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Short communication

Spontaneous seizure remission following status epilepticus in drug-resistant epilepsy due to focal cortical dysplasia

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ABSTRACT

We describe a patient with chronic pharmacoresistant epilepsy related to right parietal focal cortical dysplasia (FCD), who became seizure-free following an episode of convulsive status epilepticus (SE). Magnetic resonance imaging (MRI) and fludeoxyglucose positron emission tomography (FDG-PET) were performed before and after SE. Longitudinal MRI scans showed a stable appearance of the FCD with no new signal change. However, diffusion tensor imaging showed altered white matter fiber tract orientation in posterior cortices, especially in proximity to the lesion, at 3 years post-SE. FDG-PET showed more widespread hypometabolism 3 years after SE. The unusual occurrence of spontaneous seizure remission following SE in the context of FCD-related epilepsy, in association with neuroimaging evolution, suggests possible cerebral reorganization triggered by SE as a mechanism in this case.

1. Introduction

Focal cortical dysplasias (FCD) are typically associated with epilepsy that often begins in childhood, the majority of cases proving to be pharmacoresistant (Semah et al., 1998). Because of poor response to antiepileptic drug (AED) treatment, surgery is often required, and spontaneous remission of epilepsy without surgery in these cases is very rare (Callaghan et al., 2007).

We report the case of a patient with chronic drug-resistant epilepsy related to focal cortical dysplasia, in whom an episode of convulsive status epilepticus (SE) was followed by complete and sustained remission of seizures, with longitudinal modifications in neuroimaging.

2. Clinical case

A 27-year-old right handed woman first presented seizures as an infant aged 8 months with eyelid clonus. Seizure expression evolved to altered contact and hypotonic falls by age 18 months. A period of seizure freedom was achieved with vigabatrin, taken until age 6 years. The patient remained seizure free off anti-epileptic drugs (AED) till aged 9 years, at which time seizures re-appeared, mainly nocturnal, characterised by unpleasant cephalic sensation, pressure in throat then loss of consciousness with hyperkinetic movements of all 4 limbs and ocular clonus, often progressing to secondary generalised tonic-clonic seizures (GTCS). Cerebral magnetic resonance imaging (MRI) performed at age 14 years in 2004 demonstrated right parietal focal cortical dysplasia (FCD), likely Type II (Fig. 1A) and fludeoxyglucose positron emission tomography (FDG-PET) showed hypometabolism in the same region (Fig. 2A). Because of resistance to AED polytherapy, presurgical evaluation was undertaken, including stereoelectroencephalography (SEEG) in 2010, which showed interictal discharges in relation to the FCD. However, despite prolonged SEEG recording during 3 weeks, only one spontaneous seizure was recorded, which showed some non-habitual semiological features and no clear electrophysiological change preceding clinical onset. The result of SEEG was therefore considered inconclusive and insufficient to permit surgical planning. Although a second SEEG exploration was subsequently proposed the patient declined this, and treatment was thereafter based on medical therapy alone. Transient improvement in seizure frequency and severity was seen during the 2 months following SEEG, but her epilepsy remained

