RADY 401 Case Presentation
Cheyenne Bricken, MS4
July 18, 2023
Focused patient history and workup

20yF with history of anti-NMDAR encephalitis on monthly IVIG, catatonia, and seizures who presented to UNC ED one month after uncomplicated SVD (at 38w1d) with symptoms of altered mental status and behavioral disturbance raising concern for relapse of anti-NMDAR encephalitis

• ED Vitals: BP 133/110 | Pulse 132 | Temp 36.4 C | RR 24 | SpO2 100%

• Mental Status and Neuro Exam Key Findings: Alert and oriented only to name and DOB with difficulty following commands. Recent and remote memory appeared impaired. Cranial nerves intact. Sensory and motor exam normal.

• Initial labs unremarkable
List of imaging studies

Neuroimaging during previous admission 6mo ago (when patient was found to have Anti-NMDAR encephalitis):

• CT head without contrast (unremarkable)
• MRI head without contrast

Neuroimaging this admission:

• cEEG overnight with diffuse slowing (nonspecific)
• MRI head without contrast [admission]
• MRI head with and without contrast [infectious workup recommended by ID due to onset of autonomic instability with fever]
• MRI head with and without contrast [new upper motor neuron symptoms: +Babinski, clonus]
MRI brain from previous admission

Non-contrast axial T2 weighted:

Non-contrast axial T2 - FLAIR:

Water signal is suppressed → dark CSF

Abnormal findings:
Scattered hyperintense foci in white matter

*T2 – FLAIR most sensitive sequence for white matter hyperintensity*

*T1 and diffusion sequences unremarkable for this patient*
Non contrast axial T2 – FLAIR: midventricular level

At time of admission:

No acute abnormalities (foci of hyperintensity still present)

Hospital day 9 (after fever onset):

Significant increased area of white matter hyperintensity

Hospital day 46 (after concern for new UMN symptoms):

Improvement in white matter hyperintensity with some baseline still present

T1 and diffusion sequences remain unremarkable
No abnormal enhancement with contrast
Non contrast axial T2 – FLAIR

Hospital Day 9

White matter hyperintensities:
Can be associated with infectious damage, traumatic injury, autoimmune disorders, ischemia, migraines, cognitive changes... NON-SPECIFIC
Non contrast axial T2 - FLAIR

*Hospital day 9 at level of pons and midbrain*

Source: e-Anatomy Atlas²
Patient treatment and outcome

49 day hospitalization

• Fluctuating behavioral symptoms and autonomic instability

• Treatment included:
  • IV and oral steroids, PLEX, IVIg, Rituximab for anti-NMDAR encephalitis
  • Empiric acyclovir for HSV encephalitis
  • Ativan, Haldol, Risperdal for catatonia and agitation

• MRI did not change patient management

• Determined to be anti-NMDAR encephalitis relapse (original leading differential diagnosis)

• At time of discharge, condition improved with resolution of catatonic and UMN symptoms along with improvement in dysautonomia
Anti-NMDAR Encephalitis

• Considered a paraneoplastic autoimmune neurological disorder
  • Ovarian teratomas most associated

• Symptoms include psychiatric changes (AMS, catatonia, bizarre behavior, delusions etc.), seizures, autonomic instability

• Diagnosis confirmed by presence of NMDAR antibodies in CSF

• MRI findings nonspecific and not diagnostic (normal in up to 50%)
  • Most common MRI abnormality: T2-FLAIR hyperintensities

• Most patients respond to immunotherapy

• Psychiatric symptoms may be only symptom with relapse
Discussion: Imaging Appropriateness

**Variant 1:**
Acute mental status change. Increased risk for intracranial bleeding (ie, anticoagulant use, coagulopathy), hypertensive emergency, or clinical suspicion for intracranial infection mass, or elevated intracranial pressure. Initial imaging.

<table>
<thead>
<tr>
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<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT head without IV contrast</td>
<td>Usually Appropriate</td>
<td>♦ ♦ ♦</td>
</tr>
<tr>
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<td>O</td>
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<td>MRI head without and with IV contrast</td>
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</tr>
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<td>May Be Appropriate</td>
<td>♦ ♦ ♦</td>
</tr>
<tr>
<td>CT head with IV contrast</td>
<td>Usually Not Appropriate</td>
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**Variant 2:**
Acute or progressively worsening mental status change in patient with a known intracranial process (mass, recent hemorrhage, recent infarct, central nervous system infection, etc). Initial imaging.

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**Source:** American College of Radiology

Reasons patient received imaging:
1. Acute onset of AMS in patient undergoing treatment for known anti-NMDAR encephalitis
2. Infectious workup for infectious encephalopathy when patient spikes fever in setting of AMS
3. New UMN symptoms
## MRI Cost:

### MRI head without contrast

<table>
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<th>Facility Name</th>
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<th>Ancillary Service</th>
<th>Charge</th>
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<tr>
<td>UNC Hospital</td>
<td>611</td>
<td>MRT</td>
<td>70551</td>
<td>$3,535.00</td>
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### MRI head with and without contrast

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**Total: $13,095**

### MRI Radiation: None

Source: UNC Health
UNC Top Three: Anti-NMDAR Encephalopathy

• Negative imaging does not rule out anti-NMDAR encephalopathy
• Patients must be monitored closely for changes in behavior, as this often indicates a relapse necessitating treatment
• Remember association with ovarian teratomas
References

7. UNC Health Standard Charges & Shoppable Services Data by Hospital. 2023 Jan. Retrieved from: https://rca.centaurihs.com/ptapp/#d4ccc071fab9c79f17e52d5b243ef668affc5e569afa907c5b481f0a89284