

Status Epilepticus Amauroticus Revisited

Ictal and Peri-ictal Homonymous Hemianopsia

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Objective: To describe the clinical, electrographic, and radiographic features of status epilepticus amauroticus, or homonymous hemianopsia associated with partial status epilepticus, in 3 patients with subsequent resolution of radiographic abnormalities and visual deficits.

Design: Case series.

Setting: Rancho Los Amigos National Rehabilitation Center in Downey, California, and the Los Angeles County + University of Southern California Medical Center.

Patients: One patient with a single remote seizure and 2 patients with symptomatic partial epilepsy all presented with homonymous hemianopsia.

Intervention: Continuous electroencephalographic monitoring, magnetic resonance imaging, and antiepileptic medical therapy for status epilepticus.

Main Outcome Measures: Neurologic examination, electroencephalography, and magnetic resonance imaging.

Results: The association of homonymous hemianopsia and restricted diffusion on magnetic resonance imaging

led to an initial diagnosis of ischemic infarction in 2 cases despite atypical diffusion-weighted imaging patterns. However, continuous electroencephalogram demonstrated focal epileptiform discharges in 2 cases and repetitive focal seizures in another, suggesting a diagnosis of status epilepticus amauroticus. Homonymous hemianopsia resolved in all 3 patients after escalation of the dosage of anticonvulsant therapy. Follow-up magnetic resonance imaging and electroencephalogram demonstrated complete or near-complete resolution of associated abnormalities.

Conclusions: Status epilepticus amauroticus is an uncommon but important cause of homonymous hemianopsia, and it should be considered in any patient with a history of seizures, fluctuating visual symptoms, or atypical patterns of restricted diffusion involving the occipital cortex. Continuous electroencephalographic monitoring is an important diagnostic tool for the diagnosis of status epilepticus amauroticus, which may have a favorable prognosis when treated with aggressive anticonvulsant therapy.

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SINCE THE ASSOCIATION BETWEEN blindness and epilepsy was first described by Gowers¹ in 1881, a variety of ictal or postictal visual symptoms have been described, including positive phenomena such as flashes or scintillations, and negative phenomena such as homonymous hemianopsia or cortical blindness. Cases of documented blindness due to seizures with electroencephalogram (EEG) correlates are rare, and we are aware of only a single previous report with corresponding magnetic resonance imaging (MRI) abnormalities.² Here, we describe 3 cases of status epilepticus amauroticus in which homonymous hemianopsia was the presenting sign of partial status epilepticus, along with correlative EEG and MRI abnormalities.

REPORT OF CASES

CASE 1

A 65-year-old man with diabetes mellitus and a remote history of 1 seizure was admitted for intravenous antibiotic treatment of right foot osteomyelitis. Several days after hospital admission, the patient reported decreased vision and scintillations. Ophthalmologic evaluation and retinal photography revealed proliferative diabetic retinopathy. Approximately 2 weeks after the onset of visual symptoms, the patient had a convulsion. The following day, neurologic examination revealed a right homonymous hemianopsia. The result of cerebrospinal fluid analysis was normal and MRI of the brain demonstrated hy-

perintense signal on diffusion-weighted imaging (DWI), fluid-attenuated inversion recovery (FLAIR), and T2-weighted sequences in the left parieto-occipital cortex with variable hypointensity and iso-intensity on apparent diffusion coefficient (ADC) (**Figure 1A**). Electroencephalogram demonstrated continuous, focal epileptiform discharges over the left posterior temporal lobe (**Figure 2A**). Levetiracetam therapy and empirical treatment for encephalitis were initiated; epileptiform discharges terminated after the addition of treatment with phenytoin sodium. Follow-up EEG was normal, and MRI 1 week later showed resolving abnormalities (**Figure 1A**). The patient's visual deficit resolved within 1 month.

