Sudden bilateral vision loss as the sole manifestation of posterior reversible encephalopathy syndrome from acute uremia

Clinical case report

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Abstract

Rationale: Posterior reversible encephalopathy syndrome (PRES) is a cliniconeuroradiological entity associated with vasogenic edema. Symptoms may include headache, seizures, altered mental status, and visual impairment. Patients with PRES generally present with neurological deficits.

Patient concerns: Here, we report an unusual case of a 42-year-old man who presented with sudden bilateral vision loss without any other neurologic symptoms.

Diagnoses: He was diagnosed with PRES secondary to acute uremia.

Interventions and outcomes: Our patient experienced a dramatic improvement in visual acuity, blood chemistry values, and magnetic resonance imaging findings following repeated hemodialysis.

Lessons: Sudden bilateral vision loss may be the sole manifestation of PRES, particularly in patients with risk factors for PRES. Awareness of this variation of the clinical symptoms of PRES is important to facilitate its recognition.

Abbreviations: MRI = magnetic resonance imaging, PRES = posterior reversible encephalopathy syndrome.

Keywords: acute uremia, posterior reversible encephalopathy syndrome, vision loss

1. Introduction

Posterior reversible encephalopathy syndrome (PRES) has been associated with a typical pattern of relatively symmetric vasogenic edema involving the subcortical white matter.\textsuperscript{[1]} The common causes of PRES are hypertensive encephalopathy, eclampsia, and the use of cytotoxic and immunosuppressive drugs.\textsuperscript{[1]} Uremic encephalopathy is one of the common causes.\textsuperscript{[2,3]} Patients typically experience a combination of headache, nausea, vomiting, seizures, altered mental status, and visual impairment.\textsuperscript{[1]} Here, we present an interesting case of a patient with sudden bilateral vision loss as the sole manifestation of PRES secondary to acute uremia. This study was conducted in accordance with the ethical standards stated in the Declaration of Helsinki and with the approval of the Institutional Review Board of Seoul St. Mary’s Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea.

2. Case history

A previously healthy 42-year-old man came to the emergency department because of a sudden decrease in the visual acuity of both eyes upon waking that morning. According to the patient, his visual acuity was good in both eyes before the onset of the symptoms. He had no other neurologic symptoms, such as headache, nausea, vomiting, seizure, and altered mental status. His blood pressure was slightly elevated to 150/80 mm Hg, although he has never had a diagnosis of hypertension. There were no signs of optic neuropathy or retinopathy on ophthalmologic examination, and the optic disc appeared normal in both eyes. Laboratory tests showed an increased blood urea nitrogen (216.4 mg/dL), creatinine (24.28 mg/dL), and serum potassium (8.3 mEq/L). On initial T2 fluid-attenuated inversion recovery and “diffusion-weighted” imaging, there were small, patchy, nodular high signal intensities in the both occipital cortices (Fig. 1). These imaging findings were felt to be most consistent with PRES.
The patient was admitted to the intensive care unit and underwent repetitive hemodialysis. Kidney biopsy revealed marked acute tubular necrosis. The cause of acute tubular necrosis was unclear.

His visual acuity improved gradually every day. After 7 days of hemodialysis, the patient’s visual acuity was 20/20 in both eyes and his blood urea nitrogen and creatinine level had decreased (43.6 and 9.54 mg/dL, respectively). Follow-up brain magnetic resonance imaging (MRI) 9 days later showed marked regression of the previously noted high signal intensity lesions in both occipital cortices, most consistent with PRES.

3. Discussion

PRES has been associated with a typical pattern of relatively symmetric vasogenic edema involving the subcortical white matter.\(^\text{1}\) Common causes of PRES are pre-eclampsia/eclampsia, hypertensive encephalopathy, immunosuppressive drugs, and uremic encephalopathy. The occipital and parietal lobes are most typically affected.\(^\text{1,2,4}\) The pathogenesis of PRES is not understood completely and remains controversial. Suggested pathophysiologic mechanisms of PRES include failed autoregulation and endothelial abnormality.\(^\text{5}\)

In general, PRES is characterized by headache, seizures, altered mental status, visual impairment, focal neurological signs, and reversible cerebral lesions on imaging.\(^\text{1}\) However, our patient had the sole manifestation of sudden bilateral vision loss. We were only aware of his renal dysfunction after blood tests because he had no other neurological symptoms suggestive of uremic encephalopathy. Brain MRI on admission revealed vasogenic edema involving both occipital cortices, suggestive of PRES. The resolution of his visual impairment and MRI findings after repetitive hemodialysis further support the clinical diagnosis of PRES secondary to acute uremia, although visual disturbance was his only neurological symptom.

The location and the extent of PRES lesions vary.\(^\text{6,7}\) Even though the extent of vasogenic edema was mild in our case, we speculated that his visual loss was severe initially because the lesions were located in the occipital lobe. Although our patient did not have severe neurologic manifestations, such as headache, focal or generalized seizure, or altered mental status, his MRI findings and response to treatment for uremia were consistent with PRES. Sudden bilateral vision loss was the only presenting symptom of PRES secondary to acute uremia. We assumed that he was diagnosed with PRES early due to the location of the cerebral lesions prior to worsening of the vasogenic edema and development of severe neurologic manifestations.

4. Conclusions

Early recognition of PRES is important for prompt treatment by eliminating factors that cause PRES, such as uremia, hypertension, cytotoxic and immunosuppressive drugs. To date, this is an unusual case of PRES with sudden bilateral vision loss as the only presenting symptom. Clinicians should be aware that presenting signs and symptoms of PRES vary greatly depending on the location and extent of the lesions, and visual impairment may be the sole manifestation in some patients with PRES.

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