

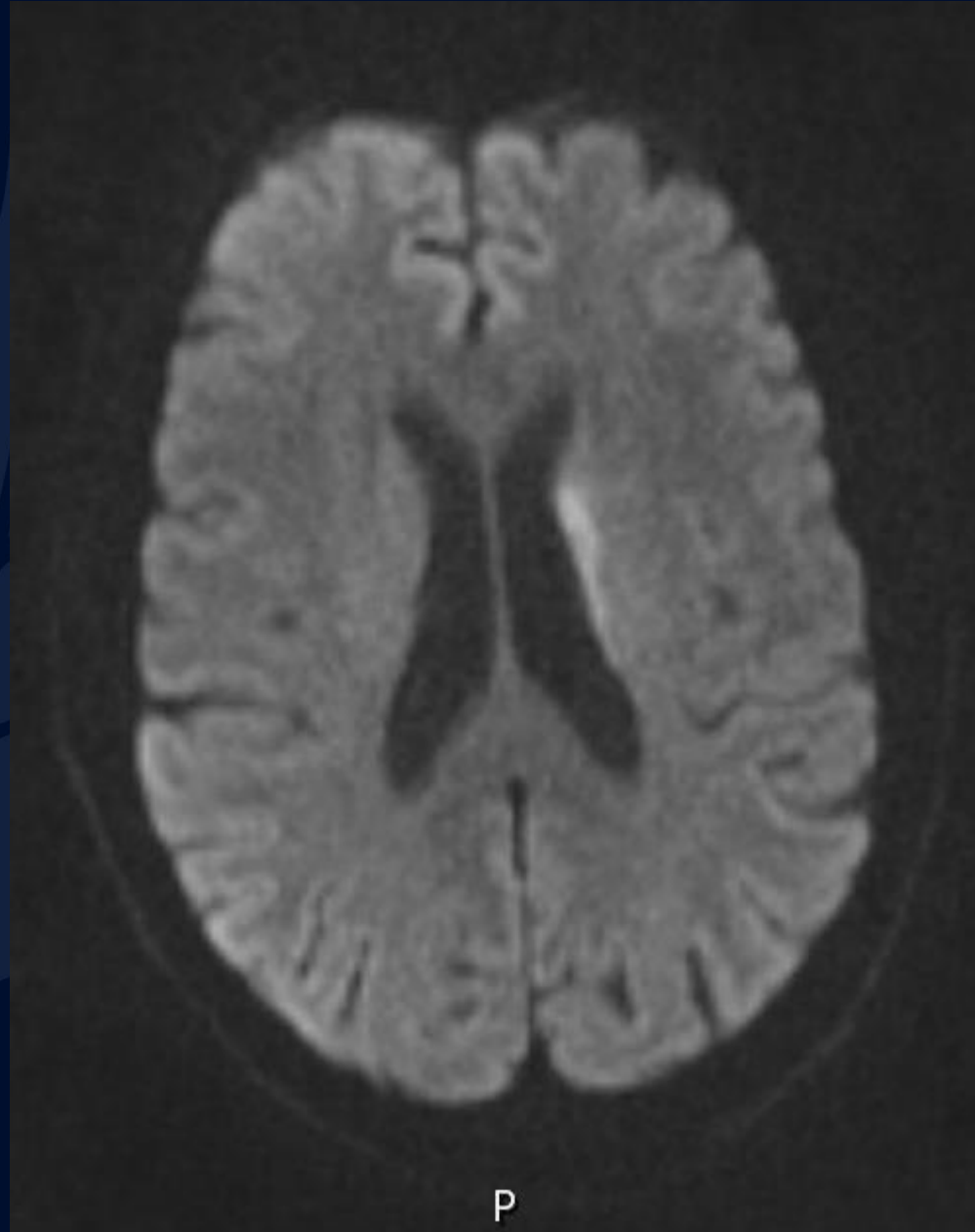
64-year-old female with rapidly progressive gait instability and cognitive decline for 8 weeks

