

Transient Attenuation of Visual Evoked Potentials during Focal Status Epilepticus in a Patient with Occipital Lobe Epilepsy

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Abstract-

Purpose: Seizures originating in the occipital areas are relatively uncommon. They are usually characterized by visual hallucinations and illusions or other symptoms related to the eyes and vision.

Case Report: In a 54-year-old woman with occipital lobe epilepsy, complex visual hallucinations, illusions, and migraine-like headache constitute the major clinical manifestations. During focal status epilepticus, ictal electroencephalography revealed rhythmic focal spikes in the right occipital region, rapidly propagating to the right parietal and contralateral occipital areas. Ictal brain single-photon emission computed topography revealed hyperperfusion of the right occipital region. Using a full-field pattern-shift visual evoked potential (VEP) study, we found that the P100 responses on both sides were markedly attenuated in amplitude during occipital focal status epilepticus, whereas the latencies of the VEPs were normal. The amplitude and morphology of P100 responses on both sides, however, returned to the normal range 7 days after cessation of the seizures.

Conclusion: In addition to clinical seizure semiology, scalp EEG, SPECT and neuroimaging studies, VEP studies may be used as a supplementary examination tool to provide further information in the patients with occipital lobe seizures or epilepsies.

Key Words: occipital lobe epilepsy, visual evoked potentials, visual hallucinations, visual illusions, focal status epilepticus.

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INTRODUCTION

Seizures originating in the occipital lobe area are relatively uncommon. They are usually characterized by visual hallucinations and illusions or other symptoms related to the eyes and vision.⁽¹⁾ As occipital lobe epilep-

sy may emulate other epileptic syndromes and rapidly propagate outside the occipital lobe, they could be an under-recognized problem.⁽¹⁾ The seizure semiology, interictal and ictal electroencephalography (EEG) and neuroimaging studies may help in the diagnosis of occipital lobe epilepsy and identification of the underly-

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ing pathology.^(1,2)

The occipital lobe is the main center of the visual system. Visual evoked potential (VEP) recordings have been widely applied to evaluate patients with visual problems in many neurological disorders.⁽³⁾ However, the experiences of VEPs in patients with occipital lobe epilepsy are limited, particularly those during the ictal state. Herein, we describe the seizure semiology and ictal as well as interictal findings of EEG, single-photon emission computed topography (SPECT) and VEPs in a patient with occipital lobe epilepsy. We show that the P100 amplitude of VEP responses was transiently attenuated during occipital focal status epilepticus.

CASE REPORT

A 54-year-old woman suffered from acute onset of visual hallucinations and migraine-like headache in the evening before admission. The visual hallucinations usually lasted several minutes in duration but would cluster with a frequency higher than ten times per hour. The visual symptoms consisted of strong flashing lights, phosphenes and colored spots, circles, lines or characters in Chinese or English (Fig. 1C). These hallucinations usually appeared in her left visual field, and then would spread to involve the entire visual field, even when her

