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Early axial involvement in progressive supranuclear palsy with progressive gait freezing: A rare case report

İlerleyici yürüme donması ile seyreden progresif supranükleer felçte erken aksiyal tutulum: Nadir bir olgu sunumu

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ABSTRACT

Progressive supranuclear palsy with progressive gait freezing (PSP-PGF) phenotype represents a unique presentation pattern emerging with selective involvement of axial locomotor networks in the early stages, where clinical findings may manifest before structural imaging biomarkers become evident. This case report presents a 68-year-old male patient unresponsive to standard levodopa therapy, characterized by a two-year insidious and progressive course of recurrent and complete blockages in gait initiation, a tendency toward retropulsion, and marked axial rigidity, while limb parkinsonism remained minimal and tremor was absent. This case emphasizes the importance of differential diagnosis in PSP cases presenting with early-onset predominant freezing of gait.

Keywords: Axial rigidity, freezing of gait, interpeduncular fossa enlargement, mesencephalic contour flattening, progressive supranuclear palsy.

ÖZ

Yürüme donması-baskın progresif supranükleer palsi fenotipi (PSP-PGF), erken dönemde aksiyel lokomotor ağların seçici tutulumu ile ortaya çıkan ve klinik bulguların yapısal görüntüleme biyobelirteçlerinden önce belirginleşebildiği özgün bir sunum paternini temsil eder. Bu olguda, iki yıllık sinsi ve ilerleyici süreçte yürüme başlatmada tekrarlayıcı ve tam blokajlar, retropulsiyon eğilimi ve belirgin aksiyel rijidite ön planda olup, ekstremite parkinsonizminin minimal düzeyde kaldığı ve tremorun eşlik etmediği, standart levodopa tedavisine cevapsız 68 yaşında erkek bir hasta sunuldu. Bu olgu erken dönemde baskın yürüme donması ile seyreden PSP olgularında ayırıcı tanının önemini vurgulamaktadır.

Anahtar sözcükler: Aksiyal rijidite, yürüme donması, interpedinküler fossa genişlemesi, mezensefalik kontur düzleşmesi, progresif supranükleer palsi.

Progressive supranuclear palsy (PSP) is a neurodegenerative tauopathy characterized by central 4R-tau deposition, presenting with parkinsonism, postural instability, vertical gaze restriction, and frontal-subcortical cognitive impairment.[1] The Movement Disorders Society (MDS) diagnostic criteria provide a framework for early diagnosis and investigation by subdividing the phenotypic heterogeneity of PSP into subtypes along the axis of oculomotor dysfunction, postural instability, akinesia, and cognitive impairment.[2] The progressive supranuclear palsy with progressive gait freezing (PSP-PGF) presentation is characterized by difficulty in initiating early gait and poor response to levodopa, requiring detailed evaluation and close monitoring, as it can be confused with Parkinson's disease from a clinical perspective.[3] This case report describes a levodopa-refractory PSP-PGF presentation with prominent gait-initiation failure and axial involvement, in which conventional high-resolution magnetic resonance imaging (MRI) demonstrated mesencephalic structural abnormalities consistent with early involvement of the axial locomotor network.

CASE REPORT

A 68-year-old right-handed male patient presented with a two-year history of insidious-onset and gradually progressive gait disturbance, characterized predominantly by marked freezing

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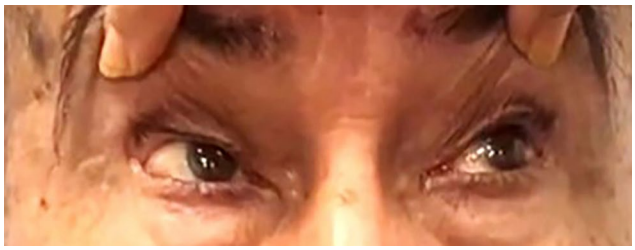

Figure 1. Clinical photograph of the patient attempting upward gaze, demonstrating bilateral supranuclear upgaze limitation: voluntary elevation of the globes is markedly restricted despite manual eyelid retraction by the examiner, a feature consistent with vertical supranuclear gaze impairment seen in progressive supranuclear palsy.

