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Pilomotor seizures associated with sequential changes in magnetic resonance imaging

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ABSTRACT – Piloerection is rarely described in seizures. This symptom has been most frequently observed in patients with temporal lobe epilepsy and is rarely the principal clinical feature of seizures. No specific etiology of epilepsy associated with pilomotor seizures has been reported. We present the first case of a patient who experienced sudden and transitory epilepsy with pilomotor seizures occurring several times a day for months, and associated with sequential changes of the left hippocampus demonstrated by magnetic resonance imaging.
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Key words: pilomotor seizures, transient MRI signal abnormality, hippocampus atrophy, autonomic seizures, TLE

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Pilomotor seizures are classified as a subtype of autonomic seizures, so-called cutaneous autonomic seizures (Baumgartner *et al.* 2001). Piloerection is rarely reported as an ictal manifestation. It may occur unilaterally or bilaterally. Lateralizing and localising value of ictal piloerection is still the subject of debate (Loddenkemper *et al.* 2004). However, this symptom has been most frequently observed in patients with temporal lobe epilepsy. No specific etiology of epilepsy associated with pilomotor seizures has been found.

We describe an additional case with pilomotor seizures as the main feature of temporal lobe epilepsy, studied with video-EEG recording, and associated with sequential changes in magnetic resonance imaging (MRI).

Case report

A 35-year-old, right-handed woman, with no relevant clinical history, was referred for sudden epilepsy with a seizure frequency reaching 30 per day. They lasted less than 90 seconds without loss of consciousness. They consisted of bilateral sensations of chill, associated with coloured phosphenes in the right hemi-field, nausea, thoracic compression, followed a few seconds later by piloerection involving both arms and legs. Results of neurological and general examination were normal.

Routine laboratory studies, including white cell count, showed normal results.

Standard EEG recording showed slow waves on the left temporal region. MRI performed one week after the first seizure, showed increased signal intensity of the left hippocampus in T2 and FLAIR-weighted images *(figure 1)*.

She received antiepileptic drugs, which decreased the severity and the frequency of seizures. The number of seizures decreased from 30 to 10 per day.

Three and six months after the onset of epilepsy, follow-up MRI showed a slight regression of the signal abnormality in the left hippocampus (*figure 2*A, B).

Video-EEG performed six months after the start of the disease recorded six seizures, each lasting less than one minute. These seizures consisted of sensations of chill ascending from the feet to the whole of the body, associated with piloerection on the left arm. Ictal EEG showed diffuse flattening of the electrical activity followed by a rhythmic slow activity with a maximum amplitude on the left central and temporal area (*figure 3*A, B and *figure 4*A, B). Interictal video EEG was normal.

One month later, seizures ceased after adjustment of the antiepileptic medication. The patient became pregnant and had a healthy child.

Two years after the onset of epilepsy, MRI showed a clear decrease of the signal abnormality in the left hippocampus, however atrophy of the left hippocampus was observed *(figure 5)*. The patient remains seizure-free with carbamazepine, 600 mg/d.

Discussion

Pilomotor excitation as an ictal sign has been rarely well studied with video-EEG recordings. Several case reports and experimental findings confirm piloerection may be induced by epileptic discharges (Stefan *et al.* 2002). It could also be a secondary induced sensation occurring during seizures, in particular those associated with psychic symptoms such as feelings of fear. However, pilomotor excitation may be the first clinical symptom of seizure, as reported in five of out 25 patients with pilomotor seizures recorded at the Cleveland Clinic Foundation between 1994 and 2001 (Loddenkemper *et al.* 2004).

Ictal piloerection is usually associated with other symptoms. Most of them are autonomic signs: flushing, pallor, sweating, feeling of warmth or cold, shivering. In many cases, these autonomic signs are associated with other, non-autonomic ictal phenomena related to the onset and propagation of the ictal discharge: sensory hallucinations, feeling of fear, automatisms, loss of consciousness (Baumgartner *et al.* 2001, Loddenkemper *et al.* 2004).

Seizure consisting of piloerection as the principle ictal manifestation is very uncommon. Less than 10 cases have been reported in the literature (Roze *et al.* 2000, Loddenkemper *et al.* 2004).