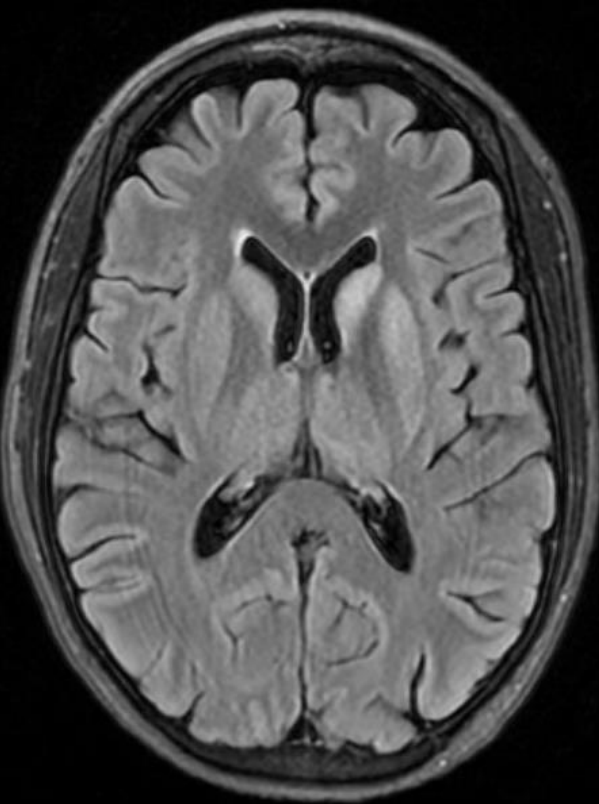
Maleek A. Masood, M3

CTA

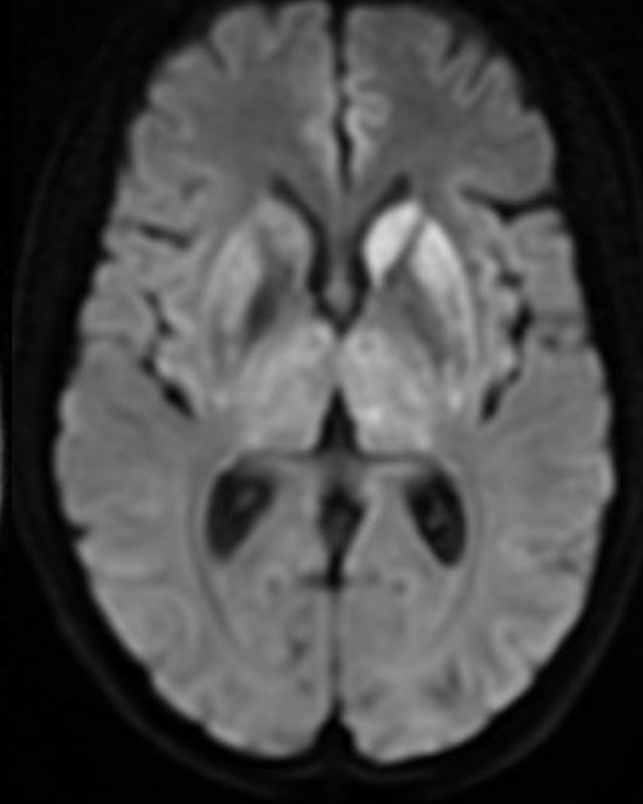


DWI





T2 Flair



DWI

A large, stylized oak leaf graphic in a dark blue color, positioned on the left side of the slide. It features detailed vein patterns and a lobed edge.

?

