

Postpartum Blindness: Two Cases

Kimberly A. Chambers, MD
Terry Wayne Cain, MD

*From the Department of
Emergency Medicine,
University of Texas–Houston
Medical School, Houston, TX.*

We present 2 cases, one eclamptic patient and one noneclamptic patient, of headache, cortical blindness, and seizures. Both patients demonstrated findings consistent with posterior leukoencephalopathy syndrome. Posterior leukoencephalopathy syndrome is a rapidly evolving neurologic condition that is characterized by headache, nausea and vomiting, seizures, visual disturbances, altered sensorium, and occasionally focal neurologic deficits. Posterior leukoencephalopathy syndrome can be triggered by numerous conditions, including preeclampsia-eclampsia, and can be seen in the postpartum period. It is characterized predominately by white matter vasogenic edema of the occipital and posterior parietal lobes. This condition can be difficult to differentiate clinically from cerebral ischemia, and magnetic resonance imaging with diffusion-weighted imaging and apparent diffusion coefficient are needed to do so. In most cases of posterior leukoencephalopathy syndrome, the prognosis is excellent, with full resolution of symptoms.

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INTRODUCTION

Headache associated with true cortical blindness is a rare event. The differential for headache with neurologic symptoms in the postpartum period includes preeclampsia, eclampsia, migraine, stroke, cerebral artery dissection, cerebral venous thrombosis, and posterior leukoencephalopathy syndrome. The clinical presentation of 2 patients and the differential diagnosis, management, pathophysiology, and prognosis of posterior leukoencephalopathy syndrome are discussed.

CASE REPORT

Case 1

A 31-year-old black woman, 7 days postpartum, presented with the chief complaint of “I’ve gone blind.” She had been seen for a severe headache and discharged from another emergency center earlier in the day after receiving ketorolac, meperidine, and promethazine. After she returned home, she slept for an hour and, on awakening, was blind. She reported nausea but no other symptoms. She had a past medical history of migraine but was taking no medications for that or any other condition.

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On physical examination, vital signs were as follows: blood pressure of 146/85 mm Hg, pulse rate of 64 beats/min, respiratory rate of 18 breaths/min, and oral temperature of 37.4°C (99.3°F). She was alert and oriented to person, place, and time. Pupils were equal, round, and reactive from 5 mm to 3 mm bilaterally. Extraocular movements were dysconjugate but intact. Visual fields revealed no light perception. The results of ophthalmoscopic examination were unremarkable, as was the remainder of the physical examination.

CBC count, electrolytes, calcium level, liver function test results, and coagulation study results were all within normal limits. Urinalysis demonstrated 2+ protein but was otherwise unremarkable.

During her emergency department (ED) evaluation, the patient had a witnessed tonic-clonic seizure with a subsequent postictal period. She was treated with benzodiazepines and magnesium sulfate. The results of head computed tomographic scanning were normal. She was admitted to the hospital and underwent magnetic resonance imaging (MRI), which revealed bilateral occipital lobe edema with a hyperintense T2 signal. Additionally, there were hyperintense signals in the frontal lobe, basal ganglia, and bilateral hemispheres. There was no evidence of ischemia or central venous thrombosis on MRI.

No further seizure activity was witnessed during her hospital course. Within 24 hours, her vision had completely returned.

Case 2

A 15-year-old patient presented with severe headache 2 days postpartum after a normal spontaneous vaginal delivery with epidural anesthesia. She reported worsening of her headache with sitting up. She denied facial or extremity swelling, chest or abdominal pain, blurred vision, or lateralizing neurologic findings.

Her pregnancy was unremarkable for preeclampsia or other complications. She had no past medical history, had no allergies, and took no medications or illicit drugs.

On physical examination, she was a nontoxic-appearing female. Her vital signs were as follows: blood pressure of 130/64 mm Hg, pulse rate of 100 beats/min, respiratory rate of 18 breaths/min, and an oral temperature of 36.8°C (98.3°F). She did not have orthostatic hypotension. Initial neurologic examination included cranial nerves, reflexes, strength, sensation, Romberg, and Babinski, results of which were all normal. She had no nystagmus or extremity edema. The remainder of the physical examination was unremarkable.

