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Case report

BENIGN OCCIPITAL EPILEPSY OF CHILDHOOD: PANAYIOTOPOULOS SYNDROME IN A 3 YEAR OLD CHILD

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ABSTRACT

Panayiotopoulos syndrome (PS) is a relatively frequent and benign epileptic syndrome seen in children in the age group of 3-6 years and is characterised by predominantly autonomic symptoms and/or simple motor focal seizures followed or not by impairment of consciousness. Although multifocal spikes with high amplitude sharp-slow wave complexes at various locations can be present in the EEG, interictal electroencephalogram (EEG) in children with this particular type of epilepsy characteristically shows occipital spikes. This syndrome has known to be a masquerader and can imitate gastroenteritis, encephalitis, syncope, migraine, sleep disorders or metabolic diseases. In the absence of thorough knowledge of types of benign epilepsy syndromes and their various clinical presentations, epilepsy such as PS can be easily missed. The peculiar aspects of this type of epilepsy in children should be known not only by paediatricians but also by general doctors because a correct diagnosis would avoid aggressive interventions and concerns on account of its benign outcome. In this case study, we report a case of PS in a 3 year old child.

Keywords: Benign occipital epilepsy, Panayiotopoulos syndrome, Autonomic symptoms, Emesis, EEG

INTRODUCTION

The International League Against Epilepsy in their expert consensus has given due importance for the various benign childhood seizures which have good prognosis.[1] PS is a common idiopathic childhood-specific seizure disorder formally recognized by the league and is included in the category of benign epilepsy syndromes and is recognized worldwide for its autonomic presentations.[2,3] This early-onset benign childhood seizures was described by Panayiotopoulos.[4] It has been defined by Panayiotopoulos as consisting of brief, infrequent attacks or prolonged status epilepticus and characterized by ictal deviation of the eyes and/or head and vomiting, occurring in children usually between the ages of 3 and 7 years. [5]

Seizures are usually followed by postictal headache and are often associated with interictal occipital rhythmic paroxysmal EEG activity that appears only after eye closure.[5]The PS has excellent prognosis and parents can be definitely reassured about its benign course.[4,6-9] The risk of developing seizure disorder in later life is negligible. [6] Detection of occipital epilepsy at very early stage is needed to successfully treat this condition and allay the fears of the parents and care givers of these children with PS. In this case report, we discuss about the occipital epilepsy in a 3 year old girl child.

CASE REPORT

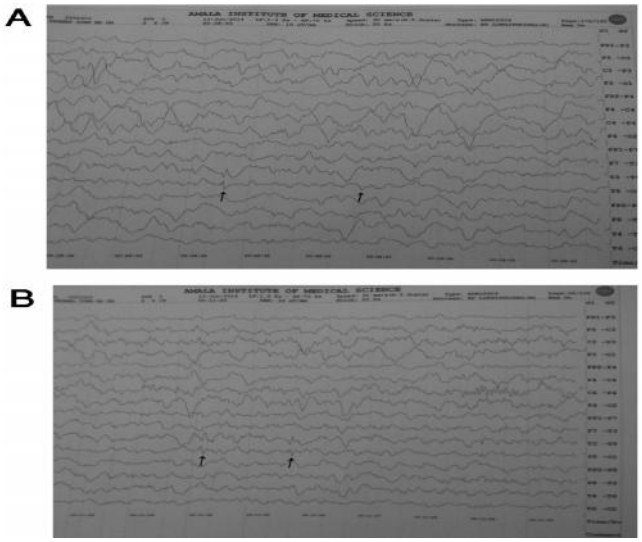
A 3-year-old girl, only sibling from a poor socioeconomic family of a non-consanguineous couple, presented in the out-patient with complaints of becoming limp after sudden episode of vomiting, followed by up rolling of eyes, stiffening of the both upper limbs and lower limbs and a brief period of drowsiness. The child was happily playing in the house about half an hour back. There was no associated fever, trauma, ear discharge, no common paediatric illnesses like diarrhea, dysuria, cough, running nose, wheezing and throat pain.

A detailed history was taken. The child was born of non-consanguineous parents, full term normal vaginal delivery, with a birth weight of 2.215 kg. She was immunized to date and had normal milestones of development. The history revealed that she had similar episodes of vomiting especially getting up from sleep and having deviation of eyes to one side, becoming limp and followed by drowsiness for few minutes in the past from the age of 1 ½ years old. Overall she had 5-6 such episodes and 3 times she had these episodes when she was sleeping. There was no associated fever during these episodes. Two times she had stiffening of all the limbs with deviation of eyes to one side, and followed by drowsiness. There was no focal type of seizures in this child. The parents attributed these to indigestion and gave home remedies as always there was vomiting and tiredness following the episodes. The child then used to play around normally. One month back the child was seen by a local doctor who advised EEG and it was done which was reported as normal and parents were advised follow up.