Cruetzfeldt Jakob Disease

CTA

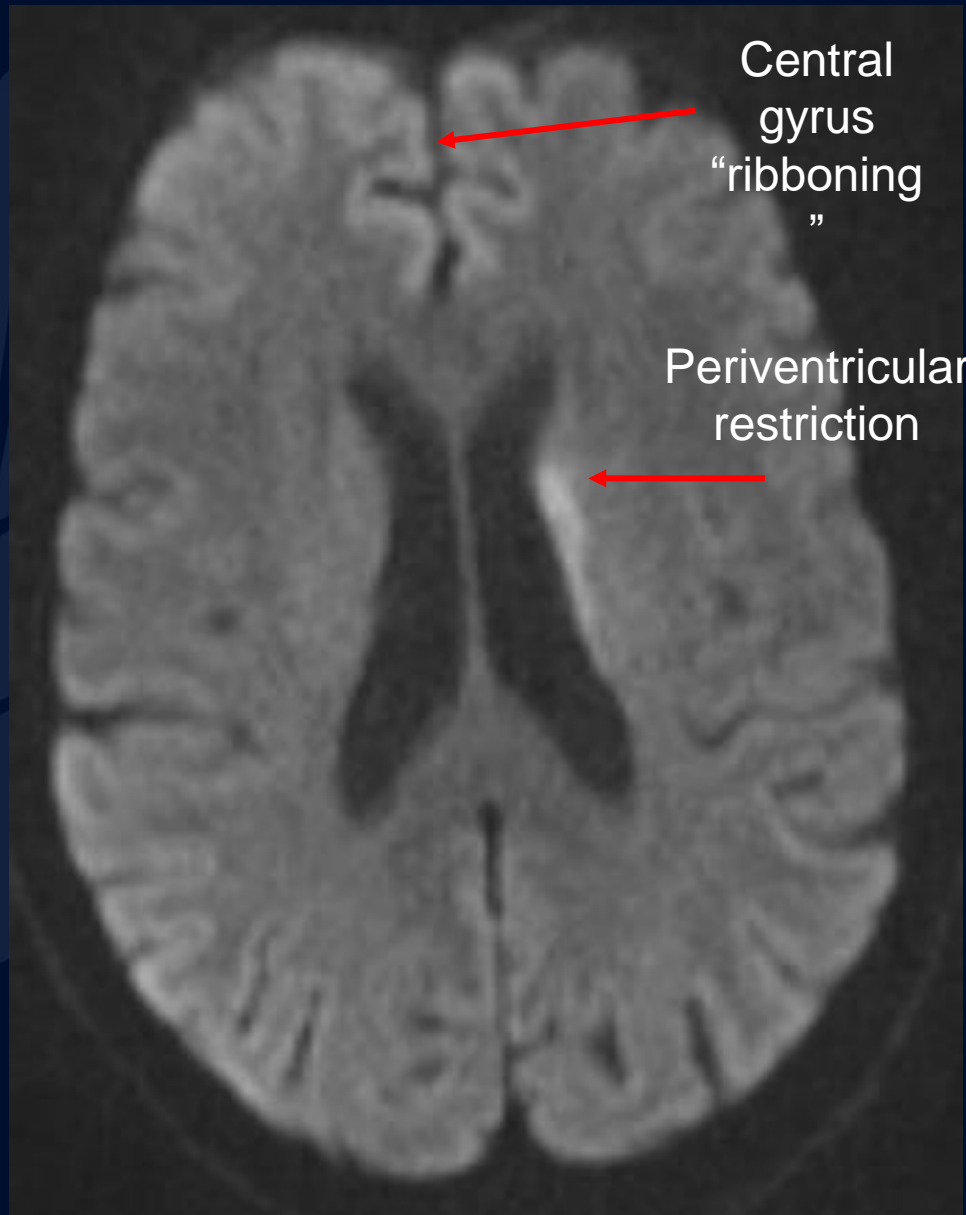
No large vessel
occlusion

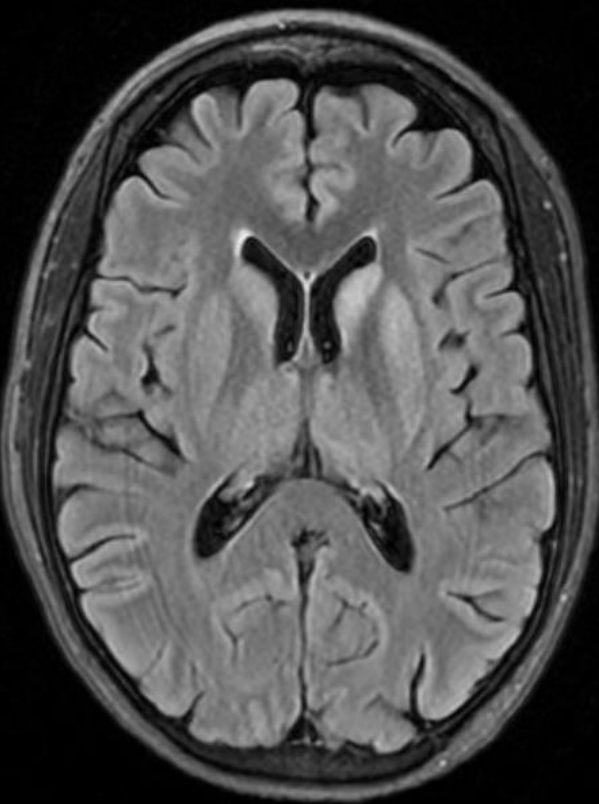
No critical stenosis

No vertebral artery
dissection



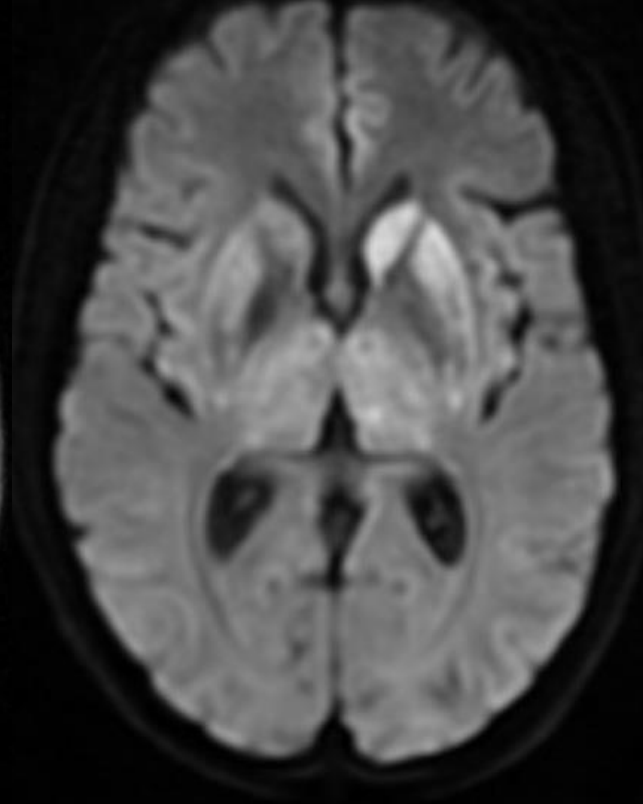