The patient was treated with 500 mg of caffeine administered intravenously and a normal saline solution bolus for suspected postepidural headache. Her headache resolved after therapy.

Before disposition, she began having eyelid twitching and complained of blurred vision. A full neurologic examination was repeated, and the patient was found to have nystagmus to the left, which extinguished after 30 seconds, and left homonymous hemianopsia. The remainder of the neurologic examination was normal. One milligram of lorazepam was administered intravenously.

Urinalysis revealed no protein or infection. CBC count, electrolyte level, liver function test results, and calcium level were unremarkable. Head computed tomographic scanning showed bilateral cerebral edema in the occipital lobe.

The patient continued to have multiple focal seizures without loss of consciousness or postictal period. Members of the neurology and obstetrics departments both evaluated the patient in the ED. She was given 2 more doses of lorazepam and a bolus of magnesium sulfate.

She became unresponsive, and respiratory depression was noted. An arterial blood gas measurement was sent as bagging was initiated, and it revealed the following: pH of 7.07, PO₂ of 157 mm Hg, and PCO₂ of 101 mm Hg. The patient was intubated without difficulty through a rapid sequence induction.

The cause of her initial ventilatory failure was the subject of some debate among the various specialists. It was due either to nontonic-clonic status epilepticus or to excess sedation from the medications. Carbon dioxide narcosis probably perpetuated the coma. The patient was loaded with phenytoin at the same time the carbon dioxide was being blown off by using mechanical ventilation. Thirty minutes after intubation, the patient was following commands.

She was admitted to the ICU, continued on magnesium sulfate, and started on dexamethasone for cerebral edema and heparin for possible cerebral venous thrombosis.

MRI, which included magnetic resonance venography, revealed no evidence of thrombosis. There were extensive white matter and cortical signal abnormalities involving the parietal and occipital regions bilaterally, which were considered “highly suggestive of preeclampsia” by the neuroradiologist.

When extubated on the first admission day, she was found to be blind. Her blindness gradually resolved over 24 hours. After 4 days of evaluation, the patient

was discharged on phenytoin for persistent, right-sided, focal facial seizure activity.

DISCUSSION

Posterior leukoencephalopathy syndrome is a reversible neurologic condition characterized by a headache followed by neurologic deterioration, including decreased alertness, altered mental status, seizures, and cortical visual disturbances, including blindness.¹ Posterior leukoencephalopathy syndrome is characterized by the presence of white matter edema predominantly affecting the occipital and parietal lobes. Posterior leukoencephalopathy syndrome has occurred in patients with hypertensive encephalopathy, renal disease, immunosuppression, and eclampsia in the postpartum period.²⁻⁴ Subsequent cases have been documented in other conditions, including collagen vascular disorders, thrombotic thrombocytopenic purpura, AIDS, and acute intermittent porphyria.⁵

The differential diagnosis for posterior leukoencephalopathy syndrome includes stroke, cerebral venous thrombosis, encephalitis, and demyelinating disorders. Early recognition of posterior leukoencephalopathy syndrome is paramount because the treatment of posterior leukoencephalopathy syndrome can be relatively contraindicated in some of these conditions. In particular, one would not necessarily want to treat hypertension (which could possibly decrease cerebral perfusion pressure and consequently blood flow) if the cortical blindness is being caused by stroke. Early imaging is essential to make this differentiation.

Computed tomography is not helpful for differentiation because several of these conditions are represented as hypodense parenchymal abnormalities. Hence, MRI is the modality of choice (Figure).^{1,6} This becomes particularly apparent when trying to differentiate posterior leukoencephalopathy syndrome from an acute ischemic event. Proper MRI evaluation should include echo planar diffusion-weighted imaging and apparent diffusion coefficient techniques.⁶ Diffusion-weighted imaging is dependent on the microscopic random motion of water molecules. Diffusion of water decreases in the early phase of ischemia, which demonstrates a hyperintense signal. The apparent diffusion coefficient, which displays the diffusion component only, is hypointense. In posterior leukoencephalopathy syndrome, the opposite occurs: the diffusion-weighted image is hypointense, whereas apparent diffusion coefficient imaging is hyperintense (Table).

