Sleep-related Screaming as a Manifestation of Frontal Lobe Epilepsy in a 8 Year-Old Girl: Case Report with Diagnostic Polysomnographic and Neuroradiological Findings

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Abstract- We report the case of a 8-year-old girl with a 3-month history of sleep-related screaming during both the night and daytime naps. A terrified expression accompanied the episodes of screaming, often with dream mentation and clouded consciousness. Her parents witnessed up to 5-6 episodes every night, with each episode lasting for less than 1 minute. A polysomnographic study documented six episodes of paroxysmal screaming without limb movement, with all episodes emerging from stage 2 non-REM sleep, and episode duration usually lasting 30-60 seconds. Suppression of the generalized background EEG activity occurred for 5-10 seconds before the onset of epileptiform activities (a run of sharp waves) and the onset of screaming. Brain MRI revealed right orbitofrontal lobe cortical dysplasia. Therapy with oxcarbazepine, 300 mg at bedtime, fully suppressed the sleep-related events, with a prompt relapse whenever the medication was not taken. Daytime wakeful seizures eventually appeared, and the girl had a seizure attack characterized by generalized choreiform and dystonic movements lasting for less than one minute, without any postictal confusion. Her consciousness was fully preserved during the attack. This patient had typical and atypical features of frontal lobe epilepsy.

Key Words: Nocturnal screaming, Parasomnia, Frontal lobe epilepsy, NFLE (nocturnal frontal lobe epilepsy), Sleep-related/nocturnal seizures, Polysomnography, Oxcarbazepine

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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 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.

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Extreme restlessness, excessive swallowing movements, nightmares, and sleepwalking may represent one phenomenon of seizures that are masked by sleep[3]. Sleep-related epilepsy with bizarre behaviors is usually caused by frontal lobe epilepsy (FLE) or temporal lobe epilepsy (TLE). FLE and TLE[4] can be contrasted. FLE present bizarre behaviors, with frequent short attacks and a rapid recovery, and preserve some degree of consciousness. Repetitive, and stereotypical behaviors characterize the clinical presentation without change in scalp EEG; the aura and postictal periods can be masked by sleep. Frontal cortical dysplasia is found in some FLE patients[5].

We report a case of FLE with cortical dysplasia in the right orbitofrontal lobe cortical dysplasia, presenting with sleep-related screaming and a good response to oxcarbazepine, 300 mg at bedtime.

CASE REPORT

A 8-year-old girl presented with sleep-related screaming for 3 months since July 2007. Her parents witnessed up to 5-6 episodes every night sleep (sometimes more than 10 episodes), even during daytime naps, with each episode lasting for less than 1 minute. The manifestations during sleep were paroxysmal episodes of screaming with terrified expression and dream-like visual hallucinations containing people staring at or grimacing at the girl. The episode duration was approximately 30-60 seconds. No unusual or bizarre movements were noted by her parents. If the girl was awakened by her parents at the end of a screaming episode, she would have partial awareness of the event, including visual hallucinations. The patient could not remember the sleep-related events upon awakening in the morning.

She had been sent to a pediatric neurology clinic for evaluation and underwent a daytime awake and sleep EEG study. Repetitive spike and slow waves were noted over F4-C4 and F3-C3 during the sleep, period without screaming or any bizarre behavior movement. Sleep-related screaming subsided after oxcarbazepine therapy, 300 mg at bedtime.

The patient then presented to the sleep center for polysomnography (PSG). Prior to the onset of sleep-related screaming, she had no identified abnormal sleep history, from early childhood, positive medical or psychiatric history, nor any positive family history. No current history of restless legs syndrome, leg jerking during sleep, or snore was noted. There was no precipitating medical or psychosocial event preceding the emergence of the sleep-related screaming. Her usual sleep time was from 10:00 p.m. to 7:00 a.m., including holidays. No complaint of daytime sleepiness was reported by the patient or her parents.

A comprehensive questionnaire covering lifetime sleep-wake, medical and psychiatric history, and review of systems was completed. The girl and her parents were interviewed. Neurological examinations and psychiatric interviews were conducted. An overnight, hospital-based, PSG monitoring, utilizing standard recording and scoring methods[6], was then performed on this patient after discontinuation of oxcarbazepine for one month. The PSG monitoring included eye movement, expanded EEG (seizure montage) with a fast paper speed, submental and leg electromyograms, airflow, chest and abdomen respiratory effort, electrocardiogram, and continuous time-synchronized audiovisual recording.

