



## Case Report

# Slow pseudoperiodic lateralized epileptiform discharges in nonconvulsive status epilepticus in a patient with cerebral palsy and a large central meningioma



Y.Z. Imam <sup>a,\*</sup>, D. Deleu <sup>a,b</sup>, B. Mesraoua <sup>a</sup>, A. D'souza <sup>a</sup>, H. Al Hail <sup>a</sup>, P.W. Kaplan <sup>c</sup>

<sup>a</sup> Department of Neurology (Medicine), Hamad Medical Corporation, Doha, Qatar

<sup>b</sup> Weill Cornell Medical College – Qatar, Doha, Qatar

<sup>c</sup> Department of Neurology, Johns Hopkins University School of Medicine, Baltimore, MD, USA

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## ABSTRACT

The presence of cerebral palsy and that of slow growing brain tumors are risk factors for convulsive and nonconvulsive status epilepticus. Nonconvulsive status epilepticus (NCSE) needs electroencephalographic (EEG) monitoring to be confirmed as it may be clinically subtle. Furthermore, it may present with a variety of ictal EEG morphologies. We report a case of a patient with cerebral palsy and a large central meningioma. Electroencephalogram showed a slow pattern of periodic lateralized epileptiform discharges (PLEDs) (a pattern considered as being situated in the ictal–interictal continuum) on an alpha background. The patient was treated for NCSE successfully with benzodiazepines followed by up-titration of his antiepileptic drug doses.

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## 1. Introduction

The term nonconvulsive status epilepticus (NCSE) is defined as “a range of conditions in which electrographic seizure activity is prolonged and results in nonconvulsive clinical symptoms” [1,2]. The clinical features of NCSE may be subtle and diverse, and confirmation relies on the electroencephalogram (EEG). Several approaches to establishing criteria for NCSE in adults have been published (Table 1) [1,3,4], and some clinical states may lie along an ictal–interictal continuum. Periodic discharges are often regarded on the interictal end of the spectrum, but when accompanied by clinical features including impaired vigilance and when signs resolve proximate to treatment, then this lends evidence to the ictal nature of periodic discharges along this spectrum. Periodic lateralized epileptiform discharges (PLEDs), in particular, have sparked controversy with regard to being part of the ictal vs. interictal spectrum. Periodic lateralized epileptiform discharges can occur in the temporal vicinity of an overt seizure, thus reflecting an ictal–interictal continuum phenomenon that can continue for weeks (chronic PLEDs) in conscious patients with retained alpha or basic rhythm [5]. However, when they occur in a patient with altered level of consciousness, some authors regard it as ictal [6–14]. Since changes in mental status are common in epileptic encephalopathies and developmental delay such as in

patients with cerebral palsy, it can be difficult or impossible to determine whether they are in status epilepticus [15], unless there is a discernible clinical improvement with treatment. We report the case of a patient with chronic epilepsy and cerebral palsy (CP) who presented with deterioration from an already impaired baseline. Although the EEG revealed slow PLEDs which are not typical of NCSE, the patient showed a good clinical and EEG response to benzodiazepines with PLED regression.

## 2. Case study

This 49-year-old man with cerebral palsy, mental retardation, and epilepsy residing in a long-term care facility for the prior 13 years had been awake, noncommunicative, but responsive to auditory and visual stimuli.

He was quadriparetic with spasticity and flexion contractures in the upper and lower limbs. He was bedridden and entirely dependent on caregivers for activities of daily living.

He was treated with phenytoin and levetiracetam and had yearly breakthrough seizures over the prior 10 years.

He had recently developed fever and dyspnea with a drop in oxygen saturation and was suspected of having aspiration pneumonia complicated by sepsis. He was given intravenous antibiotics, fluids, and supplemental oxygen and was transferred to our facility.

