Teaching Video NeurolImage: Clenched Fists as an Unusual Presentation of Focal Neuromyotonia

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Figure 1 Active Finger Extension From the Resting Clenched Fist State

(A–C) Remarkably delayed and incomplete relaxation (pseudomyotonia) of forearm wrist/finger flexors before treatment, taking approximately 12 seconds to complete the maneuver. (D, E) Significant improvement to 4 seconds following treatment observed in both hands; only left hand shown.

A 72-year-old woman with a history of asthma and bronchiectasis presented with a 7-year history of bilateral hand cramps, initially affecting the right, frequently resulting in clenched fists. Examination revealed clenched fists bilaterally in the resting state and pseudomyotonia with incomplete relaxation following active finger extension (figure 1, A–C; video 1). Myokymia and percussion myotonia were not observed. No weakness was elicited in the muscles. Needle EMG of the forearm flexors demonstrated bursts of spontaneous high-frequency waning discharges typical of neuromyotonia (figure 2; video 2). Significant clinical improvement was observed following treatment with botulinum toxin injections and oxcarbazepine (figure 1, D–F).

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Figure 2 Neuromyotonic Discharges

Spontaneous bursts of high-frequency (120–180 Hz) waning amplitude discharges on needle EMG of flexor carpi ulnaris muscle.

The current case illustrates an unusual but treatable presentation of the rare focal form of neuromyotonia,^{1,2} and should be differentiated from other conditions that may have similar presentations such as myotonic dystrophy and other nondystrophic myotonias, ulnar neuropathies, and Dupuytren contracture, the latter of which may result in unnecessary surgical intervention. As with the current case, patients previously reported with this condition tend to be older women with a history of chronic obstructive pulmonary disease (COPD) managed by inhaled β_2 sympathomimetics. Whereas the pathomechanisms remain elusive, enhanced axonal hyperexcitability secondary to COPD-induced hypoxemia as well as hyperpolarization from sympathomimetics, through their effects on voltage-gated Na⁺/K⁺ pumps, may result in ectopic firing in high-frequency

bursts.^{1,2} Other cases have also been found in association with a more generalized voltage-gated potassium channel antibody autoimmunity,³ although these antibodies were absent in the current case.

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Disclosure

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Appendix Authors

Name	Location	Contribution
Roshan Dhanapalaratnam, MBBS	Institute of Neurologic Sciences, Prince of Wales Hospital	Designed and conceptualized manuscript, drafted manuscript
Raymond Schwartz, PhD, MBBS	Southern Neurology, Kogarah; Sydney University, Australia	Patient management, final edit of manuscript
William Huynh, PhD, MBBS, BSc	Brain and Mind Centre, University of Sydney; Southern Neurology, Kogarah, Australia	Design and conceptualized manuscript, patient management, drafted and final edit of the manuscript

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