Parkinsonism and Related Disorders 78 (2020) 200–203

Contents lists available at ScienceDirect

Parkinsonism and Related Disorders

journal homepage: www.elsevier.com/locate/parkreldis

Video-tutorial for the Movement Disorder Society criteria for progressive supranuclear palsy

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ARTICLE INFO

Keywords:
Progressive supranuclear palsy
Clinical diagnostic criteria
Phenotype
Video tutorial

ABSTRACT

Background: The International Parkinson and Movement Disorder Society-endorsed Progressive Supranuclear Palsy Study Group published clinical diagnostic criteria for progressive supranuclear palsy in 2017, aiming to optimize early, sensitive and specific diagnosis.
Objective: To assist physicians in the application of these criteria, we developed a video-based tutorial in which all core clinical features and clinical clues are depicted and explained.
Methods: Patients provided written informed consent to the publication of their videos. High-quality videos along with essential descriptions were collected by the study group members. Most educational videos were selected in a structured consensus process.
Results: We provide 68 videos of all core clinical features and clinical clues defined by the diagnostic criteria, along with instructive descriptions of the depicted patients, examination techniques and clinical findings.
Conclusions: This comprehensive video-based tutorial will support physicians in the application of the diagnostic criteria of progressive supranuclear palsy.

1. Introduction

Progressive supranuclear palsy (PSP) is a neuropathologically defined disease entity [1]. The pathological hallmarks of PSP are intracellular aggregations of hyperphosphorylated four-repeat tau protein in the form of neurofibrillary tangles, tau deposits in astrocytes ("astrocytic tufts") and oligodendroglia ("coiled bodies") [2].

The Movement Disorder Society (MDS) criteria for the clinical diagnosis of PSP (MDS-PSP criteria) were published in 2017 [1]. Diagnostic challenges addressed in these criteria include the heterogeneity of clinical features associated with PSP, clinical overlaps with and distinctions from other diseases, and the limited additional value of diagnostic biomarkers [1].

The previously used Neurological Disorders and Stroke and Society

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https://doi.org/10.1016/j.parkreldis.2020.06.030
Received 21 November 2019; Received in revised form 19 June 2020; Accepted 27 June 2020
Available online 22 September 2020

Table 1
MDS-PSP-criteria – 'core clinical features' and supportive 'clinical clues'.

Core clinical features	
Domain: Ocular motor dysfunction	
O1	Vertical supranuclear gaze palsy
O2	Slow velocity of vertical saccades
O3	Subtle ocular motor dysfunction, *i.e.*
O3-1	frequent macro square wave jerks, *or*
O3-2	"eyelid opening apraxia"
Domain: Postural instability	
P1	Repeated unprovoked falls within 3 years
P2	Tendency to fall on the pull-test within 3 years
P3	More than 2 steps backward on the pull-test within 3 year
Domain: Akinesia	
A1	Progressive gait freezing within 3 years
A2	Parkinsonism, akinetic-rigid, predominantly axial and levodopa-resistant
A3	Parkinsonism, with tremor and/or asymmetric and/or levodopa-responsive
Domain: Cognitive dysfunction	
C1	Speech/language disorder, *i.e.* at least 1 of the following features:
C1-1	Nonfluent agrammatic variant of primary progressive aphasia (nfaPPA), *or*
C1-2	progressive apraxia of speech (AOS)
C2	Frontal cognitive/behavioral presentation, *i.e.* at least 3 of the following features:
C2-1	Apathy
C2-2	Bradyphrenia
C2-3	Dysexecutive syndrome (e.g., pathological Luria sequence)
C2-4	Reduced phonemic verbal fluency (e.g., reduced "D, F, A or S" words per minute)
C2-5	Impulsivity, disinhibition, or perseveration (e.g., socially inappropriate behaviors, overstuffing the mouth when eating, motor recklessness, applause sign, palilalia, echolalia)
C3	Corticobasal syndrome, i.e. at least 1 sign each from the following 2 groups
C3-1	Cortical signs
C3-1a	Orobuccal or limb apraxia
C3-1b	Cortical sensory deficit
C3-1c	Alien limb phenomena (more than simple levitation)
C2-2	Movement disorders signs
C3-2a	Limb rigidity
C3-2b	Limb akinesia
C3-2c	Limb myoclonus
C3-2d	Limb dystonia
2. Clinical clues	
CC1	Levodopa-resistance
CC2	Hypokinetic, spastic dysarthria
CC3	Dysphagia
CC4	Photophobia

