Pediatric Epilepsy
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Case Study

Sophia is a 7 year old Caucasian female who presented to her primary care physician with concerns of incontinence in sleep. Sophia is an otherwise healthy child, with uncomplicated birth history, appropriate developmental progression with regards to early milestones, and previously diurnal/nocturnally toilet trained. No report of recent illness, infection or ill exposures. Sophia has had no polyuria, polydipsia or change in her eating habits; her weight is at the 50th% for age.

Sophia had been exceeding the expectations of school. Her teachers report her being focused the first two quarters, but have found her attention to be somewhat off over the last few weeks. Teachers have asked parents if Sophia has been getting good rest as she has been falling asleep in class.

Sophia’s mother and father note that she has had several episodes of urinary incontinence each week over the last 3-4 weeks. Parents have limited fluids prior to bed without improvement in the frequency of events. One night shortly after Sophia had laid down to sleep, mother heard a thump and went to check on her. Sophia’s eyes were wide open and deviated left, left arm was stiffened with rhythmic shaking, Sophia’s mouth was described as “sort of twisted” and Sophia was drooling. The event lasted 90 seconds. Following, Sophia was disoriented and very sleepy. Father reports his sister had similar events as a child.

Sophia’s neurological exam was normal, therefore her pediatrician made a referral to pediatric neurology. Given Sophia’s presentation, what differential diagnoses would one have for Sophia?

Case Discussion

Given Sophia’s history, the pediatric neurologist followed up by performing an EEG (electroencephalogram). High voltage interictal (defined as time between attacks or convulsions) right and bilateral centrotemporal spikes were noted during drowsiness and sleep. These interictal spikes suggest potential for epilepsy which is focal in nature. The pediatric neurologist started Sophia on oxcarbazepine (Trileptal) 150 mg twice daily given presenting history and EEG findings consistent with Benign Epilepsy with Centrolateral Spikes (BECTS). Since the start of medication, parents have not appreciated any seizure like activity and Sophia has had no further episodes of urinary incontinence in sleep. Sophia’s teachers feel her attention and performance has returned to normal.

What is BECTS?

BECTS, also known as benign rolandic epilepsy, is an epileptic syndrome with onset between 3 and 13 years of age and peak incidence at 7-8 years of age. It is often genetically transmitted as an autosomal dominant trait (Fenichel, 2009). Frequently, children diagnosed with BECTs have a close relative with history of febrile seizures or epilepsy. Without medication therapy, ~10% of patients with BECTS will have only one seizure in their lifetime, 70% have isolated infrequent seizures and 20% will have frequent seizure activity (Fenichel, 2009). Typically seizures associated with BECTs spontaneously stop following puberty. While termed benign, these seizures can appear scary and threatening to parents as well as the child experiencing them. With anti-epileptic medication, 20% of children with BECTS will have isolated seizures and 6% will have frequent seizures. The majority of patients with BECTS have seizures only in sleep, however 15% will have seizures while awake and 15% while both awake and asleep (Fenichel, 2009).

Typically seizure presentation with BECTs wakes the child from sleep. There may be associated paresthesias or numbness around the mouth as well as ipsilateral “mouth twisted” twitching of the face, mouth and pharynx resulting in drooling and inability to speak. Consciousness is often preserved. Daytime seizures typically do not generalize, however nocturnal seizures may spread to limb movements or even a generalized tonic clonic seizure presentation. Most of these type seizures will self-resolve within 1-2 minutes. Children with BECTS may be at greater risk for cognitive or behavioral problems, centered primarily around focus, reading and language processing difficulty. Therefore, it is important following diagnosis to monitor school function (Fenichel, 2009).
Epilepsy Discussion

Seizures are defined as a sudden, excessive, rapid discharges of neurons in the gray matter leading to clinical manifestations that may or may not impair or alter level of consciousness. Seizures are sudden and transient and may involve motor, psychic and sensory phenomenon (Fenichel, 2009). Following the first unprovoked seizure, recurrence risk ranges from 27-76%. When there are two or more unprovoked seizures, diagnosis of an epilepsy syndrome may be made by pediatric neurologist based upon presentation, careful attention to history and observation through video and electroencephalogram (Shovron, 2011). BECTs, which was discussed in the above case presentation, is classified as a complex partial form of epilepsy. The International League Against Epilepsy has defined epilepsy in terms of 1.) at least two or more unprovoked seizures occurring at least 24 hours apart 2.) one unprovoked seizure and probability of further seizures similar to the general recurrence risk of ~60% 3.) diagnosis of an epilepsy syndrome (Fisher et. Al, 2014). Seizures may be classified as complex partial or generalized depending on the parts of the brain involved. This determination can be made based upon clinical presentation and EEG findings. Medication therapy is tailored to the individual and the type of seizures he/she is having.

Resources:
