AMSER Case of the Month February 2025

38 year-old male with left hemiparesis and seizure

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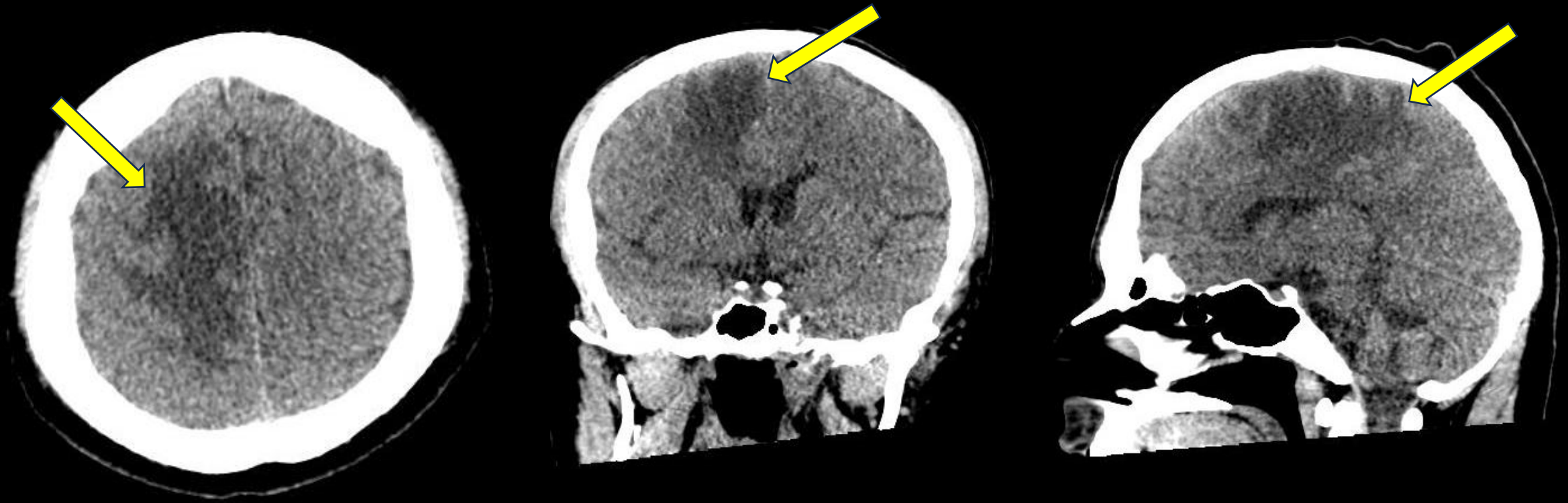
Patient Presentation

- HPI: 38-year-old male with PMH of HTN, HLD, DM1 complicated by ESRD on hemodialysis since 2023, presented with 2-week left hemi body weakness and 1-month onset of seizure, blurry vision, and headache. Patient was transferred from outside medical center to our neurosurgery service for workup of right-sided brain lesion found on MRI. Non-contrast head CT and CTA of the head and neck found vasogenic edema without large vessel occlusion or stenosis.
- PE: pertinent for reduced muscle strength in left UE and LE 1-4/5, 60% sensation in left UE and LE compared to the right side.

Pertinent Labs

- Cr 7.63 (H), CRP 0.42, ESR 7
- CSF studies: lactate 1.9, pyruvate 0.134 (H), protein 36, glucose 101, RBC 151, negative gram stain and culture, negative for Cryptococcus, CMV, West Nile, Meningitis/Encephalitis panel, unremarkable cytopathology
- Serology studies: negative for ANCA, ANA, cardiolipin antibodies, beta 2 IgG/IgM, and cryoglobulin

Initial head CT obtained on admission



Axial, coronal and sagittal head CT images demonstrate a large area of vasogenic edema throughout the paramedian right frontal and parietal lobe

What Imaging Should We Order Next?

