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Idiopathic Childhood Occipital Lobe Epilepsies in Turkish Children

Türk Çocuklarında İdiopatik Çocukluk Çağı Oksipital Lob Epilepsisi

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ABSTRACT

Purpose: Two forms of idiopathic occipital lobe epilepsy can be distinguished: an early onset or Panayiotopoulos type (PS), and a late onset or Gastaut type (GS). The aim of this study was to identify clinical manifestations and outcome in Turkish children with idiopathic occipital lobe epilepsy.

Material and Methods: Thirty nine children (27 boys, 12 girls) were divided into the PS and GS type, according to the classification for epileptic seizures of the International League Against Epilepsy.

Results: Among the 39 patients, 27 (69.3%) were boys and 12 (30.7%) were girls and the mean age at the seizure onset was 77.38 ± 27.33 months (36-145 months). The PS type patients consisted of 27 children (19 boys, 8 girls). The average age of seizure onset in PS type patients was 60.76 ± 14.21 months (range, 36-94 months). The GS type patients consisted of 12 children (8 boys, 4 girls), and seizure onset was 107.7 ± 18.8 months (range, 72-145 months). Ictal vomiting was more common in the PS type patients, and visual symptoms were more common in the GS type patients. We detected that after treatment, 27 patients (100%) in PS type and 10 patients (83.3%) in GS type became seizures-free. Two patients continued having seizures in GS type.

Conclusions: The average age of seizure onset was much younger in the PS than in the GS. Also, in idiopathic occipital lobe epilepsy, the PS type has better seizure-free and prognosis than the GS.

Key words: Idiopathic childhood occipital epilepsies, clinical findings, children.

ÖZET

Amaç: İdiopatik çocukluk çağı oksipital lob epilepsisinin iki farklı tipi tanımlanmıştır: erken başlangıçlı veya Panayiotopoulos tip (PS) ve geç başlangıçlı Gastaut tip (GS). Bu çalışmanın amacı, idiopatik çocukluk çağı oksipital lop epilepsisi tanısı alan Türk çocuklarında klinik bulguları ve seyri ortaya koymaktır.

Materyal ve Metod: Otuz dokuz çocuk (27 erkek, 12 kız), Uluslarası Epilepsi ile Savaş Derneği (İLAE) tarafından önerilen epileptik nöbet sınıflamasına göre PS ve GS tip olarak ayrıldı.

Bulgular: Otuz dokuz hastanın, 27'si (69.3%) erkek ve 12'si (30.7%) kız idi ve ortalama nöbet başlangıç yaşı 77.38 ± 27.33 ay (36-145 ay) idi. Hastaların 27'si (19 erkek, 8 kız) PS tip idi. PS tip hastaların ortalama nöbet başlangıç yaşları 60.76 ± 14.21 ay (36-94 ay) idi. GS tip 12 hastanın (8 erkek, 4 kız) idi ve nöbet başlangıç yaşı 107.7 ± 18.8 ay (72-145 ay) idi. PS tip hastalarda iktal kusma ve GS tip hastalarda vizüel semptomlar daha sıktı. Tedavi sonrası PS tip hastaların 27'si (100%), GS tip hastaların 10'u (83.3%) nöbetsiz duruma gelmişti. GS tip hastalardan ikisinin nöbetleri devam ediyordu.

Sonuç: Nöbet başlangıç yaşı ortalaması PS tipte, GS tipe göre daha erkendir. Ayrıca idiopatik çocukluk çağı oksipital lop epilepsisi olan, PS tipinin prognozu ve nöbetsizlik durumu, GS tipine göre daha iyidir.

Anahtar Kelimeler: İdiopatik çocukluk çağı oksipital lob epilepsisi, klinik bulgular, çocuklar.

INTRODUCTION

Occipital seizures originate from an epileptic focus confined within the occipital lobes.

