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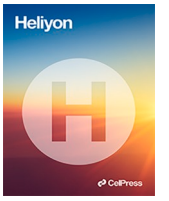

Case report

A case of bilateral cortical blindness followed by generalised tonic-clonic seizure epilepsy in a patient with posterior reversible encephalopathy syndrome

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) in end-stage kidney disease (ESKD) is rare, with ocular symptoms as the first manifestation being even rarer. Here, we report a case of PRES in a patient with ESKD, characterized by sudden binocular blurred vision followed by epilepsy, to improve the understanding of this syndrome among nephrologists and ophthalmologists. A 50-year-old female requested an ophthalmic consultation due to bilateral vision loss followed by generalised tonic-clonic seizures. One month before onset of current illness, she developed ESKD secondary to rapid progression of previous ANCA vasculitis associated renal damage. Latter magnetic resonance imaging confirmed the diagnosis of PRES. Two weeks later, the patient's vision fully recovered. **Conclusion**: PRES is not an etiological diagnosis but a neuroimaging sign. In addition, PRES is a danger signal that is usually reversible if recognized and treated early, and can be life-threatening if treatment is delayed.

1. Introduction

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey et al., in 1996, and its main clinical features include seizures, headaches, blurred vision, and confusion [1,2]. It should be emphasized that this syndrome is not an etiological diagnosis but a neuroimaging sign [1]. So far, the exact cause of PRES is unknown, but it often co-occurs with a variety of acute and marked arterial hypertension induced by severe preeclampsia, eclampsia, acute end-stage kidney disease (ESKD), and acute hepatic porphyria [1–3]. Tawati DA et al. reported that PRES was diagnosed in 51.4 % of 342 pregnant women with eclampsia, followed by 19.8 % of 121 pregnant women with severe preeclampsia [1]. Similarly, Pischik E et al. found that PRES occurred in 42 % of patients with acute encephalopathy induced by acute hepatic porphyria [2]. However, in a retrospective study in Southwest Ireland over a ten-year period, Canney M et al. showed that PRES occurred in only 0.84 % of patients with ESKD [3] (see Fig. 1)

Here, we report a case of PRES in a patient with ESKD, characterized by sudden binocular blurred vision followed by epilepsy, to improve the understanding of this syndrome among nephrologists and ophthalmologists.

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2. Patient information

A 50-year-old female requested ophthalmic consultation due to sudden bilateral vision loss followed by generalised tonic-clonic seizure epilepsies (two seizures, 90 seconds each, half an hour apart). She also complained of headaches, altered consciousness, and vomiting. On arrival, her blood pressure was 160/85 mmHg and best-corrected visual acuity of each eye was light perception. However, she denied a medical history of arterial hypertension or eye disease. A week prior to admission, she developed ESKD (serum creatinine of 1046 μmol/L) secondary to rapid progression of previous ANCA vasculitis-associated renal damage and was started on intravenous methylprednisolone (500 mg daily for 3 days), followed by five sessions of plasma exchange (once time every two days), followed by intravenous methylprednisolone (250 mg daily for 3 days) plus cyclophosphamide (400 mg ONCE), to decrease high MPO level (261AU/mL). Along with the above treatments, two sessions of hemodialysis (once every two days) were also performed due to extracellular fluid volume expansion and poorly controlled hypertension, and oral hydroxychloroquine (200 mg twice daily) was started on admission. After the above treatment measures, her serum creatinine level gradually improved to 380 μmol/L, and serum C-reactive protein level returned to normal from the previous 62 mg/L.

However, three days before onset of present illness, she suddenly had blurry vision, followed by two seizures. About 3 hours later, her blood pressure peaked at 215/109 mmHg and she was urgently administered intravenous nicardipine (2 mg every hour) to control her blood pressure. Then, her blood pressure had gradually decreased to 163/87 mmHg. Later magnetic resonance imaging of the FLAIR sequence showed high signal areas in both occipital lobes (Fig. A, white arrows), while T2 weighted imaging showed a suspected high signal intensity in the same areas (Fig. B), indicating a diagnosis of PRES. Subsequently, her systemic medication was adjusted to the following regimen: sequential reduction of oral prednisolone acetate (from 80 mg on, reduction by 10 mg every 3–5 days), intravenous cyclophosphamide (400 mg, once per month), oral nifedipine controlled release tablet (30 mg, once daily), and oral hydroxychloroquine (200 mg, twice daily). Two weeks later, the patient's vision had fully recovered, and a repeat MRI (Fig. C and D) demonstrated the complete resolution of these abnormal areas.