ACR Appropriateness Criteria

Variant 3: **Adult. Suspected intraaxial brain tumor based on prior imaging. Pretreatment evaluation.**

Procedure	Appropriateness Category	Relative Radiation Level
MRI head perfusion with IV contrast	Usually Appropriate	O
MRI head without and with IV contrast	Usually Appropriate	O
MR spectroscopy head without IV contrast	May Be Appropriate	O
MRI complete spine without and with IV contrast	May Be Appropriate	O
MRI functional (fMRI) head without IV contrast	May Be Appropriate	O
MRI head perfusion without IV contrast	May Be Appropriate	O
MRI head without IV contrast	May Be Appropriate	O
MRI head without IV contrast with DTI	May Be Appropriate	O
Fluciclovine PET/MRI brain	May Be Appropriate	☢☢☢
Fluciclovine PET/CT brain	May Be Appropriate	☢☢☢☢
MRI complete spine with IV contrast	Usually Not Appropriate	O
MRI complete spine without IV contrast	Usually Not Appropriate	O
MRI head with IV contrast	Usually Not Appropriate	O
CT head with IV contrast	Usually Not Appropriate	☢☢☢
CT head without and with IV contrast	Usually Not Appropriate	☢☢☢
CT head without IV contrast	Usually Not Appropriate	☢☢☢
DOTATATE PET/CT brain	Usually Not Appropriate	☢☢☢
DOTATATE PET/MRI brain	Usually Not Appropriate	☢☢☢
FDG-PET/CT brain	Usually Not Appropriate	☢☢☢
FDG-PET/MRI brain	Usually Not Appropriate	☢☢☢

This follow-up imaging modality was ordered by the neurosurgeon

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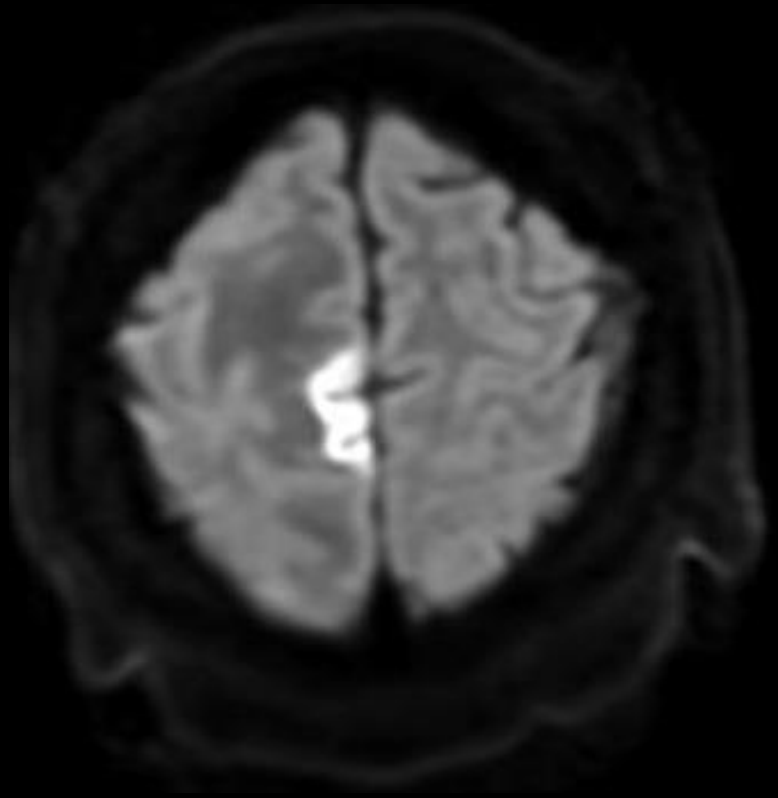
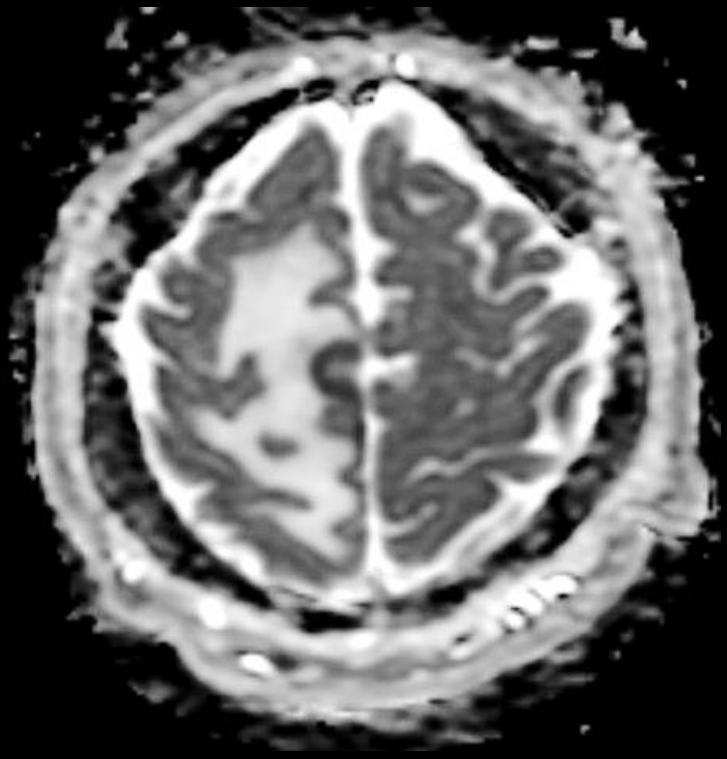
Findings (unlabeled)

