CASE REPORT

Early ophthalmologic manifestations of progressive supranuclear palsy: a case report with updated narrative literature review

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Abstract

Objective: To highlight early ophthalmologic manifestations of progressive supranuclear palsy and emphasize their diagnostic value in clinic-radiological correlation and early disease recognition.

Methods: A 61-year-old male presenting with progressive visual disturbance, ophthalmoplegia, postural instability, and recurrent backward falls was evaluated through comprehensive ophthalmological examination and brain magnetic resonance imaging.

Results: Ophthalmological examination revealed impaired smooth pursuit, abnormal saccades, and supranuclear horizontal and vertical gaze palsy with preserved vestibulo-ocular reflexes, while visual acuity and fundus findings were normal. Brain magnetic resonance imaging demonstrated characteristic midbrain atrophy with preserved pontine structures, including the hummingbird, Mickey Mouse, and morning glory signs, supporting the diagnosis of progressive supranuclear palsy.

Conclusions: Progressive supranuclear palsy is frequently underrecognized in its early stages due to overlap with Parkinson's disease. Detailed ocular motor examination combined with neuroimaging is crucial for early and accurate diagnosis. Increased awareness of ophthalmic manifestations may enhance early recognition and multidisciplinary management.

Keywords: Progressive supranuclear palsy, Ophthalmic manifestations, Ocular motor dysfunction

Received 22 february 2026 / Accepted 13 july 2026

Introduction

Progressive supranuclear palsy (PSP), also known as Steele–Richardson–Olszewski syndrome, is a neurodegenerative parkinsonian disorder with a prevalence of five to six persons per 100,000 people. It is characterized by early postural instability, impaired voluntary vertical gaze, axial rigidity, dysarthria, and mild cognitive decline. The wide variability in clinical examination findings often leads to underrecognition or misdiagnosis of PSP [1,2].

It represents an important differential diagnosis of Parkinson's disease, accounting for approximately 6% of degenerative parkinsonism cases referred to specialist clinics [2]. PSP comprises several clinical phenotypes with overlapping features, and in its early stages is frequently misdiagnosed as idiopathic Parkinson's disease. Given the average survival of 5–10 years after diagnosis, early recognition and appropriate management are essential [3]. At the time PSP is diagnosed, patients are often already approximately three years into their disease course, representing nearly half of its overall duration. This delay in diagnosis arises from multiple factors, including postponed consultation with primary care providers, under recognition of early clinical features, and frequent misdiagnosis as depression or Parkinson's disease [4].

This review aims to evaluate the spectrum of ophthalmological manifestations in PSP and to highlight their diagnostic value in early disease recognition and clinic-radiological correlation.

Methodology

This study consists of a case report accompanied by a narrative literature review. To provide an updated overview of the ophthalmologic manifestations of progressive supranuclear palsy (PSP), a thorough search has been conducted across research databases such as PubMed and Google Scholar. The search included articles published in English up to December 2025 using combinations of the keywords "progressive supranuclear palsy," "PSP," "ophthalmologic manifestations," "ocular manifestations," "ocular motor abnormalities," "vertical gaze palsy," "saccades," and "magnetic resonance imaging." Original research articles, review articles, case reports, and clinical guidelines relevant to the ophthalmologic manifestations, diagnosis, neuroimaging, and management of PSP were included. Non-English publications, conference abstracts without full-text availability, duplicate publications, and articles not directly related to the ophthalmologic aspects of PSP were excluded. Additional relevant articles were identified through manual screening of the reference lists of selected studies.

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Case Representation

Patient's Information / Anamnesis

A 61-year-old man with no prior medical history presented with progressive visual field loss over one year, worsening in the preceding six months. He reported difficulty moving his eyes, compensating with head movements, and frequently collided with objects and lost balance while walking.

The patient reported recurrent backward falls over two years, worsening in the past eight months, along with rapid stepping, generalized rigidity, and an apathetic facial expression. He denied headache, trauma, dyspnea, resting tremor, or other systemic symptoms.

Clinical Findings:

Ophthalmological examination revealed a best-corrected visual acuity of 6/7.5 in both eyes. Overall anterior and posterior segment examinations were within normal limits, suggesting that the patient's visual symptoms were not attributable to primary ocular pathology.

