Higher-level gait disorder as a presenting manifestation of progressive supranuclear palsy: a video case report

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Introduction. Frontal gait disorder/gait apraxia is a higher-order motor deficit with various causes, characterized by difficulties with gait initiation, such as freezing or ignition failure. We aimed to report a patient who presented with progressive higher-level gait disorder and fall episodes as the initial manifestations of progressive supranuclear palsy (PSP). Patient data were obtained from medical records from the Department of General Medicine, Burdwan Medical College & Hospital (Burdwan, West Bengal, India).

Case report. A 58-year-old previously healthy woman presented with a gait disorder and fall episodes. Detailed neurological examination highlighted characteristic facial appearance (wide-eyed staring, furrowing of the forehead with a frowning expression, and fixed expression of the lower face). She was hypokinetic-rigid with symmetrical signs and predominant axial rigidity with retrocolic trunk and neck posture. Gait examination revealed a higher-level gait pattern characterized by an exhibition of profound start hesitation requiring assistance from nearby objects/persons. Once walking was underway, steps became relatively better, but ineffective gait re-emerged when she attempted turning. She had short strides, freezing, broad stance base, disequilibrium, slow leg movement, shuffling, and loss of normal fluidity of trunk and limbs. Postural reflexes were impaired. Brain magnetic resonance imaging revealed atrophy of the midbrain, dilated aqueduct of Sylvius and third ventricle, atrophy of frontal lobes and typical hummingbird sign. Diagnosis of probable PSP was finally made.

Conclusions. Several etiologies, including PSP, should be considered in appropriate clinical contexts if gait examination demonstrates a higher-order gait disorder.

Key words. Gait. Higher-level gait disorder. Movement disorder. Presenting manifestation. Progressive supranuclear palsy. Video case report.

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Introduction

Progressive supranuclear palsy (PSP), a tauopathy subtype, is a rapidly progressive neurodegenerative disorder with akinetic-rigid parkinsonism characterized by insidious onset and predominantly symmetrical axial involvement. Backward unprovoked falls and gait difficulties are PSP's most common initial manifestation. Other standard features are generalized motor slowing, cognitive impairment, ocular motor disturbances, sleep disorders, and poor response to levodopa [1]. Although vertical supranuclear gaze palsy is a hallmark of PSP, it can appear several years into the disease delaying the diagnosis [1,2]; slowed vertical saccades may be the only eye movement manifestation early on [3].

Frontal gait disorder/gait apraxia is a higher-order motor deficit with various causes (Table I), characterized by difficulties with gait initiation, such as freezing or ignition failure [1,4].

We aimed to report a 58-year-old previously healthy woman who presented with progressive higher-level gait disorder and fall episodes as the presenting manifestation of PSP.

Patient data were obtained from medical records from the Department of General Medicine, Burdwan Medical College & Hospital (Burdwan, West Bengal, India).

Case report

Detailed neurological examination highlighted characteristic facial appearance: wide-eyed staring, furrowing of the forehead with a frowning expression (procerus sign), and fixed expression of the

Vídeo. Gait examination revealed a higher (complex, integrative) order gait pattern characterized by an exhibition of profound start hesitation (as if the feet were glued to the floor) requiring assistance and support from nearby objects/persons with reduced upper body movements. The patient has short strides, freezing, broad stance base, disequilibrium, slow leg movement, shuffling, and loss of normal fluidity of the trunk and limbs. There is also severe difficulty in sitting over a chair. Once walking is underway, steps become relatively better, but ineffective gait re-emerges when she attempts turning. *(Click on the image to play video).*

