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Posterior Reversible Encephalopathy Syndrome

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Outline

- Case
- Brief overview of PRES
- Typical Imaging features with anatomy review
- Imaging Differential Diagnosis



Case: Presentation

- 41 year old female presented to the ED with headache, blurred vision, and unclear left leg weakness
- Recent history of falls and vertigo
- Possible seizure episodes consisting of urinary incontinence and “foaming at the mouth” over the past 5 days



Case

- Physical exam: limited due to lack of patient effort and functional overlay.
- Pertinent positives: Subjective left leg weakness; Anion gap acidosis on routine labs
- Vital signs: T: 97 HR: 89-130 RR: 16 SaO₂: 95% on room air.
- BP was 128/75 upon admission, but later spiked to 200/100 requiring transfer to the MICU



Case: Differential

- Infectious—encephalitis/meningitis
- Ischemic/Infarct
- Trauma/hemorrhagic
- Cerebral vein thrombosis
- CNS vasculitis
- Demyelinating condition
- Malignancy
- ...the list goes on....

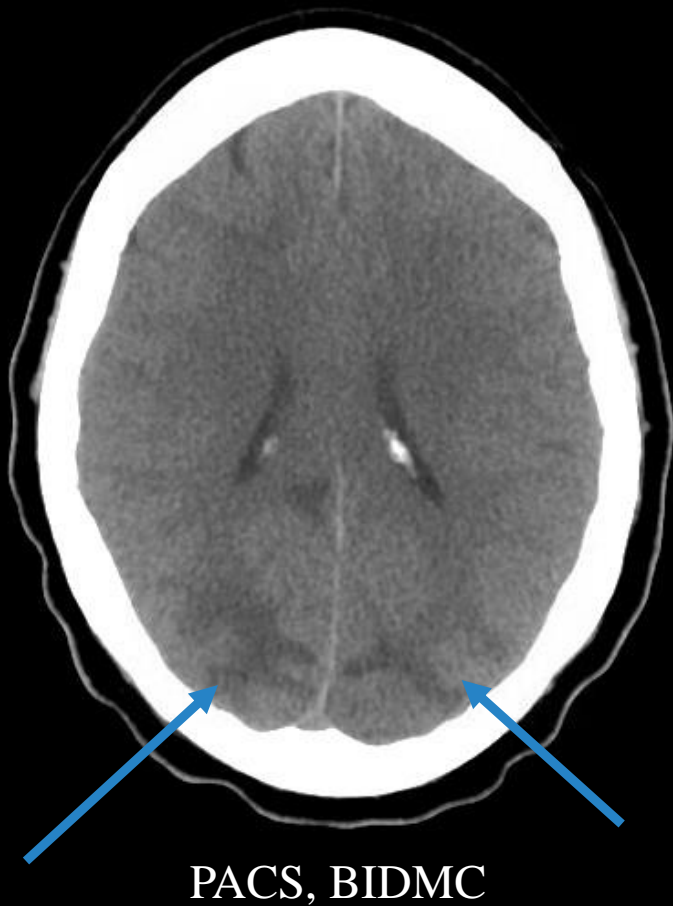


Case: Imaging

- Step 1: rule out acute hemorrhage!



Imaging—Non-Contrast Head CT



- No evidence of acute bleed
- No evidence of evidence of fracture
- No midline shift or evidence of intracranial hypertension
- Multiple subcortical white matter hypoattenuating regions involving the occipital, parietal, and frontal lobes