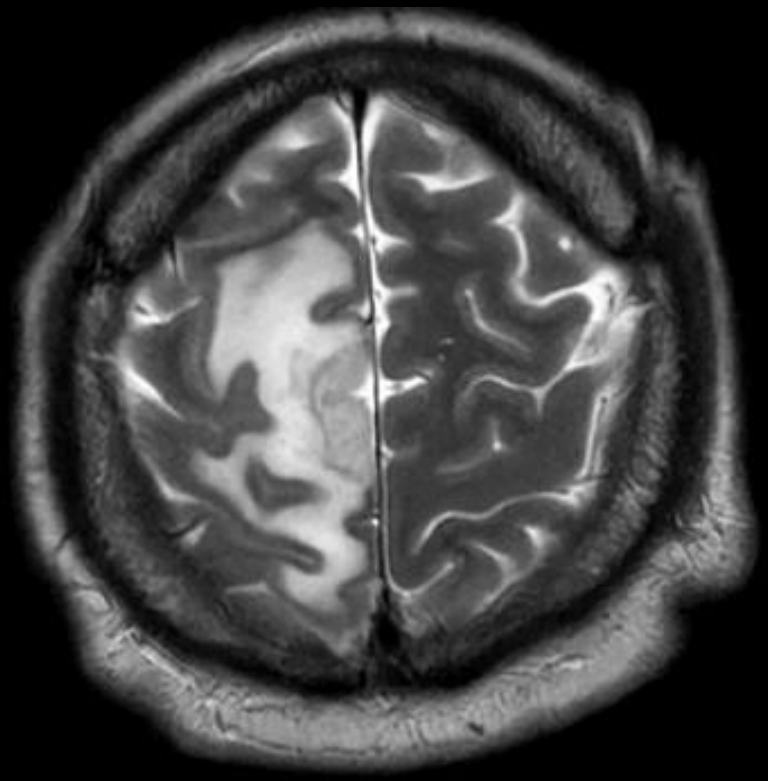


DWI

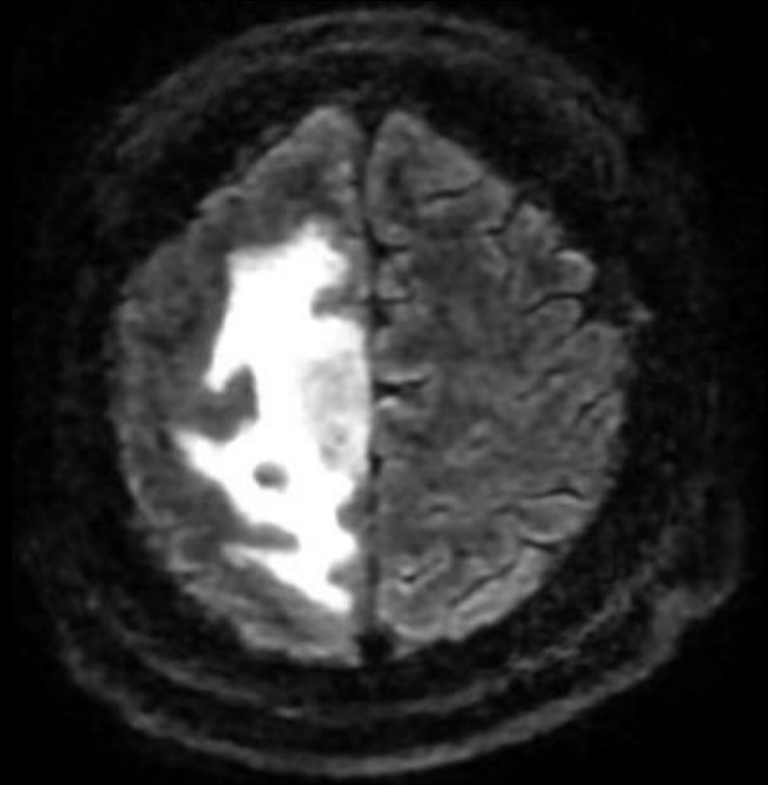


ADC

Findings (unlabeled)