lower face. The patient was hypokinetic-rigid with symmetrical signs and predominant axial rigidity with retrocolic trunk and neck posture. Gait examination revealed a higher-level gait pattern characterized by an exhibition of profound start hesitation requiring assistance from nearby objects/persons. Once walking was underway, steps became relatively better, but ineffective gait re-emerged when she attempted turning. She had short strides, freezing, broad stance base, disequilibrium, slow leg movement, shuffling, and loss of normal fluidity of trunk and limbs. Postural reflexes were impaired. There was severe difficulty standing from sitting and difficulty turning over in bed (Video). However, frontal signs such as paratonic rigidity, grasp reflexes, urinary incontinence, and features of frontal cognitive impairments (e.g., dysexecutive and personality changes and impulsivity) were absent except for a progressive apathetic state. Vertical saccades were impaired, with relatively normal amplitudes and horizontal saccadic movements. Square wave jerks and vertical supranuclear gaze palsy were evident. Brain magnetic resonance imaging revealed atrophy of the midbrain, dilated aqueduct of Sylvius and third ventricle, and atrophy of frontal lobes, and typical hummingbird and morning glory signs (Figure). Diagnosis of probable PSP was finally made [2,5].

Discussion and conclusions

Gait should always be examined meticulously in every patient with a history of unprovoked falls. Clinical examination of gait patterns can be a reliable clinical aid in clinical and research settings and may help formulate different rehabilitation approaches for these patients. All the etiologies described in table I should be considered in appropriate clinical contexts if gait examination demonstrates a higher-order gait disorder. In our case, a comprehensive clinical examination, including the gait patterns and MRI findings, gave us essential clues to reach a diagnosis of PSP. Of note is that quantitative gait evaluation can distinguish PSP patients from Parkinson's disease patients even at the earliest stages of the disease (Table II) [6].

Regarding the course of the different PSP subtypes, natural history studies have mainly focused on the PSP-Richardson's syndrome subtype (vertical ocular motor dysfunction, early onset postural instability, and falls) [1]. In one series of 100 pathologically confirmed PSP cases that included patients with PSP-Richardson's syndrome, PSP-parkinsonism, PSP-postural instability, PSP-ocular motor, PSP-corticobasal syndrome, PSP-frontotemporal dementia, and unclassified phenotypes, the mean disease duration (± standard error of the mean) for all subtypes was 8.7 (±0.4) years, ranging from 2 to 28 years [7]. Individuals with the PSP-Richardson's syndrome phenotype had the shortest mean disease duration (7.3 ± 0.6; range 4 to 17 years). Meanwhile, individuals with the PSP-parkinsonism subtype (clinical phenotype resem-

Table I. Main differential diagnoses of higher-level gait disorders.

1. Normal-pressure hydrocephalus
2. Other subtypes of communicating hydrocephalus in adults
3. Vascular parkinsonism
4. Progressive supranuclear palsy
5. Corticobasal degeneration
6. Multiple system atrophy
7. Advanced Parkinson's disease
8. Frontal lobe tumours
9. Frontotemporal dementia
10. Alzheimer's disease
11. Creutzfeldt-Jakob disease
12. Juvenile Huntington's disease (Westphal variant)
13. Wilson disease
14. Diffuse axonal injury
15. Cerebral anoxia
16. Neurosyphilis

bling Parkinson's disease, later development of symptoms of PSP-Richardson's syndrome) had the longest disease duration (12.8 ± 1.5; range 4 to 28 years). It is likely that those with the PSP-progressive gait freezing subtype (presentation with an isolated gait disorder with start hesitation and progressive freezing of gait) also have a longer disease duration, according to data from a pathologic case series (mean disease duration of 13 years; range 5 to 21 years) [8], and from case reports revealing disease durations up to 15 years [9].

Predictors of shorter survival in PSP, derived from cohorts of individuals with pathologically proven PSP or in-life PSP-Richardson's syndrome diagnoses using prior PSP diagnostic criteria, include the PSP-Richardson's syndrome subtype (versus PSP-parkinsonism) and early dysphagia, cognitive symptoms, and falls [10].

Figure. Magnetic resonance imaging of the brain revealing hummingbird sign-on mid-sagittal T_1 weighted imaging (a) and T_2 weighted imaging (b) and morning glory sign on axial T_2 weighted imaging (c). The head position concerning the cervical spine gives us an idea that the patient has active retrocollis.

