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CASE REPORT

Autoimmune encephalitis with posterior reversible encephalopathy syndrome: A case report

Shu-Juan Dai, Qiu-Jian Yu, Xiao-Yan Zhu, Qun-Zhu Shang, Ji-Bo Qu, Qing-Long Ai

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Abstract

BACKGROUND

Posterior reversible encephalopathy syndrome (PRES) is a neuroimaging-based syndrome and is associated with multifocal vasogenic cerebral edema. Patients with PRES frequently demonstrate headache, seizure, encephalopathy, altered mental function, visual loss and so on. We here report a patient who showed persistent neurologic deficits after PRES and was ultimately diagnosed with autoimmune encephalitis (AE).

CASE SUMMARY

This case exhibits a rare imaging manifestation of anti-casper 2 encephalitis which was initially well-matched with PRES and associated vasogenic edema.

CONCLUSION

AE should be further considered when the etiology, clinical manifestations, and course of PRES are atypical.

Key Words: Autoimmune encephalitis; Posterior reversible encephalopathy syndrome; Neuroimaging; Immunotherapy; Blood-brain barrier; Case report

Core Tip: 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.

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INTRODUCTION

A 37-year-old man visited the Department of neurological care unit in the First Affiliated Hospital of Kunming Medical University complaining of a general fatigue for 1 wk, aggravated with disturbance of consciousness for 1 d. He had no past medical history. When admitted to the hospital, his Glasgow Coma Scale (GCS) score was 9 (E4/V1/M4), he showed a normal blood pressure, no cervical resistance, increased muscle tone, hyperreflexia and hyperactivity ankle clonus. At presentation, brain magnetic resonance imaging (MRI) showed bilateral multifocal vasogenic edema especially in his bilateral occipital lobes, which is compatible with posterior reversible encephalopathy syndrome (PRES) (Figure 1A). Spot-like microbleeds were found on susceptibility weighted imaging (SWI) mapping (Figure 1C). During admission, follow-up MRI at 1 wk showed reduced vasogenic edema in both cerebral hemispheres considerably, with less prominent microbleeds on SWI (Figure 1B and D). Serum and cerebrospinal fluid (CSF) autoantibody tests using a cell-based immunocytochemistry kit (Shaanxi MYBiotech Co., Ltd.) showed the presence of anti-Casper 2 antibody with tilter of 1: 3.2 in CSF and 1: 100 in blood serum. The patient's CSF profile was otherwise normal (red blood cell 20/μL; white blood cell 1/μL; protein 0.25 g/L; and glucose 4.5 mmol/L) with no evidence of infection. The patient finally diagnosed as an anti-Casper 2 autoimmune encephalitis (AE). After intravenous immunoglobulin (IVIG) for 5 d (400 mg/kg/d), his GCS score increased to 10 (E4/V1/M5), which means patient's movement was improved. Unfortunately, due to economic problem, he didn't perform electroencephalography (EEG) and electromyography (EMG). After 3 wk, he was transferred to local hospital.

CASE PRESENTATION

Chief complaints

A general fatigue for 1 wk, aggravated with disturbance of consciousness for 1 d.

History of present illness

There is no special illness except the chief complaints.

History of past illness

There is no history of past illness.

Personal and family history

There is no personal and family history.

Physical examination

The patient's GCS score was 9 (E4/V1/M4), he showed a normal blood pressure, no cervical resistance, increased muscle tone, hyperreflexia and hyperactivity ankle clonus.

Laboratory examinations

Serum and CSF autoantibody tests using a cell-based immunocytochemistry kit (Shaanxi MYBiotech Co., Ltd.) showed the presence of anti-Casper 2 antibody with tilter of 1: 3.2 in CSF and 1: 100 in blood serum. The patient's CSF profile was otherwise normal (red blood cell 20/μL; white blood cell 1/μL; protein 0.25 g/L; and glucose 4.5 mmol/L) with no evidence of infection.

