Progressive Supranuclear Palsy

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Progressive Supranuclear Palsy (PSP) is a rare, degenerative neurological disorder predominantly affecting **adults over the age of 60**. Categorised as an **atypical parkinsonian syndrome**, PSP is distinguished by its **characteristic motor, cognitive, and behavioural features**. Clinical Neuropsychologists play a vital role in recognising the cognitive and neuropsychiatric profile, as well as contributing to differential diagnosis alongside neurological assessment.

What Causes Progressive Supranuclear Palsy?

Progressive supranuclear palsy (PSP) is a **neurodegenerative disorder**—meaning it involves the gradual deterioration of specific parts of the brain. The exact cause is **not fully understood**, but current research points to the following:

Abnormal Tau Protein Accumulation

The hallmark of PSP is the accumulation of **abnormal tau protein** within brain cells. Tau is a protein normally present in nerve cells (neurons) that helps stabilise their internal structure. In PSP, the tau protein becomes defective and forms twisted filaments or "tangles" inside nerve cells. This abnormal accumulation impairs and eventually kills targeted brain cells, especially in the **brainstem**, **basal ganglia**, and **frontal lobes**. This selective degeneration results in the classic features of PSP: motor impairment, postural instability, eye movement problems, and cognitive changes.

Because tau protein changes are central to the disease, PSP is classified as a **primary tauopathy** . This distinguishes it from other neurodegenerative diseases like Parkinson’s (which are usually classified as synucleinopathies). Some other diseases, such as Alzheimer’s, also involve tau pathology but with different distribution and co-existing protein abnormalities.

Contributing Factors

- **Genetics:** PSP is generally a **sporadic** condition, meaning it is not usually inherited. However, some genetic variations (such as in the MAPT gene, which encodes the tau protein) might increase susceptibility. Familial cases are very rare.
- **Environmental factors:** No clear environmental risk factors (such as toxins or infections) have been consistently identified, although research is ongoing.
- **Age:** Risk rises sharply after age 60.

Core Symptoms and Clinical Presentation in Progressive Supranuclear Palsy

Motor and Oculomotor Features

- **Postural Instability and Falls**: One of the hallmark features of PSP is early onset of postural instability, often resulting in unexplained backward falls within the first year of symptoms.
- **Supranuclear Gaze Palsy**: Impaired voluntary control of vertical (mainly downward) gaze is a defining feature. Patients often have difficulty looking down on command, which can contribute to falls and complicate daily functions such as eating or reading.
- **Axial Rigidity and Bradykinesia**: There is a pronounced stiffness, particularly affecting the neck and trunk, alongside slowed movement initiation and execution.
- **Masked Facial Expression**: A characteristic, often startled, appearance due to frontalis muscle involvement is common.

Cognitive and Behavioural Changes

- **Slowed Processing Speed**: Patients may exhibit markedly delayed response times, although accuracy can be preserved if given sufficient time. This is particularly evident during neuropsychological testing.
- **Executive Dysfunction**: Impairments in planning, problem-solving, abstract reasoning, and verbal fluency are frequently observed, reflecting frontal lobe involvement.
- **Apathy and Personality Change**: Apathy, disinhibition, irritability, and emotional lability are common. Some patients may develop depressive features, although anxiety and agitation are less frequent.

- **Memory Deficits**: Memory impairment is often secondary to executive dysfunction, particularly inefficient retrieval, and can be exacerbated by susceptibility to interference.

Neuropsychological Assessment Considerations in Progressive Supranuclear Palsy

Clinical neuropsychologists should adapt their test administration in several ways:

- **Allowing Extended Time**: Due to severe bradyphrenia, results may better reflect capacity if additional response time is permitted.
- **Functional and Behavioural Observations**: Nonverbal cues, motor sequencing, and saccadic eye movement observations may offer diagnostic clues.
- **Cognitive Profile**: Focus on thorough assessment of executive, attentional, and visuospatial domains. Memory tasks should consider both encoding and retrieval aspects.

Diagnostic Considerations and Differentials

Differentiating PSP from other neurodegenerative conditions is essential, as several disorders present with overlapping features:

Key Differential Diagnoses

- **Parkinson's Disease (PD)**: Unlike PD, PSP typically does not respond robustly to levodopa, presents with early falls, and exhibits symmetrical rigidity without the rest tremor and asymmetry more common in PD.
- **Corticobasal Degeneration (CBD)**: Both share asymmetric motor features and apraxia, but PSP more consistently shows vertical gaze palsy and early postural instability.
- **Multiple System Atrophy (MSA)**: MSA often presents with prominent autonomic dysfunction and cerebellar features, while PSP's ocular findings and backward falls are distinctive.
- **Dementia with Lewy bodies (DLB)**: PSP can be misdiagnosed as DLB, but DLB presents with fluctuating cognition, visual hallucinations, and Parkinsonism, usually without the classic vertical gaze palsy of PSP.

Pathology and Brain Imaging in Progressive Supranuclear Palsy

Pathologically, PSP is classified as a tauopathy, with abnormal tau protein accumulation affecting the brainstem and subcortical structures, disrupting cortical-subcortical networks. MRI in advanced cases may reveal midbrain atrophy (sometimes called the "hummingbird sign") and frontal atrophic changes, but imaging can be normal in early disease.

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An "almost certain" diagnosis of progressive supranuclear palsy © Radiopaedia

Clinical Management

There is currently no cure for PSP. Treatment is symptomatic, focusing on physiotherapy for mobility, speech therapy for dysarthria and swallowing issues, and occasionally pharmacological management of behavioural symptoms. It is crucial for neuropsychologists to assist in patient and carer education, support cognitive interventions, and contribute to multidisciplinary care planning.

Key References

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