

Late-Onset Childhood Occipital Epilepsy. An unusual case in adolescence and differential diagnosis with migraine

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ABSTRACT

Introduction: The new proposed classification of ILAE Task Force (2001) proposes that the occipital epilepsies should be split into two subtypes: an early-onset benign childhood occipital epilepsy (or Panayiotopoulos type) and late-onset childhood occipital epilepsy (or Gastaut type). Migraine with visual aura must be considered as a differential diagnosis in childhood and adolescents with occipital epilepsy without motor phenomena. **Objective:** The goal of our paper is to report the case a 16-year-old female, with normal psychomotor development, that during the lunch time presented an event characterized by the vision of multiple colored spots which were moving horizontally and vertically and also in circles through the visual field. Minutes after the visual event, the patient referred to a severe diffuse throbbing headache with frontal predominance. During the clinical investigation was submitted to a video-electroencephalogram exam for 12 hours with, revealing occipital sharp-waves discharges in occipital right region as well as in occipital left region. **Conclusion:** We reported of such classic type of epileptic syndrome in a patient in the unusual age of onset, the end of adolescence, considering the differential diagnosis with migraine with visual aura.

Key words: Late-onset childhood occipital epilepsy, migraine with visual aura.

RESUMO

Epilepsia occipital da infância de início tardio

Introdução: A nova proposta de classificação da ILAE (2001) propõe que as epilepsias occipitais sejam classificadas em dois subtipos: epilepsia occipital benigna da infância de início precoce (ou tipo Panayiotopoulos) e epilepsia occipital benigna da infância de início tardio (ou tipo Gastaut). A migrânea com aura visual deve ser considerada como um diagnóstico diferencial nas crianças e nos adolescentes com epilepsia occipital sem fenômenos motores associados. **Objetivo:** relatar o caso de uma paciente do sexo feminino de 16 anos, com desenvolvimento neuropsicomotor normal, que durante o almoço apresentou um evento caracterizado pela visão de múltiplas manchas coloridas que se movimentavam no sentido horizontal e vertical e também em círculos no seu campo visual. Minutos após este evento visual, a paciente passou a referir cefaléia difusa com predomínio frontal. Durante a investigação clínica foi submetida à realização de vídeo-eletrencefalograma com 12 horas de duração revelando descargas de ondas agudas ora na região occipital direita ora na região occipital esquerda. **Conclusão:** apresentamos um tipo clássico de síndrome epilética iniciando em uma idade pouco usual, o final da adolescência, considerando o diagnóstico diferencial com a migrânea com aura visual.

Unitermos: Epilepsia occipital da infância de início tardio, migrânea com aura visual.

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INTRODUCTION

In 1982, based on 36 patients, Henri Gastaut¹ reported a new type of epilepsy which was then named “benign partial epilepsy with occipital spike-waves” and later occipital epilepsy Gastaut type. This type of epilepsy is featured by visual hallucinations and favorable outcome.

According to the Commission on Classification and Terminology of the International League Against Epilepsy – ILAE (1989) there are two distinct types of benign idiopathic epilepsy in childhood, the benign childhood epilepsy with centrotemporal spikes and childhood epilepsy with occipital paroxysms.² The new classification of ILAE Task Force (2001) proposes that the occipital epilepsies should be split into two subtypes: an early-onset benign childhood occipital epilepsy (or Panayiotopoulos type) and late-onset childhood occipital epilepsy (or Gastaut type).³

The epilepsies considered benign must present the following essential features: a) a well defined clinical and electroencephalographic aspects; b) favorable clinical outcome (probably the seizures are related to a brain maturation process); c) tendency to remission during or after adolescence (age-related epilepsies); d) genetic influence (not being identified potentially epileptogenic focal structural lesions in cerebral cortex).⁴

Theoretically, the occipital idiopathic epilepsy with strong genetic component, and the symptomatic occipital epilepsies with identified structural lesions should have different clinical features and evolutions. However, in clinical practice the differential diagnosis between both is frequently complex, especially when the structural lesions are small and can not be identified by MRI equipment commonly used nowadays.

In addition, migraine with aura must be considered as a differential diagnosis, particularly in female adolescents with predominant visual manifestations, headache and without motor phenomena

The peak age of ictal manifestations is around 8 years.^{1,2} Although a late-onset childhood occipital epilepsy may have its beginning during adolescence. The reason that motivated this publication was the presentation of such classic type of epilepsy in a patient in the unusual age of onset, the end of adolescence. This paper was approved by the Pequeno Príncipe Hospital Committee of Ethics (register number 0612-08). An informed consent was presented and signed by the patient’s legal responsible allowing publication of the case report.

