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CASE REPORT

A child with Epileptic and nonepileptic Head Drops

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ABSTRACT

This paper aims to document a case of head drops with distinct epileptic and nonepileptic phases. A baby, born at term and weighing 2.9 kg, developed seizures and hypoglycemia (blood sugar 39 mg/dl) at 24 hours of age. She received special care for 24 hours and advice to continue phenobarbital for one year. Tonic-clonic seizures recurred at six weeks of age and were controlled by a combination of four-drug anticonvulsants. The treatment was tapered off after two years of seizure-free interval. At seven years of age, the tonic-clonic convulsions reappeared, only to be replaced by head drops after eight months of intensive antiseizure treatment. The head drops were nonepileptic for two years and epileptic later on before subsiding altogether with a new antiseizure combination. Cognition rapidly improved afterward.

Keywords
Epileptic head drops; Nonepileptic head drops; Smartphone videos for epilepsy diagnosis.

Introduction

Many paroxysmal events may be mistaken for epilepsy. A study observed that 30% of children (55/184) referred with epilepsy diagnosis had a nonepileptic event (NEA) (1). Correct diagnosis of NEA may avoid potentially harmful antiepilepsy treatment and high emotional costs to the patients and their families. It may also do away with restrictions on the child's lifestyle, such as restrictions on daily routines such as sports activities. NEA is an event that may resemble an epileptic seizure but is not associated with abnormal cortical electrical discharges. Head drops (HD) are a nonpsychogenic type of NEA (2). HD is an abrupt and repetitive neck flexion associated with drooping eyelids. The motor component of a non-epileptic attack (NEA) mimics epilepsy. In a nonepileptic HD, the head's fall and rise occur at the same velocity, unlike in an epileptic seizure, where the fall of the head is typically more rapid than the rise of the head (3). Video–EEG monitoring is a gold standard test to distinguish NEA from electrographic seizures (4), despite the cost involved and the time consumed. In low-resource settings, smartphone videos may be a good proxy for video-EEG monitoring.

We report a case where the child has gone through distinct epileptic and nonepileptic phases. A new set of antiepilepsy drugs led to the cessation of both epilepsy and head drops.

Case report

A nine-year-old boy, born from a non-consanguineous marriage, presented to the Primary Health Centre (PHC), Kamshet, for paroxysmal events. The mother showed smartphone videos that revealed abrupt neck flexion associated with drooping eyelids and brief periods of decreased responsiveness. There were no tonic-clonic limb movements or rolling of eyeballs. The mother noticed 10-12 events daily, of which 6-8 occurred in the morning, soon after getting up. There was a history of falls to the ground and injuries during severe attacks.

The boy was born at term and weighed 2.9 kg after an uneventful pregnancy and childbirth. The mother, not investigated for gestational diabetes in this pregnancy, was diagnosed with diabetes mellitus in the subsequent pregnancy. The baby had a seizure, not very severe or prolonged, with a blood sugar level of 39 mg/dl at 24 hours of age and requiring admission to the special care unit. No treatment details were available. On discharge, the baby received phenobarbital. Severe tonic-clonic seizures occurred at six weeks of age. Stepping up the dose of phenobarbitone or the addition of sodium valproate and clobazam did not help at this stage. The seizures ceased only after the addition of lamotrigine and impanel. EEG showed bilateral epileptiform activity, as mentioned in the discharge card, but no details were available. An MRI scan report mentioned hippocampal atrophy and flattening of the dentate gyrus. Antiseizure treatment continued for two years of a seizure-free period and a normal EEG.

The course was uneventful up to the age of seven years, except for slow growth and development. The child started walking at two years old, was very cranky, and did not mix with his peers. At six years he showed no interest in reading and writing. At seven years, he fell to the ground following a major attack of tonic-clonic seizures and upward rolling of his eyes, followed by 5-6 episodes over a month. A four-drug intensive antiepilepsy treatment controlled the seizures. However, the HD continued. On average, 8-10 attacks occurred daily in the morning, of which 3-4 were severe, probably following the circadian cortisol surge (5), and the subsequent ones were restricted to head nods (video 1). The child preferred to remain seated for 2-3 hours after getting up for fear of falls. The HD was also more frequent and severe after an afternoon nap. Spending time on a mobile phone and activities such as reading and writing often induced HD, but the events were fewer in front of a television screen or when in a happy mood.

When presented to the PHC, the child appeared sleepy and drooled saliva. Besides this, there was

no noticeable finding. The action plan included stopping clobazam and impanel, continuing valproic acid and phenobarbital, and administering a mega-dose of vitamin D, 600,000 IU.

The frequency of HD remained the same. However, his alertness and cognitive function started to improve steadily. The mother was apprehensive about a complete stoppage of antiseizure treatment. After about six months, the nature of the 'attacks' changed entirely. Tonic-clonic seizures emerged along with HD (video 2). Suspecting epileptic head drops, the child received carbamazepine and topiramate in addition. The HD and the seizures subsided, followed by a rapid improvement in cognition.

Discussion

The child, an infant of a diabetic mother, had hypoglycemic seizures in the neonatal period. The seizures recurred in infancy and at seven years of age. The seizures were replaced by nonepileptic head drops for two years (video 1) and appeared later as epileptic HD (video 2). The addition of a new set of antiepilepsy drugs, namely carbamazepine and topiramate, led to a complete cessation of seizures and HD. The response to antiseizure treatment may suggest that the epileptic and nonepileptic phases had common origins. It is also possible that the HD phase (video 1) was a manifestation of a partially controlled epileptic phase. The diagnosis of epilepsy and nonepileptic events was made by examining the home videos provided by the mother. This fact emphasizes the usefulness of smartphone videos in low-resource areas where the internet has a broader reach.

The diagnosis of epilepsy is generally based on clinical history taking and interictal EEG. The diagnostic value of a single interictal EEG is low. EEG may be normal in children with epilepsy and may be epileptiform in 2–5% of children without epilepsy (1). Interobserver agreement among child neurologists about the diagnosis of the first-time paroxysmal event was only fair to moderate (6). The rate of misdiagnosis of epilepsy in a national sample of difficult-to-treat patients from a developed country is exceptionally high (1). At times, it may be challenging to differentiate epilepsy from NEAs. The latter tend to occur in clusters with a benign course (7). Children with epileptic HD had a higher prevalence of facial expressions and subtle myoclonic movements. (3). In one study, home videos of four paroxysmal ocular deviations suspected of being epileptic seizures were diagnosed with nonepileptic events (2). In the same study, three cases of head drops, referred to with a diagnosis of nonepileptic events, turned out to be epileptic spasms.

Conclusion

In our case, with the background of earlier epileptic seizures, HDs were initially misdiagnosed as refractory seizures in the absence of vide-EEG monitoring. This led to the addition of multiple antiseizure drugs, and as a result, the child was sleepy and had drooling saliva at the time of the first presentation at the PHC. The sequence of events began with tonic-clonic seizures and abnormal EEG in early infancy, followed by multiple nonepileptic events in the form of HD every day. The reappearance of epileptic HD and, finally, response to a new combination of antiseizure treatment is noteworthy. HD in the child was likely a phase of partially controlled seizures that responded to the new combination of antiseizure drugs.

Conflict of Interest:

None.

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Videos 1 and 2:

Head drops1.mp4

Head drops 2.mp4