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Tremor and Other Hyperkinetic Movements

Case Reports

A Novel Presentation of an Ocular Geste Antagoniste in Cervical Dystonia: A Case Report

James T. Boyd*, Timothy J. Fries, Keith J. Nagle & Robert W. Hamill

Department of Neurological Sciences, University of Vermont College of Medicine, Burlington, Vermont, United States of America

Abstract

Background: A geste antagoniste or sensory trick is a well described phenomenon associated with primary cervical dystonia. Craniocervical tactile stimulation or stereotyped limb movements allow patients to transiently ameliorate dystonic activation of cervical musculature.

Case Report: We report a patient with primary cervical dystonia who presented with a novel "ocular" geste antagoniste. Through a sensory trick of tonic left eye deviation, the patient transiently reduces cervical dystonic activity (improved range of motion and reduced dystonic tremor). Multi-channel surface electromyography and video are used to illustrate these findings.

Discussion: This case presents a unique clinical observation of specific voluntary eye movements attenuating cervical dystonia. The phenomenon is phenotypically consistent with previously described limb sensorimotor tricks.

Keywords: Cervical dystonia, sensory trick, geste antagoniste, ocular, oculomotor

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*To whom correspondence should be addressed. E-mail: james.boyd@vtmednet.org

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Introduction

Cervical dystonia represents one of the most common forms of primary focal dystonia. A geste antagoniste or sensory trick is a well described phenomenon associated with cervical dystonia. Craniocervical tactile stimulation or stereotyped limb movements allow patients to transiently ameliorate dystonic activation of cervical musculature. We report a unique case of a patient with cervical dystonia capable of attenuating dystonic symptoms through left gaze deviation, in a pattern consistent with previously described sensory tricks.

Case report

A 50-year-old man presented with a 12-year history of primary cervical dystonia. Initial examination at our movement disorders center revealed a 10° left rotation, hypertrophy, and over activation of the right sternocleidomastoid (SCM) and left splenius capitis (SC) muscles, and horizontal head tremor. Saccadic and pursuit eye movements were normal. A mild increase in blink frequency was observed without other evidence of blepharospasm. A null point of dystonic activity was present with greater than 20° of left rotation from the midline. The patient has achieved sustained benefits from quarterly onabotulinum toxin injections.

During the first 4 years of symptoms, the patient benefited from the use of a geste antagoniste with light tactile stimulation against the chin. Our astute patient later identified a unique trick to reduce his dystonic symptoms. Through visual fixation on the left upper rim of his eye glasses, the patient experienced a reduction in subjective neck tightness, improved ability to turn right, and reduced head tremor. Benefits were lost if the patient lost fixation and/or changed direction of gaze.

Informed consent was obtained from the patient. Electrophysiological evaluation of the phenomenon was performed utilizing simultaneous video, multi-channel surface electromyography (EMG), and peri-ocular surface electrodes (electro-oculography [EOG]). Recordings were

performed on NCI (Network Concepts, Inc.). Madison, Wisconsin, USA. Dimensions video-digital electroencephalography equipment. EOG and EMG tracings were filtered at 0.5 Hz LFF (low frequency filter) and 70 Hz HFF (high frequency filter). Muscle activity was referenced to a common average reference that included bilateral mastoids and the cranial vertex. The patient performed kinetic and isotonic craniocervical muscle activation in a variety of situations, including with and without the use of the described ocular maneuver and extreme lateral tonic eye deviations (with and without eye closure). Potential asymmetries in the amplitude of cervical muscle activity were anticipated due to the effects of therapeutic injections of onabotulinum toxin within 12 weeks of the recordings.