Following a mild generalized tonic–clonic seizure, his examination remained persistently unchanged but he was now unresponsive to

\* Corresponding author at: Neurology Section, Department of Medicine, Hamad Medical Corporation, P.O. Box 3050, Doha, Qatar. Tel.: +974 55246887.

E-mail address: [yahyazakaria200x@yahoo.com](mailto:yahyazakaria200x@yahoo.com) (Y.Z. Imam).

**Table 1**

EEG criteria for nonconvulsive status epilepticus (NCSE).

Adapted from Raoul Sutter and Peter W. Kaplan. [3]

- 1) Frequent or continuous focal electrographic seizures, with ictal patterns that wax and wane with change in amplitude, frequency, and/or spatial distribution.
- 2) Frequent or continuous generalized spike-wave discharges in patients without a previous history of epileptic encephalopathy or epilepsy syndrome.
- 3) Frequent or continuous generalized spike-wave discharges, which show significant changes in intensity or frequency (usually a faster frequency) when compared with baseline EEG, in patients with an epileptic encephalopathy or epilepsy syndrome.
- 4) PLEDs or BIPEDs that occur in patients in coma in the aftermath of a generalized tonic-clonic status epilepticus (subtle status epilepticus).
- 5) EEG patterns that are less easy to interpret including: frequent or continuous EEG abnormalities (spikes, sharp waves, rhythmic slow activity, PLEDs, BIPEDs, GPEDs, triphasic waves) in patients whose EEGs show no previous similar abnormalities in the context of acute cerebral damage (e.g., anoxic brain damage, infection, trauma).
- 6) Frequent or continuous generalized EEG abnormalities in patients with epileptic encephalopathies in whom similar interictal EEG patterns are seen, but in whom clinical symptoms are suggestive of NCSE.

auditory, visual, and noxious stimuli, with a fluctuating level of alertness. Plasma phenytoin level was low at 34.8  $\mu\text{mol/L}$  (normal: 40–80  $\mu\text{mol/L}$ ). Chest X-ray revealed bilateral lower lobe infiltrates. Nonconvulsive status epilepticus was suspected, and EEG (Fig. 1) showed intrusions of excess slow left-sided periodic lateralized epileptiform discharges (PLEDs) on a background of normal alpha pattern. No limb jerking or eye deviation/movements were noted.

Administration of 1 mg of IV lorazepam resulted in resolution of the “slow” PLEDs (Fig. 2) and improvement in the patient’s neurological

status, which now became responsive to auditory, noxious, and visual stimuli that occurred in a matter of 15 min after administration of lorazepam. By the next morning, his condition had greatly improved and he was near his baseline status. He received additional intravenous loading doses of phenytoin and levetiracetam on the same day, and his maintenance doses were adjusted.

