Paroxysmal Nonepileptic Spells in Adolescence

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Abstract

Paroxysmal nonepileptic spells refer to events that clinically can resemble epileptic seizures but are not caused by abnormal epileptiform activity in the brain. These spells can often be difficult to diagnose owing to clinical similarities, but a correct diagnosis is needed to prevent unnecessary treatment with antiepileptic drugs. We review the most common diagnoses in adolescents that can be mistaken for epilepsy, including migraine, syncope, movement disorders, sleep disorders, and psychologic disorders.

Keywords
- paroxysmal nonepileptic
- spells
- psychogenic seizures

Introduction

Paroxysmal nonepileptic spells are often difficult to diagnose in pediatric patients owing to overlapping clinical features with epileptic seizures. A diagnosis of epilepsy is typically associated with pharmacologic and social sequelae, and an incorrect diagnosis can create an unnecessary burden highlighting the need for proper identification of paroxysmal nonepileptic spells. The adolescent population can present a further unique challenge compared with younger patients, and the common etiologies of nonepileptic events can often differ. These events can be seen in both psychiatric and nonpsychiatric conditions with the most common paroxysmal spell with a psychiatric basis being psychogenic nonepileptic seizures (PNES).[1] The objective of this review is to identify common mimickers of epilepsy in adolescents, including migraine, syncope, movement disorders, sleep disorders, and psychologic disorders.

Migraine

Children and adolescents can have a wide variety of presenting symptoms with headaches, which makes the initial diagnosis difficult. In addition, migraine equivalents involving recurrent intermittent episodes without a clinical headache can be seen in 70% of children and adolescents with headaches.[2] Given similar neurologic and autonomic features of both migraines and epilepsy, it is important to understand clinical features that are more indicative of a certain diagnosis. Furthermore, an overlap can be seen, as postictal headaches are often reported in patients with epilepsy.[3]

A common form of headache in adolescents is migraine and at times symptomatology can resemble characteristics of seizures. For example, visual auras can be present in both migraines and occipital seizures; however, distinguishing features include an aura of longer duration in migraine that can propagate over several minutes and tend to spread over the visual field. Patients often describe a serrated or zigzag appearance to the visual disturbance.[4] This is in contrast to epileptic occipital auras, which tend to be stereotypical and brief, often with colored circular patterns.[5] Auras associated with migraine are also typically followed by a headache with typical migrainous features. In similar fashion, migrainous somatosensory or auditory auras tend to last for longer periods with greater simplicity compared with brief stereotypical symptoms that are more complex in focal epilepsy. Atypical migraines, such as basilar-type migraine or acute confusional migraine, can cause prolonged confusion that can mimic nonconvulsive status epilepticus or perhaps a prolonged postictal state. Basilar-type migraine can also trigger vasovagal syncope or possibly even convulsions.[6] Acute confusional migraine occurs more commonly in children and adolescents with the majority of patients being younger than 21 years.[7] Given the clinical similarities, it has been suggested that acute confusional migraine might be a variant of transient global amnesia seen more commonly in adults.[8] The level of associated dysfunction can vary from mild behavioral changes to even coma. In addition to confusion, language deficits can develop and last for hours, making the differential

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concerning etiology upon initial presentation vast, including ingestions, infections, metabolic derangements, stroke, or postictal changes.[9] Electroencephalography (EEG) recordings can possibly reveal bitemporal or bioccipital slowing, though these changes, if present, resolve after the episode has resolved.[10] Even for pediatric patients with common migraines, 36% can have EEG abnormalities during a migraine, and 16% have abnormalities while headache free.[11] Other atypical forms of migraine such as familial hemiplegic migraine or alternating hemiplegia can be seen in adolescents but tend to present initially in the earlier childhood year.

It is also possible to have nonorganic movement disorders associated with migraines. A study from Mayo Rochester[12] in 2015 identified functional movement disorders in 4% of chronic migraine patients and 1% of episodic migraine patients with the most prominent movement being nonepileptic shaking spells.

Syncope

Syncope refers to a sudden loss of consciousness, often secondary to a reduction in cerebral perfusion. Syncope is commonly confused with seizure and presents a dilemma for the clinician, especially when the onset of the event was not witnessed. However, obtaining a careful and complete history often leads to clinical clues that can distinguish these two etiologies. Neurocardiogenic syncope is the most common type of syncope seen during childhood and adolescence, and is considered a reflex syncope with transient disturbances in autonomic control.[13] This form of syncope can occur with the patient standing or sitting, and is often preceded by a prodrome of lightheadedness, nausea, and a gradual fading of binocular vision. Associated pallor and diaphoresis can also occur. Vasovagal syncope may be seen in adolescents as well and is usually precipitated by events such as pain or fear.[14] Positive motor activity can be seen with various forms of syncope and should not automatically indicate a primary epileptic event. During prolonged periods of hypoperfusion versive eye movements, tonic stiffening and clonic jerks can occur.[15] After the syncopal event, patients may feel tired for some time, though the return to baseline generally is quicker than that seen with epileptic events.[13]

