Childhood Occipital Epilepsy Of Gastaut: A Case Report

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L'épilepsie occipitale de type Gastaut: A propos d'un cas

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RÉSUMÉ

Prérequis : L'épilepsie occipitale idiopathique de l'enfant de type Gastaut est rare. Il s'agit d'un syndrome épileptique bien défini dans le groupe des épilepsies focales de l'enfant. Il se manifeste sur le plan clinique par des crises partielles simples, constituées essentiellement par des symptômes visuels, pouvant être suivis de généralisation secondaire.

But: Rapporter l'observation d'une épilepsie occipitale idiopathique de l'enfant de type Gastaut.

Observation: Nous rapportons le cas d'une fille âgée de 10 ans, sans des antécédents pathologiques notables. À l'âge de 9 ans, l'enfant a commencé à avoir des crises épileptiques dans un contexte d'apyrexie. Les premières crises consistaient en de plusieurs épisodes de perte de conscience avec hypotonie et amnésie post critique. Le deuxième type de crises était trois épisodes d'hallucinations visuelles complexes durant 10 minutes. Un mois plus tard, un troisième type de crises généralisées tonico-cloniques est apparu. L'examen neurologique et les bilans biologiques de routine étaient normaux. La neuroimagerie était normale. L'EEG interitique a montré des bouffées d'ondes lentes à front raide généralisées à l'hyperventilation. Elle a été traitée par du Valproate de Sodium, avec disparition des crises. L'enfant est actuellement asymptomatique.

Conclusion: Ce syndrome épileptique doit être identifié, et traité tôt. En effet, sans traitement approprié, l'évolution peut être marquée par l'installation de pointes ondes continue pendant le sommeil avec détérioration cognitive.

Mots-clés

Epilepsie focale idiopathique, Crises occipitales, Hallucinations visuelles, Enfance

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SUMMARY

Background : Idiopathic childhood occipital epilepsy of Gastaut is a rare but well defined syndrome within the group of idiopathic focal epilepsies in childhood. Clinical manifestations are characterized by simple partial seizures with mainly visual symptoms followed by secondary generalization.

Aim: Report of a case of Childhood Occipital Epilepsy.

Case report: We report a case of 10-year-old-girl, with no history. At the age of 9 years, the patient started experiencing repetitive events without fever. The first event consisted of several episodes of loss of consciousness with hypotonia and post ictal amnesia. The second type was three episodes of stereotyped, elaborated complex visual hallucinations, during 10 minutes. One month later, a third type of tonico-clonic generalized seizures appeared. Neurological examination and routine laboratory investigation were normal. Brain neuroimaging was normal. Interictal EEG showed bilateral discharges of slow generalized waves activated by hyperventilation. She has been treated by Sodium Valproate. All seizures stopped and she remained free of seizures.

Conclusion: This epileptic syndrome must be identified, and treated without delay. In fact, without appropriate treatment, the evolution may lead to a continuous spike-wave during the sleep with cognitive deterioration.

Key-words

Focal idiopathic epilepsy, Occipital seizures, Visual hallucinations, Childhood

مرض صرع أساسي للأطفال متعلق بمؤخر الرأس من صنف" قاستو" : دراسة حول حالة طفل

الباحثون : هـ .المرابط الخياري ، هـ .البطي شحيدر ، أ .المرابط .

تستعرض دراستنا حالة فتات بدأت بالتعرض إلى ثلاثة أنواع من الصرع في التاسعة من عمرها .في البداية كانت نوبات الصرع تتمثل في فقدان الوعي مع استرخاء في العضلات .النوع الثاني من النوبات يتمثل في تخيلات بصرية تدوم عشرة دقائق ،ترى فتيات يقمن بتقديم الكعك لها .في الشهر التالي ظهر نوع ثالث من الصرع يتمثل في نوبات تقلصية ارتجاجية .الفحص الطبي و الفحوصات البيولوجية و التصويرية كانت سليمة .التخطيط الدماغي اظهر أمواج بطيئة في التنفس السريع .خضعت الفتاة للعلاج ب "فالبر ووات دو صوديوم "مما أدى إلى اختفاء النوبات .كان الدواء ناجعا و أصبحت الفتاة سليمة من كل أعراض مرض الصرع.

استنتجنا أن هذه الفتاة لها نوع نادر من الصرع ،مرض صرع أساسي للأطفال متعلق بمؤخر الرأس من صنف" قاستو ."النوبات الجزئية البسيطة البصرية تمثل أهم مظهر لهذا المرض .يمكن أن تصبح النوبات كلية. العلاج المناسب أساسي لتفادي ظهور نشاط دماغي حاد و أمواج متواصلة في النوم مما قد يؤدي إلى مساس بالقدرات الذهنية.

الكلمات الأساسية : مرض صرع جزئي أساسي؟ نوبات متعلقة بمؤخر الرأس - تخيلات بصرية -طفولة-

