Progressive Neuropsychological Deficits Secondary to an Acute Disseminated Encephalomyelitis Attack 11 Years Later Complicated by Possible New Pathology

Mohammadi Khan, M.A., Matthew S. Phillips, M.A., Jessica Yang, M.A., Joseph W. Fink, Ph.D., ABPP-CN
The University of Chicago Medicine, Department of Psychiatry and Behavioral Neuroscience, Adult Neuropsychology Clinic

Objective

- Acute Disseminated Encephalomyelitis (ADEM) often presents as an abrupt, brief, and intense attack causing widespread inflammation and demyelination to the central nervous system, usually with antecedent events (e.g., infectious event or vaccination) preceding the illness onset.
- Majority of symptoms are more systemic rather than focal. This includes fever, headache, nausea/vomiting, along with subtle symptoms of encephalopathy manifested as decreased level of consciousness varying from lethargy, irritability, sleepiness, to coma and seizures. Multifocal neurologic symptoms may include hemiparesis, cranial nerve palsies, and other movement disorders. Behavioral changes likely also dominate the clinical picture[1,2].
- The diagnosis of ADEM is based upon clinical and abnormal craniospinal MRI findings of diffuse, large, poorly demarcated lesions (>1-2 cm) in the white, and sometimes gray matter, especially of the basal ganglia. This includes the periventricular regions and subcortical areas[1,3], along with some spinal cord lesions.
- Most patients typically make a complete neurological recovery, usually within an average 23.1 day duration of acute hospitalization and care following an attack[3]. However, a few may demonstrate long-term, persistent impairments[3].

Participant

- A 64-year-old white woman presented for a neuropsychological evaluation (2021) following a progressive 1-year decline in her memory functions and motor abilities, which overlay persisting residual deficits from an ADEM attack she had in January 2010.
  - Cognitively, her current symptoms appeared to worsen in November 2020, nearly 11 years after her ADEM attack. Her symptoms consisted of increased forgetfulness, difficulty learning novel tasks, confusion, disorientation, and sometimes incomprehensible speech.
  - Physically, she was experiencing approximately 1-2 falls per month and having difficulties with her mobility.
  - Emotionally, she was experiencing greater embarrassment due to her challenges, leading to increased defensiveness and agitation.
  - Functionally, nearly 11 years following the attack, she reported continued reliance on her family in completing most of her instrumental ADLs and some of her basic ADLs. Her husband helped her compensate and significantly supports her functioning throughout the day.
- Initial neuroimaging following the attack (2010) revealed extensive bilateral white matter (WM) lesions, that eventually appeared less conspicuous over the span of a few months. Whereas current neuroimaging (2021) (despite not explicitly commenting on the resolution status of her lesions), continued to reveal widespread hyperintense signals in WM regions, progressing cerebral atrophy, and the possibility of superimposed normal pressure hydrocephalus (i.e., ventriculomegaly).