It most often occurs in patients with temporal lobe epilepsy (Stefan *et al.* 2003). However, the generator of ictal piloerection remains unclear. It has also been observed in seizures with frontal or parietal onset. The amygdala, anterior insula, anterior cingulate cortex and posterior orbitofrontal cortex are interconnected with the central

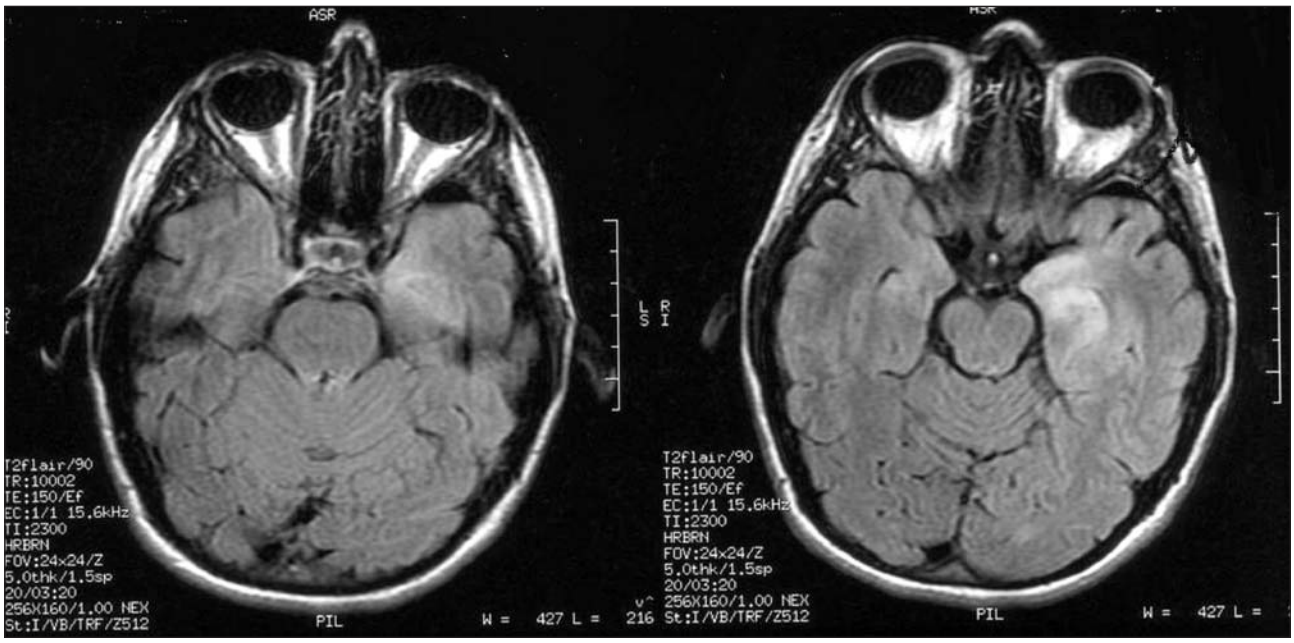

Figure 1. MRI performed seven days after the start of epilepsy: increase signal intensity of the left hippocampus in axial FLAIR-weighted images (hippocampic plan).

