May 14th, 11:30 AM - 1:30 PM

CHILDHOOD ABSENCE WITH POLYSPIKE ICTAL-ONSET

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Abstract

Polyspike ictal-onset absence seizures have been reported in adult patients with idiopathic generalized epilepsy but are a unique pattern in the pediatric population. Absence seizures are usually associated with generalized spike and wave pattern. However, we present the case of a 10- year-old girl with Trisomy 21, speech, and cognitive delays who presented with atypical absence seizure associated with an unusual electrographic pattern of polyspike ictal-onset. Recognition of this unique pattern is essential as it can have therapeutic and prognostic implications.

Introduction

Absence seizures in children have been extensively described in the setting of unknown etiology, structural, or genetic generalized epilepsies. Several types of absence seizures have been described like typical absence, atypical absence, absence with eyelid myoclonia, and myoclonic absence as outlined by the International League Against Epilepsy [1]. Ictal patterns in these children vary widely according to seizure type and epilepsy syndrome. The pediatric literature on the ictal EEG [2] recording for absence seizures describes 2.5 to 4 Hz generalized spike-and-wave (GSW) discharges in typical absence seizures, generalized polyspike waves at 3 to 6 Hz with eyelid myoclonia and slower frequencies of <2.5 Hz in atypical absence seizures. There is no existing pediatric literature of polyspike ictal-onset for absence seizures, nor are there specific EEG features of epileptiform activity in Down syndrome. Tatum [3] & Fakhoury [4] et al. have described polyspike ictal-onset of absence seizure in adults. However, this is a novel EEG finding not previously described in children. We describe here a unique EEG finding of our patient and review the literature on polyspike ictal patterns.

Case Presentation

The patient is a 10-year-old girl with trisomy 21 adopted four years ago from China. Due to her adoption, there is no information regarding her perinatal or family histories. The child lives with both adoptive parents, attends homeschooling, and is doing well as per her mother and regularly participates in soccer. Her surgical history was significant for laryngoscopy, bronchoscopy, nasal endoscopy, tonsillectomy, and PDA repair. She presented to the neurology clinic at Children's Mercy Hospital due to new-onset episodes of staring, which started a month prior. Her mother described these as a complete behavioral arrest accompanied by eyes rolling upward. These events typically happen multiple times daily and last less than 30 seconds with no post-ictal changes. No automatisms or body jerks are associated with the seizures and no reported history of Generalized Tonic-Clonic (GTC), or other forms of seizures. Her physical exam shows typical features of Down syndrome, and neurological examination was significant for mild developmental delay involving all areas of developmental milestones, especially in speech. Magnetic resonance imaging of the brain was unrevealing.

An EEG was done to characterize the events [figure 1], and multiple stereotypical episodes were captured lasting 4-8 seconds. Clinically, she was noted to have eye rolls associated with behavioral arrest and unresponsiveness. She was unable to recall the word given during the episodes. Ictal EEG recordings showed initial high voltage generalized 14-18 Hz polyspikes, which occurred in isolation or followed by 2-3 Hz generalized spike and wave discharges (Fig. 1). The faster ictal discharges were the dominant ictal activity and appeared to be embedded in some ictal SW discharges. Interictal EEG recordings showed few bursts of high voltage 4 to 6 Hz irregular GSW, 3 to 4 Hz polyspike and wave complex (PSW), as well as fragmentary bursts noted both during the awake and asleep state. Background rhythm revealed a poorly sustained 7-7.5 Hz posterior dominant rhythm for age as well as diffuse moderate voltage 4-6 Hz slowing in all the head regions. No abnormalities were elicited with the photic stimulation or hyperventilation.

Our patient most likely has a new onset of genetic generalized epilepsy with exclusive absence seizures at age ten. Based on the clinical profile and EEG findings, the child was started on Levetiracetam in the last clinical encounter with ongoing breakthrough seizures for which medication are optimized.

