

# Posterior reversible encephalopathy syndrome with spinal cord involvement but without hemisphere lesions

## A case report

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### Abstract

**Rationale** Posterior reversible encephalopathy syndrome (PRES) was termed by Hinchey<sup>[1]</sup> in 1996. Patients have a reversible vasogenic brain edema in imaging and acute neurological symptoms such as headache, seizures, encephalopathy, and visual disturbances when suffering from hypertension, pre-eclampsia/eclampsia, renal failure, immunosuppressive medications, autoimmune disorders, sepsis, thrombocytopenia, hypocalcaemia, alcohol withdrawal, and many other potential causes.<sup>[2,3]</sup> de Havenon A et al<sup>[4]</sup> have proposed a new syndrome named PRES with spinal cord involvement (PRES-SCI). The patients with PRES-SCI have similar symptoms these of PRES. Patients have neurologic signs with the spinal cord involved and lesions in magnetic resonance imaging (MRI) extending to the cervicomedullary junction, usually with extreme elevation in blood pressure and a history of hypertensive retinopathy.<sup>[4]</sup> We administrated a young patient whose condition was consistent with PRES-SCI except for the hemisphere lesions.

**Patient Concerns** A 20-year-old Asian male patient was admitted for a 1 week history of blurred vision and weakness of the limbs. He has had poorly controlled hypertension for 1 year before admission. In emergency room, his blood pressure could raise to 260/140mmHg. Neurological examinations and cerebral spinal fluid tests were negative. The MRI of the brain and spinal cord showed reversible lesions in the medulla and upper cervical spinal cord that extended to the lower thoracic spine.

**Diagnosis** Taking into account the characteristic lesions in the MRI as well as the reversible course upon treatment, he was diagnosed PRES-SCI.

**Interventions** He was treated with medicines for the hypertension.

**Outcomes** His symptoms rapidly improved and finally the lesions on the MRI of the brain and spinal cord disappeared.

**Lessons** Clinicians should suspect PRES-SCI when patients have mild or no neurologic signs accompanied with extreme elevation in blood pressure and lesions in spinal cord. Spinal lesions alone may be a subtype of PRES-SCI.

**Abbreviations:** CT = non-enhanced computed tomography, FLAIR = fluid-attenuated inversion recovery, MRI = magnetic resonance imaging, PRES = posterior reversible encephalopathy syndrome, PRES-SCI = posterior reversible encephalopathy syndrome with spinal cord involvement.

**Keywords:** hypertension, posterior reversible encephalopathy syndrome, spinal cord

## 1. Introduction

Posterior reversible encephalopathy syndrome (PRES) was termed by Hinchey et al<sup>[1]</sup> in 1996. Patients with PRES have

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a reversible vasogenic brain edema in imaging and acute neurological symptoms such as headache, seizures, encephalopathy, and visual disturbances while suffering from hypertension, pre-eclampsia/eclampsia, renal failure, immunosuppressive medications, autoimmune disorders, sepsis, thrombocytopenia, hypocalcaemia, alcohol withdrawal, and many other potential causes.<sup>[2,3]</sup> The magnetic resonance imaging (MRI) findings corresponding to edema are classically located in parieto-occipital lobes, but often the frontal and temporal lobes, basal ganglia, cerebellum, and brainstem are also involved.<sup>[4]</sup>

Thanks to advanced and more readily available imaging modalities, PRES is becoming increasingly diagnosed. de Havenon A et al<sup>[4]</sup> has proposed a new syndrome named PRES with spinal cord involvement (PRES-SCI). Patients with PRES-SCI have similar symptoms these of PRES. Patients have neurologic signs with the spinal cord involved and lesions in MRI extending to the cervicomedullary junction, usually with extreme elevation in blood pressure and a history of hypertensive retinopathy.<sup>[4]</sup> The lesions can be located in the brain or the spinal cord or both.

