Electroclinical reasoning report

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Epilepsia partialis continua triggered by traumatic hand injury: a peripheral tuning of brain excitability?

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Introduction

Epilepsia partialis continua (EPC) is characterized by continuous focal jerking of a body part, usually the hemiface or a limb, lasting hours, days or years (Guerrini, 2009). In the absence of a gross focal lesion, microscopic dysplasia should be suspected (Desbiens et al., 1993). It is recognized that focal cortical dysplasia (FCD) in the rolandic cortex may remain dormant for decades, awaiting a trigger to generate EPC (Hemb et al., 2014). Once triggered, however, the continuous jerking may never stop. Here, we address these issues reporting the case of a man who developed EPC following a peripheral hand injury.

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Case report

A 50-year-old man presented with a 3-week history of continuous myoclonic jerking of the left upper limb, less than 24 hours after a traumatic injury to his left hand. Continuous jerking worsened with repetitive voluntary movements, but never disappeared, even during sleep (video S1), without response to medical treatment.

Ten years earlier, the patient had a right temporal lobectomy for refractory complex partial seizures, precipitated by an unpleasant feeling in the left hand. Surgery significantly improved the complex partial seizures, but somatosensory auras persisted.

Despite negative initial investigation, the patient underwent awake epilepsy surgery. Acute ECoG revealed continuous, rhythmic spiking in the sensorimotor cortex, predominating in the bottom of the central sulcus, which was resected. Histopathology showed abnormal radial lamination and neuronal distribution in micro-columns, which was compatible with FCD IA. Movements of the hand completely recovered after seven days. Two weeks after surgery, EEG showed runs of electrographic seizures in the right temporal lobe, which had never been seen before rolandic resection.

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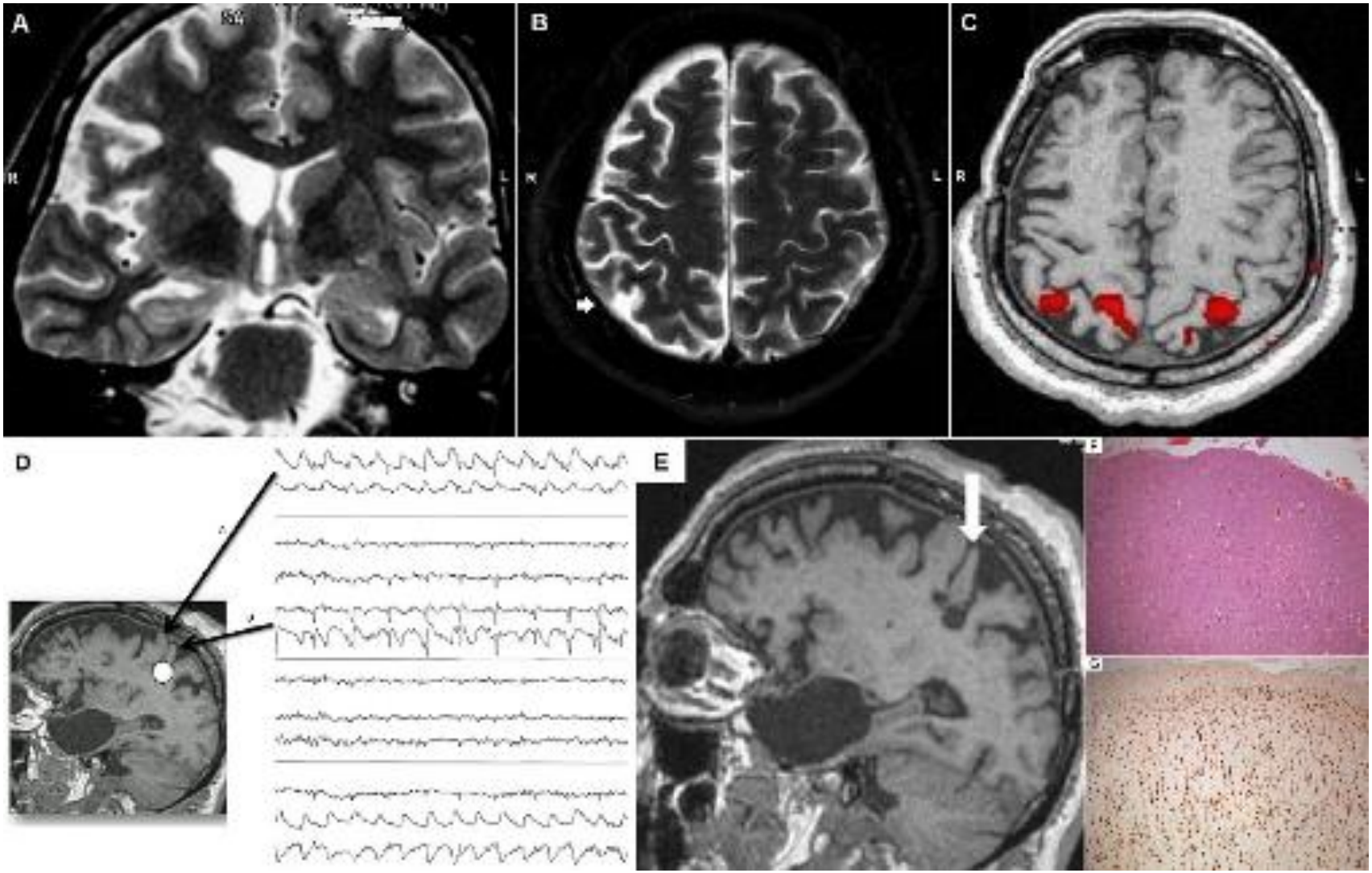