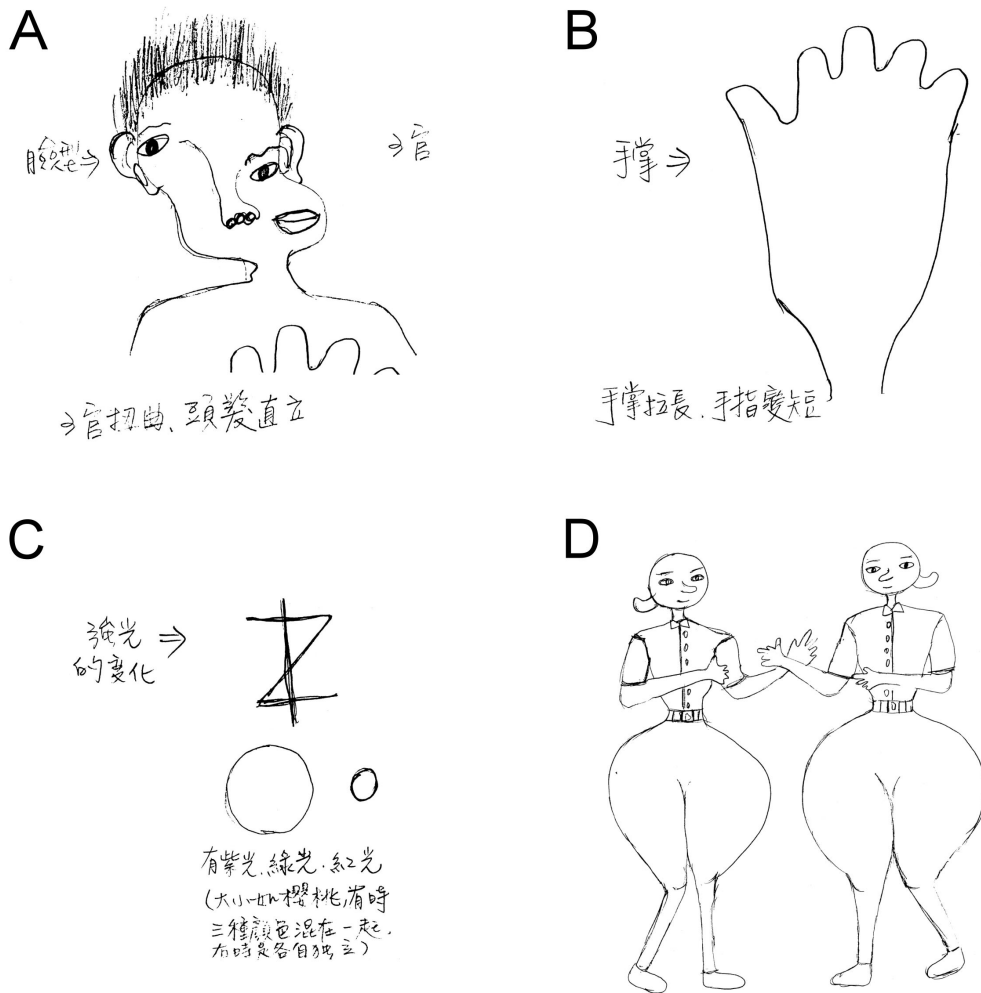


Figure 1. The patient drew her visual hallucinations and illusions. As she saw it, the persons (A) or objects (B) were distorted in size, shape, color or clarity. (C) Strong flashing lights, phosphenes and colored spots, circles, lines or characters were seen by the patient during the seizures. (D) She also saw two small persons jumping and rapidly fighting each other.

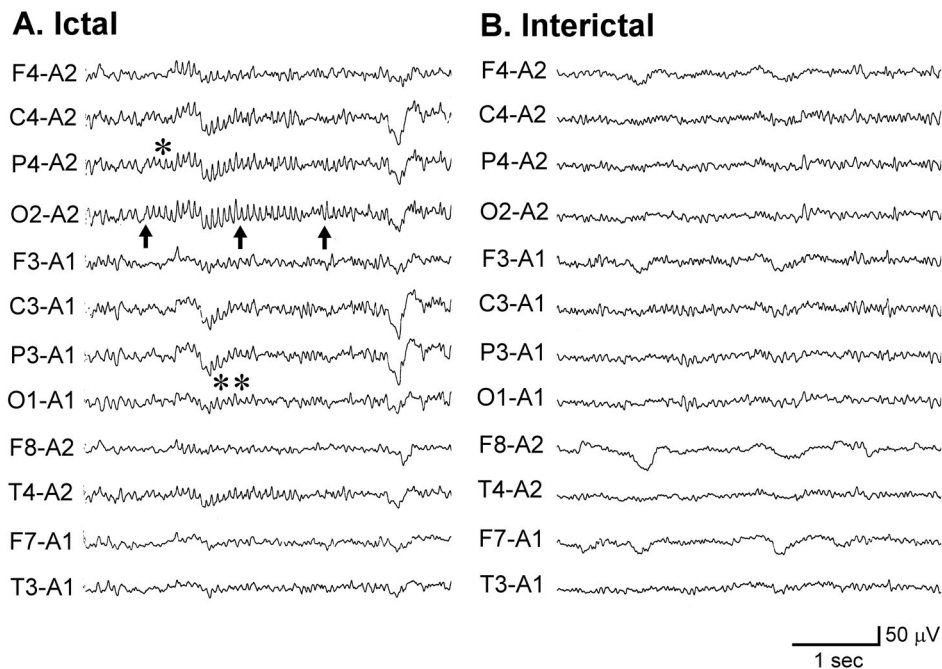


Figure 2. Ictal (A) and interictal (B) EEG in the patient. The ictal EEG shows focal spikes in the right occipital region (arrows), rapidly propagating to the right parietal (*) and contralateral occipital areas (**).

