCA) territory. (B and C) Cerebral angiogram on admission showing stenosis at the A2 portion of the right anterior cerebral artery (B) and occlusion at the proximal M1 portion of the left MCA (C). (D) Thyroid scintigram on admission showing diffuse high uptake of technetium-99m in the bilateral thyroid lobes. (E and F) Cerebral angiogram 17 months after stroke showing the progression of stenosis at the right A1 and A2 (E), occlusion at the left M1, and newly developed moyamoya vessels (F).

suddenly lifted her right leg slightly on day 25, and she bent the right knee and lifted the right arm on day 43.

Seventeen months were required for normalization of thyroid function. Then, on angiography, net-like collateral moyamoya vessels were seen to have proliferated in the left MCA territory, and the right ACA stenosis grew more severe (Fig 1, E,F). There was no steno-occlusion in the vertebrobasilar circulation. The misery-perfusion phenomenon persisted on positron emission tomography. Superficial temporal artery–MCA bypass surgery was performed. She has been free from recurrent stroke for 3 years since initial stroke onset.

Discussion

Progressive steno-occlusion of the bilateral distal internal carotid arteries, proximal ACAs, and/or MCAs, in conjunction with specific complications, including Graves' disease, is called moyamoya syndrome. Excessive thyroid hormones may cause stroke by increased sensitivity of the sympathetic nervous system and alteration of cerebral vascular reactivity¹ and augmentation of cerebral metabolism and oxygen consumption.² Of more than 30 reported cases of moyamoya syndrome with Graves' disease,^{1–9} few

showed symptomatic recovery with amelioration of arteries' stenosis after antithyroid therapy.^{8,9}

This report is unique in that neurologic recovery occurred right after initiation of antithyroid medication without radiological improvement. Radiologically undetectable recovery of microcirculation might have resulted in neurologic improvement.

References

1. Kushima K, Satoh Y, Ban Y, et al. Graves' thyrotoxicosis and moyamoya disease. *Can J Neurol Sci* 1991;18:140–142.
2. Tendler BE, Shoukri K, Malchoff C, et al. Concurrence of Graves' disease and dysplastic cerebral blood vessels of the moyamoya variety. *Thyroid* 1997;7:625–629.
3. Sasaki T, Nogawa S, Amano T. Co-morbidity of moyamoya disease with Graves' disease. Report of three cases and a review of the literature. *Intern Med* 2006;45:649–653.
4. Im SH, Oh CW, Kwon OK, et al. Moyamoya disease associated with Graves disease: special considerations regarding clinical significance and management. *J Neurosurg* 2005;102:1013–1017.
5. Nakamura K, Yanaka K, Ihara S, et al. Multiple intracranial arterial stenoses around the circle of Willis in association with Graves' disease: report of two cases. *Neurosurgery* 2003;53:1210–1215.

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6. Hsu SW, Chaloupka JC, Fattal D. Rapidly progressive fatal bihemispheric infarction secondary to moyamoya syndrome in association with Graves thyrotoxicosis. *Am J Neuroradiol* 2006;27:643-647.
7. Endo H, Fujimura M, Niizuma K, et al. Efficacy of revascularization surgery for moyamoya syndrome associated with Graves' disease. *Neurol Med Chir* 2010;50:977-983.
8. Wakamoto H, Ishiyama N, Miyazaki H, et al. The stenoses at the terminal portion of the internal carotid artery improved after initiation of antithyroid therapy: a case report. *No Shinkei Geka* 2000;28:379-383.
9. Utku U, Asil T, Celik Y, et al. Reversible MR angiographic findings in a patient with autoimmune Graves disease. *Am J Neuroradiol* 2004;25:1541-1543.