Case Reports in Neurology

Case Rep Neurol 2020;12:18–23

DOI: 10.1159/000505230
Published online: January 15, 2020

Single Case – General Neurology

A Case of Complex and Abnormal Behaviors at Night: The Role of the Epilepsy Monitoring Unit in Diagnosis

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Keywords
Parasomnia · Epilepsy · Epilepsy monitoring unit · Diagnosis

Abstract
Complex nocturnal behaviors associated with sleep have many potential causes, including parasomnias and epilepsy. Although the type of event and description can frequently lead to a diagnosis, sometimes it is challenging clinically to determine the cause of the behaviors, requiring a more in-depth investigation. We report the case of a 29-year-old woman with a long history of complex abnormal behaviors and visual hallucinations at night. An extensive clinical evaluation failed to reveal a definitive cause of these episodes, prompting a 3-day epilepsy monitoring unit admission. During the stay, several events were captured on video electroencephalography, leading to a conclusive final diagnosis. This case highlights the challenging task of finding a definitive diagnosis in cases of complex nocturnal behaviors and the potential role of an admission to an epilepsy monitoring unit to help diagnose the cause of these behaviors.

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Introduction

Complex nocturnal behaviors associated with sleep have many potential causes. Parasomnias are undesirable physical events or experiences that occur during sleep and sleep-wake transitions. They are characterized by complex movements, behaviors, emotions, perceptions, dreams, and autonomic nervous system activity, and are believed to originate from sleep-wake state dissociation or state instability [1]. Epilepsy is also commonly on the differential for complex nocturnal behaviors from sleep. Frontal lobe epilepsy, temporal lobe epilepsy, benign focal epilepsy of childhood, and sleep-related hypermotor epilepsy tend to manifest with seizures occurring predominantly during sleep. Seizures occurring predominantly during sleep represent up to 12% of people with epilepsy [2].

The pattern of behavior can frequently lead to a diagnosis, but sometimes more detailed investigations are required to reach a final diagnosis. We report the case of a 29-year-old woman with a long history of complex visual hallucinations and abnormal behaviors at night. An extensive clinical evaluation failed to distinguish between parasomnias and epilepsy, prompting a 3-day epilepsy monitoring unit (EMU) admission. During the stay, several events were captured on electroencephalography (EEG) and video leading to a conclusive final diagnosis. This case highlights the challenging task of finding a definitive diagnosis for complex nocturnal events associated with sleep.

Case Report

A 29-year-old woman was referred for evaluation of hallucinations and abnormal behaviors during sleep. The patient reported a long history of somniloquy (sleep-talking), bruxism, somnambulism (sleep-walking) as a child, and vivid dreams. At the age of 22, she started having episodes of visual hallucinations 1–2 h after falling asleep, between 12 and 2 a.m. She would awaken and see images that were somewhat threatening but not stereotyped; the images tended to follow certain themes such as her husband's face being visually altered, a stranger being in her bed, or seeing chairs around the bed. The images were complex and well formed, tended to be in black/white and could appear anywhere in the visual field.

Generally, during these episodes the patient would scream, appear frightened with her eyes open, and sometimes verbalize "Oh my God, oh my God, don't you see them?" Motor manifestations such as chewing, grinding teeth, lip smacking, or repetitive squeezing of her arm or leg with her right hand were occasionally noted. Rarely, she would walk out of the bedroom. She was confused upon awakening from these episodes. Her husband would try to convince her that the hallucinations were not real, after which she would fall back asleep. The events lasted less than 1 min, occurred 3–4 times per week, and typically occurred once per night during the first half of the night. The patient was amnestic at times of the events.

The spells sometimes appeared triggered by her husband getting into bed and possibly causing her to arouse. Other triggers included sleep deprivation and stress. There were no daytime spells, incontinence, tongue biting, episodes of loss of time, or other spells suspicious of unwitnessed seizure activity. She had no epilepsy risk factors. She had no other past medical history and was on no medications. Physical examination was unremarkable and body mass index was 22.9 kg/m^2.

The complex nocturnal visual hallucinations were thought to most likely reflect hypnopompic (sleep to wake transition) and hypnagogic (wake to sleep transition) hallucinations associated with several non-rapid eye movement sleep (NREM) parasomnias, such as

confusional arousals, night terrors, and somnambulism. Less likely, they were thought to represent epileptic seizures or peduncular hallucinosis from a brainstem or thalamus lesion. Brain magnetic resonance imaging with and without contrast was performed and was normal. The patient was subsequently admitted to the EMU to assist in the classification of events and rule out epilepsy.

During the 3-day EMU admission, two typical events were captured out of sleep, the first one characterized by awakening from sleep with confusion associated with an auditory hallucination and the second one awakening from sleep with confusion trying to speak with her husband. The events did not appear stereotyped in appearance and there was amnesia. Both events arose from stage N3 sleep and there was no associated epileptiform activity on EEG, (Fig. 1, 2). Given the lack of stereotypy and lack of epileptiform activity on EEG, the events were concluded to represent NREM parasomnias, confusional arousals with associated hypnopompic hallucinations, as opposed to seizure.

