A Case Report on Progressive Supranuclear Palsy
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Abstract
Progressive supranuclear palsy (PSP) is a quite rare neurodegenerative disease that affects mainly the brain stem, basal ganglia, and cerebellum in the central nervous system. PSP is classified in the rubric of parkinsonism plus syndromes, which causes progressive degeneration in the midbrain areas those controls eye movements. PSP often goes under diagnosed, hence making it essential for physicians to be well-known of this disorder. We present a case of PSP of a 65-year-old female, which came primarily with reiterative falls due to postural instability and speech disturbances. Radiodiagnosis with magnetic resonance imaging (MRI) of brain diagnosed a classical sign of PSP.

Keywords: Hummingbird; Penguin sign; Progressive supranuclear palsy; Mickey mouse sign; Neurodegenerative disease

Introduction
Three Canadian Physicians, Richardson J. C., Steele J., and Olszewski J., in 1963, had reported eight cases with a syndrome comprising supranuclear ophthalmoplegia, pseudobulbar palsy, nuchal dystonia, and dementia, which they titled as "a study of eight cases of heterogeneous system degeneration"[1]. Then it was named Steele–Richardson–Olszewski disease and this neurodegenerative syndrome is now called progressive supranuclear palsy (PSP)[2]. Since it shares some features of Parkinsonism; it may be sometimes grouped in "parkinsonism plus syndromes"[3]. The exact cause of this disease is not known; evidence suggests it is due to abnormal deposition of tau protein in neuronal tissues. PSP is a disease of middle or late age, which affects both sexes coequally after the sixth decade of life, with a prevalence rate of 5-6 per 100,000 and median survival of 7-12 years from the period of its diagnosis[4]. The presentation in patients suffering from PSP may vary like as in they can present with complaints of dysarthria, multiple falls with direction of fall will be backwards, they will have a downward gaze palsy, rigidity will be predominantly axial, and eventually will have cognitive impairment[5]. Magnetic resonance imaging (MRI) is always
important and helpful in making this diagnosis of PSP. We present a case of early onset PSP presenting with an abnormal eye movement and repeated falls.

Case presentation
A 60-year-old middle aged lady from rural village of Latur, Maharashtra, presented with her brother to the outpatient department with drooling of saliva, not able to maintain posture after standing with neck and body leaning backward, coughing while eating food and drinking water and soiling her clothes and fixed upward gaze with onset six months history prior to presentation. The patient suffered multiple falls at home due to lack of ability to look downwards. As days passed with her illness, she inclined to be silent and uncommunicative. She remained distant and showed lack of friendliness and lost interest in routine activities. She had difficulty in swallowing and needed help as the disease progressed. Sleep and appetite were impaired with no previous history of hallucinations, forgetfulness, and urinary incontinence. Family history was insignificant. No treatment had ever been sought.

Investigations and clinical findings
On general examination vital parameters were normal with no orthostatic hypotension. A dinner fork deformity in right and left wrist joint due to recurrent falls were noticed while other systemic examination was normal. On examining the nervous system, pupils were bilaterally equal and reactive to light with no nystagmus, but eyes were fixed with an incessant staring upward gaze, (Figure 1) depicting supranuclear horizontal and vertical gaze palsies. Dolls eye reflex was present. Axial rigidity was noticed in the form of nuchal rigidity, stiffness of the back muscles and tone was increased in all four limbs, with no tremor and grade 4 power existed in all the four limbs. All deep tendon reflexes were brisk and plantar were flexors. Bradykinesia with difficulty in walking was present. Glabellar tap was positive, sensory examination was normal and there were no cerebellar signs.

Figure 1: Supranuclear ophthalmoplegia
On mental status examination, the patient was conscious and followed all the commands. Memory was intact and dysarthria was present, mini mental status examination revealed pronounced micrographia with score of 20/30. Fundus examination was normal. Routine investigations were within normal limits. MRI brain showed a characteristic “Humming Bird sign/ Penguin sign” in axial view which means atrophy of the midbrain tegmentum, with a relatively preserved pons, decreased midbrain-to-pons ratio with a superior aspect concavity, resembling the head and body, respectively, of a humming bird along with atrophy in the frontal lobe (Figure 2) and in axial MRI brain in figure 3 shows selective atrophy of midbrain with preservation of tectum and “Mickey mouse sign”.

![Figure 2: Hummingbird sign](image)

![Figure 3: Mickey mouse sign](image)
Therapeutic Intervention

This was mainly symptomatic in the form of citicoline (400 mg twice daily), piracetam (800 mg twice daily), and clonazepam (0.5 mg at night) for sleep. Levodopa and carbidopa (110 + 10 mg in two divided doses) was added by the neurophysician. Physiotherapy was started.

Outcome and follow-up

After two weeks of treatment minimal improvement was observed. Sleep improved slightly but communication remained poor. The poor prognosis was explained to the relatives.

