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Original Research Paper

Neurology

DILEMMA BETWEEN PARKINSONISM AND PROGRESSIVE SUPRANUCLEAR PALSY

Dr. Ajinkya Kadam	Department Of Medicine Pacific Institute Of Medical Sciences, Udaipur.
Dr N. K. Gupta	Department Of Medicine Pacific Institute Of Medical Sciences, Udaipur.
Gargi Gupta	Department Of Medicine Pacific Institute Of Medical Sciences, Udaipur.

KEYWORDS :

INTRODUCTION

Atypical parkinsonian syndromes are a subset of progressive neurodegenerative disorders that present with Parkinsonian features but differ from idiopathic Parkinson's disease (PD) in their clinical course, underlying pathology, and response to treatment. Progressive Supranuclear Palsy (PSP) is one of the key syndromes within this category. PSP is characterized by its hallmark symptoms, which includes rigidity, bradykinesia, and postural instability, alongside prominent supranuclear gaze palsy, particularly affecting vertical eye movements (Batheja et al., 2023).

Unlike PD, PSP often leads to rapid disease progression and exhibits a range of cognitive and behavioral changes that can significantly impact the quality of life. Patients with PSP typically PD, present with early falls, gait disturbances and difficulties with the eye movement control (Rowe et al., 2021). Cognitive decline may include executive dysfunction and personality changes, which are distinct from the motor symptoms commonly associated with PD.

Pathologically, PSP is defined by the presence of abnormal tau protein deposits, which accumulate in specific brain regions such as basal ganglia, brainstem and frontal cortex (Coughlin et al., 2020). These tauopathies result in progressive neuronal loss and contribute to the distinctive clinical features of PSP. Despite advancements in research, treatment options remain limited, focusing primarily on managing symptoms and improving patient comfort. Accurate diagnosis of PSP and differentiation from other forms of atypical Parkinsonism is crucial for the effective management and patient care.

This case report highlights a patient with atypical presentation of PSP, emphasizing the diagnostic challenges and the importance of recognizing key clinical features for early diagnosis. Through this report, we aim to contribute to the growing body of literature on PSP, providing insights into its clinical course and management, and to raise awareness among clinicians for better identification and care of affected individuals.

We reported a case of a 64-year-old man with previous normal neurological function who developed tremors, cramps and gait abnormality (short stepping gait).

Case Report

History

A 64-year-old male with past medical history of percutaneous transluminal coronary angioplasty (PTCA) performed 6 years back, presented to the medicine outpatient department with involuntary shaking of B/L hand with twitching of both upper and lower limbs over the preceding one year. The physical examination showed presence of involuntary rhythmic oscillatory movements of both hands on rest, with short stepping gait with heel touching to ground and with no swinging of arm while walking with glabellar sign/Myerson sign – persistent blinking with no rigidity and bradykinesia with no limb or trunk ataxia.

General examination-

The patient was well oriented to time, place, person with normal speech (GCS = 15), had an erect posture with involuntary hand movements observed on action and mask like facial expression. All cranial nerves were intact on examination and no nystagmus was observed. Bilateral wheezing present on upper, middle and lower lobe for which the patient was taking foracort (200) inhaler.

Motor examination-

The muscle bulk was normal in size and was symmetric with involuntary rhythmic movements of both upper limbs on rest with low frequency and low amplitude. Slight resistance was observed in all four limbs through whole range of movements. Muscle power, finger to nose test, heel to shin test were all normal at the time of examination. Rocket sign which is sign of motor recklessness was seen to be positive for this patient.

Sensory system examination – light touch, pain temperature, vibration, joint sense – felt at all points symmetrically.

Cerebellar sign- no dysmetria (patient was able to perform finger to nose test and heel to shin tests) with no dysdiadochokinesia (patient able to perform rapid alteration movements-pronation and supination of the hands) no sensory ataxia (patient was able to maintain balance with feet together and eyes closed). Impaired tandem walking with swing toward right side.

Investigation-

- Routine CBC, LFT, serum electrolyte was done
- MRI brain was suggested to rule out Parkinson and atypical Parkinson.

Mri Brain –

Chronic ischemic changes in fronto-parietal region along with mild cerebral atrophy was observed. The midbrain appears mildly atrophic. Further, dedicated imaging with Parkinson protocol was advised for further evaluation.

Magnetic Resonance Parkinsonism Index (MRPI)-

Indicative ratio was calculated to be 19.9 (normal value = 13.55), which strongly suggested the development of PSP (figure 1).

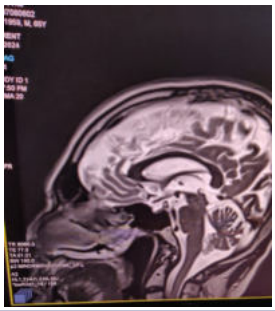

Figure 1: Sagittal T1- weighted showing decreased midbrain to pons area ratios and flattening of superior aspect of the midbrain resulting in the Hummingbird sign (arrow)

Management

Initially the patient was started on the medication prescribed by the cardiologist in review of PTCA, multivitamin injection was added for weakness.

Vitals of the patient at the time of admission:

BP: 139/76 mmHg, P.R: 78/min, SpO_2: 98%, R.R: 18/min, RBS: 146 mg/dl

The patient was prescribed multivitamins along with hypertensives which included Jupiros Gold, Tolol and Sacurise for the management of PTCA. Further the drug Pacitine was added after the diagnosis of early PSP. The patient condition was stable at the time of discharge.

DISCUSSION

Progressive Supranuclear Palsy (PSP) presents significant challenges in clinical diagnosis and management, primarily due to its overlap with other neurodegenerative disorders such as Parkinson's Disease (PD) and multiple system atrophy (MSA). This case report underscores the complexities involved in distinguishing PSP from these conditions, particularly in the early stages when symptoms can be nonspecific or misleading.

The patient in this case, exhibited a range of symptoms, including tremors, cramps and gait abnormality (short stepping gait), which initially led to a differential diagnosis that included PD. However, the subsequent development of involuntary movements along with mask like expressions and early postural instability, shifted the clinical suspicion towards atypical parkinsonism. The parkinsonism index as calculated by measuring the ratio of pons to midbrain in midsagittal plane multiplied by the ratio of average width of middle cerebellar peduncles and average width of superior cerebellar peduncles, which was found to be abnormal (19.9, normal value = 13.55). These findings align with the diagnostic criteria for PSP, which further emphasize vertical gaze palsy and a progressive course leading to severe disability within a few years of onset.

Despite the patient receiving symptomatic treatment, the progression of PSP is inevitable, highlighting the current limitations in therapeutic options. Unlike PD, where dopaminergic therapies can provide significant symptom relief, PSP does not respond well to these treatments, reflecting its distinct pathophysiology involving tau protein accumulation rather than dopaminergic neuron loss (Batheja et al., 2023).

The absence of effective disease- modifying therapies in PSP underscores the need for continued research into its underlying mechanisms. Biomarkers, imaging techniques and a better understanding of tau pathology may offer hope for earlier diagnosis and the development of targeted treatments. This case also highlights the importance of a multidisciplinary approach in managing PSP, given the complex array of motor, cognitive, and behavioral symptoms that the patient experiences.

In conclusion, PSP remains a diagnostic and therapeutic challenge, with the clinical course that rapidly leads to severe disability. Early recognition of PSP- specific features is crucial for accurate diagnosis and appropriate management, although the current lack of effective treatments remains a significant hurdle. Future research aimed on understanding the pathogenesis of PSP and developing novel therapies is essential for improving outcomes in this devastating disease.

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