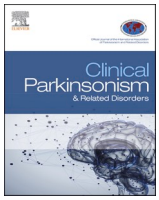
PRDOA 10 (2024) 100238

Contents lists available at ScienceDirect

Clinical Parkinsonism & Related Disorders

journal homepage: www.sciencedirect.com/journal/clinical-parkinsonism-and-related-disorders



Case Reports

The selfie sign in the diagnosis of functional tremor

Ekhlas Assaedi *, Xin Xin Yu, Junaid Siddiqui, Umar A Shuaib

Neurological Institute, Cleveland Clinic, Cleveland, OH, USA

ARTICLE INFO

Keywords:
Tremor
Functional movement disorder
Parkinson
Distractibility
Sign

ABSTRACT

Functional tremor (FT) is the most common phenotype of functional movement disorders (FMD). Its diagnosis can often be challenging. While positive signs such as tremor variability, distractibility, and entrainment support a diagnosis of FT, these diagnostic clues may not always be present and can be challenging to assess. In this case series, we identify another examination technique which could be of value when assessing FT. In our Movement Disorders clinic, charts were retrospectively reviewed for relevant clinical information. Video examinations were conducted. Obtained videos were either synchronous, via the use of screen recording software during telehealth visits or asynchronous, from self-recorded home videos. In both settings, patients were instructed to self-record their tremor using their phone cameras. Three patients with FT or comorbid FT were identified as demonstrating a unique examination sign. Videos showed an improvement or suppression of the tremor when the phone was held by the affected hand. When compared to a patient with tremor-dominant Parkinson's disease serving as a control, this "selfie sign" was not observed. These observations are preliminary and larger studies are needed to confirm the usefulness of the selfie sign in diagnosing FT. Patient-recorded videos of their tremor can be a convenient and practical way of evaluating suspected FT, especially when paroxysmal or variable symptoms limit the usefulness of classic signs often assessed in the clinic.

1. Introduction

Tremor is the most common movement disorder [1]. Among functional movement disorders (FMD), functional tremor (FT) is the most prevalent form [2]. However, its diagnosis can often be challenging [3,4]. Current concepts of FMD encourage a diagnosis by inclusion [5]. Identifying positive signs such as tremor variability, distractibility, entrainment, and incongruence supports FT diagnosis.

A growing body of literature highlights the role of telemedicine in the care of patients with Parkinson's disease and hyperkinetic disorders [6]. Synchronous videoconferencing, conducted over various platforms such as Zoom or Microsoft Teams, can be utilized for clinical follow up, to administer an abbreviated neurological examination or movement disorders scales, or to counsel [7–10]. Examination can also be supplemented by asynchronous videos recorded by a personal recording device. These can be particularly helpful when dealing with a paroxysmal movement disorder or a hyperkinetic disorder with variability and fluctuations [6]. Here, we identify an examination technique which can be of value when assessing patients with FT and propose that it be referred to as 'the selfie sign'. To the best of our knowledge, this sign has not been reported previously.

1.1. Case 1

A 54-year-old, right-handed gentleman presented with a three-year history of intermittent, bilateral involuntary movements of the hands and occasionally the legs. This occurred in the context of a history of hypertension, chronic inactive hepatitis B virus infection, diabetes, depression, chronic abdominal pain, right C3-C4 radiculopathy, and pancreatic exocrine insufficiency secondary to chronic pancreatitis treated with pancreatectomy. His medications included: amlodipine, amitriptyline, fluoxetine, zolpidem, gabapentin (total of 1800 mg/d), glucagon nasal spray, insulin, and lipase-protease-amylase. His mother had a history of tremor.