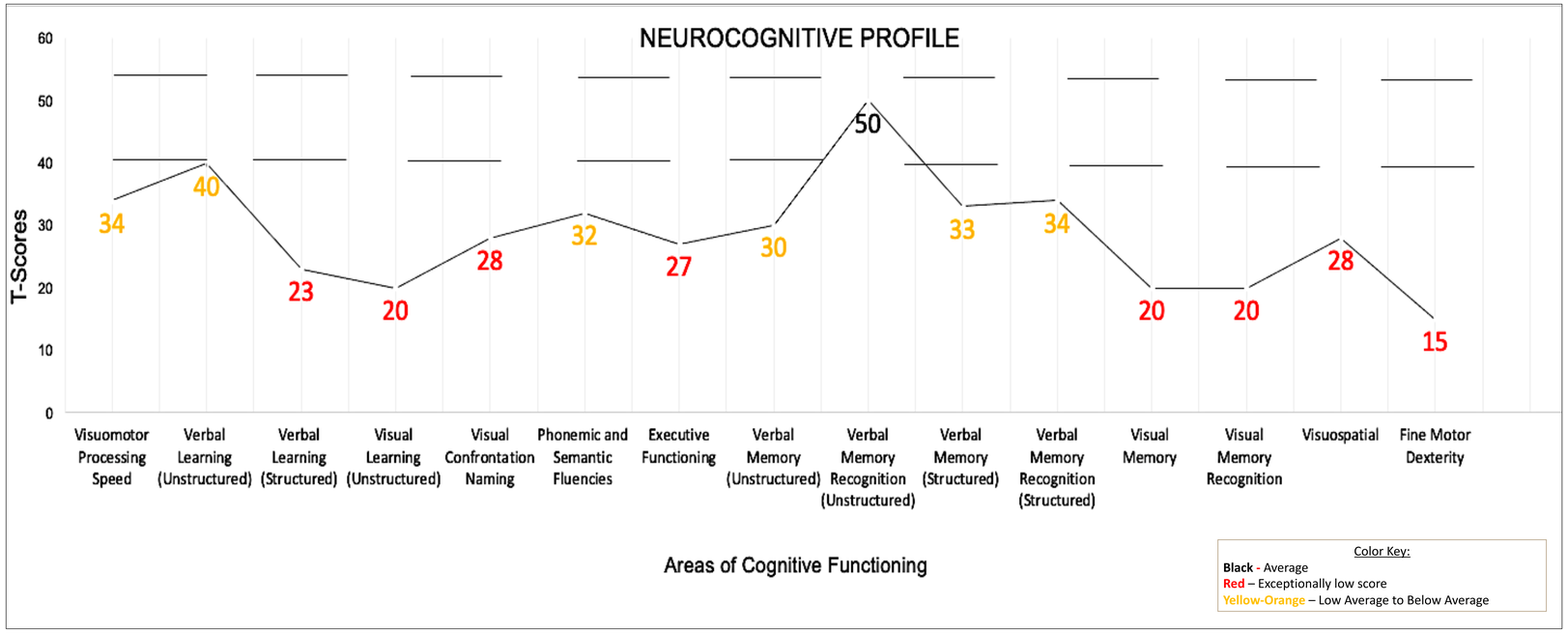

Figure 1: General domain-level results of the patient's 2021 neuropsychological evaluation performance. Results in this table were derived from the following measures: CVLT-II, WMS-IV Logical Memory, RCFT, RBANS Digit Span and Coding Subtests, Stroop Color-Word Test, Trail Making Test, BNT, and DKEFS Verbal Fluency and Design Fluency.
T-Scores represent domain-level averages.

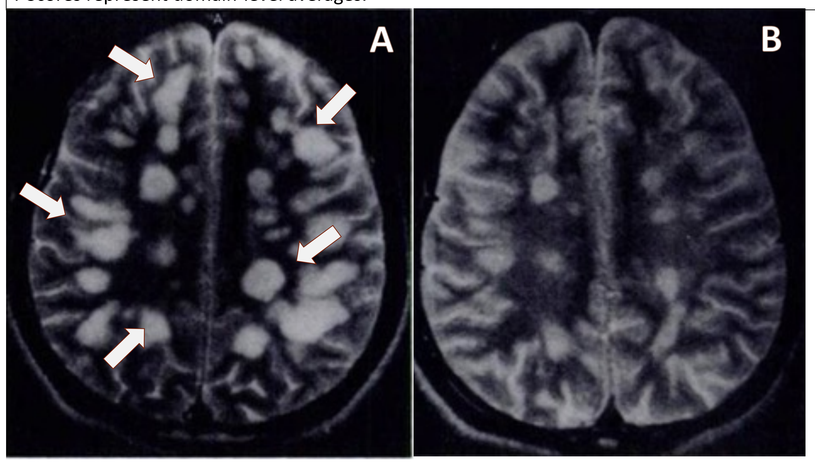

Figure 1: Pictured above is an example of bilateral symmetric presentation of ADEM in a 21-year-old man[4].

Picture A: A T2-weighted axial MR image at admission. It shows bilateral, confluent, well-defined lesions of varying sizes in the cortex, subcortical white matter, and deep white matter.

Picture B: A T2-weighted axial MR image (obtained at 3-month follow-up) showed significant reduction in the size, number, and signal intensity of the lesions[4].

Results

- Overall, her neuropsychological profile (with no prior baseline) was marked by substantial impairments in immediate and delayed verbal/visual memory, attention, language, motor abilities, visuospatial skills, executive functions, along with mild anxiety particularly related to physiological symptoms of anxiety. Most of her cognitive profile was generally below average to exceptionally low.

Conclusions

- This neurocognitive profile displayed atypical findings in patient recovery following an ADEM attack. Given the test results and the continued presence of extensive hyperintense signals in WM regions, it is possible her initial brain lesions (presumably due to ADEM) remained unresolved. This, along with the possible intervening effects of new pathology (e.g., NPH and possible vascular changes) are likely exacerbating her existing deficits related to ADEM.
- Given the patient's current neurocognitive status, this case illustrates the importance of integrating comprehensive neuropsychological services in the recovery period of an intense attack that involves the CNS. This allows for the establishment of a baseline for longitudinal comparative purposes, in addition to helping better parse out precise areas of neurocognitive change following the ADEM attack, and to track later cognitive trajectory.

Contact

Mohammadi Khan, M.A.
UChicago Medicine, Adult Neuropsychology Clinic; The Chicago School of Professional Psychology
mkhan13@bsd.uchicago.edu
mkhan1@ego.thechicagoschool.edu

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