

A Case of Posterior Reversible Encephalopathy Syndrome Presenting with Isolated Diplopia

Masayuki Hata^{1*}, Akio Oishi¹, Yasuo Kurimoto², Shiro Yamamoto³ and Nobuo Kohara³

¹Department of Ophthalmology, Kobe City Medical Center General Hospital, Japan

²Department of Ophthalmology, Institute of Biomedical Research and Innovation Hospital, Japan

³Department of Neurology, Kobe City Medical Center General Hospital, Japan

Abstract

Background: Posterior Reversible Encephalopathy Syndrome (PRES) is a clinical entity characterized by a unique pattern of vasogenic brain edema mainly caused by eclampsia, immune suppressing drugs, or severe hypertension. The disease typically affects parieto-occipital lobe as the nomenclature indicates. Here we report a man with isolated concomitant strabismus, who was initially suspected as having glioma but was finally diagnosed with PRES.

Methods: A 43-year-old man complained of horizontal double vision for a week. He had no other neurological symptom, including headache or confusion. He had a history of hypertension but had no medication. On examination, he showed 10 to 12 prism diopter of exotropia in all gaze directions without any restriction of eye movement. The sudden onset of the symptom and the history of hypertension urged us to take head MRI, which showed poorly-demarcated T2-high intensity area in extensive brainstem and bilateral cerebellum. The image suggested a plausible diagnosis of brainstem glioma.

Results: He was admitted for planning brain biopsy. Since his systolic blood pressure was over 240 mmHg, he was to undergo blood pressure control before the biopsy. As the pressure decreased to around 180 mmHg, his symptom improved gradually. The lesion diminished in 2 weeks. Finally, he was diagnosed with PRES due to hypertension judged from the clinical course.

Conclusions: PRES can affect any locations in central nervous system including brainstem as shown in the present case. And of note, the patients with brainstem variant of PRES may present only minimal symptoms such as diplopia in the present case. Differentiation from tumors or infarction is very important to avoid unnecessary and invasive interventions. One useful characteristic is the clinical radiologic dissociation. Once brainstem variant of PRES is diagnosed, to monitor and control blood pressure is important because it is often caused by severe secondary hypertension.

Keywords: Posterior reversible encephalopathy syndrome; Brainstem involvement; Hypertensive encephalopathy; Diplopia; MRI; Clinical-radiologic dissociation

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical

entity characterized by a unique pattern of vasogenic brain edema mainly caused by eclampsia, immune suppressing drugs, or severe hypertension [1]. Patients present acute neurological symptoms and magnetic resonance imaging (MRI) shows characteristic findings that disappear in parallel with the clinical symptoms. The disease typically affects parieto-occipital lobe as the nomenclature indicates.

Despite its rarity, PRES may affect other areas than posterior lobe. Some reported the atypical variant of PRES in which the main lesion was in the brainstem [2-6]. The patients show various combinations of neurological symptoms including visual symptoms, but rarely complain of diplopia [2]. Here we report a man with isolated concomitant strabismus, who was initially suspected as having brainstem glioma but was finally diagnosed with PRES.

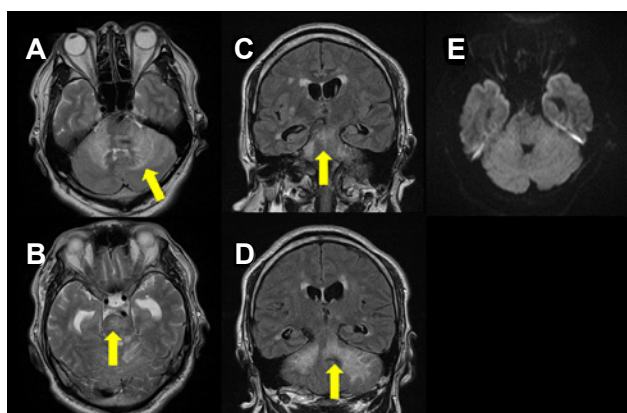


Figure 1: (Magnetic resonance imaging) A, B, C, D: In cerebral magnetic resonance (MR) images, T2-weighted Images (A, B) and fluid-attenuated inversion-recovery (FLAIR) images (C, D) on first visit day revealed poorly-demarcated high intensity area (arrow) in brainstem and bilateral cerebellar hemisphere. E: In diffusion-weighted imaging (DWI) on first visit day, there was no abnormality in the lesions, as were detected in FLAIR images.

