LATE EXACERBATION OF RASMUSSEN ENCEPHALITIS WITH CHILDHOOD ONSET

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ABSTRACT

We describe the case of a 21 year old woman that presented for epilepsia partialis continua syndrome that she had been having in the last 8 months. Slight diminished speech fluency, cortical discriminative sensibility impairment and neuroradiological features of hemispheric atrophy pointed to the diagnosis of Rasmussen Encephalitis. We highlight an unusual prolonged interval between the first focal seizures and the onset of the suggesting clinical picture, that consisting an evidence for the clinical variability of this disease.

Key words: epilepsia partialis continua, Rasmussen encephalitis, focal encephalitis

BACKGROUND

Rasmussen's Encephalitis (RE) is a rare syndrome characterized by intractable seizures, often associated with epilepsia partialis continua (EPC), and symptoms of progressive hemispheric dysfunction [1]. Although adolescent and adult onset cases have been described [2,3] it usually occurs in children [4,5].

We present a case with EPC at the age of 21, with past history of simple partial motor seizures at the age of 9, which was diagnosed as RE. We correlated the clinical picture with previously reported cases, and found mixed characteristics from childhood and adult onset RE.

CASE PRESENTATION

A 21 years old woman was admitted for involuntary movements in the right upper limb, especially of the hand and fingers, which she had been having continuously for 8 months.

She had been diagnosed with partial motor seizures at the age of 9 when Carbamazepin 600 mg/day was started and she was seizure free untill the age of 15, when she presented a few episodes of clonic movements in her right leg. Gabapentin 3x300 mg/day and Clonazepam 2x0.5 mg/day and were added-on and she became seizure free for the next 4 years. At the age of 19, the partial motor seizures recurred, 10-20 episodes/day of 1-2 minutes length. Brain MRI showed a small T2 and FLAIR hyperintense lesion without Gadolinium enhancement in the left occipital cortex. Fig 1. EEG revealed theta activity, as well as spikes and sharp-waves mainly located over the left parieto-occipital region.

At the age of 21 she presented an 8 months course of continuous intractable clonic movements of her right upper limb so that the diagnosis of epilepsia partialis continua was made. Physical examination revealed an agonist/antagonist muscle contraction involving the right upper limb, especially the hand and fingers. The continuous jerks totally disabled the patient as regarding the motor function of her right upper limb.The patient could partially supress the clonic movements by firmly holding the arm or by pressing it against a surface. The myoclonic jerks were absent during sleep. Slight impairment of cortical sensory functions and diminished speech fluency were noticed too. The EEG revealed a diffuse left hemisphere slowing consisting in theta-delta waves, no ictal activity. CSF cell count (5 cells/μl) and protein levels (20 mg/dl) were normal.

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MRI disclosed gliosis in the left occipital cortex and slight atrophic changes of the whole left hemisphere. Fig 2. All 3 of Part A diagnostic criteria for RE were present [4].

Intravenous immunoglobulin (IVIG) 400 mg/kc 5 days/months was administered for several months, followed then by Azathioprine 1.5 mg/kg per day. The myoclonic jerks diminished in amplitude and area during the next 6 months. A pyramidal syndrome with increased tendon reflexes and right Babinski sign developed. Atrophy of the right limbs was noticed after 12 months. During the next 24 months the patient was stable, except for 3 episodes of increased myoclonic jerks with different localization: the leg, the upper arm, the thorax, or the face on the right side.

CONCLUSIONS AND DISCUSSIONS

Disease onset was characterized by focal motor seizures. The most prominent feature of the case was EPC that developed 12 years later. EPC jerks involved different regions, but they were strictly unilateral. Neurological deficits were minimal at the moment of EPC, a moment when imagistical findings of atrophy pointed to the diagnosis. That sustains the statement that the "myoclonic" fenotype is characterized by less severe and later developed neurological deficits were [3] as compared to the "epileptic" phenotype.

The long delay (12 years) between the first seizure and the onset of EPC and neurological deficit consists an usual finding in children, where the prodromal stage is very short, ranging from 15 days to 24 months, with a mean of 6.8 months [3], the longer course being more common in adulthod onset.

The initial lesion was occipital, although this location appears to be more common in the adolescent and adult onset form [5], while the temporal region is characteristic in childhood RE [2].

Childhood onset with late exacerbation consists a particular type of evolution in childhood RE, pointing to the the clinical variabiliy of the disease.

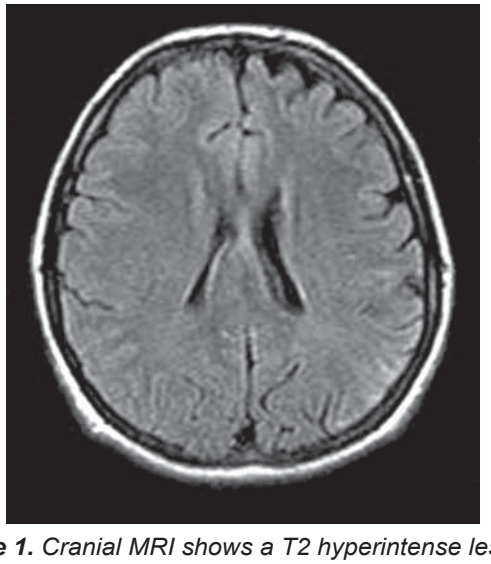

Figure 1. *Cranial MRI shows a T2 hyperintense lesion in the left parieto-occipital region.*

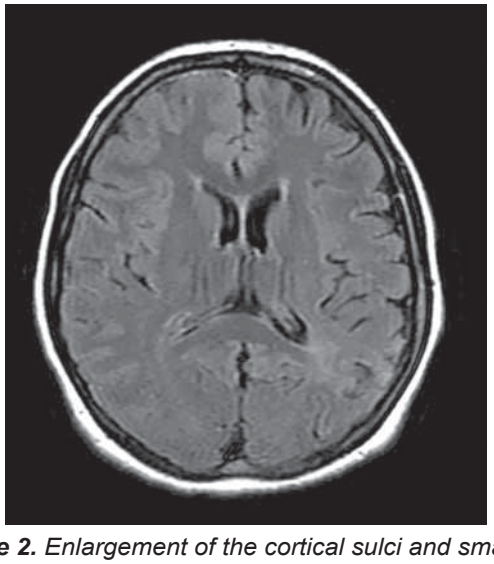

Figure 2. *Enlargement of the cortical sulci and smaller size of the left hemisphere.*

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