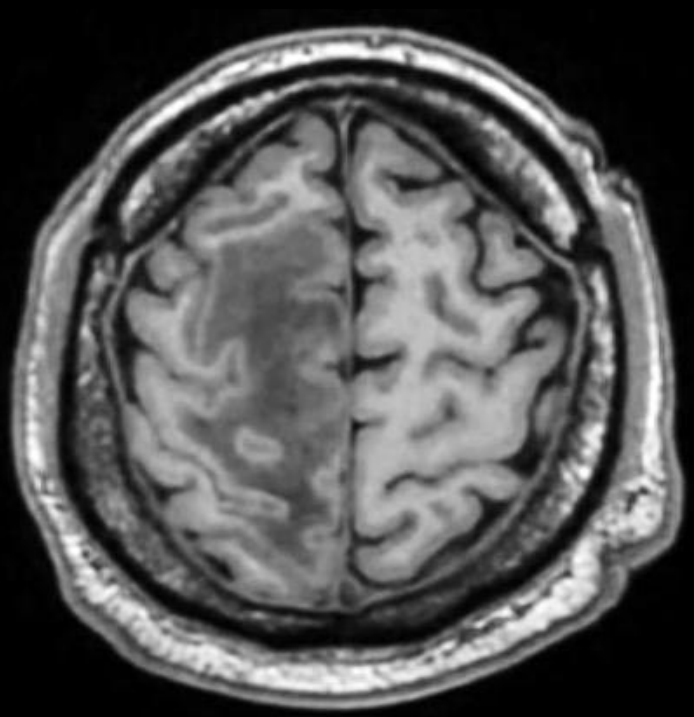


T2

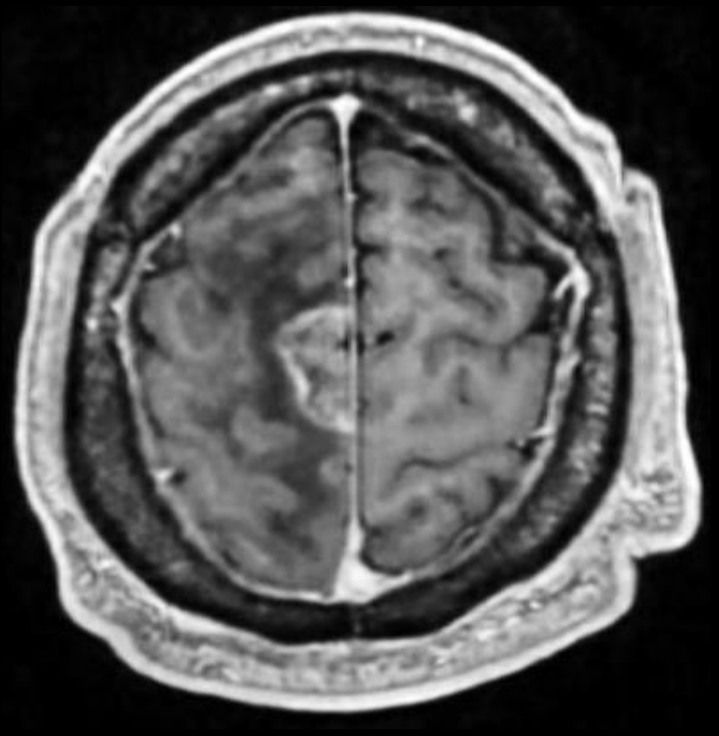


FLAIR

Findings (unlabeled)