eyes were closed. The objects looked distorted in size, shape, color or clarity (Fig. 1A and B). Sometimes, she seemed to see two small persons jumping and rapidly fighting each other in front of her (Fig. 1D), especially when the other symptoms were more severe. During the attacks, she was sensitive to strong lights and large sounds. The headache was located in the right parietal-occipital areas and was pulsating in nature, associated with nausea but not vomiting. The headache would usually aggravate following the visual symptoms or when moving or changing position. There was no loss of consciousness, limb convulsion or facial twitching.

According to the history, she had her first seizure characterized by flashes of lights and subsequent generalized convulsions. She had received phenytoin therapy (300 mg/day) for 3 years and remained seizure free. The patient's gestational and neonatal history was unremarkable, with normal developmental milestones. There was no history of major systemic or infectious diseases, nor a family history of epilepsy.

During admission, the headaches and visual halluci-

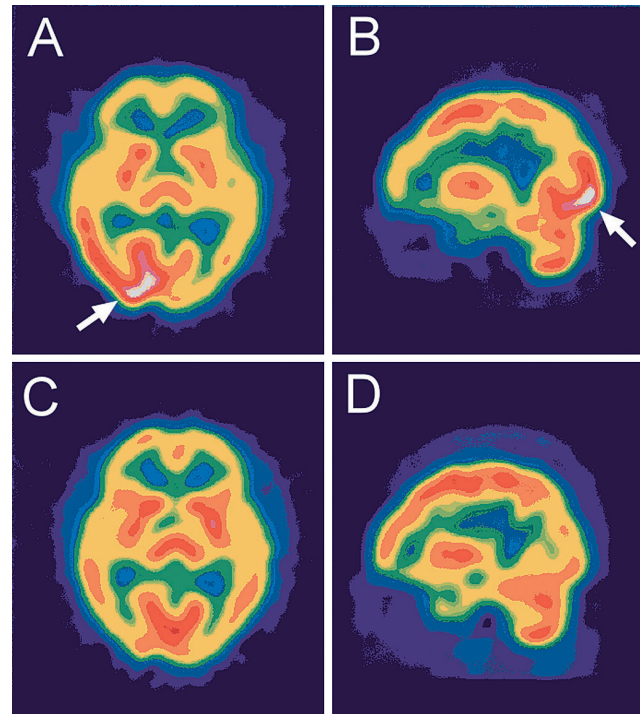


Figure 3. Ictal (A) and interictal (B) brain ^{99m}Tc SPECT in the patient. Ictal SPECT revealed regional hyperperfusion in the right occipital region (arrows).

nations became more frequent and persisted for nearly a whole day. An ictal EEG study revealed rhythmic focal spikes in the right occipital region (Fig. 2), accompanied by clinical visual hallucinations. The spikes also propagated to the right parietal lobe and contralateral occipital area (Fig. 2). Brain magnetic resonance imaging showed a cystic lesion in the right occipital lobe. Ictal brain ^{99m}Tc SPECT revealed a hyperperfusion state in the right occipital area, whereas the interictal scan showed hypoperfusion in the same region (Fig. 3).

We also performed pattern-shift VEPs for this patient during the occipital focal status epilepticus and 7 days after cessation the seizures. VEPs were obtained by using a black-and-white checkerboard displayed on a

television screen. Recordings were performed in a dark room after monocular full-field stimulation with the active scalp electrodes at O1, O2 and Oz, referenced to linked-ears (A1+A2). The ground electrode was placed around the forehead. The frequency limits were set at 1-100 Hz and the analysis time was 250 ms. A total of 200 responses were averaged. We demonstrated that the latencies of pattern-shift VEPs were within normal limits, but the amplitude of P100 was markedly attenuated during the attacks (Fig. 4A). A follow-up VEP study 7 days after cessation of the seizures showed that the P100 on both sides had returned to normal (Fig. 4B).