The child on admission was tired, but was conscious. On examination, she was afebrile, signs of meningeal irritation were absent, central nervous system examination was normal, neurocutaneous markers were absent, fundus examination was normal. Other systemic examinations were normal. Laboratory investigations showed hemoglobin (11.7 g/dl) with low indices, total leucocyte count (11,550/cumm), neutrophils (75%), lymphocytes (22%), platelets (210000/µl), ESR (35mm at1 hr), serum calcium (10 mg%), SGPT (28 U/l), serum electrolytes levels were normal.EEG was done (Figure- 1A and B) and reported as symmetrically distributed normal sleep activities, with activation of rare sharp wave discharges arising from the left occipital region. An awake record could not be obtained. The diagnosis of PS was made based on the clinical history and EEG which showed the predominantly occipital spikes. (Fig 1) She was started on carbamazepine with increasing the dose schedule to her required weight. The child did not have any allergic reaction to the drug and did not progress to autonomic instability. She and her parents were given excellent emotional and pschycological supportive care.

After completion of 5 days of observation for her symptoms and any allergy to the she was discharged on day 6 with improvement in clinical conditions on multivitamins, hematinics and deworming drugs with an advice to follow-up .

Fig1: A and B): EEG of the child showing the occipital spikes (arrow heads).



DISCUSSION

PS described by Panayiotopoulos[4] is a common autonomic childhood epileptic syndrome with a significant clinical, pathophysiological characteristics and is multifocal.[10] PS is now formally recognized as a distinct clinical entity within the spectrum of benign focal epilepsies of childhood.[11] PS affects 13% of children aged 3 to 6 years who have had 1 or more afebrile seizures and 6% of such children are in the 1- to 15-year age group.[6,7,12] Autonomic epileptic seizures and autonomic status epilepticus are the cardinal manifestations of Panayiotopoulos syndrome.[12]. The main aspect of PS is that irrespective of their location at onset, there is activation of autonomic disturbances and emesis, to

which children are particularly vulnerable. These symptoms and pattern of autonomic seizures and autonomic status epilepticus in PS do not occur in adults and are very specific to childhood. [12]

PS is often confused with occipital epilepsy and acute non-epileptic disorders such as encephalitis, syncope, cyclic vomiting or atypical migraine even with characteristic clinical and EEG manifestations.[13]The clinical and EEG features of PS is due to a maturation-related diffuse cortical hyperexcitability. [4,6] This diffuse epileptogenicity which may be unequally distributed, is predominating in one area of the brain, and is often posterior. The explanation for the characteristic involvement of emetic and the autonomic systems may be attributed to epileptic discharges which are generated at various cortical locations and this in turn influence the children's vulnerable emetic centers and the hypothalamus.[4,6]The diagnosis is based entirely on clinical presentation and EEG.[12]

PS has some of the key clinical features which are often present as single, focal seizures with an unusual constellation of autonomic, mainly emetic, symptoms, associated behavioral changes, and sometimes seizure like clinical manifestations such as unilateral deviation of the eyes and convulsions. [3,4,7,8,9,13] The emetic triad in PS (nausea, retching, vomiting) culminates in vomiting in 74% of the seizures; in others, only nausea or retching occurs, and in a few, vomiting may not be present. Other autonomic manifestations include pallor, mydriasis or miosis, flushing or cyanosis thermoregulatory and cardiorespiratory alterations. Frequently incontinence of urine and/or feces, hypersalivation, cephalic sensations, and modifications of intestinal motility are also seen.[9] Half of the convulsions end with hemiconvulsions or generalized convulsions. Two thirds occur during sleep as was seen in our child for about three times. Autonomic status epilepticus enveals then. The seizures usually last for 5–15 min, but half of them are prolonged, sometimes for hours, constituting autonomic status epilepticus. The patient recovers within a few hours. Even after the most severe seizures episodes and status.[12]

