



CASE REPORT

Ictal paralysis with tonic eye gazing mimicking a pontine infarction

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KEYWORDS

Focal akinetic seizure;
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Summary

Purpose: Concomitant positive and negative motor phenomena in a single seizure have not been reported before.

Method: We used an extensive history review, neurological examination, EEG, MRI and SPECT study to demonstrate a rare combination of motor presentations as an ictal phenomenon.

Result: A 64-year-old male was brought to the emergency room with dizziness, progressive drowsiness and left hemiparesis. A spontaneous eye deviation to the left side with nystagmus was observed. A right pontine lesion was tentatively diagnosed. However, a focal motor seizure of the patient's left face and limbs occurred 3.5 h later. A brain MRI revealed a high signal in the right amygdala, hippocampus and thalamus, instead of the pons. An EEG showed periodic epileptic discharges in the right posterior temporal parietal region. Regional hyperperfusion was found by brain SPECT. The level of consciousness improved dramatically after adequate phenytoin treatment.

Conclusion: A posterior temporal–parietal seizure can present with a prolonged ictal paralysis, a positive ocular nystagmoid deviation and an altered level of consciousness. The EEG is essential for a correct diagnosis, especially with a negative or an unexplainable MRI study. The SPECT has an additional role for the differential diagnosis.

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Introduction

Seizures are frequently associated with positive phenomena, including motor (tonic, clonic, myoclonic

and automatic) and non-motor (elementary sensory and experiential) symptoms.¹ A short-lasting transient negative motor symptom, i.e., limb paralysis, is rarely reported as an ictal phenomenon.²

Focal atonic seizures are partial seizures with ictal paresis or paralysis of one or more parts of the body, which must be distinguished from the more common postictal Todd paralysis. Ictal paralysis

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(usually hemiparesis) may be preceded or accompanied by a somatosensory aura.¹

However, a prolonged focal limb weakness associated with a positive eye sign (nystagmoid deviation) has not been reported before. We describe the history, electroencephalography (EEG), magnetic resonance imaging (MRI) and single photon emission computed tomography (SPECT) of a patient whose seizure phenomenon mimicked a pontine lesion as the initial presentation.

Case report

A 64-year-old hypertensive and diabetic male had a history of an old right parietal lobe hemorrhage about half a year previous. He had no motor weakness, but sensory disturbance at that time. Later, the neurological deficits resolved completely and he did not receive medical follow-up thereafter. On 15 March 2006, he was brought to the emergency room with the presentations of a sudden onset of progressive drowsiness and left-side limb weakness. He was in his friend's yard and complained of dizziness and sleepiness after arising from a squatting position. After resting for several minutes, he found his left-side limbs were mildly weak and he could step into the car. He was driven home by his friend. On arrival, he could not get up from the seat and needed others to carry him out. He could correctly recall his son's phone number and called asking for help. Because of the progressive limb weakness, he was sent to the emergency department immediately. The initial vital signs were: body temperature 36.8 °C, heart rate 106 bpm, respiration rate 20 min⁻¹ and blood pressure 255/128 mmHg. He did not complain of headache or nausea. The neurological examinations revealed a drowsy consciousness with impaired orientation and spontaneous verbal expression. He could barely follow verbal commands. A neglect sign to simultaneous deep pain stimulation was absent. The pupil sizes were normal and reacted promptly to the light. His head tended to turn to the left side. He was unable to look to the right side and the eyes deviated to the left side with a spontaneous horizontal nystagmus to the left side. The bilateral face seemed weak with a diminished facial expression to pain stimulation. His left upper limb was completely paralyzed. The left lower limb was only able to move slightly. His tendon reflexes were diffusely absent. The plantar reflexes showed no response bilaterally. The bilateral limbs could sense pain to a similar degree. Laboratory examinations revealed an elevated sugar level of 412 mg/dl, using the

