Case Report

Unilateral Symptomatic Intracranial Arterial Stenosis and Myopathy in an Adolescent with Graves Disease: A Case Report of an High-resolution Magnetic Resonance Imaging Study

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Vascular and muscular involvements in Graves disease (GD) are rare. Here, we report a case of a 17-year-old patient with unilateral symptomatic middle cerebral artery stenosis concurrent with GD and myopathy. He presented with a 1-day history of acute severe right-sided hemiparesis and aphasia and a 3-week history of high metabolic syndrome. The pathogenesis of the stenosis is most likely vasculitis rather than atherosclerosis, based on contrast-enhanced high-resolution magnetic resonance imaging showing concentric wall enhancement. We suggest that lipid storage myopathy is secondary to GD, and it is likely mitochondrial dysfunction or immune dysfunction induced by GD responsible for the myopathy and that magnetic resonance spectroscopy (MRS) is capable of establishing the diagnosis of myopathy. Thus, MRS can be used for follow-up evaluations of the myopathy along with the pathology biopsy. **Key Words:** Adolescent—infarction—middle cerebral artery—Graves disease—muscular diseases—magnetic resonance imaging.

Graves disease (GD), a common autoimmune disorder of the thyroid with genetic susceptibility, is mainly induced by infection and trauma and often results in hyperthyroidism. During the past few years, an increasing number of case reports concerning hyperthyroidism, together with intracranial arterial disease, cerebral infarction, or transient ischemic attack, have been published, suggesting a growing interest in GD and vascular diseases.[1-3] To the best of our knowledge, there has been no report of a youth suffering from an ischemic stroke, GD, and myopathy simultaneously. The present work is likely the first case with these complicated issues and provides an initial exploration of the potential etiology and pathogenesis of the disease.

Case Report

A 17-year-old Asian boy without apparent vascular risk factors initially presented with a 1-day history of acute severe right-sided hemiparesis and aphasia. Previously, the patient had experienced bilateral lower extremity weakness early in the morning 3 times during March and April 2012, which was resolved in approximately 10 minutes after receiving traditional Chinese medical treatment.

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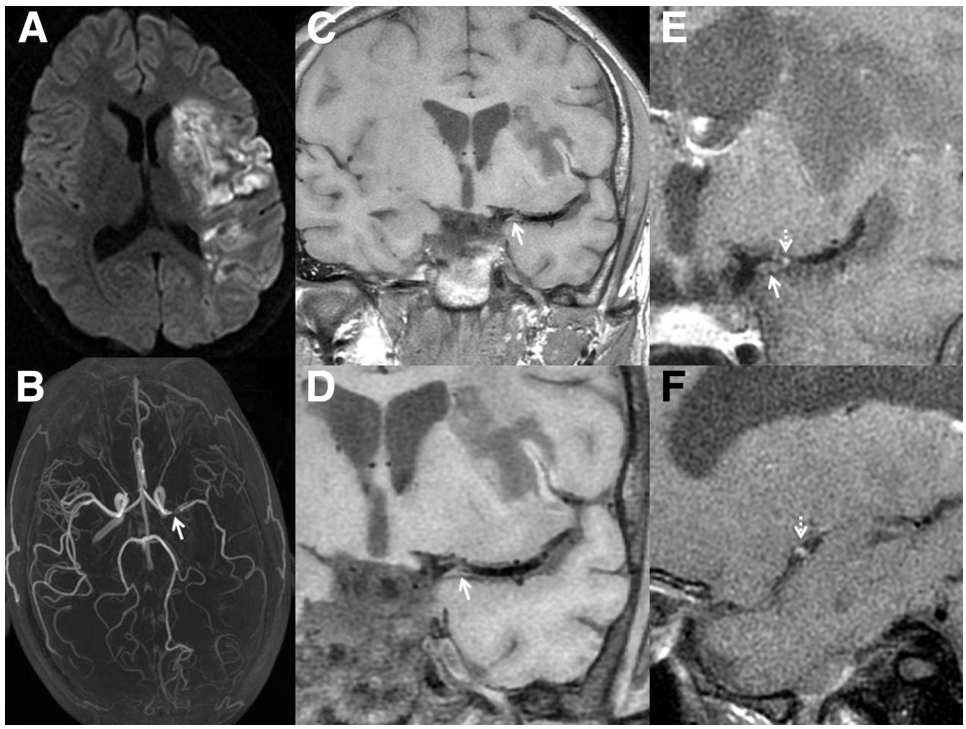

