Reversible Cerebral Vasoconstriction Syndrome following Guillain–Barré Syndrome: A Rare Complication

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To the Editor: Reversible cerebral vasoconstriction syndrome (RCVS) is well characterized by severe headaches, with or without other acute neurological symptoms, and diffuse segmental vasoconstriction of cerebral arteries, which resolves within 3 months. It has been reported predominantly in middle age, and the syndrome is more common in women than in men.[1] Neuroradiological features of RCVS include convexity subarachnoid hemorrhage, intracerebral hemorrhage, cerebral infarction, and posterior reversible encephalopathy syndrome (PRES).[1] Although the pathological process of RCVS remains uncertain, dysregulation of intracranial arterial tone is thought to be the common underlying mechanism. Up to our knowledge, RCVS occurring in the setting of Guillain–Barré syndrome (GBS) is rarely reported.[2] We describe a GBS patient who develops RCVS following treatment with intravenous immunoglobulin G (IVIG).

A 60-year-old diabetic woman presented with symmetrical numbness and weakness in all the four limbs for 1 week. There was no history of hypertension. On admission, her blood pressure and pulse were 120/70 mmHg and 82 beats/min, respectively. Neurological examination revealed left facial paralysis, flaccid weakness in all the four limbs (magnetic resonance cholangiography Grade 2/5), and disappeared deep tendon reflexes. Light pain and touch sensation was reduced in a stocking distribution, and proprioception was impaired distally. Initial laboratory studies on admission yielded serum potassium 3.2 mEq/L, random blood glucose 7.1 mmol/L, total cholesterol 6.21 mmol/L, and low-density lipoprotein 4.21 mmol/L. Magnetic resonance imaging (MRI) of the cervical spinal cord revealed normal finding. Nerve conduction studies showed prolonged latencies, reduced conduction velocity, and absent F waves with reduction in the amplitude of compound muscle and sensory nerve action potentials. Cerebrospinal fluid analysis (CSF) showed distinctly albuminocytological dissociation with a protein level of 96 mg/dl and a normal cell count. She was diagnosed with GBS and subsequently administrated with IVIG (0.4 g/kg over 5 days). On the 6th day after admission, she complained of blurred vision and dizziness. Brain MRI demonstrated asymmetric hyperintense signal abnormalities in bilateral occipital lobes on fluid-attenuated inversion recovery (FLAIR) and T2-weighted images with no diffusion restriction, consistent with vasogenic edema [Figure 1a-c]. Five days later, she developed bilateral loss of vision. A repeated brain MRI showed bilateral cortical and subcortical hyperintensity involving the parieto-occipital areas on FLAIR and diffusion-weighted images, with a decreased apparent diffusion coefficient [Figure 1d-f], whereas the second neuroimaging findings were in accordance with cytotoxic edema. We, therefore, conducted a cerebral angiography in order to identify the cause of infarction. Computed tomography angiography (CTA) demonstrated diffuse vasoconstriction involving in anterior and posterior circulation [Figure 1i]. Although she received rehabilitation treatment for 4 weeks, the limb weakness still aggravated and her daily activities relied on her family members. After a 3-month follow-up, magnetic resonance angiography (MRA) showed resolution of the diffused segmental narrowing of cerebral arteries [Figure 1j].

Initially, the typical clinical, electrophysiological, and CSF features of our case met the diagnostic criteria for GBS. Several days later, neurological examination showed visual impairment and MRI demonstrated asymmetric vasogenic edema in bilateral occipital lobes, which might suggest a diagnosis of PRES. As deterioration of vision, a repeated MRI showed infarction in bilateral parieto-occipital lobs and CTA revealed diffuse vasoconstriction involving in anterior and posterior circulation. After a 3-month follow-up, MRA revealed resolution of the narrowing cerebral arteries. In addition, based on the detailed comparison between axial MR scans at baseline and at follow-up, blood vessel diameter of cerebral arteries such as bilateral anterior cerebral arteries, bilateral posterior cerebral arteries, and basilar artery indeed dilated [Figure 1g and 1h]. Hence, the diagnosis of RCVS can be established.

