CASE REPORT

A Rare Case of Steel Richardson Syndrome: A Variant of Progressive Supranuclear Palsy

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ABSTRACT

Progressive supranuclear palsy (PSP) is a rare neurodegenerative disorder characterized by progressive loss of balance, walking, and eye movements. We present a case of a 65-year-old male who presented with classic Richardson Steel Syndrome, a variant of PSP, accompanied by significant ocular involvement. The patient demonstrated typical clinical features including postural instability, supranuclear gaze palsy, and cognitive impairment. Ocular manifestations included slowed vertical movements and convergence insufficiency. This case underscores the importance of recognizing PSP variants and highlights the challenges in management, particularly regarding ocular symptoms.
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INTRODUCTION

Progressive supranuclear palsy (PSP) is a rare neurodegenerative disorder with an estimated prevalence of 5.8 cases per 100,000 individuals over 40 years of age (1). It is characterized by progressive impairment of balance, walking, and eye movements. PSP encompasses several clinical subtypes, including Richardson Steel Syndrome, which is characterized by early postural instability, supranuclear gaze palsy, and cognitive impairment (2). Although ocular involvement is a hallmark feature of PSP, its presentation can vary significantly. Here, we describe a case of Richardson Steel Syndrome with notable ocular manifestations and discuss the diagnostic challenges and management strategies associated with this rare condition.

CASE REPORT

A 65-year-old male presented to our OPD with a one-year history of progressive gait instability, frequent falls, difficulty in walking fast, climbing stairs and slurred speech. The patient reported difficulty in initiating movements, particularly when getting up from squatting position while working at his Farm. Additionally, he complained of visual disturbances, including blurred vision and diplopia, difficulty in seeing objects in down gaze particularly when reading or watching television. His past medical history was significant for systemic hypertension and Type 2 Diabetes Mellitus, with no family history of any neurological disorders.

On examination, the patient demonstrated bradykinesia, axial rigidity, and impaired postural reflexes. Notably, he exhibited a stooped posture with a tendency to fall backward, suggestive of early postural instability. Neurological examination revealed supranuclear gaze palsy, characterized by impaired voluntary vertical eye movements with preserved horizontal movements. The patient also displayed a prominent frontalis release sign, consistent with the classic features of Richardson Steel Syndrome. Moreover, cognitive assessment using the Mini-Mental State Examination (MMSE) revealed a score of 22/30, indicating mild cognitive impairment.

Ophthalmological evaluation revealed slowed vertical movements and impaired convergence, with preserved horizontal eye movements. Fundoscopic examination was done and found to be normal, ruling out significant retinal pathology. Magnetic resonance imaging (MRI) of the brain demonstrated midbrain atrophy with preservation of pons, giving the characteristics 'hummingbird sign'. Ocular findings and MRI findings favoured a diagnosis of Richardson Steel Syndrome- a variant of PSP. The patient was started on a trial of Levodopa/Carbidopa, but there was minimal improvement in motor symptoms. Antidepressants were prescribed for depressive symptoms, which showed moderate improvement. Additionally, the patient was advised regular physiotherapy, occupational therapy and speech therapy. The patient was followed up every

three months. Despite the multidisciplinary approach, his condition progressively worsened, consistent with the natural course of PSP.

DISCUSSION

Progressive supranuclear palsy is a clinically and pathologically heterogeneous disorder, often presenting with a wide spectrum of symptoms and disease trajectories. PSP is characterized by the accumulation of hyperphosphorylated tau protein, forming neurofibrillary tangles. These tangles lead to neuronal loss, gliosis, and atrophy, particularly affecting the brainstem, basal ganglia, and frontal cortex.(2) Richardson Steel Syndrome, a variant of PSP, is distinguished by its characteristic clinical features, including early postural instability, supranuclear gaze palsy, and cognitive impairment.(2) The diagnosis of PSP remains primarily clinical, relying on the recognition of typical signs and symptoms. However, distinguishing PSP from other parkinsonian syndromes can be challenging, particularly in the early stages of the disease (1).

