Neuropsychological Profile of Early Onset ADEM and AMSAN

Palmeri, M.J.[1,2], Weiss, E.F.[1], McGinley, J.[1], Bronshteyn, D.[1], & Masur, D.[1], Facchini, R.[1]

[1]Montefiore Medical Center, Department of Neurology, Division of Neuropsychology

[2]Ferkauf Graduate School of Psychology, Yeshiva University

Background

- **Acute Disseminated Encephalomyelitis (ADEM)** is an inflammatory autoimmune disease of the CNS of abrupt onset resulting in multifocal demyelinating lesions of the gray and white matter. Sudden and rapidly worsening neurological symptoms can be observed (e.g., seizures, diminished sensation, weakness of extremities, tingling sensations, etc.).
- **Acute Motor Sensory Axonal Neuropathy (AMSAN)** is a rare variant of Guillain-Barré syndrome causing muscle/sensation difficulty.
- **A nine-year-old girl, repeating 3rd grade (+IEP; 12:1:1 class), presented for neuropsychological evaluation with a complicated medical history.**

Brief History

- Right-handed, nine-year old girl.
- Born full term; no complications. Met all milestones on time or early up to 18 months.
- At 18 months, developed ataxia with worsening gait and falls. Found to have demyelinating lesions in the left frontal lobe and upper cervical spine with cervical cord signal changes.
- History included feeding problems/dysphagia, delays in speech with oromotor problems, and notable personality changes (i.e., agitation). Symptoms waxed and waned, improving with IVIG treatment.
- Initially thought to be Transverse Myelitis in the setting of ADEM. Subsequent EEG supported a diagnosis of AMSAN.
- She received formal schooling through 1st grade in a rehabilitation hospital setting.
- Presently receives weekly occupational, physical and speech therapies.
- Struggles with gross/fine motor coordination skills and speech.

Observations

- Significant limitations to her attention regulation capacity and her ability to sustain effort; conducted testing over 4 sessions to accommodate.
- Slightly awkward gait; difficulty with fine motor skills.
- Somewhat immature in communication content appearing much younger than her age
- Poor volume control.

WISC-V	SS/ss
Verbal Comprehension	98
Similarities	10
Vocabulary	9
Visual Spatial	89
Block Design	6
Visual Puzzles	10
Fluid Reasoning	85
Matrix Reasoning	7
Figure Weights	8
Working Memory	91
Digit Span	9
Picture Span	8
Processing Speed	80
Symbol Search	8
Coding	5
Full Scale IQ	83
General Ability Index	80
Cognitive Proficiency	82

WJ-IV	SS
Letter-Word Identification	80
Applied Problems	**72**
Spelling	81
Passage Comprehension	**63**
Calculation	**63**
Writing Samples	**66**
Oral Reading	**68**
Sentence Reading Fluency	82
Math Facts Fluency	**69**
Sentence Writing Fluency	**69**
Word Attack	**48**
Academic Fluency	73
Academic Applications	**63**
Broad Achievement	**68**

NEPSY-II	ss
Finger Tapping Sequence Combined	**4**
Speeded Naming Correct Total	**<1**
Inhibition – Naming/Inhibition	**3**
Oromotor Sequences	**<1**
Word Generation – Initial Letter	5

WRAML-2	SS
Verbal Memory	108
Visual Memory	82
Attention/ Concentration	100
General Memory	95

CPT-3
Significant Elevations indicating impulsivity, inattention, and weak sustained focus

BASC-3
At Risk in: Attention Problems and aspects of Adaptive Functioning

Results

- Neuropsychological evaluation revealed overall cognitive function to be in the low average range.
  - Variable processing speed.
- **Below age and grade level performances in all major academic areas** with varying capacity within each domain.
- Extensive **attention regulation difficulty** requiring continual prompting and redirection.
- **Significant oromotor and gross motor coordination difficulties.**
  - Problems with articulation although language functions were generally intact.
- Learning and memory were intact.
- **Mild self-esteem issues** and immature social relatedness.

Conclusions

- **Findings revealed significant areas that require intervention.**
- Evidenced **global academic delay, secondary to neurologic injury.**
- Behavior during testing, test performances, supported by parent and teacher interview, indicate the presence of **ADHD** that interfere with her performances.

- ***This case demonstrates the importance of neuropsychological evaluation in pediatric paraneoplastic motor/sensory motor syndromes.***

DIAGNOSES:

- **Attention-Deficit Hyperactivity Disorder, combined type presentation** (DSM-5: 314.01)

References

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