***Corresponding author:** Masayuki Hata, Department of Ophthalmology, Kobe City Medical Center General Hospital, 4-6 Minatojima-nakamachi, Chuo-ku, Kobe 650-0046, Japan, Tel: 81-78-302-4321; Fax: 81-78-302-2487; E-mail: masayuki_lemmonsoda_0611@hotmail.com

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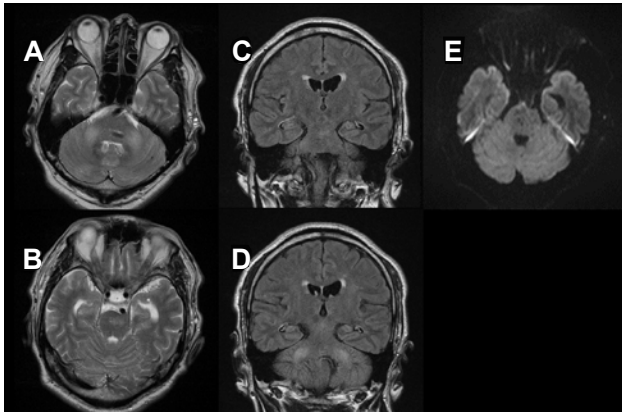


Figure 2: (Magnetic resonance imagings) **A-D:** T2-weighted images (**A, B**) and FLAIR weighted images (**C, D**) performed 2 weeks after the admission revealed diminution of abnormal findings. **E:** DWI images revealed no abnormality.

Case Report

A 43-year-old man complained of horizontal double vision lasting for a week. He suddenly found the symptom when he was fishing. There was no diurnal variation. He had no other neurological symptom, including headache or confusion. He had a history of hypertension but had no medication.

On examination, his corrected visual acuity was 1.2 OD and 1.5 OS. Light reflex was prompt and complete. He showed 10 to 12 prism diopter of exotropia in all gaze directions without any restriction of eye movement. He had no other neurological finding including cranial nerve involvement and ataxia. Ophthalmoscopy showed no change in his fundus.

The sudden onset of the symptom and the history of hypertension urged us to take head MRI, which showed poorly-demarcated T2-high intensity area in pons and bilateral cerebellum (Figure 1A and Figure 1B). The image suggested a plausible diagnosis of brainstem glioma.

He was admitted for planning brain biopsy. Since his systolic blood pressure was over 240 mmHg, he was to undergo blood pressure control before the biopsy. As the pressure decreased to around 180 mmHg, his symptom improved gradually. His exotropia and the abnormal findings in MRI diminished in 2 weeks (Figure 2A and Figure 2D). Finally, he was diagnosed with PRES due to hypertension judged from the clinical course. Further workup for hypertension revealed aldosterone-secreting adrenal tumor, and laparoscopic adrenalectomy was performed.

Discussion

PRES is often associated with an abrupt increase of blood pressure and typically involves occipital lobe. Although the pathophysiology of PRES is not completely understood, the failure of the autoregulation for cerebral blood flow, and vasogenic edema with endothelial injury are considered to be the underlying causes [1]. In the present case, T2-weighted MR images revealed diffuse hyperintensity area in the brainstem and cerebellum whereas diffusion-weighted imaging (DWI) findings were normal (Figure 1A-Figure 1E). The normal DWI findings were consistent with vasogenic edema but not cytotoxic edema seen in infarction.

The mechanism of posterior dominant involvement is considered to be related to a sparse distribution of sympathetic nerve in the vertebra basilar circulation, in contrast to the anterior cerebral circulation, which is richly innervated by sympathetic nerves [3].

Brainstem involvement of PRES is rare but certainly occurs, usually in patients with extreme hypertension [2-6]. The patients typically are in their fourth decade and have secondary hypertension as in the present case. Headache (73%), nausea or vomiting (43%), blurred vision (34%), abnormal gait (26%), coma (23%), seizure (17%), and paresis (9%) are general symptoms and they are generally mild. Concerning visual symptoms, blurred vision caused by hypertensive retinopathy is frequently seen but diplopia is rarely reported despite that the center of eye movement control lies in the brainstem [3]. Blurred vision can be caused by hypertensive retinopathy, but we doubt some patients express the diplopia as blurred vision. We should consider the possibility of PRES in patients with only mild visual symptoms such as diplopia.

In this case, the patient showed same extent of exotropia in all gaze directions without any restriction of eye movement. It indicated that the mechanism of his horizontal diplopia was not due to ocular motor nerve palsy but due to be supra nuclear impairment.

Both occipital and brainstem lesions are within the territory of the vertebra basilar artery, and the reason why lesion develops in brainstem but not in occipital lobes is unclear. The definitive mechanism of brainstem involvement without occipital lobes remained unknown.

Another clinical point in our case was the lack of symptoms corresponding to the extensive brainstem and cerebellum lesion on MRI. Some reports on brainstem involvement in PRES have been mentioned about this and called clinical-radiologic dissociation [3,6]. In this case, he presented diplopia without any other symptoms, which was considered to be an example of clinical-radiologic dissociation.

It is important to distinguish PRES from other disease, including cerebral infarction, brain tumor, and demyelinating disorder because the treatment for each disease is completely different. Though MRI findings are certainly useful for distinguishing them, the differentiation is sometimes quite difficult. The clinical-radiologic dissociation is useful information in differentiating from these diseases.

In conclusion, brainstem variant of PRES may present only minimal symptoms such as diplopia. Differentiation from tumors or infarction is very important to avoid unnecessary and invasive interventions. One useful characteristic is the clinical-radiologic dissociation.

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