
Neurology Publish Ahead of Print
DOI:10.1212/WNL.0000000000207412

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

Author(s):

Andrea Stabile, MD¹; Silvana Franceschetti, MD, PhD¹; Francesco Deleo, MD¹; Roberta Di Giacomo, MD¹; Giuseppe Didato, MD¹; Chiara Pastori, MD¹; Ferruccio Panzica, MSc²; Marco De Curtis, MD¹; Flavio Villani, MD³; Laura Canafoglia, MD¹

Corresponding Author:

Francesco Deleo, franc.deleo@gmail.com

Affiliation Information for All Authors: 1 Epilepsy Unit, Fondazione IRCCS Istituto Neurologico Carlo Besta, Member of the ERN EpiCARE, Milan, Italy; 2 Clinical Engineering, Fondazione IRCCS Istituto Neurologico Carlo Besta, Milan, Italy; 3 Division of Clinical Neurophysiology, IRCCS Ospedale Policlinico San Martino, Genova, Italy

Neurology® Published Ahead of Print articles have been peer reviewed and accepted for publication. This manuscript will be published in its final form after copyediting, page composition, and review of proofs. Errors that could affect the content may be corrected during these processes. Videos, if applicable, will be available when the article is published in its final form.

Equal Author Contribution:

Contributions:

Andrea 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

Silvana Franceschetti: Drafting/revision of the manuscript for content, including medical writing for content; Study concept or design; Analysis or interpretation of data

Francesco 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

Roberta 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

Giuseppe 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

Chiara 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

Ferruccio Panzica: Drafting/revision of the manuscript for content, including medical writing for content; Analysis or interpretation of data

Marco De Curtis: Drafting/revision of the manuscript for content, including medical writing for content; Study concept or design; Analysis or interpretation of data

Flavio 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

Laura 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

Figure Count:

1

Table Count:

0

Search Terms:

[284] EEG; see Epilepsy/Seizures (S), [64] Epilepsy semiology, [69] Video/ EEG use in epilepsy, [132] Autoimmune diseases, [162] Dystonia

Acknowledgment:**Study Funding:**

This work was supported supported by the Italian Ministry of Health (RRC).

Disclosure:

The authors report no relevant disclosures.

Preprint DOI:**Received Date:**

2022-09-18

Accepted Date:

2023-03-30

Handling Editor Statement:

Submitted and externally peer reviewed. The handling editor was Resident & Fellow Section Editor Whitley Aamodt, MD, MPH.

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

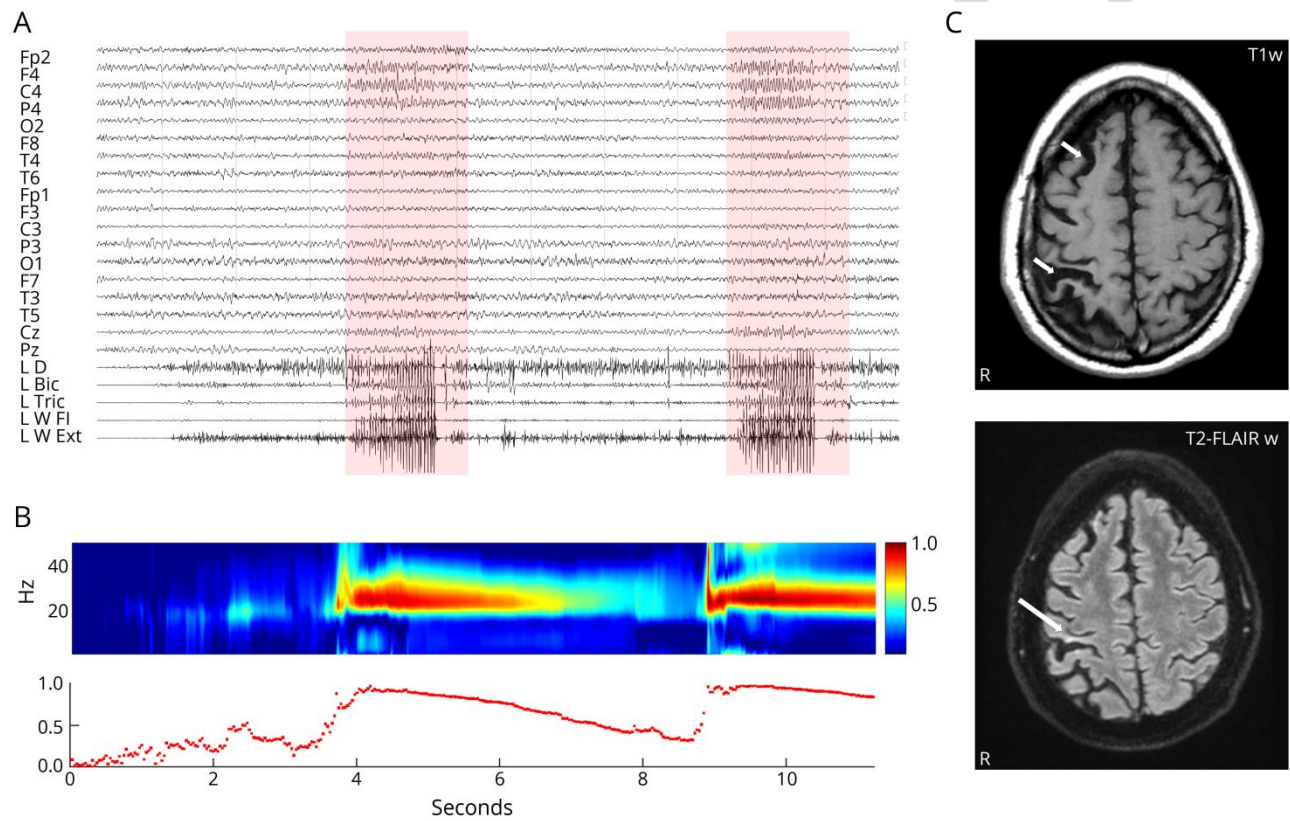
Video 1 Legend

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).



WNL-2023-000254_slides ---<http://links.lww.com/WNL/C830>

WNL-2023-000254_sup ---<http://links.lww.com/WNL/C831>

WNL-2023-000254_vid1 --- <http://links.lww.com/WNL/C832>

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):1803-8. doi: 10.1016/j.clinph.2014.01.005.

ACCEPTED

Neurology®

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

Andrea Stabile, Silvana Franceschetti, Francesco Deleo, et al.

Neurology published online May 18, 2023

DOI 10.1212/WNL.0000000000207412

This information is current as of May 18, 2023

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/early/2023/05/18/WNL.0000000000207412.citation.full
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Autoimmune diseases http://n.neurology.org/cgi/collection/autoimmune_diseases Dystonia http://n.neurology.org/cgi/collection/dystonia EEG; see Epilepsy/Seizures http://n.neurology.org/cgi/collection/eeg_see_epilepsy-seizures Epilepsy semiology http://n.neurology.org/cgi/collection/epilepsy_semiology Video/ EEG use in epilepsy http://n.neurology.org/cgi/collection/video_eeg_use_in_epilepsy
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2023 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