Two forms of occipital lobe epilepsy can be distinguished: a benign, idiopathic form and a symptomatic form. The benign form was first described by Gastaut as Benign Epilepsy with Occipital Paroxysms (BEOP)[1]. In 2001, the Commission on Classification and Terminology of the International League Against Epilepsy proposed that childhood epilepsy with occipital paroxysms should be classified into two syndromes: early onset (Panayiotopoulos type-PS) and late onset (Gastaut type-GS)[2].

In PS, children present at a mean of 4.7 years (range 1-14 years) with rare seizures which are prolonged and nocturnal. Seizures begin with autonomic features such as vomiting, pallor and sweating followed by tonic eye deviation, impairment of consciousness and may evolve to a hemi-clonic or generalized convulsion. Autonomic status epilepticus may occur. Prognosis is excellent and treatment often unnecessary[3]. Gastaut syndrome presents at a mean of 8 years (range 3-16 years). Seizures are frequent, brief and diurnal. They comprise simple partial seizures characterized by initial visual hallucinations such as phosphenes and/or ictal blindness and illusions; post-ictal headache is common. Impairment of consciousness is rare unless associated with hemi-clonic or generalized convulsions. The prognosis is less predictable, with the remission rate at approximately 50-60% of cases, often within 2-5 years of onset[2,4].

In this study, we evaluated clinical and outcome data derived from our experience of 39 patients with BEOP.

MATERIAL and METHODS

This study was conducted a retrospective study of 39 children diagnosed with BEOP and started on an antiepileptic drug (AED) in the pediatric neurology department of our hospital.

Children were included in the idiopathic occipital lobe epilepsy group when no abnormalities of neurologic examination, psychomotor/mental status, neural development, and neuroimaging studies were present. The patients can be further divided into two subgroups, that is, early-onset group and late-onset group according to clinical manifestations.

The clinical data analyzed in each patient involved sex, family history of epilepsy, history of febrile seizure, consanguinity, age at seizure onset, semiology of seizures, frequency of seizures, interictal electroencephalogram findings obtained while awake and asleep, AED treatment, duration of starting first treatment, and seizure outcome.

Antiepileptic treatment was discontinued who remained seizure-free during at least two years of follow-up.

RESULTS

A total of 39 patients, 27 (69.3%) boys and 12 (30.7%) girls with a mean age of 88.6 ± 32.76 months (45-163 months) at time of evaluation, were studied. The mean age of onset of seizures was 77.38 ± 27.33 months (36-145 months).

Of these 39 patients, 27 (69.2%) (19 boys, 8 girls) satisfied the criteria for PS type; the average age for onset of seizures was 60.76 ± 14.21 months (range, 36-94 months). The remaining 12 (30.8%) patients (8 boys, 4 girls) met the criteria for GS type; the average age for onset of seizures was 107.7 ± 18.8 months (range, 72-145 months).
All 27 children with PS except four (14.8%) had seizures during sleep. The most common ictal symptoms in PS type were impairment of consciousness (100%), and ictal vomit (85.1%). The other common ictal manifestations included lateral gaze deviation in 19 (70.3%), pallor in 17 (62.9%), ictal nausea in 10 (37.1%), headache in 3

(11.1%), hemiconvulsions in 7 (25.9%), secondary generalized seizures in 9 (33.3%), and visual symptoms in 1 (3.7%). The most common ictal symptom in GS type was visual symptoms (100%). This visual symptoms were included elementary visual hallucinations in nine patients (75.0%), blindness or blurring of vision in eight (66.7%), and complex visual hallucinations in 2 (16.7%). Other ictal sypmtoms were headache in three (25.0%), and secondarily generalized tonic-clonic seizures in six (50.0%). In PS type, single seizure was in 4 (14.8%), and two or more seizures were 23 (85.2%), whereas in GS type, all patients had two or more seizures. The demographic and clinical data for the patients are listed in (Table 1).

Interictal electroencephalography revealed normal background activity for all children. All children in both type had epileptiform discharges over occipital or posterior temporal-parietal areas (Figure 1). All patients underwent cranial magnetic resonance imaging normal structural findings.