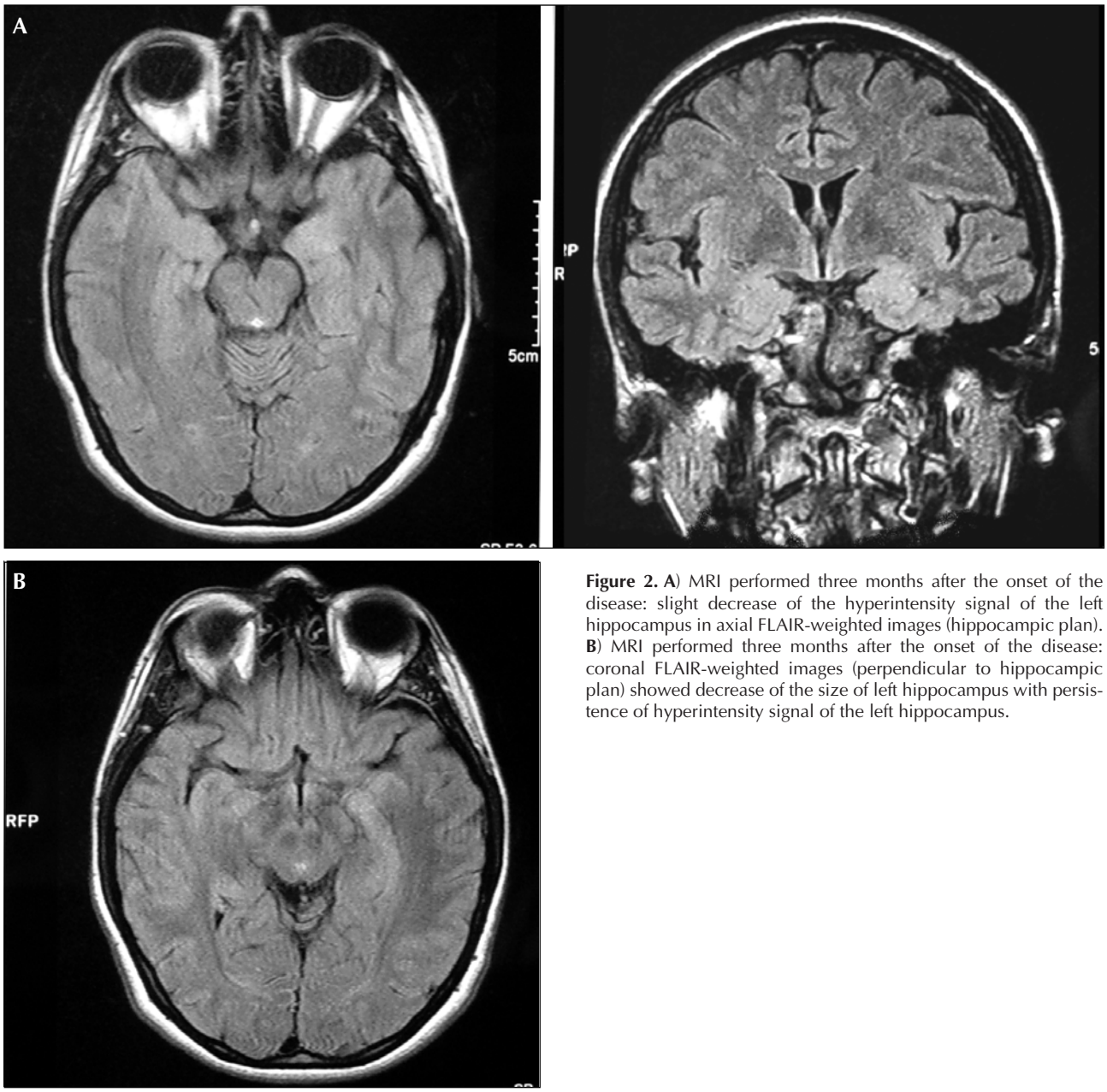

Figure 2. A) MRI performed three months after the onset of the disease: slight decrease of the hyperintensity signal of the left hippocampus in axial FLAIR-weighted images (hippocampic plan). **B**) MRI performed three months after the onset of the disease: coronal FLAIR-weighted images (perpendicular to hippocampic plan) showed decrease of the size of left hippocampus with persistence of hyperintensity signal of the left hippocampus.

autonomic network: (Devinski *et al.* 2004). The autonomic network includes the hypothalamus, periaqueductal gray matter, parabrachial region in the pons, solitary tract nucleus and ventrolateral medulla with specific organization (Benarroch 1993). Electrostimulation or seizures spreading in the central autonomic network can modify autonomic functions. These autonomic changes can induce cardiovascular, respiratory, gastrointestinal, cutaneous, pupillary, urinary and genital, manifestations. Piloerection has been elicited by stimulation of multiple sites: insula, hippocampus, amygdala, hypothalamus, midbrain and medial prefrontal cortex in humans (Fish *et al.* 1993). Seizures originating in the mesial temporal area may spread to the insula inducing autonomic signs. In our observation, electroclinical and neuroimaging data analysis might suggest involvement of the left mesiotemporal and insula during the epileptic discharges. All these areas are interconnected with the central autonomic network.

Piloerection may be localized, with the possibility of secondary spreading to another, homolateral or contralateral area of the body. It may also be generalized from the start of the seizures. Unilateral or initially unilateral piloerection is usually associated with an ipsilateral, epileptogenic focus (Loddenkemper *et al.* 2004).