Other forms of syncope are less common in adolescents but should be considered in the right clinical setting. Cardiogenic syncope can be associated with ominous signs such as syncope upon exertion and should promote a cardiac evaluation.[6] This should not be confused, however, with postexertional postural hypotension that can occur and produce syncope and is less concerning.[16] Hyperventilation that can occur during panic attacks can also lead to cerebral vasoconstriction and decreased cerebral blood flow resulting in syncope.[17]

Testing for evaluation of syncope should be individualized for the patient and directed based on the historical clues. Electrocardiography can be useful for evaluation of cardiac arrhythmias, and orthostatic blood pressures can easily be obtained in patients with postural syncope. The use of routine EEG is questionable, given the low yield of information in between events.[18] However, in patients with recurrent events, prolonged video EEG monitoring can be valuable when events are captured. During syncope, the EEG tends to demonstrate generalized slowing, and with severe events can reveal background suppression.[19] Further testing, such as tilt testing or continuous Holter monitoring, should be determined based on individual cases.

Movement Disorders

Movement disorders are rarely confused with seizure activity, though some overlap in symptoms can occur. For example, in patients with tics or Tourette syndrome, atypical motor movements can be confused for simple focal seizures or myoclonic jerks. Tics are defined as rapid, recurrent motor movements or vocalizations that wax and wane over time. Tics tend to be motor or vocal and can be further divided into complex or simple depending on the complexity of tics.[20] These movements affect up to 1% of the population and there is a male predominance.[21,22] Although the mean age of onset is 5 years, the severity of the tics is often most prominent between 8 and 12 years of age.[23] After these years, there tends to be an improvement in both intensity and frequency of the tics and some children, by the age of 18 years, no longer have any impairment from their tics.[24] However, there remains a population of patients who continue to have severe and debilitating tics into adulthood. Diagnosis is based on clinical findings and certain clinical clues can help differentiate tics from other movement disorders and seizures. Patients with tics tend to report brief, rapid, nonrhythmical recurrent movements that can be temporarily suppressed, though this suppression leads to an uncomfortable sensation. This awareness of a tic being a self-directed conscious movement that is performed to satisfy an internal urge is expressed in the majority of patients with tics by the age of 12 years.[25] Through careful obtainment of these historical clues, differentiation between tics and epileptic seizures is usually clear.

Paroxysmal dyskinesias represent another movement disorder that at times can be confused for seizure activity. Paroxysmal kinesigenic choreoathetosis or dyskinesia is characterized by brief episodes of choreoathetotic or dystonic movements, often initiated by a triggering event such as movement.[26] The episodes can be unilateral or bilateral and typically are not associated with alteration in consciousness. These events do not represent an epileptic process and EEGs tend to be normal[27]; however, there have been reports of effective treatment with various antiepileptic medications.[26] Typical age at presentation is between 6 and 15 years.[28] Paroxysmal dystonic choreoathetosis is a similar phenomenon but is not triggered by movements.

Sleep Disorders

Sleep disorders can be confused with epilepsy when the patient presents with paroxysmal recurrent events. Some epilepsy syndromes, such as nocturnal frontal lobe epilepsy, can include nocturnal seizures often with bizarre descriptions and should be considered in patients with stereotypical

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events during sleep.[29] Common sleep phenomenon seen in younger children such as night terrors is not typically seen in the adolescent age group. The possibility of epilepsy may be raised with parasomnias such as sleep walking and talking, or with other sleep disorders such as narcolepsy and periodic limb movements.

Narcolepsy is a rapid eye movement sleep disorder characterized by excessive daytime sleepiness, cataplexy, sleep paralysis, and hypnagogic hallucinations; however, only a small percent of pediatric patients will report all of these symptoms.[30] When the symptoms occur independently, these phenomena can be mistaken for epilepsy. For example, hypnagogic hallucinations seen in narcolepsy can be simple or complex and occur as the patient is falling asleep or upon awakening. However, the timing of these hallucinations as well as the patients insight into what is occurring suggests a diagnosis of narcolepsy.[31] An association between narcolepsy and epilepsy has not been established and the two etiologies can be differentiated with the help of polysomnography and multiple sleep latency tests.[30]

Periodic limb movements of sleep are characterized by involuntary movements of the limbs occurring predominantly during non–rapid eye movement sleep. The majority of the cases will involve the legs only.[32] Typical movements include dorsiflexion of the ankle with extension of the great toe and can be unilateral or bilateral.[33] Clinical history and polysomnography should help differentiate these movements from epileptic events.

