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Case Reports

EARLY ONSET BENIGN OCCIPITAL EPILEPSY (PANAYIOTOPOULOS SYNDROME): REPORT OF A CASE

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ABSTRACT

Seizure disorders are the most common neurological illnesses in infants and children.

Presented is an 8 year old boy with nocturnal vomiting episodes, found to have EEG characteristics of early onset benign occipital epilepsy, better known as Panayiotopoulos syndrome.

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Keywords: Panayiotopoulos Syndrome (PS), Occipital Epilepsy (OE), Benign Cortical Epilepsy.

INTRODUCTION

Seizure disorders are the most common neurological problems in infants and children. "Benign", or idiopathic (genetic) epilepsies prevail in this age group. Of benign *partial* epilepsies, benign partial epilepsy with centro-temporal spikes (Rolandic Epilepsy) is the most common, followed by benign occipital epilepsy (OE). The latter has been divided into 3 groups, based on age of onset, clinical and EEG characteristics[1] (see the Discussion).

An 8 year old boy with clinical and EEG features of "early onset" benign occipital epilepsy, better known as Panayiotopoulos Syndrome (PS), is presented.

CASE REPORT

In June 2004, M.J., an 8 year old Iranian boy presented to the author's private clinic with a chief complaint of 3 annual episodes of vomiting at sleep since 3 years ago. For each episode, he sat up in the bed in the middle of the night, eyes deviated to one side with pallor and some lip cyanosis, minimal hand movement, and he vomited. He was unresponsive, or "minimally responsive" during the episodes; no generalized body spasm, urinary or fecal incontinence, or post-ictal headache occured. He was the first twin of an uncomplicated pregnancy and delivery. Past history was unremarkable; one cousin had a history of seizure disorder. No history of migraine headache in the family. No history of photosensitivity. General and neurological examination was normal. An awake-to-sleep electroencephalogram (EEG), (Neuroscan LT2001, USA) done after a brief period of sleep deprivation revealed (Figure 1): a) normal background activity, and reactivity to eye opening and closure, 2) right -sided, medium voltage, 1-2 Hz sharp & slow waves at right occipital area seen at photic stimulation, and when the patient went to sleep,when it continued as a train of activity. His brain magnetic resonance imaging (MRI) proved normal, and he was sent home, reassured, with a diagnosis of early-onset benign occipital epilepsy (Panayiotopoulos Syndrome, PS). So far, he has remained asymptomatic.

DISCUSSION

3 variants of benign occipital epilepsy (OC) with occipital spikes have been described:[1]

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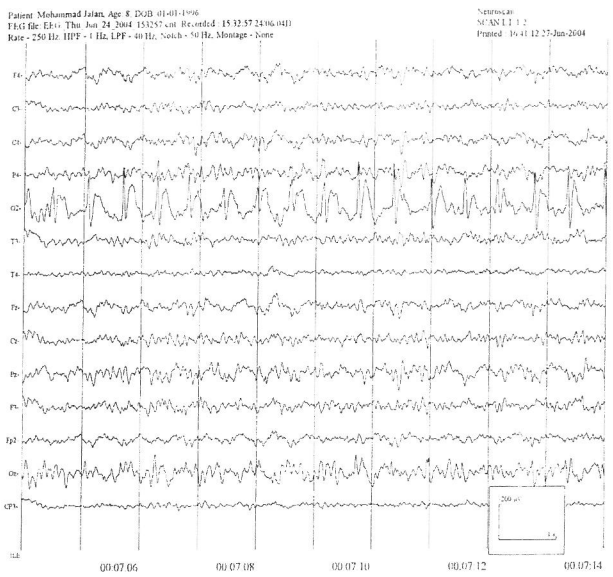


Fig.1. EEG finding.

1. Late-onset , "Gastaut type": mean age of onset of 6 years, positive family history of epilepsy or migraine headaches, visual phenomena (transient amaurosis, elementary or formed visual hallucinations,visual illusions). The seizures occur alone, or are followed by hemisensory, hemiclonic, complex partial, or generalized tonic-clonic seizures. One third of the patients experience severe, long-lasting migraine-like headaches, accompanied by nausea or vomiting. Ictal EEG discharges are characteristically localized to one occipital lobe, and interictal EEG spike-wave discharges are bilateral or unilateral, occur only with eye closure, and are abolished with eye fixation.[2,3,4]

2. Early-onset OC (Panayiotopoulos syndrome, PS)[14]: It is common (13% of 3-6 year olds & 6% of 1-15 year olds),[10] onset is between 1 to 14 years, average age 4 to 5 years. Episodes occur relatively rarely; up to 30 % of children have only a single episode, with mean number of 3 in the course of the disease. Two-thirds of the episodes are nocturnal, start with feeling of sickness and nausea, most often followed by vomiting, deviation of the eyes, in most patients. Loss, or obtundation of, consciousness may then often be seen, followed in some cases by unilateral clonic convulsions. Some patients may have only vomiting (ictus emeticus). Other atypical symptoms may include: headache at the beginning, pallor, mydriasis, syncope, visual symptoms, speech arrest, and hemiconvulsions. Characteristically, the patient is normal after the attack.[1,10] PS seems to have a benign course. Remission usually occurs within 2 years of onset. A third of these children have only a single attack, and only 5-10% have more than 10 seizure episodes. One-fifth may develop other seizure types, usually Rolandic, but these also remit before the age of 16,[10] Interictally,1 to 3 Hz bilateral, or as in our case, unilateral[11] occipital, often high amplitude spike waves are seen, in clusters, or singly, but more characteristically continuously, with eye closure, more in sleep, and are abolished with eye opening (fixation-off phenomenon).[1] EEG may be normal while awake-as in our case- and a sleep EEG may be required.[10] Ictal EEG shows a mixture of repetitive slow wave and spikes contralateral to the eye deviation.[5,6,7,8,9] Because of infrequent attacks, treatment with anti-epileptic drugs (usually carbamazepine) is usually unnecessary.[10]

3. Idiopathic photosensitive occipital epilepsy ("Guerrini type"): Photic stimulation induces focal occipital seizures, manifesting as often brightly colored visual hallucinations, blurring of vision, or blindness. They may last for several minutes up to several hours, and may be accompanied by headache. Such seizures may be induced by television viewing or video-game playing. Prognosis is favorable, and avoidance of precipitating light and administration of valproate is advised.[12,13]

The symptoms presented, along with the EEG suggest PS, although the EEG lacks the fixation-off phenomenon (occipital paroxysms in the EEG when eyes are closed, or when fixation and central vision are eliminated).[14] He was reassured of the favorable prognosis and was put to regular follow-ups.

CONCLUSION

The case presented is an example of PS, a benign condition in children, the diagnosis of which reassures the parents, and obviates further anti-epileptic treatment.

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