Differential Diagnosis

- ~~Trauma/hemorrhagic~~
- Ischemic/Infarct
- Infectious—encephalitis/meningitis
- Cerebral vein thrombosis
- CNS vasculitis
- Demyelination
- Malignancy
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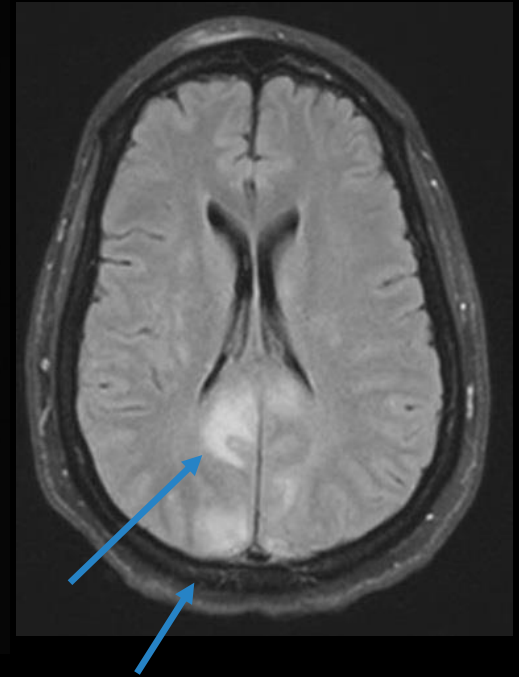
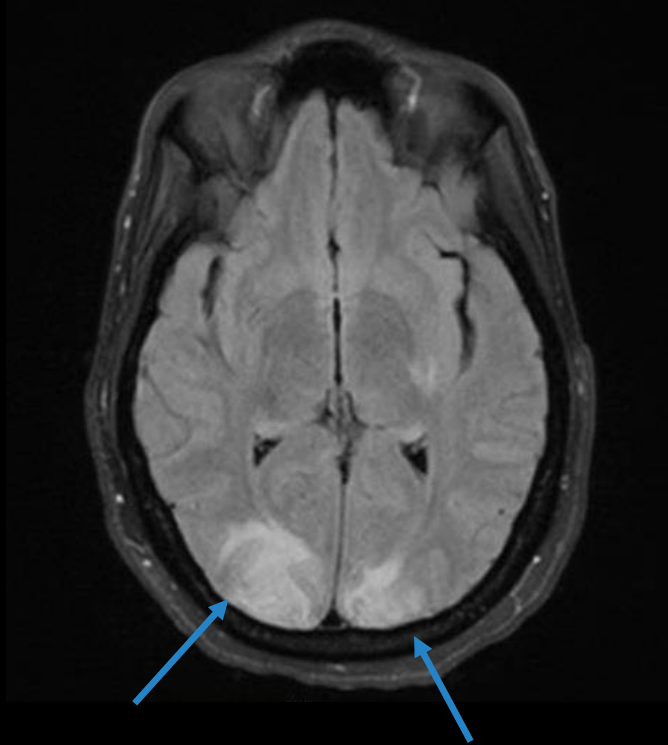


Imaging

- Step 1: rule out acute hemorrhage with non-contrast head CT
- Step 2: More refined information with MRI



Imaging--MRI



FLAIR

Hyper-intensities in the bilateral occipital, bilateral posterior frontal, posterior right cingulate, and bilateral basal ganglia. Most likely diagnosis—PRES.



PRES: Overview

- Posterior Reversible Encephalopathy Syndrome
- Does not have to be posterior; not always reversible
- Two theories as to the pathophysiology of PRES
 - Acute increase in blood pressure → Hyper-perfusion
 - Endothelial dysfunction → Hypo-perfusion → reflex hypertension
- Believed that edema is concentrated in the posterior hemispheres due to decreased autoregulation of blood pressure and sympathetic tone in the posterior fossa



PRES: Aggravating factors

- Sudden increases in blood pressure
- Immunosuppressant drugs
- Renal Failure
- Certain Chemotherapies
- Autoimmune disorders (50% of people with PRES)



Symptoms of PRES

- Headache
- Visual Disturbances
- Confusion
- Tonic-Clonic Seizures
- Focal Neurologic Deficits



Diagnostic Complications

- Clinical criteria is non-specific
- No gold standard with only retrospective information
- Diagnosis does change management
- This makes imaging extremely important!