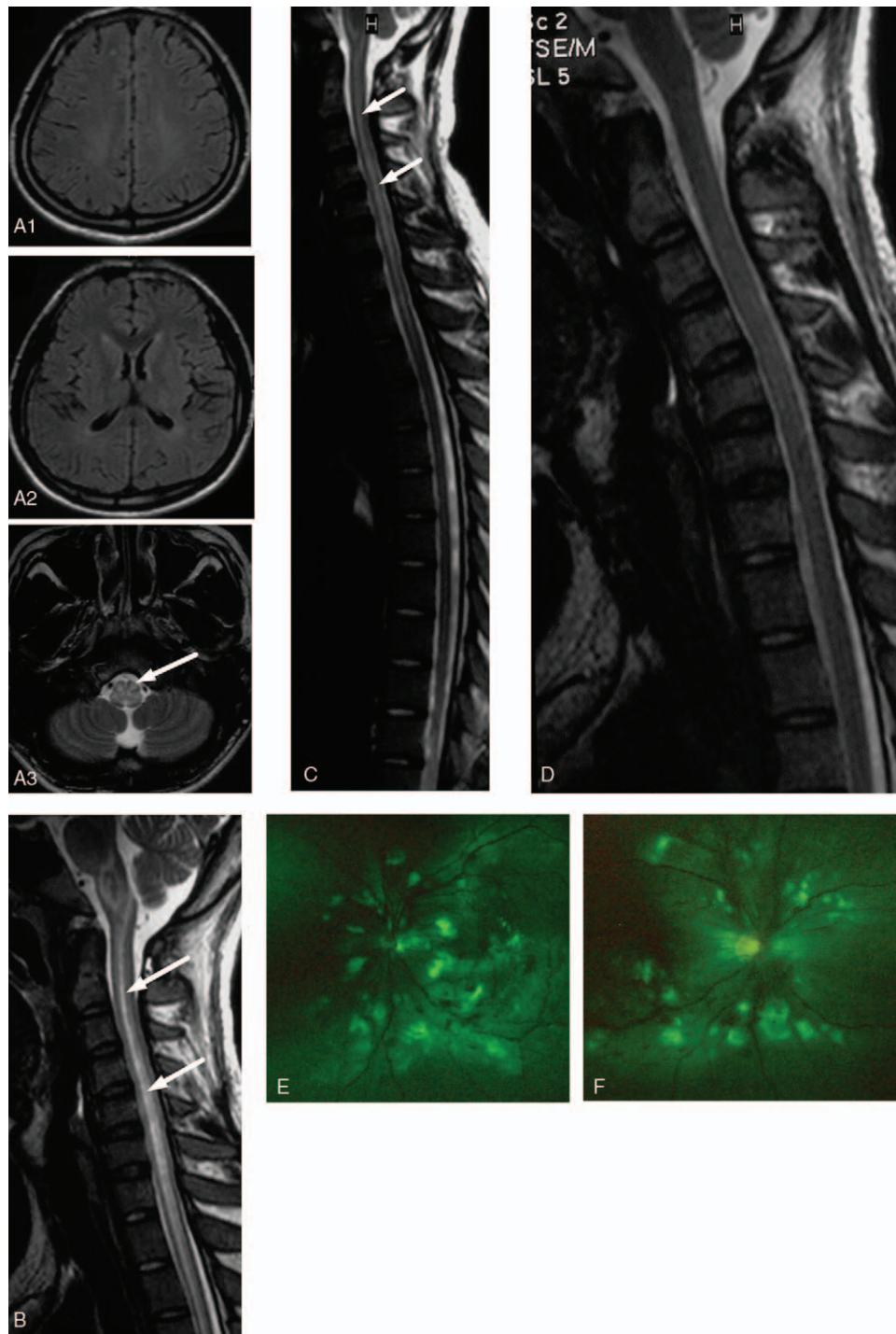
We report a young patient whose condition was consistent with PRES-SCI but without the hemisphere lesions.

## 2. Case description

An appropriate written informed consent was obtained from the patient reported in this study.

### 2.1. Patient information

This 20-year-old Asian male patient was admitted for a 1 week history of blurred vision, abrupt headache, and weakness of the limbs. He has had poorly controlled hypertension for 1 year and



**Figure 1.** Characteristic imaging findings in PRES-SCI without hemisphere lesions involved. (A) Axial FLAIR of hemisphere and T2 of medulla. (A1, A2) There was no hyperintensity of the bilateral parietal and occipital lobes in FLAIR brain MRI. (A3) Hyperintensity of the brain only limited in the medulla in T2. (B) Axial FLAIR hyperintensity of the medulla, and sagittal T2 hyperintensity in upper cervical spinal cord extending to the lower thoracic spine on admission. (C, D) follow-up MRI on the 3rd (C) and 10th (D) day of the spinal cord. Sagittal T2 of the medulla and cervical spinal cord after admission shows resolution of the prior hyperintensity (compare to A3 and B), which was also true on the thoracic spine MRI (not shown). (E, F), Scanning Laser Ophthalmoscope examination showed hypertension related retinopathy in both eyes (E, left; F right). Fundus photograph of the eyes shows optic nerve edema, macular star pattern of exudates in the macula, and cotton-wool spots and flame hemorrhages. MRI = magnetic resonance imaging, PRES-SCI = posterior reversible encephalopathy syndrome with spinal cord involvement.

mild kidney disease for about 1 month. His blood pressure is usually between 210/110 and 220/120mm Hg.