An electroencephalogram is the only investigation with abnormal results, usually showing multiple spikes in various brains locations.[12] Multifocal spikes that predominate in the posterior regions characterize the EEG. [6]The EEG variability in our child of 3 years is showing the characteristic occipital spikes from the left occipital region. The EEG done 5 months back was normal in our child. PS is the second most frequent benign syndrome of childhood after rolandic epilepsy, which primarily affects 15% of children at a peak onset at age 7–9 years. [1] Another epileptic syndrome categorized with PS and rolandic epilepsy is the Gastaut type childhood occipital epilepsy[2], manifesting with frequent and brief visual seizures. However, this is rare, of uncertain prognosis, and markedly different from PS, despite common interictal EEG manifestations of occipitalspikes.[6] Occipital spikes in non-epileptic children with defective vision, occipital slow spike-and-wave found in some patients with the Lennox-Gastaut syndrome, focal epilepsy due to occipital lesions, seizures originating in the temporal lobe secondary to an occipital abnormality, and complicated or basilar migraine must be considered in the differential diagnosis.[5]

There are typical and atypical case of PS.[15,17,18] Lada et al [15] conducted a retrospective study of 43 patients with PS who were seizure free >2 years. In their analysis girls predominated, as in our child was a girl. The first seizure was seen in 5 years of age. 86% had emesis as the symptom with the seizures. Seizures during sleep (84%) were more common than those in wakefulness. EEG showed occipital spikes in more than 50% of patients. Prognosis was excellent and 80% children have been free of seizures for ≥ 2 years as is in a typical case of PS.[15] Değerliyurt et al[16] did a case series study of patients with PS and postulated that PS is associated with high rates of febrile convulsions. Afebrile convulsions/epilepsy, migraine, and breath-holding spells in the patients and families suggested the importance of genetic factors [17]. Febrile seizures are to be considered in the differential diagnosis because the recovery of consciousness from seizure is fast and control of the seizure is paramount, uncomplicated usually.[18] Ferrieet al. [17] postulated an atypical evolution of PS in a case report.

The management of PS is not complicated. Education and knowledge about PS is the cornerstone of management. Control of the seizure is paramount. Prophylactic treatment with antiepileptic medication may not be needed for most patients. The emphasis is

on treatment of possible fever and mainly of the underlying illness. One third (30%) of the seizures are relatively brief and self-limited. They subside spontaneously within 2–10min. The other two thirds (70%) have long-lasting seizures (>10 min) or status epilepticus (>30 min to hours). These should be appropriately and vigorously treated as for status epilepticus.[19,20] Parents of children with recurrent seizures should be advised to place the child on its side or stomach on a protected surface and administer a preparation of intravenous rectal benzodiazepine (BZD). In an emergency facility, the child's airway should be kept clear, oxygenation maintained, and intravenous or rectal antiepileptic drug (AED) given to halt the seizures. A BZD is probably the first choice. The great majority with PS do not need AED treatment even if they have lengthy seizures or have more than two recurrences. There is no increased risk of subsequent epilepsy or neurologic deficit. If a child has multiple recurrences (only about 5% exceed 10 seizures) and if the parents too worried prophylaxis can be given. Continuous prophylaxis consists of daily medication with any AED with proven efficacy in partial seizures. Although, there is no evidence of superiority among monotherapy with phenobarbitone, carbamazepine (CBZ), sodium valproate or no treatment in PS, most authors prefer CBZ. [14] Our child was started on oxcarbazepine, a structural derivative of CBZ with no side effects since last 1 month. Autonomic status epilepticus in the acute stage needs thorough evaluation; aggressive treatment may cause iatrogenic complications including cardiorespiratory arrest.[12] The adverse reactions of the antiepileptic drugs such as severe allergic reactions, abnormal liver function tests and idiosyncratic reaction should be kept in mind and monitored.[14]

The prognosis of PS is excellent [4,6-9] The lengthy seizures and status do not have any adverse prognostic significance, and the risk of developing epilepsy in adult life is probably no more than that of the general population.[6] One third of patients (27%) have a single seizure only, and another half (47%) have two to five seizures. Only 5% have >10 seizures, but outcome is again favorable. Remission usually occurs within 1 to 2 years from onset.[6]

CONCLUSION

PS is a common cause of epilepsy in children and a knowledgeable doctor does not miss it. Physician education of PS and recent guidelines on epilepsy management is vital in detecting PS at very early stage, so further lifesaving interventions can be done and prevent delay in the treatment administration. Multiple antiepileptic drugs use is required in only in a small proportion of patients. Seizures in PS, like febrile convulsions, despite their excellent prognosis, are a frightening experience for the in experienced parents, who often think that their child is dead or dying. Parents of young children should have general information by the family doctor regarding PS. Parental education and a supportive group comprising the paediatrician, neurologist, nursing staff and the social worker can help and reassure these distort parents as was done in our child who is doing fine with no recurrence in the last 1 month.

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