To the Editor

Clinically Silent Posterior Reversible Encephalopathy in Guillain-Barré Syndrome

Posterior reversible cerebral vasoconstriction syndrome (PRES) is a clinicoradiologic syndrome characterized by headache, encephalopathy, visual disturbances and seizures, associated with a transient vasogenic oedema predominantly located in the posterior circulation system on brain imaging. The most frequent aetiologies include hypertension, eclampsia, cytotoxic or immunosuppressive drugs, and sepsis. Some cases of PRES have described in association with Guillain-Barré syndrome (GBS). Here we report the case of a woman who presented with a GBS, dysautonomia, and an asymptomatic incidental finding on brain magnetic resonance imaging (MRI) suggestive of PRES, but without any encephalitic symptom suggestive of that syndrome.

Case Report

A 67-year-old woman with a one week history of limb paresis was admitted with moderate neck pain followed by sudden tetraparesis. Neurological examination revealed flaccid weakness in all four limbs and no cranial nerve involvement. Cervical spinal cord MRI revealed no spinal cord abnormalities but led to the incidental discovery of clinically silent PRES (Figure). The patient’s cerebrospinal fluid revealed albuminocytologic dissociation with a protein level of 67 mg/dl and a normal cell count and normal glucose. Nerve conduction studies and electromyography were also consistent with the diagnosis of GBS and she was treated with intravenous immunoglobulin. Serology was negative for HIV, hepatitis B and C, Mycoplasma pneumoniae, Borrelia, Chlamydia, syphilis, Epstein-Barr virus and Cytomegalovirus. Wide fluctuations of blood pressure occurred over the following days despite continuing antihypertensive medication and a transient episode of altered mental functions was observed. However, she did not develop headache, seizure or visual disturbance and brain MRI abnormalities rapidly resolved.

Discussion

This case description focuses on the MRI features of PRES, but yet not fulfilling the clinical definition of this syndrome. Indeed, a transient alteration of mental function does not qualify. One could argue that the term "clinically silent PRES" to describe this case is a misnomer without the appropriate clinical symptoms to accompany the MRI findings, and a more appropriate radiological description might include transient bilateral occipital edema rather than "PRES" for this case. Nevertheless, almost ten cases of PRES in association with GBS have been previously reported in the literature, although most of them were symptomatic. Parmentier et al recently reported similar findings in a 28-year-old woman with GBS and moderately elevated blood pressure. One likely explanation is that brain imaging is rarely performed in GBS, reducing the likelihood that a diagnosis of associated PRES will be made.

Although the underlying physiopathology supporting the association between PRES and GBS remains unclear, the most accepted mechanism is dysautonomia induced by GBS. It is the presumably mechanism in our case, with wide fluctuations of blood pressure leading to failed cerebral autoregulation and vasogenic edema. Alternative hypotheses may include endothelial dysfunction in keeping with elevated cytokines levels in plasma and cerebrospinal fluid, leading to altered integrity of the blood-brain barrier.

In conclusion, this case emphasizes the fact that PRES may be clinically silent, and may therefore be more common than previously reported.

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References