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Case Report

Sporadic nocturnal frontal lobe epilepsy: A consecutive series of 8 cases

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ABSTRACT

Objective: To present findings on a series of cases of sporadic nocturnal frontal lobe epilepsy (NFLE), a form of NFLE that is infrequently reported, in contrast to familial (autosomal dominant) NFLE. Both forms of NFLE need to be distinguished from parasomnias, nocturnal temporal lobe epilepsy, and other nocturnal disorders.

Methods: Eight consecutive cases of sporadic NFLE were evaluated at a sleep clinic in Taiwan. All patients had clinical evaluations, daytime waking and sleeping EEGs, brain MRIs, and overnight video-polysomnography (vPSG) with seizure montage.

Results: Gender was equal (four males, four females); mean age was 18.4 yrs (range, 7–41 yrs). Age of NFLE onset was by puberty. Premorbid history was negative for any neurologic, medical or psychiatric disorder. NFLE subtypes: nocturnal paroxysmal dystonia, $n=6$; paroxysmal arousals, $n=2$. MRI brain scan abnormalities with clinical correlates were found in one patient. Daytime awake EEGs were negative for ictal/interictal activity in all patients, but two patients had daytime sleep EEGs with interictal epileptiform EEG activity. During vPSG studies, three of eight patients with NFLE seizure events had concurrent epileptiform EEG activity, and two patients had interictal epileptiform EEG activity during their vPSG studies. No case had a spontaneous remission. Anticonvulsant therapy was highly effective in all eight cases (>75% reduction in seizure frequency).

Discussion: These cases confirm that sporadic NFLE closely resembles familial NFLE, and comprises a set of distinct clinical manifestations, with variable intensity, and variable scalp EEG epileptiform abnormalities across sleep and wakefulness, which have previously been identified in Caucasian patients from Europe and North America.

1. Introduction

In 1881, Gowers documented that 21% of epilepsy patients had seizures exclusively during sleep [1]. Subsequent studies of subjects with either partial or generalized seizures have estimated that the relative incidence of seizures during sleep is from 7.5% to 30% [2].

Some types of sleep-related epilepsy present with bizarre behaviors or vocalization, but without convulsion-like movement, tongue biting or upward gaze. Extreme restlessness,

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excessive swallowing movements, nightmares, and sleepwalking may represent various phenomena of seizures that emerge from sleep [3]. Since nightmares and sleepwalking can be components of the nocturnal symptom complex, parasomnias can be incorrectly diagnosed as the basis for these sleep related behaviors and disturbed dreams, and then subsequently incorrectly treated. This scenario poses a major challenge for sleep clinicians. However, epilepsy with bizarre behaviors is usually caused by frontal lobe epilepsy (FLE) or temporal lobe epilepsy (TLE). FLE and TLE can be contrasted [4]. FLE presents with bizarre behaviors, frequent short attacks, rapid recovery, and substantial preservation of consciousness. Repetitive and stereotypical behaviors characterize the clinical presentation without change in scalp EEG; the aura and the postictal periods can be masked by sleep. This scenario further increases the risk of incorrectly diagnosing a parasomnia as the basis of these abnormal nocturnal behaviors.

On the basis of the different intensity, duration and features of the motor patterns, Provini et al. [5] classified the nocturnal FLE (NFLE) epileptic seizures into three groups, according to Montagna [6]: (1) Paroxysmal Arousals (PA) with brief (<20 sec) episodes in which patients suddenly open their eyes, raise their heads or sit up in bed with a bizarre posture of the limbs, staring around with a frightened or surprised expression, and sometimes screaming; they then return to sleep. (2) Nocturnal Paroxysmal Dystonia (NPD) with a longer duration (20 sec–2 min) and more complex behaviors characterized by wide-ranging, often violent, and sometimes ballistic movements, with dystonic posturing of the head, trunk and limbs, such as head rotation, torsion of the trunk and choreo-athetoid movements of the arms and legs, with vocalization. (3) Episodic Nocturnal Wandering (ENW) with duration of episodes lasting up to 1–3 min, for which the characteristic feature is stereotypic paroxysmal ambulation during sleep, often with agitation and accompanied by screaming and bizarre, dystonic movements.

Sporadic (i.e. non-familial) NFLE is rarely reported, in contrast to familial NFLE, and therefore is poorly understood. We now report eight consecutive cases of sporadic NFLE, which also comprise the first case series from Taiwan (or any other Asian country), and provide clinical and vPSG findings, particularly vis-à-vis findings reported in Caucasian populations. The differential diagnosis, particularly focused on nocturnal temporal lobe epilepsy [7,8] and the parasomnias, will be discussed. One case in this series reported herein has previously been reported (patient 4) [9].

2. Materials and methods

Eight consecutive patients with equal 4:4 sex ratio and mean age of 18.4 yrs (range, 7–41) presented to the sleep clinic of one author (S-B Y) from July 2006 to November 2011 on account of nocturnal paroxysmal episodes suggestive of NFLE. These eight patients completed a comprehensive questionnaire covering lifetime sleep-wake, medical and psychiatric history, and review of systems. The patients and, when applicable, their caregivers were interviewed. They also received a full neurological examination by a pediatric or adult neurologist. Routine daytime awake and sleep EEG recordings were also performed for these patients. Table 1 contains the clinical data, including results of brain MRIs.

An overnight, hospital-based, vPSG monitoring, utilizing standard recording and scoring methods [10], was then performed on these patients after discontinuation of antiepileptic drugs for at least one day, except patient 8 who had recurrent attacks several times daily in wakefulness and sleep beginning shortly on the day of medication discontinuation. The PSG monitoring included an electrooculogram (EOG), expanded EEG (seizure montage) with a 1 cm/sec recording speed, submental and bilateral anterior tibialis electromyograms (EMGs), nasal-oral airflow, chest and abdomen respiratory effort, electrocardiogram, and continuous time-synchronized audiovisual recording.

3. Results

Seven of eight patients manifested one or more nocturnal attacks during vPSG monitoring. Patient 8 (who was maintained on anticonvulsant medication) did not have an attack, but she had interictal epileptiform discharges (spike and waves) during the overnight vPSG study, and attacks were viewed with event video home recording provided by her family. These eight cases were classified as PA (two cases) and NPD (six cases). All eight cases had sporadic NFLE, without any positive family history.

The nocturnal paroxysmal episodes had been present for up to 18 yrs (mean 9.6 yrs) before the current reported evaluation; age at presentation ranged from 7 to 41 yrs (mean 18.4 yrs). Mean duration of the seizure history was 9.6 yrs (range, 2–18 yrs). Seizure frequency in all eight patients was several attacks nightly, during nearly every night. All eight patients were unaware of their nocturnal motor manifestations, and so medical consultation was sought by their families who had observed the recurrent episodes. All eight patients had undergone neuroradiological examination, viz. brain MRI. Abnormalities with clinical correlates were detected in one case, involving a right orbitofrontal lobe cortical dysplasia in patient 4.

Two or more seizures with a stereotypic motor pattern were recorded in six of eight patients, and in one patient a single episode was recorded; patient 8 had no seizure episode attack during vPSG recording, but with clear-cut interictal epileptic EEG activity during the vPSG recording. Three patients also had occasional seizures during daytime wakefulness, similar to their seizures during sleep. In these three cases, however, daytime seizures were sporadic with low frequency.

Six PA episodes were recorded from two patients, lasting a mean of 11.0 sec in patient 5 and 11.5 sec in patient 6. In the PA episodes, the first movement usually involved the upper limbs: the patients suddenly raised their arms while asleep,