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

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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 (Fig1A-C, Video1). Myokymia and percussion myotonia were not observed. No weakness was elicited in all muscles. Needle EMG of the forearm flexors demonstrated bursts of spontaneous high-frequency waning discharges typical of neuromyotonia (Fig2, Video2). Significant clinical improvement was observed following treatment with botulinum-toxin injections and oxcarbazepine (Fig1D-F). The current case illustrates an unusual but treatable presentation of the rare focal form of neuromyotonia, and should be differentiated from other conditions that may have similar presentations such as myotonic dystrophy and other non-dystrophic myotonias, ulnar neuropathies and Dupuytren's contracture – the latter which may result in unnecessary surgical intervention. As with the current case, patients previously reported with this condition tend to be older females with a history of COPD managed by inhaled beta-2-sympathomimetics. Whilst 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. Other cases have also been found in association with a more generalised VGKC-antibody autoimmunity although these antibodies were absent in the current case.

Teaching Slides-http://links.lww.com/WNL/B378

Video 1-http://links.lww.com/WNL/B379

Video 2-http://links.lww.com/WNL/B380
References


Figure and Video Legend

Figure/Video 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 manoeuvre; (D-E) Significant improvement to 4s following treatment observed in both hands; only left hand shown.
Figure/Video 2. Neuromyotonic discharges. Spontaneous bursts of high frequency (120-180Hz) waning amplitude discharges on needle EMG of flexor carpi ulnaris muscle.

Appendix 1: Authors

<table>
<thead>
<tr>
<th>Name</th>
<th>Location</th>
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</tr>
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<tbody>
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