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ÇOCUKLUK ÇAĞI OKSİPİTAL EPİLEPSİ: 19 HASTANIN KLİNİK DEĞERLENDİRİLMESİ

CHILDHOOD OCCIPITAL EPILEPSY: CLINICAL EVALUATION OF 19 PATIENTS

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ÖZ

AMAÇ: Çocukluk çağı oksipital epilepsi (ÇOE) başlıca idiyopatik (İOLE) ve semptomatik oksipital lob epilepsisi (SOLE) olarak ayrılabilir. İOLE, Panayiotopoulos (PS) ve Gastaut sendromundan (GS) oluşmaktadır. Bu çalışmada çocukluk çağı oksipital epilepsisini sınıflandırmak ve klinik özelliklerine göre gruplar arasındaki farklılıkları araştırmak amaçlandı.

GEREÇ VE YÖNTEM: Çocukluk çağı oksipital epilepsi tanılı 19 hasta çalışmaya dahil edildi. Hastaların tıbbi kayıtları değerlendirildi. Demografik verileri, iktal semptomları, nörolojik muayeneleri, elektroensefalografi ve manyetik rezonans görüntüleme(MRG) bulguları, aile öyküleri, febril nöbet ve tedavi yanıtları açısından analiz edildi.

BULGULAR: İdiyopatik oksipital lob epilepsi tanılı 6 hasta ve SOLE tanılı 13 hasta mevcuttu. SOLE ile karşılaştırıldığında, IOLE tanılı tüm olguların nörolojik muayeneleri ve beyin MRG'leri normal saptandı (p=0.044 ve p=0.009). Tüm gruplarda en sık iktal semptom jeneralize nöbetti (%100 IOLE, %75 SOLE). Ancak, IOLE ve PS grubunda en sık izlenen otonomik nöbet iktal kusma idi (2/6, 2/4; %33, %50). Nokturnal nöbetler, SOLE'den daha sık IOLE'de gözlendi (1/13, 3/6; %8, %50). Aile öyküsü (1 olgu, %25) ve febril nöbet (1 olgu, %25) sadece PS'de saptandı. Psikomotor/ mental retardasyon, SOLE'de IOLE'den daha sık saptandı (8/13, 2/6; %62, %33). Tedavi yanıtlarına göre nöbetler tek bir antiepileptik ilaç ile tüm PS tanılı olgularda ve iki antiepileptik ilaç ile tüm GS tanılı olgularda kontrol altına alındı. Diğer taraftan, SOLE tanılı olguların %38'i üç veya daha fazla antiepileptik ilaç ile tedavi edildi.

SONUÇ: Normal nörolojik muayene ve nörogörüntüleme IOLE'ni SOLE'den ayırmada esas özelliklerdir. İktal kusma IOLE ve PS içinde en dikkat çekici otonomik nöbettir. Psikomotor/ mental retardasyon SOLE'de IOLE'den daha sık gözlenmesine rağmen ÇOE tanılı hastalar dikkatlice izlenmelidir. İdiyopatik oksipital lob epilepsisi SOLE'den ve PS de GS'den daha iyi tedavi cevabına sahiptir.

ANAHTAR KELİMELER: Oksipital epilepsi, Çocuk, Panayiotopoulos, Gastaut

ABSTRACT

OBJECTIVE: Childhood occipital epilepsy (COE) can mainly be divided into idiopathic (IOLE) or symptomatic occipital lobe epilepsy (SOLE). Idiopathic occipital lobe epilepsy consists of Panayiotopoulos (PS) and Gastaut syndromes (GS). In this study, we aimed to classify COE and investigate the segregations between the groups according to clinical features.

MATERIAL AND METHODS: Nineteen patients with COE were enrolled. Medical records of the patients were evaluated. Demographic data, ictal symptoms, neurological examination, brain magnetic resonance imaging (MRI) and electroencephalography (EEG) findings, family history, febrile seizure, and treatment response were analyzed.

RESULTS: There were 6 patients diagnosed with idiopathic occipital lobe epilepsy and 13 patients diagnosed with SOLE. Compared to the SOLE, all patients of the IOLE had a normal neurological examination and MRI findings (p=0.044 and p=0.009). The most frequent ictal symptom was generalized seizures in all groups (100% IOLE, 75% SOLE). However, ictal vomiting was the most frequent autonomic seizure in IOLE and PS (2/6, 2/4; 33%, 50%). Nocturnal seizures were observed more frequently in IOLE than SOLE (1/13, 3/6; 8%, 50%). The rate of family history (1 patient, 25%) and febrile seizures (1 patient, 25%) were found in only PS group. Psychomotor/mental retardation was more common in SOLE than IOLE (8/13, 2/6; 62%, 33%). According to treatment outcomes, the seizures were controlled with one antiepileptic drug (AED) in all patients of PS and two AEDs in all patients of GS. On the other hand, 38% of patients in SOLE were treated with three or more AEDs.

