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Imaging of White Matter Matters

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Year IV

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Agenda

- Our Patient: Presentation
- Normal White Matter (WM) Features & Imaging Characteristics
- Framework for Evaluation of WM Disease
- Diagnoses by Distribution
- Our Patient: Suspected Diagnosis
- Summary



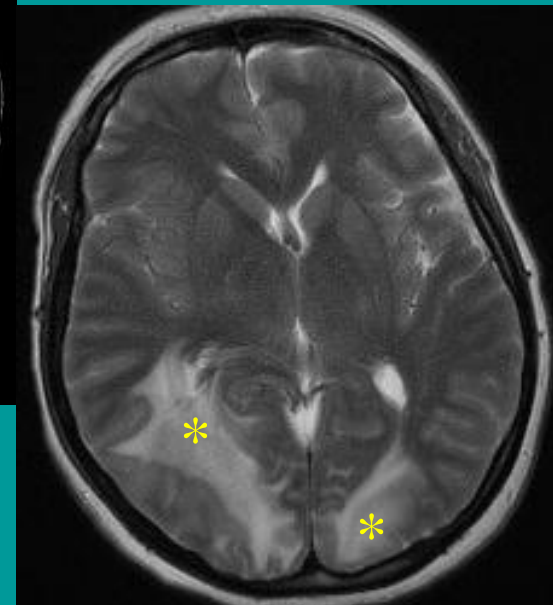
Our Patient: Presentation & Selected Imaging

- 60 year old female with hypertension to 160/100, headache & altered mental status.
- **Head non-contrast computed tomography (C-):** Nonspecific hypodensity in right parietooccipital region
- **Head Magnetic Resonance Imaging (MRI):** T2-hyperintensity in bilateral parietooccipital regions



Axial C- CT

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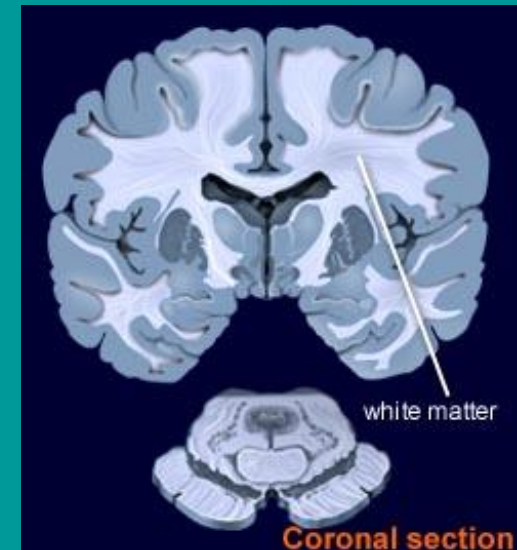
Axial T2 MRI

Let's continue to view the normal features of WM & a framework for evaluating WM disease.

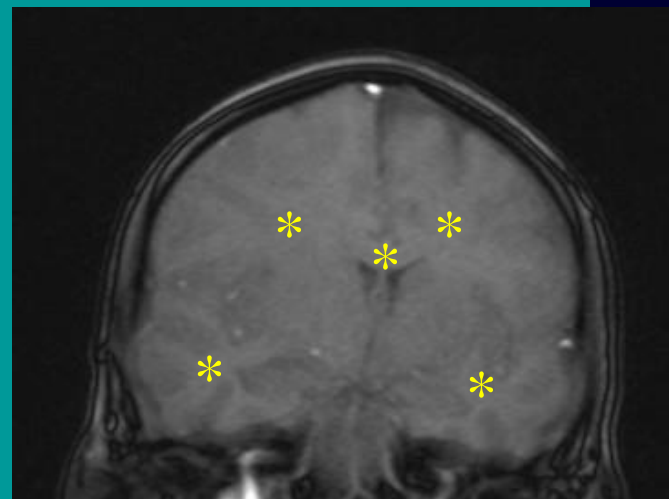


Companion Patient #1: Normal WM: Features & Imaging Characteristics

- In the brain, **WM is located centrally**
- WM contains **myelinated axons**
 - Composed of hydrophobic lipids
 - **High in fat & low in fluid** relative to gray matter (GM)
- **WM on CT**
 - hypodense to GM
- **WM on MRI**
 - **Hyperintense to GM on T1**
 - Hypointense to GM on T2