All patients but one had received treatment with antiepileptic drugs for seizure control. In the PS patients, AED treatment was started in 25/27 patients (92.5%) with single drugs, such as carbamazepine (15 patients 60.0%), valproic acid (8 patients 32.0%), or levetiracetam (2 patients 8.0%).

In the GS patients, 7 of 12 patients (58.3%) were treated with a single drug, and 5 (41.7%) with two ore more drugs (carbamazepine, valproic acid, levetiracetam). In monotheraphy most using AEDs were carbamazepine (71.4%) and valproic acid (28.6%).

After treatment, 27 patients (100%) in PS type and 10 patients (83.3%) in GS type became seizures-free. Two patients continued having seizures in GS type.

Table 1. Clinical features of PS type and GS type of occipital lobe epilepsy

Parameters	**Panayiotopoulos type**	**Gastaut type**
Age at onset (mean ± S.D. months)	60.76 ± 14.21 months	107.7 ± 18.8 months
Sex (male/female)	19/8	8/4
Febrile seizure (yes/no)	5/22	1/11
Family of epilepsy (yes/no)	5/22	2/10
Consanquinity (yes/no)	4/23	5/7
Seizures		
Nocturnal seizure	23/27	0/12
Single seizure	4/27	0/12
Two or more seizures	23/27	12/12
Ictal manifestations		
Impairment of consciousness	27/27	8/12
Ictal vomit	23/27	0/12
Ictal pallor	17/27	0/12
Ictal nause	10/27	0/12
Lateral gaze deviation	19/27	0/12
Headache	3/27	5/12
Visual symptoms	1/27	12/2
Treatment		
One AED	25/27	7/12
Two AEDs	2/27	5/12
Discontinuation of AED	15/27	3/12
Prognosis		
Seizure-free	27/27	10/12

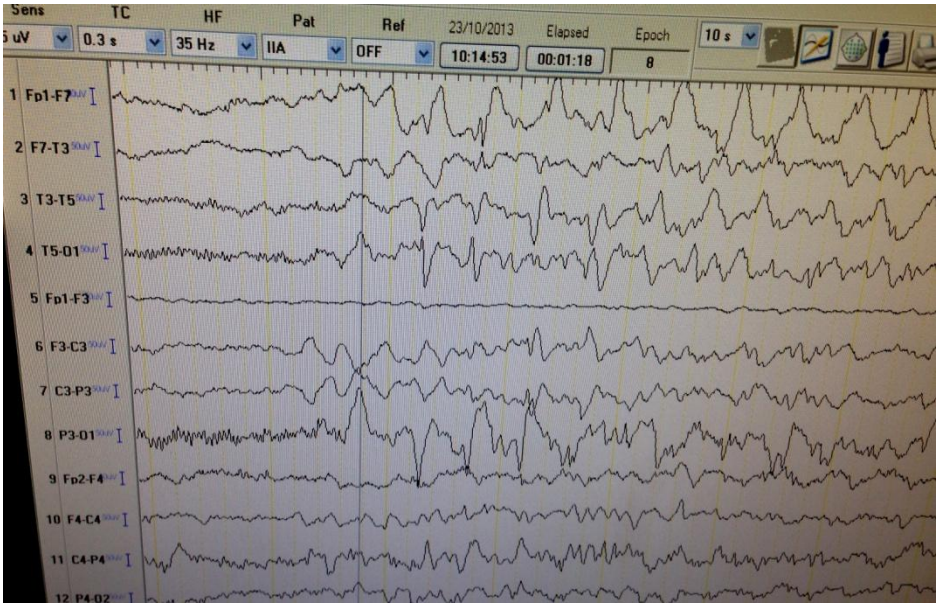

Figure 1. Epileptiform discharges on EEG.

DISCUSSION

Prevalence of PS may be high, probably affecting 13% of children 3-6 years old with one or more non-febrile seizures and 6% of the age group from 1 to 15 years[5,6]. The childhood epilepsy with occipital paroxysms described by GS is a rare syndrome (2-7% of benign partial epilepsies)[1]. The prognosis of the PS type is invariably excellent, with remission usually occurring within 1 year of onset, and in some children treatment might not be necessary. The prognosis of GS type is not as predictable as that of rolandic epilepsy and PS. Although there is a good response to treatment with carbamazepine, 30-40% of patients continue to have seizures[4].

