Atypical Parkinson’s Disease

Authors

- Dr. Nidhi Hooda (PG Resident, Department of Medicine, ABVIMS and Dr. RML Hospital)
- Dr. Aanchal Arora (Assistant Professor, Department of Medicine, LHMC and Dr. RML Hospital)
- Dr. Vishakha Mittal (Associate Professor, Department of Medicine, ABVIMS and Dr. RML Hospital)
- Dr. Moti Lal Negi (Professor, Department of Medicine, ABVIMS and Dr. RML Hospital)

Case History

- 52 year old female with no known previous co-morbidities presented to our hospital with complaints of unsteadiness of gate and difficulty in reaching objects for 6 months followed by slurring of speech for 5 months.

- These symptoms started gradually and was progressive.

- She had tendency to fall forwards and had episodes of sudden syncope lasting for few seconds after changing her position.

- There was no history of alcohol consumption, drug intake, no history of genetic diseases or similar disease in her family.

Examination

- Patient was conscious, oriented and co-operative
  - HR: 112/min, regular ; RR: 16/min ; SpO2: 98% on RA
  - BP: 134/84 mmHg (Supine position)
    108/70 mmHg (On standing after 3 minutes)
- NERVOUS SYSTEM EXAMINATION:
  - Higher mental function : Scanning speech present, rest normal
  - Cranial nerves examination : Normal
  - Motor system (Power, bulk & tone) : Normal
  - Sensory examination : Normal
  - Reflexes
    1) DTR: Upper limb – Normal, Lower limb – Pendular knee jerk present
    2) Superficial reflexes: Normal, B/L plantar flexor

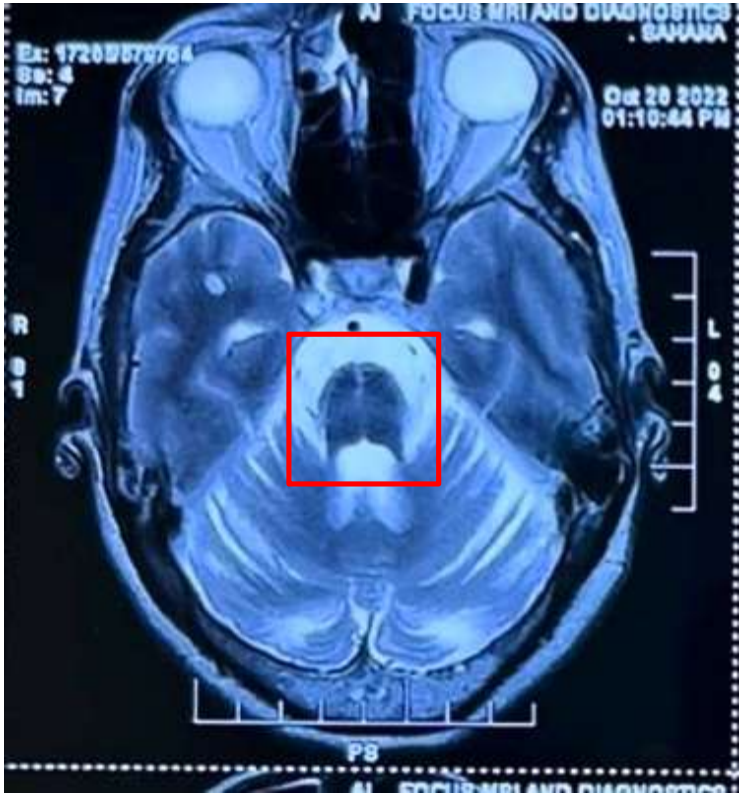
- Cerebellar examination:
  - Bilateral finger-nose-test abnormal
  - Bilateral past pointing present
  - Bilateral dysdiadochokinesia present
  - Bilateral heel shin test abnormal
  - Titubation present
  - Bilateral intention tremor present
  - Wide based gait present; tandem walking impaired
  - Nystagmus absent
- Cranium/ Spine : Normal, No meningeal signs
- CVS, R/S, ABDOMINAL EXAMINATION : Normal