Discussion

The case presented has unique features because of the presence of multiple parasomnias in the same patient and the complex nocturnal behavior resemblance to a nocturnal seizure disorder. The patient presented with symptoms of somniloquy, bruxism, somnambulism, confusional arousals, and night terrors, which are NREM parasomnias. She also experienced symptoms suggestive of hypnopompic visual and auditory hallucinations. The symptoms suggestive of a nocturnal seizure disorder were stereotyped automatic movements, hypermotor activity, and amnesia for the episode. Ultimately, based on the findings of confusion, hallucinations and partial amnesia arising from NREM sleep (stage N3) during the EMU admission, a diagnosis of confusional arousals with prominent hypnopompic hallucinations was made.

Parasomnias are undesirable physical events or experiences that occur during entry into sleep, within sleep or during arousal from sleep. They can occur during NREM, REM and sleep-wake transitions. They are characterized by complex sleep-related movements, behaviors, emotions, perceptions, dreams, and autonomic nervous system activity. The sleep and wake states are believed to occur on a spectrum rather than a completely dichotomous state; parasomnias are believed to be originating from a state dissociation or state instability [1].

Confusional arousals are characterized by mental confusion or confusional behaviors without terror, autonomic activation or ambulation outside of the bed [1]. NREM parasomnias such as somnambulism, sleep terrors and confusional arousals are usually considered as distinct diagnoses, but more likely exist on a continuum and represent a defective arousal mechanism [3], and may co-exist in the same patient, as was the case with our patient. Overlap parasomnia disorder includes one or more manifestations of an REM parasomnia seen in combination with a NREM parasomnia or rhythmic movement disorder. This condition is male predominant. Most cases begin during childhood or adolescence. It can be idiopathic or secondary to an underlying structural lesion or neurodegenerative disorder. A study by Schenck et al. [4] revealed that patients with idiopathic parasomnia overlap disorder have a significantly lower age of onset, compared to secondary parasomnia overlap disorder.

The most striking aspect in this case was the report of terrifying hypnopompic auditory and visual hallucinations associated with confusional arousals arising from NREM. An extensive review of the literature provides little insight into this phenomenon. One similar report comes from Mantoan et al. [5], who reported adult-onset NREM parasomnia with hypnopompic hallucinatory pain. Similar to our case, the episodes arose within 1–2 h of sleep onset and

were associated with looking terrified and initial lack of responsiveness. Polysomnography (PSG) showed an increased arousal index, but no evidence of a primary sleep disorder.

Hypnopompic hallucinations are less common than hypnogogic hallucinations and are reported in 7–13% of the population; both are more common in women than men [6]. These typically occur in the setting of sleep deprivation, alcohol ingestion, REM-suppressing medications and/or a change in sleep schedule, and may be repetitive in nature but not stereotyped [7]. Hypnopompic hallucinations are not typically associated with intermittent amnesia and typically resolve with turning the light on [7]. Thirty percent of these hallucinations are reported as frightening [6]. Nielsen and Zadra [8] described terrifying hypnagogic hallucinations as frightening dreams similar to those occurring in REM sleep. However, in this description, the phenomenon is described as arising from sleep-onset REM sleep instead of regular REM periods and to have an immediate recollection of the disturbing content, neither of which appears to fit our case.

Complex gestural automatisms, autonomic activation and mood changes can be seen in frontal lobe seizures. Seizures involving orbitofrontal region can be associated with gestural automatisms, olfactory hallucinations and illusions as well as autonomic signs. Opercular seizures can be seen associated with mastication, salivation, swallowing, gustatory hallucinations, fear and autonomic phenomena. Patients with frontal lobe epilepsy may not experience postictal confusion and the duration of the seizures can be short, compared to seizures arising out of other parts of the brain [9]. The ictal and interictal EEG could be completely normal in this condition, which makes the diagnosis even more challenging [10].

Autosomal dominant nocturnal frontal lobe epilepsy was initially described by Lugaresi and Cirignotta [11] in 1981, and is characterized by a genetic defect in the nicotinic acetylcholine receptor. This condition typically manifests in the second decade. The patients can experience multiple seizures per night (clustering), and the frequency tends to decrease during adulthood. Occasionally seizures can occur during wakefulness. Semiology comprises of hyperkinetic activity, bimanual and bipedal automatisms, vocalizations such as screaming, laughing, autonomic activation, and epileptic nocturnal wandering reminiscent of sleep walking. Awareness may or may not be preserved.

As there was concern for seizures, the decision was made to proceed directly with video EEG (vEEG) monitoring instead of a PSG. It can be difficult to differentiate parasomnias from nocturnal seizures, and vEEG is the gold standard for making a diagnosis of epilepsy. The benefits compared to PSG include an expanded EEG montage allowing for increased sensitivity for detecting ictal and interictal activity, and prolonged monitoring as the study is run 24 h a day – often for multiple days [12]. As continuous pulse oximetry, electrooculogram and electromyogram can be easily added to the montage, the only major limitation of vEEG compared to PSG is a lack of respiratory monitoring of apneas and hypopneas. Since the patient was very low risk for obstructive sleep apnea (normal weight, without excessive daytime sleepiness, snoring or witnessed apneas, and a STOP BANG [13] score of 0/8) and had no other symptoms suggesting other organic sleep disorders, a PSG was not performed at the time.

