

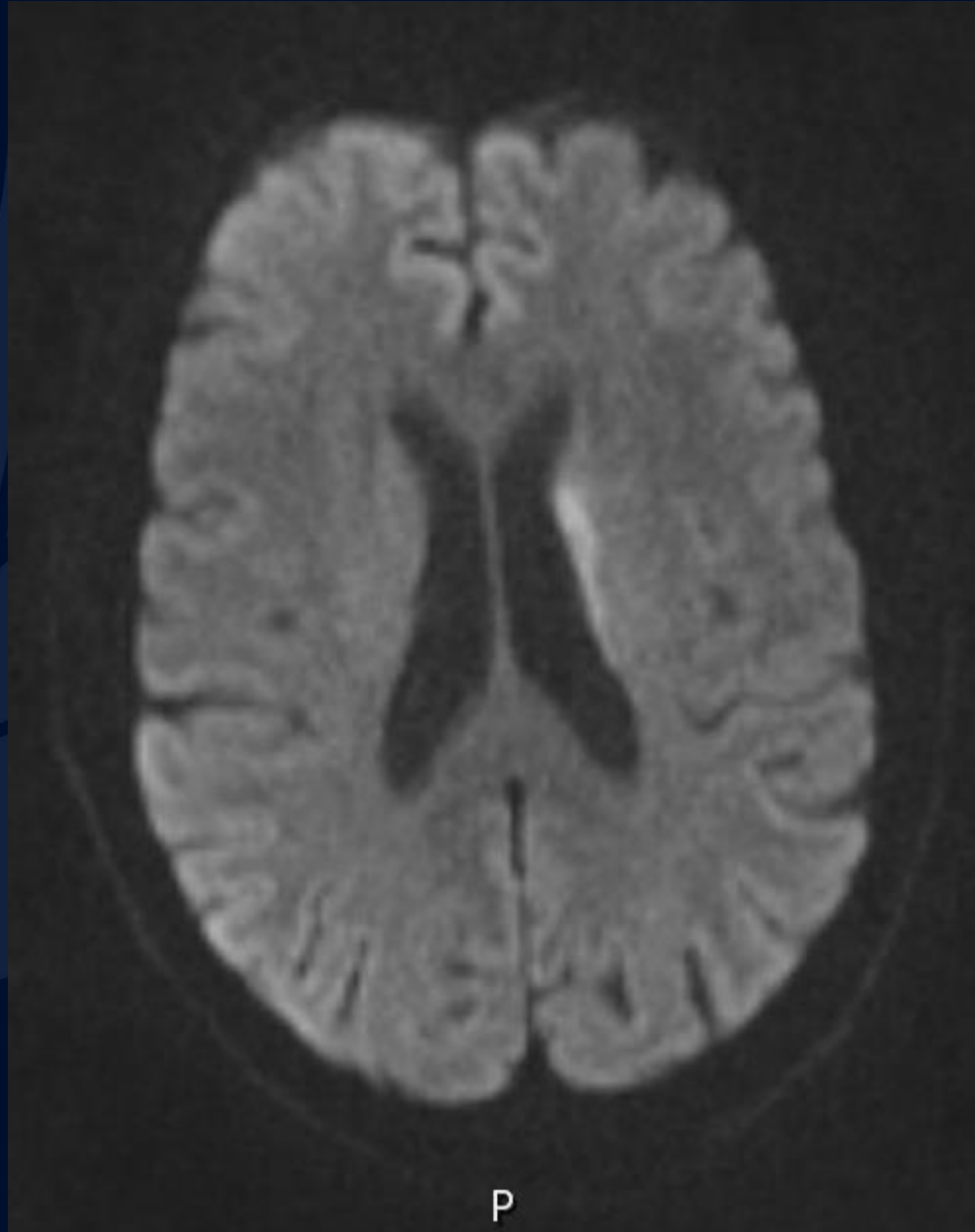
64-year-old female with rapidly progressive gait instability and cognitive decline for 8 weeks

