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Clinical Observations

Anti–*N*-Methyl-D-Aspartate Receptor Encephalitis and Rasmussen-like Syndrome: An Association?

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ABSTRACT

BACKGROUND: *N*-methyl-D-aspartate (NMDA) receptor encephalitis is an immune-mediated condition that has a broad spectrum of manifestations, including seizures, coma, psychosis, and focal neurological deficits. Although usually a diffuse process, unihemispheric involvement mimicking early stages of Rasmussen encephalitis can occur. Rasmussen's encephalitis is a unique syndrome characterized by progressive hemiplegia, drug-resistant focal epilepsy, cognitive decline, and hemispheric brain atrophy contralateral to the hemiplegia. **PATIENT DESCRIPTION:** We describe a two-year-old girl with progressive right weakness and epilepsia partialis continua, concerning for early Rasmussen's encephalitis, who tested positive for anti-NMDA receptor antibodies. She experienced complete clinical recovery after immunotherapy. Anti-NMDA receptor antibodies were absent at three weeks and again at one year after the first treatment of intravenous immunoglobulin. **CONCLUSIONS:** There are few reports of Rasmussen-like encephalitis in individuals with anti-NMDA receptor antibody positivity. Thus the clinical significance of this association is yet to be determined. In addition, several other antibodies have been documented in individuals with Rasmussen encephalitis. The lack of a consistently reported antibody in Rasmussen encephalitis patients and the temporary nature of the anti-NMDA receptor antibody in our patient raise the following question: Is the presence of anti-NMDA receptor antibodies the cause of the symptoms or secondary to the pathogenic process?

Keywords: Rasmussen syndrome, NMDA receptor, AMPA receptor, glutamate, encephalitis, pediatrics, autoimmune

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Patient Description

This previously normal two-year-old girl developed subacute onset of progressive right hemiparesis, speech regression, and right body focal seizures described as frequent right arm or leg twitching. Electroencephalography (EEG) showed continuous delta rhythm in the left hemisphere (Fig 1). Numerous right-sided twitching episodes were captured on EEG without any clear ictal change. Seizures remained refractory to oxcarbazepine and levetiracetam. Her contrast-enhanced brain magnetic resonance imaging (MRI) was normal. The possibility of stage I Rasmussen encephalitis was suggested by her clinical presentation, and a positron emission tomography (PET) study was performed, which demonstrated diffuse left hemispheric hypermetabolism (Fig 2).

Given the clinical picture and PET findings, the child was readmitted to the hospital for further autoimmune evaluation and immune therapy. Cerebrospinal fluid (CSF) was submitted for the following studies: chemistry, glutamate receptor 3 (GluR3) antibody, *N*-methyl-D-aspartate receptor (NMDAR) antibody, cytology, flow cytometry, oligoclonal bands, myelin basic protein, Immunoglobulin G index, electrophoresis, and a viral panel. In addition, serum NMDAR antibody was submitted. Once these studies were submitted, intravenous immunoglobulin (IVIG) was started; dosing was 2 g/kg divided over five days.

Unfortunately, there was not enough CSF to perform the GluR3 and NMDA studies. The other CSF studies were normal. The serum NMDAR antibody study was positive, however. The serum NMDAR antibody titer value was not available. Abdominal and pelvic ultrasound examinations did not reveal an ovarian teratoma.

When she was seen for follow-up three weeks later, her speech had improved and she was using her right side more. She still had occasional twitching of the right side. Physical examination was notable for increased tone of the right upper extremity, a mild right pronator drift, and decreased speed of rapid alternating movements on the right. Serum anti-NMDA receptor antibody was resent and MRI was repeated. The

