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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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.
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.
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.