CASE 2

A 36-year-old man with drug-resistant epilepsy since childhood and left hippocampal sclerosis presented with headache, subjective episodes of horizontal diplopia, and nausea. Neurologic examination demonstrated a right homonymous hemianopsia. The result of cerebrospinal fluid analysis was normal. Magnetic resonance imaging showed cortical hyperintensities of the left posterior temporal, parietal, and occipital lobes on DWI and FLAIR sequences, with a mild corresponding increase in ADC (**Figure 1B**). The overall radiologic impression favored subacute ischemic infarction, and comprehensive evaluation identified a patent foramen ovale that was closed for possible paradoxical embolism. However, follow-up MRI on hospital day 5 showed progression of DWI and FLAIR abnormalities with cortical edema and adjacent leptomeningeal enhancement. Close review of his medical history revealed that the patient had erroneously stopped levetiracetam treatment 2 weeks prior to presentation. Routine EEG showed sharply contoured focal slowing over the left temporal-occipital region, and treatment with levetiracetam was restarted (**Figure 2B**). The patient's homonymous hemianopsia resolved during several weeks, and follow-up MRI showed nearly complete resolution of cortical abnormalities (**Figure 1B**).

CASE 3

A 35-year-old man with a history of human immunodeficiency virus, toxoplasmosis encephalitis, and postinfectious seizure disorder presented with frequent simple partial seizures involving right-sided jerking. Examination was significant for a left homonymous hemianopsia, an extension of his baseline left upper quadrantanopsia. Magnetic resonance imaging demonstrated subtle DWI and FLAIR hyperintensity of the right occipital lobe, in addition to multifocal areas of stable encephalomalacia. Video EEG monitoring revealed brief repetitive seizures arising from the right occipital lobe, which gradually resolved after escalation of treatment with antiepileptic medications (**Figure 2C**). Neurologic examination several weeks after hospital discharge demonstrated a baseline sectantanopsia. Two years later, the patient had identical symptoms and MRI revealed a pattern of hyperintensity on trace DWI (isointense on ADC) within the temporal and occipital cortices in addition to more focal FLAIR hyperintensity and

