Neuropsychological Sequelae of Opsoclonus-Myoclonus Syndrome: A Case Series
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Conclusions
In general, OMS results in reduced global intellect, problems with speech and language, and behavioral dysregulation. Both cases presented with reduced intellectual function, largely consistent with published data on OMS. Fine motor performance was dependent upon treatment response, especially in Case 2. Behavior dysregulation was present in Case 2, but varied depending on treatment regimen (e.g., dexamethasone). Cognitive prognosis is heavily dependent upon management of OMS-related symptoms. Given the rarity of OMS, greater awareness of the disorder and its clinical manifestations is needed.

References

Introduction
Opsoclonus-Myoclonus Syndrome (OMS) is a rare movement disorder which typically presents in infancy. It has been described as a paraneoplastic disorder and can be a precursor to neuroblastoma, although some cases are chronic and never progress to a malignant state. Clinical features of the disorder include erratic eye movements and generalized myoclonus. Corticosteroid treatment along with immunosuppressive and immunomodulatory therapies are standard. Research regarding cognitive functioning in OMS is limited but has shown that lower intellectual performance, speech impairments, and behavioral problems are most commonly observed. Unfortunately, cognitive outcome is generally poor, despite early treatment.

Case 1
Clinical History
A 3-year-old Caucasian boy diagnosed with OMS at 23 months of age. Presenting symptoms included leg pain and bilateral leg tremor. Motor development regressed and speech articulation was delayed. Also present were autosomal recessive polycystic kidney disease, which was asymptomatic, and Caroli Syndrome, which presented as bleeding esophageal varices requiring two transfusions. Remission was achieved after five months of treatment. He was asymptomatic at the time of neuropsychological evaluation.

OMS Treatment
Cytoxan x 6 months
Monthly IVIG infusions (ongoing at time of evaluation)
ACTH
Daily Aldactone & Bactrim

Neurocognitive Test Results
DAS-II
Verbal Reasoning=80
Nonverbal reasoning=81
General Conceptual Ability=79
Bracken Basic Concepts Scale-3-R:
School Readiness Composite=12 (Age Equiv 3:10)
ABAS-II & BASC-2 WNL

Speech articulation and visual-motor integration were weaknesses. No emotional or behavioral problems were present.

Case 2
Clinical History
A 6-year-old Caucasian boy diagnosed with OMS at 13 months of age. Presenting symptoms coincided with 12-month vaccines and included regression in motor skills and oculomotor abnormalities. Multiple relapses occurred despite treatment and a neurologic relapse was ongoing at Time 1.

OMS Treatment
Rituximab x 3 courses
Low-dose IV cyclophosphamide
ACTH x 2 years
Monthly IVIG infusions (ongoing at Time 1 and Time 2)
Dexamethasone pulses (ongoing at Time 2)
Daily Tenex (for sleep)

Neurocognitive Test Results
Time 1: Attention, executive functioning, and motor dexterity were impaired. Mild weaknesses were observed in verbal memory, expressive vocabulary, and visuospatial skills. Difficulties with behavioral regulation and speech articulation were also noted.

Time 2: At re-evaluation four months later (after dexamethasone treatment), improvements in verbal memory and motor dexterity were observed.

References