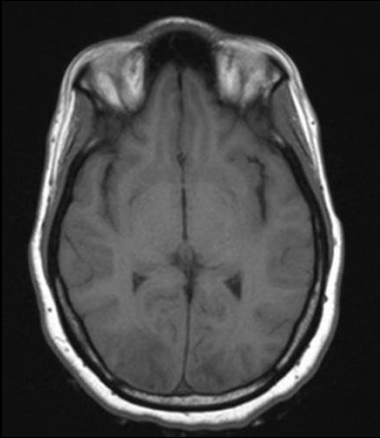


Typical Imaging Features of PRES

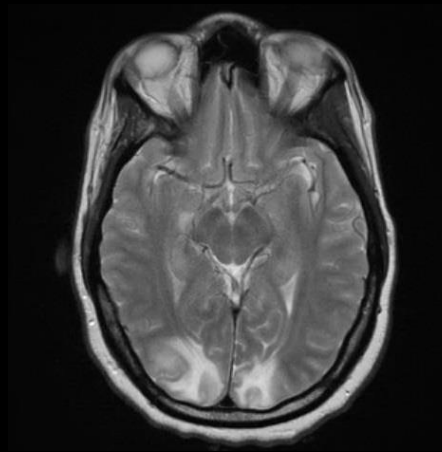
- Bilateral vasogenic edema in the parietal-occipital regions
- Affects both the cortical and the sub-cortical regions
- Bright on T2 sequences, FLAIR is the most sensitive.
- Contrast enhancement is inconsistent
- Hallmark → imaging changes are reversible



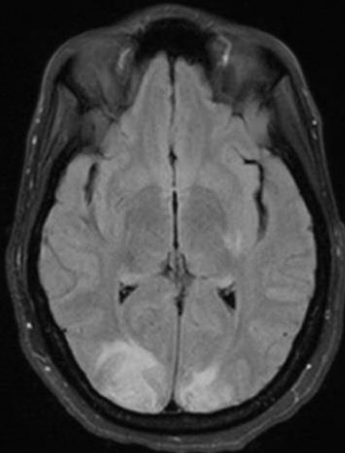
Typical Imaging Features of PRES



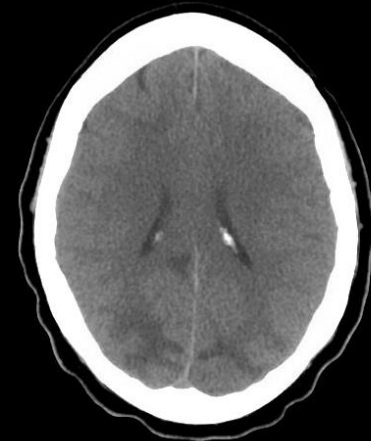
T1: Hypointense



T2: Hyperintense



FLAIR: Hyperintense



CT: Hypoattenuating¹⁶



Follow-up imaging: our patient



Original CT: 12/2015



Follow up: 12/2016

Resolution of prior imaging findings!



Alternative Imaging Features

- Main distribution patterns
 - 1. Parietal-occipital bilaterally
 - 2. Watershed distribution over the full hemisphere
 - 3. Superior frontal sulcus