Legend: For detailed operationalized definitions of the 'core clinical features' and supportive 'clinical clues' see the original MDS-PSP criteria publication [1]. In the definition of C3, "limb dystonia" was unintentionally omitted in the original publication [1] and shall be considered as 4th qualifying movement disorders sign for corticobasal syndrome.

for Progressive Supranuclear Palsy diagnostic criteria [3] had lower sensitivity, because they relied exclusively on the identification of patients with Richardson's syndrome [4,5]. The MDS-PSP criteria have improved sensitivity by taking into account the broader spectrum of clinical presentations of PSP [6,7]. Consequently, their application is more complex compared to prior criteria.

The MDS-PSP criteria are based on 'mandatory inclusion criteria', 'mandatory exclusion criteria', 'context-dependent exclusion criteria', 'core clinical features', and 'supportive features' (including 'clinical clues' and 'imaging findings') [1]. Core clinical features (Table 1) are defined in four functional domains, i.e. ocular motor dysfunction, postural instability, akinesia, and cognitive dysfunction, labelled O, P, A, or C, respectively. In each domain, three levels of core clinical features are defined, labelled 1, 2 or 3, respectively, with lower numbers indicating a higher diagnostic certainty to a diagnosis of PSP [1]. Operationalized definitions for the core clinical features and clinical clues were developed based on a systematic analysis of the literature, analysis of a large clinico-pathological cohort and expert consensus [1]. Specific combinations of core clinical features and clinical clues indicate the degree of diagnostic certainty and the predominance types of PSP patient [1].

To facilitate the application of the MDS-PSP criteria and to promote best clinical practice in PSP diagnosis, the MDS-endorsed PSP Study Group developed this video-based tutorial, in which the examination techniques and characteristic pathological signs for the diagnostic features are illustrated.

2. Methods

The MDS-endorsed PSP Study Group formed a steering committee (G.U·H., G.R. and V·I.) to coordinate the development of the video tutorial. The steering committee established guidelines for videotaping of the core clinical features and clinical clues, as defined in the MDS-PSP criteria. All videos had to be taken in high-definition; the area of interest had to be bright with minimal shadows; the background had to be clean and non-distracting. Existing videos were identified from the study group members' archives, and new videos were recorded. All patients provided written informed consent for publication of their videos. Videos meeting these guidelines were uploaded to a secure, password-protected online platform. Information about age, clinical phenotype and diagnostic certainty of the patients were assembled. In a first review, the steering committee evaluated quality and content of all uploaded videos. Underrepresented clinical features were identified and the study group members were encouraged to provide additional videos of these specific features. In a second review, the steering committee proposed an initial selection for the tutorial, based on technical and educational quality, eliminating videos not meeting the pre-specified quality criteria. All study group members were then asked to provide written feedback on this initial selection. All comments were taken into consideration and the final selection of videos was made through Delphi consensus. Videos were cut to present the most instructive content in a compact time frame using *OpenShot* Video Editor. Title captions at the beginning of each video were introduced using *MLTmelt* and *ImageMagic*.

3. Results

From over 100 submitted videos, 68 representative videos were selected to depict the core clinical features' and supportive 'clinical clues' (Table 1), as defined and operationalized in the MDS-PSP criteria [1].

