Movement Disorders
CLINICAL PRACTICE
CASE SERIES

Characteristic "Forcible" Geste Antagoniste in Oromandibular Dystonia Resulting From Pantothenate Kinase-Associated Neurodegeneration

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Abstract: Geste antagonistes are usually considered typical of primary dystonia, although rarely they have been described in secondary/heredodegenerative dystonias. We have recently come across a particular geste antagoniste in 5 of 10 patients with pantothenate kinase-associated neurodegeneration (PKAN) who had prominent oromandibular involvement with severe jaw-opening dystonia. It consists of touching the chin with both hands characteristically clenched into a fist with flexion at the elbows. Because of the resemblance of this geste antagoniste with the praying-like posture of *Mantis religiosa*, we coined the term "mantis sign." Reviewing videos of PKAN cases in literature, including what is considered the first cinematic depiction of a case of this disorder, 3 additional cases with akin maneuvers were identified. In contrast, examining 205 videos of non-PKAN dystonic patients from our database for the presence of a similar maneuver was unrevealing. Thus, we consider the mantis sign to be quite typical of PKAN and propose it to be added as a clinical hint toward diagnosis.

The movement disorder of dystonia is characterized by sustained or intermittent muscle contractions that lead to abnormal and often repetitive movements and/or postures.[1] In some cases, the use of simple maneuvers, usually related to ephaptic contact with the dystonic body region, lead to an impressive temporary improvement of symptoms. These maneuvers are called gestes antagonistes (GAs) or sensory tricks.[2,3] GAs are usually considered in the context of primary dystonias.[1,3] However, their presence has also been described in secondary[4] as well as heredodegenerative conditions to include pantothenate kinase-associated neurodegeneration (PKAN).[5] In addition to typical GAs, atypical tricks have also been described in the literature (for review, see a previous work[6]). Forcible tricks (GA with additional use of force) are considered atypical sensory tricks and have been reported to be more prevalent in severe forms of dystonia than classic sensory tricks.[7]

We have recently observed, in 5 of 10 genetically proven PKAN patients (4 of them previously published elsewhere),[8] a characteristic GA maneuver associated with severe jaw-closing or -opening dystonia and retrocollis. It consisted of repetitive chin touching with the dorsum of one or both hands, which were clenched in dystonic flexion (see Video, Segment 1; Fig. 1). In addition to light touch in all cases, force was applied during the maneuver onto the chin to correct the abnormal jaw position (atypical GA). Reviewing available videos of PKAN cases in the literature,[9,10] 3 additional cases with akin maneuvers were identified, including what is considered the first cinematic depiction of a case of this disorder published by Vincent and van Bogaert[10] (see Video, Segments 2 and 3). The phenomenology of these "forcible tricks" resembled the praying-like posture of the praying mantis insect. Therefore, we propose naming this specific GA described here the "mantis sign."

The classic phenotype of PKAN resulting from mutations in the *PANK2* gene is characterized by early-onset generalized dystonia with prominent oromandibular involvement leading to dystonic jaw opening or jaw closing, anarthria, and severe dysphagia.[11] Lingual dystonia with tongue protrusion and repetitive touching of the face have been suggested as helpful clues

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Keywords: PKAN, dystonia, geste antagoniste.
Relevant disclosures and conflicts of interest are listed at the end of this article.
Received 18 January 2014; revised 11 April 2014; accepted 12 April 2014.
Published online 16 May 2014 in Wiley InterScience (www.interscience.wiley.com). DOI:10.1002/mdc3.12035

doi:10.1002/mdc3.12035

Figure 1 Praying-like GA maneuver in PKAN patients.

for the diagnosis.[5,11] Dystonic opisthotonus was recently suggested to be a red flag for clinicians to suspect neurodegeneration with brain iron accumulation syndromes.[12] Although typical GAs have been previously demonstrated in PKAN patients,[5,9] and are also shown in one of our cases presented here (gentle forehead touch to overcome blepharospam: case 4; see Video), these are rare. It thus appears that the mantis sign proposed here could be an additional diagnostic clue for PKAN. Reviewing 205 videos of our patients with other forms of dystonias, including 10 with prominent oromandibular involvement and, particularly, jaw-opening/-closing dystonia, but also videos from the literature we were not able to identify a similar maneuver. Whether it has been underreported or is indeed specific for PKAN is still thus unclear. However, clinicians should be aware and recognize the mantis sign proposed here as another red flag facilitating diagnostic considerations and prompting further examinations to include neuroimaging for the search of additional characteristic signs.[5,11]

Author Roles

(1) Research Project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript: A. Writing of the First Draft, B. Review and Critique.

I.N.P.: 1A, 3A, 3B
N.K.: 1B, 1C, 3A, 3B
C.G.: 1A, 1C, 3A, 3B
M.S.: 1B, 3B
N.D.: 1B, 3B
K.P.B.: 3B
V.S.K.: 1A, 1B, 1C, 3B

Disclosures

Funding Sources and Conflicts of Interest: The research is funded by the Ministry of Science, Republic of Serbia (project no.: 175090). The authors report no conflicts of interest.

Financial Disclosures for previous 12 months: I.N.P. has received honoraria from Boehringer Ingelheim and GlaxoSmithKline (GSK) and been employed by Institute of Neurology, CCS, Belgrade. N.K. has been employed by Institute of Neurology, CCS, Belgrade. C.G. has received grants for travel from Actelion, Ipsen, Pharm Allergan and Merz Pharmaceuticals and been employed by Institute of Neurology, University College London (UCL), London, UK. M.S. has received honoraria from Boehringer Ingelheim and GSK and has been employed by Institute of Neurology, CCS, Belgrade. K.P.B. has received personal compensation for serving on the scientific advisory boards of GSK and Boehringer Ingelheim; funding for travel from GSK, Orion Corporation, Ipsen, and Merz Pharmaceuticals, LLC; speaker honoraria from GSK, Ipsen, Merz Pharmaceuticals, LLC, and Sun Pharmaceutical Industries Ltd.; grant support through Dystonia Society UK, the Wellcome Trust MRC strategic neurodegenerative disease initiative award (reference no.: WT089698), a grant from the Dystonia Coalition, and a grant from Parkinson's UK (reference no.: G-1009); received research support from Ipsen and from the Halley Stewart Trust; has been employed by Institute of Neurology, UCL, London, UK; receives royalties from the Oxford University Press; and serves on the editorial boards of *Movement* Disorders and *Therapeutic Advances in Neurological Disorders*. V.S.K. has served on the regional advisory board of Boehringer Ingelheim and the local advisory board of Pfizer; has received payment for lectures from Novartis, Boehringer, Abott, GSK, Pfizer, Hemofarm-Stada, and Akaloid; has received grant sup-

doi:10.1002/mdc3.12035

port from the MNTR Serbia project (grant no.: 175090); and has been employed by Institute of Neurology, CCS, Belgrade. N.D. has been employed by Institute of Neurology, CCS, Belgrade.

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Supporting Information

A video accompanying this article is available in the supporting information here.

Video. Segment 1: Four patients with generalized dystonia and prominent oromandibular involvement with characteristic "forcible" GA consisting of repetitive chin touching. Self mutilation is present in cases 2 and 3. Segment 2: Siblings with PKAN with a similar maneuver previously published by Adamoviceva et al.[9] Segment 3: Akin maneuver in the case reported by van Bogaert and Vincent, considered to be the first video-recorded case of PKAN.[10]

doi:10.1002/mdc3.12035