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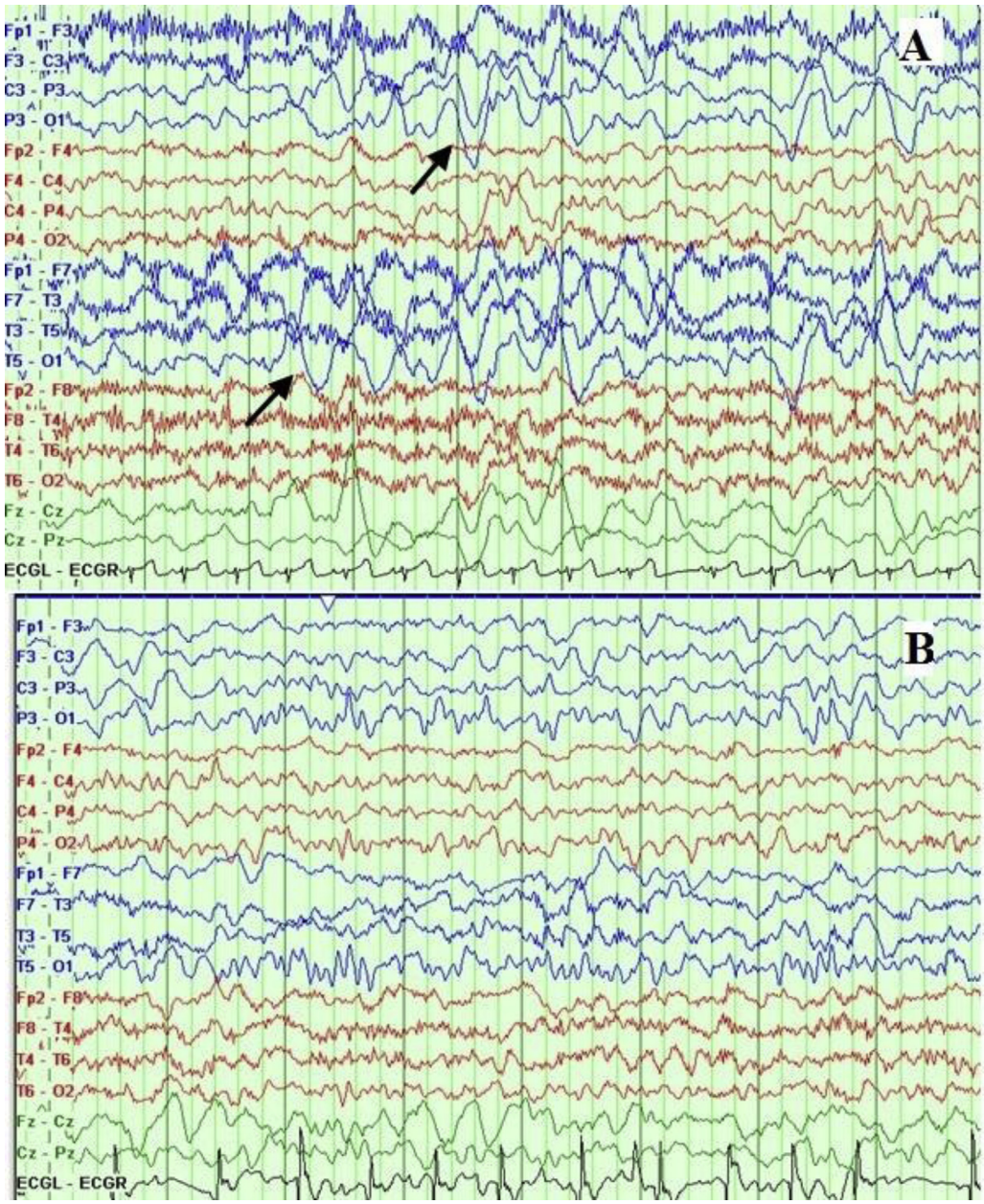

FIGURE 1.
(A) Initial EEG at the time of presentation showing left-sided slowing (black arrows). (B) EEG after six months of treatment with intravenous immunoglobulin documents resolution of the slowing. (The color version of this figure is available in the online edition.)

repeat NMDA receptor antibody was negative. Brain MRI showed no atrophy but had areas of increased cortical thickness within the left frontal, parietal, and temporal lobes on fluid-attenuated inversion recovery (FLAIR) sequence (Fig 3). Repeat EEG continued to show left slowing. The decision was made to continue IVIG 2 g/kg every four to five weeks for one year.

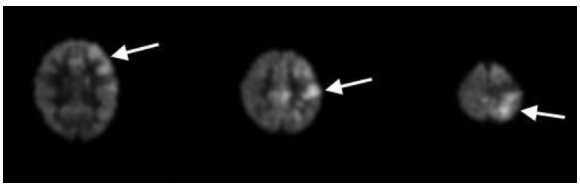

FIGURE 2.
Positron emission tomography—documents increased fludeoxyglucose uptake (white arrows) involving the left temporal and frontal lobes.

Her speech rapidly improved and was normal by three months after the initial treatment. Her weakness improved more slowly than her speech, but it too was normal seven to eight months after initial treatment. Body twitching episodes had resolved four months after the initial treatment. Her parents self-discontinued antiepileptic drugs eight months after the initial treatment. Her EEG had normalized ten months after the initial IVIG treatment. A serum anti-NMDA receptor antibody was sent for a third time 11 months after treatment and it was unremarkable. Pelvic and abdominal ultrasound at 12 months was normal. In total she received six doses of IVIG. Now more than three years after her initial IVIG treatment, she remains asymptomatic with normal developmental milestones in accordance with the Denver II Developmental screening checklist.

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