Uncovering Sleep-Related Hypermotor Epilepsy in a Child with Behavioral Changes

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Case

A 9-year-old male presented with 5 months of sudden onset changes in behavior, along with falling out of bed and sleepwalking, and most recently observed seizure-like activity. No clear triggers, trauma, or infection reported.

1. Behavioral changes

Hypersexuality, destruction of property, inappropriate language, and physical aggression. Reading capabilities and cognitive motor skills also regressed and he can no longer tie his shoes.

2. Semiology

Multiple episodes of nighttime full-body shaking, stiffness, and rolled back eyes that last for 3-4 seconds, followed by a period of terrified screaming and sleepwalking lasting 3-5 minutes. Upon awakening, he is amnestic and drowsy for 10-15 minutes.

3. Past Medical History

•Attention Deficit Hyperactivity Disorder
•Autism Spectrum Disorder
•Dyslexia

4. Family History

•Father: ADHD
•Paternal cousin: epilepsy

5. Developmental history

• 33-week premie, has a twin sister with dyslexia
•Mild speech delay

6. Physical Exam

Normal

7. Mental Status Exam

Behavior: Disinhibited, demanding
Psychomotor Activity: hyperactive, unable to sit still
Mood: irritable and excited
Affect: labile
Eye contact: limited
Attention: decreased

8. Differential Diagnoses

Parasomnias
nocturnal panic attacks
autoimmune encephalitis,
psychogenic nonepileptic seizures
Sleep-related hypermotor epilepsy

9. Work up

•CBC, CMP, TSH, UA, CT and MRI brain: normal.
•Autoimmune encephalitis panel: negative.
•CSF studies: normal
•EEG: bifrontal spike and wave discharges.

Disclosures

Authors report no disclosures.

Sleep-related hypermotor epilepsy (SHE) is a rare neurological disorder that can present with behavioral symptoms such as disinhibition and aggression, and cognitive dysfunction.

Diagnosis is made by capturing hypermotor seizures associated with epileptic discharge or interictal epileptiform abnormalities on EEG.

Oxcarbazepine is effective in seizure and behavior control.

Background

- Sleep-related hypermotor epilepsy (SHE) is a rare neurological disorder with an estimated minimum prevalence of 1.8/100,000 that manifests in childhood and adolescence.
- Seizures occur during sleep, and predominantly arise from the frontal lobes (Tinuper, 2016).
- Behavioral disturbances, such as disinhibition and aggression, and cognitive dysfunction can be seen (Braakman, 2011).
- Seizures may not be noticed early on, and behavioral changes may be the only presenting symptom to the clinic.

Discussion

- Given typical seizure semiology, behavioral changes related to frontal lobe dysfunction, cognitive decline, EEG findings; and unrevealing CSF analysis and antibody testing, patient was diagnosed with SHE.
- SHE seizures involve hyperkinetic movements, asymmetric tonic or dystonic posturing, vocalization, and epileptic nocturnal wandering.
- The cause of SHE is often unknown. SHE is suggested to be associated with ADHD (Zhang, 2012). Diagnosis is made by capturing hypermotor seizures associated with epileptic discharge or interictal epileptiform abnormalities on EEG (Tinuper, 2016).
- SHE can cause cognitive and behavioral issues that are difficult to treat and typically require lifelong medication for seizure control. Oxcarbazepine has been found to be effective in controlling seizures (Raju, 2007), resulting in an improvement in behavior, as was observed in this patient.

Conclusion

- In patients with behavioral changes and sleep-related symptoms, seizures should be ruled out.
- In the presence of seizure-like behavior, SHE should be kept in differentials and video-EEG should be obtained.
- Patients with SHE may achieve good seizure and behavioral control with Oxcarbazepine treatment.

References

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