Alternative Imaging Features

Parietal Orbital Pattern

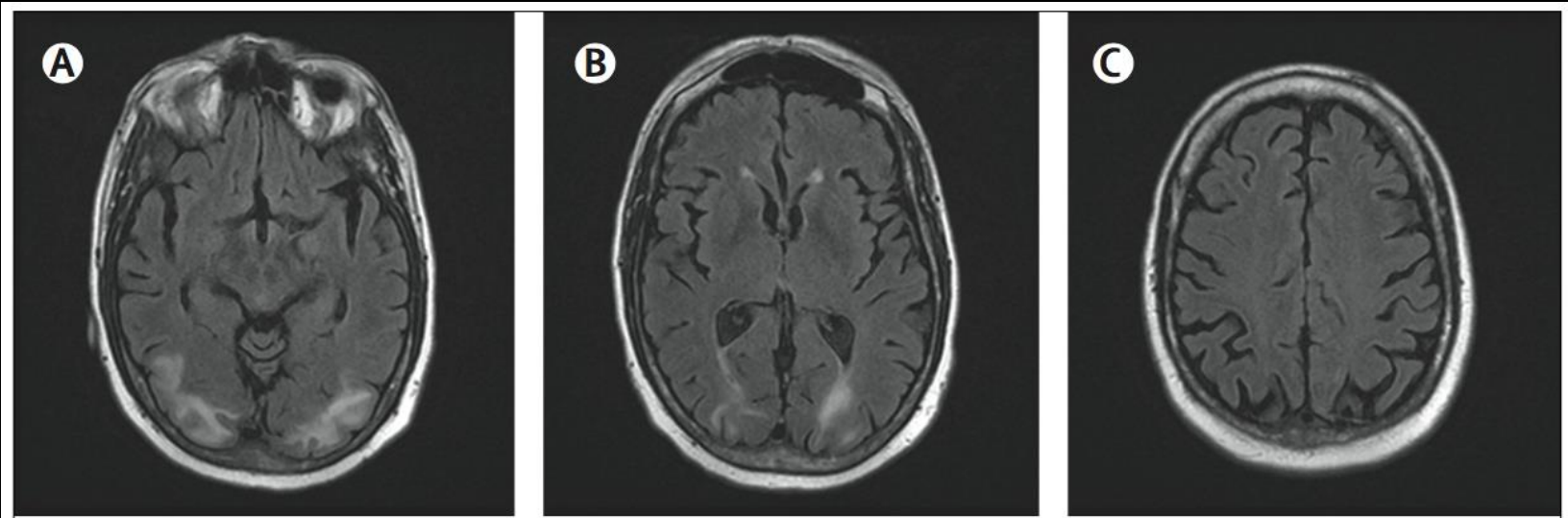


Image obtained by Petrovic et al. 2011



Alternative Imaging Features

Watershed Distribution

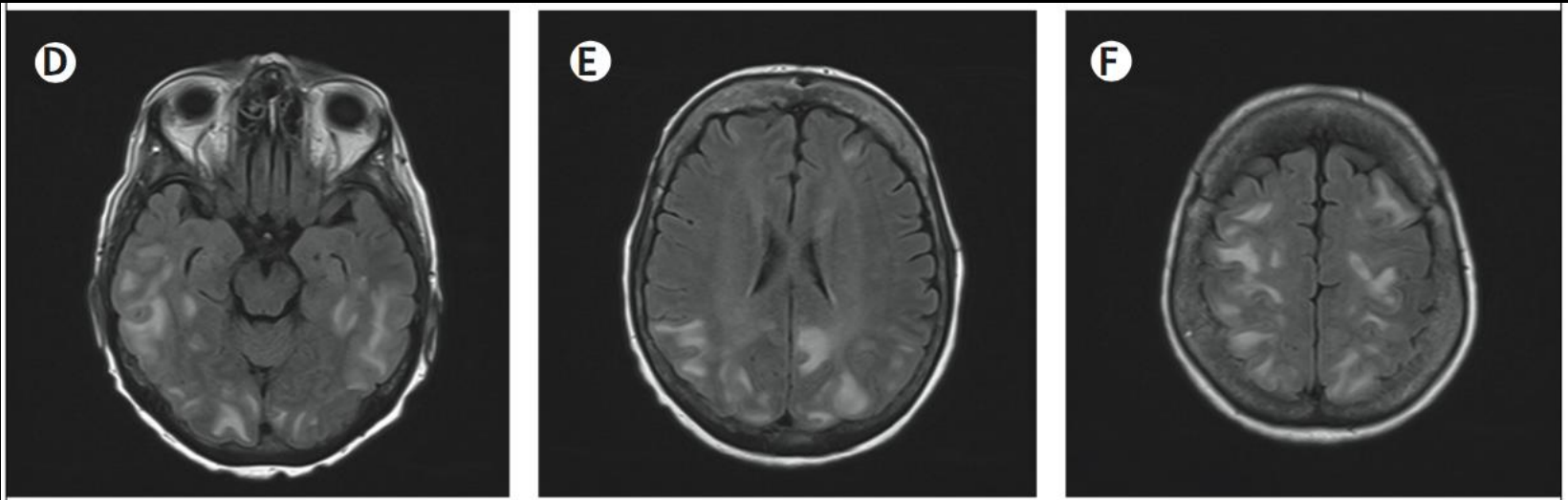


Image obtained by Petrovic et al. 2011



Alternative Imaging Features

Superior Frontal Sulcus Pattern

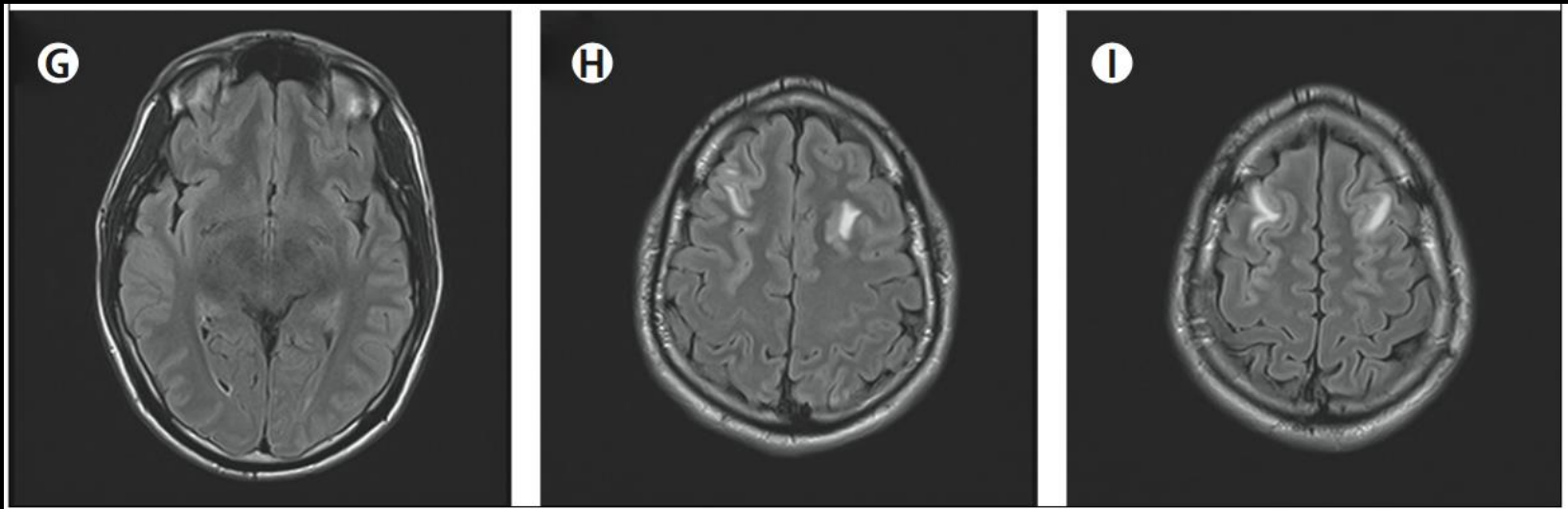


Image obtained by Petrovic et al. 2011




Best Imaging Modalities for PRES

- No strict criteria due to limited research
 - MRI FLAIR is considered the most sensitive
 - Many patients receive a CT because of acute onset headache
 - Additional imaging (such as CTV or MRV) may be necessary depending on the specific clinical presentation



Differential Diagnosis of Imaging

- Trauma/hemorrhagic  Rule out with CT
- Ischemic/Infarct
- Cerebral vein thrombosis
- CNS vasculitis
- Progressive Multifocal Leukoencephalopathy
- Autoimmune encephalitis: Acute disseminating encephalomyelitis
- Malignancy: Gliomatosis cerebri