Diagnostic Assessments:

Standard automated perimetry revealed asymmetric visual field loss, with moderate to severe patchy sensitivity reduction in the right eye and diffuse depression in the left eye. Spectral-domain optical coherence tomography demonstrated preserved retinal nerve fiber layer thickness and largely intact macular structure bilaterally. Fundus examination was unremarkable in both eyes. Overall, structural findings were relatively preserved despite marked functional impairment, suggesting predominant involvement of supranuclear visual pathways.

Supranuclear ocular motor dysfunction was evident, characterized by marked impairment of voluntary vertical gaze, particularly downgaze, with relative sparing of horizontal eye movements. Despite restricted voluntary eye excursions, vestibulo-ocular reflexes were preserved, indicating intact nuclear and infranuclear pathways and supporting a supranuclear origin. Fixation instability with saccadic intrusions, along with eyelid retraction and a reduced blink rate, produced a characteristic staring appearance. A mild exotropia measuring approximately 15 degrees was noted, likely reflecting impaired supranuclear control of ocular alignment. In addition, the patient exhibited a reduced blink rate and a characteristic staring facial expression

Ocular motor examination revealed slow saccadic eye movements with reduced velocity and amplitude. Attempted vertical gaze demonstrated curved, oblique trajectories consistent with a round-the-house sign. In contrast, vestibulo-ocular reflexes were preserved.

Other neurological examination demonstrated impaired coordination and postural instability, with a tendency to fall backward. Gait was abnormal. Motor examination revealed mildly reduced muscle strength, graded as 4/5 in all extremities.

Figure 2. Nine-gaze examination demonstrates preserved horizontal gaze, restricted voluntary vertical gaze, and manifest exotropia of approximately 15° in primary position.

Brain MRI demonstrates selective midbrain atrophy



Fig. 1. Color fundus photography of (a) the right eye (OD) and, (b) the left eye (OS)