at gait initiation and recurrent falls, most of which occurred backward. He lived in a rural area and was actively engaged in farming. His medical history was notable for diabetes mellitus, coronary artery disease treated with coronary artery bypass grafting, and bilateral burr-hole drainage surgeries performed several years earlier for bilateral subdural hematomas. There was no family history of parkinsonism or other neurodegenerative disorders. Before referral, he had undergone a standardized levodopa trial for approximately one year, during which both dose escalation and adequate treatment duration were ensured; however, no clinically meaningful motor benefit was observed. At presentation, his pharmacological regimen consisted of levodopa/benzerazid 125 mg three times daily and amantadine 100 mg twice daily, in addition to antihypertensive and antidiabetic medications. On neurological examination, speech was characterized by hypophonia, reduced rate, and reduced fluency, without aphasic features. Oculomotor examination revealed a clear supranuclear limitation of upward gaze, accompanied by mildly slowed saccades, preserved convergence, and absence of spontaneous or gaze-evoked nystagmus. Motor examination demonstrated normal muscle strength in the upper extremities (Medical Research Council grade 5/5), whereas the lower extremities showed mild relative weakness related to rigidity, with proximal and distal muscle groups graded approximately 4+/5. Muscle tone was increased in a plastic-stiff pattern, predominantly affecting the axial musculature and lower limbs. Coordination testing disclosed a brady-ataxic component, with mild slowness and delayed target acquisition on finger-nose and heel-to-shin testing. Rapid alternating movements revealed slight impairment of pronation-supination, and tandem gait was unstable even over short distances, with evident disruption of step sequencing. Extrapyramidal examination highlighted marked axial rigidity, moderate bilateral lower-limb rigidity, and bradykinesia that was most prominent during movement initiation and repetitive motor tasks, while only mild involvement of the right upper limb was noted. Facial expression was mildly

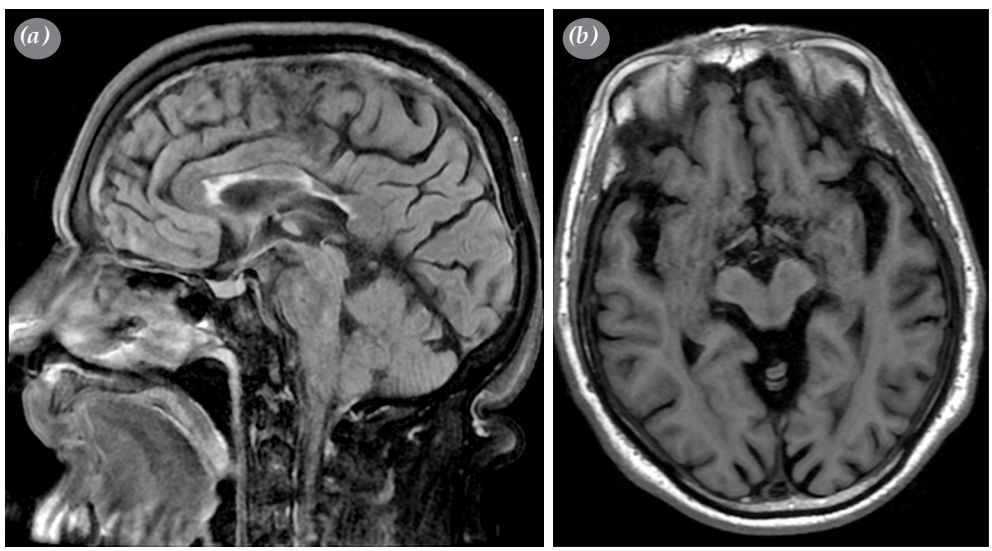

Figure 2. (a) Sagittal T1-weighted MRI and **(b)** axial T1-weighted MRI demonstrating flattening of the mesencephalic contour with loss of anteroposterior convexity and widening of the interpeduncular fossa on the sagittal plane, along with a pattern of global cortical atrophy and lateral ventricular enlargement on the axial plane.