Since Calabrese *et al*. first proposed the name and diagnostic criteria of RCVS in 2007, the clinical course, possible triggers,

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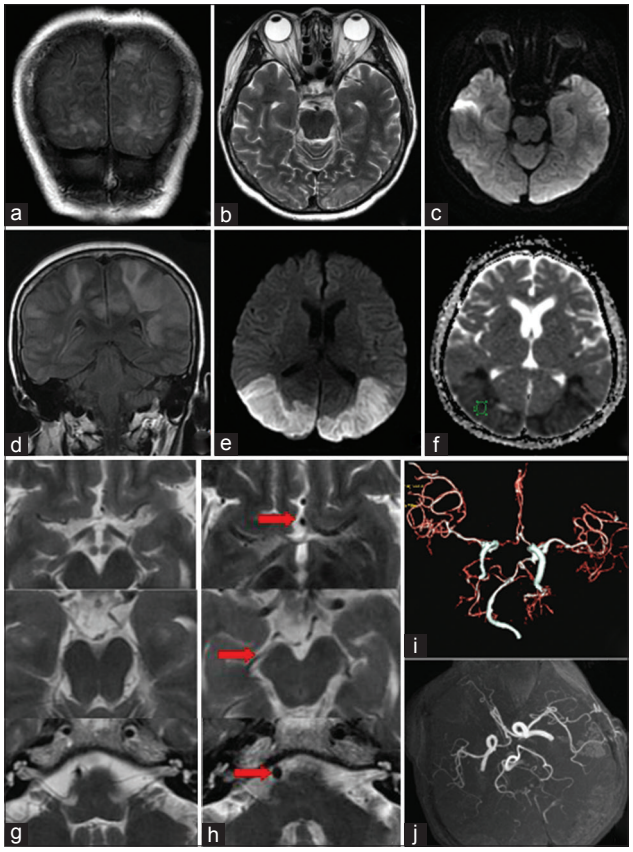

Figure 1: Brain MRI on day 6 after admission demonstrated asymmetric vasogenic edema in bilateral occipital lobes (a-c). On day 11, a repeated MRI showed infarction in bilateral parieto-occipital lobs (d-f). Based on the comparison between axial MR scans at baseline and at 3-month follow-up, blood vessel diameter of cerebral arteries obviously dilated (red arrows) including bilateral anterior cerebral arteries, bilateral posterior cerebral arteries, and basilar artery (g and h). On day 11 after admission, CTA demonstrated diffuse vasoconstriction of intracranial vessels (i). After a 3-month follow-up, MRA showed resolution of the narrowing cerebral arteries (j). MR: Magnetic resonance; MRI: MR imaging; MRA: MR angiography; CTA: Computed tomography angiography.

and neuroimaging features of RCVS have been extensively characterized. They put more emphasis on the importance of potential triggers, time course, neuroimaging features, and excluding other etiologies.[3] Although thunderclap headache is the most common symptom, it is not necessary for the diagnosis. A diverse group of possible triggers have been proposed, including vasoactive agents, pregnancy and postpartum state, metabolic and endocrine disturbances, autoimmune associations, and vascular abnormalities.[3] Despite the pathophysiologic mechanism of RCVS remains unknown, it is considered that dysfunction of cerebral vascular tone plays a crucial role. Several hypotheses have been speculated to explain the underlying pathophysiologic mechanism of RCVS. First, RCVS occurring in the setting of blood pressure fluctuation, use of sympathomimetic vasoactive substances, and pheochromocytoma supports the role of sympathetic overactivity in its pathogenesis. Second, a significant association between RCVS and PRES suggests the importance of endothelial dysfunction in the disease. Nowadays, a leading theory of the pathophysiological changes underlying PRES purports that rapidly developing hypertension causes hyperperfusion, which could lead to endothelial dysfunction and breakdown of the blood–brain barrier. PRES-like reversible cerebral edema is encountered in anywhere from 9% to 38% of patients with RCVS, while most patients with PRES (>85%) demonstrate some element of RCVS-like cerebral vasoconstriction when conventional angiography is performed.[4] Furthermore, various hormonal and biochemical factors, such as estrogen, endothelin-1, serotonin, nitric oxide, and prostaglandins, have been found to correlate with disease severity.[4]

Up to our knowledge, previously reported cases of RCVS complicating with GBS are rarely documented in the literature. Dysfunction of cerebral autoregulation induced by hypertension is considered as a presumptive mechanism for contributing to RCVS.[2] As arterial hypertension, probably due to autonomic dysfunction, is frequently associated with GBS, it may be an important provoking factor of RCVS. Furthermore, increased levels of cytokines or other GBS-linked factors may also play a role in the pathogenesis. In addition, IVIG therapy has been independently associated with complications such as PRES and ischemic stroke which may result from increased serum viscosity, platelet activation, intravascular hypercoagulopathy, and necrotizing microangiopathy.[5] We, therefore, speculate that GBS-related dysfunction of cerebral autoregulation and IVIG therapy are potential triggers for developing RCVS in our patient.

In conclusion, the pathophysiologic mechanism of RCVS occurring in the setting of GBS remains uncertain. Both the clinician and radiologist should be aware of a concurrent occurrence of GBS with RCVS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initial will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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