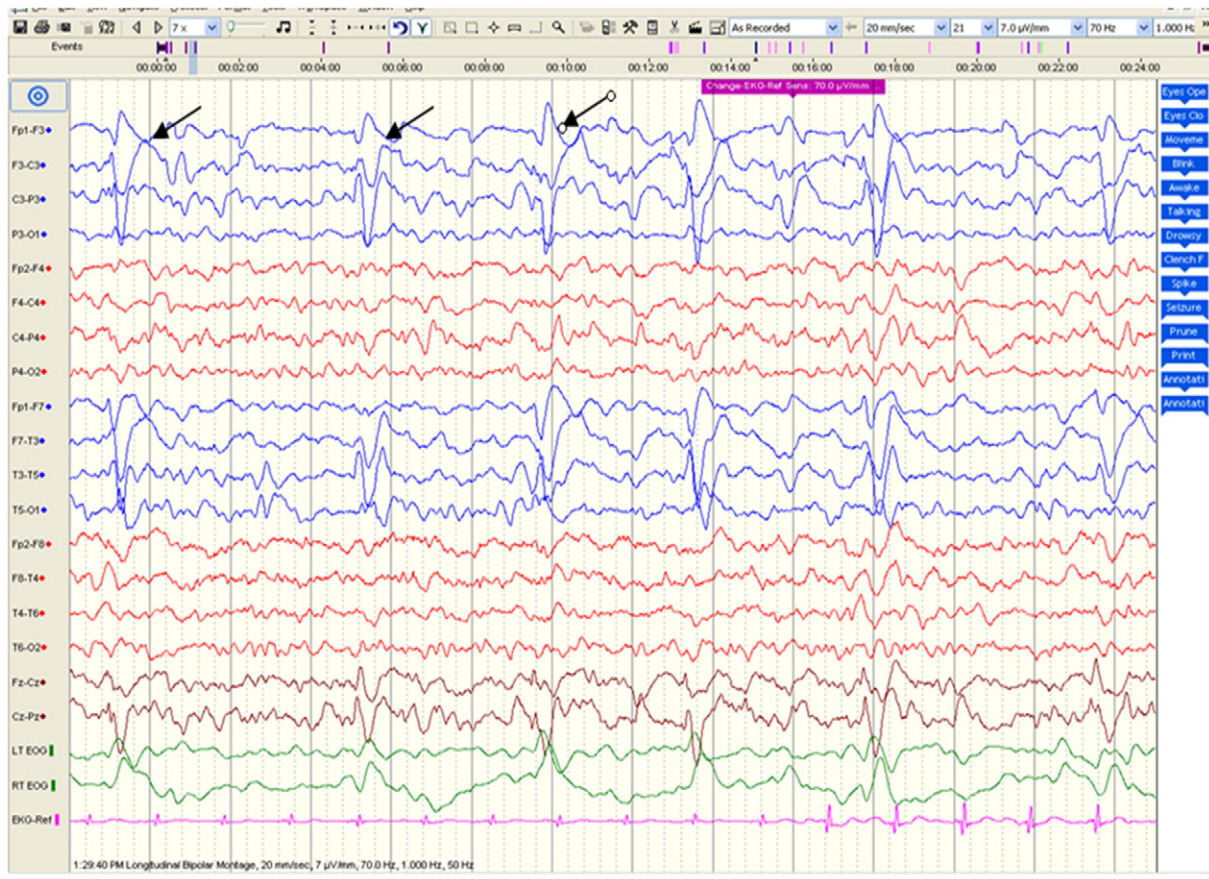
Brain CT showed a large central meningioma as well as a small posterior fossa meningioma (Fig. 3). Review of his old brain CT films (done seven years ago) showed no such lesions.

Because of his poor quality of life and the risks involved, neurosurgery was not considered an option, and the patient was managed medically with antiepileptic medication. Seizure and PLED regression were achieved both clinically and electroencephalographically (Fig. 4), and the patient was discharged to a long-term care facility for further care.

### 3. Discussion

This patient with chronic epilepsy and cerebral palsy (CP) showed deterioration from a baseline that was already impaired, a finding described in NCSE in those with developmental disability [1,15]. The EEG findings were not typical of NCSE in that slow PLEDs lie in the gray zone along the ictal–interictal continuum.

Cerebral palsy is a complex, heterogeneous lifelong condition with significant disability and long-term challenges that persist into adult life. Epilepsy is reported in 35% to over 60% of patients with CP [16,17] and is more frequent in patients with more severe disability [16]. Sixteen percent of children with CP and epilepsy may develop nonconvulsive and/or convulsive status epilepticus [18]. Brain tumors, particularly primary ones, are associated more frequently with both convulsive and nonconvulsive status epilepticus [19,20].



