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Case Report

A patient with a 44-year history of epilepsia partialis continua caused by a perirolandic cortical dysplasia

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Abstract

Epilepsia partialis continua (EPC), or Kojevnikov's syndrome, is a rare epileptic syndrome arising from a variety of lesions in the perirolandic area. We report herein a 46-year-old woman with drug-resistant EPC due to a cortical dysplasia in the left frontoparietal region. For 44 years she has suffered continuous right-sided jerks, particularly in the right arm and hand, with an average frequency of 10–20 jerks per minute. During EEG recordings her jerks were associated with spikes and sharp waves over the left frontocentroparietal region, sometimes also with bursts of high-voltage generalized spike–wave complexes with a maximum bicentrally, followed by an electrodecrement. Despite the continuous jerks she is independent in daily life activities, and she considers the jerks not severe enough to justify surgery, i.e., multiple subpial transections.

Keywords: Epilepsia partialis continua; Kojevnikov's syndrome; Focal cortical dysplasia

1. Introduction

Persisting simple partial seizures with focal motor symptoms have been named epilepsia partialis continua (EPC), or Kojevnikov's syndrome [1]. Such an ongoing epileptic condition may arise from lesions not only in the primary motor cortex (Brodmann's areas 4 and 6), but also in the postcentral gyrus (Brodmann's areas 1, 2, and 3). This is due to the fact that a considerable portion (~40%) of corticospinal and corticobulbar fibers originate from the parietal lobe [2]. EPC may have different etiologies, infections, tumors, and cerebrovascular events being the most common [3]. The persistent or intermittent stereotyped, focal or regional, irregular clonic jerks usually last hours, weeks, months, and even years [3,4]. Prognosis and treatment are determined mainly by the nature of the underlying lesion.

Here we present a unique patient with a large perirolandic cortical dysplasia giving rise to drug-resistant EPC that has been ongoing for 44 years.

2. Case report

The patient is a woman born in 1958 after an uneventful pregnancy and labor. She had delayed psychomotor development; she was not able to sit before she was 1 year old, and she could not walk alone until she was 3 years old. When attending school at 7 years of age, she wrote with her left hand. She was apparently weak and clumsy with her right hand, and in gymnastics and in children's play she was somewhat slow and unsteady, particularly in the terrain. She completed 9 years of compulsory school, but her school achievements were poor.

After school she was a factory worker for 25 years, but for the last 2 years has been unemployed. She is married and has given birth to two healthy children.

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From 6 months to 2 years of age, she had recurrent febrile convulsions, about which there is no information on focality. At 2 years of age she developed frequent, afebrile right-sided clonic jerks. These seizures were not accompanied by loss of consciousness. Ever since she has had persisting right-sided focal motor seizures daily. The jerks do not occur during sleep. Her jerks are most prominent in the right hand and arm, but occasionally may affect the right leg and face and, very seldom, the left side. Routinely, the frequency of the jerks averages 10/minute. However, when she is stressed, deprived of sleep, or in the presence of flickering light, the frequency increases to 20–30 jerks/minute. She has never had a secondary generalized tonic–clonic seizure. In her daily life she uses mainly her left hand. When she uses her right hand to carry objects, she does not drop them, but she may spill coffee, soup, etc.

Her ongoing jerks have been drug resistant. Despite doses of phenobarbital, phenytoin, carbamazepine, and topiramate giving serum concentrations in the upper therapeutic range, her clinical situation has been unaffected. However, an attempt to withdraw carbamazepine, a drug currently in use, resulted in exacerbation of the seizures. During an EEG recording, she received 2 mg clonazepam rectally without any apparent effect, neither clinically nor electroencephalographically.

A clinical examination disclosed a modest hypotrophia with a central paresis of her right arm and hand, and a slight ataxia assessed by Romberg's test. Her language was unaffected. Twenty-six-channel digital scalp EEGs and video telemetry revealed high-voltage (11–12 Hz) background activity with a higher amplitude over the left hemisphere than the right, and with an unusual maximum over the anterior regions. Over the left frontocentroparietal region there were frequently occurring spikes and sharp waves. From time to time, bursts of generalized high-voltage spike–wave complexes appeared, with a maximum in the frontocentral regions, often followed by an electrodecrement. These electroencephalographic events corresponded to jerks in the patient's right arm and hand (Fig. 1). Our clinical neurophysiologist (H.K.) claimed that these EEG abnormalities strongly indicated an underlying focal cortical dysplasia (FCD).

