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Case Report

Sleep- Related Hypermotor Epilepsy: A Rare Case

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Abstract

Sleep-related hypermotor epilepsy is a subgroup of sleep-related epilepsies and is very rare among focal epilepsies. It is a type of epilepsy that can be confused with the primary diseases of sleep and is diagnosed late. Here, we present a case diagnosed with electroencephalography (video-EEG) monitoring 13 years later.

Keywords: Epilepsy, sleep-related hypermotor epilepsy, video-EEG

INTRODUCTION

When sleep-related epilepsy is mentioned, epilepsies in which all or more than 90% of seizures occur during sleep come to mind. Sleep-related hypermotor epilepsies are a subgroup of sleep-related epilepsies.

Semiological features with evident hypermotor behaviors are very characteristic of seizures. Seizures in clusters for a short time are often seen during sleep at night. It may originate from the frontal lobe or, more rarely, from the extra frontal lobe.[1]

Sleep-related hypermotor epilepsies are quite rare among focal epilepsies, and their prevalence is 1.8/100 000.[2] It peaks in childhood and adolescence and often occurs during the Non- Rapid Eye Movements (NREM) sleep phase and causes poor sleep quality.

Sleep-associated hypermotor epilepsy is a late-diagnosed type of epilepsy and can be confused with primary sleep disorders. This article found it appropriate to present a case with recurrent sleep episodes for 13 years. She was followed up with the diagnosis of non-epileptic psychogenic seizures without treatment but was diagnosed correctly only with video-EEG monitoring.

CASE PRESENTATION

A 22-year-old female patient with a known diagnosis of epilepsy and using antiepileptic therapy presented to the clinic because of an increase in the frequency of seizures over the last 2 weeks. Involuntary movements started when she was nine, such as leaping in her hands and feet, often occur during falling asleep and waking up from sleep. A few years later, she had a generalized seizure accompanied by loss of consciousness, jaw locking, and convulsions throughout the body. Valproic acid was started with the diagnosis of epilepsy in the patient who had no pathology in EEG and magnetic resonance imaging. After 2-3 years of use, another physician said that she had psychogenic epilepsy and discontinued her treatment. During the drug-free period, the frequency of seizures was increased, and lamotrigine was started. She had seizures once a year or every two years for about 4-5 years under the lamotrigine treatment. However, recently, when she was stressed, her seizures started to be 1-2 times a week.

The patient was hospitalized in the video-EEG unit for further examination and treatment; sharp wave discharges were noted in the bilateral parieto-occipital regions and were more prominent on the right in the interictal EEG (Figure 1A and B). She had 22 seizures, with a frequency of 5-8 per night, during the 4 days in the video-EEG monitorization. During these seizures, it was noted that she woke up suddenly from light sleep. She sometimes covered her mouth with her right hand and sometimes with both hands. Motor movements as pedaling both feet were accompanied by fearful facial expressions. It was observed that after these seizures in which awareness was lost, the patient recovered quickly and fell asleep again in a short time. When the patient was questioned retrospectively, she stated that she was unaware of what had happened. At the beginning of these attacks, which lasted between 30 and 70 seconds, rapid generalized activity was observed in the EEG, and epileptic activity appeared in the temporal region of the right hemisphere in the later periods of the seizure, mixed with rhythmic theta activity (Figure 1C and D). When we questioned the patient retrospectively, her attacks, which her relatives described as jumping, were similar to those seen during hospitalization and repeated almost every night. With the diagnosis of sleep-related hypermotor epilepsy, we planned to increase the dose of lamotrigine and switch to carbamazepine if it did not benefit when it reached the adequate dose.