finger stick method. The serum ketone bodies and osmolality were not checked at that time. Although hyperglycemia can be associated with focal neurological symptoms and signs, the finding of a gaze palsy to the right side, together with a spontaneous nystagmus, a left hemiparesis and a depressed consciousness level, suggested a right pontine lesion involving the paramedian pontine reticular formation, central vestibular pathway, pyramidal tract and ascending reticular activating system. A brain CT revealed an old cerebral malacia lesion in the right parietal lobe. No intracranial hematoma or obvious hypodensity lesion could be found. Insulin and normal saline were given to lower the blood sugar level and enhance hydration, under the impression of acute pontine infarction and hyperglycemia.

After admission to the intensive care unit about 3.5 h from the onset, he suffered a motor seizure presenting as left facial and upper limb clonic movements with a loss of consciousness. The seizure stopped 2 min later. He appeared generally flaccid with comatose consciousness post-ictally. The serum biochemistry studies revealed hyperosmolar hyperglycemia (osmolality 300 mosmo/kg H₂O, sugar 440 mg/dl) without detectable ketone bodies. Lorazepam 2 mg was given intravenously. Phenytoin was prescribed with a daily dose of 300 mg. Enhanced hydration and insulin therapy was also given. For the hyperglycemic effect of phenytoin, the finger sugar and insulin were closely monitored during the treatment. Two hours later, his consciousness improved to the pre-seizure state, but was not completely normal. The follow-up sugar level, using finger sticks, was 286 mg/dl. Later, about 9 h from the onset, the left hemiparesis improved to the point of being able to moderately resist the testing maneuvers. EEG revealed periodic lateralized epileptiform discharges (PLED) in right temporoparietal region (Fig. 1). A brain MRI study performed on the second day showed an absence of ischemic change in the pons. Cytotoxic edematous lesions in the right temporal lobe, along with the body and tail of the hippocampus and the ipsilateral pulvinar nucleus of the thalamus, were found (Fig. 2). A brain SPECT performed on the third day showed a hyperperfusion area in the right temporo-parietal cortex (Fig. 3). During these days, the patient felt drowsy and irritable. He was disoriented to place and could not completely follow up verbal commands. He had the visual hallucination of seeing his sons crying outside of his room. The follow-up EEG on the fourth day was similar to the first one. The serum level of phenytoin was 3.49 µg/ml. A loading of 800 mg of phenytoin was given intravenously. His consciousness became