During the overnight PSG study, the patient exhibited six episodes of screaming that each time arose in stage 2 Non-REM sleep, with each episode lasting 30-60 seconds. The patient did not awake after any of the six episodes, and had no subsequent recall of the events. The six episodes of screaming occurred during the following times, with the first episode appearing 3.5 minutes after sleep onset at 10:26:50 p.m.: 1st episode: 10:30:20 p.m. to 10:31:00 p.m.; 2nd episode: 11:07:20 p.m. to 11:08:11 p.m.; 3rd episode: 1:11:10 a.m. to 1:11:45 a.m.; 4th episode: 2:21:40 a.m. to 2:22:21 a.m. ; 5th episode: 3:37:25 a.m. to 3:38:12 a.m. ; 6th episode: 4:52:49 a.m. to 4:53:36 a.m. The mean interval between the six screaming episodes was 76.2 minutes, and the mean screaming behavior duration was 43.5 seconds. The patient had 4 REM sleep periods during the PSG study, and the appearance of REM sleep in relation to the seizures was as follows: first REM sleep period occurred between the second and third seizures; second REM sleep period occurred between the third and fourth seizures; third REM sleep period occurred between the

fourth and fifth seizures; fourth REM sleep period occurred after the sixth seizure.

Suppression of the generalized background EEG activity occurred for 5-10 seconds before the onset of epileptiform activities (a run of sharp waves) and the onset of screaming during the attack episode. Occasional interictal epileptiform discharge (burst sharp waves) was also noted. With the morning awakening, the patient had no memory for any screaming episodes in the previous night.

Figs. 1-2 illustrate one such episode from stage 2 sleep. Total sleep time was 8 hours, 8.5 minutes; time spent of screaming in stage 2 sleep was 4.3 minutes during a mixed sleep/wakefulness state that accounted for over 1.1% of the sleep architecture; total time in bed was 8 hours, 21.5 minutes. Sleep architecture contains stage 1: 0.2%, stage 2: 65.1%, stage 3/4: 23.6%, REM sleep: 8.3%, screaming behavior time: 1.1%, wake time: 1.0% and movement time: 0.1%. Therefore, REM sleep was reduced almost three-fold of the normal range, with increased stage 2%, and normal amount of delta (stage 3/4) sleep. There was no snoring, apneas/hypopneas, oxygen desaturations, periodic breathing, periodic limb movements, or excessive arousals during the PSG study, apart from the 6 episodes of screaming. Autonomic activation (tachycardia, tachypnea changes) was not prominent surrounding these episodes, in contrast to typical sleep terrors (a disorder of arousal in Non-REM sleep).