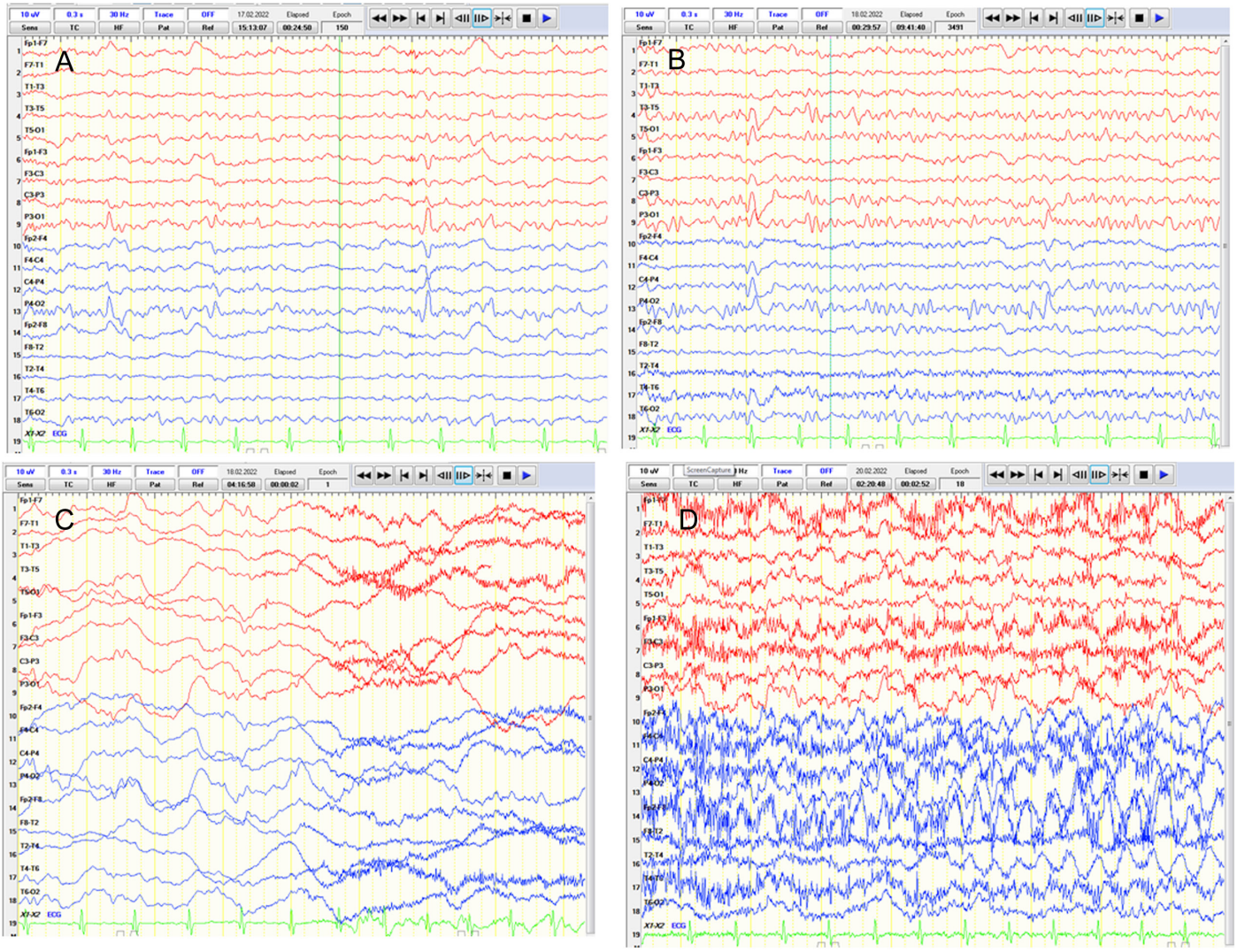

Figure 1. Sharp wave activity in the parieto-occipital region in the right hemisphere in the interictal EEG (A, B). Rhythmic delta activity in the right hemisphere in ictal EEG (C, D).