MRI, magnetic resonance image.

hypomimic. Postural reflex testing using the pull test elicited mild-to-moderate instability, with a characteristic retropulsive tendency. Gait assessment showed a slightly forward-flexed posture, short shuffling steps, reduced stride length, and frequent complete motor blocks at gait initiation. Turning was particularly impaired, requiring pauses, corrective small steps, and external support, with a consistent tendency toward backward imbalance. Nonmotor symptom assessment was unremarkable, except for constipation. The MDS-Unified Parkinson's Disease Rating Scale-III score was 17, and the modified Hoehn and Yahr stage was 3.0. Magnetic resonance angiography demonstrated no significant intracranial or extracranial large-vessel stenosis, occlusion, or aneurysmal change. Periventricular and deep white matter hyperintensities were mild and age-appropriate (Fazekas grade 1), insufficient to account for the gait disorder. Conventional brain MRI revealed progressive global cortical atrophy and widening of the interpeduncular angle compared with prior imaging, along with chronic sequelae of previous bilateral subdural hematomas. Although quantitative measurements of midbrain and pons areas remained within reference ranges, serial imaging demonstrated progressive flattening of the mesencephalic contour and enlargement of the interpeduncular fossa. Susceptibility-weighted imaging showed residual hemosiderin deposits along the convexities related to prior hemorrhage. Preservation of the nigrosome-1 signal supported a pattern of suprasegmental and axial network involvement rather than primary presynaptic dopaminergic degeneration. Taken together, the constellation of early and dominant freezing of gait, axial rigidity, supranuclear vertical gaze limitation, minimal limb parkinsonism, tremor absence, levodopa unresponsiveness, and evolving mesencephalic morphological changes was considered most consistent with the PSP-PGF phenotype. A written informed consent was obtained from the patient.

DISCUSSION

The underlying pathophysiology of PSP-PGF goes beyond isolated nigrostriatal dopaminergic degeneration, reflecting a multi-level neurodegenerative process involving fronto-subcortical circuits, mesencephalic locomotor areas, and brainstem networks responsible for postural control and gait automaticity.[4] Specifically, loss of functional connectivity between the supplementary motor area, prefrontal cortex, and pedunculopontine nucleus (PPN) constitutes the core mechanism for gait initiation difficulty and impaired axial motor control. This network-based dysfunction also explains the limited response to dopaminergic therapies.[5]

In the PSP-PGF phenotype, "freezing of gait" is conceptualized as a loss of automaticity resulting from impaired integration between cortical executive areas and brainstem locomotor pattern generators, rather than as basal ganglion output dysfunction.[5] Degeneration in the PPN and cuneiform nuclei, which constitute the mesencephalic locomotor region, is associated with early axial rigidity, retropulsion, and gait initiation blocks. This is consistent with a clinical pattern in which extremity parkinsonism remains minimal and tremor is absent.

Quantitative biomarkers used in structural MRI gain diagnostic specificity in advanced stages of the disease, but their sensitivity may remain limited in early PSP-PGF cases.[6] In this context, subtle morphological changes, such as mesencephalic contour flattening and enlargement of the interpeduncular fossa, can be considered radiological clues of early mesencephalic involvement, even if they remain below volumetric thresholds. The progression of these morphological changes on serial imaging increases their diagnostic value when interpreted in conjunction with clinical findings. Although clinically it is difficult to differentiate PSP-PGF from freezing of gait seen in Parkinson's disease, early-onset difficulty with gait initiation, marked axial involvement, levodopa unresponsiveness, and progressive brainstem morphological changes provide strong evidence in favor of the PSP spectrum.

In conclusion, through this case, we aim to highlight the PSP-PGF variant, a rare clinical form of PSP, and to contextualize it within current advances in the understanding of PSP clinical spectrum. Early recognition of this phenotype is clinically important for predicting prognosis and initiating non-pharmacological approaches targeting balance, gaze stabilization, and gait automaticity in a timely manner.

Author Contributions

I.C.B., E.D.U., Z.Y., S.S.Ç.: Concept and design, data collection and processing, analysis and interpretation, literature search, critical review; E.D.U., S.S.Ç.: Supervision; I.C.B., E.D.U.: Writing.

Conflict of Interest

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Data Sharing Statement

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