52 Stable Cognitive Impairment and Increased Psychiatric Symptoms in a Patient with Neuropathologically Progressive Fahr's Disease After 2-Year Repeated Neuropsychological Assessment

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Objective: Fahr's disease is a rare genetic neurological disorder characterized by abnormal idiopathic calcification of the basal ganglia, typically with extrapyramidal symptoms, speech difficulty, behavioral disturbances, and progressive neurologic dysfunction. A small number of case reports have explored the neuropsychological profile of Fahr's disease patients, and even fewer have followed the course of neuropsychological functioning over time.
Participants and Methods: A 53-year-old Asian woman presented for a neuropsychological re-evaluation (2021) after experiencing a recurrence of memory difficulties and mood changes. Relevant medical history was significant for systemic lupus erythematosus (SLE) and Fahr's disease. Following an episode of acute confusion, the patient underwent a head CT (2019) which revealed extensive calcification throughout the cerebellum, central pons, and periventricular and subcortical white matter, suggestive of Fahr's disease. Two months later, she underwent an initial neuropsychological evaluation (2019), which demonstrated prominent attention and processing speed deficits contributing to variably impaired new learning and memory along with spatial planning and problem-solving difficulties. The etiology of her cognitive deficits was determined to likely reflect metabolic and immune instability, consistent with her history of SLE and Fahr's disease. An updated CT (2021) revealed increased calcification throughout the bilateral corona radiata, basal ganglia, cerebellar hemispheres, and midbrain, which was determined to be compatible with progressive Fahr's disease.
Results: The patient's neurocognitive profile from current neuropsychological testing (2021) was marked by notable deficits in attention and processing speed, delayed memory, problem solving, visuospatial reasoning, and motor dexterity. Compared to her initial evaluation, her cognitive profile remained stable save for a slight decline in processing speed. The largest change was seen within the psychiatric domain. Self-reported depressive symptoms involving anhedonia, concentration difficulties, and anxiety symptoms involving nervousness and tension were more pronounced in her current evaluation. In addition, she endorsed an increase in apathy compared to her initial evaluation.
Conclusions: The cognitive profile seen in this patient is consistent with the current literature relating to the clinical sequelae of Fahr's disease in patients that eventually went on to develop dementia. Despite an increase in brain calcification seen on CT imaging over an 18-month interval, the patient's neurocognitive profile remained relatively stable. An increase in psychiatric symptoms appeared to be the most prominent change over repeated neuropsychological assessment, which elucidates the heterogenous course of Fahr's disease from a neuropsychological perspective. Further exploration of this disorder is warranted to better understand the clinical progression of symptoms over time.

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53 Concurrent Cognitive Predictors of School Age Academic Functioning in Children with Neurofibromatosis Type 1

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Objective: Children with neurofibromatosis type 1 (NF1) commonly have academic problems. While the neuropsychological profile of children with NF1 is variable, NF1 results in difficulties in a variety of cognitive domains including intellectual functioning, attention, executive functioning. Previous studies have suggested that cognitive functioning may relate to academic functioning in children with NF1;

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