



Autoimmune Encephalitis Mimicking PRES: A Case Report and Literature Review

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Abstract

Posterior Reversible Encephalopathy Syndrome (PRES) is associated with many diverse clinical comorbid, the most common of which are hypertension, eclampsia, renal failure and immunosuppressive treatment. PRES is a neuroimaging-based syndrome and is associated with multifocal vasogenic cerebral edema. Patients with PRES are frequently manifested by headache, seizure, encephalopathy, altered mental function, visual loss, etc. We here report a patient who showed persistent neurologic deficits after PRES and was ultimately diagnosed with Autoimmune Encephalitis (AE).

Case Presentation

A 37-year-old man visited the Department of NCU in First Affiliated Hospital of Kunming Medical University complaining of a general fatigue for 1 week, aggravated with disturbance of consciousness for 1 day. He had no medical history. When admitted to the hospital, his GCS score was 9 (E4/ V1/ M4), he showed a normal blood pressure, no cervical resistance, increased muscular tension, hyperactivity of tendon reflexes and ankle clonus. At presentation, brain MRI showed bilateral multifocal vasogenic edema especially in his bilateral occipital lobes, which is compatible with PRES (Figure 1A). Meanwhile, spot-like microbleeds can be found on SWI mapping (Figure 1C). During admission, follow-up MRI at 1 week showed reversed vasogenic edema in both cerebral hemispheres, with decreased microbleeds on SWI (Figure 1B, 1D). Serum and CSF autoantibody tests using a cell-based immunocytochemistry kit (Shaanxi MYBiotech Co., Ltd.) showed the presence of anti-Casper 2 antibody with titer of 1:3.2 in CSF and 1:100 in blood serum. The patient's CSF profile was otherwise normal (red blood cell 20/ μ L; white blood cell 1/ μ L; protein 0.25 g/L; and glucose 4.5 mmol/L) with no evidence of infection. The patient was diagnosed with anti-Casper 2 AE. After immunotherapy, his symptoms partly resolved. However, due to economic problem, he didn't perform EEG and EMG. After 3 weeks, he was transferred to local hospital.

Discussion

This case exhibits a rare imaging manifestation of anti-Casper 2 encephalitis which was initially well-matched with PRES and associated vasospasm. Generally, PRES is predicted to be both clinically and radiologically reversible and especially has a good prognosis. One of the major causes of PRES is acute hypertension. Nevertheless, patients with normal BP who accompanied with systemic autoimmune disorders can also perform classic PRES radiologically. Jaeho Kim et al. [1] proposed a case of PRES patient with anti-LGI 1 antibody who's MRI showed apparent vasospasm edema. It is presumed that factors such as Tumor Necrosis Factor alpha (TNF- α) and Interleukin- 1 (IL-1), that lead to PRES can activate the immune system and release other cytokines. These cytokines produce expression of adhesion molecules (vascular cell adhesion molecule 1 and intercellular adhesion molecule 1), which cooperate with leukocytes and lead them to produce Reactive Oxygen Species (ROS) and proteases that result in endothelial damage and consequent fluid leakage. TNF- α and IL-1 can furthermore stimulate astrocytes to secrete Vascular Endothelial Growth Factor (VEGF), which deteriorates the form of junctions of the brain vasculature. Thus, these cascades result in vasogenic edema. In conclusion, endothelial hypotheses may be considered the most relevant in PRES patients with autoimmune disorders [2]. Meanwhile, PRES might cause the breakdown of Blood-Brain Barrier (BBB) and the disorganization of brain tissue [3]. In this case, it can be either presumed that BBB breakdown could uncover neuronal membrane antigen epitopes, such as Casper 2, and further induce a process of autoimmune inflammatory encephalitis. More experiments should be

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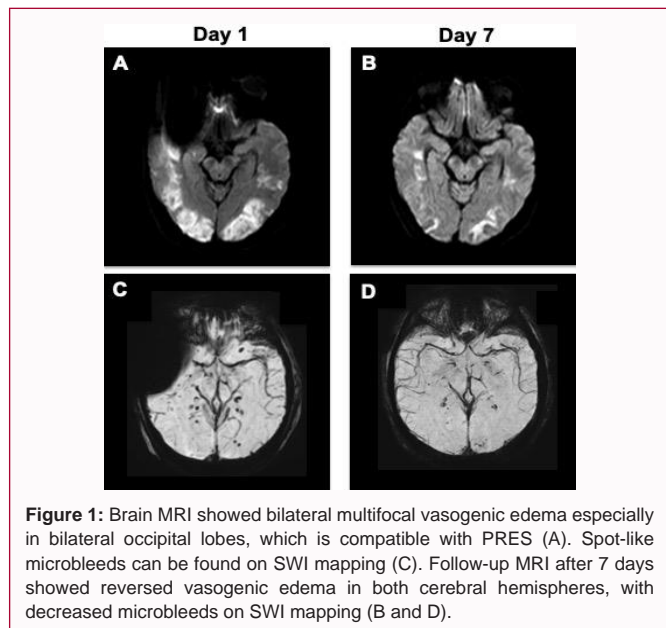
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taken in vitro and in vivo to further test the pathogenesis associated PRES-AE. Clinical observations should also be emphasized when a patient original with PRES neuroradiological features. PRES has been reported in patients with Acute Demyelinating Encephalomyelitis (ADEM), Multiple Sclerosis (MS), other systemic autoimmune encephalitis (e.g. Hashimoto's disease, systemic lupus erythematosus, Behcet's disease), and paraneoplastic encephalitis [4]. In the present case, patient was diagnosed with Casper 2 AE rather than PRES due to the effectiveness of immunotherapy.

In conclusion, AE can mimic PRES radiologically. AE should be further considered when the etiology, clinical manifestations, and course of PRES are atypical. Persistent encephalopathic symptoms, imaging abnormalities in the multiple cortical and subcortical areas, and specifically, autoantibody analysis can be the evidences of AE. Last but not the least, immunotherapy and relevant systemic supportive treatment such as antiepileptic treatment, can lead to a better prognosis.

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