

Posterior reversible encephalopathy syndrome

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CASE

A 76-year-old woman with a history of hypertension, pulmonary embolism (on apixaban), DM type 2 with neuropathy and nephropathy, gout, GERD, CHF, COPD, dyslipidemia, CKD, and anxiety presented to the emergency department (ED) with new onset seizures and confusion. She had been recently discharged from the hospital after receiving treatment for acute respiratory failure. In the ED the patient had tonic-clonic movements of the right upper extremity and nystagmus. She did not respond to voice but did withdraw from pain. Her pupils were unequal but did respond to light. Both toes were up going on plantar stimulation. In the ED she was given lorazepam and fosphenytoin and was intubated for airway protection. Computed tomography of the head revealed new bilateral symmetrical low density changes in both occipital lobes (Figure 1) suggestive of posterior reversible encephalopathy syndrome (PRES). Magnetic resonance imaging (MRI) showed bilateral, symmetrical, white matter edema of the frontal, parietal, and occipital lobes consistent with PRES (Figure 2). Her initial blood pressure in the ICU was 179/118 mmHg. Management in the intensive care unit included levetiracetam for seizure prophylaxis, blood pressure control, and correction of metabolic abnormalities. She recovered and was extubated.

DISCUSSION

ETIOLOGY

Posterior reversible encephalopathy syndrome is an acute neurologic disorder characterized by vasogenic brain edema.¹ Common risk factors for the development

of PRES include, but are not limited to, immunosuppressive therapy, eclampsia, renal failure, and hypertensive encephalopathy. The pathophysiology is uncertain and has two competing theories for its etiology. Both theories involve the autoregulatory system of the brain. The first postulates that severe hypertension leads to autoregulatory vasoconstriction in the brain, which then leads to ischemia and edema. This theory has flaws since prolonged vasoconstriction would potentially lead to infarction which is not reversible. In addition, 15-20 percent of patients with PRES are normotensive or hypotensive.¹ The second theory postulates that rapidly developing hypertension overwhelms cerebral blood flow autoregulation and causes hyperperfusion and break down of the blood brain barrier. This leads to extravasation of plasma and macromolecules into the interstitial space causing cerebral edema. This theory does not explain PRES in the 15-20 percent of patients without hypertension. However, pronounced fluctuations in blood pressure, rather than absolute hypertension, could lead to the development of PRES.

CLINICAL PRESENTATION

The symptoms of PRES usually develop rapidly over hours to days. The clinical presentation commonly includes headache, seizures, altered mental status, and visual disturbances. Seizures are tonic-clonic in up to 65-70 percent of patients and are often the presenting symptom.¹ Headaches in PRES patients are usually described as constant and dull. A “thunderclap” headache should raise suspicion for cerebral vasoconstriction, associated with PRES. The altered mental status ranges from confusion and agitation to coma in severe cases. Visual disturbances can include hallucinations, cortical blindness, visual field defects, and changes in visual acuity. The fatality rate associated with PRES ranges from 5-15 percent.² It is, therefore, important to recognize this clinical presentation as quickly as possible.

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Figure 1. Patient's CT scan of the head without contrast showing new interval developing bilateral, symmetric low density changes of both occipital lobes. Slight prominence of ventricles and sulci pattern consistent with diffuse atrophy.

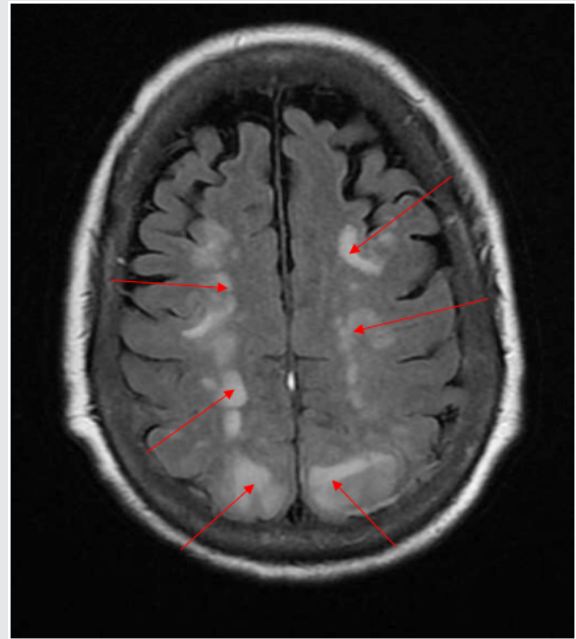


Figure 2. The patient's MRI showed bilaterally symmetric white matter edema in the frontal, parietal and occipital lobes appearing moderate in severity.

RADIOLOGICAL PRESENTATION

While PRES can be suspected based on clinical presentation, imaging is essential to exclude alternative diagnoses. Neurological imaging in PRES reveals bilateral vasogenic edema. The classic involvement of this edema includes the posterior parietal and occipital lobes, generally sparing the calcarine and paramedian occipital lobes. Both white matter and grey matter may be involved depending on severity, though typically the edema only involves the subcortical white matter. Three distinct radiographic patterns have been described in up to 70 percent of patients with PRES: 1) a dominant parietal-occipital pattern; 2) a superior frontal sulcus pattern most often isolated to the mid and posterior regions of the sulcus; and 3) a holo-hemispheric watershed pattern which shows linear involvement of the frontal, parietal, and occipital lobes at the watershed zones. Computed tomography is often the first test ordered and does show vasogenic edema in some cases; MRI is more sensitive and therefore preferred over CT. A fluid attenuated inversion recovery (FLAIR) MRI should be

obtained because it is the most sensitive for detecting lesions in the cortical and subcortical areas. There is no imaging gold standard for diagnosing PRES, so imaging findings should be combined with clinical features when making a diagnosis.

Keywords: posterior reversible encephalopathy syndrome, vasogenic edema, seizures

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