www.brainexplorer.org/glossary/white_matter.shtml



Coronal T1 MRI

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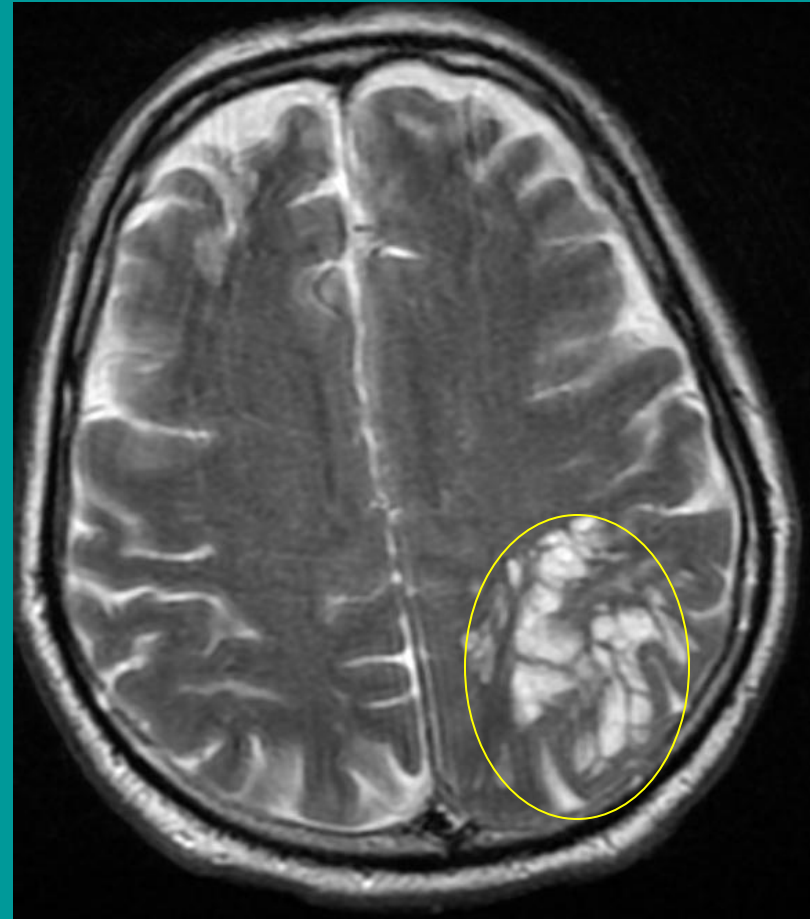
WM Disease: Framework for Narrowing the Differential Diagnosis

- **Clinical**
 - **Patient demographics**
 - Age
 - Gender
 - Patient history & physical exam
 - Laboratory results
- **Radiological**
 - **MRI** more sensitive than CT for evaluation of WM disease
 - **Distribution** throughout the brain on imaging
 - Multifocal
 - Confluent
 - Selective



Companion Patient #2: Perivascular spaces of Virchow-Robin: Normal Mimic of WM Disease

- Extensions of subarachnoid space surrounding small blood vessels
- Commonly visualized in
 - Brainstem
 - Basal ganglia
 - **Subcortical WM**
- Isointense to CSF on all pulse sequences
 - **Hyperintense to brain parenchyma on T2**
 - Signal nulls out with CSF signal on FLAIR
- Range from 1-2 mm to more than 1 cm in size



Let's continue to view examples of multifocal WM diseases.



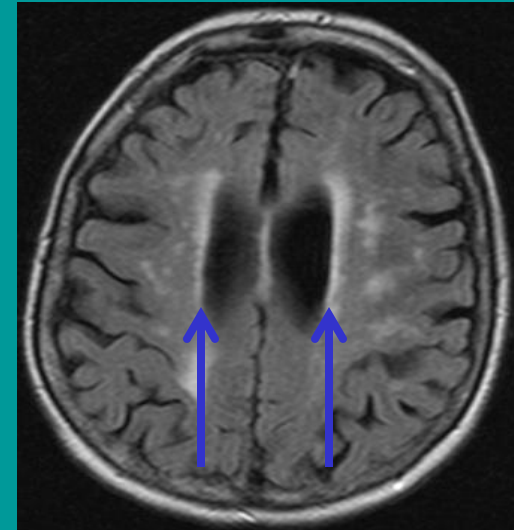
WM Lesion Distributions: Multifocal

- Multifocal examples
 - Small Vessel Ischemic Disease (SVID)
 - Multiple Sclerosis (MS)
 - Diffuse Axonal Injury (DAI)
 - Progressive Multifocal Leukoencephalopathy (PML)
- Confluent examples
 - Leukodystrophies in pediatric patient
 - Chemotherapy & radiation changes
- Selective examples
 - Central Pontine Myelinolysis (CPM)
 - Marchiafava-Bignami disease
 - Wallerian degeneration
 - Posterior Reversible Encephalopathy Syndrome (PRES)

