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

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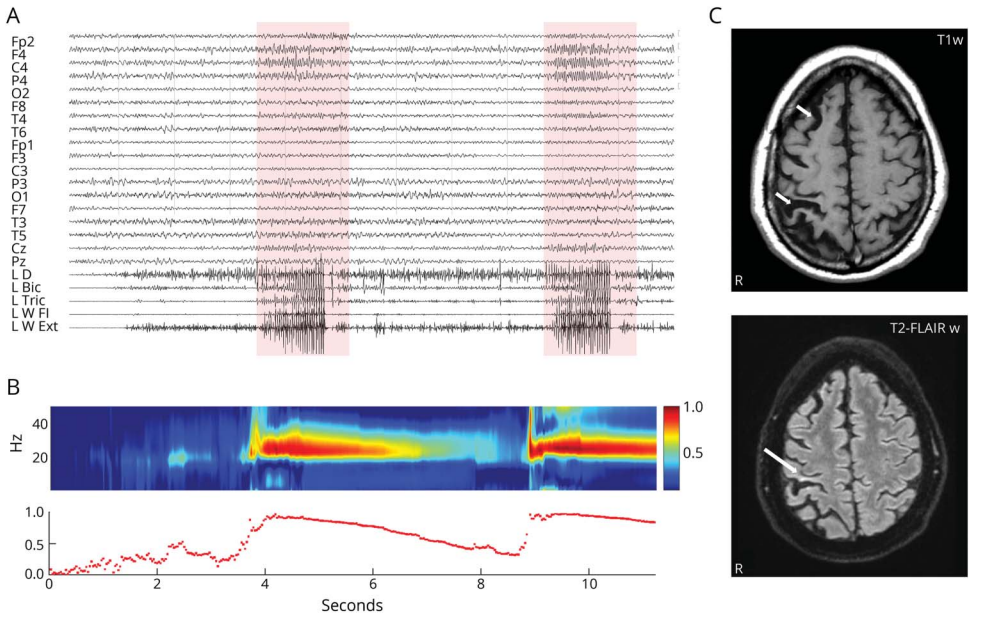
Figure 1 EEG-Polygraphic Recording, CMC Analysis, and Brain MRI



(A) Two reflex seizures over the right central leads associated with muscular bursts (boxes). (B) Sudden increase of C4/left wrist flexor muscles CMC[2] during voluntary movement to seizure shift. (C) Brain MRI shows (top) right hemisphere atrophy and (bottom) signal hyperintensity in the right postcentral gyrus (arrows). CMC = corticomuscular coherence.

Case Report

A 35-year-old right-handed man with late-onset Rasmussen encephalitis[1] involving the right hemisphere reported focal aware seizures with motor onset, rare focal-to-bilateral tonic-clonic seizures, and epilepsia partialis continua to the left upper limb (eAppendix 1 and eFigures 1–3, links.lww.com/WNL/C831). 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 corticomuscular coherence 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

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Video

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(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 antiseizure medications while periodic IV immunoglobulin administration resulted in a transient beneficial effect.

Author Contributions

A. Stabile: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; analysis or interpretation of data. S. Franceschetti: drafting/revision of the manuscript for content, including medical writing for content; study concept or design; analysis or interpretation of data. F. Deleo: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; analysis or interpretation of data. R. Di Giacomo: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; analysis or interpretation of data. G. Didato: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; analysis or interpretation of data. C. Pastori: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; analysis or interpretation of data. F. Panzica: drafting/revision of the manuscript for content, including medical writing for content; analysis or interpretation of data. M. De Curtis: drafting/revision of the manuscript for content, including medical writing for content; study concept or design; analysis or interpretation of data. F. Villani: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; analysis or interpretation of data. L. Canafoglia: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; analysis or interpretation of data.

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References

1. Doniselli FM, Deleo F, Criscuolo S, et al. MRI in late-onset Rasmussen encephalitis: a long-term follow-up study. *Diagnostics*. 2022;12(2):502. doi:10.3390/diagnostics12020502
2. Panzica F, Canafoglia L, Franceschetti S. EEG-EMG information flow in movement-activated myoclonus in patients with Unverricht-Lundborg disease. *Clin Neurophysiol*. 2014;125(9):1803-1808. doi:10.1016/j.clinph.2014.01.005

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