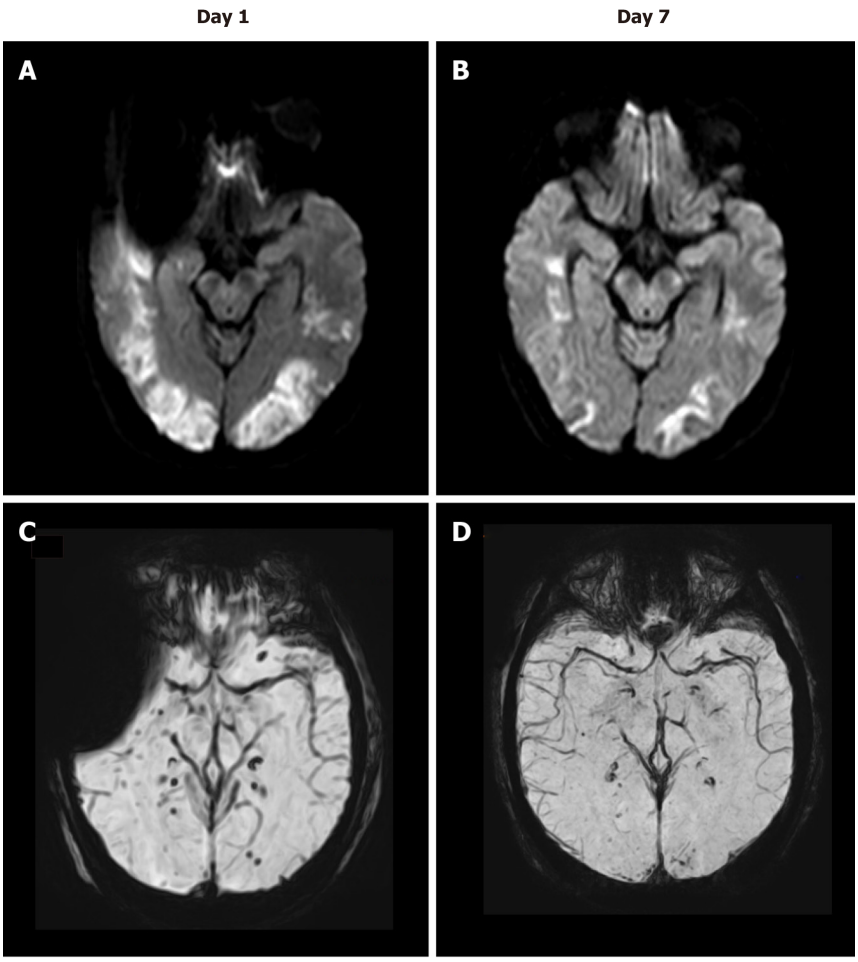

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Figure 1 Brain magnetic resonance imaging showed bilateral multifocal vasogenic edema especially in bilateral occipital lobes. A: Compatible with posterior reversible encephalopathy syndrome; B and D: Follow-up magnetic resonance imaging after 7 d showed significantly reduced vasogenic edema in both cerebral hemispheres, with decreased microbleeds on susceptibility weighted imaging (SWI) mapping; C: Spot-like microbleeds were found on SWI mapping.

Imaging examinations

Brain MRI showed bilateral multifocal vasogenic edema especially in his bilateral occipital lobes, which is compatible with PRES (Figure 1A). Spot-like microbleeds were found on SWI mapping (Figure 1C). During admission, follow-up MRI at 1 wk showed reduced vasogenic edema in both cerebral hemispheres considerably, with less prominent microbleeds on SWI (Figure 1B and D).

FINAL DIAGNOSIS

Anti-Casper 2 AE.

TREATMENT

IVIG for 5 d (400 mg/kg/d).

OUTCOME AND FOLLOW-UP

After intravenous immunoglobulin (IVIG) for 5 d (400 mg/kg/d), his GCS score increased to 10 (E4/V1/M5), which means patient's movement was improved. Unfortunately, due to economic problem, he didn't perform EEG and EMG. After 3 wk, 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. Patients with normal BP who accompanied with systemic autoimmune disorders can also produce features of classic PRES radiologically. Tetsuka and Ogawa[1], proposed a case of PRES patient with anti-LGI 1 antibody whose 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 secret vascular endothelial growth factor (VEGF), which deteriorates the form of junctions of the brain vasculature. 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 dis-organization of brain tissue[3]. In this case, it can be either presumed that BBB breakdown could uncover neuronal membrane antigen epitopes, such as Caspr 2, and further induce a process of autoimmune inflammatory encephalitis. More experiments should be taken *in vitro* and *in vivo* to further test the pathogenesis associated PRES-AE. Clinical diagnosis should also be made cautiously when a patient original has 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 lupuserythematosus, Behcet's disease), and paraneoplastic encephalitis[4].

CONCLUSION

In the present case, the patient was diagnosed as Caspr 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. At last, immunotherapy and relevant systemic supportive treatment such as antiepileptic treatment, can lead to a better prognosis.

FOOTNOTES

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