Case Report

Panayiotopoulos syndrome: A benign childhood epilepsy syndrome masquerading as head injury

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ABSTRACT

Panayiotopoulos syndrome (PS) is a benign childhood epilepsy syndrome characterized by autonomic seizures and electroencephalogram (EEG) showing predominant occipital paroxysms. PS may easily be confused with many non-epileptic conditions such as atypical migraine, gastroenteritis, or syncope. We here present a 5 ½-year-old boy who presented to us with a history of head injury followed by vomiting, seizures, and prolonged unresponsiveness. Neuroimaging was normal, and on detailed history, it was clear that autonomic symptoms preceded the head injury and so an EEG was done. Interictal EEG showed bilateral occipital spikes and wave discharges with characteristic "fixation-off sensitivity." Hence, the seizure semiology along with the EEG pattern helped to clinch the diagnosis. Parents were counseled about the benign nature of the condition.

Key words: Autonomic symptoms, Childhood, Occipital epilepsy, Panayiotopoulos syndrome

Panayiotopoulos syndrome (PS) is an idiopathic childhood epileptic syndrome recognized by the International League against Epilepsy in 2001 [1]. Autonomic seizures are the sine qua non of PS [2]. PS supposedly affects up to 13% of children aged 3–6 years [3], yet it is underdiagnosed and underreported [4]. It is a benign self-remitting condition with spontaneous remission within 2–3 years from its onset. Due to the prominent dysautonomic features and the unusual symptomatology of PS, it can be confused with other non-epileptic conditions such as syncope, gastroenteritis, encephalitis, and migraine. Therefore, a detailed history and evaluation will help to diagnose this condition and counsel the parents about its good prognosis which goes a long way in allaying the parental anxiety.

CASE REPORT

A 5 ½-year-old boy came to emergency with alleged history of accidental hitting of head on the door knob, followed by vomiting, up rolling of eyes, deviation of mouth and head to right side, and stiffening and jerking of whole body lasting for 15–20 min, was aborted with IV midazolam in emergency.

On examination, his vitals were stable; no signs of external injury, pupils were equal and reactive. As he was in postictal state, detailed neurological examination could not be done at that time.

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The child was shifted to ward and given a loading of IV phenytoin (20 mg/kg). In view of head injury, urgent non-contrast computed tomography (NCCT) brain was done. Child became normal with no neurological deficit after postictal sleep and neurosurgical review was also normal. As NCCT report revealed suspicious mild subdural hematoma and appeared non-conclusive, magnetic resonance imaging (MRI) brain was also done and was normal. At this point, we revisited the history as knocking the head on door knob appeared to be a trivial injury. Question was – did the minor head injury lead to seizure or vice versa? Hence, a detailed history was taken and the sequence of events was noted. The child started with feeling uneasy, nauseous (Aura), while he walked toward the door, he started vomiting and in the process, he bent down and knocked the door knob. He had one more episode of vomiting (ictal vomiting) followed by deviation of mouth to right side and stiffening of whole body. The event lasted long, 15–20 min by which time, he arrived at emergency and was given injection midazolam and transferred to the ward. The development history, birth history, and family history were insignificant. Hence, a routine awake and sleep video electroencephalogram (EEG) was done.

EEG showed a normal background with a posterior dominant rhythm (PDR) of 9 Hz, reactive to eye opening. However, frequent paroxysms of bilateral occipital spike-wave discharges (Fig. 1), which accentuated in sleep, were seen. A characteristic finding of "fixation-off sensitivity" (occipital spikes disappeared on eye opening and reappeared on eye closure) was also seen (Fig. 2). Sleep

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features were present bilaterally and synchronously and activation procedures such as hyperventilation and photic stimulation did not add any additional information. Therefore, the clinical presentation of autonomic symptoms with head version and progressing to tonic-clonic seizures and characteristic electrographic findings helped us to finally diagnose the case as PS which was initially thought to be a case of head injury. Parents were counseled about the benign nature of this age-dependent childhood epilepsy syndrome. However, parents were anxious about recurrence and opted for antiepileptic therapy. The child was started on oxcarbazepine and discharged. He had two breakthrough seizures after 2–3 months when his antiepileptic drug (AED) dose was adjusted, and thereafter, he has been seizure free for 1 ½ years and is doing well.