During maximal right head turning (position of maximal dystonic symptoms), the patient was asked to fixate gaze to both lateral extremes. A change in gaze fixation toward the left was associated with relative reduction in firing of dystonic agonist (right SCM and left SC) and antagonist muscles. Similar reductions in firing were identified with the eyes closed and sustained eye deviation (confirmed with peri-ocular surface electrodes) (Figure 1). A reduction in phasic EMG firing and clinical reduction in tremor were also identified during right turning with left eye deviation. The time required to deviate the head from mid-position to a maximal right turn was reduced to 0.6–0.9 seconds from 2.0 to 3.0 seconds (Video 1). Our patient did not gain clinical benefit or reduced EMG activity from thinking about left visual fixation or deviation of the eyes, a phenomenon previously demonstrated with gestes involving the limbs.[1] The maximal amplitude of surface EMG activity on the right SCM and left splenius capitis were consistently reduced relative to their contralateral counterparts, presumably due to previous botulinum toxin injections.

Discussion

The "ocular geste antagoniste" showed many of the accepted clinical and neurophysiological findings previously described with gestes in cervical dystonia. Electrophysiological evaluation revealed that the effectiveness of the geste was dependent on ocular position and not visual input. Our patient presented with a novel maneuver to reduce dystonic symptoms that in many ways paralleled the well-described geste antagoniste.[2–4] Previous studies of gestes have shown an associated clinical reduction in tonic and phasic muscle contraction,