Differential Diagnosis of Imaging

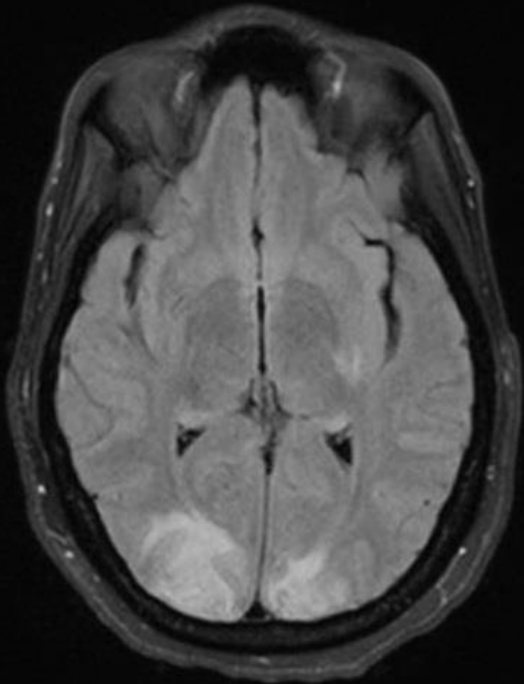
Infarct

- Ways to differentiate
 - Vascular territories
 - DWI restriction

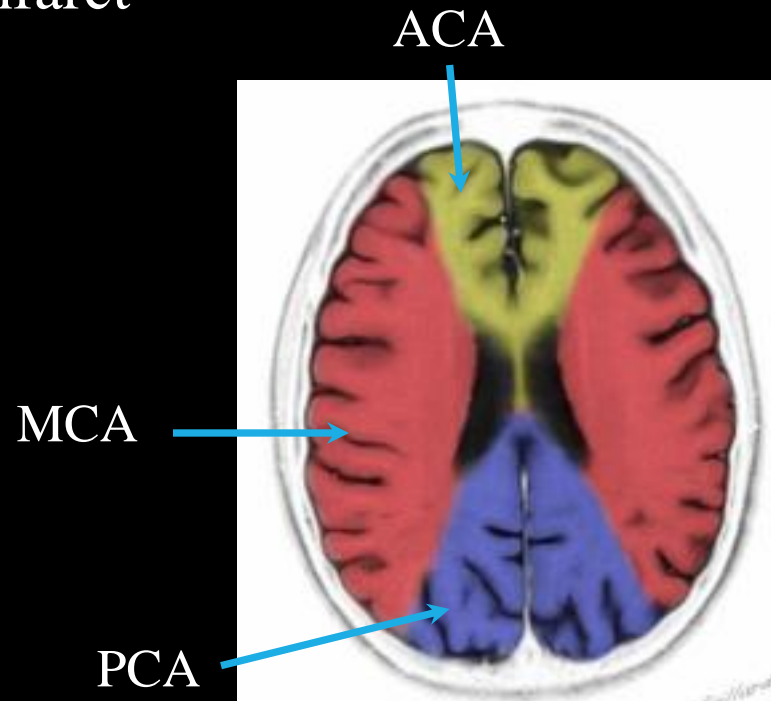


Differential Diagnosis of Imaging

Infarct



Our Patient
PACS, BIDMC



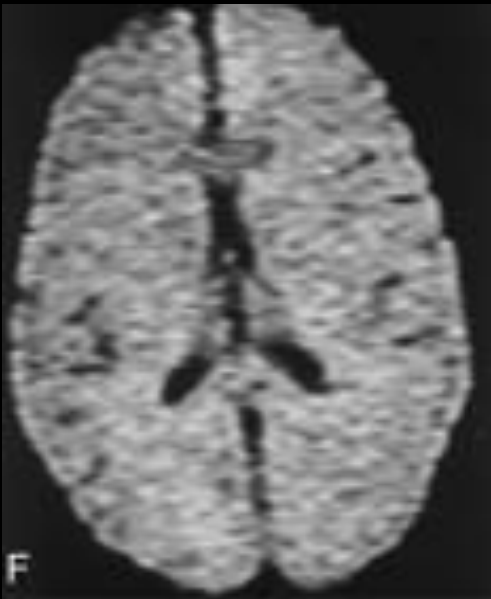
Vascular Territories
Obtained from www.radiopedia.com