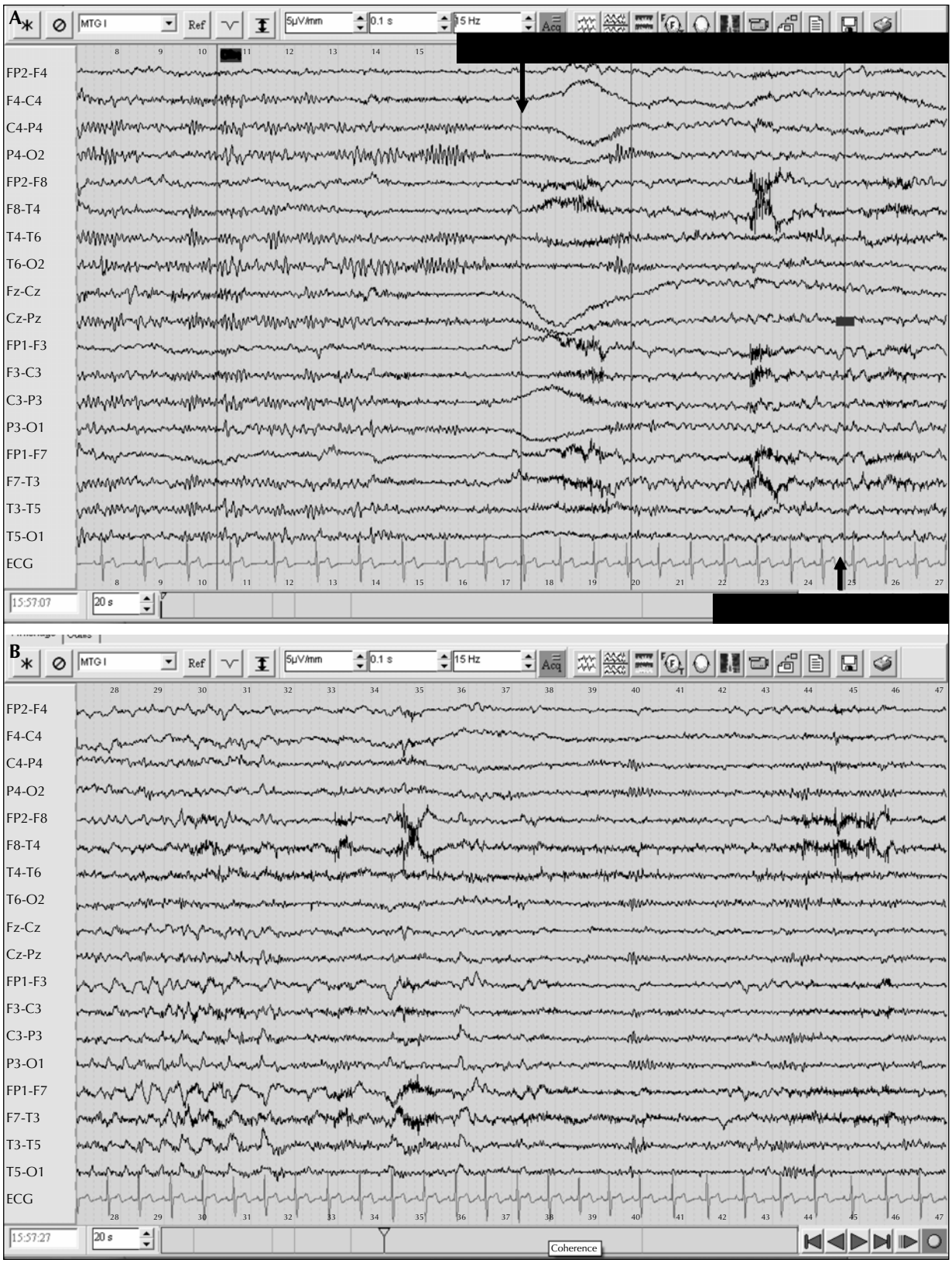

Figure 3. A, B). Ictal video-EEG recording showed diffuse flattening of the electrical activity followed by a rhythmic slow activity with maximum amplitude on the left central and temporal area.