Provided history could reflect intermittent tremor or myoclonus. However, the movements in question were not evident when examined in the office, but mild asterixis was present. His neurological examination, including Archimedes spiral drawing, was otherwise normal. He was weaned off gabapentin due to suspicion for medication-induced myoclonus, but his symptoms did not improve. Given his medical history, he was screened for secondary causes of myoclonus. His laboratory investigations were normal except for a mildly elevated hemoglobin A1C, gamma-glutamyl transferase, and alkaline phosphatase. Copper

* Corresponding author at: Center for Neurological Restoration, 9500 Euclid Ave, Cleveland, OH 44195, USA.
E-mail address: assaede@ccf.org (E. Assaedi).

https://doi.org/10.1016/j.prdoa.2024.100238
Received 20 September 2023; Received in revised form 3 December 2023; Accepted 23 January 2024
Available online 26 January 2024
2590-1125/© 2024 Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

and ceruloplasmin were normal. Before expanding workup or initiating symptomatic treatment, the patient was asked to videotape his abnormal movement at home. His self-recording showed (Video 1, Segment 1) an asymmetric action tremor which affected the right arm more severely. It had variable frequency and moderate amplitude. When he alternated using his hands to document the tremor of the contralateral hand, the left hand tremor attenuated and the right hand tremor improved, as inferred from the quality of the video. The diagnosis of FT was made based on the inconsistent clinical features and the lack of metabolic abnormalities relevant to his phenomenology.

1.2. Case 2

A 54-year-old, right-handed lady was referred for the evaluation of a two-year history of gait impairment and weakness, numbness, and pain affecting the right leg. Her initial neurological examination showed severe weakness (Medical Research Council score 0 out of 5) of the right lower extremity (LE) with a positive Hoover's sign and patchy non-dermatomal pinprick sensory loss. She was able to ambulate independently with an antalgic gait. Her symptoms were previously investigated with nerve conduction studies and electromyography of the right LE and an MRI of the brain and the spine. Tests were non-revealing. Her past medical history included hypertension and she was treated with chlorthalidone. She was diagnosed with functional neurologic disease and was referred to a multidisciplinary team which included pain management, physical therapy, and occupational therapy.

She was followed longitudinally and was noted, a year later, to develop new onset tremor and slowing of movements. Examination was conducted via a telemedicine platform, and the patient used her mobile phone to attend. Exam (not shown) demonstrated right, non-decremental slowing of the repetitive movements and a low-frequency tremor. Right-hand tremor was less prominent when it was the recording hand, and it showed distractibility when carrying out motor tasks with the left hand.

1.3. Case 3

A 59-year-old, left-handed female was seen for a second opinion on her tremor. She was interested in assessing candidacy for deep brain stimulation (DBS). Tremor involved her head, voice, and hands. Tremor onset was at the age 35, starting with the hands. It progressed gradually. She had a family history of tremor. Alcohol may have helped her tremor to some extent. She was previously treated with propranolol with mild improvement. Topiramate was not well tolerated. Her initial examination showed a moderate amplitude action tremor which affected the left arm more severely. Her tremor disappeared when either hand was engaged in Luria's sequence. She had a head tremor without dystonic posturing. She was referred for videolaryngoscopy with stroboscopy to evaluate her voice tremor further. It showed a mild tremor of the vocal cords during phonation with a normal and complete pattern of glottic closure. Given reports of fluctuating tremor severity, she was asked to record her more severe tremor at home. Self-recorded video (not shown) revealed a jerky left-hand tremor which intermittently attenuated when it was recording or when the right hand was engaged in manual tasks. Given the variability and distractibility of her jerky tremor, her condition was felt to be most in keeping with essential tremor with comorbid FT. Decision was made against proceeding with DBS surgery. Propranolol dose was titrated further and the patient was followed in the FMD multidisciplinary clinic.

1.4. Case 4

A 68-year-old, right-handed gentleman with Parkinson's disease was evaluated as a control subject. He had a one-year history of right-hand tremor. Examination showed mild rest and postural tremor. It was accompanied by mild bradykinesia. When examined utilizing a telehealth platform on the patient's phone (Video 1, Segment 2), the recording right hand showed a persistent, mild, slow, postural tremor.