CASE REPORT

L.P.R., 16-year-old female, normal psychomotor development. In August 2007 during the lunch time the patient presented an event characterized by the vision of

multiple colored spots which were moving horizontally and vertically and also in circles through the visual field. Minutes after the visual event, the patient referred to a severe diffuse throbbing headache with frontal predominance. Although there are no reports of losing consciousness she could not remember the following events. At the same day during awareness an electroencephalogram and a MRI were performed and both were normal. Migraine with visual aura was the hypothetical diagnosis and due to a history of obesity a treatment with topiramate (50 mg per day) was chosen. After one month the patient had a similar event followed by ictal blindness and tonic-clonic seizure. A new electroencephalogram was performed during sleep (phase 1 and 2) for 30 minutes, which was normal again. After the second event topiramate was changed to oxcarbazepine. The patient had presented severe drowsiness with 600 mg per day, and the oxcarbazepine was changed to fenobarbital (150 mg at night). She persisted with drowsiness and after five months presented other event with the same features as the previous ones. At this time the patient was referred to Pequeno Príncipe Hospital in Curitiba, Brazil, for a new evaluation and following treatment.

The history of a bariatric surgery in the past and some aspects of anxiety and signs of depression hindered the conduction of the case in that moment. However the patient was always cooperative with the exams and the treatment.

In April 2008, still using fenobarbital, the patient was submitted to a video-electroencephalogram exam for 12 hours with electrodes placed in accordance with the international 10-20 system. The exam revealed occipital sharp-waves discharges with high electronegativity alternately in occipital right region as well as in occipital left region and sometimes synchronously in both occipital regions. The background activity was considered normal during the resting awake state as well as at sleep phase 1 and 2 and slow wave sleep. A blockage of dominant rhythm was observed after the proof of eye closure. Voluntary hyperventilation and intermittent photic stimulation with a strobe light (1 to 21 Hz) did not trigger any specific abnormality. Fixation-off sensitivity was not observed during the exam.

After the exam, fenobarbital was replaced by carbamazepine with slow increase of the dosage to avoid the side effects occurred previously when the patient was being treated with oxcarbazepine. At the moment the patient has been treated with carbamazepine 400 mg per day and it has achieved the complete control of seizures.

DISCUSSION

The late-onset childhood occipital epilepsy, described by Gastaut¹ more than 25 years ago, is considered a rare

type of age-related epilepsy, with its peak in the beginning of school age. Cases beginning after adolescence are extremely rare and of difficult diagnosis.

Clinical aspects: this epileptic syndrome is characterized by partial visual seizures, predominantly manifested with elementary visual hallucinations, ictal blindness and less frequently with complex visual hallucinations and visual illusions. The visual events, including the visual hallucinations, can be followed by holocranian headache, of frontal predominance and severe intensity. The seizures usually occur during the day, are frequent and short, lasting less than 5 minutes.^{1,5} According to Panayiotopoulos,⁵ the elementary visual hallucinations in late-onset childhood occipital epilepsy may present different features, such as single or several colored images, generally circular, moving horizontally through the visual field. The images may be either multiplied in size and increase or decrease during the seizure. Elementary visual hallucinations are rarely followed by the complex hallucination type such as micropsia, macropsia and metamorphopsia as well as by later evolution to tonic-clonic or hemiclonic seizures. Those events are more frequent in symptomatic epilepsies than in the idiopathic types. The sensation of ocular movements (in the absence of detectable motion) may correspond to an ictal event, preceding or not a secondary generalization. The ictal and pos-ictal blindness, preceding or succeeding visual hallucinations are common symptoms in late-onset childhood occipital epilepsy.^{5,6}

Deviation of the eyes with or without ipsilateral turning of the head is also a frequent symptom, occurring in two thirds of the patients.^{1,5,7} The ictal vomiting, usually seen in Panayiotopoulos syndrome, is a rare symptom in late-onset childhood epilepsy and can be used as differential diagnosis criteria.⁸

Lee et al.⁹ evaluated the clinical aspects of 54 children with occipital lobe epilepsies, where 26 patients had symptomatic type, 18 had Panayiotopoulos type and 10 had Gastaut type. The study demonstrated that the nocturnal events were predominant in Panayiotopoulos type, the daily events were more common in the Gastaut type and secondary generalization was more frequent in patients with symptomatic epilepsies.

The differential diagnosis of idiopathic and symptomatic occipital epilepsy is not usually complex. In a study carried out by Fogarasi et al.,¹⁰ with video-EEG, 110 epileptic events were analyzed in 18 children with ages range from 3 to 81 months and diagnosed as symptomatic occipital epilepsy. The authors concluded that childhood symptomatic occipital epilepsy presents a varied semiology, presenting different types of epileptic events such as generalized tonic-clonic seizures, myoclonic seizures, staring, oral automatisms, oculo-cephalic deviation and frequent episodes of behavior's alterations.