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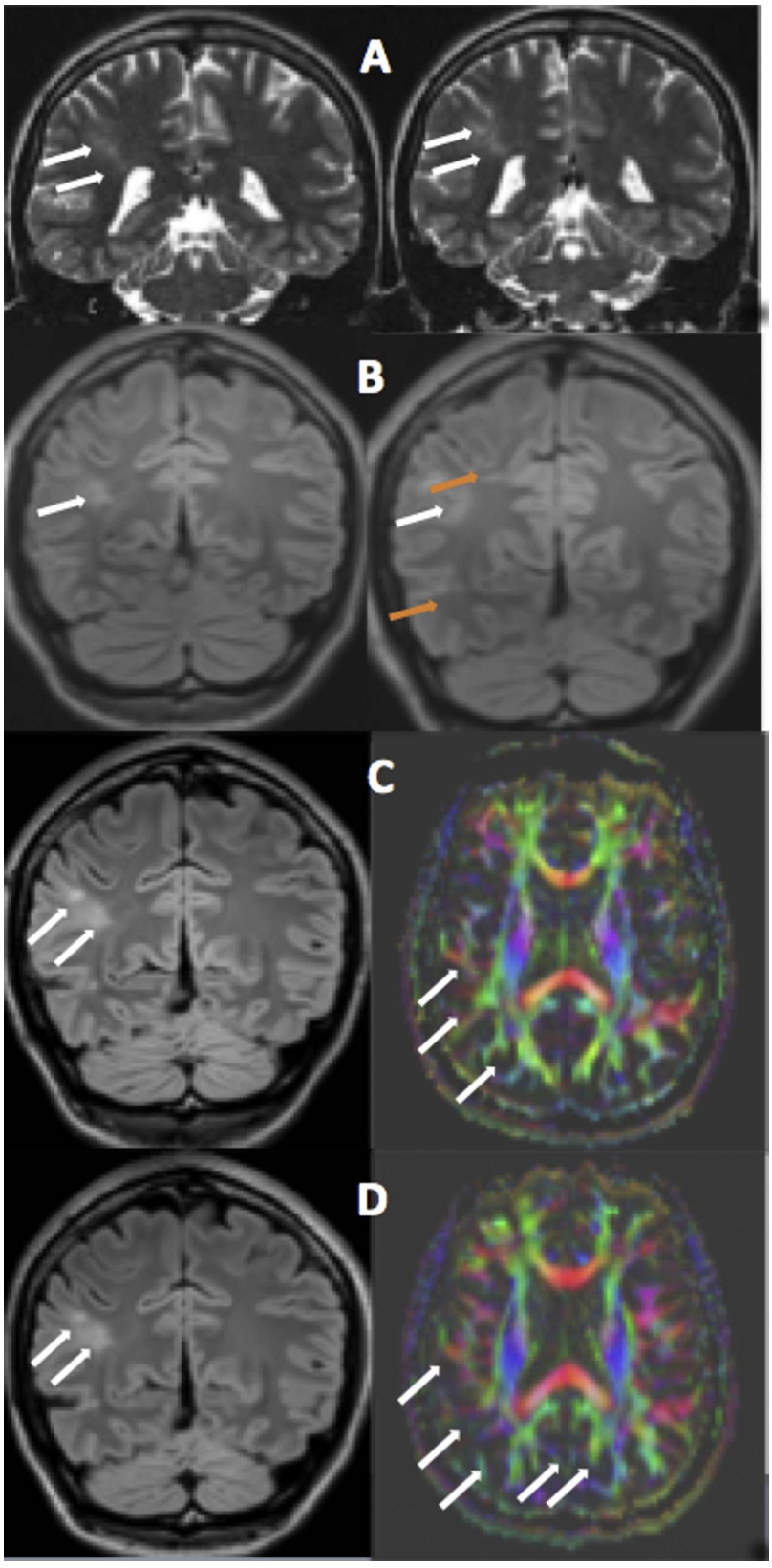

Fig. 1. A. One year prior to SEEG: coronal T2 images showing focal cortical dysplasia characterised by slightly increased signal that extends from the cortex to the lateral ventricle (white arrows). B Eighteen months after SEEG (before SE): coronal FLAIR images. Focal cortical dysplasia showing bright signal, similar to prior MRI (white arrow). Also note linear increased signal above the FCD, related to SEEG electrode (orange arrows). C One month after SE. Left image: coronal FLAIR image showing similar appearance of FCD. Right image: axial color FA image showing posterior asymmetry, with different orientation of white matter tracts underlying the cortical abnormality, compared to contralateral region. D Three years after SE. Images obtained with same machine and same acquisition parameters as image in Fig. 1C (Siemens Skyra 3T). Left image: coronal FLAIR image showing similar appearance of FCD. Right image: axial color FA image showing modification of the orientation of white matter tracts in right posterior cortex underlying the FCD but also contralateral mesial posterior cortex, compared to the previous MRI shown in Fig. 1C. The important feature to note is the change in position of colored fibers between the 2 images, representing altered orientation of white matter tracts, suggesting evolution over time. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

active with seizure frequency of several nocturnal seizures per month, often secondary GTCS. She also developed fairly disabling migraines around this time. While baseline cognitive function was normal, her epilepsy severity required that stopping her university course. Multiple unsuccessful combinations of AED were tried, the most efficacious being topiramate and vigabatrin. Topiramate had a beneficial effect on migraine frequency. No period of seizure freedom of more than 14 days occurred between ages 9 and 24 years.

In 2013 at the age of 24 years, during a febrile illness, the patient presented a cluster of nocturnal GTCS evolving to generalised