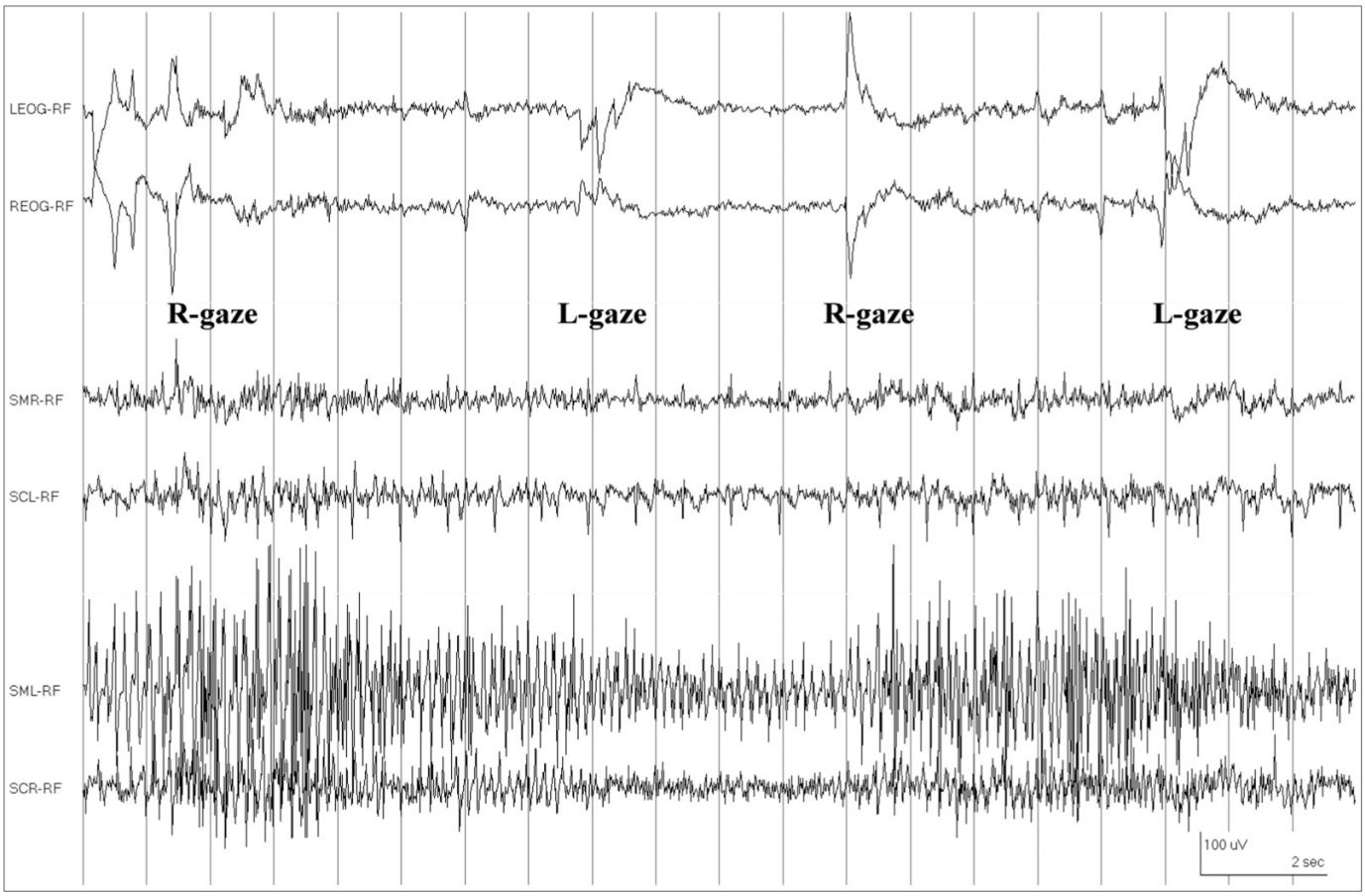

Figure 1. **Peri-ocular and multi-channel surface electromyography are recorded during isotonic right head turning, eye closure, and alternating gaze.** A relative reduction in amplitude of cervical muscle activity is demonstrated in association with left gaze. Right (R), left (L), electro-oculography (EOG), sternocleidomastoid (SM), splenius capitis (SC), average reference (RF).

Video 1. **Ocular Geste Antagoniste in Cervical Dystonia.**

reduction in EMG activity, and the immediate return of symptoms following cessation.[3]

Several potential mechanisms of action are considered when discussing the effect of eye movements on cervical dystonia. Though exquisitely integrated with vestibular, visual, and craniocervical proprioceptive systems, extraocular muscle activity is generally thought of as a point of efferent action.[5–9] The closed feedback loop of the vestibulo-ocular and cervico-ocular reflexes allows for rapid synchronization to maintain stabilization of visual input. The open feedback loop of visual processing and resultant correction of saccadic and optokinetic eye movements allows visual points of interest to remain on the fovea.[5,6] Less well described is the suspected existence of oculomotor proprioceptive afferent pathways, capable of generating excitatory and inhibitory effects on cervical muscle activity.[10,11] Human and animal studies have shown an association of voluntary horizontal shifts of gaze with activation of ipsilateral posterior cervical muscles. Previous authors have postulated that descending eye muscle proprioceptive information may modulate second order vestibular neurons; resulting in direct or indirect influence on the medial vestibulospinal and reticulospinal tracts.[12,13] In animal studies, the trigeminal ganglion and the spinal trigeminal nucleus have been demonstrated through anatomical tracing studies to receive projections from eye muscle proprioceptors. The existence of similar pathways in humans is yet to be confirmed.[11] The work of Shaikh and colleagues supports the existence of a shared neural integrator responsible for coordinated control of head and eye movements. Dysfunction of the proposed neural integrator (including circuits of the interstitial nucleus of Cajal and the nucleus of Darkschewisch) may contribute to cervical dystonia.[14]

The consideration that visual fixation was necessary for our patient to perform his geste was refuted by the ability to maintain the effect during eye closure. This finding would support implication of eye muscle proprioceptors. Our patient displayed a reduction in posterior cervical muscle tonic activity ipsilateral to eye deviation, suggesting that this is not an amplification of previously demonstrated reflexive eye–head coupling.[12] There are two possible mechanisms of action for the "ocular geste antagoniste". First, there may be influences of eye proprioceptive input on higher cortical sensorimotor activity, similar to those demonstrated with functional imaging during traditional sensory tricks.[15,16] Second, there may be less well understood effects of eye movement or oculomotor proprioception on local brainstem circuits and neural integrators. Amplification or aberrations of normal reflexive responses may occur due to compensatory mechanisms in dystonia.

The authors would like to propose that ocular proprioceptive input may provide another potential mechanism for modification of motor output to the cervical musculature in primary cervical dystonia. Owing to the difficult to elicit nature, the ocular geste may be common but unrecognized in cervical dystonia. Identification of prevalence and investigation may facilitate further understanding of the pathophysiology of cervical dystonia.

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