INTRODUCTION

Intravenous immunoglobulin (IVIg) is a preparation fractionated from pooled human plasma, to contain primarily immunoglobulin G (IgG). IVIg is increasingly used as an effective treatment for an expanding list of autoimmune diseases. Most adverse effects of IVIg are mild and transient and IVIg is considered generally safe [1]. Thromboembolic complications are recognised but rare, and have been reported to occur in patients with vascular risk factors [2]. There have been only five previous reports of cerebral infarction following IVIg therapy, with reported latencies of 2 to 10 days following infusion [3]. We report the occurrence of cerebral infarction after a longer latency following IVIg therapy for Miller Fisher syndrome (MFS) in a patient with no previous vascular risk factors.

A previously well, 44-year-old Sri Lankan man presented with perioral and acral paraesthesiae for 3 days associated with disabling episodic frontal headaches and vomiting. He was afebrile and there was no recent history of fever or symptoms of infection. His general and neurological examinations were normal. His blood counts, inflammatory markers (erythrocyte sedimentation rate, C-reactive protein), renal and liver function tests were normal. A non-contrast-enhanced computed tomography scan of his brain showed no abnormality. Two days after admission to hospital, he developed a right lower motor neurone (LMN) facial paralysis, left perioral and acral paraesthesiae for 3 days associated with disabling episodic frontal headaches and vomiting. His pupils were 3mm bilaterally and reacting to light. Muscle power in his upper and lower limbs was 4+/5 and all deep tendon reflexes were easily elicited. A day later, he developed bilateral LMN facial paralysis, bilateral complete external opthalmoplegia with bilateral partial ptosis and bilateral dilated pupils with no reaction to light. His muscle power and tendon reflexes remained unchanged, but he was ataxic. His vital lung capacity was 2000mL. Contrast-enhanced magnetic resonance imaging and magnetic resonance angiography (MRA) of his brain, and electroencephalogram (EEG) were normal. Nerve conduction studies showed focal segmental demyelination with sural sparing. His cerebrospinal fluid (CSF) protein was elevated at 207mg/dL, with no associated cells in the CSF. He was treated with IVIg at 0.4g/kg/day (36g/day) for 5 days. Two days later, he was noted to have global areflexia. He had evidence of syndrome of inappropriate secretion of antidiuretic hormone and required fluid restriction for correction of electrolytes. His blood pressure showed fluctuations from 180/100mmHg to 100/80mmHg and he had a persistent tachycardia. From day 4 of IVIg, he showed improvement in general health, eye movements, facial weakness and incoordination. He was discharged from hospital 11 days after admission. Since he had several high blood pressure readings he was prescribed telmisartan 40mg twice a day.

On review 3 weeks later, he appeared well with normal eye and facial movements and normal coordination, but complained of persistent headache of 2 days. Optic fundi were normal. His muscle power was almost 5/5 but he had global areflexia. He was noted to have had low blood pressure recordings on home monitoring of 100 to 110/60 to 80mmHg and the telmisartan was reduced to once daily with the proviso of stopping completely after further monitoring. On returning home after review, he had difficulty in expressing speech and had complained of worsening headache. He was admitted to hospital the next day with recurring secondary generalised seizures and was found to have expressive aphasia and a right homonymous hemianopia. His blood pressure was 100/60mmHg. Brain imaging showed evidence of a left peri-ocipital infarct with haemorrhagic transformation (Figure 1) and the EEG showed left posterior sharp wave discharges. The MRA and venogram were normal. An electrocardiogram, echocardiogram, blood investigations including thrombophilia screening (activated partial thromboplastin time, prothrombin time/international normalised ratio, thrombin time, bleeding time/clotting time and platelet count), plasma glucose and lipid profile, and carotid duplex scan were normal. He was treated with intravenous midazolam, oral sodium valproate and clozapam for seizures; the telmisartan was omitted and intravenous saline was given to restore his blood pressure to 130/80mmHg. He did not have further seizures but complained of increasing headaches, which subsided with 2 days of mannitol and intravenous dexamethasone. He was discharged from hospital 3 days later and had a modified Rankin score of 2 on admission.
Deep Cerebral Venous Thrombosis  Hypointense signal in areas of infarct Hyperintense signal in areas of infarct Restricted diffusion in case of acute or subacute infarct Filling defect on MRV, most commonly in straight sinus No enhancement Wernicke’s Encephalopathy Hypointense signal lesions Hyperintense signal lesions Possible restricted diffusion No characteristic findings Lesions may enhance Extrapontine Myelinolysis Hypointense signal lesions Hyperintense signal lesions Possible restricted diffusion No characteristic findings Lesions show variable enhancement Wilson’s Disease Hypointense signal lesions Hyperintense signal lesions Possible restricted diffusion in early phase No characteristic findings No enhancement Creutzfeldt-Jakob Disease Hypointense signal lesions Hyperintense signal lesions No restricted diffusion No characteristic findings No enhancement Bilateral Thalamic Glioma Isointense lesions Hyperintense signal lesions No restricted diffusion No characteristic findings No enhancement

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Table 3: Functions of Thalamic Nuclei

<table>
<thead>
<tr>
<th>Function</th>
<th>Thalamic Nuclei</th>
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<tbody>
<tr>
<td>Visual</td>
<td>Medial Geniculate, Pulvinar, Lateral Dorsomedial</td>
</tr>
<tr>
<td>Auditory</td>
<td>Lateral Geniculate</td>
</tr>
<tr>
<td>Somatosensory</td>
<td>Ventral Posteromedial, Ventral Posterolateral</td>
</tr>
<tr>
<td>Motor</td>
<td>Ventral Lateral, Ventral Anterior</td>
</tr>
<tr>
<td>Autonomic control and emotion</td>
<td>Medial Dorsomedial</td>
</tr>
</tbody>
</table>

ABBREVIATIONS

MRA  magnetic resonance angiography
MRI  magnetic resonance imaging
ADC  apparent diffusion coefficient
FLAIR fluid-attenuated inversion recovery
TEE  transesophageal echo
PFO  patent foramen ovale
MRV  magnetic resonance venography
DWI  diffusion weighted images

REFERENCES