Case Report

Nocturnal Frontal Lobe Epilepsy Presenting as Excessive Daytime Sleepiness

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Abstract

Excessive daytime sleepiness (EDS) is common in the general population. Etiologies include insufficient sleep and primary sleep disorders. Due to its high prevalence, physicians often overlook EDS as a significant problem. However, EDS may also be the presenting symptom of seizures, in particular Nocturnal Frontal Lobe Epilepsy (NFLE). Due to the clinical similarity between the nocturnal behaviors of NFLE and parasomnias, and poor patient-related history, NFLE remains a challenging diagnosis. We report the case of a patient with NFLE who presented with a primary complaint of EDS, and discuss the differential diagnosis and evaluation of patients with EDS associated with nocturnal behaviors. In the context of a patient presenting with EDS and stereotyped nocturnal events, clinical suspicion should be high for NFLE.

Keywords: Epilepsy, excessive daytime sleepiness, hypersomnia, parasomnia, seizures

Introduction

Excessive daytime sleepiness (EDS) is reported by approximately 10% of the US population. The management of EDS depends on its etiology, which commonly includes insufficient sleep, circadian rhythm disorders, sleep apnea, and parasomnias.[1,2] A rarer cause of EDS is sleep-related epilepsy, as in individuals with Nocturnal Frontal Lobe Epilepsy (NFLE). Clinical suspicion for NFLE must remain high, as diagnosis is challenging due to its variable presentation, similarity to parasomnias and vague patient-reported history.

Herein, we discuss a patient who presented with complaints of EDS associated with stereotyped episodes during sleep, the combination of which led to the diagnosis of NFLE.

Case Report

A 23-year-old graduate student reported a six-year history of worsening EDS, with an elevated Epworth Sleepiness score of 13 (abnormal ≥ 11). This was associated with movements during sleep, in which his roommate observed him sitting up in bed, looking around, and returning to sleep within one minute. He also had up to seven episodes of bicycling movements with tonic posturing of the left arm and leg nightly, for which he was amnestic.

He did not have any previous medical history or use any medications. Social history was negative for tobacco or alcohol use. He had one maternal aunt with epilepsy.

Neurological exam, routine laboratory testing and MRI of the brain with and without gadolinium were normal.

A sleep diary obtained two weeks prior to polysomnography (PSG) excluded insufficient sleep time (mean sleep duration was 549 ± 74 minutes).

Attended overnight PSG showed normal REM sleep latency (68 minutes) and multiple spontaneous arousals (10.0/hrs) during non-REM sleep. No other primary sleep disorders were seen. Multiple sleep latency testing was consistent with objective hypersomnia, with a mean sleep latency of 4.8 minutes (abnormal < 8 minutes), without sleep-onset REM periods.

A full set of EEG channels was recorded during PSG. Stereotyped clinical seizures were captured, consisting of repetitive blinking, head turning and left arm elevation, and corresponded with frontally predominant epileptiform activity on EEG.

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Treatment was started with topiramate, which was titrated to 200 mg at night with improvement of sleepiness and elimination of nocturnal paroxysmal arousals and tonic posturing events.

Discussion

Daytime sleepiness is defined as "the inability to stay awake and alert during the major waking episodes of the day, resulting in unintended lapses into drowsiness or sleep."[3] EDS has diverse etiologies, including sleep deprivation, untreated primary sleep disorders, metabolic disturbances, medications, and psychiatric and neurological disorders, including seizures.[1]

EDS as the presenting symptom of NFLE was originally reported in 1986.[4] Subsequent series demonstrated EDS in up to 75% of patients.[5] Some patients, while complaining of EDS, were unaware of their nocturnal seizures; they came to medical attention at the request of others who witnessed their disturbed sleep.[6] Anecdotal cases of EDS improvement with medical management of NFLE has also been reported.[6]

NFLE seizures present a diagnostic challenge in the evaluation of EDS, and are at risk of being misdiagnosed for various sleep disorders, or even pseudoseizures. This is due to the diversity of NFLE manifestations, which can involve movements such as kicking, thrashing and wandering; and vocalizations, including screaming and roaring. The brief and frequently occurring forms of NFLE may be discounted as restless sleep.[7]

Non-REM and REM parasomnias should be considered in the differential diagnosis of NFLE. The non-REM parasomnias (confusional arousals, somnambulism and sleep terrors) tend to occur early in the night during slow-wave sleep. Somnambulism may range from semi-purposeful walking to more complex tasks such as driving; distressed cries and aggressive behavior may also occur.[7] In REM sleep behavior disorder (RBD), muscle tone is pathologically retained during REM sleep, allowing dreams to be "acted out". These manifestations can resemble NFLE; however, RBD usually occurs in the early morning and can be associated with vivid dream recall.

Other potential confounders include shared precipitating factors (stress, sleep deprivation, alcohol) between NFLE and NREM parasomnias; the increased prevalence of somnambulism and epilepsy in the families of patients with NFLE;[8] and poor history of nocturnal events as provided by the patient. In a large series of NFLE, 72% of patients were unaware of their nocturnal motor manifestations.[6]

Distinguishing NFLE from a parasomnia often requires referral to a specialist in epilepsy, sleep medicine or both. Diagnostic testing involves prolonged video-EEG and/or PSG to capture and characterize nocturnal events. As an abbreviated number of EEG channels are used during routine PSG, it is important that the PSG is ordered with a full set of EEG channels. Both clinical and EEG characteristics have been identified to distinguish between NFLE and parasomnias; therefore, obtaining a detailed history, which often requires supplemental information from family or friends, is as important as diagnostic testing.

Characteristics which may distinguish between NFLE and parasomnias include: Mean event duration longer for parasomnias (60 versus 37 seconds); event onset during non-REM sleep stage, with 100% of parasomnias presenting during slow-wave-sleep versus 13% of NFLE (which typically presents during non-REM stages 1-2); and the absence of clear seizure activity on EEG during parasomniac events versus 38% of NFLE seizures.[9] Inconclusive EEG patterns in NFLE seizures have been noted in other series, likely related to the insensitivity of scalp EEG for detecting frontal lobe seizures.[6] Clinical features which strongly favor a parasomnia include: Yawning, nose rubbing, rolling over, external triggers, waxing-waning pattern, physical/verbal interaction, and sobbing. Behaviors after NFLE seizures, such as staring, fumbling and partially interactive speech, are generally not useful differentiators, due to their similarity to the manifestations of parasomnias.[9]

Management of NFLE requires treatment with anti-epileptic drugs (AEDs) to control seizure activity. While topiramate has demonstrated efficacy in NFLE,[10] alternative AED choices would also have been valid. Deciding which AED to start is based upon several factors, including patient co-morbidities, AED side effect profiles, and reproductive considerations in women. In addition, control of seizures may require more than one AED. Given the complexity of AED regimens and the potential for seizures to evolve or change over time, long-term management of NFLE should be referred to an epilepsy specialist.

Conclusion

In the context of a patient presenting with EDS and stereotyped nocturnal events, clinical suspicion should be high for NFLE. Referral to a specialist in epilepsy and/or sleep medicine is recommended, and a thorough evaluation should be undertaken, including a detailed clinical history, and either PSG to exclude primary sleep disorders, video EEG monitoring to capture the stereotyped episode, or both.

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