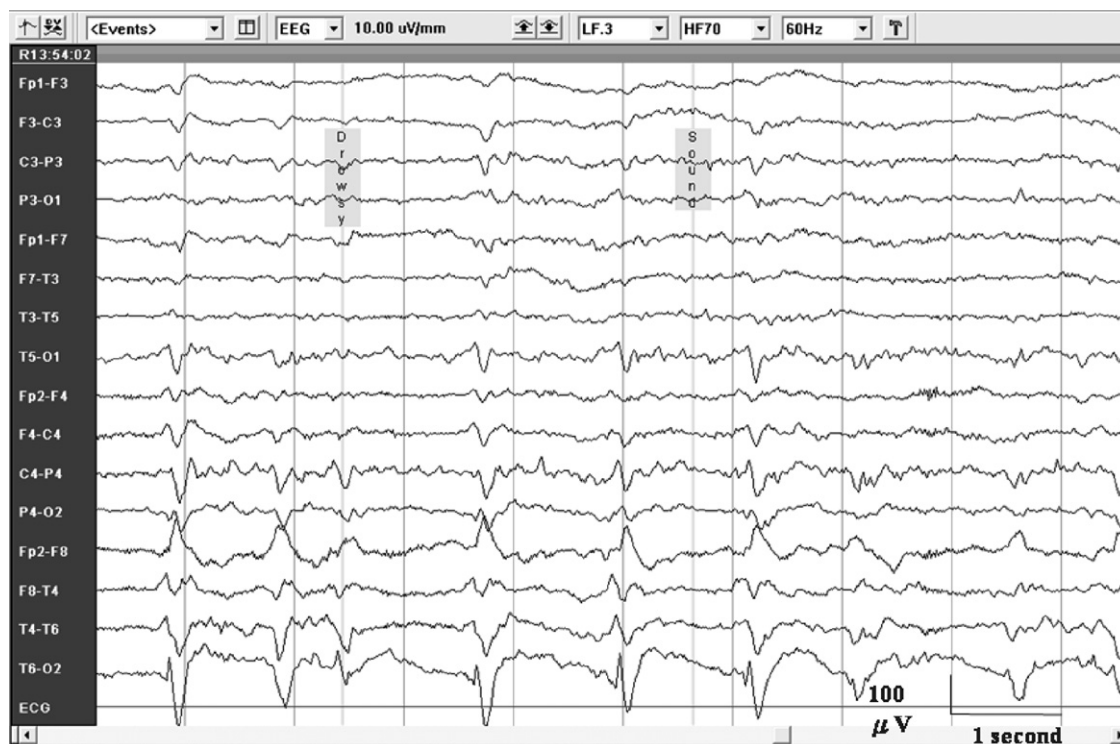


Figure 1 The EEG in confusion state revealed periodic epileptic discharges in the right temporal–parietal region.

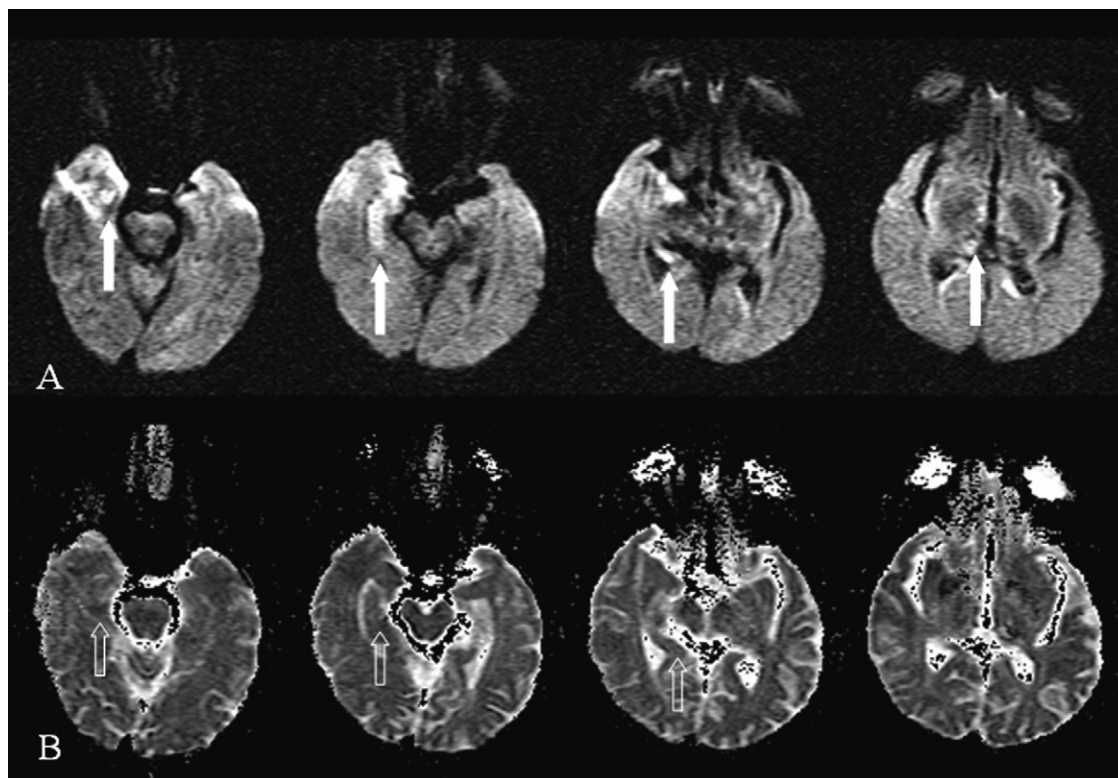


Figure 2 DWI high signal (solid arrow) and ADC low signal (arrow) lesions in the right temporal lobe, along with the body and tail of the hippocampus and the ipsilateral pulvinar nucleus of the thalamus. (A) Diffusion-weighted image (DWI). (B) Apparent diffusion coefficient (ADC) map.