The patient received intravenous phenytoin therapy immediately after admission, and the seizures gradually

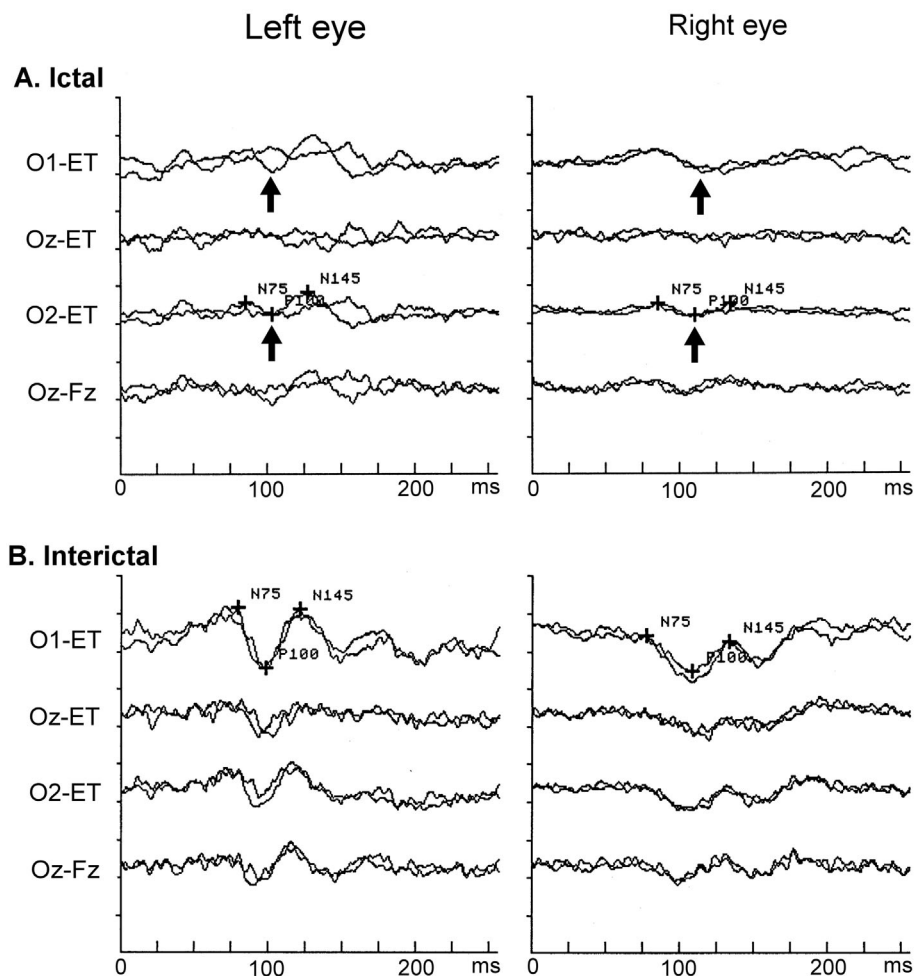


Figure 4. Ictal (A) and interictal (B) full-field pattern-shift visual evoked potentials (VEPs) in the patient. (A) There was a marked attenuation of P100 amplitude on both sides (arrows) during the ictus. (B) The follow-up VEP study 7 days after cessation of seizures showed normal amplitude, morphology and latency of P100 responses on both sides. ET: linked-ears reference.

decreased. Under the combination therapy of phenytoin (300 mg/day) and topiramate (300 mg/day), the patient reached a seizure-free state and was then followed at the out-patient department.

DISCUSSION

Visual hallucinations or illusions are the hallmark of occipital seizures.^(1,4,5) In this patient, the characteristics of visual phenomena during attacks included flashing lights, phosphenes, colored spots, lines or geometric forms, and illusions with distorted objects with regards to size (macropsia or micropsia) and shape (metamorphopsia). Ictal EEG showed focal rhythmic spikes in the right occipital lobe, accompanied with visual hallucinations and illusions that led to the diagnosis of occipital lobe epilepsy. In addition, regional hyperperfusion in the right occipital area on ictal SPECT and a good response to antiepileptic drug therapy further confirmed our diagnosis.