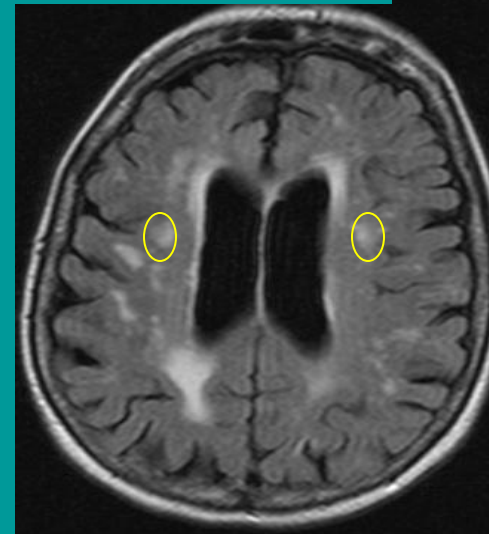


Companion Patient #3: SVID

- Risk factors similar to those of cardiovascular (CV) disease
- Locations
 - **Periventricular**: associated with age
 - **Deep**: associated with other CV risk factors
 - Subcortical WM: common in women
- Associations
 - Cognitive decline
 - Dementia
 - Gait disturbances
 - Stroke
 - Mortality



Axial FLAIR MRI

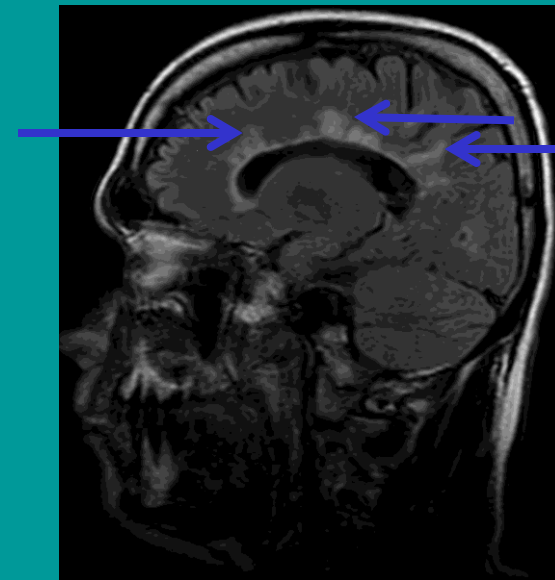


Axial FLAIR MRI

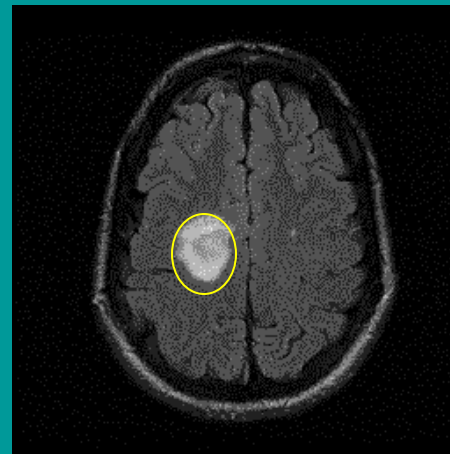


Companion Patient #4: MS

- Autoimmune demyelinating disease
- Most common in middle-aged females
- Sagittal FLAIR images useful to evaluate
 - lesions within the corpus callosum
 - Dawson's Fingers
- Often enhancement acutely
- Multiple subtypes/variants
 - i.e. Balo's concentric sclerosis
- Head MRI
 - 80 % sensitive & 90 % specific
- McDonald's criteria
 - 2 MRI lesions
 - 2 clinical episodes



