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Seminars in
Pediatric
Neurology

Bizarre Semiology and Medically Intractable Seizures

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Case Report

A 15-year-old right hand–dominant young man with medically intractable epilepsy presented to our center to be assessed for surgical candidacy.

His seizures began at 5 years of age. Semiology consisted of manual motions where he would grab his groin and rock back and forth. These events were witnessed during the day and lasted for 20-30 seconds. Seizure frequency was regular but not more often than twice per day. During the episodes, he seemed aware, and there was no postictal confusion. Multiple medication trials were tried, and he ultimately achieved successful control with carbamazepine monotherapy and remained seizure free for the next 3 years.

At 8 years of age, there was seizure reoccurrence, but with altered semiology. He experienced a premonitory aura, after which he would locate to a safe place to go lie down before the start of the seizures. There was hypermotor semiology, which consisted of limb flailing, squealing, and ended with laughter. Compared with prior seizures, the frequency was increased in that he would have seizures 2-3 times per day for duration of 15-20 seconds. Again, there was a quick return to normal baseline cognition immediately after the event with little to no postictal phase. The seizures occurred predominantly during the day but did occur during sleep as well. Further therapeutic trials with multiple antiepileptic medications, including carbamazepine, clonazepam, lamotrigine, and gabapentin, failed to adequately control seizures. An electroencephalogram (EEG) was performed during one of these episodes, which did not show any electroencephalographic discharge. At that point, there was doubt as to whether these were truly seizures, and he was referred to psychiatry and treated for a conversion disorder. After 6 months of unsuccessful psychiatric intervention, he was referred to another neurologist, who diagnosed frontal lobe epilepsy (FLE).

The young man had been monitored twice in an Epilepsy Monitoring Unit before presenting to our center. EEG results were suggestive of frontal lobe seizures but failed to show lateralization. A 3-T magnetic resonance imaging (MRI) was performed, which was reported as normal. Frequent seizures persisted without any sustained interval of seizure freedom. Antiepileptic regimen at presentation included lacosamide 250 mg twice daily, oxcarbazepine 1200 mg twice daily, and ezogabine 200 mg 3 times daily. Ezogabine decreased his frequency of daytime seizures from daily to once every 2 weeks, but he continued to have nightly seizures. Although he denied any specific aura, his mother reported that he would still be able to get himself to a safe place and in a lying position most of the time. If he were not already lying down, he would fall over, writhe about, flail all of his limbs, squeal, and occasionally laugh at the end. There were no lateralizing features such as head or gaze version nor were there any neurologic signs thereafter. He was able to speak clearly immediately after. These seizures lasted for 15-20 seconds and were still without postictal symptoms. He had no loss of bowel or bladder function and did not bite his tongue during these events. There had been a single generalized tonic-clonic seizure, which occurred in the setting of a medication wean, and lasted 4-5 minutes. He had never been in status epilepticus.

The pregnancy had been unremarkable, and he was born at full term by spontaneous vaginal delivery, weighing 4.16kg. He reached all developmental milestones at the expected age. He had no history of febrile seizure, head trauma, central nervous system infection, autoimmune/inflammatory, or neoplastic or acquired cerebrovascular insult.

Socially, he has many friends and enjoys sports. However, he does have some trouble with mathematics and academic issues secondary to difficulty with focusing and attention.

Family history was positive for maternal aunt with febrile seizures as a child. Otherwise there is no history of seizures or neurologic conditions in the family.

On examination, the patient was normocephalic with a head circumference of 58.0 cm. No neurocutaneous

