Relapsed Autoimmune Encephalitis Complicated by Seizure: A Case Report

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Background

- Immune mediated encephalitis includes both paraneoplastic encephalitis and autoimmune encephalitis.
- The most studied autoimmune encephalitis, anti-NMDA receptor encephalitis (NMDA-R encephalitis), was first characterized by Dr. Josep Dalmau in 2007.
- Incidence is rare, affecting 1.5 people per 1 million yearly.
- Typically presents in children and adults younger than 45. 37% of cases occur in patients 18 years old and younger.
- Female to male ratio of approximately 8:2.
- 80% of patients improve with immunotherapy; recovery may take as long as 18 months.
- Relapse may occur in 12-24% of cases.

Case Description

- 58 year old male with a history of NMDA-R (N-methyl-D-aspartate receptor) Encephalitis presents with acute onset altered mental status, aphasia, right sided weakness, and seizure.
- Stroke was deemed unlikely, as physical exam was more suggestive of encephalitis versus complex partial status epilepticus affecting the right upper extremity and face.

Case (continued)

- Infectious work up was negative, and MRI did not show progression of prior area of encephalitis.
- EEG showed lateral periodic discharges; Keppra dose increased.
- CT of chest, abdomen, and pelvis did not show malignancy, ruling out paraneoplastic encephalitis.
- Treatment for NMDA-R encephalitis with Solumedrol, plasma exchange, and Rituximab was initiated and followed by a course of inpatient rehabilitation.

Discussion

- The clinical course of autoimmune encephalitis, specifically NMDA receptor encephalitis, varies.
- A patient may present with headache or fever followed by psychiatric disturbances, insomnia, seizure, memory deficits, decreased level of consciousness, dyskinesias, autonomic instability, and language dysfunction.

Discussion (continued)

- Probable diagnosis of NMDA-R encephalitis is made using several criteria, including clinical and laboratory findings, along with the exclusion of other diagnoses.
- Definitive diagnosis additionally requires the presence of IgG anti-GluN1 antibodies.

Conclusion

- Fewer than 5% of NMDA-R encephalitis cases occur in patients who are 45 years and older.
- Unlike paraneoplastic encephalitis, autoimmune encephalitis often responds to treatment.
- Management focuses on the removal of offending antibodies and immunosuppression followed by treatment with Physical Therapy, Occupational Therapy, and Speech Language Pathology to address resultant deficits.

Images

Diagnostic criteria for NMDA-R Encephalitis

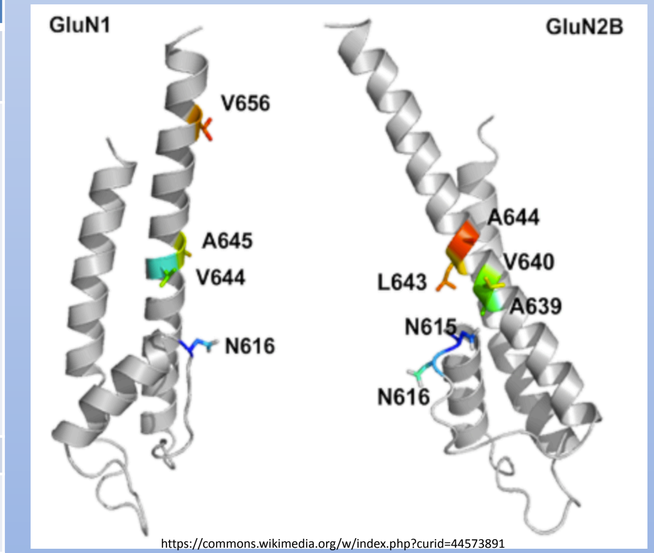
Probable Diagnosis of NMDA-R Encephalitis must meet all 3 criteria:

1. **Rapid onset (< 3 months) of at least four of the six symptom groups below:**
   - Abnormal behavior or cognitive dysfunction
   - Speech dysfunction
   - Seizures
   - Movement disorder, dyskinesias, or rigidity
   - Decreased level of consciousness
   - Autonomic dysfunction or central hypoventilation.
2. **At least one of the following lab findings:**
   - Abnormal EEG (focal or diffuse slow or disorganized activity, epileptic activity, or extreme delta brush)
   - CSF with pleocytosis or oligoclonal bands
3. **Reasonable exclusion of other disorders.**

Definite Diagnosis of NMDA-R Encephalitis

1. IgG anti-GluN1 antibodies in the presence of one or more of the six major symptom groups after reasonable exclusion of other disorders.

Schematic diagram of NMDA receptor



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