In conclusion, in this case we confirmed the presence of NREM parasomnia – confusional arousals associated with prominent hypnopompic hallucinations – arising from stage N3 sleep. vEEG allowed us to rule out epilepsy as a cause of the patient's complex behaviors, and to differentiate between REM and NREM parasomnias. The patient also had multiple other parasomnias based on history. This case highlights the utility of vEEG monitoring in diagnosing atypical parasomnias and in differentiating between epileptic events and parasomnias in patients with complex nocturnal behaviors, and the possibility of encountering multiple coexisting parasomnias in the same patient.

Statement of Ethics

There are no ethical conflicts to declare.

Disclosure Statement

The authors have no conflicts of interest to disclose.

Funding Sources

The authors received no financial support for the research, authorship, and publication of this article.

Author Contributions

The authors conceived and designed the manuscript, analyzed the data, wrote the manuscript, revised and gave approval to the final version, and are accountable for all aspects of the work.

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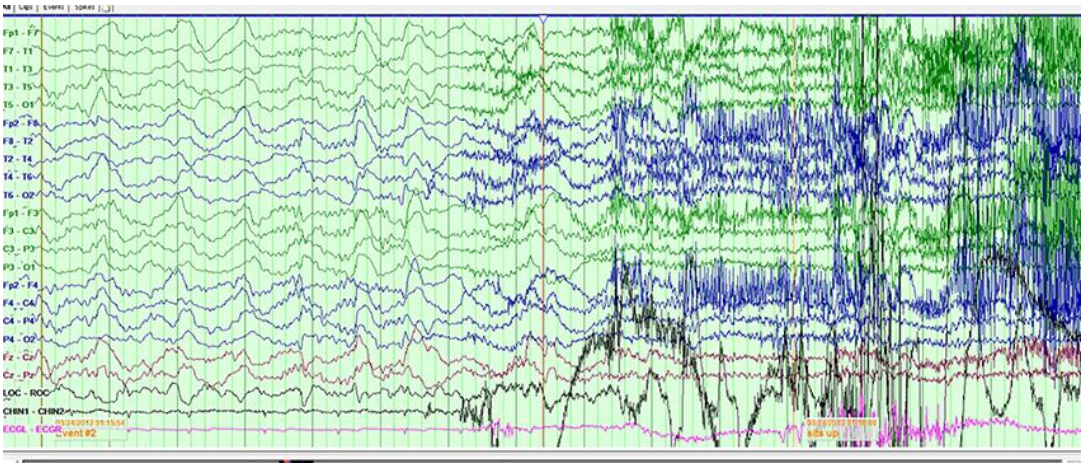

Fig. 1. Onset of event #1 from N3 (slow-wave) sleep. No evidence of epileptiform discharges before or during onset.

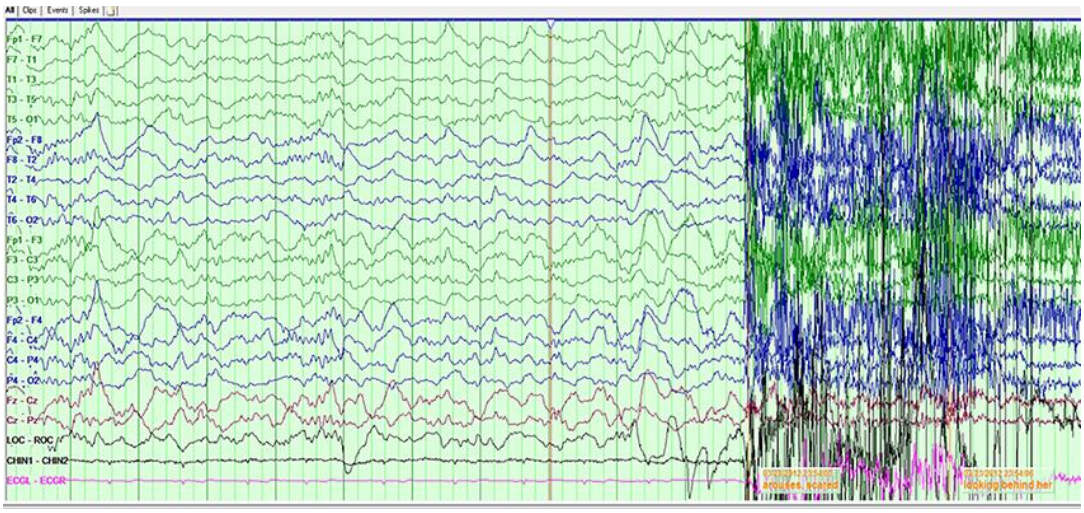

Fig. 2. Onset of event #2. Once again, event arising from N3 sleep and absent epileptiform discharges.