Multiple System Atrophy

Dementia Neuropsychology · https://neuro.vip/neuropsychology/dementia-neuropsychology/multiple-system-atrophy/

Multiple System Atrophy (MSA) is an **uncommon yet significant** neurodegenerative disorder that poses substantial diagnostic and management challenges for clinicians, including neuropsychologists. As a member of the "**Parkinson's plus**" syndromes or atypical parkinsonian disorders, MSA warrants close attention for its **complex symptom profile** and implications for differential diagnosis.

MSA is a **progressive neurological disease** that typically manifests in **mid-to-late adulthood**. It is characterised by **varying degrees of autonomic dysfunction, parkinsonism, cerebellar ataxia, and pyramidal signs**. Historically, MSA encompassed three separate syndromes—Shy-Drager syndrome, olivopontocerebellar atrophy, and striatonigral degeneration—which are now understood to be different clinical expressions of the same underlying pathology.

The condition is **sporadic** and has **no known cure**, with a generally **poor prognosis and rapid progression** compared to idiopathic Parkinson's disease. Mean survival after diagnosis is approximately 6–7 years, though this may vary.

Clinical Presentation in Multiple System Atrophy

Core Symptom Domains

1. Parkinsonism

Most individuals with MSA develop parkinsonian symptoms such as rigidity, bradykinesia (slowness of movement), and postural instability. Unlike Parkinson's disease, responsiveness to levodopa is usually limited.

2. Autonomic Dysfunction

Autonomic failure is a hallmark of MSA and can include orthostatic hypotension (drop in blood pressure upon standing), urinary incontinence or retention, erectile dysfunction, and impaired gastrointestinal motility. These symptoms may present early and can be very disabling.

3. Cerebellar Features

Cerebellar involvement manifests as ataxia, gait unsteadiness, slurred speech (dysarthria), and

difficulty with fine motor coordination.

4. Pyramidal Signs

Some patients display upper motor neuron signs, including spasticity and hyperreflexia.

Cognitive and Neuropsychological Profile in Multiple System Atrophy

On Neuropsychological Assessment, cognitive deficits in MSA tend to be **less severe and less prominent, especially in the early stages, than in disorders such as** dementia with Lewy bodies or Alzheimer's disease. Nevertheless, subtle difficulties in **executive functioning, attention, verbal fluency, and processing speed** can emerge over time. Patients with the parkinsonian subtype (MSA-P) are more likely to develop pronounced cognitive symptoms than those with the cerebellar subtype (MSA-C).

Neuropsychologists may encounter:

- Reduced verbal and phonemic fluency
- Slowed mental processing
- Impaired attention and working memory
- executive dysfunction (e.g., difficulty with planning, set-shifting, and inhibition)

Significant memory impairment, global dementia, or prominent visuospatial deficits are less common and should prompt consideration of alternative diagnoses.

Differential Diagnosis in Multiple System Atrophy

Given its varied presentation, MSA can be misdiagnosed or confused with several other neurological conditions. Key differentials include:

- **Parkinson's Disease**: Both present with parkinsonian signs, but MSA tends to progress more rapidly, responds less to dopaminergic medication, and features more prominent early autonomic and cerebellar involvement.
- **Progressive Supranuclear Palsy (PSP)**: PSP often presents with early gait instability, falls, and vertical gaze palsy. Eye movement abnormalities and early cognitive changes, particularly in executive function and language, are more pronounced in PSP.
- **Dementia with Lewy Bodies (DLB)**: DLB is distinguished by fluctuating cognition, well-formed visual hallucinations, and early dementia. REM sleep behaviour disorder is common in both MSA and DLB, but the cognitive profile in DLB is usually more severe.

- **Corticobasal Degeneration (CBD)**: CBD may overlap with MSA in terms of asymmetrical parkinsonism and apraxia but is often distinguished by pronounced cortical features such as limb apraxia and alien limb phenomena.
- **Pure Autonomic Failure**: Presents with isolated autonomic symptoms without motor or cerebellar signs.

Neuropathology and Brain Imaging in Multiple System Atrophy

MSA is pathologically characterised by **widespread glial cytoplasmic inclusions** (mainly composed of **alpha-synuclein**) in the brain, leading to degeneration in multiple neural systems, including the **basal ganglia, cerebellum, pons, and spinal cord**. Imaging may show atrophy in the cerebellum or putamen, but these findings are not exclusive to MSA.

Embedded media: https://radiopaedia.org/cases/71750/play?lang=gb

Brain imaging of a real multiple system atrophy Case in a 70-year old with "diagnosis certain" © Radiopaedia

Clinical Neuropsychologist’s Role in Multiple System Atrophy

Clinical neuropsychologists should be vigilant in:

- Recognising early cognitive changes that might impact daily functioning, especially executive dysfunction and processing speed deficits.
- Monitoring for acute or evolving mental status changes that may differentiate MSA from other conditions.
- Advising on cognitive strategies, compensatory techniques, and psychoeducation for patients and families.
- Collaborating within multidisciplinary teams to support diagnosis, optimise management, and address the psychosocial consequences of progressive disability.

Summary

Multiple System Atrophy poses significant challenges due to the complexity of its presentation and the overlap with other neurodegenerative diseases. For neuropsychologists, awareness of its

cognitive and behavioural manifestations, alongside motor and autonomic symptoms, can contribute crucially to the diagnostic puzzle and the holistic management and compensatory strategies of affected individuals.