These videos and the corresponding legends are found in the online supporting material.

We did not provide videos for the features apathy (part of the frontal cognitive/behavioral presentation C2) and limb rigidity (part of corticobasal syndrome C3), because these features turned out to be difficult to visualize in a video.

For certain clinical features more than one representative video was selected to show their variable quality and severity.

4. Discussion

An early and accurate clinical diagnosis of PSP is increasingly important to provide patients with reliable information about prognosis, treatment options, and access to clinical trials [2]. The MDS-PSP criteria take into account early clinical features as well as the broad clinical spectrum of PSP, creating the basis for an earlier and more sensitive, but still specific diagnosis of PSP [6,7]. Therefore, it is now most important to increase awareness for the MDS-PSP criteria and the clinical spectrum of PSP among physicians.

For this purpose, we established this comprehensive video-based

tutorial for the correct application of the MDS-PSP criteria. This tutorial aims to reach physicians and clinician scientists from all levels of experience, to assist in the standardized application of the MDS-PSP criteria by displaying the clinical features associated with PSP pathology.

This video tutorial does not display 'mandatory inclusion criteria', 'mandatory exclusion criteria', 'context-dependent exclusion criteria', and supportive 'imaging findings', which are also essential components of the MDS-PSP criteria. Therefore, we wish to emphasise the need to refer to the original publication of the MDS-PSP criteria [1] and to the more recently published guidelines for a correct application of the diagnostic criteria [8].

We expect this video-based tutorial not only to facilitate the application of the MDS-PSP criteria, but moreover to increase awareness among clinicians of the broad spectrum of clinical presentations of PSP, fostering an early, sensitive and still specific clinical diagnosis.

Author roles

1. Project: A. Conception, B. Organization, C. Execution;
2. Manuscript Preparation: A. Writing the First Draft, B. Review and Critique.

Vassilena Iankova: 1B, 1C, 2A
Gesine Respondek: 1A, 1B, 1C, 2B;
Gerard Saranza: 1C, 2B;
Cèlia Painous: 1C, 2B;
Ana Cámara: 1C, 2B;
Yaroslau Compta: 1C, 2B;
Ikuko Aiba: 1C, 2B;
Bettina Balint: 1C, 2B;
Nikolaos Giagkou: 1C, 2B;
Keith A. Josephs: 1C, 2B;
Mika Otsuki: 1C;
Lawrence I. Golbe: 1C, 2B;
Kailash P Bhatia: 1C, 2B;
Maria Stamelou: 1C, 2B;
Anthony E Lang: 1C, 2B;
Günter U. Höglinger: 1A, 1B, 1C, 2B;

Financial disclosures of all authors for the preceding 12 months

Vassilena Iankova has nothing to disclose. Gesine Respondek has nothing to disclose. Gerard Saranza has nothing to disclose. Cèlia Painous has received support from Boston and has a competitive Río Hortega predoctoral grant from the Spanish Ministry of Health.

Ana Cámara has received funding or support from Abbvie and Medtronic as well as from the H2020 lPI043760 grant. She has a competitive PERIS grant from the Department of Health of the Generalitat de Catalunya (SLT008/18/00191).

Yaroslau Compta has served as a consultant for Abbvie and Zambon, and has received honoraria for scientific presentations from Abbvie, Alter, Bial, Medtronic, Merz, Teva, UCB, and Zambon. He is currently an associate editor of Parkinsonism & Related Disorders (Elsevier). He has competitive grants from Instituto de Salud Carlos III (ISCIII) / Spanish Ministry of Health (PI17/00096) and the European Commission H2020 program (lPI043760). YC's center receives support from the CERCA program of Generalitat de Catalunya and the Maria de Maeztu program of the Spanish Ministry of Education, and is part of the European Reference Network for Rare Neurological Diseases (ERN-RND).