DISCUSSION

The diagnosis of a patient group with episodes of abnormal motor movements during night sleep was named hypnogenic paroxysmal dystonia in 1981 and nocturnal paroxysmal dystonia in 1986. In the 1990s, its epileptic origin was understood, and it was called "nocturnal frontal lobe epilepsy."[3-5] However, it was suggested to use the term "Sleep-related hypermotor epilepsy" in 2014 after it was observed that the seizures were not only caused by the frontal lobe but also could originate from the temporal, opercular-insular, and sometimes the parietal region. Besides, it was noticed that the episodes were not only during night sleep but can also occur at short naps during the day.[1]

MAIN POINTS

- Sleep-related hypermotor epilepsies are a subgroup of sleep-related epilepsies.
- It is necessary to combine EEG recordings with clinical findings.
- Video-EEG is a valuable diagnostic method in the differential diagnosis of sleep-related hypermotor epilepsy.

The diagnosis is made clinically according to the primary clinical features. The seizures must last for less than 2 minutes with abrupt onset and termination; the most common hypermotor events, stereotypic motor patterns, and seizures are frequently seen during sleep. Clustering of seizures is characteristic but not essential. Preservation of awareness during the episode, absence of ictal and interictal EEG disorders, the extra frontal origin of the seizures, and accompanying mental involvement do not exclude the diagnosis.[1]

The diagnosis of sleep-related hypermotor epilepsy is clinically made based on semiological features and history. Hypermotor, exaggerated hyperkinetic movements such as kicking, cycling, and complex body movements are typical. Emotional facial expression and vocalization also accompany this semiology. Sometimes, it can be in the form of asymmetrical tonic posture and dystonic movements.[6] Three different categories are defined according to their level of precision: probable (witnessed)—basic clinical characteristics and the presence of a person who witnessed the seizure, clinical (video documented) —the diagnosis is made by the presence of 1 or 2 seizure recordings, definite (video-EEG documented)—1 or 2 stereotypical recordings and ictal or interictal epileptiform activity. However, the patient's interictal

EEG may be normal, and scalp EEG recording may not be informative during seizures. Therefore, it is necessary to combine EEG recordings with clinical findings. When polysomnography is performed, it may not be distinguished from parasomnias due to intense motion artifact, and even parasomnia attacks may accompany it. It has also been reported that parasomnia coexistence is common in cases with sleep-related hypermotor epilepsy. Patients' responsiveness to carbamazepine or other antiepileptic treatments also support the diagnosis. Still, the fact that they may be resistant to drugs in 30% of cases must be kept in mind.[1]

Sleep-related hypermotor epilepsy may be mistakenly confused with primary sleep disorders such as restless leg syndrome, sleep-associated rhythmic movement disorder and parasomnia, paroxysmal movement disorder, and non-epileptic psychogenic seizure. In our patient, attacks during falling asleep and waking up, described as jumping, were considered non-epileptic psychogenic seizures. Therefore, treatment was given only for generalized seizures that occurred during sleep. The patient's sleep quality was impaired, and she had excessive daytime sleepiness from time to time. The diagnosis of Sleep-Related Hypermotor Epilepsy was made because the attacks were stereotypical, started during sleep, showed clustering, were short lasting, and consciousness was quickly regained at the end of the attack and then fell asleep again. In addition, EEG abnormality was also detected concurrently with hyper motor movements.Since the attacks occur during sleep, we avoided paroxysmal movement disorder and non-epileptic psychogenic seizures. Restless Legs Syndrome diagnosis was ruled out due to a lack of discomfort in the legs and a desire to move before falling asleep and at rest.. The diagnosis of parasomnia was dismissed because the attacks are stereotypical and short lasting, and show clustering.

CONCLUSION

Sleep-related hypermotor epilepsy is a picture that should be considered in the presence of abnormal movements during sleep. Experiences of clinicians and EEG records are critical because misdiagnosis and treatment can negatively affect patients' lives. Video-EEG is a valuable diagnostic method in the differential diagnosis of epilepsy; the contribution of long-term monitoring comes to the fore, especially in recognition of attacks that occur during sleep.

Informed Consent: Written informed consent was obtained from patients/ patients' parents/ the parents of the patients/patient who participated in this study.

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