# *Clinical Case Report*

## Posterior reversible encephalopathy syndrome presenting with seizures

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**ABSTRACT**

**Hyperperfusion syndrome, previously known as posterior reversible encephalopathy syndrome (PRES), is a clinico-radiological entity with characteristic features on neuro-imaging. It is believed to be caused by vasogenic oedema, predominantly in the posterior cerebral hemispheres. We report the case of an elderly man who presented with convulsions and was diagnosed to have PRES due to hypertension.**

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### INTRODUCTION

Hyperperfusion syndrome, previously named posterior reversible encephalopathy syndrome (PRES), is an increasingly recognized clinical and radiological entity. It was first described by Hinchey *et al*. in 1996.[1] Initially, the syndrome was believed to be secondary to arterial hypertension, with or without hypertensive encephalopathy, renal disease or immunosuppressive therapy. It has recently been described in a variety of conditions, including eclampsia, haemolytic uraemic syndrome, connective tissue diseases, vasculitis, malignancies being treated with chemotherapy, transfusions, thrombotic thrombocytopenic purpura and porphyria.[2–7] We describe a patient who presented to us with this unusual disorder.

### THE CASE

A 64-year-old man presented to the emergency department of our hospital in status epilepticus. He had apparently been well till 3 hours earlier, when he experienced diminution of vision. This was followed by 10–12 episodes of generalized tonic–clonic seizures, each lasting for 5–7 minutes, with the patient failing to regain consciousness in between. There was no history of trauma, headache, vomiting or fever. He was not known to have diabetes mellitus or hypertension and there was no past history of tuberculosis.

On examination, the patient was drowsy with a Glasgow coma scale of 11/15. He was afebrile, and his pulse rate was 80 per minute and blood pressure 200/160 mmHg. Examination of the chest, cardiovascular system and the abdomen was normal. Examination of the nervous system was remarkable only for bilateral extensor plantar responses. There were no signs of meningeal irritation. Examination of the optic fundus was normal.

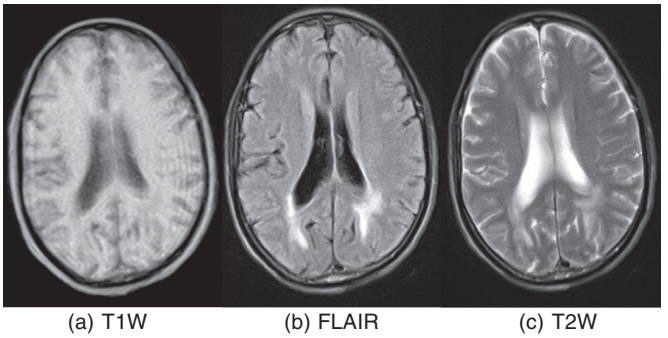
Seizures were brought under control with benzodiazepines and anti-epileptic medications. The patient regained consciousness after 2 hours, but was not fully oriented.

A haemogram, tests of renal and hepatic function, an electrocardiogram and a chest X-ray were all normal. A magnetic resonance imaging (MRI) scan of the brain, carried out on the day of admission, showed bilateral hyperintense signals in the temporo-occipital white matter and the centrum semiovale on T2-weighted and fluid-attenuated inversion recovery (FLAIR) images. These lesions were iso-intense to hypo-intense on T1-weighted images (Fig. 1).

The patient's blood pressure was gradually controlled with intravenous labetalol, followed by oral antihypertensive agents. He became fully oriented after about 24 hours, but there was a loss of recent memory. He was discharged after a week with a prescription for two oral antihypertensive drugs—telmisartan (40 mg once daily) and hydrochlorothiazide (12.5 mg once daily).

### DISCUSSION

Patients with the hyperperfusion syndrome may present with headache, seizures or focal neurological deficits. Isolated seizures as the presenting feature have been reported in 23%–28% of patients.[8] Secondary generalized seizures are common and present in 53%–62%.[1,8] Status epilepticus, defined as continuous seizure activity for at least 5 minutes (continuous) or as more than two motor seizures without full recovery of consciousness in the interval (intermittent), has been described in 3%–13% of patients.[8,9] Visual abnormalities are common; blurred vision has been reported in 26%–67% of patients, visual neglect in 4%–27%, homonymous hemianopia in 4%–20%, visual hallucinations in 3%–5% and



