ALABAMA ACADEMY OF NEUROLOGY

AAN 2022 Poster Presentation

Abstract 22-02

Title: Case of Anti-NMDA Receptor Encephalitis Presenting in a Toddler with Hemorrhagic Cavernomas

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Introduction/Background: Anti-N-methyl-D-aspartate (anti-NMDA) receptor encephalitis signifies an autoimmune antibody-mediated neuropsychiatric disease that often presents with a set of well-described clinical characteristics and other times manifests with more rare features. The heterogeneity of patient presentation can propose a diagnostic challenge to even the best clinical neurologist. Anti-NMDA receptor encephalitis should be considered for patients who possess an alternative existing diagnosis that shows atypical progression because early recognition and treatment of the disease can help reduce long-term complications.

Description: We illustrate a 14 month-old previously healthy boy with anti-NMDA receptor encephalitis who first presented with focal seizures. Initial neurologic imaging revealed intracranial hemorrhage with underlying cavernous malformations. He responded well to a single anti-seizure agent, but re-presented one week later with transient weakness that was ultimately attributed to worsening intracranial hemorrhage with surrounding edema. Upon his third presentation, he developed dyskinesias, sleep dysfunction, autonomic instability, cognitive changes, and motor regression, prompting further work-up with lumbar puncture. Cerebrospinal fluid analysis showed a positive NMDA antibody titer of 1:40. Treatment with intravenous steroids, plasma exchange (PLEX), and intravenous immune globulin (IVIg), followed by infusions of Rituximab and Cyclophosphamide resulted in gradual, marked clinical improvement.

Discussion and Conclusion: This case study and literature review explores the relationship between cavernous malformations, intracranial hemorrhage, and anti-NMDA receptor encephalitis, and how these diagnoses respond to escalating immunomodulation therapies. The temporal relationship of two rare co-existing pathologies suggests a pathophysiologic mechanism between the two, and this relationship is compared to that of herpes simplex encephalitis and subsequent anti-NMDA encephalitis. Consideration of this entity should be made when the neurologic examination does not follow an expected course of a previously established diagnosis. With timely recognition and aggressive treatment approaches, patients can achieve substantial clinical improvement.

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