Sagittal FLAIR MRI

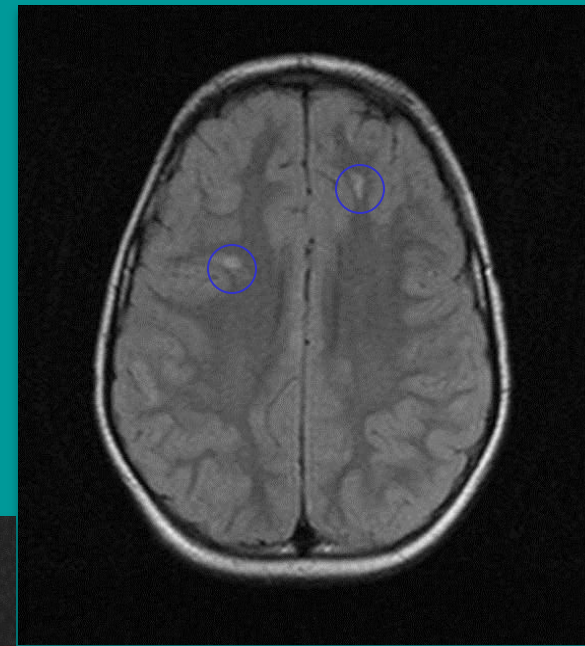


Axial FLAIR MRI

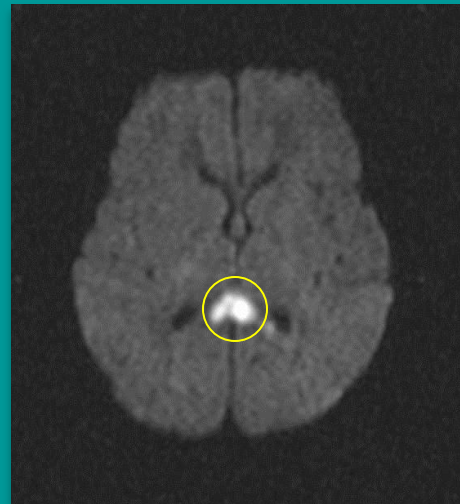


Companion Patient #5: DAI

- Associated with severe shear forces that occur with rapid deceleration
 - i.e. motor vehicle collisions
- Affects interfaces with disparate densities
 - GM/WM interfaces
- Graded by location
 - Frontotemporal
 - Corpus callosum
 - Brainstem
- May show **restricted diffusion on DWI**



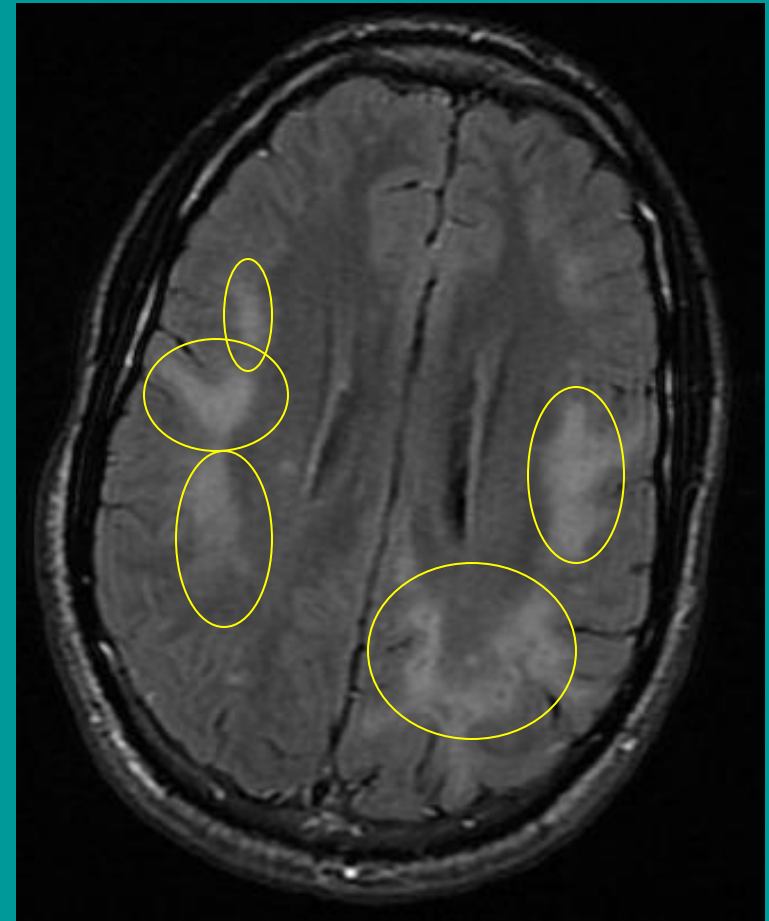
Axial FLAIR MRI



Axial DWI MRI

Companion Patient #6: PML

- Involves reactivation of the John Cunningham (JC) virus in immunocompromised patients
- Male predominance
- Often involves
 - **subcortical WM**
 - corpus callosum
 - GM in up to 50%
- Enhancement is rare
- As with other multifocal processes, regions may become confluent over time.



Axial FLAIR MRI



Now that we have viewed examples of multifocal WM lesions, let's continue to view examples of WM diseases that are more characteristically confluent in appearance.