## 2.2. Clinical findings

In the emergency room, his blood pressure rose to 260/140mm Hg. Neurological examinations did not find any positive symptoms. Immunological blood tests were normal. Non-enhanced computed tomography (CT) showed no evidence of cerebral hemorrhage or subarachnoid hemorrhage. Magnetic resonance imaging (MRI) of the brain (Fig. 1A(A1, A2)) on the admission showed that there were no lesions in either hemisphere, though MRI of the spinal cord on admission (Fig. 1B) and the third day (Fig. 1C) showed T2-weighted images and fluid-attenuated inversion recovery (FLAIR) hyperintensity in the medulla, and upper cervical spinal cord that extended to the lower thoracic spine. Scanning laser ophthalmoscope examination showed hypertension related retinopathy in both eyes (Fig. 1E-F). A lumbar puncture had normal opening pressure, unremarkable cell indices, and no evidence of demyelination.

## 2.3. Diagnostic assessment

After management of blood pressure, his symptoms rapidly improved. Taking into account the characteristic lesions in the MRI as well as the reversible course upon treatment, he was diagnosed PRES-SCI.

## 2.4. Therapeutic interventions

He was treated with Valsartan (40 mg per day) and Amlodipine Tablets (I) (Novartis Pharma Stein AG, Switzerland) (5 mg per day) for the hypertension, his symptoms rapidly improved. Ten days after admission, a follow-up MRI of the spinal cord showed disappearance of previous imaging finding in T2-weighted image in the medulla and spinal cord (Fig. 1D). Considering the characteristics of clinical and imaging performance, an atypical variant of PRES was the most plausible diagnosis. One year after being discharged from hospital, the patient remained asymptomatic.

## 3. Discussion

Posterior reversible encephalopathy syndrome with spinal cord involvement (PRES-SCI) was 1st proposed by de Havenon A et al<sup>[4]</sup>. The PRES-SCI is considered as a unique disorder which affects the spinal cord and classically displays posterior hemispheric lesions and is thought to be a subtype of PRES. The PRES-SCI is very rare and usually seen in young male patients with severe acute hypertension that clinically manifests as headache, nausea/vomiting, visual disturbance, and hypertensive retinopathy, renal failure, but not necessarily seizure.<sup>[4,6]</sup>

The most frequently proposed mechanism of PRES is a failure of posterior circulation autoregulation in severe hypertension which break through the normal cerebral blood flow.<sup>[4,5]</sup> The vertebrobasilar system is considered particularly sensitive to the sudden change of blood pressure due to the reduced innervation of sympathetic nerves. The anterior spinal artery arises from the vertebrobasilar system, which may explain our patient's lesions in the cervical cord. The prognosis of PRES is mainly decided by the underlying condition, since most neurological symptoms are reversible in the majority of patients.<sup>[7-8]</sup> Patients diagnosed with PRES-SCI usually have good prognosis and typically resolve within a few weeks after appropriate treatment. Appropriate

reduction of blood pressure may prevent lesion progression from vasogenic edema into cytotoxic oedema, infarction or even permanent neurological deficits.<sup>[9]</sup>

Unlike other classical PRES cases, which have lesions involving posterior structures of the brain, posterior hemisphere lesions including bilateral occipital lobes were not involved in this patient. Compared with clinical manifestation of PRES-SCI, the course which usually lasts more than 2 weeks was much shorter and imaging change was rather faster. In this case the patient's symptom resolved within a few days after gradually controlling his blood pressure and MRI lesions disappeared within 10 days. We followed up him for a year and he had no complaints. We think hemisphere lesions such as parietal or occipital location were not necessary for the clinical manifestation and diagnosis of PRES-SCI. Although rare but spinal lesions alone may indicate a subtype of PRES-SCI.

Clinicians should suspect PRES-SCI when patients have mild or no neurologic signs accompanied with extreme elevation in blood pressure and lesions in the spinal cord. Spinal lesions alone may indicate a subtype of PRES-SCI. As this subtype also includes the spinal cord lesions, when considering PRES-SCI, which, if diagnosed, would spare patients the morbidity of a standard myelitis workup and empiric treatment.<sup>[10]</sup>

We found and diagnosed an unusual case of PRES-SCI clearly. However, since we have only 1 patient of this subtype, there is little opportunity to do further analysis.

## Author contributions

**Conceptualization:** Liming Zhang.

**Data curation:** Xun Wang.

**Investigation:** Dawei Dai.

**Resources:** Fan Cao.

**Supervision:** Xun Wang.

**Writing – original draft:** Lu Liu.

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