A Case of Progressive Supranuclear Palsy/Steele–Richardson–Olszewski Syndrome

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ABSTRACT
Progressive supranuclear palsy (PSP) is an uncommon neurological disorder, the hallmark of which is supranuclear ophthalmoplegia involving vertical gaze. Other clinical features include pseudobulbar palsy (dysphagia and dysarthria), neck dystonia (retrocollis), bradykinesia, postural instability, and repeated falls occurring early in the course of the disease, personality changes, a staring unblinking facies, mild dementia, and cerebellar and corticospinal tract signs. Magnetic resonance imaging (MRI) of the brain on midsagittal images may reveal a characteristic atrophy of the midbrain in a shape that suggests a bird, particularly a humming bird. The PSP may resemble Parkinson’s disease (PD), but the pathophysiology is distinct from PD. Here, we report a rare case of a 72-year-old man who came with difficulty in naming objects and persons, dysphagia, dysarthria, difficulty in vertical gaze, and history of recurrent fall even while in sitting down position. He was diagnosed as PSP based on clinical examination and neuroimaging. The PSP has poor prognosis.

Keywords: Hummingbird sign, Mickey mouse ears sign, Progressive supranuclear palsy, Steele–Richardson–Olszewski syndrome.

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INTRODUCTION
Progressive supranuclear palsy is an uncommon brain disorder that affects movement, control of walking (gait) and balance, speech, swallowing, vision, mood, behavior, and thinking.

The PSP was first described as a distinct disorder in 1964, when three scientists published a paper that distinguished the condition from PD. It was referred to as the Steele–Richardson–Olszewski syndrome reflecting the combined names of the scientists who defined the disorder.

CASE REPORT
A 72-year-old male patient came with complaints of
- History of recurrent falls even while in sitting down position, progressed over 1 year
- Difficulty in downward gaze since 2 months
- Difficulty in naming objects and persons since 2 months
- Dysarthria since 1 month
- Dysphagia since 2 weeks

On examination, patient’s vitals were stable. Clinical examination revealed:
- Loss of vertical and downward gazes
- Loss of horizontal gaze to left
- Difficulties with convergence (convergence insufficiency),
- Narrow-based gait
- Sway while turning to walk
- Action tremors on outstretched hands
- Bilateral brisk knee jerks and equivocal plantars
- Bilateral palpmamental reflex

His mini mental state examination score was 25/30 showing mild cognitive impairment.

INVESTIGATIONS
All the lab parameters were within normal limits. Magnetic resonance imaging of the brain showed the prominence of ventricular system, basal cisterns, and cortical sulci, suggestive of cerebral atrophy with age-related changes in periventricular white matter (Fig. 1). Midsagittal images showed characteristic atrophy of the midbrain in a shape that suggests a bird, particularly a humming bird.
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the midbrain with relative preservation of the pons, the “hummingbird sign” (Fig. 2). This is a typical pattern of MRI in PSP and relates to midbrain atrophy. Axial images showed “Mickey mouse ears” sign (Fig. 3), which reflects selective atrophy of the tegmentum, with relative preservation of the tectum and cerebral peduncles.

**COURSE DURING HOSPITALIZATION**

Based on clinical features and MRI findings, the patient was diagnosed as PSP. He was started on a combination of Levodopa (100 mg) with Carbidopa (25 mg) 8 hourly. He was also given balance and gait training. Ophthalmic evaluation was done and ortho-optic exercises were taught to avoid recurrent falls. His relatives were educated about the progression of the disease and the care to be provided to the patient in future. He was discharged with advice of follow-up after 3 months. On follow-up, he came with deterioration of upward and downward gaze palsy, which was persistent.

**DISCUSSION**

The PSP, also known as Steele–Richardson–Olszewski syndrome, is a neurodegenerative disease that affects cognition, eye movements, and posture. The classical diagnostic feature of PSP is paresis of conjugate gaze. Initially, there is a problem with looking up and down on command (saccade), and as the condition advances, there is difficulty in following objects up and down (pursuit). Although the disturbance with gaze is first apparent in the vertical plane, ultimately horizontal gaze also becomes affected as in the present case.

Interruption of the saccadic and pursuit pathways before they reach the eye-movement generators results in loss of voluntary eye movements. There is an overreaction of the frontalis muscles giving “astonished facial expression” or “perpetual surprise” (as opposed to the lack of facial expression/hypomimia seen with PD) and extension of the neck to compensate for the weakness of upward conjugate eye movement. In PSP, the typical neck posture is one of extension rather than flexion, as in PD. Patients with supranuclear palsy usually have an akinetic syndrome involving all limbs, with prominent rigidity of the neck and impairment of righting reflexes. As the condition advances, there is intellectual impairment. The falling in PSP is more often backward, probably because of retrocollis (tendency to tilt the head backward). In PD, the patient tends to fall forward as if chasing center of gravity. Present case had history of recurrent falls as early symptom.

Two main clinical subtypes have been described:

1. Richardson’s syndrome (54%), which features early appearance of falls, absence of tremor, symmetry of signs, and poor response to Levodopa.
2. Progressive supranuclear palsy–parkinsonism (32%), characterized by delayed onset of falls, presence of tremor, asymmetry, and response to Levodopa.

The clinical picture of PSP often overlaps with that of PD and, hence, is usually referred as a form of atypical Parkinsonism. The PSP is the most common of the so-called Parkinson plus syndromes.

Several features, however, may alert the astute clinician to the possibility of PSP. These include early instability and falls, especially in the first year of symptom onset, speech and swallowing difficulties early in the disease course, florid frontal lobe symptomatology, such as apathy, impaired abstract thought, decreased verbal fluency or frontal release signs (bilateral palmomental reflex was evident in present case), and a predominantly axial pattern of Parkinsonism. Unique MRI features of midbrain atrophy, eye signs, and falling history clinch the diagnosis. Other degenerative conditions that can
manifest a supranuclear vertical gaze disorder are corticobasal–ganglionic degeneration, Lewy body disease, Parkinson disease, and Whipple disease, but never to the extent seen in PSP. The PSP is tauopathy, while PD is ubiquitinopathy.8 In PSP, abnormal tau is present in brain areas that are consistent with the clinical signs. The gross pathologic findings in PSP are substantia nigra and locus coeruleus pigmentation with midbrain atrophy. Variable atrophy of the palladium, thalamus, and subthalamic nucleus together with mild symmetric frontal volume loss may also be present.

The age of onset of this disease process is usually in the sixth and seventh decades (average age 63 years), and it advances more rapidly than Parkinson's disease. There is sometimes a family history suggesting autosomal dominant inheritance. Males and females are affected almost equally and there is no racial, geographical, or occupational predilection. The prevalence of PSP is age-dependent and estimated at 6 to 10% of that of PD, or 6 to 7 cases per 100,000.9 Nearly half of all patients are markedly disabled or wheelchair bound within 4 years of onset. Early onset, the presence of falls, slowness, and inability to move the eyes downward early in the development of the disease predict poor survival time.

No evidence-based therapies are available, although physical therapy with balance training and family education may be helpful.

In the early stages of clinical evolution, there may be some response to dopaminomimetic therapy, but none in later stages. Death, occurring in 2 to 12 years (the median survival is 9.5 years), is often due to the sequelae from falls or dysphagia (may cause aspiration pneumonia).7

CONCLUSION

The PSP is one of a number of diseases collectively referred to as Parkinson plus syndromes. Early falls are characteristic, especially with Richardson syndrome. A case of a 72-year-old male patient with PSP is presented. Clinical presentation and brain MRI features are described.

REFERENCES


