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A Rare Case of Progressive Supranuclear Palsy

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Abstract: ***Progressive supranuclear palsy (PSP), also known as Steel Richardson syndrome is a form of atypical Parkinsonism with readily distinguishable features from Parkinson's disease. It is a clinical diagnosis; imaging helps to differentiate mimics.1Non - pharmacological management of PSP is as important as pharmacological treatment and should be implemented early to empower patients and their families to make well - informed autonomous decisions about their care, aimed to preserve their quality oflife.[1, 5]***

Keywords: atypical parkinsonism, progressive supranuclear palsy, movement disorder, multiple system atrophy, recurrent falls

1. Background

PSP is a movement disorder characterized by recurrent falls, progressive truncal rigidity and difficulty in voluntary eye movements.[1] However, this patient had no complaint of recurrent falls, making this case all the more extraordinary. PSP frequently progresses to Multiple System Atrophy and other atypical Parkinsonian disorders or to an overlapped state with mixed features, and is therefore often misdiagnosed if the patient presents late in the clinical course of the disease.4 It mainly affects males above 50 years of age. Treatment is primarily supportive, although a levodopa trial maybe effective in some cases.[2]

2. History

A 60/M farmer, chronic bidi smoker presented with complaints of, Tremulousness of both hands at rest, insidious in onset, slowly progressing, not task specific, since 1 year.

Progressive slowness of movements in form of generalized slowing of daily activities like difficulty in turning in bed, getting up from chair, since 1 year.

Persistent mouth opening with difficulty in chewing of solid and liquid food, and drooling of saliva, since 1month.

There were no complaints of recurrent falls, forgetfulness, bowel bladder disturbances and no history of trauma.

3. Clinical Examination

General Examination:
Patient was conscious, cooperative and oriented to time, place and person, was poorly built and nourished with generalized wasting.

BP:122/80 mmHg
Pulse: 92 bpm
Temp: Normal
Spilled food on clothes
Persistent mouth opening with drooling of saliva
Bilateral tremors in both upper limbs
Staring look on face
Curved up position with inability to lie down flat in bed
Extended Neck and Head
Widened palpebral fissure
Reduced eye blinking

Signs of autonomic dysfunction were absent.
Rest of the examination was normal.

- Systemic Examination:

CNS:
Handedness: Right - handed

Higher Mental Functions	
Consciousness	Conscious
Orientation to Time/ place/ person	Oriented
Speech and Language	Dysarthia- present, hypolinetic, hypophonic, with micrographia
Sleep	6 hours, poor quality, reduced with frequent awakenings and delayed latency
Memory	Not testable

No hallucinations/illusions/delusions

Cranial nerves:
3, 4, 6: impaired convergence, slowing of saccades and pursuits, with vertical upgaze palsy, mild eyelid apraxia, normal pupil size and reactivity.
Rest: Normal

Motor functions:	Right UL	Left UL
Nutrition	POOR	POOR
Tone	INCREASED(rigidty:cogwheel)	INCREASED(rigidty:cogwheel)
Power	4	4

Motor functions:	Right LL	Left LL
Nutrition	POOR	POOR
Tone	INCREASED(rigidity:leadpipe)	INCREASED(rigidity:leadpipe)
Power	4	4

Coordination: Normal

Extra pyramidal system findings:
Bradykinesia: decreased arm swing while walking
Finger tapping, toe tapping, heel tapping suggestive of bradykinesia.

Involuntary movements:
Tremors: slow, coarse, compound, 4 - 5 Hz, present at rest, rhythmic, decreased with movement temporarily, decreases during sleep, in bilateral hands and forearm.

Oromandibular dystonia of mouth, lip and jaw, sustained, not associated with tremors, isolated, not task specific, non - progressive, no diurnal variation.

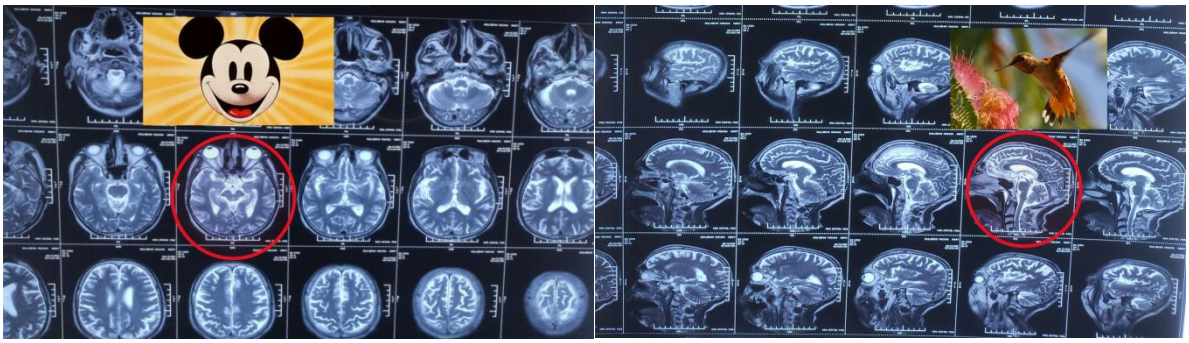
Reflexes	
Superficial	Normal
Deep	Normal
Primitive Reflexes	Absent
Glabellar Tap	Persistent blinking bilaterally beyond 4 taps

Posture, stance and gait: narrow based, slow, with reduced arm swing, en bloc - turning around, no deviation to either side.

Rest of the CNS examination was normal.

All other systems were normal.

Investigations:
All routine biochemical investigations were normal.
MRI Brain -
Humming bird sign on sagittal image (midbrain atrophy)
Mickey mouse sign on axial image.



Differential Diagnosis:
1) Atypical Parkinsonism (PSP)
2) Parkinson's disease

Management and Intervention:

Medical: Syndopa (100 plus 25) TDS trial for 1 month, Trihexyphenidyl 2mg BD
Under Trial: Amantadine, Donepezil, DBS?
Supportive: Physiotherapy, Occupational therapy, Speech and language therapy, RT

4. Conclusion

The case at hand is an infrequent presentation of a relatively familiar disease (Parkinsonism). Insidious onset, rapidly progressive, neurodegenerative disease, involving the motor system, with evidence of vertical gaze palsy and slowing of saccades and pursuits, and no evidence of sensory, or bowel bladder involvement, points towards the involvement of extra pyramidal system, indicating parkinsonism with atypical features, probably PSP. At present, therapeutic options are symptomatic and insufficient. Recenttrials have failed to provide a positive clinical outcome, however, have

led to the design of better studies that are ongoing and hold promise for a neuroprotective treatment for PSP.2

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