Discussion

In the current ILAE classification [1] , several types of absence seizures have been described like typical absence, atypical absence, absence with eyelid myoclonia, and myoclonic absence. Typical absence seizures usually present with complete impairment of awareness consisting of an abrupt start and end. The absence seizure is considered atypical if it is associated with abrupt onset, offset, and significant changes in tone. Awareness and responsiveness can be partially retained during some generalized seizures, like absence seizures with eyelid myoclonus, atypical absence, and myoclonic absence. The epileptiform discharges will differ depending on the specific epilepsy syndrome and seizure types. Typical absence seizures consist of 3-Hz frontally predominant generalized spike-and-wave (GSW) discharges in childhood absence epilepsy and relatively faster frequencies in juvenile absence seizures. In atypical absence seizures, the EEG will reveal irregular GSW discharges below 2.5 Hz. The ictal EEG pattern of eyelid myoclonia consists of generalized polyspikes waves at 3 to 6 Hz triggered by active eye-closure and photic stimulation [4]. Even though the eyelid myoclonia can be associated with generalized polyspikes, our patient did not have features of Jeavon syndrome-like eyelid myoclonia, photosensitivity, and eye closure induced epileptic paroxysms.

The current ILAE seizure classification [1] does not identify absence seizures with fast rhythmic patterns, such as those present in our patient’s EEG, as a distinct seizure type. The paroxysmal event may be classified in the category of atypical absence seizures due to unusual EEG findings that include fast activity (polyspike ictal onset) and an abnormally slow background. Atypical absence seizures are seen in cognitively impaired children with epilepsy. However, absence seizures are rare in patients with Down syndrome [6].

The pathophysiology for polyspike ictal-onset absence seizures in patients with generalized epilepsy has remains elusive [7]. Seizures in Down syndrome may be a secondary effect of both the functional and anatomical neurological decline [8]. We hypothesized that the abnormal neuronal development and the ongoing brain maturational processes in Down syndrome, affected by age, seem to be involved in the modification of the typical electroclinical pattern of absence seizures. Patients with Down syndrome typically present with infantile spasms and can have various other seizure types, including focal, myoclonic, GTC, and atonic [6]. However, our patient presents with an isolated seizure type consisting of absence seizures with eye rolls and therefore is not typical for Down syndrome-associated epilepsy.

The phenomenon of fast rhythmic activity (polyspikes) seen in our pediatric case appears in the literature to occur exclusively in adults. Fakhoury and Abou-Khalil et al. (1999) [3] reported the fast 10-15 Hz rhythmic discharges in 5 adult patients with absence seizures. They can occur in isolation or embedded in more typical SW discharges accompanying typical absence seizures. Tatum et al. (2010) [4] reported that absence seizures with polyspike onset 3-Hz GSW are seen in adult patients. This pattern may represent an intermediary form of idiopathic generalized epilepsy (IGE) that may be drug-resistant and was present in six of seven patients. Michelucci et al. (1996) described 3 of 12 patients with prolonged polyspike ictal-onset before 3-Hz GSW associated with typical absence seizures. Marini et al. (2003) [10] also suggested that patients with prolonged polyspike ictal onset 3-Hz GSW may require lifelong anti-convulsant therapy.

There are several variations of this unusual ictal manifestation that include a rhythmic fast ictal discharge for >50% of the ictus preceded the 3-Hz GSW, an intermixed polyspike pattern with 3-Hz GSW after onset, or purely a polyspike ictal-onset [3, 4, 9]. In our patient, three seizures were captured, all characterized by initial polyspikes lasting for 2-7 seconds followed by GSW, purely polyspike ictal-onset as well as polyspike with embedded GSW.

The fast rhythms may represent harmonic variations of the spike-and-wave activity or modification of the typical electroclinical pattern. Whether this polyspike pattern evolves from slow frequencies of <2.5 Hz is not proven, but in our case, it appeared as a novel ictal pattern. There have been multiple reports [11, 12] with no reference to polyspike ictal onset in a large number of children with the typical absence seizures. Even though some reports [13, 14] have classified absence seizures in patients with Lennox-Gestaut syndrome to be related with fast rhythmic activity as atypical absence seizures, or the generalized tonic seizure. However, our patient does not have any electroclinical features of Lennox-Gestaut syndrome.

