Graves disease

Graves' disease, also known as toxic diffuse goiter and Flajani-Basedow-Graves disease, is an autoimmune disease that affects the thyroid. It frequently results in hyperthyroidism and an enlarged thyroid. Signs and symptoms of hyperthyroidism may include irritability, muscle weakness, sleeping problems, a fast heartbeat, poor tolerance of heat, diarrhea, and weight loss. Other symptoms may include thickening of the skin on the shins, known as pretibial myxedema, and eye problems such as bulging, a condition known as Graves' ophthalmopathy.

About 25% to 80% of people develop eye problems.

A study indicated a higher overall autoimmune disease prevalence in unilateral than in bilateral MoyaMoya Disease MMD. Unilateral MMD may be more associated with autoimmune disease than bilateral MMD. Different pathogenetic mechanisms may underlie moyamoya vessel formation in unilateral and bilateral MMD [1)].

Diabetic ketoacidosis (DKA) is one of the precipitating factors that can evoke a thyroid storm. Thyroid storm may cause cerebral ischemia in Moyamoya disease, which can coexist in patients with Graves' disease.

Case series

2015

320 adult Chinese patients at West China Hospital were diagnosed with MoyaMoya Disease, and 29 were also diagnosed with Graves disease (GD). A total of 170 patients (25 with GD; 145 without GD) were included in this study and were followed up. The mean follow-up was 106.4 ± 48.6 months (range 6-216 months). The progression of the occlusive lesions in the major intracranial arteries was measured using cerebral angiography and was evaluated according to Suzuki's angiographic staging. Information about cerebrovascular strokes was obtained from the records of patients' recent clinical visits. Both angiographic progression and strokes were analyzed to estimate the incidences of angiographic progression and strokes using Kaplan-Meier analysis. A multivariate logistic regression model was used to test the effects of sex, age at MMD onset, disease type, strokes, and GD on the onset of MMD progression during follow-up.

During follow-up, the incidence of disease progression in MMD patients with GD was significantly higher than in patients without GD (40.0% vs 20.7%, respectively; p = 0.036). The interval between initial diagnosis and disease progression was significantly shorter in MMD patients with GD than in patients without GD (p = 0.041). Disease progression occurred in both unilateral MMD and bilateral MMD, but the interval before disease progression in patients with unilateral disease was significantly longer than in patients with bilateral disease (p = 0.021). The incidence of strokes in MMD patients with GD was significantly higher than in patients without GD (48% vs 26.2%, respectively; p = 0.027). The Kaplan-Meier survival curve showed significant differences in the incidence of disease progression (p = 0.038, log-rank test) and strokes (p = 0.031, log-rank test) between MMD patients with GD and those without GD. Multivariate analysis suggested that GD may contribute to disease progression in MMD (OR 5.97, 95% CI 1.24-33.76, p = 0.043).

The incidence of disease progression in MMD patients with GD was significantly higher than that in MMD patients without GD, and GD may contribute to disease progression in MMD patients. The

incidence of strokes was significantly higher in MMD patients with GD than in patients without GD. Management guidelines for MMD patients with GD should be developed. [2].

Eight patients with MMD and GD presenting with cerebral ischemia who were treated by direct bypass. Thyroid hormones [free thyroxin (fT4) and free triiodothyronine (fT3)], thyroid-stimulating hormone (TSH), and TSH receptor antibody (TRAb) were measured sequentially. After thyrotoxic conditions were medically optimized, revascularization surgery was performed by superficial temporal artery-middle cerebral artery (STA-MCA) double bypass in all cases. Clinical outcomes were estimated by modified Rankin scale (mRS) at discharge and 3 months after surgery.

In six patients with thyrotoxicosis, the fT4, fT3, and TRAb (range) at the onset of cerebral ischemia were 4.81-10.30 pg/ml, 13.08-31.90 pg/ml, and 3.5-83.8 IU/l, respectively. At surgery, mean (range) fT3 and fT4 were optimized to 3.02 (1.01-4.87) pg/ml and 1.09 (0.41-1.68) ng/dl, respectively. In the thyrotoxic cases, it took 70-310 days (mean, 142 days) to optimize thyroid hormones before surgery. There was no neurological aggravation after surgery, and outcome was excellent at 3 months with mRS scores ≤2 in all cases.

For MMD concurrent with GD, optimizing thyroid hormones followed by STA-MCA double bypass was successful to prevent cerebral ischemic events [3].

Case reports

A 16-year-old girl complaining of dizziness and palpitations visited the emergency department and was diagnosed with DKA combined with hyperthyroidism. A thyroid storm occurred 6 h after the start of DKA management. Her Burch and Wartofsky score was 65 points. Right hemiplegia developed during the thyroid storm, and brain magnetic resonance (MR) diffusion-weighted images revealed multiple acute infarcts in both hemispheres. MR angiography showed stenosis of both distal internal carotid arteries and both M1 portions of the middle cerebral arteries, consistent with Moyamoya disease. After acute management for the thyroid storm with methimazole, Lugol solution and hydrocortisone, the patient's neurological symptoms completely resolved within 1 month, and free T4 level normalized within 2 months. Thyroid storm may trigger cerebral ischemia in Moyamoya disease and lead to rapid progression of cerebrovascular occlusive disease. As a simultaneous occurrence of DKA, thyroid storm and cerebrovascular accident in Moyamoya disease highly elevates morbidity and mortality, prompt recognition and management are critical to save the patient's life [4].

1)
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