Conventional MRI revealed a "masslike" lesion involving the left frontoparietal region, with an inhomogenous hyperintense signal in the subcortical white matter tapering toward the ventricle on fluid-attenuated inversion recovery (FLAIR), as shown in Fig. 2A. Coro-

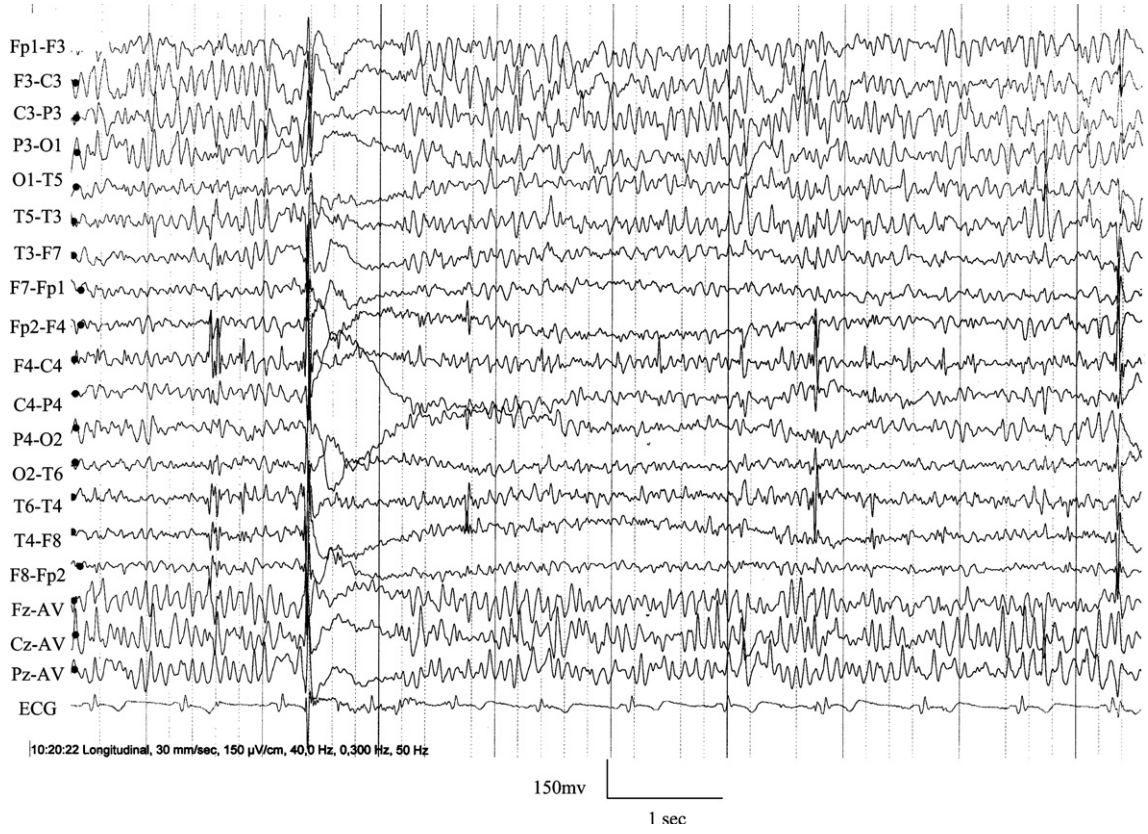

Fig. 1. This EEG from the patient with long-lasting epilepsia partialis continua shows amplitude asymmetry, with higher amplitude over the left frontocentroparietal region, compared with the right one. In addition to spikes and sharp waves in the left frontocentroparietal regions, there are bursts of generalized high-amplitude spike–wave complexes, followed by electrodecrement. These bursts corresponded to myoclonic jerks in the patient's right hand. Note that sample is shown at reduced sensitivity, 150 µV/cm.