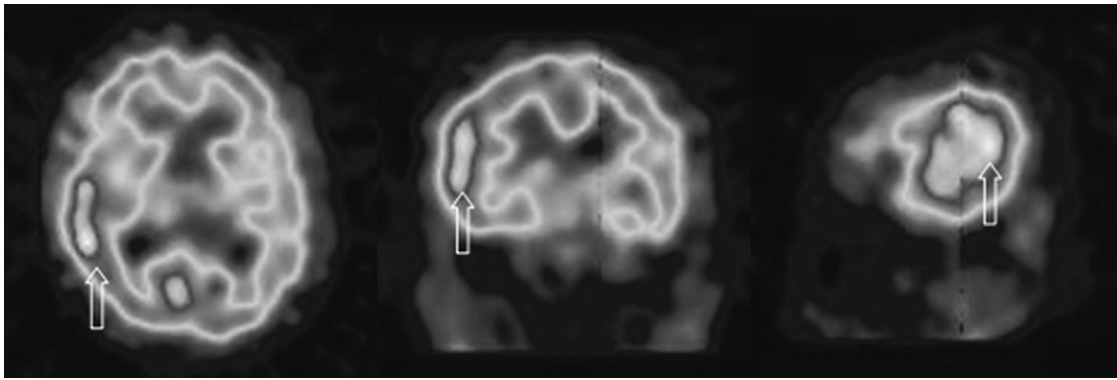


Figure 3 A ^{99m}Tc -ECD SPECT performed on the third day showed a hyperperfusion area (arrow) in the right temporal–parietal cortex.

completely alert on the fifth day. The phenytoin was shifted to carbamazepine for the hyperglycemic effect of phenytoin. The follow-up EEG showed a decreased amount of PLEDs (Fig. 4). A complex partial status epilepticus was diagnosed. A detailed history was taken by direct contact with his friend, the witness. His friend stated that the patient had arisen from the squatting position and then walked around the yard, seeming to search for things. The patient had put his helmet on and taken it off, repeatedly. The abnormal behavior, being considered a probable automatism, was not reported by the patient and his son.

Discussion

This patient's seizure initially presented with a negative motor symptom (left hemiparesis), a positive ocular sign (versive nystagmoid eye movements to the left side) and an alteration of the level of consciousness. The above combination was similar to that of patients with a right pontine infarction. But, the subsequent focal motor seizure alerted the medical staff to the possibility of a seizure with a right posterior temporal–parietal focus that was later confirmed by EEG, MRI and SPECT studies.

