Sporadic Creutzfeldt-Jakob Disease Presenting as Non-convulsive Status Epilepticus: A Case Report

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Objective

To describe a rare presentation of prion disease in a patient with new onset non-convulsive status epilepticus.

Introduction

Sporadic Creutzfeldt-Jakob disease is a rare fatal neurodegenerative disease. Symptoms include rapidly progressive dementia, myoclonus, ataxia, and visual disturbances. Seizures are uncommon[1]. To date, there has only been one case series and a few case reports[2-4]. This report demonstrates the importance of evaluating for prion disease in patients with no previous seizure history presenting with non-convulsive status epilepticus.

Medical History

- 71 yo female with diabetes, hypertension, hypothyroidism without cognitive or behavioral deficits in early 2014
- She had never traveled to Europe. No personal history of seizures. No family history of seizures or dementia
- Late 2014: Appetite declined, significant weight loss
- Jan 2015: She was aggressive with her neighbors and was arrested. She started to repeat herself often. No difficulty with naming, orientation, memory. Diagnosed with Alzheimer's dementia after having 6 months of impulsivity, aggression and perseveration.
- June 2015: Unable to do finances, spoke less often, irritable
- June 3, 2015: Family found her unresponsive with urinary incontinence. A stroke alert was activated.

Initial Exam

Vital Signs: T 97.8, HR 82, RR 18, BP 182/83, 98% O2 on RA
General: Critically ill appearing. Dehydrated.
Mental Exam: Mute, not following commands. GCS: E4V1M1
Cranial Nerves: II - No blink to threat. III,IV,VI - Right gaze preference. XI - Right head deviation.
Strength/Sensation: Flaccid paralysis. No motor response to painful stimuli axially or in the extremities.
Reflexes: 1+ of the left biceps. No clonus. Babinski sign negative
Involuntary movements: Two beats of bilateral blinking, one beat of left foot twitch.

Evaluation

Serum:
- VZV PCR, HSV 1/2 PCR, immunofixation electrophoresis, serum electrophoresis and blood cultures were negative

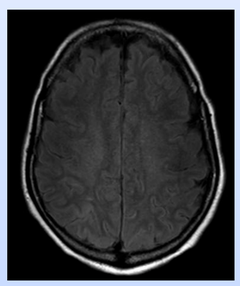
Cerebrospinal fluid:
- *Opening pressure*: 10 cm of water
- *Basic studies*: Cell count, protein, glucose, and gram stain were unrevealing
- *Further studies*: Negative for herpes, varicella and an extensive paraneoplastic panel
- *CSF 14-3-3 protein*: positive
- *CSF T-tau protein:* 1751 pg/ml (more than 1150 carries a 76% probability of prion disease)

Urine:
- Urine culture negative

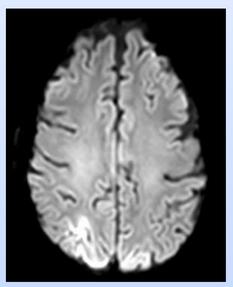
CT head: Without apparent pathology

EEG: multiple electrographic seizures during recording session

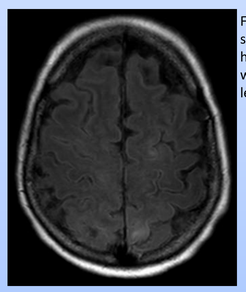
MR Imaging



FLAIR: subcortical hyperintensity with no mass lesion.



DWI: Bilateral medial parietal lobe lateral cortical restricted diffusion



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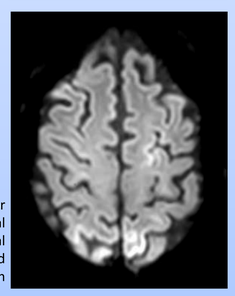

DWI: Left posterior superior frontal gyrus lateral cortical restricted diffusion

MRI brain: Bilateral medial parietal lobe and left posterior superior frontal gyrus lateral cortical restricted diffusion and adjacent T2 FLAIR subcortical hyperintensity

Management

- 0.1 mg/kg Ativan twice, bolused with levetiracetam and valproic acid without resolution of seizures
- Versed drip initiated and titrated upward to suppress electrographic seizures. Focal sharp waves continued.
- Levetiracetam, valproic acid and lacosamide were scheduled. EEG continued to show focal sharp waves.
- Versed weaned off on day 4
- Patient returned to baseline by day 7 on above scheduled antiepileptic drugs
- Repeat EEG showed periodic lateralized epileptiform discharges on day 9
- Patient was extubated, and after family discussions, hospice was consulted

Diagnosis and Discharge

Non-Convulsive Status Epilepticus (NCSE) and New Onset Refractory Status Epileptics (NORSE) criteria were met.[5&6] WHO criteria was met for probable sporadic Creutzfeldt-Jakob disease[5]. Per patient's request, she was discharged to hospice.

Conclusion

Non-convulsive status epilepticus is an extremely rare presentation of prion disease. Clinicians should consider prion disease as an etiology for NCSE and NORSE and evaluate for prions with the appropriate diagnostic tests. Unfortunately, there are no pathognomonic signs for diagnosing sCJD. Therefore, one must rely on clinical manifestations in combination with other diagnostic tests. We believe that the differential diagnosis needs to be carefully evaluated in order not to miss a possible reversible condition[7].

References

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