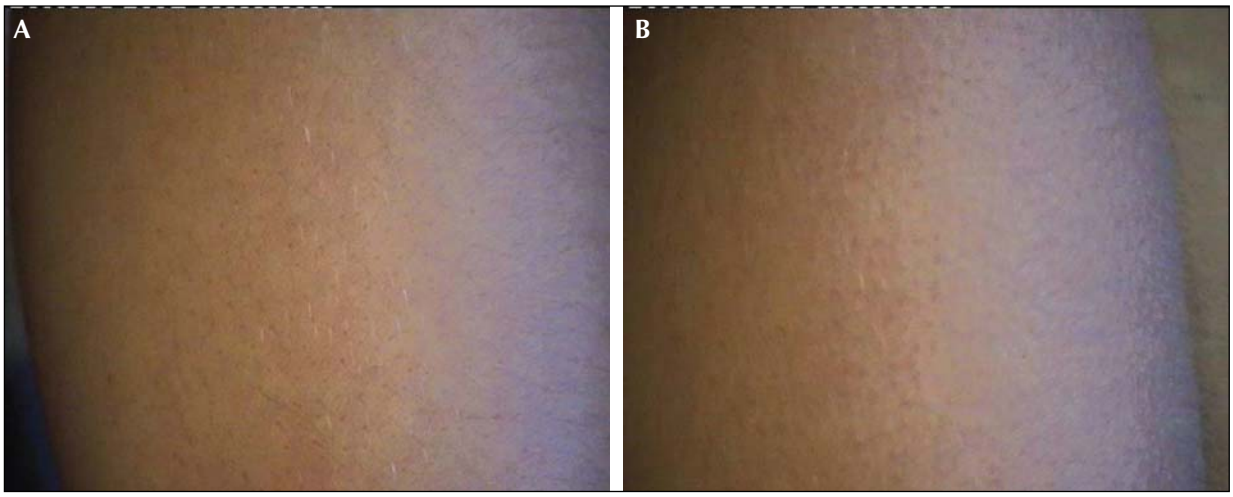

Figure 4. A) Skin of left arm of the patient before seizure. **B**) Skin of left arm of the patient during the seizure; piloerection observed.

Previous case series found that left hemispheric epilepsy is most frequent than right hemispheric epilepsy in patients with ictal piloerection or cold shiver (Stefan *et al.* 2002). This finding is still a subject of debate. Piloerection has also been observed in patients with right temporal epilepsy (Devinski *et al.* 2004).

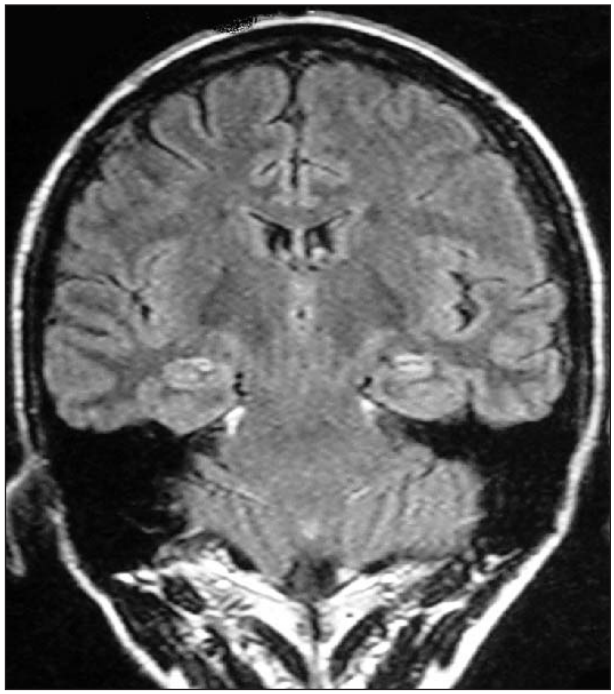

Figure 5. MRI performed two years after the onset of epilepsy. The patient is seizure-free. Clear decrease in the hyperintensity signal of left hippocampus in axial and coronal T2 and FLAIR-weighted images (perpendicular to hippocampic plan). The left hippocampus is atrophic.

No specific etiology has been found. Of the previous reported cases, etiology included tumor, post-traumatic contusion, hippocampal sclerosis or atrophy, tuberous sclerosis, cavernous angioma, temporal malformation, radionecrosis (Roze *et al.* 2000, Loddenkemper *et al.* 2004).

From our observation, the etiology of the epilepsy is unclear. Magnetic resonance imaging performed seven days after the onset of the disease showed increase signal intensity in the left hippocampus, in both T2 and FLAIR-weighted images. Follow-up MRI showed regression of this signal abnormality. Two years after the first seizure, while the patient became seizure-free, atrophy and a slightly increased signal intensity of the left hippocampus in T2 and FLAIR-weighted images were found. These abnormalities are not suggestive of an ischemic lesion. Also, a neoplastic lesion appears to be unlikely because of the spontaneous regression of the signal abnormalities with subsequent atrophy of the left hippocampus. By contrast, this finding might suggest neuronal loss (Lansberg *et al.* 1999, Meierkord *et al.* 1997) or changes in the hippocampus associated with frequent and daily seizures (Van Paesschen *et al.* 1998, Bernasconi *et al.* 2005). However we can't exclude infection or an inflammatory process because unfortunately CSF analysis was not performed at the onset of the disease (Suzuki *et al.* 1999).

Voltage-gated potassium channels antibodies were not assayed. These have been recently reported in cases of limbic encephalitis and other seizure-associated disorders. Nevertheless, our patient did not present with any clinical features of paraneoplastic or non-paraneoplastic limbic encephalitis (McKnight *et al.* 2005, Wieser *et al.* 2005, Vincent *et al.* 2004). □

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