VEPs are useful supplementary electrophysiological tools in the diagnosis of neurological disorders associated with visual problems, such as multiple sclerosis, optic neuritis, neurodegenerative diseases, or cortical blindness.⁽³⁾ However, VEPs have infrequently been studied in patients with epilepsy, particularly with occipital lobe seizures. The results of VEP studies during the ictal status of occipital lobe seizures are variable or even controversial. Interictal VEP studies on patients with child epilepsy with occipital paroxysms^(6,7) or symptomatic occipital epilepsy⁽⁷⁾ have shown that the amplitude of VEPs was increased but the P100 latency was delayed. The finding of increased amplitude of P100 is possibly ascribable to hyper-excitability of the occipital structures, whereas the prolonged P100 latency is probably related to structural changes in the occipital lobe.⁽⁷⁾

In an earlier report, Hughes et al.⁽⁸⁾ showed preserved and even increased P100 amplitude during ictal seizure activities in a patient with right occipital lobe seizures. Recently, Wang et al.⁽⁹⁾ also reported VEP findings in a patient with left occipital lobe status epilepticus induced by hyperglycemia. The amplitude of P100 was significantly higher in the right occipital lobe during status epilepticus, but became slightly higher in the left occipi-

tal area 6 months after being seizure free. Enhanced excitability or disinhibition of the epileptogenic occipital circuits has been suggested to be the causative mechanism of increased P100 amplitude.^(8,9) However, Zumsteg et al.⁽¹⁰⁾ reported a case of left occipital status epilepticus with hemi-field VEP studies, which demonstrated distorted VEP morphology topographically restricted to the seizure-generating left occipital region during right hemi-field stimulation. The VEP responses were normal 2 days later with cessation of the status epilepticus.

Our patient also had focal status epilepticus involving the right occipital lobe. The vivid visual hallucinations and complex illusions initially appeared in her left visual field, and then spread to the entire visual field. The ictal EEG showed focal spikes in the right occipital region that rapidly propagated to the right parietal and contralateral occipital areas. With full-field VEP test, we found that the P100 amplitude was markedly attenuated on both sides during the ictal period of occipital lobe status epilepticus. However, the amplitudes and morphology of P100 responses on both sides returned to normal 7 days after cessation of the seizures. Despite different types of stimulation, the transient distorted visual images and attenuated amplitude in VEPs during the ictal state are consistent with the reported case by Zumsteg et al.⁽¹⁰⁾ The exact mechanism underlying the changes of VEP responses during occipital lobe status epilepticus remains uncertain. Although the epileptogenic cortical neurons may well be abnormally excited during ictal seizures, the "excitation" or "disinhibition" of the epileptogenic circuits does not necessarily result in simultaneous augmentation of normal physiologic functions.^(11,12) Based on the VEP data, the present study provides novel evidence to demonstrate attenuation of normal VEP responses during occipital focal status epilepticus even when the patient presented with the positive phenomena such as visual hallucinations. We would therefore suggest that the attenuation or distortion of VEP responses may be related to the inhibitory effect of epileptic activities, which inhibits normal cortical functions and interferes with the neural networks for VEP generation during persistent occipital seizures. However, the exact interactions between the neural networks

responsible for VEP generation and those for epileptogenesis in occipital seizures require further investigation.

We are aware that the migraine-like headache in our patient could lead to misdiagnosis such as migraine with aura. The visual auras in classic migraine are usually achromatic, linear or zigzagged, and usually start from in the central field. In contrast, the visual symptoms in occipital lobe seizures are usually colorful, with bright balls or circles, and start in the visual field.^(13,14) The colorful and complex visual hallucinations and illusions in our patient suggested the diagnosis of occipital lobe seizures. A detailed clarification of history and drawings of the visual symptoms by the patients themselves, as demonstrated in this case report, may also be helpful for the differential diagnosis.

In conclusion, visual hallucinations and illusions may constitute the chief presentations of occipital lobe seizures. The VEP responses could be attenuated during status epilepticus involving the occipital lobe. Therefore, in addition to clinical seizure semiology, scalp EEG, SPECT and neuroimaging studies, VEP studies may be used as a supplementary examination tool to provide further information in the patients with occipital lobe seizures or epilepsies.

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