A Case Study of Progressive Supranuclear Palsy in a 68-year-old with Pronounced Frontal-Executive Deficits Complicated by Cerebrovascular Disease

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Objective

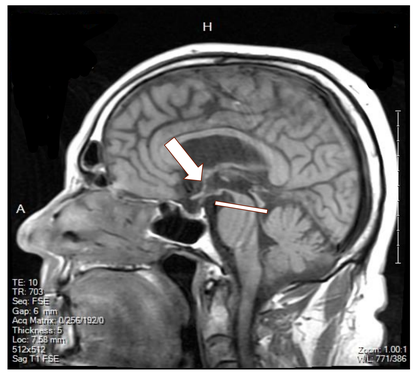
- Individuals with Progressive Supranuclear Palsy (PSP) often experience cognitive decline, postural instability and repeated falls, restricted gaze and slowing of vertical saccades, bradykinesia, and depression/behavioral changes. Their atypical parkinsonism features usually do not respond to dopaminergic therapy[1].
- Due to similar patient complaints of gait abnormalities and physical changes, behavioral and personality changes, and cognitive decline, also evident in other more well-studied neurodegenerative and movement-related disorders, PSP continues to be difficult to classify and distinguish from them. This case is an example of teasing apart and delineating these common features of neurodegenerative conditions, while illuminating frontal networks dysfunction as a forefront feature of this PSP case from a neuropsychological standpoint[1]. Further, given the high prevalence of cerebrovascular conditions in clinical populations, along with similar network involvement, cerebrovascular disease likely compounds the PSP disease course.

Participant

- A 68-year-old, white, married male with poorly-regulated hypertension and hyperlipidemia presented for a neuropsychological evaluation (April 2022) after his neurologist initially raised suspicion of possible cerebellar degeneration or multisystem atrophy based on his clinical presentation below. In the interval period prior to this current evaluation, he was diagnosed with PSP (March 2022) by a movement disorders specialist.
- In addition to reporting a progressive one-year decline in memory, motor abilities, executive functions, and a loss of independence in his activities of daily living (ADLs) (some basic and mostly instrumental ADLs), behaviorally, he also displayed significant social comportment issues with poor inhibition and impulse control. These behaviors reflected an apparent inability to organize his thinking processes and disinhibition manifested as persistent attention and concentration issues.
    - Examples of his behaviors included: Using vulgar and sexually-insinuating language with his wife in front of clinic team, making racially-pointed comments towards the examiners, and perseverating on controversial political topics despite redirection being provided.
    - He further displayed poor insight and responsiveness to the non-verbal and sometimes direct redirection and social cues in the clinic environment, but also reported being unaware of or not thinking that his behaviors were inappropriate in these and other situations that his wife was mentioning outside of the clinic setting.
- An MRI of the patient's brain completed six months prior to the evaluation simply concluded "age-related changes" by the radiologist.

Results

- The patient performed adequately on all measures of effort. He demonstrated estimated average premorbid abilities, as assessed by a measure of single-word reading. On comprehensive testing, his deficits included significant executive/attention problems, visuospatial concerns, decreased motor abilities, and variable memory functioning that were found to interfere in his ability to carry out instrumental and even some basic ADLs. Regarding memory functions, he displayed greater issues with encoding and retrieval (particularly of unstructured information), rather than a clear pattern of retention problems. Visual-centric tasks (e.g., visual learning/memory and visuomotor processing) fell largely below expectation as well. This can likely be accounted for by restricted gaze and slowing of saccades present in the patient. Aside from objective test data, subjectively the patient displayed marked disinhibition in his social and interpersonal communication abilities.
- Additionally, closer visual inspection of previous neuroimaging revealed two important neuroanatomical findings (see Figure 1).