CONCLUSIONS: Normal neurologic examination and neuroimaging are substantial features due to discrimination between IOLE and SOLE. Ictal vomiting is a remarkable autonomic seizure in IOLE and PS. Although psychomotor/mental retardation is observed higher in SOLE than IOLE, the patients of COE should be followed up carefully. IOLE has better treatment outcomes than SOLE and PS has better than GS.

KEYWORDS: Occipital epilepsy, Child, Panayiotopoulos, Gastaut

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INTRODUCTION

Occipital lobe epilepsy in children commonly is separated into two forms: idiopathic and symptomatic (SOLE) (1). In 1863, Hughlings Jackson first defined SOLE (2). Neurological deficits and abnormalities in neuroimaging play a key role in distinguishing symptomatic from idiopathic (3). Panayiotopoulos (PS) and Gastaut syndromes (GS) were called as idiopathic occipital lobe epilepsy (IOLE) until recently (4).

However, the International League Against Epilepsy (ILAE) has recently described early-onset benign childhood epilepsy (Panayiotopoulos syndrome) and late-onset childhood occipital epilepsy (Gastaut syndrome) in the self-limited occipital epilepsies of childhood (5).

Early-onset benign childhood occipital epilepsy (Panayiotopoulos syndrome) presents by ictal vomiting, tonic eye deviation with infrequent and nocturnal seizures (4, 6). The seizures commonly start at the age of 1-14 years (mean of 4.7 years) and also may complete with hemiclonic or generalized. Moreover, two-thirds of seizures occur during sleep (6).

Late-onset childhood occipital epilepsy (Gastaut syndrome) exists at the age of 3–16 years (mean of 8 years) (7). The seizures are characterized by brief, numerous and diurnal. They mostly demonstrate visual symptoms like visual hallucinations and sometimes ictal blindness. Impairment of consciousness is infrequent but visual symptoms with eyelid blinking may advance into hemiconvulsions and focal to bilateral tonic-clonic seizures (4, 6, 7).

We aimed to evaluate distinctions between IOLE and SOLE and to classify the idiopathic group as PS and GS according to their clinical manifestations.

MATERIAL AND METHODS

Between December 2017 and May 2019, the electroencephalograms including interictal spikes and/or sharp waves over occipital areas or diffuse but predominantly located in the occipital region in the department of Pediatric Neurology of Afyonkarahisar Health Sciences University Hospital were evaluated.

Total 19 children met the criteria. Medical records of these children were reviewed. Demographic data, ictal symptoms, neurological examination, brain magnetic resonance imaging (MRI) and electroencephalography (EEG) findings, family history and treatment were interpreted.

ETHICAL COMMİTTE

The study was approved by the Ethics Committee of Afyonkarahisar University of Health Sciences (03.01.2020/21).

STATISTICAL ANALYSIS

SPSS for Windows version 21.0 statistical package program was used to analyze the data. In descriptive statistics, variability criterion was given as mean ± standard error. The clinical manifestations were compared using the Mann-Whitney U test, Fisher exact test and $P < 0.05$ was considered to be significant.

RESULTS

Nineteen children with childhood occipital lobe epilepsy were included in the study. Firstly, the children were classified into two groups as idiopathic and symptomatic **(Table 1)**.

Table 1: Clinical and demographic characteristics of idiopathic and symptomatic occipital lobe epilepsy

Characteristics	Idiopathic	Symptomatic	p
Number (female/male)	6(5/1)	13(7/6)	
Age (Mean ±S.D., years)	8.48 ±4.23	5.11 ±4.37	
Neurologic examination, no. (%)			0.044
✓ Normal	6(100)	6(46)	
✓ Abnormal	0	7(54)	
Psychomotor/mental retardation, no. (%)	2(33)	8(62)	
Ictal symptoms, no. (%)			
✓ Generalized seizure	6(100)	9(75)	
✓ Focal seizure	0	3(25)	
✓ Vomiting	2 (33)	0	
Nocturnal seizure, no.(%)	3(50)	1(8)	
Febrile seizure, no.(%)	1(17)	1(8)	
Family history, no.(%)	1(17)	1(8)	
EEG, no. (%)			
✓ Occipital spikes	6(100)	12(92)	
✓ Extraoccipital spikes	0	1(8)	
✓ Abnormal background	0	1(8)	
MRG , no. (%)			0.009
✓ Normal	6(100)	3(27)	
✓ Abnormal	0	8(73)	
Tratment, no. (%)			
✓ One AED	4(67)	6(46)	
✓ Two AEDs	2(33)	2(15)	
✓ Three or more AEDs	0	5(38)	
Duration of seizure –free (mean ± S.D., months)	12.16±10.59	13.46±20.43	