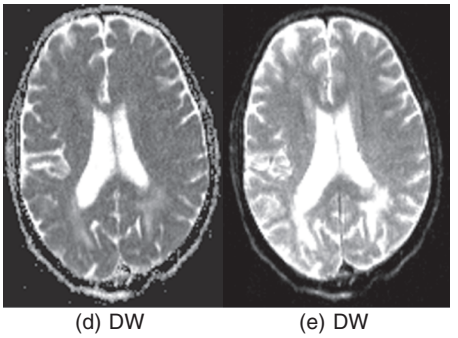




FIG 1. MRI scans of the brain showing bilaterally symmetrical hyperintense signal in the temporo-occipital lobes, peritrigonal white matter and centrum semiovales on T2-weighted, T2W (c) and fluid-attenuated inversion recovery, FLAIR (b) images, which appear iso- to hypo-intense on T1-weighted (T1W) images (a). No restriction of diffusion is seen in diffusion-weighted (DW) images (d and e).

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cortical blindness in 8%–33%.[1] Headache, nausea and vomiting have been observed among 26%–53% of patients.[1] Focal neurological signs have been reported in only 3%–17% of cases.[8] Hypertension has been present in 67%–80% of the reported cases.[1,8] However, an acute rise in blood pressure is not a common feature at presentation.

*Pathogenesis*

The brain's autoregulatory capability successfully maintains a stable cerebral blood flow among adults, despite alterations in the systemic mean arterial pressure ranging from 50 to 150 mmHg. An acute rise in blood pressure leads to stimulation of the sympathetic nerves, causing the central arterioles to constrict. When the systemic blood pressure exceeds the limits of this mechanism, breakthrough of autoregulation occurs, resulting in hyperperfusion via increased cerebral blood flow, capillary leakage into the interstitium and resultant vasogenic oedema. The predilection of hyperperfusion disorders to affect the posterior rather than anterior parts of the brain may be on account of a lower threshold for autoregulatory breakthrough in the posterior circulation.[10]

While elevated or relatively elevated blood pressure is common in many disorders of hyperperfusion, there is no apparent rise in pressure in some hyperperfusion states, such as calcineurin inhibitor toxicity. In such cases, vasogenic oedema is probably due to dysfunction of the capillary endothelium, which leads to breakdown of the blood–brain barrier. It is useful to separate disorders of hyperperfusion into those caused primarily by increased pressure and those due mostly to endothelial dysfunction, of a toxic or autoimmune aetiology.[10]

*Imaging*

Hyperperfusion syndrome was originally described as a subcortical disease of the posterior cerebrum,[1] marked by symmetrical changes both in the parietal and occipital lobes. More recently published series have shown different radiological findings and patterns, such as cortical involvement, as well as frontal or temporal lesions, or less commonly, lesions in the cerebellum, brainstem or basal ganglia.[11] Although PRES is usually bilateral, a unilateral pattern has also been described.[12]

Computed tomographic (CT) imaging of the brain may show hypodense areas in the involved regions. However, the diagnosis is established using brain MRI. T2-weighted and FLAIR MRI images show cortical and subcortical hyperintensity corresponding to areas of vasogenic oedema.[1,12] These can be distinguished from cytotoxic oedema by diffusion-weighted imaging and apparent diffusion coefficient maps (elaborated from the diffusion data).[12,13]

Previously, this classic radiographic appearance had been termed reversible posterior leucoencephalopathy. However, this term has fallen out of favour because none of its elements is completely accurate. The radiographic and clinical changes are not always reversible. The territory involved is not uniquely posterior and the disorder may also affect grey matter, unlike purely white matter, as suggested by the word 'leucoencephalopathy'.[10]

*Treatment*

Treatment should commence immediately after the diagnosis has been made. Hypertension plays a major role and the blood pressure should be lowered judiciously with intravenous agents, such as labetalol and nicardipine. Continuous cardiac and blood pressure monitoring, often through an arterial line, is also advised. The mean arterial pressure should initially be lowered by 20%, since lowering it further may cause secondary ischaemia as the blood pressure drops below the lower range of the patient's autoregulatory capability. Patients in whom the cause of the syndrome has been identified must be treated promptly. Seizures must be identified and controlled, which often necessitates continuous electroencephalographic monitoring.

*Conclusion*

Hyperperfusion syndrome is an unusual but important differential diagnosis of seizures. It is necessary to recognize and treat it promptly to ensure favourable outcomes.

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