It was demonstrated in a European longitudinal retrospective study with a group of 30 children with idiopathic occipital epilepsy that the precocious appearance of occipital's discharges is related to longer occipital seizures and to ictal vomiting. It also demonstrated that late appearance of discharges is related to previous febrile seizures and predominance of visual manifestations during seizures.¹¹

Our patient, besides the very late-onset, presented the classical signs of late-onset childhood occipital epilepsy. She had elementary visual hallucinations with colored dots and headache in the first episode, and then ictal blindness and secondary generalization in the second one. There were no reports of ictal or pos-ictal vomiting.

The most interesting aspect approached in our case was specifically the age of the seizure's onset. Tsai et al.¹² evaluated 12 children with early-onset occipital epilepsy and 14 children with late-onset occipital epilepsy. The study showed the peak age of onset in the first group was 4,9 years ($\pm 1,7$) and 8,4 years ($\pm 2,5$) in the second group. They also demonstrated that ictal vomiting, nocturnal predominance, small frequency and longer duration were marked features of early-onset occipital epilepsies, and in the late-onset type ictal headache and visual hallucinations predominated.

The late-onset childhood occipital epilepsy is considered an uncommon epileptic syndrome and cases beginning after adolescence are considered extremely rare.

Electrographic aspects: in late-onset occipital epilepsy the occipital paroxysms may be synchronous and have a bilateral occipital localization or can present maximal electronegativity alternating among the left and right occipital regions. There are some cases when the discharges may be generated at various cortical locations. The association between late-onset childhood occipital epilepsy and childhood absence epilepsy could suggest a sub-cortical epileptic mechanism that might be situated in thalamus.^{7,13} The background activity is always normal and EEG abnormalities like bursts of diffuse slowing of the background activity, occipital or extra-occipital discharges, suggest symptomatic etiology.⁹

It is not possible to differentiate between early-onset and late-onset types considering just the EEG features, however, a study conducted by Martín Santidrián et al.¹¹ showed that the migration of the occipital discharge to anterior cortex regions was significantly more common in the early-onset type.

Elimination of central vision by using means such as darkness or eyes closed corresponds to a stimulation test carried out during EEGs records in patients with suspected occipital epilepsies. The test can trigger high-amplitude discharges in occipital regions or, less frequently, generalized discharges. Evaluating 76 children with occipital paroxysms,

Martinovic¹⁴ founded fixation-off sensitivity in 42 cases. According to the author the fixation-off sensitivity was not significantly related to a specific clinical condition, as it could be encountered in idiopathic and symptomatic occipital epilepsies. The EEG findings in our patient, which were normal background activity with only occipital discharges and without focal slow waves, were typical of late-onset childhood occipital epilepsy.

Management: although there is a possibility of maintaining the patients without antiepileptic drugs in some types of idiopathic epilepsies, in cases of late-onset childhood occipital epilepsy all patients must be treated due to the tendency of diurnal seizures and frequent recurrence of the events. The seizures usually stop with appropriate doses of carbamazepine. There are no diverging points in the literature.⁵ The treatment with carbamazepine has been well tolerated by our patient until this moment, and the complete control of seizures has been achieved. We believe that parts of the side effects observed with oxcarbazepine were related to emotional aspects and anxiety on account of the treatment, as carbamazepine is being extremely well tolerated.

FINAL REMARKS

In theory, to confirm the diagnosis of childhood idiopathic epilepsies a high resolution MRI should be performed in all cases, however, this exam is not available for the majority of patients – which does not allow this diagnostic criterion to be strictly accomplished.

The differential diagnosis of occipital epilepsies is complex. The differentiation based just on clinical observations may be impossible among patients with occipital lobe seizures without motor symptoms and patients with migraine with visual aura associated to syncope.

Even with the new proposed ILAE classification,³ which has distinct criteria for each type of occipital epilepsy, the clinical differentiation remains problematic. After the evaluation of 25 children who presented suspected occipital epilepsy and discharges located on occipital lobe, Genizi et al.¹⁵ concluded that only 50% of patients fulfilled the criteria of ILAE classification to be diagnosed as having early-onset (Panayiotopoulos) or late-onset (Gastaut) forms. The other 50% had symptoms common to both forms. In an important study, Kivity¹⁶ analyzed 134 children from January 1975 to May 1997 with partial seizures associated to occipital epileptiform paroxysms. These patients were subdivided into three groups: (a) group 1, 24 patients presenting ictal visual symptoms; (b) group 2, 73 patients presenting tonic eye deviation; and (c) group 3, 38 patients with various seizures spread patterns. The mean ages of first seizures were 7 years and 11 months (group 1), 5 years and 2 months (group 2), 6 years (group 3), and the mean ages of last seizures were 10 years (group 1), 7 years

and 2 months (group 2 and 3). The study concludes that the Panayiotopoulos occipital epilepsy is the most frequent type and the symptoms and signs are sufficiently distinct among Panayiotopoulos type, Gastaut type and occipital symptomatic epilepsies to justify the separation of these three types of epileptic syndromes.

The analysis of our case shows that although the idiopathic partial epilepsy are age-related and have, in theory, time limits to occur, some cases may be present later hindering the diagnosis.

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