Discussion

The diagnosis of PSP in this case was made with the clinical research criteria given by the National Institute of Neurological Disorders and Stroke (NINDS-SPS) (Table 1)[6,7]. The Humming bird sign or Penguin sign is radiologically diagnostic of PSP. Humming bird is a small bird with a characteristically slender long beak or bill (Figure 2). The bird’s body is represented by atrophy of midbrain tegmentum seen in mid-sagittal T1-weighted images. It shows a 100% sensitivity in diagnosing PSP[8]. Generally, the upwardly convex outline of the superior aspect of the midbrain is flattened or incurved[9]. A diagnosis of PSP was made after eliminating other Parkinson’s diseases. PSP often overlaps with Parkinson’s disease, but there are differences such as an exiguous response to levodopa, peculiar pathological characteristics, and very poor prognosis[1,9,10]. To our dismay, no efficacious treatment guidelines are available for PSP. Various research showed the use of rivastigmine for cognitive enhancement and zolpidem to improve sleep, but these are anecdotal in nature[11-13]. The role of a multispecialty treatment team is a must in the management of this complex disorder. Many a times, PSP is misdiagnosed, so better understanding of PSP can help the clinicians to identify the condition. The role of neuroimaging as a tool in diagnosis is crucial along with one’s clinical judgment.

Our patient was diagnosed as PSP based on the following:

Inclusion criteria “probable”: Our patient had vertical supranuclear palsy and postural instability with recurrent falls within first year of disease onset.

Exclusion criteria: Our patient had no history suggestive of encephalitis, no cortical sensory deficits, hallucinations, delusions, cerebellar symptoms, and evidence of dysautonomia.

Supportive criteria: Our patient had symmetrical rigidity, akinesia, retrocollis, early cognitive impairment, dysarthria, poor response to levodopa.
Table 1: In 2003, the NINDS SPSP and the Scientific Issues Committee of the Movement Disorders Society formed a task force to evaluate the above diagnostic criteria\textsuperscript{[14]}

<table>
<thead>
<tr>
<th>Diagnostic categories</th>
<th>Inclusion criteria</th>
<th>Exclusion criteria</th>
<th>Supportive criteria</th>
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<tbody>
<tr>
<td>Possible</td>
<td>Gradual progressive disorder with age of onset at 40 years or later</td>
<td>For possible and probable Recent history of encephalitis, alien limb syndrome, cortical sensory deficits, focal frontal and temporoparietal atrophy, hallucinations, and delusions unrelated to dopaminergic therapy, cortical dementia of Alzheimer type, prominent early cerebellar symptoms or prominent dysautonomia, or evidence of other disease that could explain the clinical features</td>
<td>Symmetric akinesia or rigidity, proximal more than distal, abnormal neck posture especially retrocolis, poor or absent response of parkinsonism to levodopa, early dysphagia and dysarthria, early onset of cognitive impairment including 2 or 3 of apathy, impairment in abstract thought, decreased verbal fluency, utilization or imitation behaviour, or frontal release signs</td>
</tr>
<tr>
<td>Probable</td>
<td>Either vertical supranuclear palsy or both slowing of vertical saccades and postural instability with falls and &lt;1 year of disease onset</td>
<td>Vertical supranuclear palsy and prominent postural instability with falls within 1st year of onset</td>
<td></td>
</tr>
<tr>
<td>Definite</td>
<td>All criteria of possible and probable PSP are met and histopathologic confirmation on autopsy</td>
<td></td>
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Table 2: Key differences in symptoms and signs in PSP and Parkinson’s disease\textsuperscript{[15]} Considerable clinical diversity in presentation of PSP as compared to parkinsonism

<table>
<thead>
<tr>
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<th>PSP</th>
<th>Parkinson's disease</th>
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<tbody>
<tr>
<td>Symmetrical</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Rigidity</td>
<td>Axial</td>
<td>Limb</td>
</tr>
<tr>
<td>Akinesia</td>
<td>Severe, global</td>
<td>Mld to moderate</td>
</tr>
<tr>
<td></td>
<td>Even in loose limbs</td>
<td></td>
</tr>
<tr>
<td>Tremor</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Falls</td>
<td>Early, spontaneous</td>
<td>Late, with freezing</td>
</tr>
<tr>
<td>Eyes</td>
<td>Vertical paresis</td>
<td>Normal*</td>
</tr>
<tr>
<td>Voice</td>
<td>Dysarthrophonia, distorted, poor volume control</td>
<td>Hypophonia, quiet</td>
</tr>
<tr>
<td>Cognition</td>
<td>Marked early executive changes, Loss of fluency</td>
<td>Subtle early executive changes or later dementia</td>
</tr>
<tr>
<td>Levodopa</td>
<td>Poor response</td>
<td>Very good response</td>
</tr>
<tr>
<td>Gait</td>
<td>Head up, sniffing the air, Leaning back</td>
<td>Head down, stooped, leaning forward</td>
</tr>
<tr>
<td>Looks like</td>
<td>Parkinson's?</td>
<td>No</td>
</tr>
</tbody>
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Conclusion
As many PSP cases are clinically misdiagnosed, a distinctive neuroradiologic feature might improvise the differential diagnosis. Here our present case report of PSP demonstrated how MRI of the brain assisted in diagnosing PSP. Therefore, by this case report we conclude that PSP can be diagnosed clinically aided with radiodiagnosis. During the same it was analyzed that non-pharmacological management of PSP is very essential as of pharmacological treatment and should be started earlier too. Although none of the pharmacologic therapy will stop progression of this disease, a multidisciplinary team can serve these patients and their caretakers with the tools to maximize quality of life and minimize wearying symptoms.

Declaration of Patient Consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Ethical Approval: N/A
Conflict of Interest: Nil
Financial Disclosure: None

References