WM Lesion Distributions: Confluent

- Multifocal examples
 - Small Vessel Ischemic Disease (SVID)
 - Multiple Sclerosis (MS)
 - Diffuse Axonal Injury (DAI)
 - Progressive Multifocal Leukoencephalopathy (PML)
- Confluent examples
 - Adrenoleukodystrophy (ALD) in a pediatric patient
 - Chemotherapy & radiation changes
- Selective examples
 - Central Pontine Myelinolysis (CPM)
 - Marchiafava-Bignami disease
 - Wallerian degeneration
 - Posterior Reversible Encephalopathy Syndrome (PRES)



Companion Pt #7: ALD

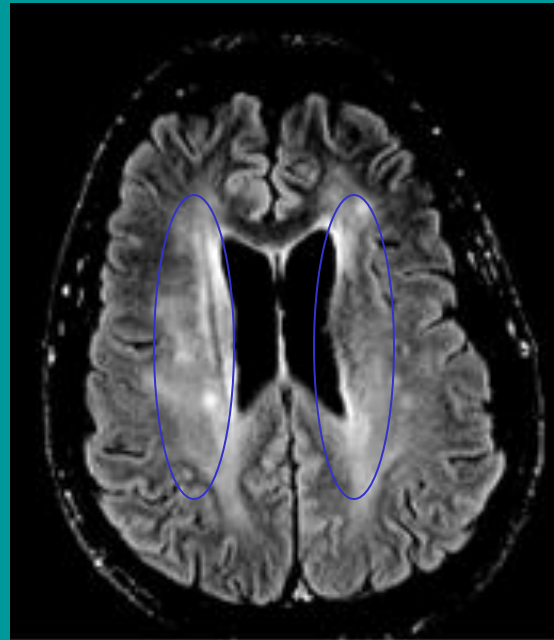
- Leukodystrophies are metabolic & often present in infancy
- **ALD**
 - **Posterior distribution** involving peritrial & occipital WM, corpus callosum & fornix
 - May show enhancement
- Metachromatic Leukodystrophy
 - Frequently diffuse
- Alexander's Disease
 - Often has a frontal distribution



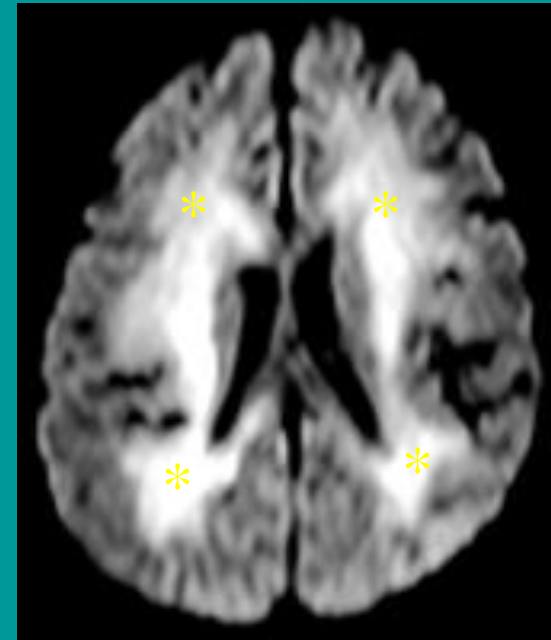
Axial T2 MRI *Courtesy of Dr. Rojas*

Companion Patient #8: Radiation & Chemotherapy Change

- Often symmetric bilaterally with scalloped outer margins
- Frequently involves periventricular WM
- Associated w/ atrophy
- Often shows variable peripheral enhancement & restricted diffusion
- PET or perfusion MRI may be helpful to distinguish from tumor



Axial FLAIR MRI *McKinney, et al*



Axial DWI MRI *McKinney, et al*



Now that we have viewed examples of confluent WM lesions, let's continue to view examples of WM diseases that each tend to be selective for a particular area of the brain.