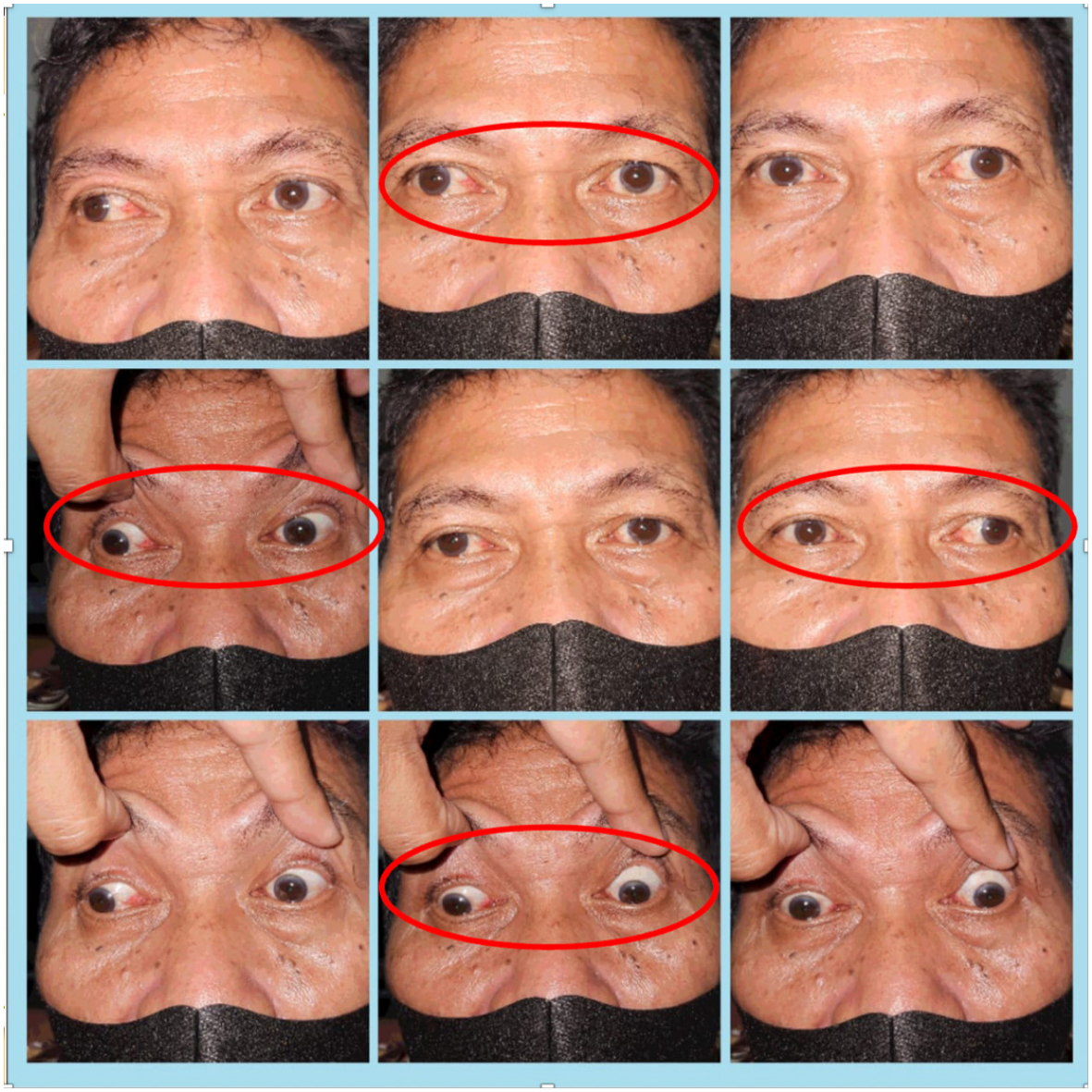


Fig. 2. Nine-gaze examination demonstrates preserved horizontal gaze,
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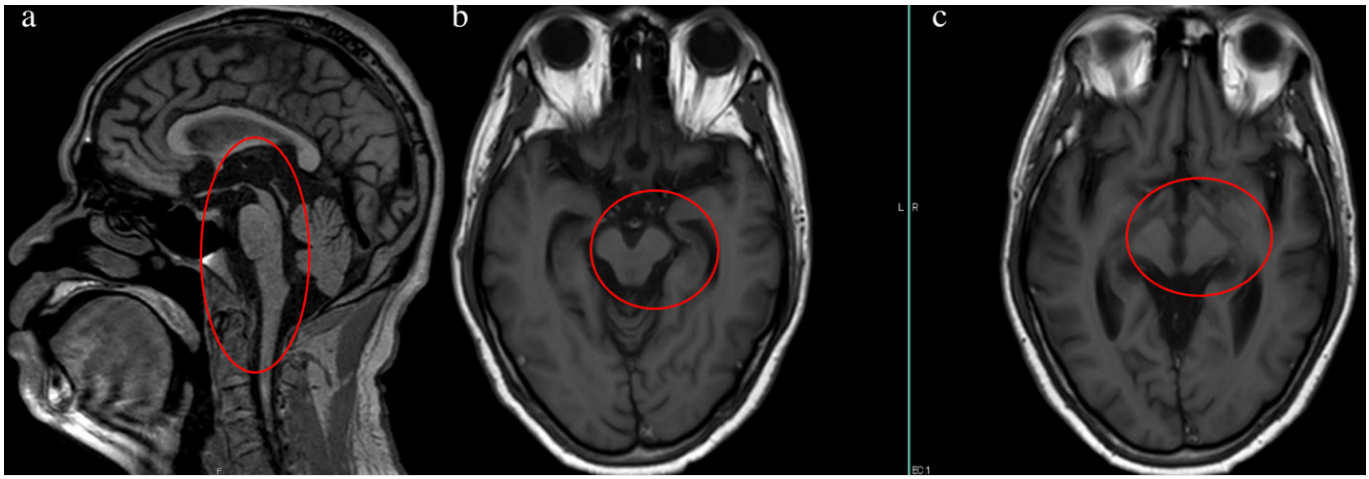


Fig. 3. Brain MRI showing selective midbrain atrophy with preserved pons and,
(a) hummingbird (penguin) sign on sagittal view,
(b) Mickey Mouse sign, and
(c) morning glory sign on axial views.

with relative preservation of the pons. On midsagittal imaging, the marked thinning of the midbrain tegmentum with a preserved pontine contour produces the classic hummingbird (or penguin) sign. Axial images at the level of the midbrain show flattening and concavity of the cerebral peduncles, resulting in the Mickey Mouse sign, while further axial sections reveal a concave configuration of the midbrain resembling a morning glory sign. These imaging features reflect preferential degeneration of the midbrain structures and thus suggest the diagnosis of Progressive Supranuclear Palsy Richardson's Syndrome (PSP-RS).

