Teaching Video NeuroImages: Cold-induced laryngeal pseudomyotonia in Isaacs syndrome

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A 36-year-old man developed progressive limb pain, excessive sweating, and muscle cramps. He described transient strained voice only after drinking cold water (video 1). Examination revealed generalized myokymia and pseudomyotonia. EMG revealed multiple spontaneous, continuous high frequency doublet and triplet motor unit discharges. Nerve conduction studies and chest CT scan were normal. CASPR2 and LGI1 antibodies were negative. Autoimmune, paraneoplastic, and other etiologies were excluded clinically as well as through additional serologic (or blood) testing and imaging. Symptoms resolved completely after IV immunoglobulin and carbamazepine for treatment of pain.

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Video

Teaching slides
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Painful cramps, myokymia, hyperhidrosis, and pseudomyotonia are cardinal features of Isaacs syndrome. Isaacs syndrome is a peripheral nerve hyperexcitability disorder often associated with antibodies targeting components of voltage-gated potassium channels; however, <50% of patients are antibody-positive.[1] Bulbar involvement is an uncommon feature, and our patient exhibits cold-induced pseudomyotonia, which has not been previously described in the literature.[2]

Author contributions

H. Morales-Briceño and J. Renan Perez drafted the manuscript and videotape. B. Balint and V.S.C. Fung examined the video of the patient and provided critical review of the manuscript.

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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