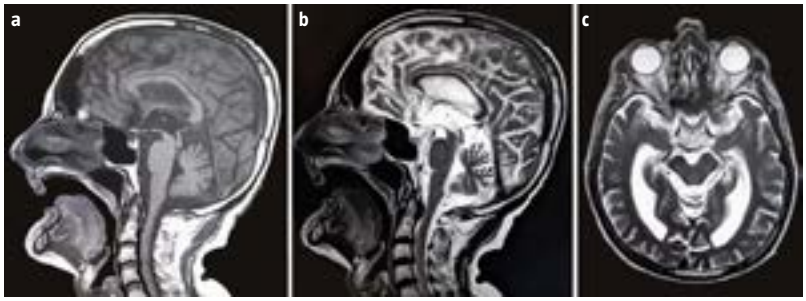

Table II. Differential diagnosis of parkinsonism and frontal gait disorder with a visual inspection of posture and gait.

	Parkinson's disease	Atypical parkinsonism	Frontal gait disorder
Stance and posture	Forward stoop, rest tremor	Extended neck and back Dystonic (axial) in some	Upper body spared. Wide based
Ignition failure	Late	Early	Very early
Arm swing	Reduced (asymmetric)	Reduced to normal (symmetric)	Compensatory superfluous movements
Stride length	Short	Shorter	Very short
Stance phase	Variable	Longest	Long
Gait speed	Progressively rapid (festination)	Slower than Parkinson's disease	
Cadence	Higher	Relatively less	None
Falls	Late	Early	Early
Freezing of gait	Late	Common	Very common
Apraxia of gait	Rare	Variable frequency	Common
Primary anatomical substrate	Nigrostriatal pathway	Basal ganglia, thalamus, midbrain	The supplementary motor area, premotor area, medial frontal gyrus, and frontal subcortical connections

Further studies addressing the Movement Disorder Society PSP criteria subtypes are warranted to understand the natural history of these different PSP subtypes and assist in counselling patients and families about expected progression and prognosis.

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Trastorno de la marcha del nivel superior como forma de presentación de una parálisis supranuclear progresiva: descripción de un vídeo caso

Introducción. El trastorno de la marcha frontal/apraxia de la marcha es un déficit motor del nivel superior con diversas causas, caracterizado por dificultades en el inicio de la marcha (congelación). Nuestro objetivo es presentar una paciente con un trastorno de la marcha del nivel superior con episodios de caídas como manifestaciones iniciales de una parálisis supranuclear progresiva (PSP). Sus datos se obtuvieron de los registros médicos del Servicio de Medicina General del Burdwan Medical College & Hospital (Burdwan, Bengala Occidental, India).

Caso clínico. Mujer de 58 años sana que consultó por un trastorno de la marcha con caídas. La exploración neurológica mostró una apariencia facial característica (mirada fija, ojos muy abiertos, ceño fruncido y expresión fija hemifacial inferior), e hipocinesia-rigidez simétrica de predominio axial (postura retrocólica del tronco y el cuello). La exploración de la marcha reveló un trastorno de la marcha del nivel superior, caracterizado por una significativa vacilación inicial, que precisaba ayuda de objetos/personas cercanos. Al iniciar la marcha, los pasos mejoraban relativamente, pero reaparecía una deambulación inefectiva al girar. Presentaba zancadas cortas, congelación, base amplia de sustentación, desequilibrio, movimiento lento de las piernas, arrastre de los pies, y pérdida de la cadencia normal del tronco y las extremidades. Los reflejos posturales estaban alterados. La resonancia magnética cerebral desveló atrofia mesencefálica, dilatación de acueducto de Silvio y III ventrículo, atrofia frontal bilateral y el signo típico del colibrí. Finalmente, la paciente fue diagnosticada de una PSP probable.

Conclusiones. Varias etiologías, incluida la PSP, deben considerarse, en el contexto clínico apropiado, si la exploración de la deambulación demuestra un trastorno de la marcha del nivel superior.

Palabras clave. Forma de presentación. Marcha. Parálisis supranuclear progresiva. Trastorno de la marcha del nivel superior. Trastorno del movimiento. Vídeo caso.