Background: This patient is a 49-year-old, right-handed, white man with 16 years of education. He was diagnosed with a progressive demyelinating disorder in June 2008. Multiple brain magnetic resonance imaging (MRI) studies over the last few years demonstrate a progressive, widespread bilateral demyelinating process that extends from the subcortical white matter to the brainstem. At present, no cause has been identified, although several etiologies have been proposed, including multiple sclerosis (MS), central nervous system lymphoma, and delayed effects of chemotherapy. He was diagnosed with Kennedy’s disease in January 2009, which was confirmed by genetic testing (47 CAG repeats). More remote medical history for this patient includes aplastic anemia (March 1991) and myelodysplasia with monosomy 7 (November 1996); both remitted after treatment. Objectives: To examine the neurocognitive profile of a man with genetically confirmed Kennedy’s disease and a co-occurring severe demyelinating disorder of unknown etiology. Methods: This patient underwent neurocognitive testing in August 2008 and August 2009 at two different medical centers. Results: The patient reported multiple physical, cognitive, and emotional symptoms often observed in patients with MS. Symptoms included cognitive problems, fatigue, depression, muscle weakness, dysphagia, bilateral tremor, neurogenic bladder, decreased vision, facial fasciculations, and decreased appetite. Symptoms have progressed over time, and he currently uses a wheelchair and is dependent for all instrumental and select activities of daily living. Neurocognitive testing in August 2008 revealed that function was largely intact, with a circumscribed deficit in processing speed. Follow-up testing (August 2009) was significant for global deficits, including impaired orientation, verbal learning and memory, and aspects of language. Conclusions: This patient shows progressive cognitive deficits, most notably in the areas of orientation, memory, processing speed, and language. This pattern of deficits and the progression documented over time is largely consistent with a subcortical neurodegenerative process. Further understanding of the etiology of this rapid demyelination may inform prognosis and treatment options.

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