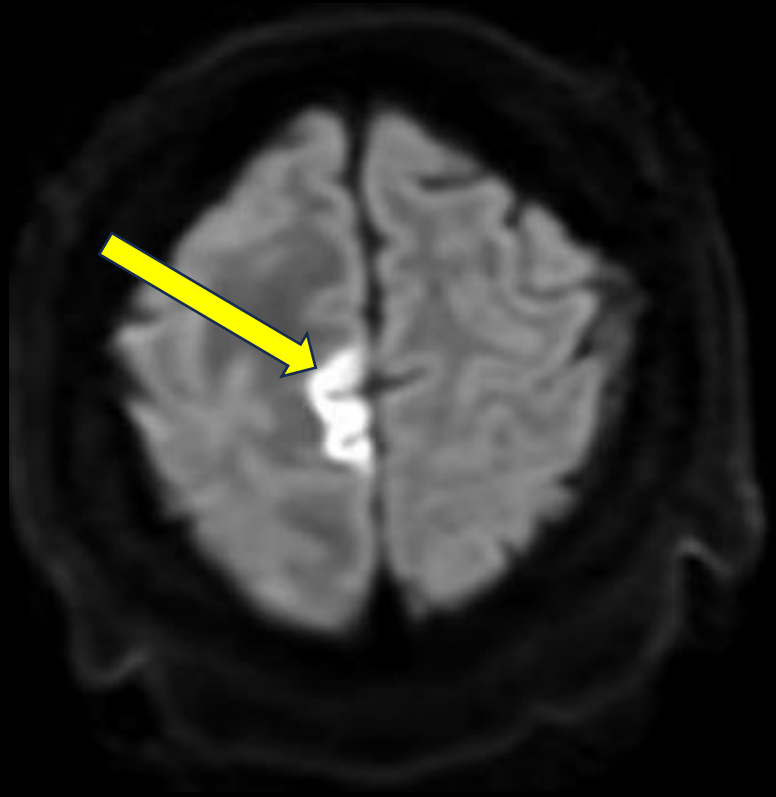


T1-Pre

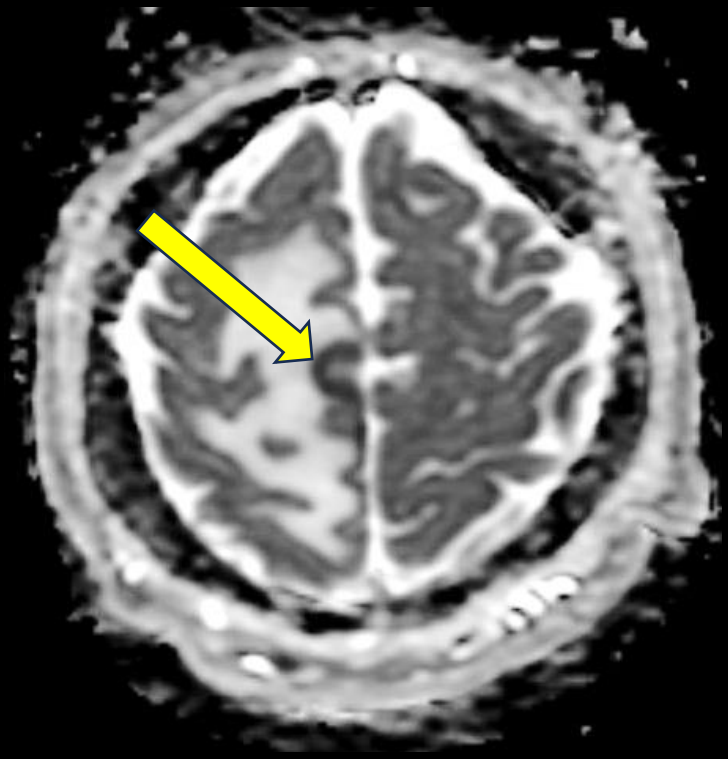


T1-Post

Findings (labeled)