Figure 1.

A. MRI T2-weighted coronal image before the first surgery with right hippocampal hyperintensity convergent with interictal and ictal scalp EEG. Note the enlarged right lateral ventricle and prominence of right hemisphere sulci;
B. MRI T2-weighted axial image 10 years later, when EPC began. Note persistent right hemisphere atrophy, particularly of the parietal cortex (arrow);
C. Axial functional MRI during EPC demonstrating intense bilateral primary sensory cortex activation.
D. Electrocorticography with continuous spike-wave activity over the right motor (arrow A) and sensory areas (arrow B), predominating on the depth of central sulcus (white circle).
E. Sagittal T1-weighted image post-resection of the depth of central sulcus (arrow).
F. F, G) Photomiographs. H & E and NeuN stains, respectively, showing abnormal radial lamination and neuronal distribution in micro-columns at 50x.

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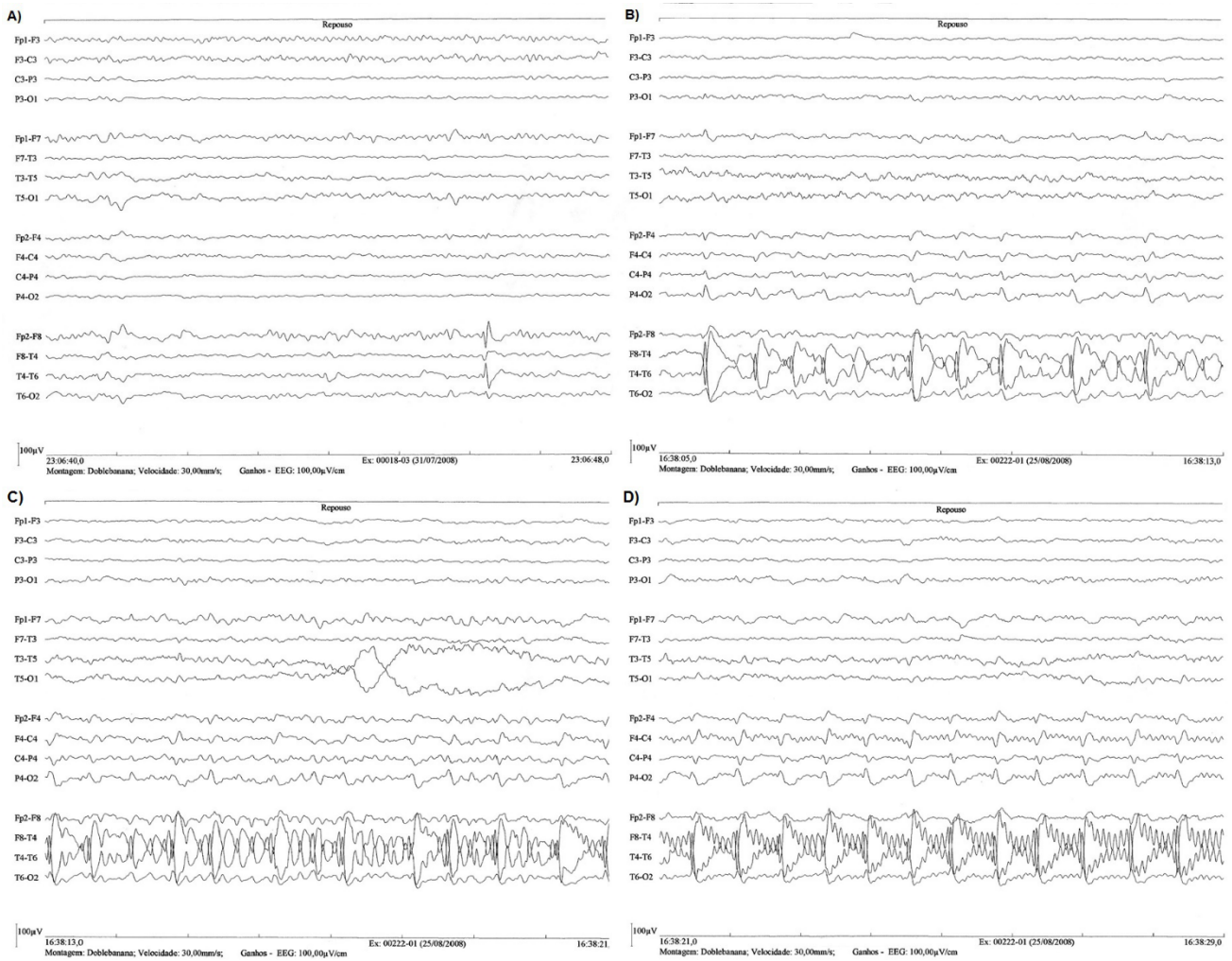
Figure 2.