Sleep starts or myoclonus may also be confused with possible epileptic activity, though certain historical clues make the diagnosis of this benign phenomenon easy to detect. Also known as benign hypnic myoclonus, this movement involves sudden diffuse jerks that occur upon falling asleep.[33] Patients at times describe a subjective sensation of falling when the jerk occurs.[32] Unlike myoclonic epilepsy, these movements are restricted to sleep and occur as the patient transitions from wakefulness to sleep.

Psychological Disorders

The most common disorder to be mistaken for seizure activity in the adolescent is PNES. PNES refers to changes in perceived consciousness or behaviors that clinically are similar to epileptic seizures, but are not associated with an ictal epileptiform discharges.[34] This is a common diagnosis encountered in epilepsy-monitoring units, given the clinical presentation of abnormal motor activity and alteration in apparent consciousness. The overall prevalence has been estimated at 2 to 33 per 100,000 with a female predominance.[35–38] There is also an increasing incidence with progressing age in childhood.[1] The true incidence in children is difficult to determine, but previous estimates have been between 1 and 9%.[39]

The correct diagnosis is often delayed, given the complexity of this disorder, and even the most trained epileptologist can mistake a nonepileptic spell for an epileptic seizure and vice versa. This is partially because there is no single characteristic that identifies a PNES. However, previous studies have identified common symptoms that can be suggestive of a psychogenic presentation. Characteristics such as asymmetrical thrashing movements, side-to-side head movements, pelvic thrusting, lack of a stereotypic pattern, waxing and waning of behaviors, sudden return to baseline followed by a prolonged generalized event, and rapid breathing at the end of the spell are clinical manifestations that can be more prominent in psychogenic seizures.[40–44] In addition, PNES tend to last longer than epileptic seizures. A review of PNES in children by Szabó et al[45] in 2012 found that PNES in children lasted on average 186 seconds longer than epileptic seizures in children. Activation procedures can also be used to induce typical events in patients with PNES.[46]

Although the above are possible characteristics of psychogenic seizures, there can be an overlap in symptoms between epileptic and nonepileptic events, often requiring video EEG monitoring to correctly differentiate between the two. Previous studies have shown that even experienced epileptologists can incorrectly distinguish between epileptic and psychogenic seizures from video recordings alone in up to 30% of cases.[47] The most common type of epileptic seizure to be mistaken for a psychogenic seizure is a frontal lobe seizure that can often include bizarre presentations.[48]

It is important to recognize these PNES symptoms and make the correct diagnosis, as an incorrect diagnosis of epilepsy can lead to treatment with unnecessary antiepileptic medication, which will also be ineffective. In addition, the diagnosis of PNES requires a different approach to treatment, including cognitive behavioral psychotherapy, stress reduction, and reassurance.[49] The communication of the diagnosis to the patient and the parents is also of vital importance with emphasis being placed that the child is not faking these events.[50] Typically, in children and adolescents, PNES are thought to be a manifestation of conversion disorder[51] and the diagnosis should be conveyed positively without blame placed on the patient.[52] Adolescence represents an awkward period of time when a child morphs into an adult both physically and psychologically. This period of change is accompanied by many psychosocial stressors such as self-image, peer pressure, sexual changes and encounters, relationships with others, and many other problems that can lead to confusion and unresolved psychological conflicts that may manifest as physical symptoms that may include PNES. A common pattern is that PNES attract much attention from family members and friends, and often result in ambulance rides, emergency department visits, medical tests, and hospitalization, which commonly serve to convince the afflicted patient that they are indeed very ill. These all commonly result in a pattern of recurring events with a self-perpetuating, increasing frequency of occurrence. Recording the typical events with video EEG is the gold standard for making the correct diagnosis and providing appropriate intervention. It is extremely important to convince the patient and family that the spells are not epileptic seizures as confirmed by the absence of epileptic activity on the EEG. This is the first step in intervention. It is next important to obtain psychological consultation to explore the potential for underlying psychological conflicts that need to be dealt

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with appropriately. Failure to complete this final step may result in persistence of PNES versus the development of other somatized complaints or disabilities. With early intervention, it appears that children and adolescents have a significantly better prognosis compared with adults with up to 81% free of PNES at 3-year follow-up.[53] Of note, it should be recognized that patients can have both epileptic and psychogenic events, therefore requiring a complex regimen to treat the individual etiologies. These children tend to have a less favorable outcome.[52]

In conclusion, paroxysmal nonepileptic spells continue to be a challenge and early identification of the correct diagnosis can minimize unnecessary investigations and treatments. These spells can have an organic or psychogenic basis and because of overlapping neurologic symptoms with epileptic seizures, diagnosis can be delayed for many years. It is important for the health care provider to be familiar with common nonepileptic spells seen in adolescents as described earlier to avoid unnecessary treatment with antiepileptic medications and to instead formulate appropriate treatment for these patients.

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