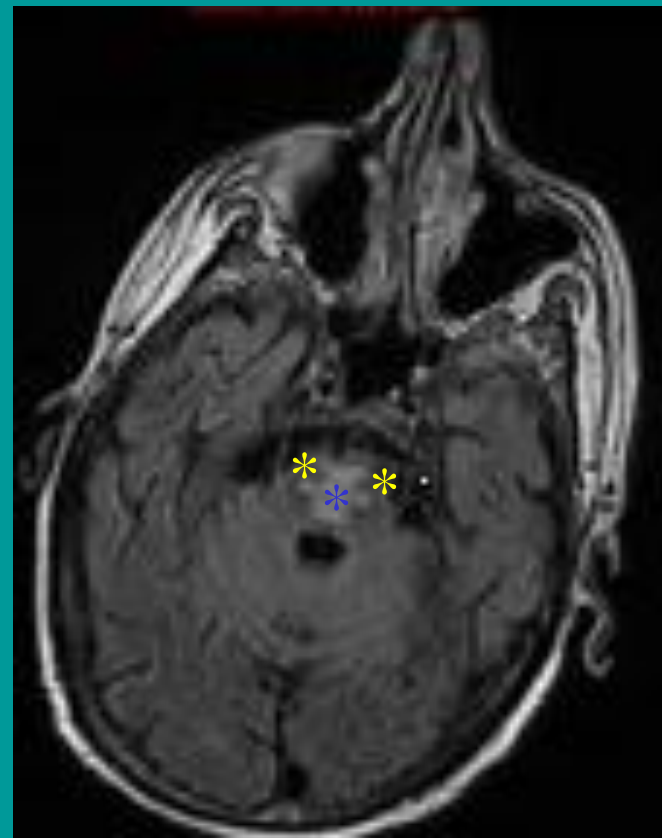
WM Lesion Distributions: Selective

- Multifocal examples
 - Small Vessel Ischemic Disease (SVID)
 - Multiple Sclerosis (MS)
 - Diffuse Axonal Injury (DAI)
 - Progressive Multifocal Leukoencephalopathy (PML)
- Confluent examples
 - Leukodystrophies in a pediatric patient
 - Chemotherapy & radiation changes
- **Selective examples**
 - **Central Pontine Myelinolysis (CPM)**
 - **Marchiafava-Bignami disease**
 - **Wallerian degeneration**
 - **Posterior Reversible Encephalopathy Syndrome (PRES)**



Companion Patient #9: CPM

- Also known as osmotic demyelination syndrome
- Associated with
 - Rapid or overcorrection of hyponatremia
 - Alcoholism, malnutrition, debilitating disease
- Early
 - Localized to the **central pons**
 - Sparing of **corticospinal tracts**
 - “**Snake eyes**” appearance
- Extra-pontine myelinolysis
 - Commonly involves midbrain & basal ganglia.



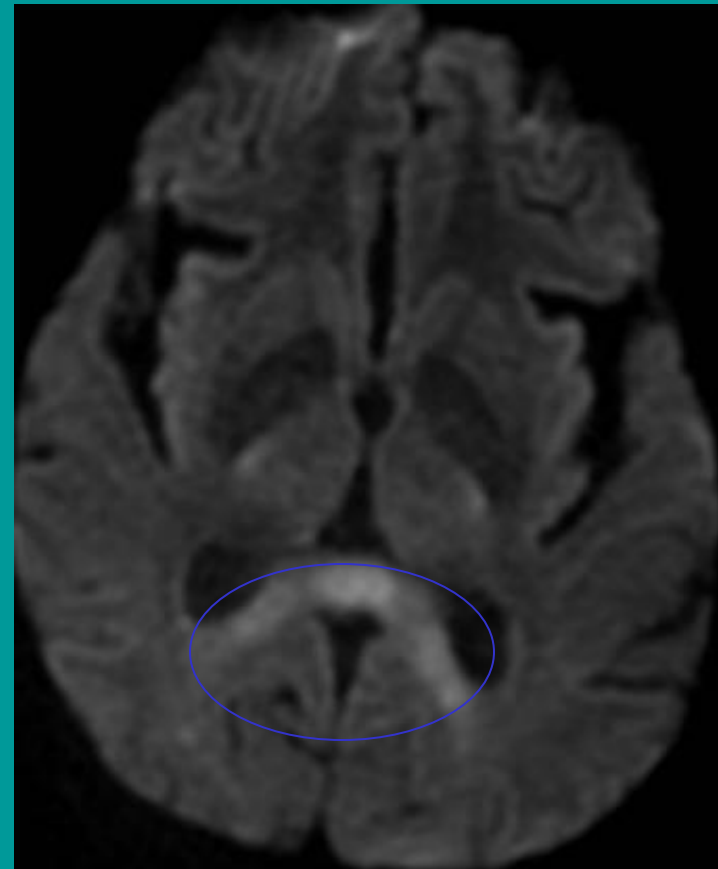
Axial T2 MRI

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Companion Patients #10: Marchiafava-Bignami Disease