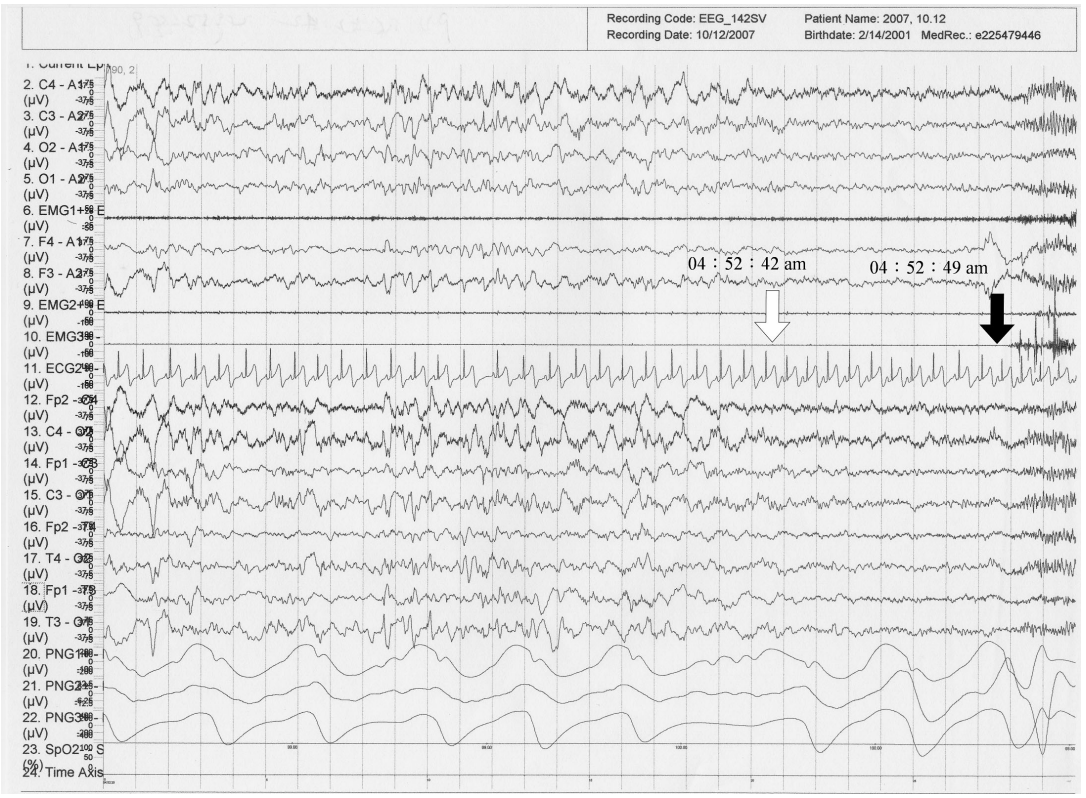

Figure 1. Nocturnal PSG (30s epoch, Ep 790) during stage 2 sleep and the emergence of a background EEG activity suppression before the screaming-behavior episode (PSG takes 30 seconds as an epoch, Ep790.2 is mean the 790th epoch and this epoch is in stage 2 sleep). Empty arrow indicates the onset time (04:52:42 a.m.) of the suppression of the background EEG activity (EEG montage, channels 12-19). Solid arrow indicates the onset time (04:52:49 a.m.) of the screaming-behavior episode beginning.The electrooculogram (channel 7-8) indicates no rapid eye movement. The electrocardiogram (channel 11) does not show an increase in heart rate during the motor activation with behavior release (i.e. screaming). Channels 20-23 represent the nasal/oral airflow, chest respiratory effort, abdomen respiratory effort and O2 saturation, which do not show any sleep apnea or oxygen desaturation.

Brain MRI study was also performed and the results revealed right orbitofrontal lobe cortical dysplasia (Fig. 3).

On the basis of her clinical history and the diagnostic PSG and neuroradiological studies, the diagnosis of FLE was confirmed. Oxcarbazepine, 300 mg bedtime therapy completely abolished the sleep-related events, with a prompt relapse whenever the patient failed to take the medication. The patient was followed at our sleep clinic every three months and was on oxcarbazepine regularly before the bedtime. At the latest follow-up visit in March 2009, daytime seizures were noted during clear-cut wakefulness. During the clinic visit, the girl had a seizure attack characterized by the onset of generalized

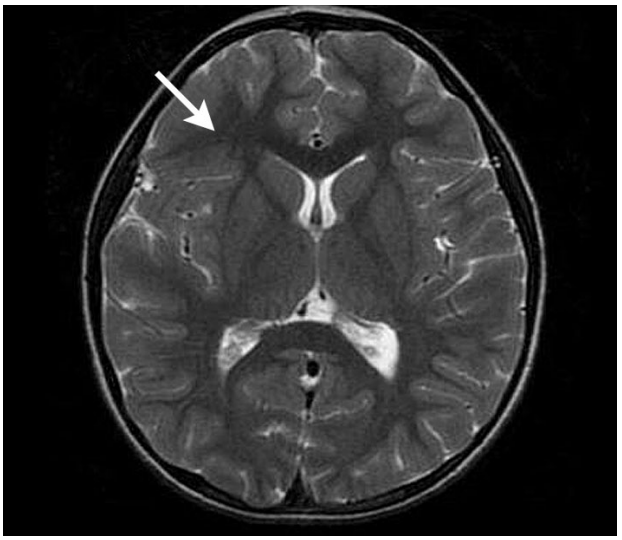

Figure 3. Brain T2W MRI demonstrating a right orbitofrontal lobe cortical dysplasia (indicated by the arrow).