Ikuko Aiba serves as a consultant for Abbvie and Biogen.Bettina Balint is supported by the Robert Bosch Foundation.Nikolaos Giagkou has nothing to disclose.

Keith A. Josephs is funded by the National Institute of Health.Mika Otsuki has nothing to disclose.

Lawrence I. Golbe receives research support from Biogen and AbbVie and serves as a consultant for CurePSP, Biogen, AbbVie, UCB, Asceneuron, GLG, Piramal, Pinteon, NeuroTrax and Novartis.

Kailash P Bhatia holds research grants from NIHR RfPB, MRC Wellcome Strategic grant (Ref. no.: WT089698) and PD UK (Ref. no.: G-1009), and has received honoraria/financial support to speak/attend meetings from GSK, Boehringer-Ingelheim, Ipsen, Merz, Sun Pharma, Allergan, Teva Lundbeck and Orion pharmaceutical companies. KB receives royalties from Oxford University press and a stipend for MDCP editorship.

Maria Stamelou serves as assistant editor in Movement Disorders Journal, has received travel and speaker honoraria from Biogen, UCB, Abbvie and Specifar, MDS and EAN, royalties from Oxford and Cambridge University Press, has served in as advisory board to Biogen and Specifar and receives research support from PPMI.

Anthony E Lang: Consultancies: Abbvie, Acorda, AFFiRis, Biogen, Bristol Myers Squibb, Intracellular, Janssen, Jazz, Lilly, Lundbeck, Merck, Ono, Paladin, Roche, Seelos, Syneos, Sun Pharma, Theravance, and Corticobasal Degeneration Solutions; Advisory Boards: Jazz Pharma, PhotoPharmics, Sunovion; Honoraria:Sun Pharma, AbbVie and Sunovion: Grants: Brain Canada, Canadian Institutes of Health Research, Corticobasal Degeneration Solutions, Edmond J Safra Philanthropic Foundation, Michael J. Fox Foundation, the Ontario Brain Institute, Parkinson Foundation, Parkinson Canada, and W. Garfield Weston Foundation; Royalties: Elsevier, Saunders, Wiley-Blackwell, Johns Hopkins Press, and Cambridge University Press.

Günter U. Höglinger has ongoing research collaborations with Orion, Prothena; serves as a consultant for Abbvie, Alzprotect, Asceneuron, Biogen, Biohaven, Novartis, Sanofi, UCB; received honoraria for scientific presentations from Abbvie, Biogen, Roche, Teva, UCB, Zambon, has received research support from CurePSP, the German Academic Exchange Service (DAAD), German Parkinson's Disease Foundation (DPG), German PSP Association (PSP Gesellschaft), German Research Foundation (DFG) and the German Ministry of Education and Research (BMBF), International Parkinson's Fonds (IPF); has received institutional support from the German Center for Neurodegenerative Diseases (DZNE).

Financial disclosure

None.

Funding source for study

The project was supported by the German Center for Neurodegenerative Diseases e.V. (DZNE), German PSP Gesellschaft, and the International Parkinson & Movement Disorder Society.

Declaration of competing interest

None.

Acknowledgements

The authors would like to thank Naomi Matsuda, PT, Yoriko Omote, PT, Hiroshi Nishimura, OT, Kousuke Kobayashi, ST, of the Department of Rehabilitation, National Hospital Organization Higashinagoya National Hospital, Nagoya, Japan and Rina Hashimoto, MD, PhD, Satoko Sakakibara, MD of the Department of Neurology, National Hospital Organization Higashinagoya National Hospital, Nagoya, Japan, for their support.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.parkreldis.2020.06.030.

Appendix

These MDS-endorsed PSP study group members contributed to this work: Anthony E. Lang, Bettina Balint, Gesine Respondek, Günter Höglinger, Ikuko Aiba, Kailash P Bhatia, Keith A Josephs, Lawrence I Golbe, Maria Stamelou, Vassilena Iankova, Yaroslau Compta.

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