

Refractory status epilepticus from NMDA receptor encephalitis successfully treated with an adjunctive ketogenic diet

Sir,
N-methyl D-aspartate receptor encephalitis (NMDARE) is characterized by the presence of NMDAR antibodies in the

CSF, and it can progress to psychosis and refractory status epilepticus (RSE). We report a case of the use of a ketogenic diet (KD) as an adjunct in RSE from NMDARE.

A healthy 21-year-old female presented to the ED with symptoms that progressed from depression, headaches, and nausea to word-finding difficulties, psychosis, and seizures. She developed SE and was intubated. A comprehensive evaluation was unremarkable, and she was transferred to our institution on hospital day 11 where an electro-encephalogram (EEG) revealed RSE. The NMDAR antibody was detected in her CSF. A thorough investigation was negative for any tumor. Despite corticosteroids, plasmapheresis, IV Ig in addition to 5 anticonvulsant medications and propofol, continuous EEG showed RSE. Ketosis was achieved after a KD (ratio of fat: Combined carbohydrate and protein of 4:1) was started (day 21). Her continuous EEG showed a significant decrease in her seizures within 2 weeks (day 35). She was weaned off mechanical ventilation and discharged to a long-term care facility on 3 anticonvulsant medications, mycophenolate mofetil, and on a KD.

Most patients with NMDARE develop a multistage illness that progresses to psychosis, memory deficits, RSE, and into a state of unresponsiveness with catatonic features.^[1] Management of NMDARE should initially focus on immunotherapy and the detection and removal of the tumor.^[2,3] Most patients receive corticosteroids, PLEX, or IV Ig as first-line immunotherapy. Additional treatment with second-line cyclophosphamide or rituximab is sometimes needed. Our patient continued to deteriorate despite these therapies. A KD, by reducing the firing of central neurons by opening K ATP channels^[4], is a very effective adjunct to antiepileptics in patients with refractory epilepsy. Side effects of KD^[5] are usually mild, predictable, and preventable, and rarely lead to diet discontinuation. Important ones include constipation, acidosis, dyslipidemia, kidney stones, and bone fractures.

To our knowledge, this is the first case report of initiating a KD as an adjunct in RSE from NMDARE.

Syed Amer, Parth Shah¹, Vishnu Kommineni¹

Department of Neurology, ¹Medicine, Mayo Clinic, Phoenix, Arizona, USA

For correspondence:

Dr. Syed Amer, Mayo Clinic, 5777 East Mayo Boulevard, Phoenix, Arizona - 85054, USA

E-mail: drsyedamer1@gmail.com

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