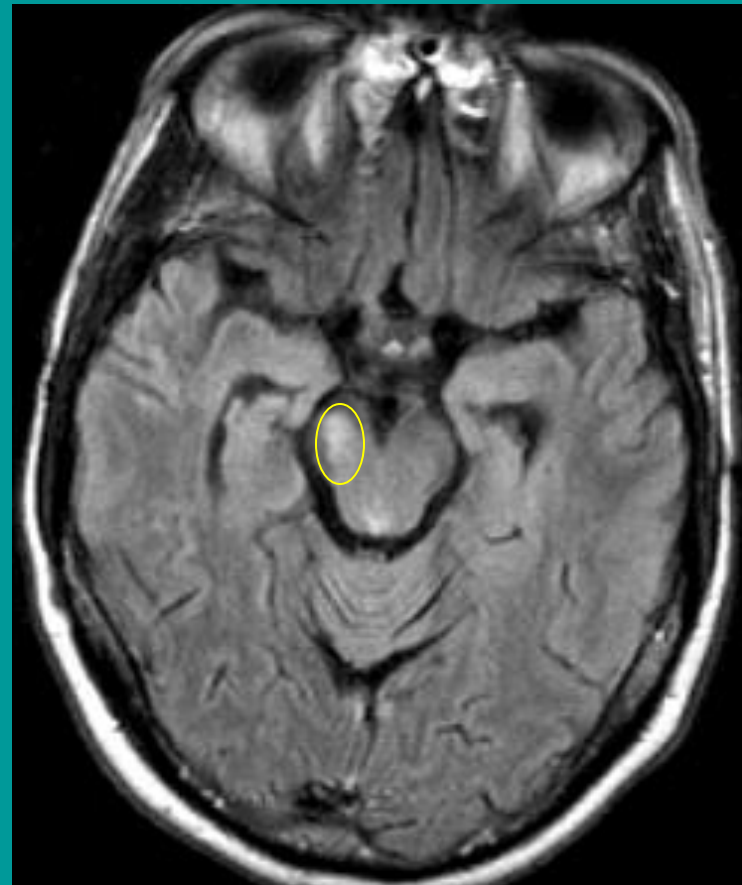
- Relatively rare syndrome leading to demyelination & atrophy
- Characteristically involves **corpus callosum**
- More common in men
- Commonly associated with alcoholism
- Often shows **restricted diffusion**





Companion Patients #11: Wallerian Degeneration

- Also known as orthograde or anterograde degeneration
- Involves injury to axons anywhere along their course
- In central nervous system (CNS), often involves **corticospinal tracts of cerebral peduncles**
- Myelin clearance by microglia in CNS is relatively slow compared to clearance by macrophages in peripheral nervous system.