Investigations

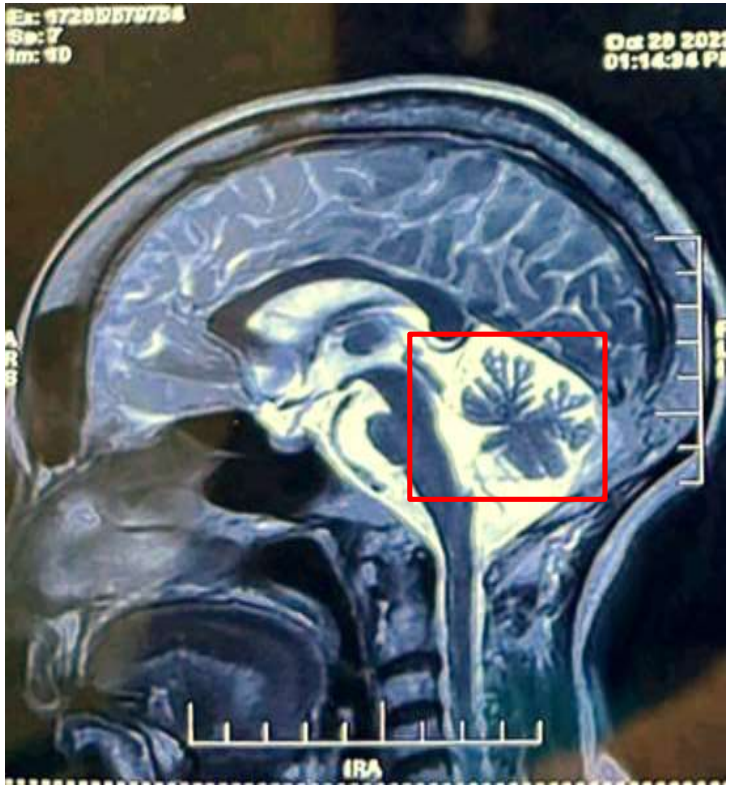
- ROUTINE Investigations
- CBC/LFT/KFT/Electrolytes – Normal
- TFT – Normal
- Vitamin B12 levels – Normal
- HIV I & II Ab – Non-Reactive
- VDRL – Non-Reactive
- CSF Studies – Normal
- ANA - Negative

- AUTONOMIC Dysfunction
- Head Up Tilt Table test : Positive
- Sympathetic Skin Response test : Normal

MRI Brain



Cruciform hyperintensity-
"HOT-CROSS BUN Sign"



Diffuse Cerebellar Atrophy

Diagnosis

- Atypical Parkinson's disease : Multiple System Atrophy (MSA-C)

Treatment

Patient was managed with non-pharmacologic measures including patient education regarding stressors of orthostatic hypotension and maintaining postural stimuli, high fluid intake (2L/D), high salt intake (10-20gm/dL), head end elevation and pharmacological measure including fludrocortisone 0.1mg/day.

Discussion

- Atypical Parkinson's diseases are neurodegenerative disorder characterised by features of parkisonism (tremor, rigidity, bradykinesia and gait abnormality) along with additional characteristics that distinguish it from idiopathic PD. It includes Progressive supranuclear palsy (PSP), Multiple system atrophy (MSA), Corticobasal degeneration (CBD), Dementia with lewy bodies(DLB) and Frontotemporal dementia.
- Multiple system atrophy (MSA) is characerised by abnormal deposition of the protein α-synuclein in the central and peripheral autonomic nervous system (synucleinopathy).
- MSA – subdivided mainly as follows:
    - Parkinsonian MSA-P (striatonigral degeneration)
    - Cerebellar MSA-C (olivopontocerebellar atrophy)

- It is important to identify the atypical features which distinguish Atypical Parkinson diseases from idiopathic PD as distinction has therapeutic and more importantly prognostic implications.
- There is no definitive therapy for MSA. Only symptomatic and supportive therapy can be provided. Overall, it has poor prognosis with mean survival of 7-9 years and mean age of onset is 54 years, most of which are sporadic.
- MSA's closest differential is Spinocerebellar ataxia type-2.
- No family history and age are against SCA in this case.
- Genetic studies have been sent and are under follow up.

- FEATURES SUGGESTING DIAGNOSIS OF MSA :

✓ Presence of cerebellar signs

✓ Presence of autonomic disturbances

✓ Rapid Progression

✓ Lack of resting tremor

✓ Lack of asymmetry

✓ Early speech and gait involvement

✓ Diffuse cerebellar atrophy

✓ Presence of Cruciform hyperintensity T2 MRI

(HOT-CROSS BUN sign indicating selective degeneration of pontocerebellar pathways)

References

1. Harrison’s Principles of Internal Medicine

2. Bradley’s Neurology in Clinical Practice

THANK-YOU