Introduction:
• This is a case presentation of a patient with a lethal disease who presented with vague neurological complaints and no initial physical exam findings, but displayed changes seen on diffusion weighted imaging (DWI) early on.

Case Summary:
• Mr. TS was a 54 year old male with past medical history of hepatitis C who presented to ALGH with memory loss and gait instability.
• Neurologic exam was completely intact.
• Initial MRI showed increased signals on DWI in the left and right parietal areas, more significant in the left. Figure 3
• Ten days later, physical exam was significant only for decreased sensation of the right lower extremity.
• EEG displayed slowing and a focal ictal event in the left frontotemporal/central region.
• Lumbar puncture (LP) was done to analyze CSF.
• Repeat MRI again showed increased signals on DWI in bilateral cerebral cortices. Figure 4
• A second LP was done and CSF sent for tau and 14-3-3 protein. Neither were identified.
• Hemiparesis, gait, and memory loss worsened.
• A second opinion was obtained at an outside hospital.
• One week later, he returned to ALGH where he acutely decompensated, became nonverbal with complete right hemiparesis, and passed away.

Results:
• Brain biopsy results reported by the National Prion Disease Pathology Surveillance Center established the diagnosis of Creutzfeld-Jakob disease (CJD).
• Autopsy tissue immunostained with 3F4, the monoclonal antibody to the prion protein, revealed granular deposits as seen in prion disease and consistent with CJD.

Conclusion:
• DWI has gained attention for earlier detection of CJD compared to T2, FLAIR, proton density, and T1 images. 3, 4
• DWI is more sensitive (83-92%) than T2 and FLAIR, especially for cortical changes. 1
• Even before findings are seen on neurological exam, EEG, or CSF analysis, DWI shows changes via increased signals. 2
• Diffusion-weighted MRI should be used as a first line diagnostic measure for patients with neurological complaints and/or exam findings to allow for a more timely diagnosis.

Figure 1: Vacuoles that create the spongiform change in CJD
Figure 2: Prion protein immunoperoxidase preparation revealing a diffuse and granular synaptic distribution pattern
Figure 3: Figure A shows a T2 MRI with normal signal intensity. Figure B is a proton density MRI with mildly increased cortical signal intensity within the right frontal lobe. Figure C is a DWI MRI that reveals more involvement in the right frontal lobe and more extensive hyperintensities in the bilateral occipital lobes. Figure D is also DWI that shows extensive cortical involvement posteriorly.

References: