

## Introduction

- Cortical ribbon sign is associated with:
  - Most commonly prion disease
  - Also, infarction, infection, hypoxia, metabolic and electrolyte derangements,
- A patient presented in status epilepticus (SE) who demonstrated cortical ribbon sign on MRI and worked up for Creutzfeldt-Jakob disease (CJD).

## Cortical Ribbon Sign

- Hyperintense signal in cerebral cortex on diffusion weighted images of MRI
- Hypothesized to be due to severe rapid loss of neurons
- On serial imaging, cortical enhancement develops into T1 hyperintensities and may resolve after multiple months
- Part of WHO guidelines for CJD diagnosis but in the correct clinical context

## Patient Presentation

- 72-year-old male who was brought to the hospital after a witnessed seizure, lasting about 45 seconds and described as generalized shivering by onlookers.
- Medical history of seizure disorder secondary to a remote stroke, Crohn's disease and treated hepatitis C
- CT head was unremarkable other than for an old right parietal infarct.
- He was started on levetiracetam.

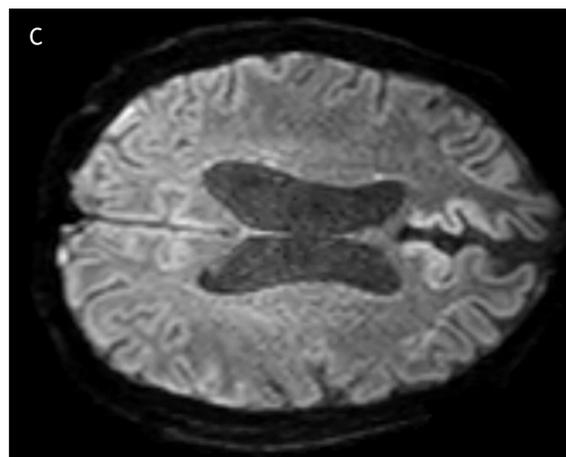
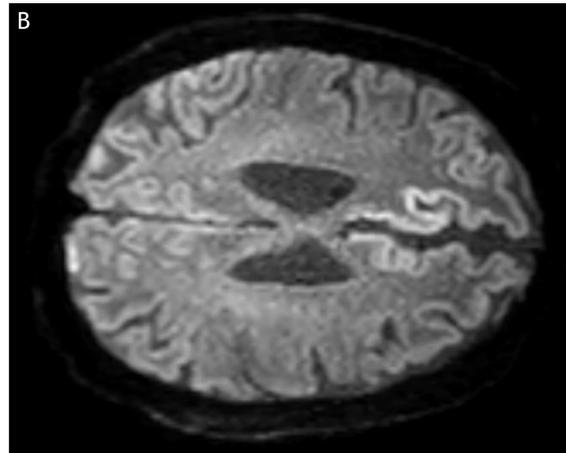
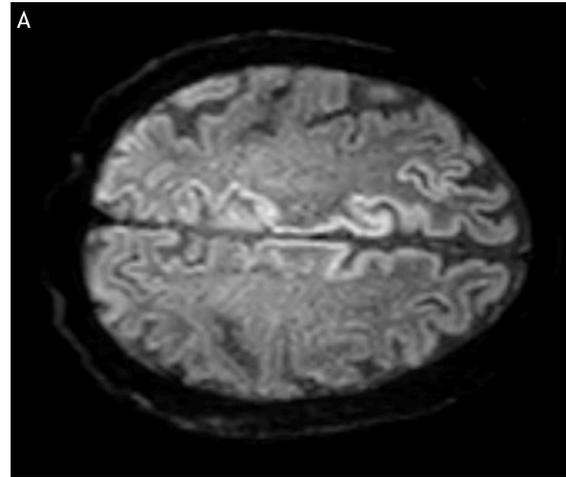


Figure 1: Diffusion weighted MRI after episode of status epilepticus

## Clinical Course

- During a routine EEG, patient had 3 further seizures with jerking movements predominantly of the upper extremities.
- A continuous EEG was started and on review of EEG, over the next 6 hours, patient was noted to have over 50 seizures.
- He was transferred to the neurological critical care unit for intubation and midazolam drip. He was also given lacosamide and additional levetiracetam. His continuous EEG showed no further seizures with some focal right sided posterior temporal slow activity.
- His MRI brain with and without contrast showed restricted diffusion in bilateral medial frontal and parietal lobes with a hyperintensity of cerebral cortical gyri of the right frontal, parietal, and occipital regions, known as the cortical ribbon sign.
- Lumbar puncture was performed to rule out infectious causes and specifically to test for protein 14-3-3 for confirmation of CJD.
- Given his poor prognosis, the family decided to palliatively extubate the patient without further workup and result from protein 14-3-3 pending, which eventually returned negative.

## Discussion

- Cortical ribbon sign is typically associated with prion disease.
- However, cortical ribbon sign is a nonspecific finding that can be seen in a wide variety of conditions that cause rapid neuronal injury, including status epilepticus, which is likely what happened in our patient.
- The patient's prognosis was grim given degree of cortical injury after several hours of difficult to control seizures
- However, suggestion of CJD as a cause was likely erroneous, especially since the patient lacked the main symptom of CJD, rapidly progressive dementia.
- The suggestion of CJD based on imaging may have contributed to the family's decision to pursue palliative extubation
- This case reinforces the importance of interpreting diagnostic testing within clinical context.