Then, the IOLE was separated into two types: Panayiotopoulos and Gastaut **(Table 2)**. The idiopathic group consisted of 6 children (1 boy, 5 girls) and the symptomatic group consisted of 13 children (6 boys, 7 girls). Follow-up duration ranged 3 - 17 months. The etiology of patients with symptomatic occipital lobe epilepsy was revealed in **(Table 3)**. The mean age in the symptomatic group was earlier than in the idiopathic group (5.11 ±4.37 years vs 8.48 ±4.23 years).

Table 2: Clinical and demographic characteristics of Panayiotopoulos and Gastaut syndromes

Characteristics	Panayiotopoulos type	Gastaut type
Number (female/male)	4(3/1)	2(2/0)
Age (Mean ±S.D., years)	8.01±5.38	9.41±0.35
Neurologic examination, no. (%)		
✓ Normal	4(100)	2(100)
✓ Abnormal	0	0
Psychomotor/mental retardation, no. (%)	1(25)	1(50)
Ictal symptoms, no. (%)		
✓ Generalized seizure	4(100)	2(100)
✓ Focal seizure	0	0
✓ Vomiting	2(50)	0
Nocturnal seizure, no.(%)	2(50)	1(50)
Febrile seizure, no.(%)	1(25)	0(0)
Family history, no.(%)	1(25)	0(0)
EEG, no. (%)		
✓ Occipital spikes	4(100)	2(100)
✓ Extraoccipital spikes	0	0
✓ Abnormal background	0	0
MRG , no. (%)		
✓ Normal	4(100)	2(100)
✓ Abnormal	0	0
Tratment, no. (%)		
✓ One AED	4(100)	0
✓ Two AEDs	0	2(100)
✓ Three or more AEDs	0	0
Duration of seizure –free (mean ± S.D., months)	12.00±9.09	12.50±17.67

Table 3: The etiology of patients with symptomatic occipital lobe epilepsy

4 patients with mental retardation
4 patients with cerebral palsy
2 patients with hydrocephalus and ventriculo with partoneal shunt
1 patient with neurogenic disease and neuromotor developmental retardation
1 patient with corpus callosum hypoplasia
1 patient with wide arachnoid cyst

However, there was no significant difference between the two groups. Neurological examination was normal in all idiopathic group but 7 patients (54%) had an abnormal neurological examination in the symptomatic (p=0.044). Psychomotor/mental retardation was observed in only two patients (33%) in the idiopathic group and 8 patients (62%) in the symptomatic group.

In the idiopathic group, 3 patients (50%) had nocturnal seizures but only one patient (8%) had in the symptomatic group. Family history and febrile seizures were reported in one patient in both groups. The most common ictal symptom in both groups was generalized seizures. While focal seizures were observed in 3 patients (25%) with the symptomatic group, ictal vomiting was noticed in two patients (33%) with the idiopathic group. Interictal EEG showed occipital spikes for all children in the idiopathic group and 12 patients (92%) in the symptomatic group.

However, EEG revealed extraoccipital spikes in one patient (8%) and abnormal background in one patient (8%) with the symptomatic group.

In the idiopathic group, all patients demonstrated normal brain MRI findings, but 8 patients (73%) showed abnormal findings (hydrocephalus, hypoxic-ischemic encephalopathy, polymicrogyria, arachnoid cyst, cerebral atrophy, hypoplasia of the corpus callosum, mega cisterna magna) in the symptomatic group (p=0.009).

In the idiopathic group, 4 patients (67%) were treated with one antiepileptic drug (AED) as levetiracetam/valproate and two patients (33%) with two AEDs (levetiracetam, valproate, carbamazepine, lamotrigine). Additionally, 6 patients (46%) were treated with one AED (levetiracetam, valproate, lamotrigine), two patients (15%) with two AEDs (levetiracetam, clonazepam, phenobarbital), and 5 patients (38%) with three or more AEDs (phenytoin, levetiracetam, clobazam, topiramate, vigabatrin, valproate, carbamazepine, clonazepam) in SOLE.

