Progressive Supranuclear palsy Diagnostic Dilemma? Parkinsonism?

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ABSTRACT

Progressive Supranuclear Palsy (PSP) is a neurodegenerative disorder with lesions in midbrain, basal ganglia and selected areas of cortex. Clinically it resembles Parkinson's disease but is different in some clinical aspects with different MR imaging features and unresponsiveness to levodopa therapy. We are reporting here a case of PSP in a 52 years old patient. The purpose of reporting this case is to highlight differentiating features between PSP, Parkinson's disease and other atypical Parkinson's syndromes clinically.

Kye-words : PSP, Progressive, Supranuclear, Palsy, Atypical Parkinson's syndrome

Introduction :

Progressive Supranuclear Palsy (PSP) is a neurodegenerative disorder characterized by lesions in brainstem (Midbrain), basal ganglia and selected areas of cortex presenting with cardinal features like opthalmoplegia, pseudobalbar palsy, postural Instability, vertical gaze palsy, cognitive Impairment, Parkinsonism unresponsive to Levodopa1. Few of these clinical features overlap with Parkinson's disease and that's why it is often misdiagnosed as Parkinson's disease. In recent years, MR imaging has improved diagnosis of PSP significantly. Till date, no confirmatory laboratory test is available for PSP. This case report focuses on clinical examination particularly because in early phases of the disease MRI brain may be normal which happened in our case.

Case Report :

52 years male, driver by occupation, was brought to Hospital, with chief complaints of slurring of speech with decreased verbal communication since 4 years, emotional lability and sense of imbalance since 2 years. His wife narrated history. Patient was apparently alright 4 years when he developed slurring of speech and progressive decrease in verbal communication although his comprehension was normal. His speech abnormality was gradual in onset and slowly progressive. He also had emotional liability in the form of inappropriate laughing spells since last 2 years. There was no history of excessive crying. He also had sense of imbalance and intermittent falls without loss of consciousness. He had difficulty in eating, as he was unable to look down. There was also history of difficulty in swallowingbut no history of nasal regurgitation. There was no history of seizures, involuntary movements, weakness in any limb, sensory loss or bladder/bowel problem. At his local town, patient was diagnosed as a case of cerebro-vascular episode (CVE) . Patient consulted many doctors including neurologist who suspected bulbar variant of Motor neuron disease.

There was no history of any co-morbid condition or any psychiatric illness. Patient was former smoker and occasional alcoholic. Sleep was normal and had normal bowel and bladder habits.

On Examination : His vitals were normal and general examination revealed no abnormality. In CNS Examination, patient was conscious and oriented. He had a stiff erect posture without stooping. He had dysarthria. Ocular examination revealed normal sized reacting pupil with vertical gaze palsy. Palatal sensations and gag reflex were absent. Rest of the cranial nerves were normal. He had rigidity in neck but tone was normal in limbs. Except for bradykinesis and brisk reflexes his rest of the motor system examination was normal. There were no s/o in co-ordination and gait was also normal. His CT Scan Brain, MRI Brain and MR

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Furrowed head classically seen in PSP

Extended posture and loss of balance

angiogram, EMG and NCV revealed no abnormality.

Diagnosis of PSP was made by using NINDS-SPSP (National Institute of neurological disorders and stroke society for PSP) criteria[2,4]. Patient was lebelled as a case of early PSP because MRI was normal. He was started onTab Syndopa 125 mg ½ tab BD, Tab Amantidine 100 mg OD, Tab Escitalopram 5 mg HS as advised by neurologist

Discussion :

PSP is also known as Steel-Richardson-Olszenski syndrome[1]. It was discovered in 1964. It is a rare disease, incidence being 2-6 / 1L population. It is one of the Parkinson plus syndromes. It is seen in middle age and elderly population and incidence is equal in both the sexes. PSP is commonly misdiagnosed as Parkinson's disease. But they respond poorly to L-dopa. Important clinical features are extended posture, axial rigidity, vertical gaze palsy. They often present with features of pseudo bulbar palsy (Progressive dysphagia, dysarthria and emotional lability), bradykinesia, absence of tremors, postural instability(Backward falls-retropulsion) with frequent falls.

Diagnosis of atypical Parkinsonism 3 should be considered when there is early speech and gait impairment with lack of tremors and lack of motor asymmetry with presence of diplopia or impaired vertical gaze, There is poor or no response to L-dopa, If dementia is the first symptom then Lewy body dementia should be suspected. If there are prominent cerebellar Features Multiple system atrophy (MSA)-C should be suspected and if there is orthostatic hypotension then MSA-P should be thought of.

The basic Pathology of PSP is Neurofibrillary degeneration by accumulation of tau proteins[5]. Usually it is sporadic but mutations in the MAPT gene are observed. Midbrain and pontine reticular formation, globuspallidus, substantianigra pars compacta and few selected areas of cortex are involved[1]. Destruction of midbrain and pontine reticular formation leads to atrophy of midbrain tagmentum and dilatation of 4th ventricle and aqueduct of Sylvius.

On MRI Brain, PSP is diagnosed by peculiar signs; ' Mickey mouse sign' seen due to reduction of anteroposterior midline midbrain diameter, at the

level of the superior colliculi on axial imaging. 'Humming bird sign' or' Penguin Silhouette' is a flattening or concave outline of the superior aspect of the midbrain on sagitalplanes of MRI brain. Midbrain to pons area ratio is reduced on the midline sagittal plane to approximately 0.12 (normal ~ 0.24). This is the most accurate imaging feature, which also helps to distinguish it from MSA-P (which shows pontine and midbrain atrophy).

Conclusion :

PSP is a rare neurodegenerative disorder which often misdiagnosed as Parkinson's disease or motor neuron disease. High index of suspicion along with proper neurological examination and typical MRI findings are important for correct diagnosis.

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