Panayiotopoulos Syndrome

MEENAKSHI GIRISH
NILOFER MUJAWAR
VITHALRAO DANDGE

ABSTRACT

Panayiotopoulos syndrome is a benign childhood epileptic illness characterized by episodic autonomic symptoms. We present a 9 year old boy with this condition, who presented with episodes of severe vomiting, fever, deviation of eyes and altered sensorium, characteristic EEG in the form of multiple occipital spikes and normal neuroradiologic and metabolic investigations.

Key words: *Children, Epilepsy, Occipital epilepsy, Panayiotopoulos syndrome.*

INTRODUCTION

Panayiotopoulos syndrome is an idiopathic childhood epileptic syndrome recently recognized by the International League Against Epilepsy. The clinical features are characterized by predominantly autonomic seizures comprising ictal vomiting, eye deviation, thermoregulatory disturbances like fever, loss of consciousness and sometimes convulsions. Awareness of this syndrome is important for several reasons. Though it has never been reported previously from India, it is stated to be a common condition(1) but underdiagnosed and underreported. It is a benign self remitting condition; and needs to be differentiated from other non-epileptic conditions with autonomic symptoms.

From the Department of Pediatrics, NKP Salve Institute of Medical Sciences, Hingna, Nagpur, 440 019, Maharashtra, India.

Correspondence to: Dr. Meenakshi Girish, 101, Shubham Enclave, Darda Marg, Rahate Colony, Nagpur, 440 022, Maharashtra, India. E-mail: min_gir@rediffmail.com.

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CASE REPORT

A 9-year-old boy was hospitalized with history of episodic stereotypic symptomatology since the age of 2 years. The frequencies of the episodes were 1-2 per year and in the last 6 months he had 3 such episodes.

The episodes would begin with sudden severe vomiting accompanied by moderate to high grade fever. The vomiting was occasionally severe enough to warrant parenteral fluid resuscitation. Sometime after the onset of vomiting the patient would develop deviation of eyes to one side lasting for approximately an hour, followed by loss of consciousness of variable duration, lasting at the minimum an hour and once extending 24 hrs. Vomiting would persist even during the period of altered consciousness. The termination of the episode would be abrupt, though drowsiness would persist for some more time. The constellation of symptoms – vomiting, fever, deviation of eyes, and loss of consciousness is seen in early onset benign occipital epilepsy, also known as Panayiotopoulos syndrome. Lending credence to this diagnosis, in the last episode, transient loss of vision and severe headache accompanied the vomiting. Investigations including blood sugar, serum electrolytes, ionic calcium, serum magnesium, serum ammonia and serum creatinine were all normal. CT scan was also normal. EEG revealed characteristic occipital spikes which disappeared on eye opening. The child has been seizure free on carbamazepine for the last one year.

DISCUSSION

Benign occipital epilepsy of childhood, characterized by occipital paroxysms on EEG was first described by Gastaut(2). It is characterized by seizures that start with visual symptoms, which often are followed by hemiclonic seizures or automatisms and, in some cases, migrainous headaches. The EEG findings include paroxysms of rhythmic occipital and posterior temporal spikes when the eyes are closed. In 1989, Panayiotopoulos described another epileptic syndrome with occipital spikes. It is an early-onset syndrome characterized mainly by ictal vomiting, head and eye deviation, and sometimes prolonged periods of loss of awareness. The

syndrome was later incorporated into the ILAE as early-onset childhood epilepsy with occipital spikes (Panayiotopoulos type). Panayiotopoulos syndrome was found to be much more common compared to the first recognized one, which is now known as late-onset childhood epilepsy with occipital spikes (Gastaut type). The clinical hallmark of this syndrome is the presence of autonomic features like vomiting.

According to a recent consensus conference(4), Panayiotopoulos syndrome is defined as a benign age-related autonomic epileptic disorder occurring in early and mid childhood with an EEG that shows shifting and/or multiple foci, often with occipital predominance**.** Atleast 5 of the following criteria need to be present to make a diagnosis of Panayiotopoulos syndrome: infrequent seizures, prolonged seizures >5 minutes, ictal vomiting, eye deviation, autonomic manifestations, behavioral disturbance and altered consciousness.

Panayiotopoulos syndrome supposedly affects upto 13% of children aged 3-6years (6% aged 1-15years) with a history of nonfebrile seizures(3). Emesis is the predominant manifestation of autonomic seizures. Other autonomic symptoms include pallor, mydriasis, urinary or fecal incontinence, thermoregulatory alterations and hypersalivation. Behavior distur-bances, headache or other nonpainful cephalic sensations are often found at the onset. In atleast a fifth of the seizures, the child becomes pale and unresponsive (ictal syncope) either before convuls-ing or in isolation. The eyes often deviate to one side (in 60%) or the patient may stare. Half of the seizures end with hemi- or generalized convulsions.

Panayiotopoulos syndrome may easily be missed or confused with many nonepileptic conditions. Mild ictal symptoms in the presence of clear consciousness would suggest trivial non-epileptic conditions such as atypical migraine, gastroenteritis or syncope, while prolonged and severe attacks may simulate life threatening insults such as encephalitis, for which many of these children are treated. Characteristically, even after the most severe symptoms the child becomes completely normal after a brief post ictal sleep. This is considered both diagnostic and reassuring.

Prognosis is invariably excellent except for the rare symptomatic cases. Only 5-10% have more than 10 seizures but the outcome is still favorable. Lengthy seizures do not appear to result in residual deficits or have adverse prognostic significance. One fifth of children with this syndrome may develop other types of infrequent seizures, usually rolandic, but these also remit before the age of 16 years. Control is easily achieved with antiepileptic drugs (especially carbamazepine) but it is unnecessary unless symptoms appear life threatening and cause parental anxiety or the seizures are very frequent.

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REFERENCES

1. Koutroumanidis M. Panayiotopoulos Syndrome. BMJ 2002; 324: 1228-1229.
2. Kaddurah Ahmad K, Bhagwan M. Benign Childhood Epilepsy. Available from URL: http://www.emedicine.com/neuro/topic641.htm# section ~Benign Partial Epilepsy of Childhood. Accessed on January 16, 2007.
3. Couvanis A. Panayiotopoulos: A benign autonomic epilepsy frequently imitating encephalitis, syncope, migraine, sleep disorder or gastroenteritis. AAP Grand Rounds 2006; 16: 69-70.
4. Ferrie C, Caraballo R, Covanis A, Demirbilek V, Dervent A, Kivity S, *et al.* Panayiotopoulos Syndrome: a consensus view. Dev Med Child Neurol 2006; 48: 236-240.