Letter to Editor

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The Cognitive Outcome of Posterior Reversible Encephalopathy Syndrome

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Posterior Reversible Encephalopathy Syndrome (PRES) is a neurotoxic and neurometabolic condition that patients typically experience a combination of headache, altered mental status, visual disturbance and seizures. PRES is associated with a number of systemic medical etiologies, including severe hypertension, pre-eclampsia/eclampsia, allogeneic bone marrow transplantation, organ transplantation, autoimmune disease, cesarean delivery, and high-dose chemotherapy [1].

Although the parietal and occipital lobes are most commonly involved, other lesions locations have been reported in the literature including the frontal lobes, basal ganglia, and brain stem [1]. There are different patterns established in the imaging features: holohemispheric watershed pattern, superior frontal sulcus pattern, dominant parietal-occipital pattern and partial or asymmetric expression of the primary patterns [1].

The posterior circulation is thought to be more susceptible to this type of damage due to less sympathetic innervation of the vertebrobasilar vasculature to protect the parenchyma from rapid increases in arterial blood pressure [2].

The diagnosis of PRES relies mainly on clinical symptoms and radiologic findings [3]. The grading of MR imaging severity may aid in determining the prognosis of patients with PRES according to Karina et al. [4], although resolution of MRI lesions is slower than clinical recovery.

There is a clinical outcome evaluation based on a study of PRES and acute toxic leukoencephalopathy [3]: grade 0, return to baseline clinical condition; grade 1, minimal residual cognitive deficit; grade 2, mild persistent neurologic deficit; grade 3, moderate persistent neurologic deficit; and grade 4, severe outcome, including no improvement, seizures, coma, or death. Glasgow Outcome Scale on day 90 (67 days after onset of severe PRES) was also adopted as the primary evaluation criterion by some investigators, especially in critically ill patients or fatal cases [5].

While the neuroanatomical features of PRES have been investigated, neuropsychological studies are needed to characterize patients' acute and chronic cognitive presentation. There may be subtle, but still debilitating, neuropsychological sequelae that persists even in patients thought to have fully resolved neurologically [6]. Given the frequent involvement of the posterior cortex, complex perceptual impairment is likely to be a lasting feature.

In addition, the presence of cortical, subcortical (e.g., basal ganglia) and white matter lesions in PRES, suggests that psychomotor and cognitive slowing, reductions in working memory, impaired and/or fluctuating attention, and memory impairment are probable features of the syndrome [6,7].

Studies reporting symptom resolution or improvement in PRES within weeks and months of onset [7] must be re-evaluated in light of the failure to include neuropsychological examination in the workup of these patients.

In the literature, there are few studies regarding to cognitive outcome of PRES, except clinical cases reports [6,8].

Initially in these cases, the patients experienced a spike in blood pressure accompanied by typical symptoms, but after weeks-months of "clinical recovery" are associated neurocognitive deficits.

A 30-year-old, Caucasian female with a high school education, after eclampsia with PRES (holohemispheric variant) continued to experience disturbances in depth perception and balance for the next 3 to 4 months and 11 months post-onset her cognitive and functional complaints included decrease attention, trouble multi-tasking, forgetfulness, slowed thinking, and mental inefficiency, as well as difficulties in describing familiar routes/directions to others and disorientation when driving. MRI was performed 19 months post-onset and showed bilateral parietal lobe encephalomalacia [6].

Next patient, is a 51-year-old, Caucasian female with a high school education. This patient suffered from lupus with PRES (parietal-occipital variant) 4 to 6 months post-onset her complaints included mild forgetfulness and reduced stamina. She was severely compromised in judging relative spatial positions and difficulty identifying alternatives approaches to solving problems [6].

A different case, is a 79-year-old, Caucasian female without school education that after mild cognitive impairment with severe PRES (bilateral holohemispheric variant) developed Alzheimer Disease (AD). This patient 1 month post-onset her Mini-Mental State Examination score was 14/30 and 2 month later, a MRI revealed medial temporal atrophy not known and a ¹¹C-PIB PET imaging showed accumulation in the fronto-parietal-temporal lobe, precuneus and posterior cingulate; 6 month post-onset the patient is institutionalized. In the neuropsychological examination were observed alterations in attention, psychomotor speed, immediate auditory memory, episodic memory, working and operational memories, wordbook, verbal IQ and learning capacity. Although this patient was homozygous for the ε4 allele of the apolipoprotein E gene (APOE), this did not influence the onset of AD. This form of presentation has not been described in the literature [8].

Other case report, is a 64-year-old Caucasian female with PRES (parietal-occipital variant). MRI revealed only cerebral atrophy, but in the second week of hospitalisation MRI revealed scattered hyperintense lesions located in the occipital and parietal lobes and both cerebellar hemispheres- picture suggested PRES. After 6 weeks of hospitalization patient condition improved, but did not restore to the premorbid state. The patient was discharged with generalised cognitive impairment [9].

These cases clearly support the need for larger case series and longer follow-up of PRES patients with both neuropsychological and neuroimaging procedures. In addition, if a significant number of PRES patients do have a persisting syndrome of spatial-perceptual and executive dysfunction, these patients must be counseled about potential long-term consequences and offered neurorehabilitation options. Failure to do so based on an a priori assumption about the reversibility of the syndrome may worsen the disablement and impede the adjustment of these patients to a new spectrum of life challenges [6].

Unfortunately PRES is not always a reversible syndrome, it is estimated that such cases percentage is about 5-15% [10]. There is probably an underlying cognitive syndrome in this entity.

In conclusion, physicians should be aware of the cognitive symptoms and imaging findings of PRES to allow to reduce

morbidity of PRES. PRES patients should be submitted to a cognitive examination with follow-up in time.

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