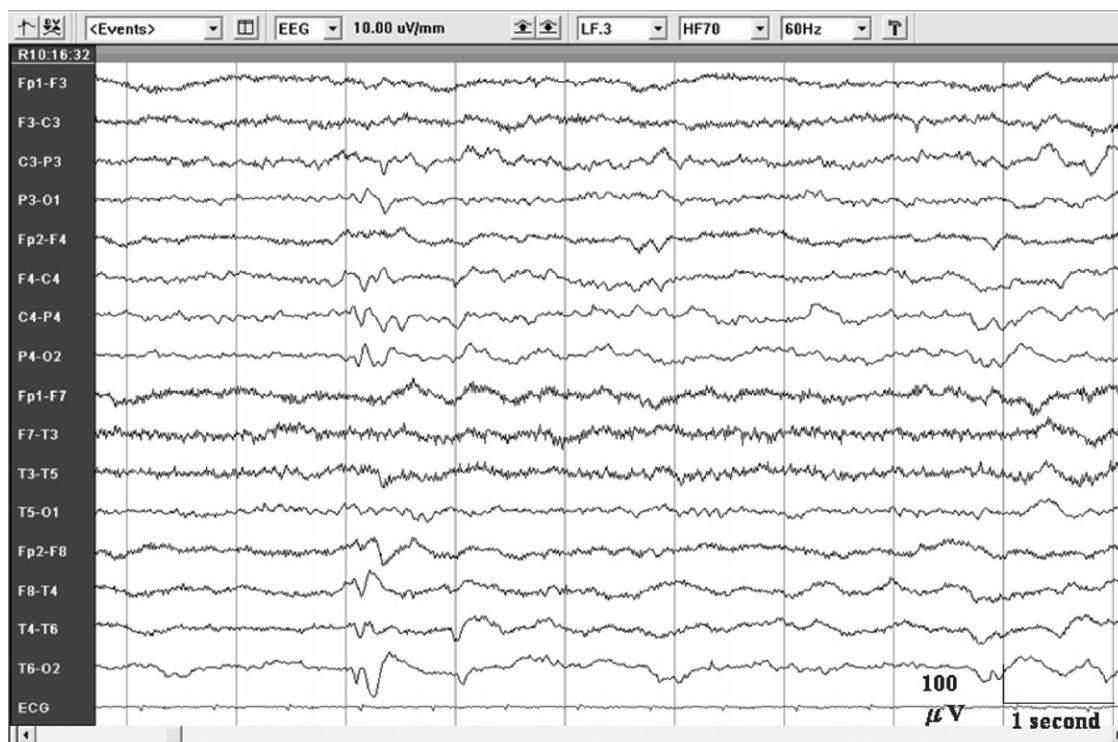


Figure 4 The follow-up EEG after adequate phenytoin treatment showed a decreased amount of periodic epileptic discharges.

Due to the prolonged and fluctuating consciousness disturbance, despite lorazepam treatment, we were concerned about the possibility of complex partial status epilepticus. In complex partial status epilepticus, a non-convulsive status epilepticus, the motor phenomena are subtle or absent. Fluctuating confusion and an altered level of consciousness were the main symptoms.³ Eye movement, including sustained gaze deviation, eyelid fluttering and persistent nystagmus, may be observed.⁴ The diagnosis of a complex partial status epilepticus necessitates an ictal EEG pattern with an effective antiepileptic drug trial.⁵ The focal periodic pattern of epileptic discharges in the EEG, fluctuating confusion and altered level of consciousness quickly and dramatically improved after an adequate phenytoin treatment that established the diagnosis.

The diagnosis of complex partial status epilepticus can also be supported by brain MRI and SPECT studies. A previous study reported that MRI findings in patients with complex partial status epilepticus could present a large area of increased signals on diffusion-weighted imaging, with decreased signals on apparent diffusion coefficient imaging with a cortical gyriform pattern.⁶ Sometimes, the ipsilateral thalamus and the contralateral cerebellum also show similar signal changes, as in the cortex.⁷ Increased perfusion had been demonstrated by perfusion-weighted imaging or by brain SPECT.⁷ A temporarily enlarged ipsilateral intracranial artery, e.g., the middle cerebral artery, may be seen on MRA.⁶ Our patient's MRI showed hyper-intensity in the right hippocampus, including the head, body and tail and in the ipsilateral thalamus. This is not seen in patients with stroke and the location could easily exclude the possibility of a pontine infarction. On the other hand, the hyperperfusion in the right temporoparietal cortex demonstrated by brain SPECT clearly disclosed the epileptic nature.

