Pearls & Oy-sters:

Frontal lobe epilepsy presenting as complex stereotyped movements

Jennifer Vermilion, MD
Jonathan W. Mink, MD,
PhD

Correspondence to
Dr. Vermilion:
Jennifer_vermilion@
urmc.rochester.edu

PEARLS

- Frontal lobe seizures have variable manifestations and can present as motor stereotypies.
- Frontal seizures that present as motor stereotypies typically have other ictal clinical features such as tonic or dystonic posturing, vocalization, autonomic signs, or an altered facial expression.
- Complex motor stereotypies typically present before 3 years of age. Onset in later childhood or adolescence suggests a different disorder.
- Capturing a spell on EEG is the key to diagnosis of frontal lobe seizures although ictal scalp EEG may lead to false-negative results.

OY-STERS

- Stereotypies are characterized by involuntary, fixed, patterned, purposeless, repetitive movements that tend to be distractible and almost invariably start prior to 3 years of age.
- It can be difficult to differentiate psychogenic nonepileptic seizures (PNES) from frontal lobe seizures solely based on history of psychiatric disorder, ictal pelvic thrusting or body rocking, or a rapid postictal recovery.
- Unlike PNES, frontal lobe seizures tend to be nocturnal, brief in duration, and stereotyped.

CASE REPORT A 14-year-old boy presented to the University of Rochester Pediatric Movement Disorder clinic for abnormal movements of 1 year's duration. He was referred to the Movement Disorder clinic for evaluation of complex tics. His movements were stereotyped and often preceded by a blank stare, sometimes associated with a "guttural sound." He also reported having a "numb" sensation prior to the movements. He denied volitional control of the movements or ability to suppress them. He reported movements only occurring during the day although parents recalled a period of nightly episodes witnessed while sharing a hotel room with him. They occurred both at home and at school. His abnormal movement

episodes were about 10–15 seconds in duration and followed by immediate return to normal activities (see video at Neurology.org). At the time of our evaluation, he was having over 10 episodes a day.

The patient's birth history was unremarkable and his developmental history was notable only for difficulty with vestibular processing and sensory integration. He was a strong student in a regular classroom setting. There was no contributory family history. Prior to our evaluation, he had a normal routine EEG done that captured both awake and sleep states. His neurologic examination was unremarkable other than a flat affect. He underwent a 24-hour ambulatory EEG that demonstrated 18 electrographic seizures arising from bilateral frontal regions with a right frontal predominance. Ten of the seizures occurred out of sleep and were not clearly recognized by the patient or family. Many of the seizures during wakefulness correlated with patient or parent button-push events. He underwent a 3T MRI scan that was normal. He was started on levetiracetam (Keppra) with resolution of spells.

DISCUSSION The semiology of frontal lobe seizures is varied and can consist of tonic spells, staring spells, hemiclonic movements, or hypermotor findings. These seizures often have an explosive onset and can be preceded by a scream. Seizures tend to be brief (less than 30 seconds in duration) and can be very frequent. Nocturnal seizures are very common. Most patients have a rapid recovery with only a brief postictal phase.¹ At this time, the etiology of our patient's seizures is not known. There are many potential causes of frontal lobe epilepsy, including genetic ones such as autosomal dominant nocturnal frontal lobe epilepsy (ADNFLE).² ADNFLE is characterized by brief nighttime seizures, positive family history, and normal neurologic examination with preserved cognitive function.³ However, daytime seizures can occur (though rarely) and mild cognitive impairment has been described. Several genes have been implicated in this condition and the diagnosis should be considered in patients with frontal lobe seizures and a normal brain MRI. However, given the negative family history and the striking predominance of daytime seizures in

Supplemental data
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From the University of Rochester, NY.

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our patient, we felt a diagnosis of ADNFLE was less likely. Thus, genetic testing has not been pursued.

Motor stereotypies are a less common presentation of frontal lobe epilepsy and can be localized to the prefrontal region.⁴ These can be confused with more typical stereotypies of childhood or motor tics. Stereotypies are characterized by involuntary, fixed, patterned, purposeless, repetitive movements that tend to be distractible.⁵ Our patient's movements had many of these features but were not clearly distractible. Stereotypies usually start prior to age 2 years and almost invariably start prior to 3 years of age. Teenage onset of symptoms argued against this diagnosis for our patient. Motor tics can be complex like our patient's movements but typically start in school-age children. The pattern is variable and they wax and wane over time. Frontal seizures that present as motor stereotypies typically have other ictal clinical features such as tonic or dystonic posturing, vocalization, autonomic signs, or an altered facial expression.² Our patient had many of these ictal features including vocalization and a smile at the end of the seizure (video).

Frontal lobe epilepsy can be difficult to diagnose in children and adults as many of the ictal features can be confused with other diagnoses, including sleep disturbances and psychogenic events.¹ Sleep behaviors tend to occur in later sleep cycles (deep non-REM phase), last longer in duration, and are less stereotyped in movements. Since most of the events reported by our patient were out of wakefulness, we had considered the diagnosis of PNES in our patient prior to obtaining his 24-hour EEG results. It can be difficult to differentiate PNES vs frontal lobe seizure solely based on history of psychiatric disorder, ictal pelvic thrusting or body rocking, or a rapid postictal recovery.⁶ Normal long-term video-EEG monitoring with captured ictal events does not exclude the

diagnosis of frontal lobe epilepsy since scalp electrode recordings are often insufficient. Characteristics to suggest frontal lobe seizures include nocturnal occurrence, shorter ictal duration, and stereotyped patterns of movements.

AUTHOR CONTRIBUTIONS

Jennifer Vermilion: examination of the patient, interpretation of the data, drafting of the manuscript, revision of the manuscript for important intellectual content. Jonathan W. Mink: examination of the patient, interpretation of the data, revision of the manuscript for important intellectual content.

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