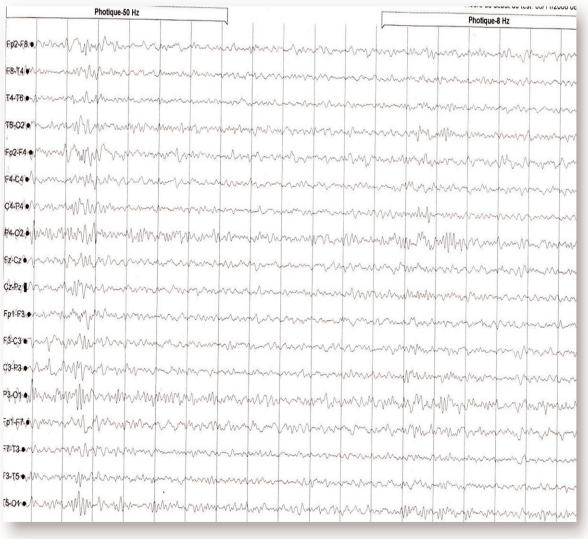
Recognizing epileptic syndromes on clinical, EEG, neuroimaging features is essential for treatment and prognosis. Benign childhood focal seizures and related idiopathic epileptic syndromes affect 20% of children experiencing non-febrile seizures (1). The common example is Benign Rolandic Epilepsy, a well known idiopathic epileptic syndrome with good outcome. The Panayiotopoulos syndrome (PS) is also a frequent benign childhood focal epilepsy. The idiopathic childhood occipital epilepsy of Gastaut (COE-G) including the idiopathic photosensitive occipital lobe epilepsy is a rare but well defined syndrome within the group of idiopathic focal epilepsies in childhood.

CASE REPORT

The patient is a 10-year-old-girl, with healthy non consanguineous parents. Pregnancy and delivery are normal, as well as the child's early development. There is no family history of epilepsy. She is currently in 5th primary scholar year.

At the age of 9 years, she started experiencing repetitive events without fever. The first event consisted of several episodes of loss of consciousness with hypotonia and post ictal amnesia. The second type was three episodes of stereotyped, elaborated visual hallucinations without loss of contact: the patient reports seeing little girls who offered her cakes. These episodes lasted 10 minutes. One month later, a third type of tonico-clonic generalized seizures appeared. Neurological examination and routine laboratory investigation were normal. Brain neuroimaging was normal. Interictal EEG showed bilateral discharges of slow generalized waves activated by hyperventilation (Figure 1). Sodium valproate was prescribed at 20 mg/kg/day. All seizures stopped and she remained free of seizures.

Figure 1 : Interitictal EEG, activation procedures, longitudinal: bilateral discharges of slow generalized waves



DISCUSSION

Our patient presented electroclinical features compatible with the diagnosis of Childhood Occipital Epilepsy (COE-G). This epilepsy syndrome, as described by Gastaut, is rare, occurring in 2 to 7% of benign childhood focal seizures (2, 3, 4, 5, 6, 7, 8). Family history, typically absent, was reported in some patients (9).

Age at onset is between 3 and 16 years. Boys and girls of all races are equally affected. Clinical manifestations are characterized by simple partial seizures with mainly visual symptoms followed by secondary generalization. Post ictal migraine headache may occur.

Visual seizures are the cardinal symptoms. They predominantly manifest with elementary visual hallucinations, complex hallucinations, blindness, or both. Elementary visual hallucinations are common (2/3 of the cases) and most characteristic of ictal symptom. They consist of stereotyped small multicolored circular patterns that often appear in the periphery of a visual field, frequently moving horizontally towards the other side. Hallucinations are usually diurnal, lasting few seconds to less than 3 minutes (1, 6). In a study of 33 patients with electroclinical criteria of COE-G, visual manifestations were the most common ictal event and may last more than 10 minutes as in our patient (7). Complex hallucinations are rare (10%) and characterized by the appearance of faces or persons with the same location and movement sequence as in elementary visual hallucinations. Blindness and/or blurring of can occur and be the only clinical manifestation (1, 2).

Deviation of the eyes associated with ipsilateral turning of the head can follow visual hallucinations. Eyelid closure and blinking; hemiconvulsions; secondarily generalized tonic–clonic seizures (GTCS) and complex focal seizures, has also been reported (7, 8).

Seizures can be triggered by intermittent luminous stimulation, such as television. They are occasionally triggered or aggravated by turning off lights, going from lighted areas to dark ones, or from dark areas to light ones (3, 8).

Interictal EEG shows occipital spikes and spike-waves with fixation-off sensitivity. Occipital paroxysms, in routine recordings, occur when the eyes are closed, and disappear or are attenuated when eyes are opened. The sporadic occipital spikes can sometimes occur only during sleep and normal EEG can be observed. Ictal EEG shows discharges of fast spike occipital activity. While the amplitude of the ictal activity increases, the frequency of repetition lessens. The ictal manifestations can have specific EEG findings: the elementary visual hallucinations are accompanied by fast spikes, whereas the complex hallucinations are accompanied by slower discharges. In oculoclonic seizures, a localized ictal fast spike rhythm may be observed before the deviation of the eyes. Temporary blindness is characterized by pseudoperiodic slow waves and spikes (1, 8). COE-G affects children with normal development. The EEG is the most useful diagnostic test. The results of all

other investigations are normal. Brain MRI may be needed for children suspected of symptomatic epilepsy caused by structural occipital brain pathology. The other etiologies to be eliminated are hyperglycemias, Lafora or mitochondrial diseases or occipital calcifications in relation with a c?liac disease (5, 6, 9).

COE-G may be misdiagnosed as Panayiatopoulos syndrome (PS). The PS is a benign age-related focal seizure disorder occurring in early and mid-childhood. It is characterized by seizures, often prolonged, with predominantly autonomic symptoms. The visual hallucinations remain less frequent. The EEG shows multifocal spikes, often with occipital predominance (1, 2, 8).

COE-G should be treated because seizures, though brief and mild, are frequent. Secondary generalization is probably unavoidable without medication. A delay in the introduction of a treatment may lead to a continuous spike-wave during the sleep with cognitive deterioration. Seizures stop or are dramatically reduced within days after appropriate treatment with carbamazepine, in more than 50% of the cases (3, 7).

Références

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