Prolonged limb paralysis has been reported in a few patients with a past history of convulsive epilepsy, but not as the first presentation.^{8–10} Epileptic ictal paralysis is an interesting special presentation of seizures. Together with cerebral ischemia and migraine, it is listed in the differential diagnosis of focal limb weakness. It can be found to precede a motor convulsion in the same limb during a seizure that often lasts less than a few minutes, or it may be associated with a simultaneous convulsion of the contralateral limbs.¹¹ Despite the fact that episodic recurrent inhibitory seizures mimicking crescendo transient ischemic attacks have been reported,¹² the initial fixed worsening paralysis (3.5 h before the onset of convulsive seizure) in association with positive ocular phenomena in our patient made the tentative diagnosis of a pontine infarction

reasonable. After the occurrence of a focal motor seizure, epileptic ictal paralysis is favored, based on the criteria, "focal paralysis immediately preceding convulsive movements in a limb", proposed by Fisher.¹¹

Focal atonic seizures may have a long duration (longer than 30 min), similar to our case's presentation and, therefore, may present as status epilepticus. In Guerrini et al.'s study, ictal EEG was recorded in 30 patients and showed spike-wave or slow wave discharges (53%) or rhythmic ictal activity (47%) in the frontal or centroparietal areas (63%), in the temporal lobe (20%), or distributed over a wide scalp region (17%) contralateral to the paralyzed limb.¹³ In our case, the EEG showed periodic epileptic discharges in the right parieto-temporal area.

The mechanisms of negative motor phenomena during a seizure are not well understood. Ictal negative motor phenomena have been reported in a frontal, central, or parietal epileptogenic focus.^{2,13,14} In Satow et al.'s report, the interictal FDG-PET disclosed hypometabolic regions consistent with the clinical and EEG findings. In our case, the ictal EEG and SPECT were localized in the right parieto-temporal area.

In the two patients with an ictal electrocorticogram, the seizure discharges involved the mesial frontal or primary sensorimotor cortices.^{2,15} Lüders et al. demonstrated a "negative motor effect" with the electrical stimulation of the inferior perirolandic cortex and the supplementary motor area.¹⁶ Matsumoto et al. observed epileptic discharges in the positive arm motor area of the right precentral gyrus and in its rostral area, but not in the negative motor area. They hypothesized epileptic activity probably inhibited the spinal motoneuron pool without eliciting excitatory activity in the corticospinal pathway¹⁵. In a stimulation study, electrical stimulation of the extrarolandic sensory areas, not the primary sensory cortex, was shown to have an inhibitory effect on voluntary movement.¹⁷ Our patient had a right posterior temporal–parietal epileptogenic focus, which might have involved the extrarolandic sensory areas that exhibited the ictal contralateral limb paralysis.

We postulate that the patients' seizure activity propagated to three areas: the mesial temporal region, resulting in an altered level of consciousness and initially abnormal behavior (wandering, searching and putting his helmet on and taking it off), which was probably one form of automatism; the right occipital region, causing a head turning and nystagmoid eye deviation to the left side and the extrarolandic sensory area, rendering his left-side limbs paralyzed. The relatively short-duration (3.5 h) ocular and limb signs improved after a focal

motor convulsion that suggested an involvement of the frontal motor cortex. However, the prolonged (3 days) alteration of consciousness might have resulted in greater metabolic stress in the right mesial temporal lobe, which showed a cytotoxic edematous change on diffusion-weighted MRI.

In conclusion, the clinical presentation of a posterior temporal–parietal seizure, an unusual combination of an ictal paralysis of the contralateral limb and a positive epileptic ocular deviation to the contralateral site without a limb convulsion, may confuse the bedside localization or lateralization of the underlying neurological pathology. A detailed history, with direct contact with a witness, emphasizing the abnormal repetitive or stereotypic behavior, is needed. An EEG is essential for a correct diagnosis, especially with a negative or an unexplainable MRI study. The brain SPECT can be helpful and the peculiar MRI pattern in the mesial temporal lobe and the ipsilateral thalamus on diffusion-weighted imaging may be a useful clue to a diagnosis of complex partial status epilepticus.

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