In both types, interictal EEGs show high-voltage repetitive spike/sharp and slow waves, suppressed by eye opening in about 80% of the affected children[1]. Gokçay et al.[7] reported male dominance in BEOP cases, whereas Du et al.[8] noted female to dominance. We found male dominance in our both groups patients with CEOP. In patients with BEOP, age of onset of epilepsy varies from 15 months to 17 years[9]. In our patients age of onset of seizures ranged from 6.5 years (3-12 years).

The distinction of PS and GS types of occipital lobe epilepsy can be made easily, because they have entirely different manifestations[2]. In the present study, the most obvious difference in ictal symptoms between the PS type and GS type. The most common ictal symptom in PS were ictal vomiting, impairment of consciousness, and lateral gaze deviation whereas ictal visuel symptoms were seen common manifestation in GS type in our study.

Ictal vomiting was detected common autonomic manifestations in PS. Other autonomic manifestations, such as pallor, nausea, and retching are also characteristic of the syndrome[5,10,11]. These autonomic signs and symptoms were not recognized before, because physicians failed to evaluate the features in detail, and parents reported only the most remarkable signs, such as motor manifestations. Deviation of

the eyes and head, impairment of consciousness, secondary generalization of the seizures are in agreement with all other authors. Caraballo et al.[12] reported ictal manifestation as pallor was 93.7%, ictal vomiting was 82.3%, ictal nause was in 20.8%, incontinence of urine was in 2.5%, and syncope-like symptoms was in 1% patients in their study. We found impairment of consciousness in 27 (100%) patients, ictal vomit in 23 (85.1%) patients, and lateral gaze deviation in 19 (70.3%) patients in PS type.

Panayiotopoulos found a more common type of childhood epilepsy with occipital paroxysms with the same occipital lobe epilepsy findings in younger children, a type in which seizures are infrequent, usually nocturnal, and marked by ictal vomiting as well as tonic eye deviation. Visual symptoms rarely occurred in these children[3,5,13]. We found frequent nocturnal seizures in the majority of our patients and one patient (3.7%) only had seizures on awaking. In literature, 10-20% of the children may have had seizures when awake only[3,14]. Ictal visual symptoms described as a rare symptom in PS type. These were not initially reported, but were later reported in literature[5]. One patient only presented visual symptoms in our study.

In PS type, age at onset is between 1 and 14 years and at peak 5 years[2,3]. Whereas in GS type, age at onset seizure is 3 to 15 years with a mean around 8 years of age[1]. Caraballo et al reported age at seizure ranged from 1.7 to 12.5 years, with a mean age of 5 in 192 patients with PS[12]. Wakamoto et al.[15] found that age at seizure onset was 10.3 years in 12 patients wit GS type. We found age at seizure onset was 5 years, and 9 years in PS and in GS, respectively. Caraballo et al.[12] and Lada et al.[16] reported a single seizure as 44.2% and 53% in children respectively in PS type. We found this rate as 14.8% in our study.

Compared with the Gastaut type, the Panayiotopoulos type has been reported to include more frequent ictal vomiting, impairment of consciousness, and nocturnal seizure but a lesser total number of seizures, visual symptoms, or postictal headache[1-3]. Similar distinctions were found in the present study.

In following up, we detected all patients in PS type and 10 patients (83.3%) in GS type became seizures-free after treatment. Two patients continued having seizures in GS type. Also, AED treatment was discontinued in 15/27 (55.5%) patients with PS type, and in 3/12 (25.0%) patients with GS type who remained seizure-free during 2 years. Caraballo et al.[12] reported that they discontinued AED in 95 (55.2%) patients with PS in their study.

In conclusion, the present study demonstrated that the main differences of presentation between PS and GS type occipital lobe epilepsy include ictal vomiting, ictal visual symptoms, and treatment response. Panayiotopoulos type occipital lobe epilepsy has a better treatment response than does the GS type.

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