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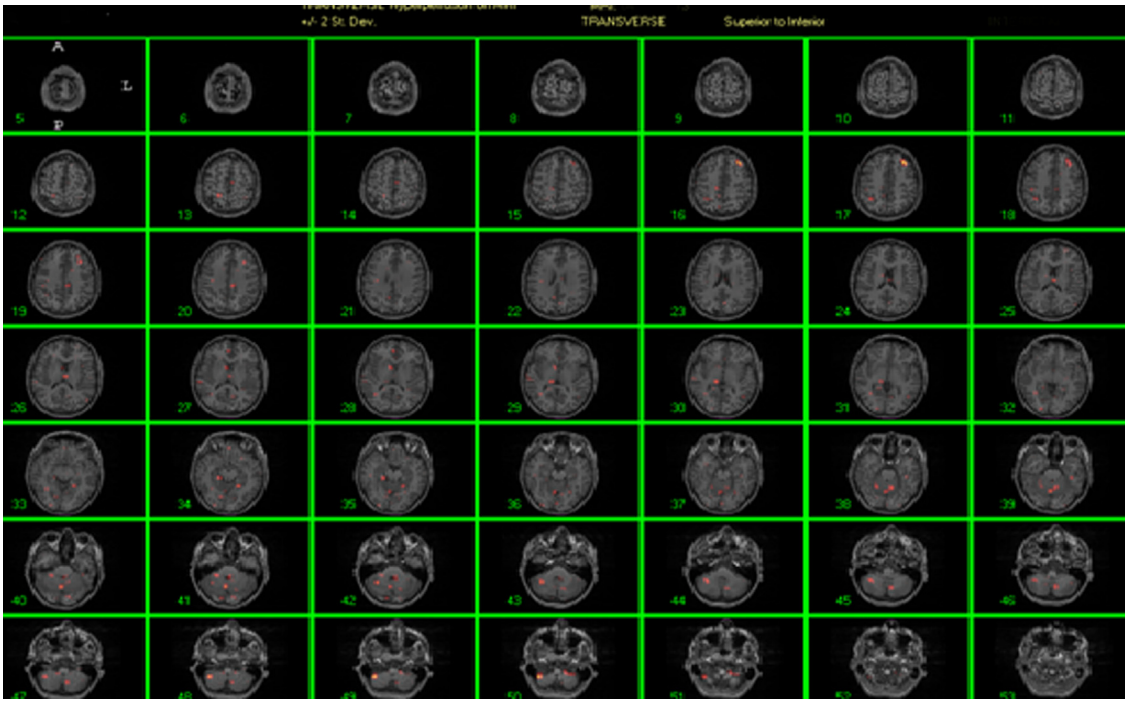

Figure 1 SISCOM analysis demonstrating a focus of ictal activity registering to the left frontal lobe anteriorly. SISCOM, subtraction ictal single-photon emission computed tomography co-registered to MRI. (Color version of figure is available online.)

stigmata were noted. Cardiopulmonary examination was benign. He was alert, oriented, and answered questions appropriately. There was no evidence of speech or language disturbance. Cranial nerves 2 through 12 were intact. Motor examination was equal and symmetric in upper and lower extremities both proximally and distally. Deep tendon

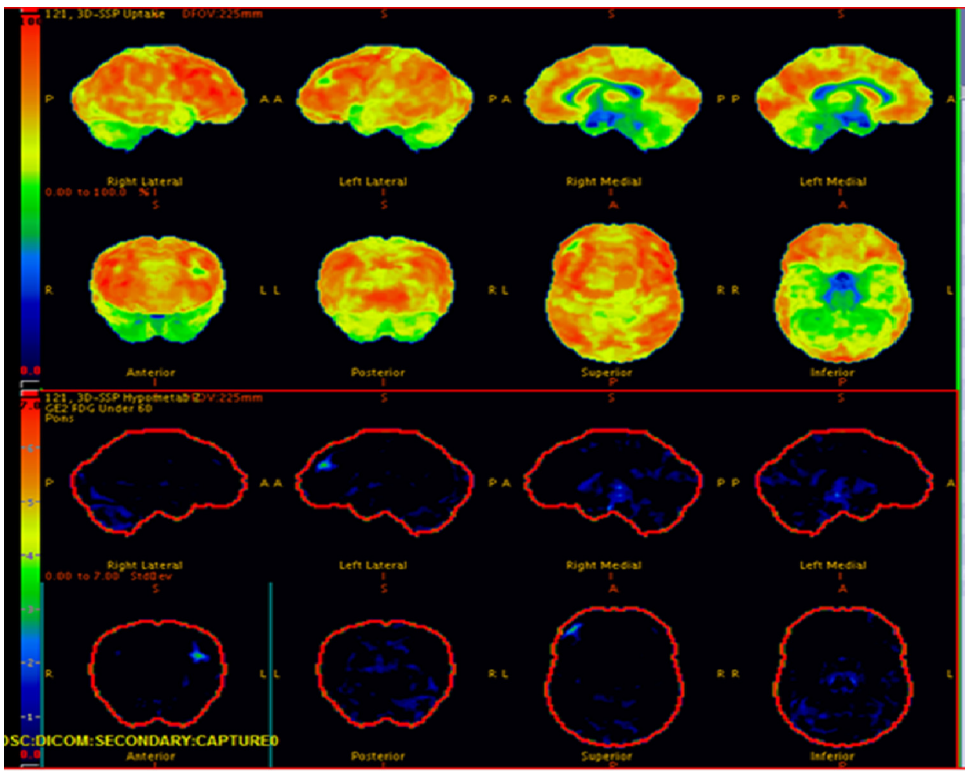

Figure 2 F-18 FDG-PET/CT showing a small area of decreased metabolism in the left anterior frontal lobe, corresponding in location to the abnormality seen on previous ictal brain perfusion scan, and represents the epileptic focus. CT, computed tomography; FDG, fluorodeoxyglucose. (Color version of figure is available online.)