**Fig. 1.** Left-sided PLEDs (arrows).



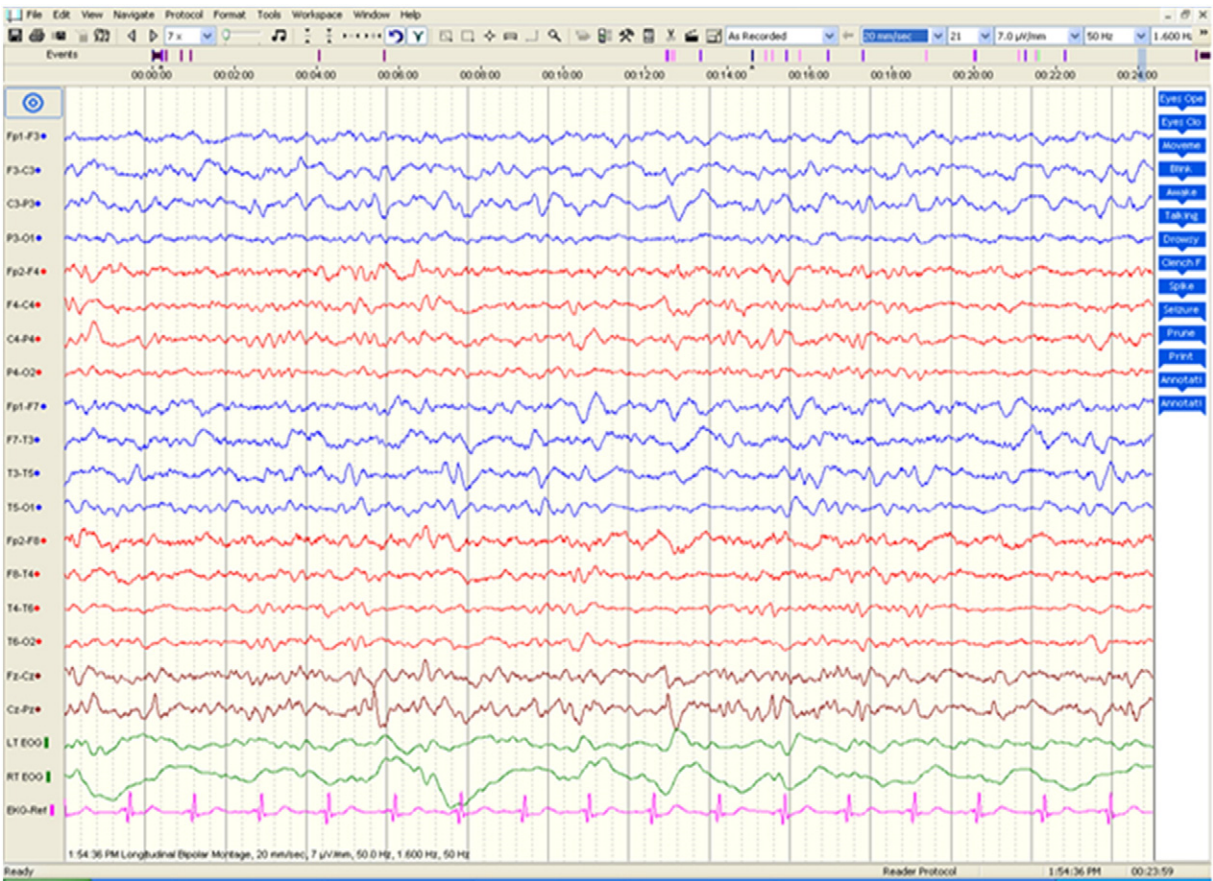


Fig. 2. After administration of 1-mg IV lorazepam, the PLEDs have regressed.

Electroencephalographic changes in mass lesions reflect irritability around the penumbra of the lesion, in this case, on the left. One can speculate that the focal structural lesion in the setting of chronic diffuse brain dysfunction, along with intercurrent infection and low AED levels,

represented the multiple risk factors seen in the recurrence of seizures and appearance of NCSE [4,21].