DWI



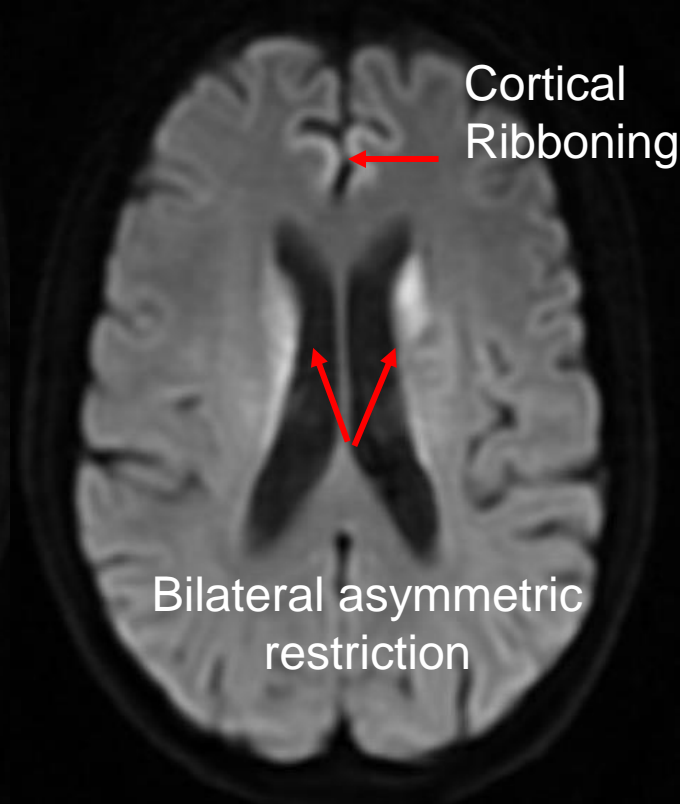


T2 Flair

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



DWI



Cortical
Ribboning

Bilateral asymmetric
restriction

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>