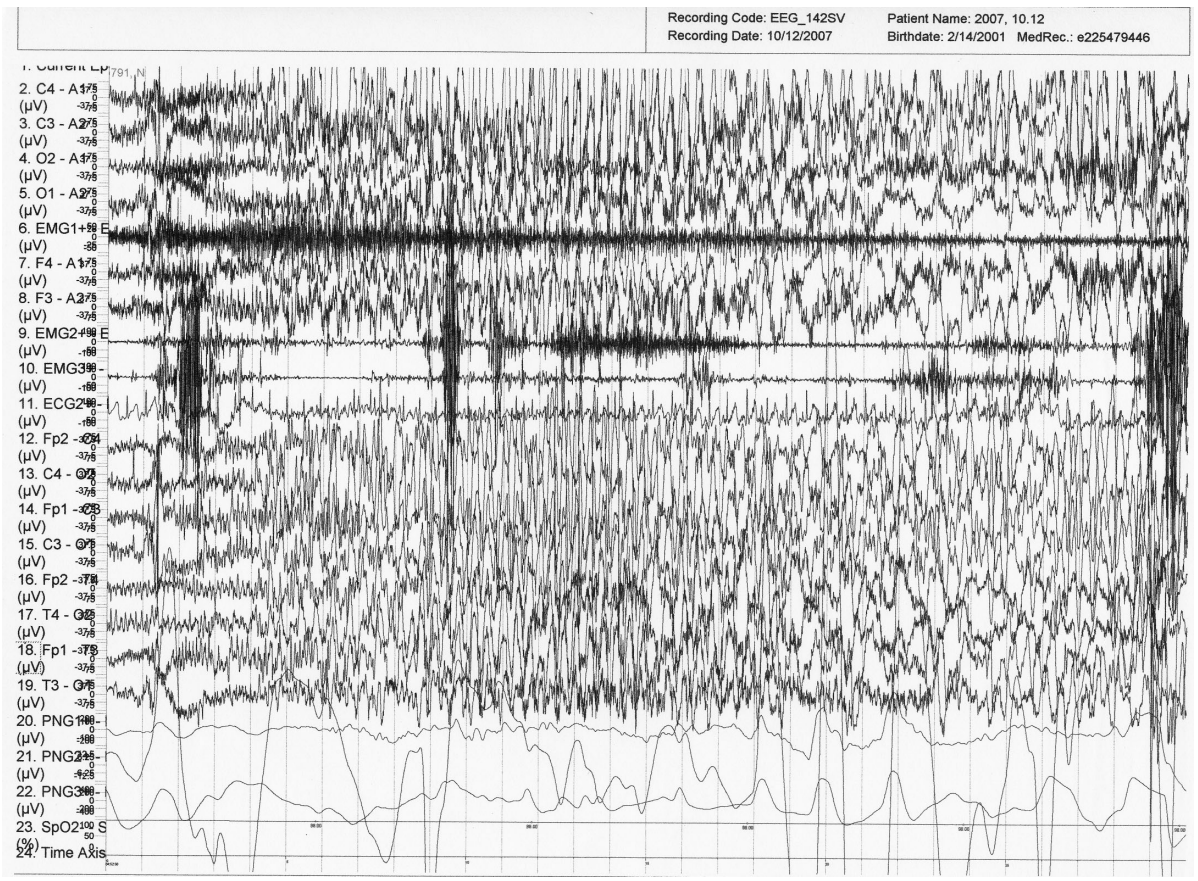

Figure 2. Nocturnal PSG (30s epoch, Ep 791) during the screaming-behavior episode with a run of sharp waves (EEG montage, channels 12-19). The electrocardiogram (channel 11) does not show an increase in heart rate during the motor activation with screaming. Channels 20-23 represent the nasal/oral airflow, chest respiratory effort, abdomen respiratory effort and O2 saturation, which do not show any sleep apnea or oxygen desaturation.

choreiform and dystonic movements lasting less than one minute and without any postictal confusion. Her consciousness seemed to be fully preserved during the attack. There was no screaming during the attack, nor any screaming reported by the parents during her wakeful seizures. Therefore, the diagnosis of FLE was further confirmed by the recent-onset of wakeful seizures.

