Clinical commentary

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Treatment responsive GABA(B)-receptor limbic encephalitis presenting as new-onset super-refractory status epilepticus (NORSE) in a deployed U.S. soldier

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Epileptic Disorders

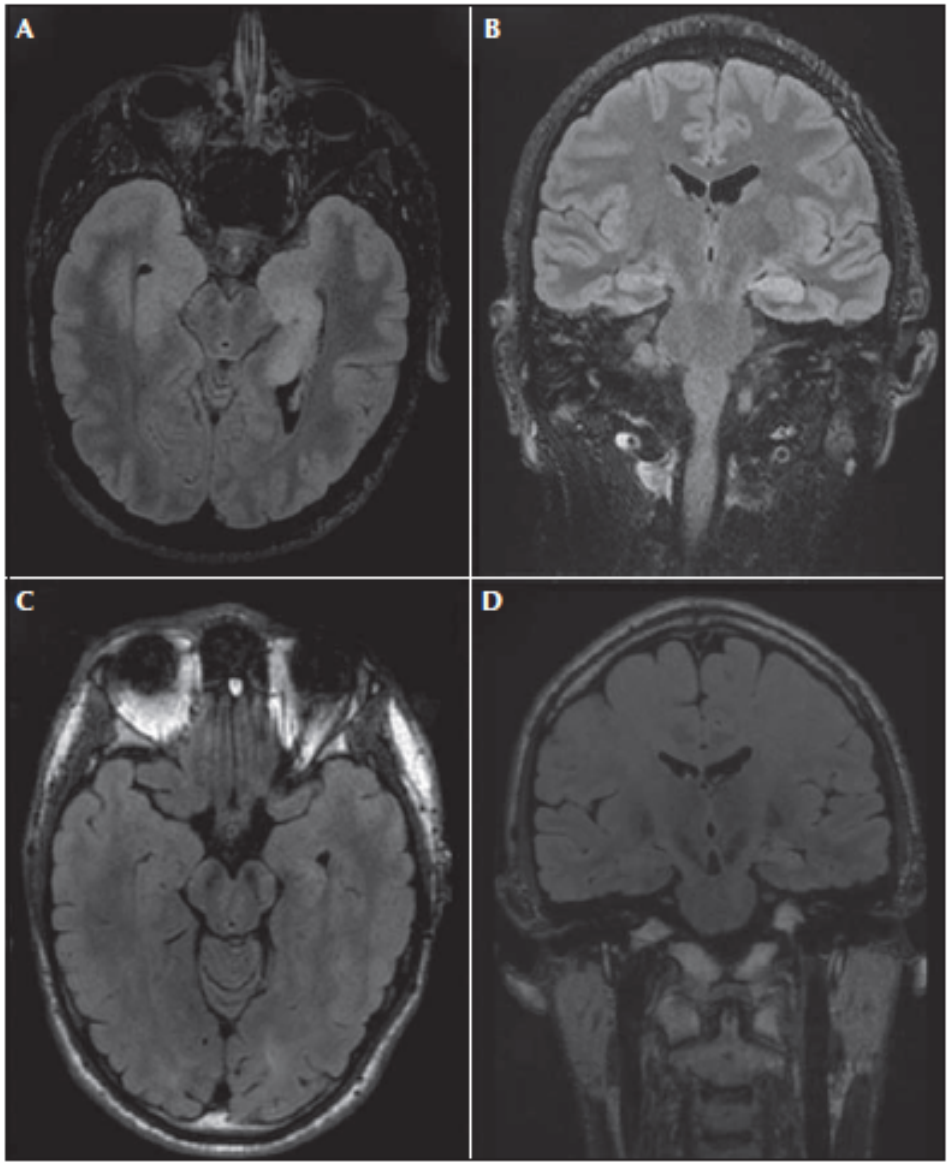

Figure 2. MRI series demonstrating limbic encephalitis and recovery seven months later, using T2-FLAIR brain MRI.
(A) Day 15 after presentation, axial-view. (B) Day 15 after presentation, coronal view. (C) Month 7 after presentation, axial-view. (D) Month 7 after presentation, coronal view.
(A, B) Asymmetric enlargement and T2 hyperintensity of the left more than the right hippocampus which may represent limbic encephalitis *vs.* postictal oedema. (C, D) demonstrate resolution of previous T2 hyperintense T2 signal seven months later.

GABA(B)-Receptor Encephalitis Presenting as NORSE

- Super-refractory status epilepticus (SE) is a stage of refractory SE characterized by SE continuing or recurring 24 hours or more after onset of anesthesia, including withdrawal/reduction of anesthesia.
- It is encountered typically in two distinct clinical situations:

  1) In patients with severe acute brain injury

  2) In patients with no history of epilepsy in whom SE develops *de novo*: new-onset refractory status epilepticus (NORSE)

- NORSE is a clinical syndrome that often carries a grave prognosis and in which a treatable etiology is often never discovered.

GABA(B)-Receptor Encephalitis Presenting as NORSE

- In patients presenting with a new refractory seizure state and limbic encephalitis, an autoimmune encephalitis should be strongly considered in the differential diagnosis.
  - This includes auto-antibodies targeting the GABA(B) receptor.
- Patients with GABA(B)-receptor limbic encephalitis can present with limbic encephalitis, seizures with or without status epilepticus (**including NORSE**), ataxia, opsoclonus-myoclonus, and Morvan syndrome.

GABA(B)-Receptor Encephalitis Presenting as NORSE

- Patients with GABA(B)-receptor limbic encephalitis should be treated aggressively to arrest seizure activity:
  - Multiple anti-epileptic drug therapy
  - Intravenous anesthesia with goal of burst suppression pattern on EEG; if necessary, to include a long-term pharmacologically-induced coma
  - Upfront immune suppression, multiple modalities if necessary
- Patients with GABA(B)-receptor limbic encephalitis should undergo rigorous cancer surveillance
  - About 50% of cases are associated with small cell lung cancer.