2. Discussion

In an international survey exploring the practices of movement disorders specialists, the presence of positive signs was considered to be necessary to make a diagnosis of definite FMD [11]. Beyond the classic features of functional neurologic disorder, many other examination signs have been previously described, which aid in diagnosing the condition and conveying the diagnosis. When evaluating functional weakness, Hoover's, reverse Hoover's, abductor sign, barre sign, or positive "make a fist" test can be seen [12–15]. In cases of hyperkinetic disorders, "stretched slinky", "whack-a-mole," and coactivation sign can be observed [4,16,17].

We observe an interesting phenomenon, which we call the selfie sign, when examining patients with FT. It is characterized by the improvement or suppression of the tremor, as assessed by the severity of the image shaking when the affected hand holds the camera for self-recoding of the movement. The proposed sign and many of the aforementioned signs share a common theme: transient distraction reveals an underlying normal function. Indeed, abnormalities of attention are thought to play a central role in the mechanism of FMD [18]. Several hypotheses and observations have been raised, including altered attentional focus and heightened self-monitoring during movement [19–21].

In conclusion, patient-recorded videos of their tremor can be a convenient and practical way of evaluating suspected FT, especially when paroxysmal or variable symptoms limit the usefulness of classic signs often assessed in the clinic. These observations are preliminary. Future studies will be needed to assess the reproducibility of the selfie sign and its usefulness as a diagnostic clue to FT and to evaluate its accuracy and specificity.

Ethical Compliance Statement

The authors confirm that the approval of an institutional review board was not required for this work. All patient data has been anonymized. Patients, appearing in the video, provided their written consent to be videotaped and to disclose information in scientific publications. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

Study funding

The authors report no targeted funding.

Financial Disclosures

The authors have no conflict of interest to disclose.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Acknowledgements

We thank our patients for facilitating this work.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.prdoa.2024.100238

References

[1] W.J. Elias, B.B. Shah, Tremor, JAMA. 311 (2014) 948–954, https://doi.org/10.1001/JAMA.2014.1397.

[2] M. Hallett, Functional (Psychogenic) Movement Disorders – Clinical Presentations, Parkinsonism Relat. Disord. 22 (2016) S149, https://doi.org/10.1016/J.PARKRELDIS.2015.08.036.

[3] P. Schwingenschuh, T.A. Saifee, P. Katschnig-Winter, A. Macerollo, M. Koegl-Wallner, V. Culea, C. Ghadery, E. Hofer, T. Pendl, S. Seiler, U. Werner, S. Franthal, N.M. Maurits, M.A. Tijssen, R. Schmidt, J.C. Rothwell, K.P. Bhatia, M.J. Edwards, Validation of "laboratory-supported" criteria for functional (psychogenic) tremor, Mov. Disord. 31 (2016) 555–562, https://doi.org/10.1002/MDS.26525.

[4] W.K. Fung, A. Fasano, E.J. Safra Program in Parkinson, G. Shulman, "Stretched Slinky" Sign Another Clue to Functional Tremor, Neurology. 12 (2022) 121–123, https://doi.org/10.1212/CPJ.0000000000200075.

[5] A.J. Espay, S. Aybek, A. Carson, M.J. Edwards, L.H. Goldstein, M. Hallett, K. LaFaver, W.C. LaFrance, A.E. Lang, T. Nicholson, G. Nielsen, M. Reuber, V. Voon, J. Stone, F. Morgante, Current concepts in diagnosis and treatment of functional neurological disorders, JAMA Neurol. 75 (2018) 1132, https://doi.org/10.1001/jamaneurol.2018.1264.

[6] R. Srinivasan, H. Ben-Pazi, M. Dekker, E. Cubo, B. Bloem, E. Moukheiber, J. Gonzalez-Santos, M. Guttman, Telemedicine for Hyperkinetic Movement Disorders, Tremor and Other Hyperkinetic Movements. 10 (2020), https://doi.org/10.5334/TOHM.534.

[7] S. Mishra, N. Dhuna, N. Lancki, C. Yeh, D.N. Larson, Telehealth utilization and patient satisfaction in an ambulatory movement disorders center during the COVID-19 pandemic, J. Telemed. Telecare. (2022), https://doi.org/10.1177/1357633X221146819/ASSET/IMAGES/LARGE/10.1177_1357633X221146819-FIG2.JPEG.

