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Case Report

Posterior reversible encephalopathy syndrome predominantly involving brainstem and spinal cord

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Abstract: We report the case of a 21-year-old female who presented with blurred vision, headache and seizure. Blood pressure is 220/110 mmHg at admission. Laboratory tests revealed renal insufficiency. Brain and cervical magnetic resonance image (MRI) showed small, isolated hyperintensity in the bilaterally cerebellum, occipital lobes and continuous central T2 hyperintensity in the whole cervical spine. After anti-hypertensive drugs and renal dialysis treatment, whose symptom and imaging manifestations improved obviously, which was confirmed diagnosis of posterior reversible encephalopathy syndrome with spinal cord involvement (PRES-SCI). To our knowledge, PRES mainly involved the brainstem and spinal cord instead of supratentorial region has few been described up to now. We present a case here with a brief discussion of clinical and radiological features.

Keywords: Hypertension, posterior reversible encephalopathy syndrome, vasogenic oedema, spinal cord, magnetic resonance imaging

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical-radiological syndrome first reported by Hinchey in 1996 [1]. It is commonly associated with acute hypertension, autoimmune disorders, renal insufficiency, eclampsia and immunosuppressants. Magnetic resonance image (MRI) of brain is characterized by vasogenic edema predominantly in parieto-occipital regions that is gradually resolved within weeks. With increasing use of MRI and more attention to the disorder in clinic, PRES involved the frontal and temporal lobes, basal ganglia, cerebellum, and brainstem were not uncommon. However, PRES mainly involved the brainstem and spinal cord have few describe up to now. We present a case in here.

Case report

A 21-year-old woman admitted to our hospital with blurred vision, headache and seizure. She had no seizure and hypertension in previous history. Physical examination revealed blood pressure at 220/110 mmHg, neck rigidity, mild weakness and brisk knee reflexes in the lower extremities. Laboratory tests revealed renal insufficiency, hyperkalemia. Renal ultrasound showed bilateral renal atrophy. Fundus examination showed bilateral hypertensive retinopathy (**Figure 1A** and **1B**). Lumber puncture had normal cell indices, protein and negative eaquaporin-4-specific immunoglobulin G (NMO-IgG), oligoclonal band (OB).

Brain MRI showed T2-weighted and fluid attenuated inversion recovery (FLAIR) multiple small, isolated hyperintensity in the bilaterally occipital lobes (**Figure 1C**), large confluent hyperintensity in pons and medulla (**Figure 1E**). Spinal MRI revealed a continuous central T2 hyperintensity in the whole cervical spine and the upper bound of the lesion was linked with medulla (**Figure 1G**). Swelling of the medulla and spinal involved were observed.

The clinical symptoms together with physical and laboratory examinations and the radiological findings were suggestive of PRES with spinal cord involvement (PRES-SCI). After the treatment with anti-hypertensive drugs and renal dialysis for eight days, the blood pressure reduced to 150/110 mmHg and the symptoms was rapidly improved. After eighteen days, repeat MRI showed complete resolution of the

