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Teaching Video NeuroImage: Reflex Seizures Mimicking Paroxysmal Dystonic Movements in a Patient With
Late-Onset Rasmussen Encephalitis

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Contributions:

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Case report

A 35-year-old right-handed man with late-onset Rasmussen encephalitis¹ involving the right hemisphere reported focal aware seizures with motor onset, rare focal to bilateral tonic-clonic seizures, and epilepsy partialis continua to the left upper limb (eAppendix 1 and eFigures 1–3 in the Supplement).

Over time, a new seizure type mimicking dystonic posturing of the left arm became recurrent (Video 1), consistently triggered by voluntary movements of the limb.

The EEG-polygraphic recording showed fast activity in the right central region associated with the clinical seizure. Time-varying cortico-muscular coherence (CMC) analysis, a method commonly applied to evaluate the functional connection between the cortex and muscles during muscle contraction, helped us identify the pattern of the paroxysmal dystonic episodes as reflex focal aware seizures (Figure 1), probably evoked by abnormal afferents to the right sensorimotor cortex during voluntary muscle activation.

As expected in this immune-mediated brain disorder, reflex seizures poorly responded to various ASMs, while periodic IV immunoglobulin administration resulted in transient beneficial effect.

Video 1 title: Video-EEG with polygraphic recording

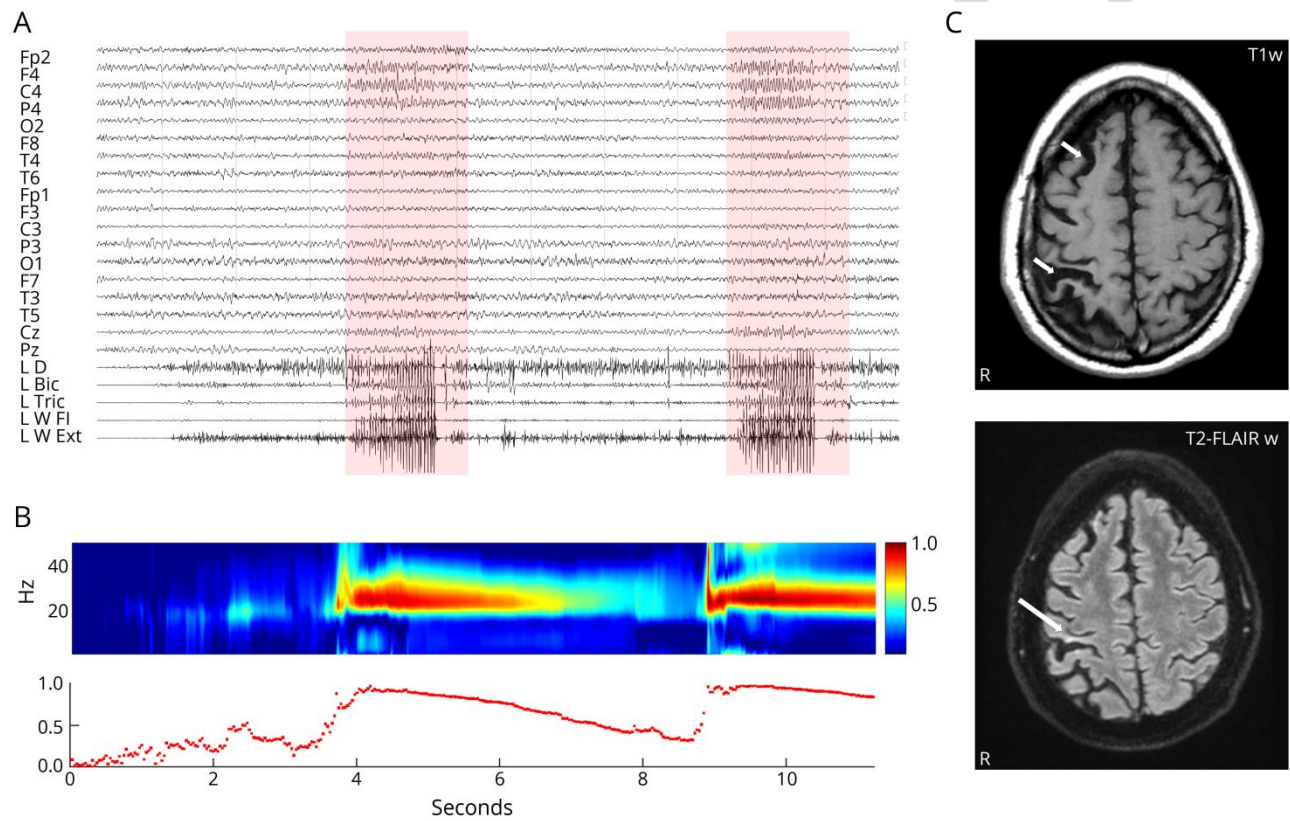
Video 1 Legend

Left deltoid (EMG1), biceps (EMG2), triceps (EMG3), flexor (EMG4), and extensor carpi (EMG5) muscles. Reflex seizures (triggered by voluntary arm lifting) with dystonic features characterized by ulnar deviation and flexion of the wrist, flexion and internal rotation of the forearm. Red arrows indicate fast activity over the right central region.

Figure 1 title: EEG-polygraphic recording, CMC analysis and brain MRI

Figure 1 legend

A: Two reflex seizures over the right central leads associated with muscular bursts (boxes).
B: Sudden increase of C4 / left wrist flexor muscles CMC² during voluntary movement to seizure shift.
C: Brain MRI shows (top) right hemisphere atrophy and (bottom) signal hyperintensity in the right postcentral gyrus (arrows).



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