[8] A. Abdolahi, N. Scoglio, A. Killoran, E.R. Dorsey, K.M. Biglan, Potential reliability and validity of a modified version of the Unified Parkinson's Disease Rating Scale that could be administered remotely, Parkinsonism Relat. Disord. 19 (2013) 218–221, https://doi.org/10.1016/J.PARKRELDIS.2012.10.008.

[9] M. Al Hussona, M. Maher, D. Chan, J.A. Micieli, J.D. Jain, H. Khosravani, A. Izenberg, C.D. Kassardjian, S.B. Mitchell, The Virtual Neurologic Exam: Instructional Videos and Guidance for the COVID-19 Era, Can. J. Neurol. Sci. 47 (2020) 1. https://doi.org/10.1017/CJN.2020.96.

[10] A. Hawkins, S. Creighton, A. Ho, B. Mcmanus, M. Hayden, Providing predictive testing for Huntington disease via telehealth: results of a pilot study in British Columbia, Canada, Clin. Genet. 84 (2013) 60–64, https://doi.org/10.1111/CGE.12033.

[11] A.J. Espay, L.M. Goldenhar, V. Voon, A. Schrag, N. Burton, A.E. Lang, Opinions and Clinical Practices Related to Diagnosing and Managing Patients with Psychogenic Movement Disorders: An International Survey of Movement Disorder Society Members, Mov. Disord. Vol. 24, No. 9, 2009, Pp. 1366–137. 24 (2009). https://doi.org/10.1002/mds.22618.

[12] O. Aboud, A. Al-Salaimeh, S. Kumar Raina, K. Sahaya, A. Hinduja, Positive clinical signs in neurological diseases – An observational study, J. Clin. Neurosci. 59 (2019) 141–145, https://doi.org/10.1016/J.JOCN.2018.10.113.

[13] C. Kobylecki, M.W. Kellett, J.P. Dick, A.G. Marshall, "Reverse Hoover's sign" demonstrated by neurophysiology in a patient with functional dystonia, Park. Relat. Disord. 29 (2016) 125–126, https://doi.org/10.1016/j.parkreldis.2016.05.005.

[14] M. Sonoo, Abductor sign: a reliable new sign to detect unilateral non-organic paresis of the lower limb, J Neurol Neurosurg Psychiatry. 75 (2004) 121–125.

[15] J. Stone, S. Aybek, Functional limb weakness and paralysis, Handb. Clin. Neurol. 139 (2016) 213–228, https://doi.org/10.1016/B978-0-12-801772-2.00018-7.

[16] J.E. Park, C.W. Maurer, M. Hallett, The "Whack-a-Mole" Sign in Functional Movement Disorders, Mov. Disord. Clin. Pract. 2 (2015) 286–288, https://doi.org/10.1002/MDC3.12177.

[17] P. Schwingenschuh, G. Deuschl, Functional tremor, Handb. Clin. Neurol. 139 (2016) 229–233, https://doi.org/10.1016/B978-0-12-801772-2.00019-9.

[18] R. McIntosh, L. McWhirter, L. Ludwig, A. Carson, J. Stone, Attention and sensation in functional motor disorder, 106 (2017) 207–215. https://doi.org/10.1016/j.neuropsychologia.2017.09.031.

[19] A.C.M.L. Huys, P. Haggard, K.P. Bhatia, M.J. Edwards, Misdirected attentional focus in functional tremor, Brain. 144 (2021) 3436–3450, https://doi.org/10.1093/BRAIN/AWAB230.

[20] F.P. de Lange, K. Roelofs, I. Toni, Increased self-monitoring during imagined movements in conversion paralysis, Neuropsychologia. 45 (2007) 2051–2058, https://doi.org/10.1016/J.NEUROPSYCHOLOGIA.2007.02.002.

[21] M.J. Edwards, J.C. Rothwell, Losing focus: How paying attention can be bad for movement, Mov. Disord. 26 (2011) 1969–1970, https://doi.org/10.1002/MDS.23920.