A) Pre-resection scalp EEG at EPC presentation, with remaining interictal right temporal discharges. Note that rolandic regions are electrically silent.

B-D) Sequential sections of scalp EEG after rolandic resection and full control of the EPC. Electrographic seizures are seen in the right temporal lobe, without clinical symptoms. This abnormality was not present before resection.



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Discussion

Reflex seizures following peripheral trauma in previously damaged brain have been rarely reported (Parsons-Smith 1948, Rae 1952, Spiller et al., 2005). Whether augmented cortical hyperexcitability following peripheral injury occurs only in the presence of already abnormal cortex is not clear and the mechanisms by which peripheral injury would trigger continuous seizures are not understood

The fact that in this patient EPC began only after peripheral injury allows us to hypothesize a delicate balance between intrinsic hyperexcitability and intrinsic inhibition in neuronal networks, such as those involved in FCD.

Once disinhibited, re-entrant synaptic loops between motor and sensory neurons may play a major role and in the patient reported here even subtle sensory stimulation or movements led to immediate exacerbation of myoclonic jerks

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Interestingly, EEGs after resection of the previously silent, dysplastic sensori-motor cortex unexpectedly showed recurrent electrographic seizures in the remaining parts of the ipsilateral, previously operated temporal lobe (figure 2, B-D). This added complexity to the pattern of disinhibition, in that a peripheral injury disinhibited a likely rolandic FCD, the resection of which disinhibited the ipsilateral temporal neocortex – indeed supporting the view that inhibition plays a central role in the clinical presentation of FCD-related networks in epilepsy (Palmini 2010). Despite the predominant view that FCD is a hyperexcitable lesion with impaired inhibition (Ferrer et al., 1992), emerging basic science data support a role of inhibition in FCD circuits that could be disrupted by surgery or massive afferent volleys. For instance, significant increase in evoked and spontaneous inhibitory postsynaptic currents was found in human dysplastic tissue, likely reflecting a decrease in transporter-mediated GABA reuptake (Calcagnotto et al., 2005)

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Conclusion

In conclusion, this patient illustrates that peripheral injuries produce afferent stimuli that could trigger seizures in dormant cortical dysplasia. This observation suggests that excitation and inhibition act in tandem in dysplastic circuits, which are vulnerable to a number of interferences that, in turn, modulate epileptogenicity.

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