DISCUSSION

For this 8 year-old girl, the nightly and multiple-times-nightly episodes of involuntary screaming emerged from sleep with subsequent partial or complete amnesia (depending on whether she was awakened by her parents after an episode). After neurologic and sleep evaluation, the case fulfilled diagnostic criteria for definitive and predominantly sleep-related epilepsy, both diurnally and nocturnally. Although sleep terrors, a non-REM sleep parasomnia, were strongly suggested by the history of sleep-related screaming episodes in a young girl, and also by the 6 screaming episodes during non-REM sleep in her PSG study, the EEG findings established the diagnosis of sleep-related epilepsy. In contrast to the disorders of arousal, including sleep terrors and sleepwalking, which predominantly emerge from slow-wave (delta) sleep[7], her sleep-related epilepsy (screaming with terrified expression) emerged exclusively in stage 2 Non-REM sleep. These very high frequency of attacks are extremely atypical for a classic disorder of arousal, such as sleep terrors. There were no identified triggers for sleep-related screaming, such as PLMs or sleep-disordered breathing. Also, the extraordinarily high frequency of her sleep-related screaming episodes is quite consistent with the diagnosis of FLE[7]. The clinical features of FLE include peculiar or bizarre behaviors that are often stereotypical; frequent, brief attacks with rapid recovery; and some preservation of consciousness. These features were eventually demonstrated by the emergence of the patient's seizures during clear-cut wakefulness. Therapy with oxcarbazepine was effective, which is also consistent with a seizure disorder.

Why certain types of seizures are facilitated preferentially by sleep remains uncertain, although several theories have been proposed. One theory concerns neuronal synchronization in which non-REM sleep activates a cortical area susceptible to epileptiform activity. Non-REM sleep is a physiological state of relative neuronal synchronization in which recruitment of a critical mass of neurons that is needed to initiate and sustain a seizure can occur[8]. Another theory concerns arousal mechanisms involving cortical hyperexcitability. Sudden synchronous excitatory input from neurons in the posterior hypothalamus (e.g., histaminergic), which project to the neocortical mantle, may facilitate seizures via exacerbation of cortical hyperexcitability[9]. A third theory addresses the anatomic substrate, i.e. the anatomical location. Anatomical location (e.g. frontal lobe) might play a critical role in this patient. In FLE, most seizures occur during sleep, while in temporal lobe epilepsy, most seizures occur during wakefulness[10]. However, a subset of patients with TLE may also have sleep-related seizures. Bernasonic et al.[4] identified a group of 26 patients with nonlesional refractory TLE in whom seizures occurred exclusively or prediminantly (> 90%) after they fell asleep or before they awakened. In addition, nocturnal TLE may have less frequent attacks, with longer durations and postictal confusion.

The bizarre behavior manifestations of FLE include uncontrollable running, yelling, vehement cursing, groaning, foot-stomping, kicking, repetitive pelvic thrusting (sexualized behavior), screaming with acute panic, and hallucinations (visual, auditory, olfactory)[11,12]. The FLE patients may retain some degree of consciousness, and the ability to follow simple commands. Nocturnal FLE has three identified subtypes according to episode duration: paroxysmal arousals (2 to 20 seconds), nocturnal paroxysmal dystonia (25 to 100 seconds), and episodic nocturnal wandering (lasting up to 3 minutes)[11,13,14]. Scheffer et al.[15] described 47 individuals with nocturnal FLE from five autosomal dominant families, and then identified these patients with autosomal dominant nocturnal FLE.

The diagnosis of sleep-related seizures should be considered in patients who have one or more of the followings: abnormal, stereotyped behavioral and/or experiential events, high-frequency events (e.g. nightly, quasi-nightly; multiple-times nightly), episodes that occur any time of night (NREM>REM sleep), similar events dur-

ing daytime (including naps), especially during tapering of medication; a positive response to a trial of antiepileptic drugs, and history of epilepsy, even if well-controlled.

Some sleep disorders are easily misdiagnosed as epilepsy, such as narcolepsy with cataplexy, rhythmic movement disorder, periodic limb movement disorder, REM sleep behavior disorder, arousal disorders, and sleep related dissociative disorders[16,17]. Even a newly identified parasomnia, "hypnagogic behavior disorder" that features complex behaviors during wake-sleep transitions in young children, can be mistaken for sleep-related epilepsy[18]. On the other hand, some forms of epilepsy can be misdiagnosed as sleep disorders, such as FLE and TLE.

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