The average duration of seizure-free was 12.16±10.59 months in IOLE and 13.46±20.43 months in SOLE. However, the average duration of seizure-free was 12.00±9.09 months in PS and 12.50±17.67 months in GS. It was not showed any statistically significant difference between the two groups.

DISCUSSION

The IOLE distinguishes from SOLE with a normal neurologic examination, absence psychomotor/mental retardation, and normal neuroimaging. However, certain outcomes of IOLE are dilemma (3). Polat et al. reported significant problems in the areas of visuomotor coordination, memory and attention with the patients of idiopathic childhood occipital epilepsy especially the late-onset form and also lower performance IQ with WISC-R (Wechsler Intelligence Scale for Children-revised edition) and significantly abnormal BVMG (Bender Visual Motor Gestalt) test with the patients of idiopathic and symptomatic groups (4). Moreover, our study showed a higher rate (62%) of psychomotor/mental retardation in the SOLE. On the other hand, 33% (two patients) of the IOLE were given educational support. These findings indicate the academic performance should follow up attentively in both groups.

As expected, compared with the SOLE, all the patients with the IOLE had normal neurologic examination and neuroimaging, respectively (p=0.044, p=0.009). PS includes predominantly nocturnal seizures (7). Similarly, we indicated more frequent nocturnal seizures in patients with PS in this study (two patients, 50%). Du et al. and various studies pointed out versive movements were more common than ictal vo-

miting into ictal symptoms however we determined ictal vomiting was more frequent in the present study (3, 8, 9). Tata et al. reported that a remarkable frequency of autonomic seizures (ictal nausea, vomiting, retching, pallor, abdominal pain, and diarrhea) in PS (10). Although the neuroanatomical and neurophysiological roots of autonomic features are undetermined in PS, it has been suggested that autonomic features including nausea and vomiting may be responsible for a maturation-related susceptibility of the central autonomic networks (11).

In the present study, we only determined ictal vomiting in patients with PS. Moreover, we did not find any other ictal symptoms (versive movements, vacant spells, visual symptoms). It can be explained the limited number of patients and the semiology overlooking by parents and people who witnessed the seizure for the first time.

Symptomatic occipital lobe epilepsies may have the typical EEG manifestations of IOLE and therefore only EEG findings cannot be suggested to separate the symptomatic from the idiopathic group (7, 12 - 14). Similarly, we could not recommend discriminating between the two groups in the present study.

Both the seizures of PS and the GS can be more easily controlled than SOLE. The prognosis of the PS is favorable and also most patients develop one and five seizures during the lifetime.

However, 25% of them may occur more frequent and/or prolonged. Remission commonly develops within 1 to 2 years, but 15% of patients may develop BECTS (Benign Epilepsy with Centrotemporal Spikes) (15).

Moreover, the prognosis of GS is uncertain, although many studies report that remission rate at approximately 50 - 60% within 2 to 4 years of onset (3, 15). In our study, the seizures were controlled with one AED in all patients of PS and two AEDs in all patients of GS. Although one or two AEDs were adequate for achieving seizure control in IOLE, three or more AEDs were used for controlling seizure in SOLE.

Although the average duration of seizure-free in IOLE was found longer than SOLE and also GS longer than PS, it was not indicated any statistically significant difference between these groups. It can possibly be explained with the smaller numbers of groups and a short follow-up duration.

A family history of IOLE is rare although it is thought that PS and GS have genetic etiology and there are studies connected to family histories of epilepsy (15). Taylor et al. reported that it cannot be suggested on familial aggregation in IOLE compared to the general population because of too small subgroups although there were five relatives with focal epilepsy in the GS and two in PS in their study (7). We showed a positive family history of epilepsy with one patient in SOLE and one patient in PS. Febrile seizures happen in nearly 16% of patients with PS and between 2% and 4% of children with GS. Taylor et al. indicated a family history of febrile seizures in 4 of the 16 families in the GS group (7). In the present study, we showed febrile seizures with one patient in PS and one patient in SOLE.

This study has some limitations. One of them was smaller numbers of groups and the other was the length of follow-up duration.

In conclusion, the present study approves the previously represented normal neurologic examination and neuroimaging separating IOLE from SOLE. Ictal vomiting is the most striking ictal autonomic seizure and nocturnal seizures more frequent in PS. This study also suggests family history and febrile seizures likely associated with PS. Additionally, the patients with SOLE and IOLE should be evaluated carefully according to academic performance. We can enounce the IOLE is better from SOLE and PS better from GS according to treatment response. Prospective studies with larger samples need investigations.

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