PRES is less likely to fall in a strict vascular distribution



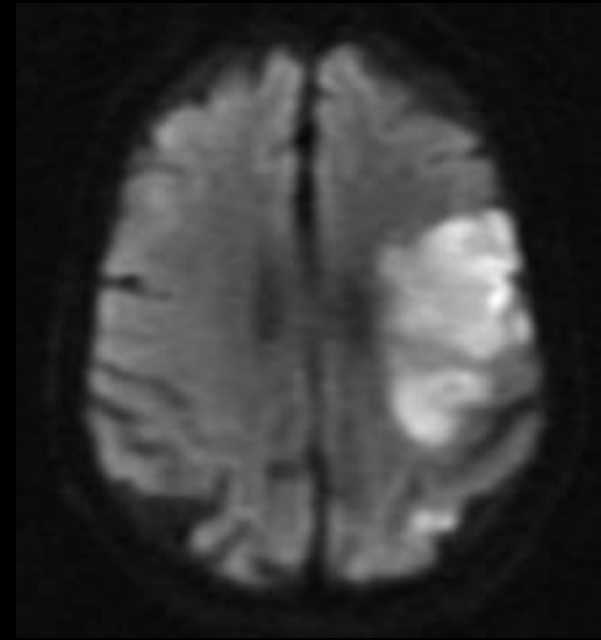
Differential Diagnosis of Imaging

Infarct



PRES on DWI

Obtained from Covarrubias,
Luetmer, and Campeau 2002



Infarct on DWI

Dr. Gagandeep Choudhary
<https://radiopaedia.org/cases/left-middle-cerebral-artery-territory-infarct>

PRES is not diffusion restricted on DWI



Differential Diagnosis of Imaging

- Trauma/hemorrhagic → Rule out with CT
- Ischemic/Infarct → Vascular distribution and DWI
- Cerebral venous thrombosis
- CNS vasculitis
- Progressive Multifocal Leukoencephalopathy
- Autoimmune encephalitis: Acute disseminating encephalomyelitis
- Malignancy: Gliomatosis cerebri