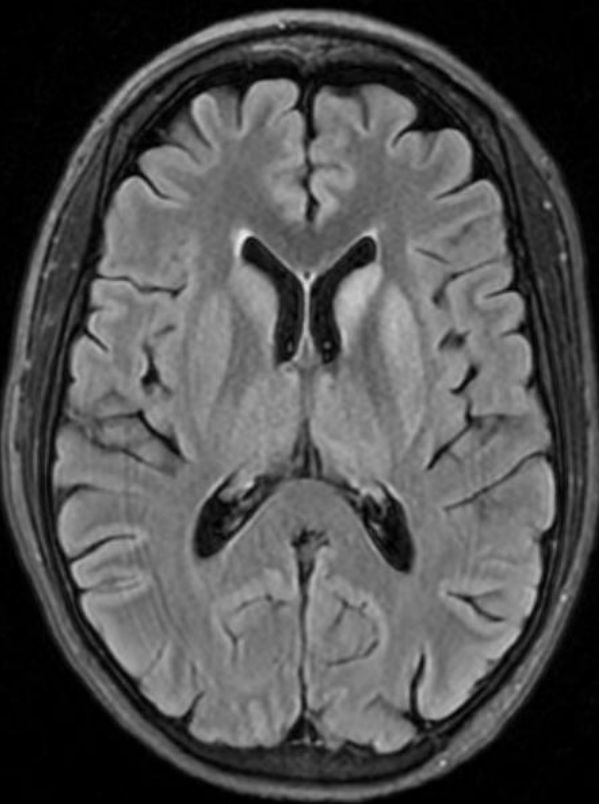
Maleek A. Masood, M3

CTA

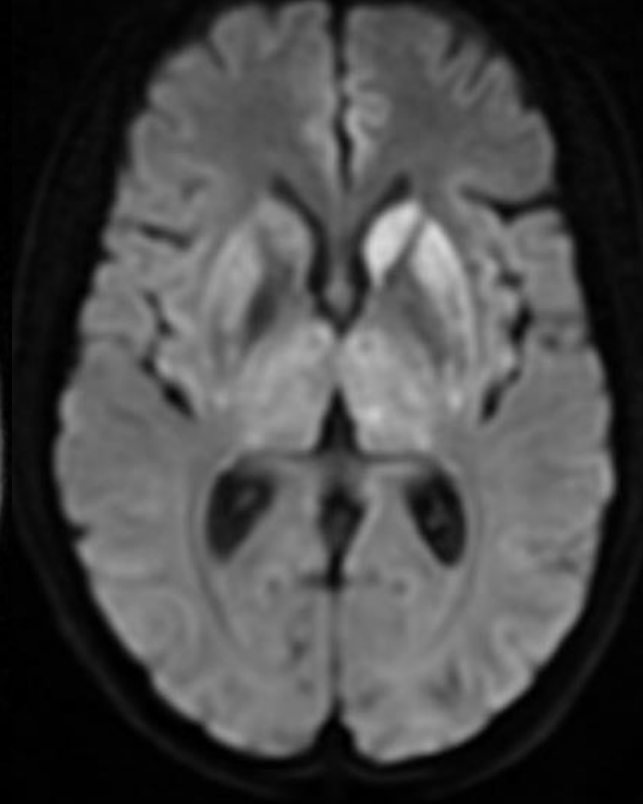


DWI

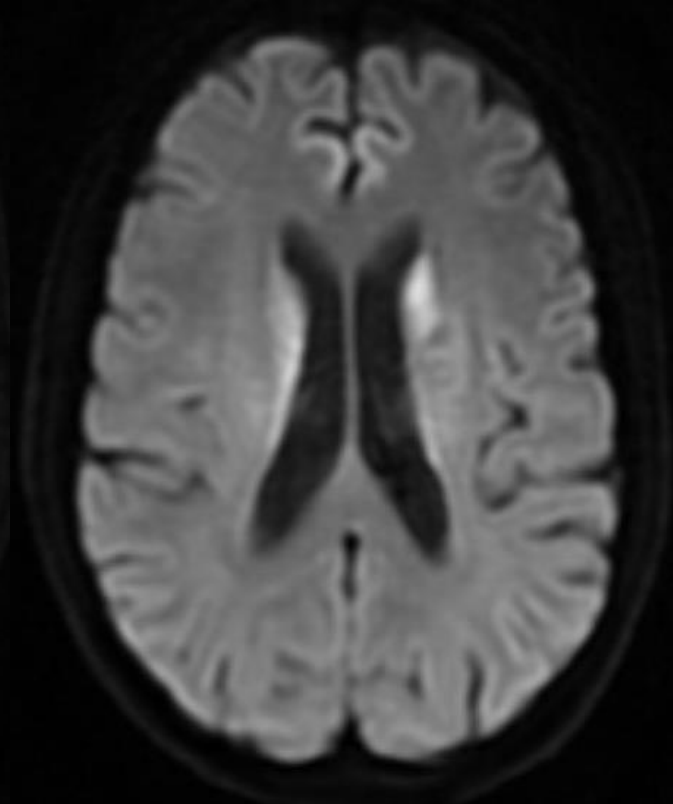




T2 Flair



DWI





?

