CASE REPORT
CASO CLINICO

Late-onset Panayiotopoulos syndrome: a case report

Sindrome di Panayiotopoulos ad esordio tardivo: un caso clinico

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KEY WORDS. – Panayiotopoulos syndrome - Epilepsy - EEG - Adolescence
PAROLE CHIAVE. – Sindrome di Panayiotopoulos - Epilessia - EEG - Adolescenza

Riassunto

La sindrome di Panayiotopoulos è un'epilessia benigna del lobo occipitale, ormai ben conosciuta, che generalmente inizia all'età di 3-6 anni. L'esordio più tardivo di questa sindrome epilettica è poco comune. Riportiamo qui le caratteristiche cliniche ed elettroencefalografiche di una ragazza con una tipica sindrome di Panayiotopoulos esordita all'età di 13 anni.

Introduction

Panayiotopoulos syndrome (PS) is a benign age-related focal seizure disorder occurring in early and mid childhood [1-3].

PS is characterized by seizures, often prolonged, with predominantly autonomic symptoms, namely emesis as the most relevant manifestation. EEG shows multifocal spikes or sharp-slow-wave complexes, often shifting from one region to another, with occipital predominance. The results of all other investigations, including also brain MRI, are normal [1-3].

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We report here the case of a girl with a typical PS, but with an unusual late onset, at 13 years of age.

Case report

A.K. is a 13-year-old Finnish girl who came to our observation with a long-lasting ictal symptomatology characterized by fever, headache, vomiting and cognitive disturbances, such as confusional state and inability to speak, occasional dyskinetic movements of the right arm. For this reason she was hospitalized and an encephalitis was suspected. Her personal and family history was unremarkable.

An interictal EEG, a brain MRI and a lumbar puncture resulted normal.

One month later, since she continued to present the same symptomatology, she was admitted again and underwent another EEG during an episode of status epilepticus which showed repetitive left occipital ictal discharges with a fast ipsilateral and a much slower contralateral diffusion (Fig. 1). This epileptic status ended after 2 hours, following an intravenous infusion of lorazepam (4 mg); therapy

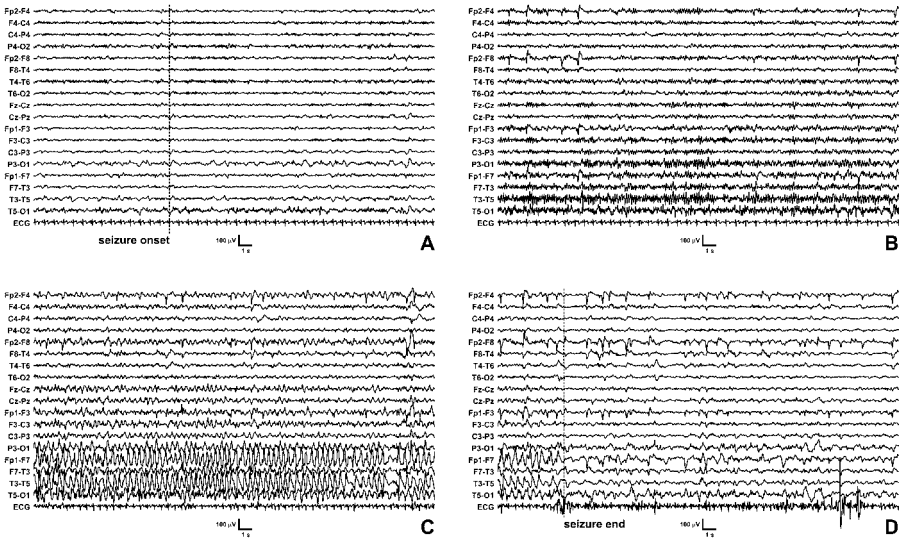

Fig. 1. A) Seizure onset characterized by the appearance of low-voltage fast activity over the left temporal-occipital regions; B) 2'20" after the seizure onset, the fast activity is diffuse over the left hemisphere; C) 4'20" after the seizure onset, a continuous discharge of spike-and-wave complexes is evident over the left hemisphere, especially over the temporal regions; D) The seizure ends almost abruptly after 5'25", and a postictal theta-delta activity is present over the left hemisphere.
(ECG = electrocardiogram).

with carbamazepine was started and all the symptoms disappeared in the next few days. Four days later, neurological examination was normal and an interictal EEG showed a slow focal activity, which was intermixed with spikes on the left occipital regions and was not interrupted by eye opening (Fig. 2). During her staying in our Department, another brain MRI was performed which resulted normal.

Discussion

Our patient presented long-lasting recurrent episodes of status epilepticus with autonomic manifestations, such as headache, vomiting, focal seizures accompanied by impairment of consciousness, an interictal EEG characterized by slow waves and spikes over the occipital regions and an ictal EEG showing ipsilateral discharges diffused to both hemispheres from the posterior regions. This clinical and EEG pattern, together with the findings of normal neurological examination, brain MRI and lumbar puncture allowed us to diagnose an occipital epilepsy with the peculiar features of the PS.

PS is a well-known childhood-related idiopathic benign focal epilepsy. Most individuals with PS have their first seizure around the age of 5 years, and three-quarters of the patients have their first seizure between the ages of 3 and 6 years [1]. This suggests a presumably genetically determined phenomenon, consisting in a diffuse cortical hyper-excitability which is related to the maturation of the brain.

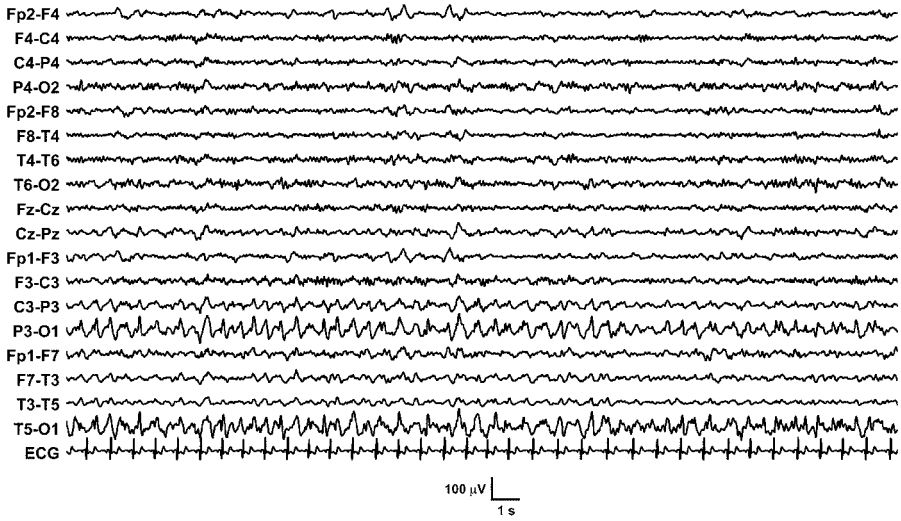

Fig. 2. The interictal EEG shows a mostly slow focal activity, intermixed with spikes on the left occipital regions (ECG = electrocardiogram).

Maturation of the brain seems to continue until near the end of the second decade of life, in fact the signal intensities in the white and deep grey matter decrease rapidly in the first decade and then gradually reach a plateau after the age of 18 years [4].

It is of interest that three main electroclinical patterns of benign partial epilepsies of adolescence have emerged from a large multicenter study. The first is characterized by the occurrence of rare somato-motor seizures and the presence of focal theta discharges over the centrotemporal regions, the second consists of versive seizures and focal spikes or sharp waves over the posterior regions, the third has a short seizure period and a normal EEG picture [5]. Then, it is reasonable to include our case with PS in these categories of benign partial epilepsies of adolescence.

The recognition of such a self-limiting clinical and EEG pattern not only in children but also in adolescents may avoid misdiagnosis and unhelpful and invasive procedures (i.e. lumbar puncture), and may allow to exclude other organic conditions, such as stroke and encephalitis.

Summary

Panayiotopoulos syndrome is a well-known benign occipital lobe epilepsy mainly starting at 3-6 years of age. A later onset of this epileptic syndrome is unusual. We report here the clinical and EEG features of a girl with a typical Panayiotopoulos syndrome with onset at 13 years of age.

The recognition of such an electroclinical pattern also in adolescents may help in excluding other organic conditions, such as stroke and encephalitis.

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