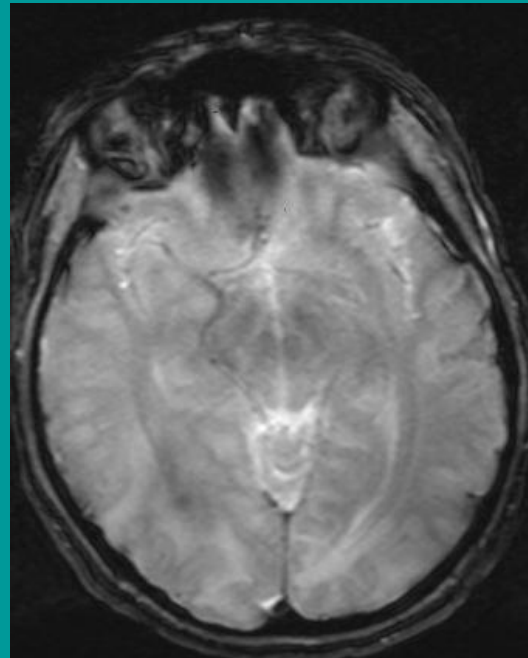


Axial FLAIR MRI

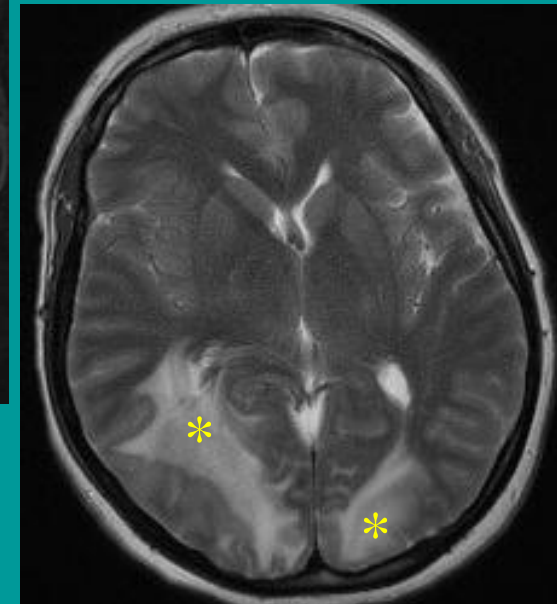


Our Patient: Suspected Diagnosis

- 60 year old female with hypertension to 160/100, headache & altered mental status.
- Absence of restricted diffusion in regions corresponding to **T2 hyperintensity**
 - No cytotoxic edema to raise concern for infarct
- Diagnosis: PRES



Axial DWI MRI *BIDMC PACS*

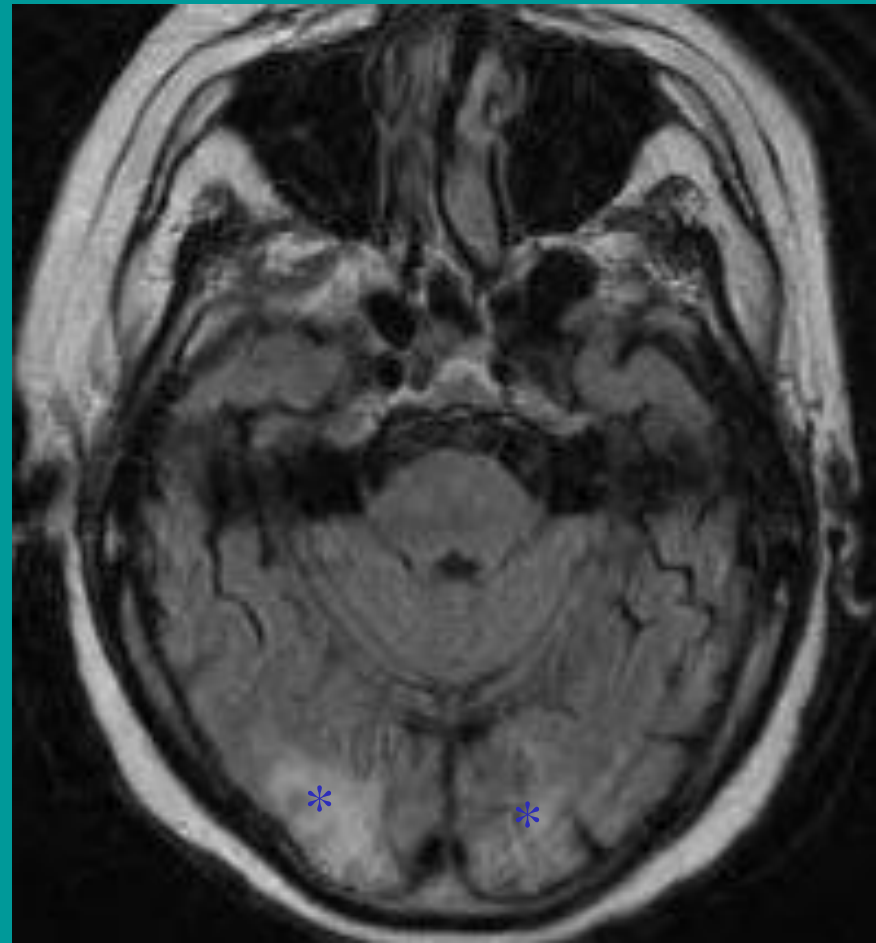


Axial T2 MRI



Companion Patient #12: PRES

- Controversial mechanism, but thought to be related to chronic hyperperfusion & blood-brain barrier breakdown
 - Results in plasma leakage & vasogenic edema
- Associated w/ HTN & immunosuppressant therapy
- Often involves bilateral occipital & parietal watershed areas
 - May involve GM & frontal lobes
- **WM injury is largely reversible.**



Summary

- Relative to GM, WM is normally high in fat & low in fluid making it hyperintense to GM on T1 & hypointense on T2
 - Demyelination leads to T2 hyperintensity
- Diagnosis of WM disease relies upon
 - Clinical features
 - Patient demographics: age, gender
 - Patient history & physical exam: signs, symptoms & chronicity
 - Laboratory results
 - Characteristic radiologic distributions
 - Multifocal: MS commonly in middle-aged females, SVID in older patients
 - Confluent: Leukodystrophies in children, radiation & chemotherapy change
 - Selective: CPM in central pons early, PRES in bilateral parietooccipital lobes



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