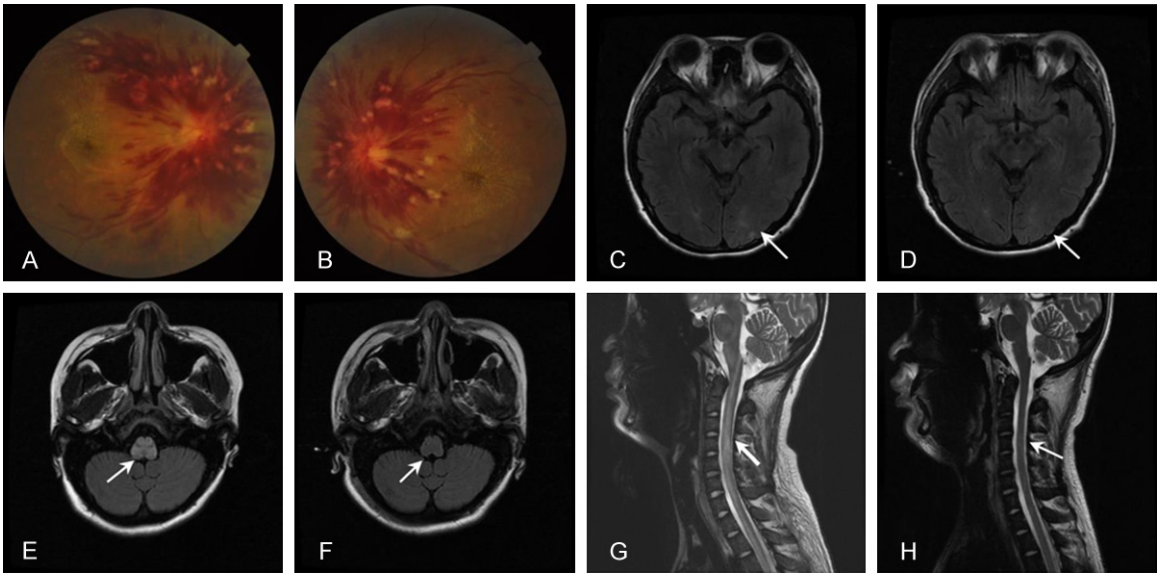

Figure 1. Fundus examination and brain MRI findings. A, B. Ophthalmologic examination displayed bilateral papill edema, flame hemorrhages and cotton-wool spots in the posterior pole. C. Axial fluid-attenuated inversion recovery (FLAIR) showed hyperintensity in the occipital lobes. D. Follow-up MRI at 18 days shows nearly complete resolution in the occipital lobes. E. Axial FLAIR showed large confluent hyperintensity and swelling in the medulla. F. Follow-up MRI at 18 days shows complete resolution of the signal abnormalities in medulla. G. Sagittal T2-weighted showed hyperintensity extending form the pons to thoracic spinal cord. H. Follow-up MRI at 18 days shows complete resolution of the T2 hyperintensity in spinal cord.

abnormalities in the brainstem (**Figure 1F**) and spinal cord (**Figure 1H**), and nearly complete resolution in the occipital lobes (**Figure 1D**), which confirmed diagnosis of PRES-SCI.

Discussion

This case experienced sudden blurred vision, headache and seizure. Blood pressure was extremely high at admission. Laboratory testing revealed renal insufficiency and hypertensive retinopathy. Together with the above-mentioned radiological findings, especially favorable clinical and radiological outcomes, the diagnosis of PRES-SCI is confirmed. The differential diagnoses include myelitis, neuromyelitis optic, central pontine myelinolysis and ischemia. This patient presented with negative NMO-IgG, with no evidence of the demyelination and inflammatory diseases in CSF, It is more important that she had a benign clinical and imaging course without specific treatment, which is unusual in the alternative disorders.

PRES-SCI is a rare PRES form, which had been only reported 12 cases up to now [2-6]. All of them had an exceptionally high blood pressure and had rapidly symptoms improved after aggressively lowering blood pressure, which revealed PRES-SCI was associated with hypertension and was in consistence with finding of the case. Because of PRES-SCI being different from classic PRES in several characteristics, Adam et al. [4] proposed to classify PRES-SCI as a new syndrome. However, some scholars think the reclassification to be unnecessary [7]. They proposed that spinal cord involvement should be regarded as part of the range of PRES because spinal cord involvement can be symptomatic or asymptomatic and it is always associated with features of brain involvement.

The exact mechanism of PRES is not completely understood and the most leading theory is breakdown of blood-brain barrier, endothelial dysfunction and a failure of cerebral blood flow auto-regulation in abrupt fluctuations of blood pressure, which caused hyperperfusion and vasogenic edema [7]. The edema in classic PRES frequently involved predominantly parieto-occipital regions, but the territories shared same vertebrobasilar blood-supply with cervical spinal cord. Therefore, similar autoregulatory dysregulation occurred in the spinal cord too. The spinal cord blood vessels, unlike the posterior cerebral, usually have dense sympathetic

innervations, which may protect the cord from the damage. Secondly, the symptoms and signs of spinal cord involvement were usually mild or absent despite extensive involvement by MRI, which led to neglect of spinal MRI. These probably account for few diagnoses of PRES-SCI so far. Therefore, combining with the current patient findings, the spinal cord MRI should be suggested in PRES even though the symptoms or signs of the spinal were mild.

This patient had two unusual MRI findings, Firstly vasogenic edema of the brainstem and spinal cord were much more serious than that of occipital lobe, which are different from the previous reports [2-6], And repeat MRI showed more complete resolution of the edema in bainstem and spinal than that of occipital lobe. Its pathophysiology mechanism is not clear and need to further research.

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Disclosure of conflict of interest

None.

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