Therapeutic Intervention / Management

The patient was managed with supportive therapy, including neuroprotective agents, and was referred to the Neurology Department for further evaluation and multidisciplinary follow-up.

Discussion and Literature Review

Although most cases of PSP are sporadic, rare familial forms have been reported, including autosomal dominant inheritance in multigenerational families. Genome-wide association studies have identified several risk genes, including RUNX2, LRRK2, APOE, MAPT, and STX6 [5].

Progressive supranuclear palsy is a primary tauopathy marked by abnormal tau accumulation and neuronal degeneration, predominantly affecting the dorsal midbrain. Disruption of vertical saccadic pathways leads to impaired vertical gaze, a hallmark clinical feature [3, 6].

Microscopically, tau is a neuronal microtubule-associated protein encoded by MAPT on chromosome 17q21 that accumulates abnormally in PSP. Grossly, the disease shows frontal and marked midbrain atrophy with involvement of the subthalamic nucleus and superior cerebellar peduncle [7].

PSP-RS is the most common and classical form of PSP, characterized by early postural instability with recurrent falls, parkinsonism, vertical gaze palsy, and cognitive or behavioral changes. It typically progresses rapidly [8]. Ocular manifestations of PSP evolve in a temporal sequence that parallels disease progression, emerging across early (<4 years), middle (4–8 years), and late (>8 years) stages [2, 9].

Table 1. Leica BOND-MAX PD-L1 immunostaining assay.

PSP subtypes	Common Ocular Abnormalities
PSP-RS	- Square-wave jerks and vertical supranuclear palsy frequently occurs in the early to middle stages - Slow saccades clearly present in early to middle stages - Round-the-house sign frequently occurs in early stage - Decreased blink rate, blepharospasm, and apraxia of eyelid opening occurs occasionally and typically in the middle to late stages
PSP-OM	- A definite vertical supranuclear palsy is present. - Vertical saccades or eyelid opening apraxia may present [10]
PSP-PI	- Slow saccades, vertical supranuclear palsy, and apraxia of eyelid opening occurs occasionally in the late stage - Square-wave jerks, round-the-house sign, decreased blink rate, and blepharospasm are absent
PSP-P	- Slow saccades and vertical supranuclear palsy occasionally occurs in the middle to late stage - Eyelid opening apraxia may present [10]
PSP-Frontal	- Although the 2017 MDS diagnostic criteria incorporate ocular motor signs (e.g., vertical supranuclear gaze palsy, slow vertical saccades, or apraxia of eyelid opening) to establish diagnostic certainty, they do not specify when these abnormalities occur [10]. - A later review found that characteristic ocular abnormalities are generally absent in PSP-F in the published literature [2].
PSP-PGF	- Slow saccades and vertical supranuclear palsy occasionally occurs in middle to late stage - Blepharospasm occasionally occurs in the late stage - Square-wave jerks, round the house sign, decreased blink rate, and apraxia of eyelid opening are absent
PSP-CBS	- Slow saccades and vertical supranuclear palsy occasionally occurs in early to middle stage
PSP-PLS	- Ocular abnormalities are absent (notably, vertical supranuclear palsy is absent even in 10 years duration)
PSP-C	- Vertical supranuclear palsy occasionally occurs in early to middle stage
PSP-SL	- Vertical supranuclear palsy occasionally occurs in middle stage - Vertical saccades or eyelid opening apraxia may present [10]

PSP-RS: PSP- Richardson's Syndrome, PSP-OM : PSP-ocular motor dysfunction, PSP-PI : PSP-postural instability, PSP-P : PSP-parkinsonism, PSP-frontal : PSP-frontal, PSP-PGF : PSP-progressive gait freezing, PSP-CBS : PSP-corticobasal syndrome, PSP-PLS : PSP-primary lateral sclerosis, PSP-C : PSP-cerebellum, and PSP-SL : PSP-speech/language disorders [2].