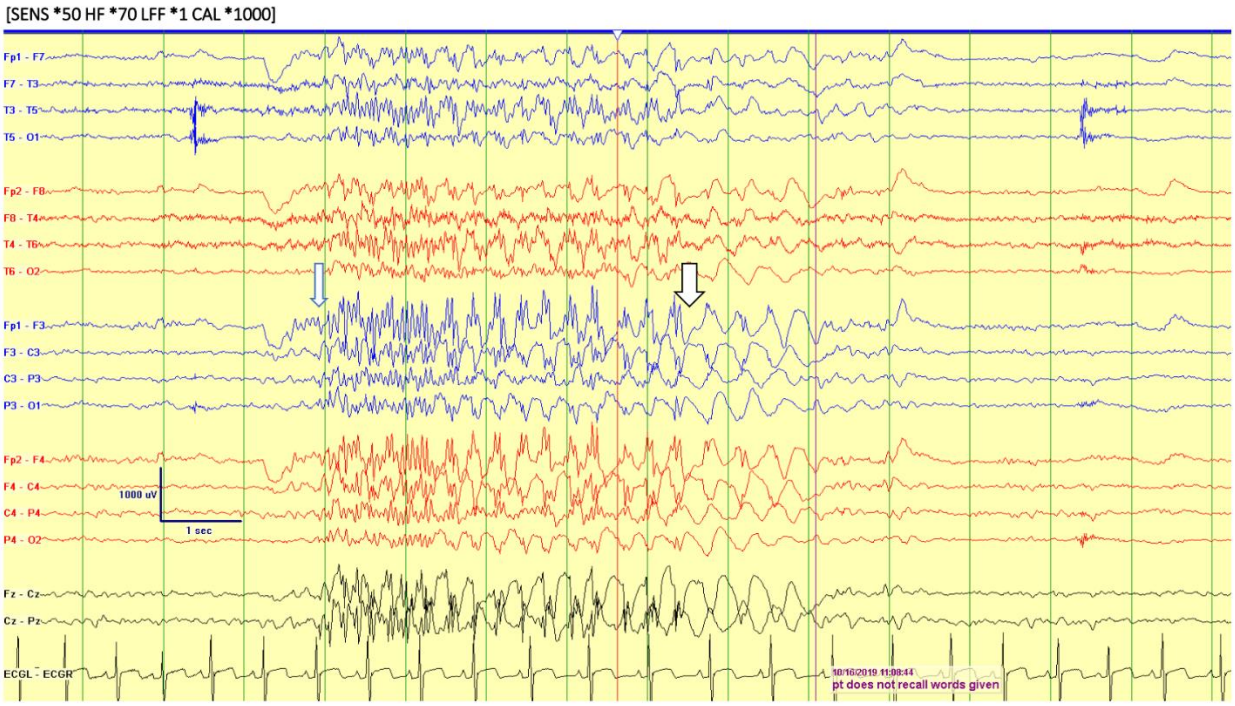
Electrographically, ictal patterns in children, can differ depending on age, seizure and epilepsy type, and epileptic encephalopathies. An understanding of the full spectrum of ictal activity in children is necessary for neurophysiologists for diagnostic and therapeutic purposes.

Conclusion

In conclusion, there is little information in the pediatric literature on the incidence of such an ictal pattern in absence seizures and their clinical significance. Recognizing this unique

pattern is of clinical importance as it is reported with an unfavorable outcome with refractory epilepsy in adult literature. This unique polyspike ictal pattern for absence seizure in the pediatric population may be a modification of the typical electroclinical pattern. It could be secondary to the brain maturational process in Down syndrome associated with cognitive impairment or an unrecognized novel EEG pattern of clinical significance.

Figure 1



Legend for figure 1:

The Ictal EEG (long arrow) with initial high amplitude frontally predominant generalized polyspikes (long arrow) followed by 2-3 Hz generalized spike and wave (short arrow). Also note the intermixed polyspikes with the spike and wave.

This absence seizure was associated with staring, subtle behavioral arrest and uprolling of eyeballs. Patient was not able to recollect the words given during this event.

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