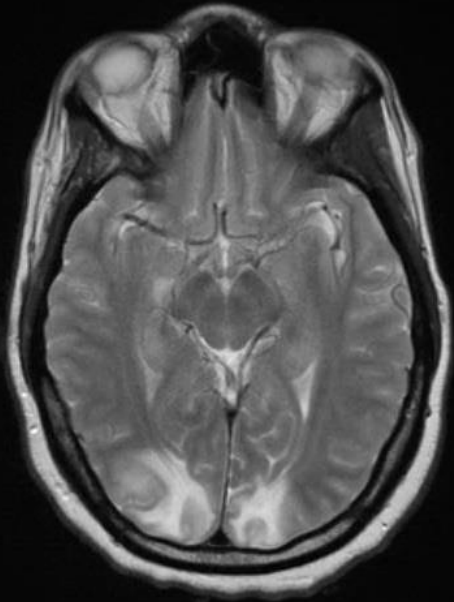
Differential Diagnosis of Imaging

Cerebral Venous Thrombosis

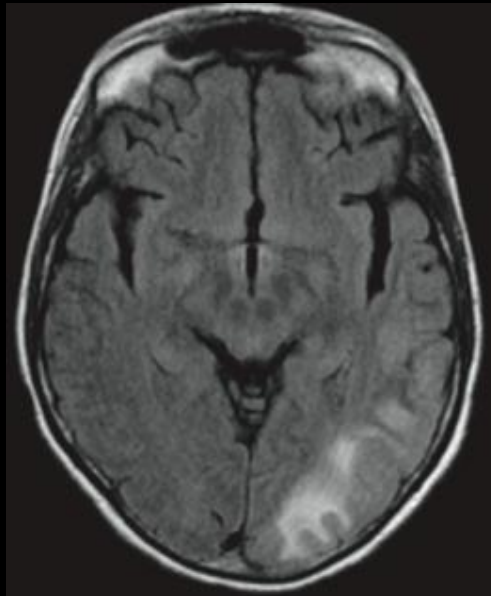
- Symptoms: vague and similar to PRES
 - Headache
 - Nausea
 - Seizures
- MRI Imaging
 - Can develop edema and show a similar imaging pattern to PRES
 - Should not be bilateral, may show hemorrhage
 - Magnetic resonance venography or CTV can confirm diagnosis if unclear



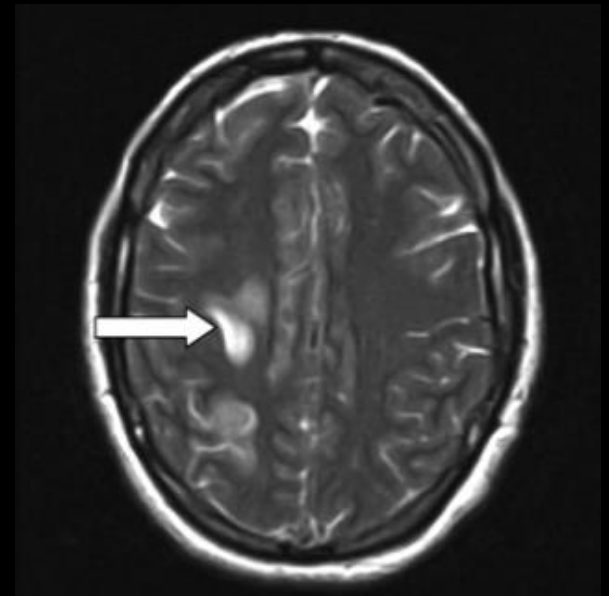
Differential Diagnosis of Imaging Cerebral Venous Thrombosis



PRES on T2 MRI
Our Patient
PACS, BIDMC



PRES on FLAIR MRI
Obtained from Stevens and
Haran, 2012



CVT on T2 MRI
Obtained from Saposnik et al.
2011



Differential Diagnosis of Imaging

- Trauma/hemorrhagic → Rule out with CT
- Ischemic/Infarct → Vascular distribution and DWI
- Cerebral venous thrombosis → MRV or CTV
- CNS vasculitis
- Progressive Multifocal Leukoencephalopathy
- Autoimmune encephalitis: Acute disseminating encephalomyelitis
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Differential Diagnosis of Imaging

- **CNS vasculitis**
 - Infarcts in different vascular territories
 - Will not be reversible
 - MRV/CTV to confirm
- **Progressive Multifocal Leukoencephalopathy**
 - Spares the cortex
 - Would not be reversible
- **Acute Disseminating Encephalomyelitis (ADEM)**
 - Also has bilateral T2 and FLAIR hyperintense lesions
 - Follows clinical signs of infection
- **Gliomatosis Cerebri**
 - Tends to be more asymmetric
 - Would not be reversible



Follow-up

- Remove aggravating factors
 - Control blood pressure
 - Take off potentially causal drugs in the acute setting
 - Treat associated conditions (i.e. renal disease)
 - Treat seizures with anti-epileptics



Prognosis

- Generally a good prognosis
- Complications include
 - Intracranial and subarachnoid hemorrhage (10-25%)
 - Brainstem compression
 - Increased intracranial pressure
- Mortality rates are variable due to the unclear diagnostic criteria for PRES



Summary

- PRES is an uncommon, but important clinical entity
- Diagnostic presentation is inconsistent and vague, giving imaging a greater role in diagnosis
- If PRES is treated it can be entirely reversed, however delayed treatment can lead to permanent neurologic injury and death



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References

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