Figure 1. *(A) Multiple infarcts are observed in the distribution of the left middle cerebral artery (MCA) on magnetic resonance (MR) diffusion-weighted imaging. (B) MR angiography shows the obvious severe stenosis located at the M1 segment of the left MCA (arrow). (C) Coronal T1WI high-resolution magnetic resonance imaging shows smooth, concentric wall thickening in the MCA at the onset of the attack (solid arrow). (D) Coronal T1WI with fat suppression demonstrates neither atherosclerosis plaques nor lipid storage existed in the vessel wall (solid arrow). (E) One year later, coronal Magnevist-enhanced T1WI demonstrates concentric wall enhancement (dotted arrow) and the expansion of the lesion (solid arrow) after treatment with methimazole, aspirin, and Lipitor. (F) Sagittal contrast-enhanced T1WI shows concentric wall enhancement more clearly (dotted arrow).*

The patient had been experiencing heat intolerance, sweating, insomnia, and weight loss for 3 weeks. Except for the symptoms listed previously, his prior medical history was unremarkable, and he has no history of inherited disease. The physical examination revealed slight proptosis and a basal metabolic rate increase of .37. In the neurologic examination, he showed mixed aphasia and spastic paralysis. The patient scored 14 points on the National Institute of Health stroke scale. The initial laboratory examination showed hyperthyroidism (serum free thyroxine, 42.25 pmol/L; free triiodothyronine, 13.706 pmol/L; and thyroid-stimulating hormone [TSH], 0.003 mIU/L), and the TSH receptor antibody and antithyroid peroxidase antibody levels were also elevated (5.66 IU/L and 54.44 IU/mL, respectively). Ultrasound imaging of the thyroid displayed a thickened echo and heterogeneous distribution in both lobes. Routine magnetic resonance imaging revealed a massive left middle cerebral artery (MCA) territory infarction, and time-of-flight magnetic resonance angiography and angiography confirmed an identical severe stenosis of the left M1 segment of approximately 1 cm in length (Fig 1, A,B). The vessel wall images of high-resolution magnetic resonance imaging (HRMRI) with fat suppression delineated the stenotic portion of the artery with smooth, concentric wall thickening and without lipid storage (Fig 1, C,D). The patient was diagnosed with an acute cerebral infarction concurrent with GD. In addition to the traditional treatment for the infarction, he received methimazole for the treatment of hyperthyroidism at that time. The patient recovered quickly, and his National Institute of Health stroke scale score decreased to 0 after being hospitalized for a month. Approximately 1 year later, a follow-up HRMRI with black-blood contrast-enhancement T1-weighted sequences showed the expansion of the lesion and concentric wall enhancement in the MCA wall (Fig 1, D,F), despite almost normal clinical and serum examinations. In addition, the patient's initial imaging showed multiple cortical necroses; as a result, we decided to perform a muscle biopsy to determine whether mitochondrial encephalopathy was present. Surprisingly, a suspected diagnosis of lipid storage myopathy (LSM) was proposed. Moreover, the findings from the magnetic resonance spectroscopy (MRS; Fig 2, A) were consistent with the muscle biopsy (Fig 2, B,C). Interestingly, however, no abnormalities were observed in the blood tests (acylcarnitine and amino acids), urine tests (organic acids), electromyography, and genetic detection tests (CPT2, CPT1A, ACADVL, ETFDH, ETFA, ETFB, ACADS, ACADM, SLC25A20, ABHD5, and SLC22A5). Our patient received a repeat MRS examination in 2014, which showed that the myopathy had significantly worsened (Fig 2, D).