DISCUSSION

In 1989, Panayiotopoulos described an age-related epileptic syndrome characterized mainly by ictal vomiting, head and eye deviation, and sometimes prolonged periods of loss of awareness. The episodes usually begin with vomiting which can be protracted, with severe retching often accompanied with pallor, mydriasis, diaphoresis, etc. [2]. Sometimes, it may also evolve into complex partial seizures with deviation of eyes and/or tonic/clonic movements lasting few minutes to sometimes over 30 min. In approximately one-fifth of the seizures, the child becomes unresponsive and flaccid (ictal syncope) before or often without convulsions. At least five of the following criteria need to be present to make a diagnosis of PS: Infrequent seizures, prolonged seizures >5 min,

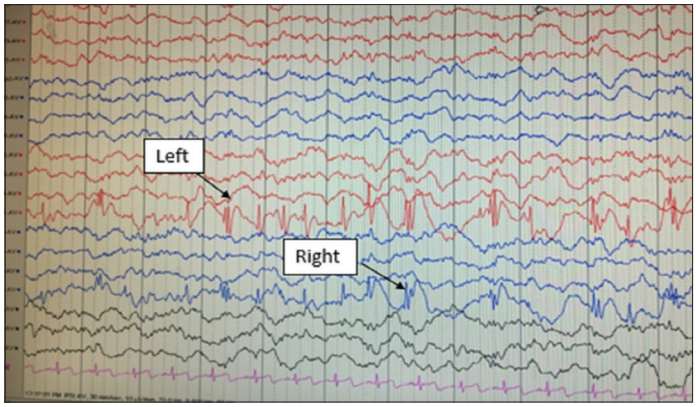

Figure 1: Bilateral occipital spikes and waves

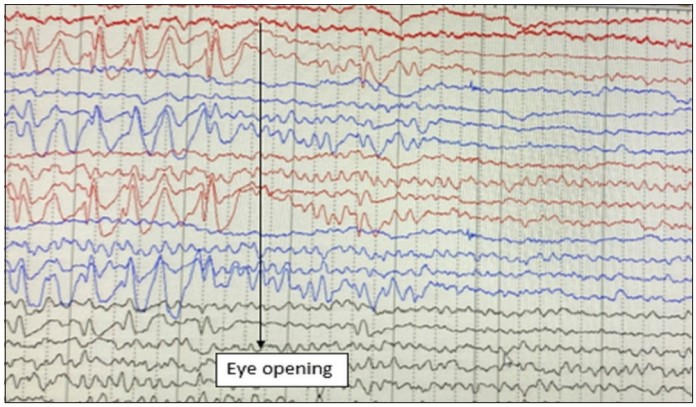

Figure 2: Fixation-off sensitivity

ictal vomiting, eye deviation, autonomic manifestations, behavioral disturbance, and altered consciousness [5]. As the predominant clinical feature of PS is emesis, it can be confused with other non-epileptic disorders with emesis as primary symptom. Thus, the close differentials of this syndrome are syncope, migraine, cyclic vomiting syndrome, motion sickness, sleep disorder, or gastroenteritis [6]. Hence, detailed history, in terms of onset and semiology, helps in differentiating this epilepsy syndrome.

The EEG is the most useful diagnostic test that shows shifting and/or multiple foci, often with occipital predominance. The characteristic fixation off sensitivity referring to disappearance of these discharges on visual fixation is a curious phenomenology seen in PS as well as in other occipital lobe epilepsies both benign and symptomatic [2]. Rest of the investigations and MRI brain is normal. Children with PS have normal developmental history. As, this is a self-remitting, benign epilepsy syndrome where a child may not have more than 2–3 seizures and sometimes a second seizure may never occur after the first one, prophylactic treatment with an AED may not be needed. Parental education is the cornerstone of management of this condition. However, antiepileptic may be given in special situation depending on the parental comfort level or severity of seizures. First line AEDs are carbamazepine, oxcarbazepine, levetiracetam, or valproate [7]. Prognosis is invariably excellent and usually remits before the age of 16 years [4].

CONCLUSION

As advancement in epileptology has helped us define epilepsy syndromes with distinct electroclinical features, one must have a high index of suspicion for PS in children presenting with autonomic symptoms and unresponsiveness as recognizing it helps in prognostication and correct choice of treatment.

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