Progressive Supranuclear Palsy (PSP) is recognized as a heterogeneous disorder with several distinct clinical phenotypes. These variants include PSP with predominant parkinsonism (PSP-P) and PSP with pure akinesia with gait freezing (PSP-PAGF), which often present with symptoms overlapping other movement disorders. Cortical and cognitive-linguistic manifestations are also prevalent, categorized as PSP with corticobasal syndrome (PSP-CBS), PSP with predominant frontotemporal dysfunction (PSP-FTD), and PSP with predominant language and speech dysfunction (PSP-PNFA and PSP-AOS). Additionally, rarer presentations involve PSP with cerebellar ataxia (PSP-C) and PSP with primary lateral sclerosis (PSP-PLS) (1).

When fully developed, the PSP-RS clinical syndrome is distinctive and usually easily differentiated from other parkinsonian disorders. However, in early or variant cases, increasing evidence suggests that biomarkers may help to improve diagnostic accuracy. Over the past decade, several potential neuroimaging, biological and neurophysiological biomarkers have been described as potentially helpful in differentiating PSP-RS from other parkinsonian syndromes. However, the diagnostic value of these biomarkers cannot be established without adequately powered studies in autopsy confirmed cases (3). Atrophy of the midbrain and superior cerebellar peduncles (SCP) has been found to be a useful marker in differentiating PSP-RS from other parkinsonian syndromes and can be tracked longitudinally using diffusion tensor imaging. Resting-state functional MRI (fMRI) is also a potentially promising imaging biomarker for PSP (3).

Ocular involvement is a hallmark feature of PSP, with vertical gaze palsy being the most prominent ocular manifestation. However, our patient exhibited additional ocular abnormalities, including impaired convergence and slowed vertical saccades. Convergence insufficiency, characterized by difficulty in maintaining binocular alignment during near tasks, has been described in various neurodegenerative disorders, including PSP, but its prevalence and clinical significance remain unclear (1). A few studies demonstrate decreased pupillary diameter in darkness (PDD) in PSP patients as pupillomotor abnormality in addition to other oculomotor signs in comparison to Parkinson's disease (PD) and Multiple system atrophy (MSA) (4). This provides a simple diagnostic test for convergence insufficiency and helps distinguish PSP from other neurodegenerative disorders. Slowed vertical saccades may also reflect dysfunction of the brainstem and midbrain structures involved in saccadic eye movements.

Management of PSP remains largely symptomatic, focusing on the reduction of motor and non-motor symptoms. Medical management with dopaminergic agonists and cholinesterase inhibitors, have shown limited efficacy in reducing symptoms and are often associated with adverse effects (2). Botulinum toxin can be considered to treat blepharospasms and other dystonic manifestations (2). Physical therapy and occupational therapy play a crucial role in managing gait disturbances and fall risk. Additionally, speech therapy may be beneficial for patients with dysarthria and swallowing difficulties. A variety of small randomized placebo controlled clinical trials have been performed of which co-enzyme Q10 showed some symptomatic relief in a 6-week study. Deep brain stimulation of the pedunculopontine nucleus has been attempted in advanced cases but with no clear benefits and unacceptable side effects (5). Three clinical trials were conducted to show mild to moderate disease modifying effects: the NNIPPS study of riluzole, a phase 2 trial of tideglusib,and a Phase 2/3 trial of davunetide, but failed to show results at the clinical endpoint (5). Despite these interventions, the prognosis of PSP remains poor, with a median survival of 6 to 9 years from symptom onset (5). A variety of new biomarkers have been described to aid in the diagnosis and evaluation of new therapeutic agents. Increasing numbers of clinical trials offers hope to the patients and their families for the possibility of effective treatment.

CONCLUSION

Our case highlights the variability of ocular manifestations in PSP, emphasizing the importance of comprehensive ophthalmological evaluation in patients with suspected PSP. Although management remains primarily symptomatic, early recognition and intervention are essential for optimizing patient care and improving quality of life. Despite advances in neuroimaging and clinical research, PSP remains

a condition with significant morbidity and mortality. Continued research into the pathophysiology and treatment of PSP is essential to improve outcomes for patients with this debilitating disorder.

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