Cruetzfeldt Jakob Disease

CTA

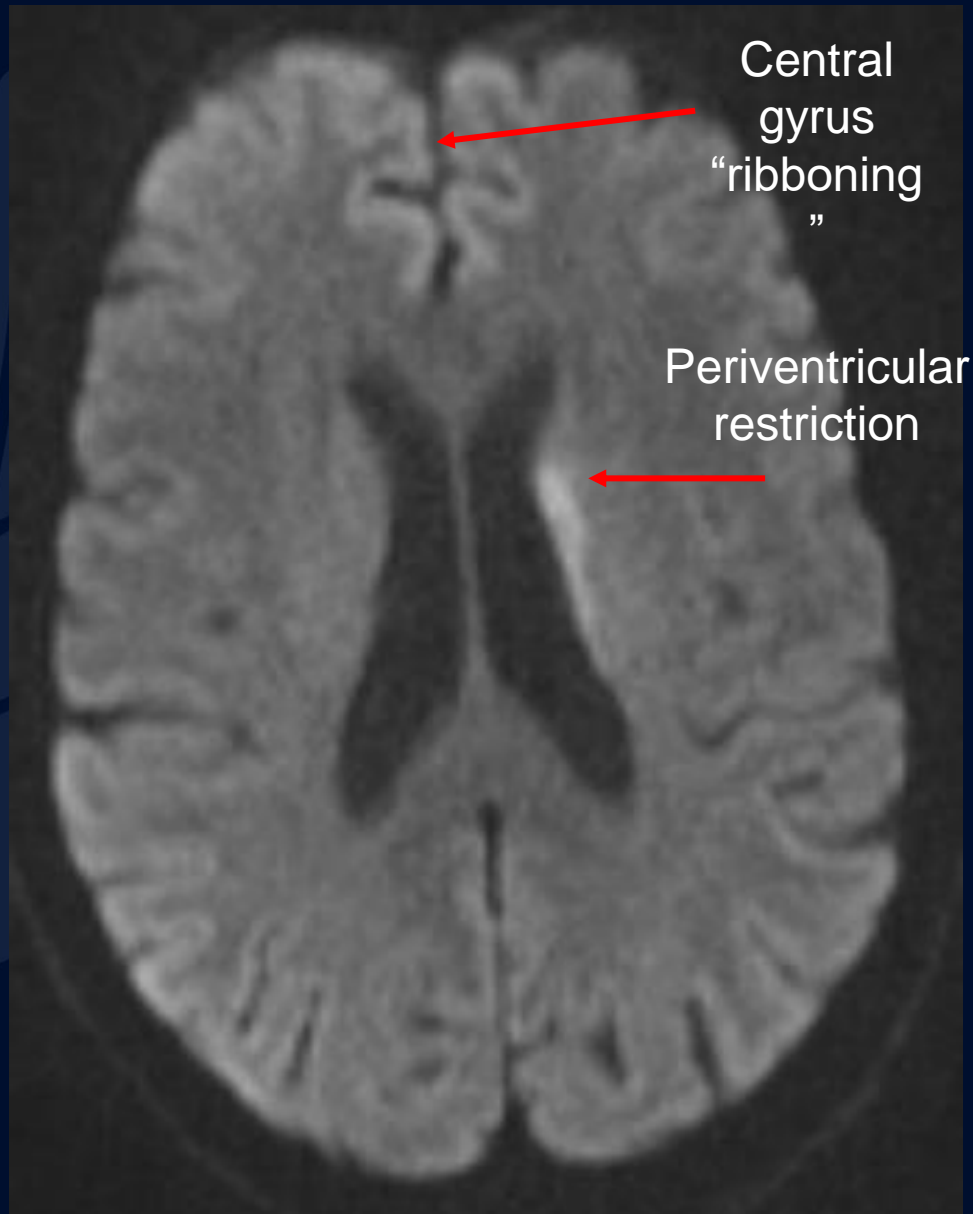
No large vessel
occlusion

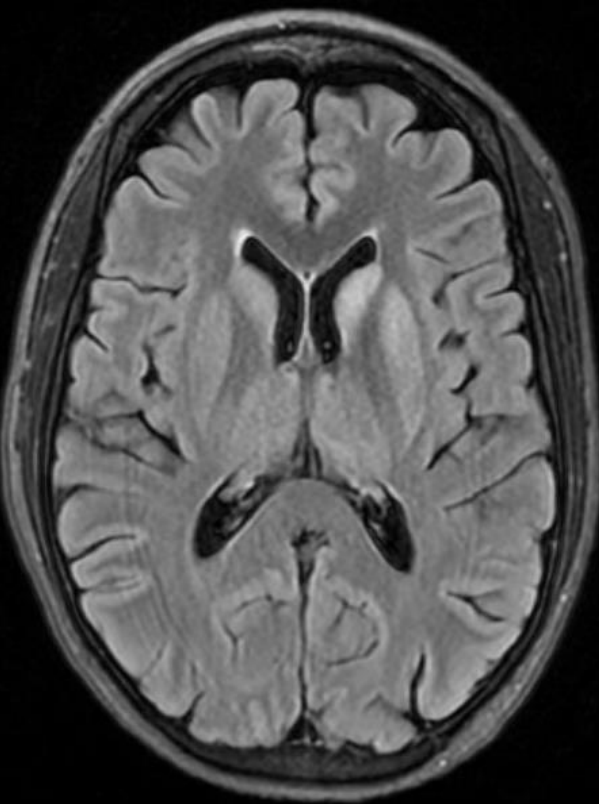
No critical stenosis

No vertebral artery
dissection

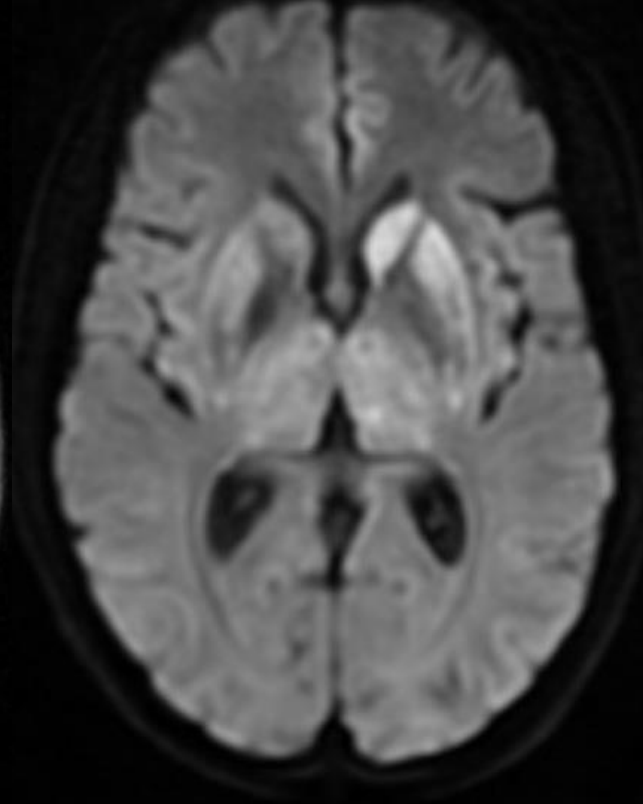


DWI

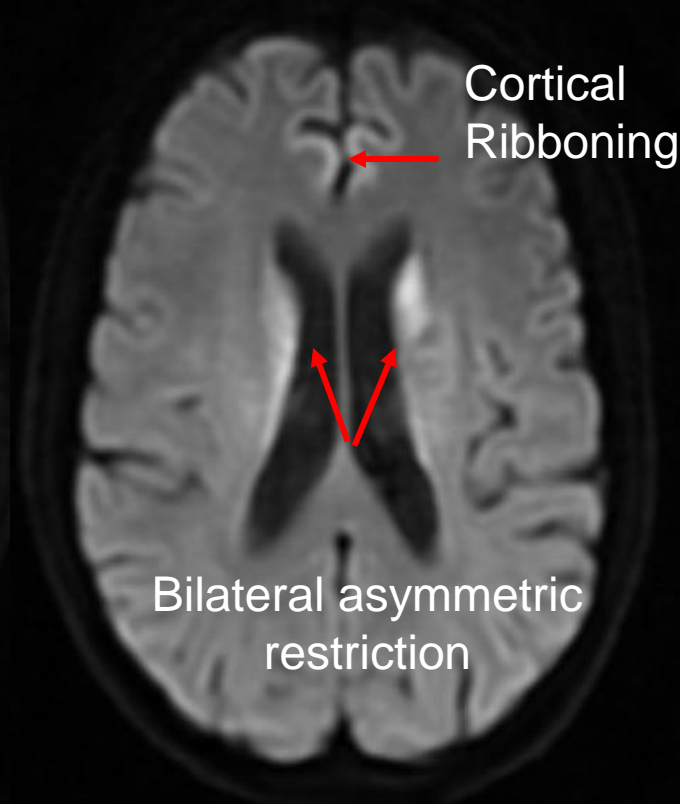




T2 Flair



DWI



Cortical
Ribboning

Bilateral asymmetric
restriction

Abnormal T2 FLAIR hyperintensity and restricted diffusion in a symmetric configuration involving the basal ganglia, thalami, external capsules, hippocampi, and insular regions.

Creutzfeldt-Jakob Disease (CJD)