Periodic lateralized epileptiform discharges (Fig. 1) consist of periodic spike-and-slow wave or sharp-and-slow wave complexes, typically

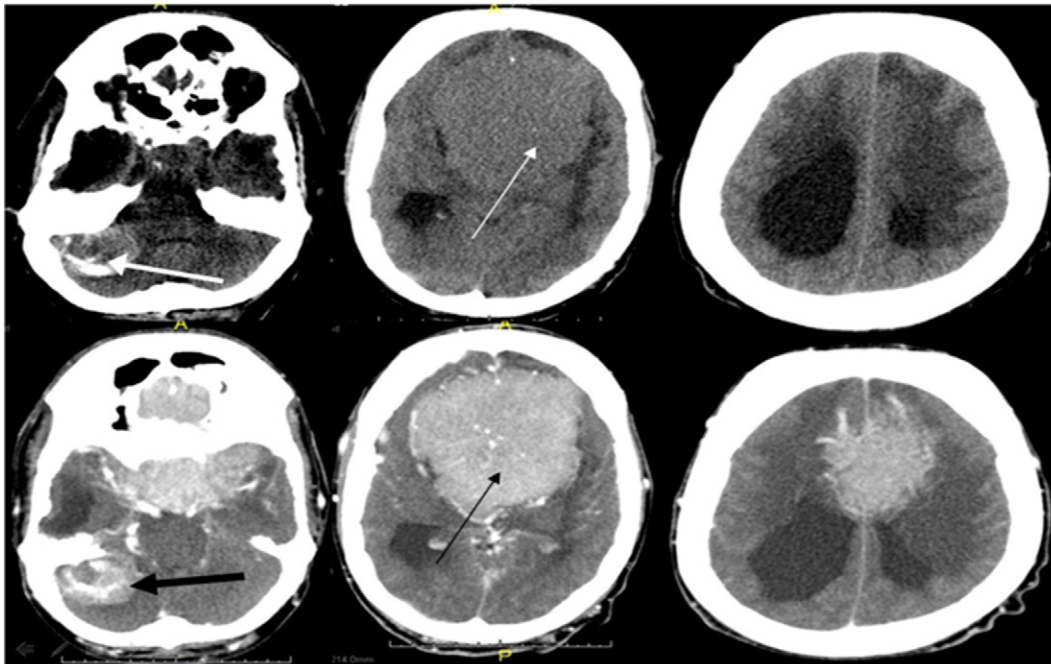


Fig. 3. Plain CT of the brain (top 3 images) and after intravenous contrast (bottom 3 images) showing a large central meningioma (thin arrow) with compression of the ventricular system and massive hydrocephalus. Also note another smaller posterior fossa meningioma (thick arrow).

