CASE REPORT

Temporary Visual Loss Due to Posterior Reversible Encephalopathy Syndrome in the Case of an End-Stage Renal Disease Patient

Michael Chia-Yen Chou^a, Chia-Yi Lee^a, and Shih-Chun Chao^{a,b,c}

^aDepartment of Ophthalmology, Show Chwan Memorial Hospital, Changhua, Taiwan; ^bDepartment of Electrical and Computer Engineering, National Chiao Tung University, Hsinchu, Taiwan; ^cDepartment of Optometry, Central Taiwan University of Science and Technology, Taichung, Taiwan

ABSTRACT

Posterior reversible encephalopathy syndrome is a clinical phenomenon associated with headache, altered mental status, seizures, and visual disturbances along with radiographic acute cerebral oedema. Several conditions are correlated with posterior reversible encephalopathy syndrome, with acute hypertension, eclampsia, and cytotoxic agents being the most common. This report presents a case of posterior reversible encephalopathy syndrome in a 45-year-old male with multiple underlying conditions, including hypertension, diabetes, and end-stage renal disease. Sudden onset of bilateral visual loss with mild systemic feature of dizziness was noted on presentation. Radiologic findings showed characteristic findings of posterior reversible encephalopathy syndrome. Following prompt control of hypertension and haemodialysis, resolution of symptoms was noted.

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Introduction

Posterior reversible encephalopathy syndrome (PRES), a disease entity first described in 1996, is a condition associated with characteristic neuroimaging findings of posterior cerebral white matter oedema. Symptoms include insidious onset of headache, decreased consciousness, confusion, visual disturbances, and seizure. The exact pathogenesis of PRES remains unclear, but it is postulated that abnormal cerebral autoregulation and endothelial dysfunction may be related.¹ PRES have been known to be associated with several conditions, most commonly hypertensive encephalopathy, eclampsia, and the use of cytotoxic and immunosuppressant drugs. Other aetiologies include autoimmune disorders, acute or chronic renal diseases, haemolytic and uremic syndromes, thrombotic thrombocytopenia purpura, blood transfusion, and contrast media exposure.² Although few cases of permanent visual loss have been reported, symptoms are often reversible with spontaneous recovery in days to weeks after correction of inducing factors.³ The aim of this report is to describe a patient of PRES

presenting as acute visual loss who is undergoing haemodialysis.

Case report

A 45-year-old Taiwanese male was admitted via the emergency department due to sudden onset of blindness, ocular pain, and dizziness. The patient had been diagnosed with both hypertension and type 2 diabetes mellitus over 10 years ago and has been taking medications for both conditions. Furthermore, he is currently undergoing regular haemodialysis triweekly for end-stage renal disease (ESRD).

At our emergency department, blood pressure was 197/87 mm Hg. Physical examination showed intact muscle power without specific neurologic deficits. Ocular examination disclosed best-corrected visual acuity (BCVA) of no light perception in both eyes, with poor pupillary light reflex. Dilated funduscopy examination showed normal optic disc, bilateral cotton-wool spots, and dot haemorrhages consistent with non-proliferative diabetic neuropathy or hypertensive retinopathy (Figure 1). No signs of retinal detachment was seen upon retinal

CONTACT Shih-Chun Chao, MD arthur27@saturn.seed.net.tw Department of Ophthalmology, Show Chwan Memorial Hospital, No. 2, Ln. 530, Sec. 1, Zhongshan Road, Changhua City, Changhua County 50093, Taiwan.

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Figure 1. Fundus photography. Slightly obscured due to lens opacity. Cotton-wool spots with dot haemorrhage was seen under indirect funduscopy examination.

examination or via B-scan ultrasound. Cortical blindness due to cerebral vascular incident was suspected; thus, non-enhanced computed tomography (CT) was arranged; results did not reveal lesions or signs of acute ischaemic or haemorrhagic stroke. Afterwards, the patient was admitted to the neurologic intensive care unit (ICU) for further observation.

At ICU, blood examination was nonspecific except for mildly elevated C-reactive protein (CRP) level. Cerebrospinal fluid sample was obtained and analysed; cell count and biochemistry were within normal range except for mild non-significant elevation of glucose and protein levels (78 and 46 mg/dL, respectively), and microbial staining and culture were negative. Magnetic resonance imaging (MRI) was performed. Diffusion-weighted (DW) images were compatible with CT findings, which was negative for posterior circulation infarction (**Figure 2**). Multiple hyperintense signal change at both cerebral watershed areas and at periventricular, parieto-occipital, cerebellar, and brainstem areas were visualised in FLAIR (fluid-attenuated inversion recovery) images (**Figure 3**). Magnetic resonance angiography showed patent vessels without narrowing or aneurysmal dilatation (**Figure 4**). After consultation with the radiologist, PRES was diagnosed. Intravenous nicardipine was given for control of blood pressure, and the patient reported improvement of visual symptoms.

Ophthalmologist was consulted for follow-up 9 days after admission. BCVA was 0.6 in the right eye and 0.9 in the left eye. Pupillary light reflex was intact with normal intraocular pressure. Visual field examination was also arranged, which initially was unreliable due to high fixation loss at the time of examination (**Figure 5**). The patient was eventually discharged due to improved visual and systemic conditions.