- **CJD:** Defined as rapidly progressive, invariably fatal neurodegenerative disorder believed to be caused by an abnormal isoform of a cellular glycoprotein known as the prion protein.
- Sporadic CJD most commonly affects the cerebral cortex and deep grey matter
 - Most common:
 - Insula
 - Cingulate gyrus
 - Superior frontal gyrus
- Treatment is supportive (life expectancy ~ 8 mo. within Sx onset):
 - Antidepressants for anxiety and depression
 - Analgesics for pain
 - Sodium valproate can aid with tremors

Imaging Findings

Usual

- Focal or diffuse, symmetric or asymmetric involvement of cerebral cortex, basal ganglia (corpus striatum), or both.
- Sparing of the perirolandic cortex
- Cerebellar atrophy

Unusual

- Involvement of the perirolandic cortex
- “Pulvinar” and “double hockey stick” signs
- DWI hyperintensity involving the cerebellum

References

- Muir, R., & Sundaram, A. (2019). Cortical Ribbon Sign in Acute Hepatic Encephalopathy with Sequential Atrophy. *Canadian Journal of Neurological Sciences*, 46(6), 758-759. doi:10.1017/cjn.2019.285
- <https://memory.ucsf.edu/dementia/rapidly-progressive-dementias/cjd-differential-diagnosis>
- <https://www.mountsinai.org/health-library/diseases-conditions/creutzfeldt-jakob-disease>
- Ho M, Jones J, Ranchod A, et al. Creutzfeldt-Jakob disease. Reference article, Radiopaedia.org (Accessed on 11 Apr 2023) <https://radiopaedia.org/articles/7269>