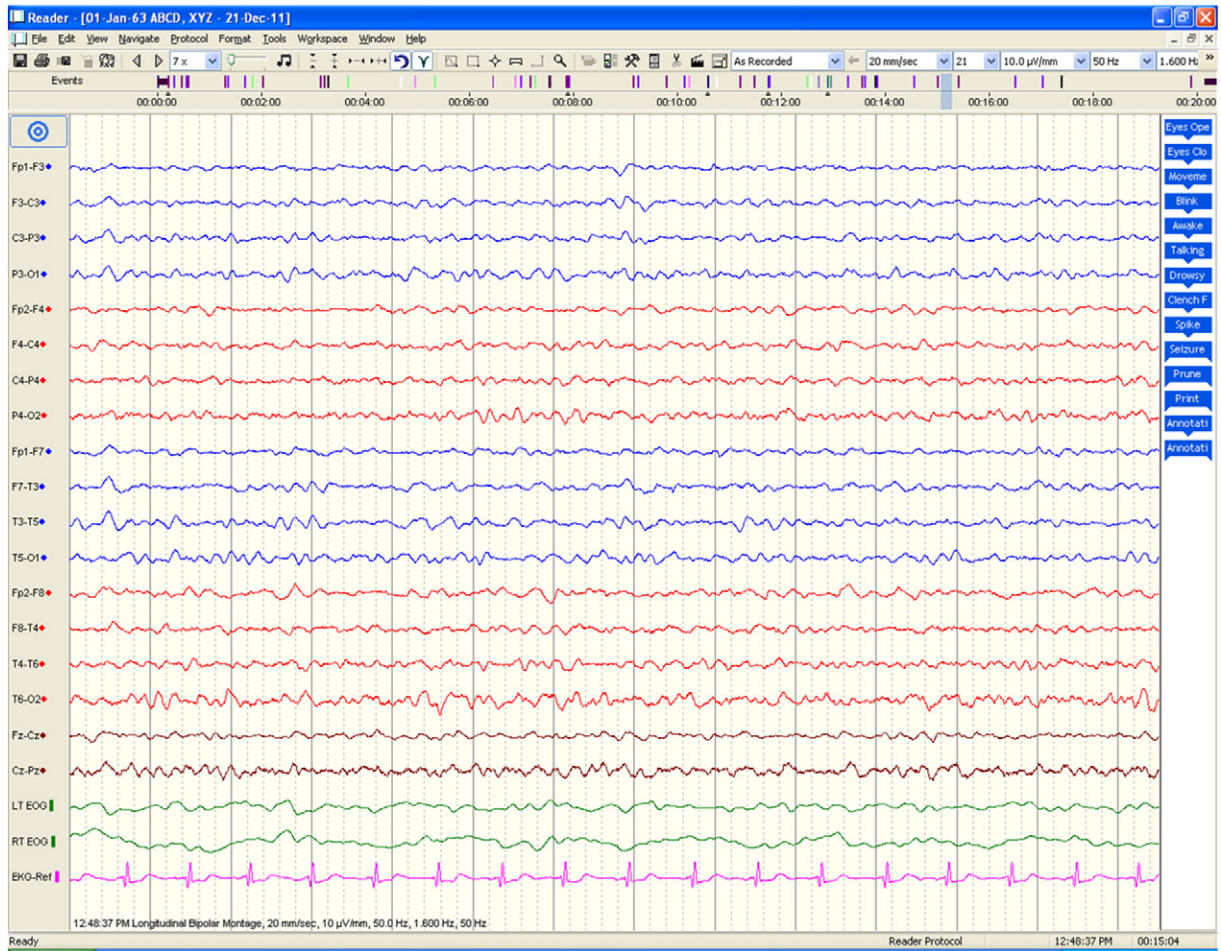


Fig. 4. EEG follow-up after 1 week which shows only diffuse slowing but no evidence of NCSE.

with a frequency of 1–2 Hz [22]. In our patient, the frequency was ~0.5 Hz. Periodic lateralized epileptiform discharges occur in a variety of conditions such as strokes, tumors, and infections; these together comprise around 50% of the underlying etiologies [20]. Slow growing extraaxial tumors such as meningiomas usually produce little EEG disturbance unless they compress and displace brain tissue or cause hydrocephalus such as in our case. Periodic lateralized epileptiform discharges are reported to support a diagnosis of NCSE if they meet certain conditions, in particular, good proximate clinical and EEG response to benzodiazepines with PLED regression [3] (Table 1). When EEG and clinical regression are not clear, SPECT or PET scanning can guide the clinician in the possible ictal (versus interictal) nature of the periodic discharges [4,23].

#### 4. Conclusion

There are diagnostic and management challenges in patients with long-term disability whose cognitive and neurological baselines are impaired and not always known to the clinical caregivers. Additionally, a diagnosis of NCSE may be difficult to make when this baseline is unknown and when the EEG findings represent a pattern that lies along the ictal–interictal continuum. Electroencephalogram can establish where along this continuum the patient resides, following which treatment that balances the risk–benefit needs of the patient, and the needs of chronic caregivers, can then be provided and monitored so as to offer the patient the optimal chance for regaining lost function with minimal risk.

#### Conflict of interest

None.

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