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Original Research Paper

General Medicine

A RARE CASE OF PROGRESSIVE SUPRANUCLEAR PALSY WITH TYPICAL HUMMING BIRD SIGN

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ABSTRACT Progressive supranuclear palsy (PSP) is a rare neurodegenerative disorder. It is one of the forms of atypical Parkinsonism, with symptoms that overlap with Parkinson's disease but progress more rapidly and respond poorly to traditional Parkinson's medications like levodopa. PSP typically affects balance, movement, speech, and swallowing, leading to frequent falls and significant disability. The hallmark of PSP is the accumulation of tau protein in brain cells, leading to their dysfunction and death, primarily in areas controlling movement and coordination

KEYWORDS : Atypical Parkinosonism (PSP), Humming Bird Sign

INTRODUCTION

Progressive Supranuclear Palsy (PSP) is a rare and debilitating neurodegenerative disorder characterized by the progressive decline of voluntary eye movements, balance, and cognitive function. This devastating condition affects approximately 5-6 people per 100,000, with a median age of onset around 63 years.

PSP is classified as a tauopathy, where abnormal tau protein accumulation in the brain leads to cellular damage and death. The disease predominantly affects the midbrain, basal ganglia, and frontal lobes, resulting in distinctive clinical features.

Key Clinical Manifestations of PSP Include:

- Supranuclear vertical gaze palsy (difficulty moving eyes upward or downward)
- Postural instability and falls
- Parkinsonian symptoms (bradykinesia, rigidity, tremors)
- Cognitive decline (executive dysfunction, memory impairment)
- Speech and swallowing difficulties
- Emotional changes (apathy, depression)

PSP's progression varies, with most patients experiencing significant decline within 5-7 years after symptom onset. Currently, there is no cure, and treatment focuses on managing symptoms and improving quality of life.

Understanding PSP's complexities is crucial for early diagnosis, optimal management, and the development of effective therapeutic strategies.

Types of PSP:

1. Richardson's Syndrome (PSP-RS): Classic PSP with prominent supranuclear vertical gaze palsy.
2. Parkinsonian Variant (PSP-P): Presents with parkinsonian symptoms, later developing gaze palsy.
3. Pure Akinesia with Gait Freezing (PSP-PAGF): Characterized by freezing of gait and difficulty initiating movements.
4. Frontotemporal Dementia with Parkinsonism (PSP-FTDP): Combines PSP with frontotemporal dementia.

Symptoms:

1. Eye Movement Difficulties:
- Supranuclear vertical gaze palsy
- Difficulty moving eyes upward or downward
- Blurred vision
2. Motor Symptoms:
- Parkinsonian rigidity
- Bradykinesia
- Tremors
- Postural instability
- Falls
3. Cognitive Decline:
- Executive dysfunction
- Memory impairment
- Language difficulties
4. Speech and Swallowing Difficulties:
- Dysarthria
- Dysphagia
5. Emotional Changes:
- Apathy
- Depression
- Anxiety

Diagnosis:

1. Clinical Evaluation
2. Neuroimaging (MRI, CT):
- Midbrain atrophy
- Hummingbird sign
3. Laboratory Tests:
- Blood tests (Ruling Out Other Conditions)
- Cerebrospinal fluid analysis
4. Genetic Testing (Tau Gene Mutations)

Treatment:

1. Medications:
- Dopaminergic agents (e.g., levodopa)
- Cholinesterase inhibitors (e.g., donepezil)
- Antidepressants
2. Rehabilitation:
- Physical therapy
- Occupational therapy
- Speech therapy
3. Lifestyle modifications:
- Regular exercise
- Balanced diet
- Stress management

Prognosis:

1. Progressive decline over 5-7 years
2. Increased risk of falls and injuries
3. Swallowing difficulties leading to malnutrition
4. Cognitive decline impacting daily activities

Current Research:

1. Investigating tau protein-targeting therapies
2. Exploring stem cell therapy
3. Developing biomarkers for early diagnosis

Case Study

66 Years old female from Rajasthan presented with Recurrent Fall since past 18 months, difficulty in talking since past 6 months, Headache since past 6 months , Blurring of Vision since past 1 month, Right Upper Limb weakness since last 20 days. No history of any chronic illness like HTN, T2DM, Seizure disorder, CVA.

Examination S/O –

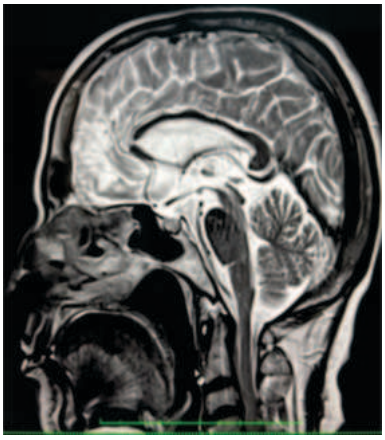
1) Backward falls and Postural instability, Unable to walk downstairs.
2) Axial Rigidity
3) Absent Facial expression with staring look (Monalisa Face)
4) Bradykinesia.
5) Freezing Gait
6) Reduced Arm Swing.
7) DTR- +3 in B/L LL
8) Plantar-B/L Extensor

Investigations

CBC- HB/WBC/PLT-11.3/11/150
RFT- UREA/CREAT-32/0.8
SODIUM/POTASSIUM-132/4.8
LFT
(T/D/I BIL)-0.48/0.25/0.23
AST/ALT -26/17
ALBUMIN/GLOBULIN/TOTAL PROTEIN-3.6/3.6/7.2
ALP-83

MRI BRAIN AND MR ANGIOGRAM OF NECK VESSELS

1) MIDBRAIN TEGMENTUM ATROPHY WITH CONCAVE UPPER BORDER OF MIDBRAIN AND TYPICAL HUMMING BIRD SIGN(MIDBRAIN TO PONS WIDTH RATIO IS 0.26), MIDBRAIN WIDTH MEASURE 4.6MM
2) ON CORRESPONDING AXIAL IMAGES DORSOLATERAL MIDBRAIN MARGIN APPEARS CONCAVE WITH DEEP INTER PEDENCULAR CISTERN POSSIBLY S/O MICKEY MOUSE SIGN.
3) GLIOTIC AREA WITH SURROUNDING HEMOSIDERIN RESIDUA IN RIGHT FRONTAL LOBE
4) SMALL GLIOTIC AREA IN LEFT BASIFRONTAL LOBE
5) MRA OF BRAIN AND NECK VESSELS DOES NOT REVEAL ANY SIGNIFICANT ABNORMALITY.



Typical Humming Bird Sign

CONCLUSION

66 Y/F came with above mentioned chief complaints. Clinical Examination and Radiological finding suggestive of PSP(TYPICAL HUMMING BIRD SIGN)

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