PSP is characterized by vertical supranuclear gaze palsy and other ocular motor signs, including fixation instability, eyelid retraction, blepharospasm, and eyelid apraxia. Decreased blink rate may cause a fixed expression and dry eye. [11] Common early oculomotor features include square wave jerks, slow and hypometric vertical and horizontal saccades, and impaired vertical optokinetic nystagmus, which typically precede overt gaze palsy and aid in early diagnosis. Recently, the "eyes on ceiling" sign has been described as an additional early feature, characterized by a persistent upward gaze during the transition from supine to sitting, occurring before complete downward gaze palsy [9, 12, 13].

Comparison of the 2017 MDS diagnostic criteria with the 2019 review reveals several differences in the reported ocular manifestations of variant PSP syndromes, likely reflecting the distinct objectives of the two publications. The former focuses on diagnostic classification, whereas the latter summarizes ocular findings reported in the published literature. Besides PSP-F, discrepancies are also observed in PSP-OM, PSP-P, and PSP-SL. The diagnostic criteria state that vertical slow saccades or apraxia of eyelid opening may be present in PSP-OM and PSP-SL, while apraxia of eyelid opening may also occur in PSP-P. However, these findings are not specifically described in the later review. The most notable discrepancy involves PSP-F: the diagnostic criteria incorporate vertical supranuclear gaze palsy, slow vertical

saccades, and apraxia of eyelid opening as features contributing to diagnostic certainty, whereas the review reports that these characteristic ocular abnormalities have generally not been observed in PSP-F based on the available published literature [2, 10].

Imaging findings in PSP are considered supportive rather than diagnostic, however, brain imaging, particularly MRI, is essential to exclude alternative conditions such as normal pressure hydrocephalus, extensive small vessel disease, leukodystrophy, or intracranial mass lesions. Midbrain atrophy is a key imaging biomarker in classic PSP and brainstem-predominant variants, reflected by an elevated MR Parkinsonism Index (MRPI) due to disproportionate midbrain atrophy, with a reduced midbrain-to-pons area ratio of approximately 0.12 compared with a normal value of about 0.24. Additional supportive measurements include a midbrain-to-pons width ratio of <0.52 and a midbrain width of <9.35 mm on the midline sagittal plane. Characteristic qualitative imaging signs include the hummingbird (or penguin) sign, characterized by flattening or a concave superior contour of the midbrain that is normally upwardly convex, the Mickey Mouse sign, defined by a reduction in the anteroposterior midline midbrain diameter at the level of the superior colliculi on axial imaging to less than 12 mm, and the morning glory sign, which reflects loss of the normal lateral convex margin of the midbrain tegmentum [14,15]. Midbrain atrophy in PSP has been shown to correlate with the severity of vertical supranuclear gaze palsy, with smaller midbrain volumes significantly associated with greater impairment of upward gaze [16].

Although no disease-modifying treatments currently exist for PSP, several options are available for symptomatic management [17]. Involuntary eyelid closure is frequently observed in PSP, ranging from mild discomfort to severe cases resulting in functional blindness. This phenomenon may arise from blepharospasm, particularly pretarsal blepharospasm, or from eyelid opening apraxia, characterized by an inability to initiate voluntary eye opening despite intact peripheral levator muscle function. Symptoms may improve with botulinum toxin injections [4]. Patients with downgaze palsy may benefit from mirror-prism glasses and tinted lenses, while single-vision reading glasses are preferred over progressive lenses. Lubricating eye drops and botulinum toxin injections can help manage dry eye, blepharospasm, and dystonia [17].

Conclusion

This case highlights the importance of detailed ocular motor examination and clinic-radiological correlation in the early diagnosis of PSP. Supportive management, including neuroprotective therapy and multidisciplinary consultation with the Neurology Department, plays a key role in symptom control and ongoing care. Increased awareness of early ophthalmic manifestations may facilitate timely referral and optimized multidisciplinary management.

Author's Contribution

AAASY (Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Visualization, Writing – original draft, review & editing)
IKA (Conceptualization, Investigation, Methodology, Resources, Supervision, Validation, Writing – review & editing)
IABMP (Conceptualization, Investigation, Methodology, Supervision, Validation; Writing – review & editing)

Conflict of Interest

The authors declare no conflict of interest.

Funding

No external funding was received.

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