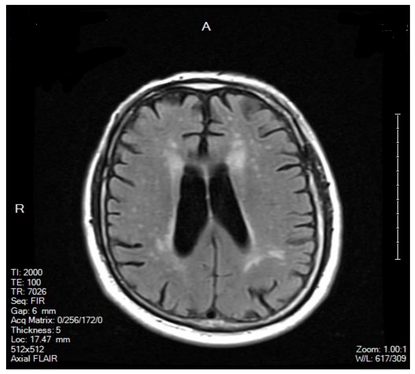
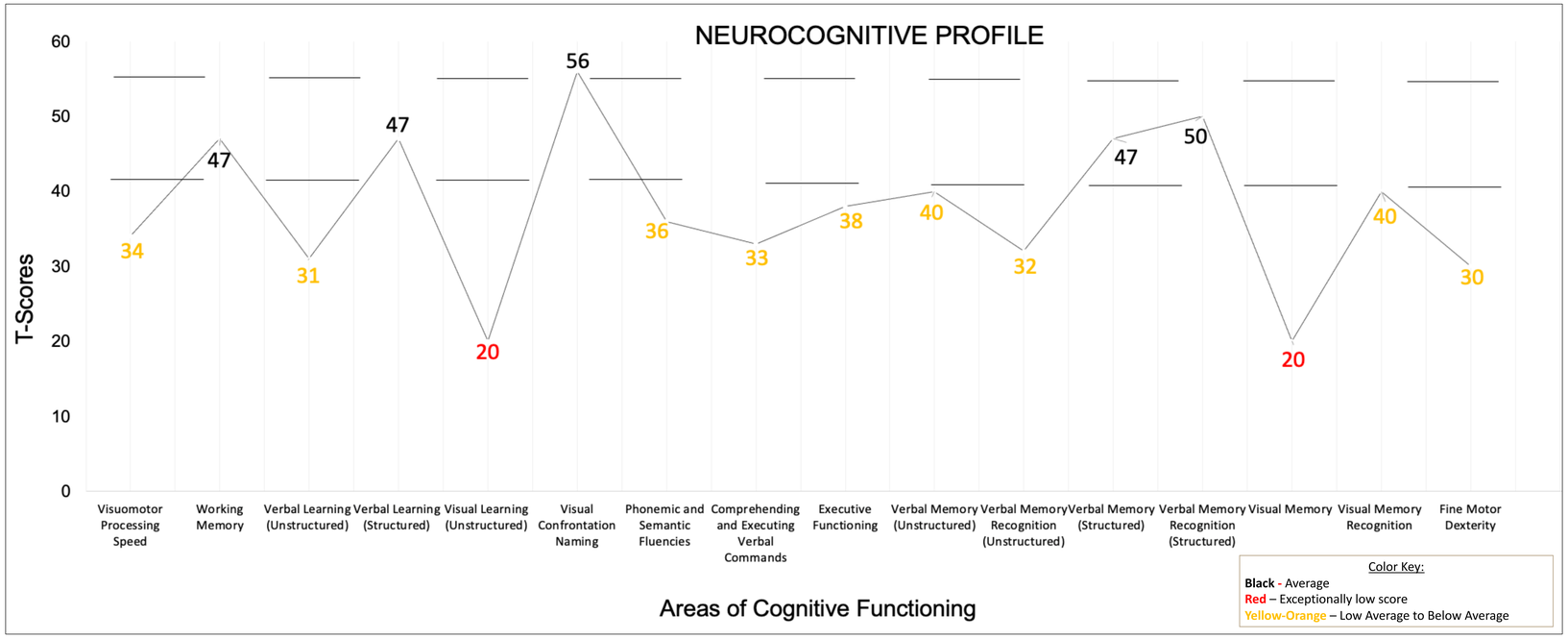


Figure 1. Despite initial neuroradiologic impressions of the patient's brain MRI, images did reveal findings that correlate to the typically more observed features of PSP.
The two important neuroanatomical changes that became apparent were:
(1) Top Image: Focal atrophy of the midbrain, resembling a so-called "hummingbird sign", on a sagittal section. The division of the white line signifies the preservation of the pons relative to the pronounced midbrain atrophy.
(2) Bottom Image: Bilateral ischemic changes in the white matter on an axial FLAIR.

Conclusions

- In distinguishing PSP neuropsychologically from other clinical disorders and neurodegenerative conditions, this case illustrates the prominence of frontal-executive deficits (e.g., disinhibition) as a particularly pronounced feature in PSP profiles. Comparatively, PSP suspected neuropathology is often thought to have greater dysfunction in striatofrontal networks[2], as opposed to reflecting retentive memory issues with hippocampal involvement[3], as is often seen in other common neurodegenerative conditions. Additionally, poorly-managed vascular conditions may act as further aggravators for individuals with PSP, superimposing on the existing attentional/executive problems present as a result of the condition.
- Given the patient's current neurocognitive status, this case illustrates the need for objective test findings to highlight cognitive deficits and behavioral changes/dysregulation when standard neuroimaging is initially reported as unremarkable.
- Additionally, this patient's neuroimaging lends support to the "hummingbird sign" (midbrain atrophy) as a useful radiologic imaging marker of PSP[4]. Although it may not entirely be clearly visible in initial stages of PSP, it may become more salient as neuropsychological dysfunction advances. Regardless, this provides another reason comprehensive neuropsychological testing is important to establish a neurocognitive and functional baseline for PSP patients for longitudinal comparative purposes.

Figure 2. General domain-level results of the participant's 2022 neuropsychological evaluation performance. Results in this table were derived from the following measures: WAIS-IV (Working Memory & Processing Speed Indices), HVLT-R, BVMT-R, Verbal Fluency (FAS & Animals), Boston Naming Test, and Wisconsin Card Sorting Test. T-Scores represent domain-level averages.



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