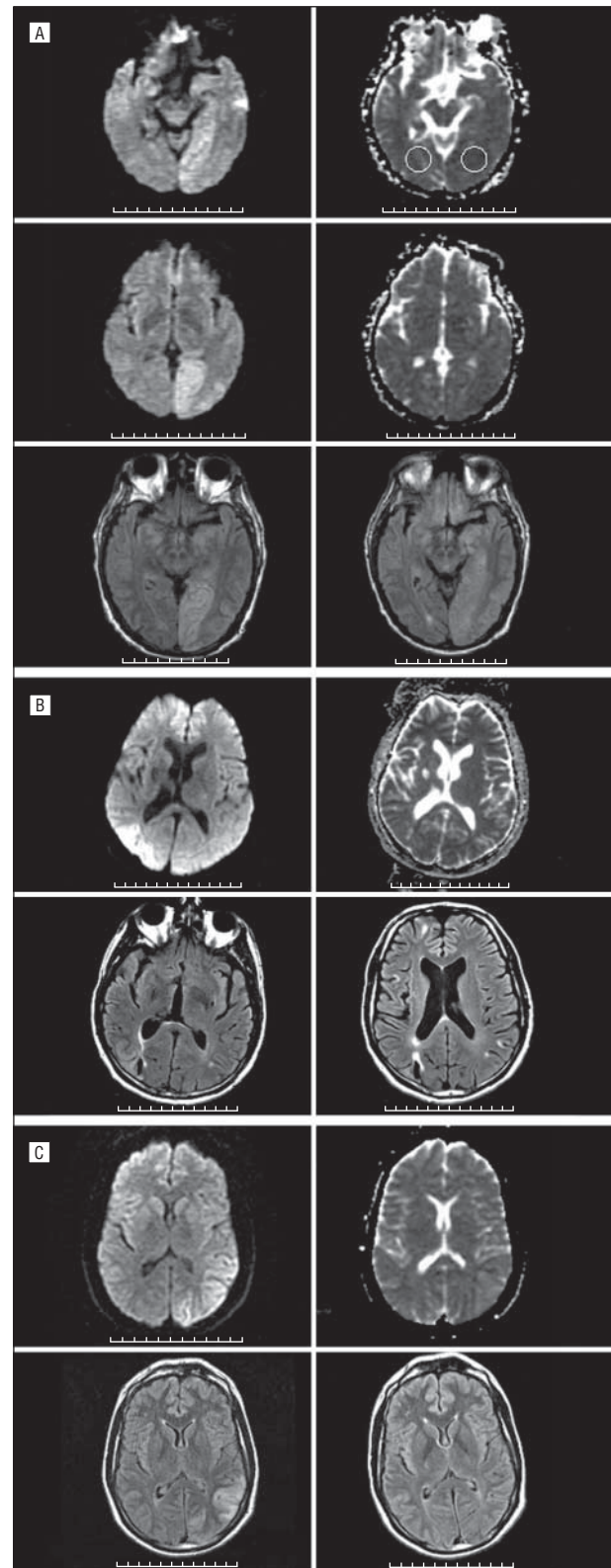


Figure 1. Neuroimaging of cases 1 (A), 2 (B), and 3 (C). Restricted diffusion on trace diffusion-weighted imaging corresponds to a 13.9% apparent diffusion coefficient reduction in a lower axial plane (A, upper right panel), while a higher axial plane reveals isointense apparent diffusion coefficient (A, middle right panel) and hyperintense fluid-attenuated inversion recovery similar to cases 2 and 3 (A-C, lower left panels). Follow-up fluid-attenuated inversion recovery images demonstrate resolution of hyperintensities at 1, 8, and 11 weeks, respectively (A-C, lower right panels).

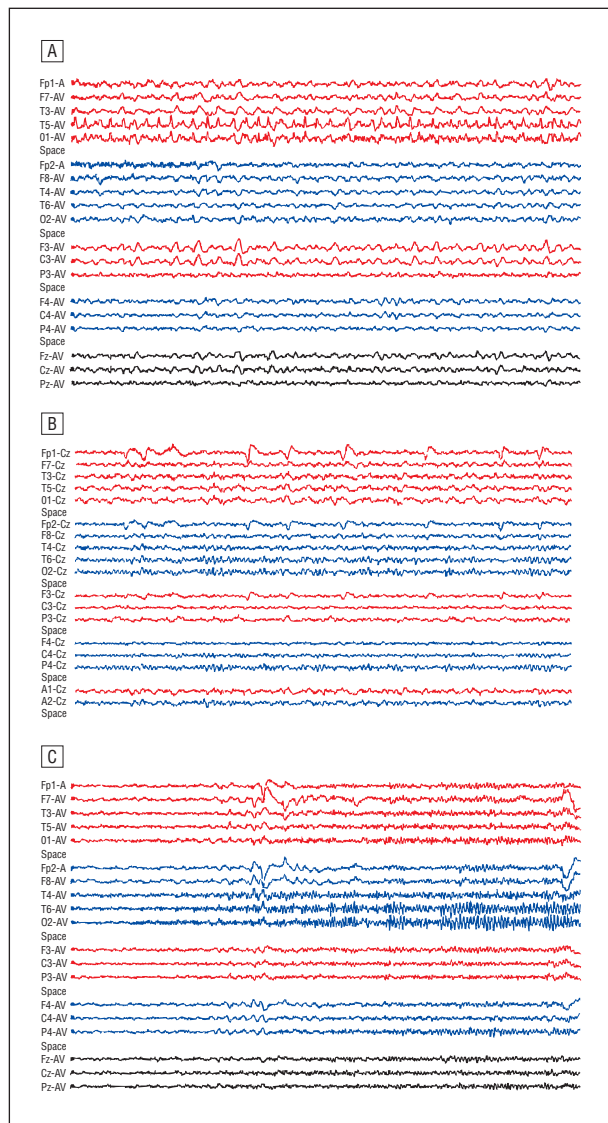


Figure 2. Ictal and peri-ictal electroencephalogram results. A, Continuous 3-Hz epileptiform discharges over the left posterior temporal region in case 1. B, Focal slowing over left temporal-occipital region in case 2. C, Right occipital seizures recurred every 2 to 5 minutes in case 3.

edema of the posterior temporal cortex (Figure 1C). Video EEG monitoring again demonstrated colocalized seizures and homonymous hemianopsia, and MRI abnormalities again resolved with escalation of the dosage of anti-epileptic medications.

COMMENT

Acute loss of vision has been recognized as a peri-ictal symptom since Gowers¹ first reported that 26 of 1000 cases of epilepsy were associated with an aura of vision loss “commonly complete and simultaneous in the 2 eyes.”^(p63) A half century later, Ayala³ coined the term status epilepticus amauroticus in describing transient blindness in a young man with poorly controlled seizures. Occasional cases of status epilepticus amauroticus have been reported,⁴⁻⁶ but to our knowledge, this is the first case series with corresponding EEG and MRI abnormalities in addition to

subsequent resolution of visual deficits and radiologic abnormalities.