Figure 2. Axial diffusion-weighted MR image. No obvious hyperintensity was seen in the posterior lobe to indicate infarctions.

Discussion

PRES is defined as a clinical syndrome with insidious onset of headache, confusion, decreased level of consciousness, visual change, and seizures. It was shown to be associated with radiologic findings of posterior cerebral white matter oedema. PRES have been attributed to several medical conditions; the primary aetiologies of PRES include acute hypertension, eclampsia, sepsis, renal disease, and the use of immunosuppressant and cytotoxic drugs.^{3–6} These conditions cause a disruption of cerebral autoregulation and endothelial dysfunction, which are believed to be the two main mechanisms of PRES.¹ An unusual rise in systemic blood pressure causes disruption of cerebral autoregulation, which leads to increased cerebral blood flow and arteriole dilation. The resulting cerebral

Figure 3. Axial FLAIR MR image of PRES showing multiple lacunar hyperintense signal changes at both cerebral watershed areas and at periventricular, parieto-occipital, cerebellar, and brainstem areas.

Figure 4. Magnetic resonance angiography showing no vascular narrowing or aneurysmal dilatation.

hyperperfusion causes breakdown of the blood-brain barrier, and subsequent cerebral oedema. Furthermore, conditions that may cause endothelial

damage and dysfunction may also play a vital role in the pathophysiology of cerebral oedema.^{1,7}

In our case, the patient presented with underlying condition of hypertension, ESRD undergoing regular haemodialysis, as well as type 2 diabetes with diabetic retinopathy. Due to the presence of elevated blood pressure upon admission, acute hypertension may be the most common and direct explanation for the patient's symptoms. However, noting the patient's history of diabetes and chronic renal disease, conditions that are known to be associated with damage of the systemic vasculature, damage of the cerebral vascular endothelium due to increased circulating toxins may cause endothelium dysfunction and become an important factor in the cause of vasogenic brain oedema. Previous reports of PRES have mostly attributed endothelial dysfunction to eclampsia, autoimmune disorders, or the use of cytotoxic agents.^{6,8}

Visual sequelae associated with PRES include cortical blindness, homonymous hemianopia, visual neglect, and blurred vision. The presence of these symptoms are often misleading, prompting misdiagnosis as ischaemic cerebrovascular events. Our patient presented with acute bilateral

Figure 5. (Top) Visual field examination showing clover-leaf pattern. (Bottom) Visual field examination at 8-month follow-up showing constricted field with retained central 10–20° vision.

cortical blindness with associated symptoms of dizziness. However, systemic features such as seizures or encephalopathy were absent, which have been reported to be as high as 74% in previous reported cases.² It should be noted that it is unusual for PRES to present with visual loss in the absence of other systemic symptoms.^{9–11}

Ocular examination of PRES is often insignificant. Funduscopy examination may reveal papilloedema accompanied by haemorrhages or exudates in the presence of raised intracranial pressure. Due to the

underlying medical history of our patient and his previous records, the presentation of cotton-wool spots and dot haemorrhage upon examination may be better explained by diabetic retinopathy rather than hypertension or PRES due to absence of marked optic disc swelling or areas of acute haemorrhage. Result of the visual field examination of our patient is also likely impertinent to PRES, as the portrayed peripheral visual defect may be associated with the pan-retinal photocoagulation treatments the patient has received during previous clinical visits.

In PRES, oedematous change is common in the posterior cerebral region, typically in the parieto-occipital region.² Imaging manifests as areas of hypointensity on CT and hyperintensity on T2-weighted MRI. In our patient, MRI examination revealed multiple lacunar hyperintense signal change at both cerebral watershed areas and periventricular, parieto-occipital, cerebellar, and brainstem areas visualised under FLAIR images, which are consistent with the diagnosis of PRES.^{2,12}

The majority of cases of PRES reported spontaneous resolution following control of blood pressure and removal of potential instigating factors.^{1,11} Generally, patients with both PRES or hypertensive retinopathy showed full recovery of vision within weeks despite severe visual impairment at onset. However, failure to identify the underlying aetiology with delay in treatment may result in permanent damage in an otherwise reversible condition. There have been documented reports of patients with PRES suffering from permanent total visual loss.^{3,13,14} Furthermore, potential serious consequences of PRES have been reported, including death and permanent neurologic disability.⁹ In one case series, elevated creatinine levels were shown to be a risk factor for death.¹⁵ Studies have suggested that cytotoxic oedema, which may ultimately result from mass effect of marked vasogenic oedema, seems to be predictive of a worse prognosis.^{4,15} Brainstem involvement were also indicative of poor prognosis.

Our patient reported pronounced improvement of visual symptoms following anti-hypertensive therapy and regular haemodialysis treatment. However, due to his multiple unfavourable underlying conditions including ESRD, diabetes, and hypertension, it is crucial to alert the patient of the potential risk of future relapses as well as more grave consequences and advise strict control of his blood pressure and blood sugar, as well as implement swift reversal of predisposing factors upon future presentation of symptoms of PRES.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

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