Whether the cerebral edema witnessed with posterior leukoencephalopathy syndrome represents a cytotoxic or vasogenic process has been the subject of much debate.^{2,6} The newer MRI techniques have been instrumental in differentiating the pathophysiology. As stated previously, diffusion-weighted imaging is sensitive to diffusion of water molecules. In cytotoxic edema, water diffusion decreases. For instance, in acute ischemia, rapid depletion of high-energy phosphates leads to cessation of Na⁺-K⁺ adenosine triphosphatase activity, trapping water intracellularly. This decreased diffusion

Figure.

Gyral hyperintensities in the occipital lobe on an MRI fluid-attenuated inversion recovery (FLAIR) image of the patient in Case 1.



Table.

Differences between the mechanism and imaging between posterior leukoencephalopathy syndrome and ischemia.

	Mechanism	DWI	ADC
PLES	Vasogenic edema	Hypointense	Hyperintense
Ischemia	Cytotoxic edema	Hyperintense	Hypointense

DWI, Diffusion-weighted imaging; ADC, apparent diffusion coefficient; PLES, posterior leukoencephalopathy syndrome.

of water molecules is represented as hyperintensity on diffusion-weighted imaging and hypointensity on apparent diffusion coefficient imaging. On the other hand, posterior leukoencephalopathy syndrome is hypointense on diffusion-weighted imaging and hyperintense on apparent diffusion coefficient. This reversal in signal intensity is consistent with extracellular edema caused by increased permeability of the blood-brain barrier, supporting a vasogenic process.

Although several mechanisms have been proposed to account for the vasogenic edema, a loss of autoregulation in cerebral arterioles has gained the most support. In hypertensive states, animal models have shown that arteriolar resistance increases proportionately as blood pressure increases. This autoregulation appears to be governed by sympathetic innervation. However, with continued or acute increases of blood pressure, eventually a threshold is reached at which autoregulation fails. Then hypertension leads to arteriolar dilatation, blood-brain barrier damage, and extravasation of albumin and intravascular volume, causing extracellular edema.⁷⁻¹²

There is also evidence of relative differences in sympathetic innervation between different regions of the brain. Regions with more sympathetic innervation are more susceptible to disruption of the blood-brain barrier. Brain white matter demonstrates regional differences in autoregulatory innervation, with the parieto-occipital and posterior frontal regions being more susceptible to increased blood pressure.⁷ This explains the propensity of posterior leukoencephalopathy syndrome to affect the parieto-occipital white matter.

In case 1, this patient did have proteinuria and hypoalbuminemia. In addition, review of nursing documentation did demonstrate an abrupt increase in blood pressure of 180/110 mm Hg. Thus, her presentation is consistent with posterior leukoencephalopathy syndrome associated with eclampsia.

Case 2 is an enigma. Although the clinical picture and MRI findings are consistent with posterior leukoencephalopathy syndrome, the cause is unclear. The final consensus by her inpatient physicians was that her condition was caused by eclampsia. However, during her hospital course, there was never a documented increase in blood pressure or facial or extremity edema consistent with even mild preeclampsia. Also, multiple urinalyses failed to demonstrate proteinuria. An HIV test was performed, and the result was negative. Other causes for posterior leukoencephalopathy syndrome, such as collagen vascular disorders, were not investigated. In conclusion, the cause of this patient's poste-

rior leukoencephalopathy syndrome still seems unclear.

The prognosis for posterior leukoencephalopathy syndrome is usually quite good. Treating the underlying problem usually leads to symptom resolution without long-term sequelae, as was seen in case 1. However, there are documented cases in which patients with posterior leukoencephalopathy syndrome have had persistent seizures requiring prolonged therapy, as was seen in case 2. In these patients, the initial MRI exhibited small areas of cytotoxic edema consistent with ischemia interposed among the larger areas of vasogenic edema. Such changes can be explained by animal trials, which demonstrated that as marked vasogenic edema developed, regional cerebral blood flow decreased, causing small areas of cerebral ischemia.¹³

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Address for correspondence: Kimberly A. Chambers, MD, Department of Emergency Medicine, University of Texas–Houston Medical School, JLL Suite 443, 6431 Fannin, Houston, TX 77030; 713-500-7585, fax 713-500-0758; E-mail kimberly.a.chambers@uth.tmc.edu.

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