While all 3 patients presented with homonymous hemianopsia and similar MRI findings, these cases highlight the variability in clinical presentation and EEG correlates in status epilepticus amauroticus. Two of our patients had active epilepsy at the time of presentation, while another reported only a single remote seizure. Homonymous hemianopsia and associated visual symptoms reportedly ranged from several days to 2 weeks. None of our patients were cognitively impaired; all 3 patients provided detailed histories and cooperated with bedside neurologic examination. However, associated clinical symptoms were variable and included fluctuations in vision, scintillations, diplopia, and simple motor seizures. With aggressive anticonvulsant therapy, visual fields returned to baseline during several weeks in all 3 patients.

Neuroimaging in all 3 patients revealed diffusion restriction characterized by hyperintensity on trace DWI with mild hypointensity, hyperintensity, and isointensity on ADC, as well as FLAIR and T2 hyperintensities along the posterior temporal, parietal, and/or occipital cortices. Subcortical white matter was relatively spared in all cases. This MRI pattern was interpreted as evidence for early subacute ischemic stroke in the first 2 cases despite extension of DWI and FLAIR hyperintensities over multiple vascular territories and prominent involvement of the cortical ribbon with relative sparing of subcortical white matter. Although arterial infarction is the most common cause of restricted diffusion, this MRI pattern is also well documented in seizures, partial status epilepticus, inflammatory disorders, neoplasm, abscesses, and prion disease.⁷ Quantitative assessment of postictal diffusion restriction has not been formally studied in humans, but animal models have demonstrated an initial mild increase in ADC, detectable 3 to 10 minutes after the onset of status epilepticus, followed by a pattern of depressed ADC that may last several days.^{8,9} Similar findings of decreased ADC have been reported in humans at 48 hours following acute symptomatic seizures and status epilepticus.^{10,11} This temporal evolution of decreasing ADC in status epilepticus contrasts with the pattern seen in ischemic stroke, where ADC is significantly decreased in the acute phase, followed by a gradual increase in ADC during several days.¹² The modest ADC changes present in status epilepticus, combined with these opposing patterns of temporal evolution in status epilepticus and ischemic stroke, limit the clinical use of ADC in the diagnosis of status epilepticus amauroticus.

Typical locations of restricted diffusion in complex partial status epilepticus include the cerebral cortex, hippocampal formations, pulvinar, and contralateral cerebellum.¹³⁻¹⁵ Restricted diffusion is considered a marker of irreversible parenchymal injury in the setting of acute ischemic stroke, yet these cases demonstrate that visual deficits and DWI abnormalities may both normalize in status epilepticus amauroticus as in other cases of partial status epilepticus. It is thought that these divergent outcomes occur because DWI abnormalities in status epilepticus result from a combination of impaired energy metabolism and hemodynamic changes rather than hypoxia/anoxia present in acute ischemic stroke.¹³⁻¹⁵

Continuous EEG supported the diagnosis of status epilepticus amauroticus in all of our cases, although electrographic patterns varied considerably. In case 1, continuous EEG revealed periodic epileptiform discharges over the left posterior temporal region corresponding to the area of DWI and FLAIR hyperintensity. Continuous EEG monitoring in case 3 demonstrated a different pattern of repetitive, brief focal seizures (Figure 2C). Electroencephalogram was delayed in the second case owing to management of a presumed ischemic stroke and likely represents an interictal or postictal study with sharply contoured focal slowing over the corresponding region of DWI and FLAIR hyperintensity.

Homonymous hemianopsia is a common presenting sign of an acute neurologic condition, associated with stroke, trauma, and tumor in more than 90% of cases in 1 large series.¹⁶ However, a clinical history of seizures, fluctuating visual symptoms, or atypical patterns of restricted diffusion should immediately raise suspicion for status epilepticus amauroticus. Importantly, continuous EEG monitoring may be required to firmly establish this diagnosis because frank seizures or epileptiform discharges may not be evident on a routine study as in case 2. Prompt recognition of status epilepticus amauroticus will prevent unnecessary interventions associated with the management of presumed ischemic stroke, and appropriate treatment with anticonvulsant therapy will likely hasten and/or improve the neurologic outcome of many patients.

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