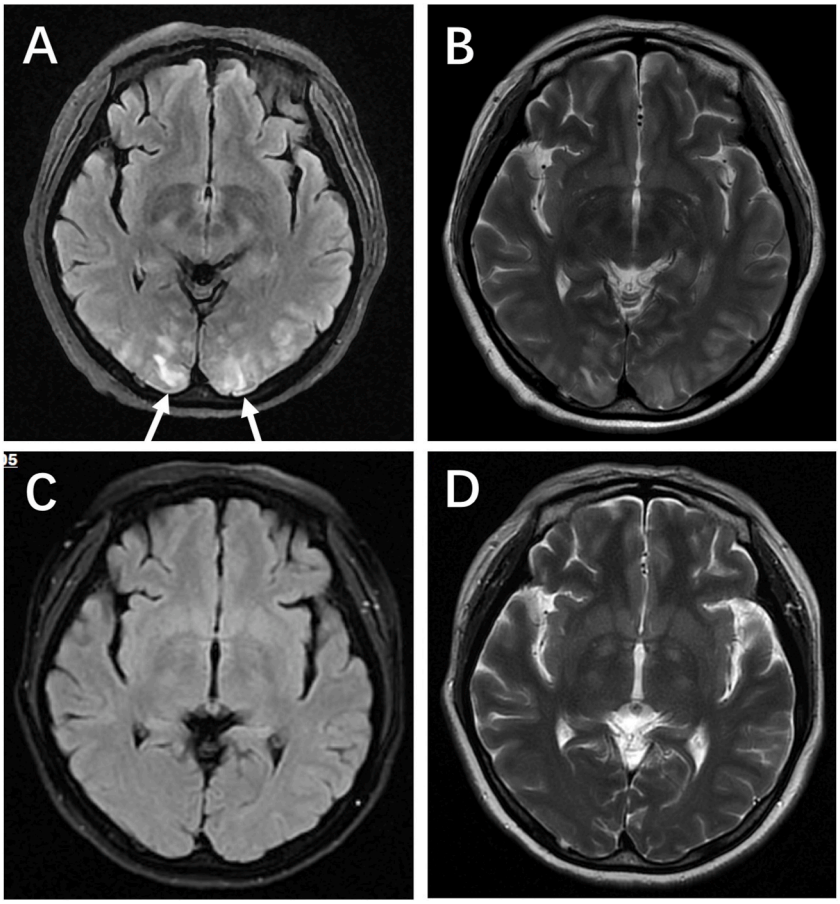

Fig. 1. Typical images of bilateral cortical blindness.
(A) Magnetic resonance imaging FLAIR sequence showing high signal areas (white arrows) in both occipital lobes. (B) T2 weighted imaging showing suspected high signal in both occipital areas. (C, D) Repeat magnetic resonance imaging demonstrating complete resolution of these abnormal areas two weeks later.

3. Discussion

In general, PRES in ESKD is rare, with ocular symptoms as the first manifestation being even rarer. As previously reported [1], seizures and headaches are the most common clinical manifestations, followed by blurred vision and altered consciousness. Another point that differs from previous literature is that PRES secondary to acute encephalopathy induced by acute hepatic porphyria often lasted 1–5 days [2], whereas the PRES we reported, secondary to ESKD, lasted only 10 hours. Also, PRES does not always occur in the posterior cortex and is not always reversible, although the words "posterior" and "reversible" are used in this terminology. All parts of the brain can be involved, most commonly in the occipital lobe, followed by the parietal and frontal lobes, and again in the pons and cerebellum [1,4]. The lesion in our case was located in both occipital lobes, which is consistent with the above results in the literature. It is important to emphasize that PRES is a danger signal that can lead to death in approximately 5.3%–40 % of the patients [1,3].

It was reported that arterial hypertension is a common cause and precipitating factor of PRES [3], which cannot be overlooked. In addition to acute and marked arterial hypertension, autoimmune diseases, immunosuppression, and severe acute respiratory syndrome coronavirus 2 may be involved in the occurrence of PERS [3,5–7]. Canney M et al. reported a case of PRES in ESKD induced by lupus nephritis [3] which was very similar to our case of PRES in ESKD induced by ANCA vasculitis-associated renal damage. The former was treated with high doses of oral glucocorticoids, nine sessions s of plasma exchange and four doses of rituximab before PERS onset. The latter also received high doses of intravenous glucocorticoids, five sessions s of plasma exchange and one dose of intravenous cyclophosphamide (400 mg) before PERS onset. Therefore, we speculate that multiple factors jointly participate in the destruction of the blood-brain barrier and automatic regulation of cerebral blood flow and ultimately cause PRES with vasogenic cerebral edema as its basic pathophysiological feature. However, our case lacked long-term follow-up results to know whether PRES episodes left behind damage to the optic nerve or visual field.

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Data availability statement

The data that support the findings of this study are available upon request from the corresponding author [SS].

CRediT authorship contribution statement

Benqi Zhao: Writing – original draft, Investigation, Data curation. **Shancheng Si:** Writing – review & editing, Writing – original draft, Supervision, Project administration.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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