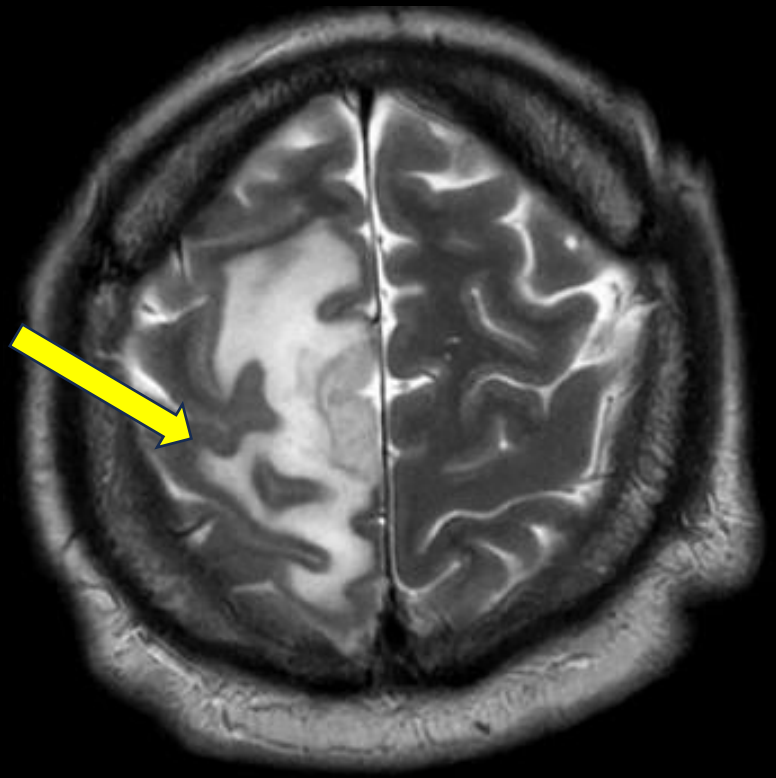
DWI



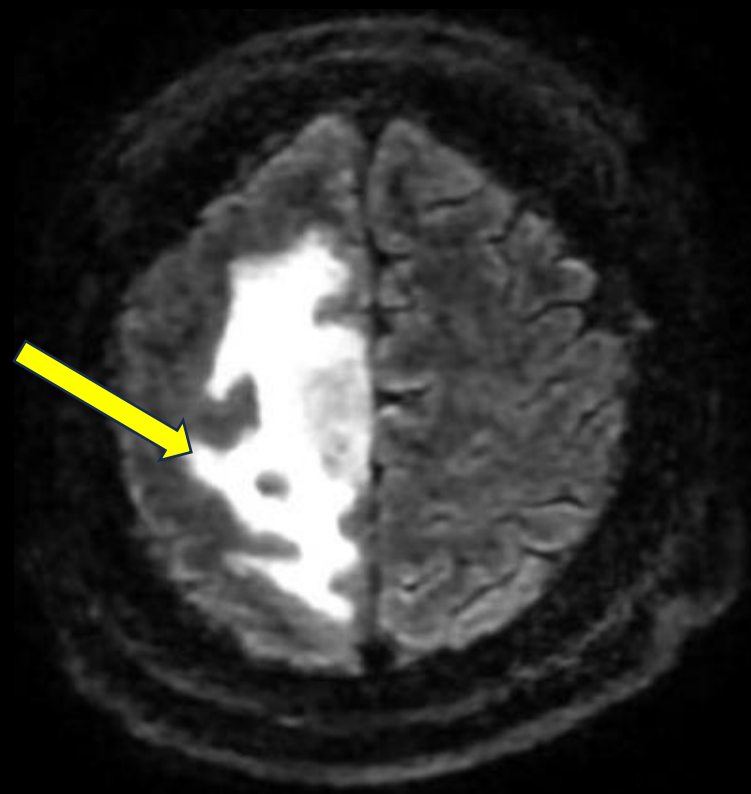
ADC

DWI and ADC images demonstrate a region of restricted diffusion in the posterior paramedian right frontal lobe

Findings (labeled)



T2



FLAIR

T2 and FLAIR images demonstrate a large area of vasogenic edema surrounding the area of restricted diffusion in the posterior paramedian right frontal lobe

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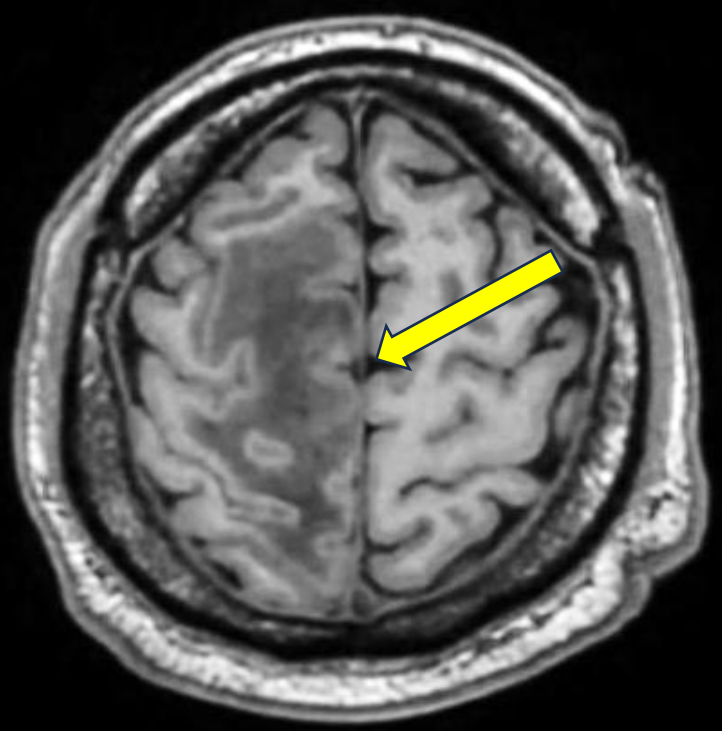
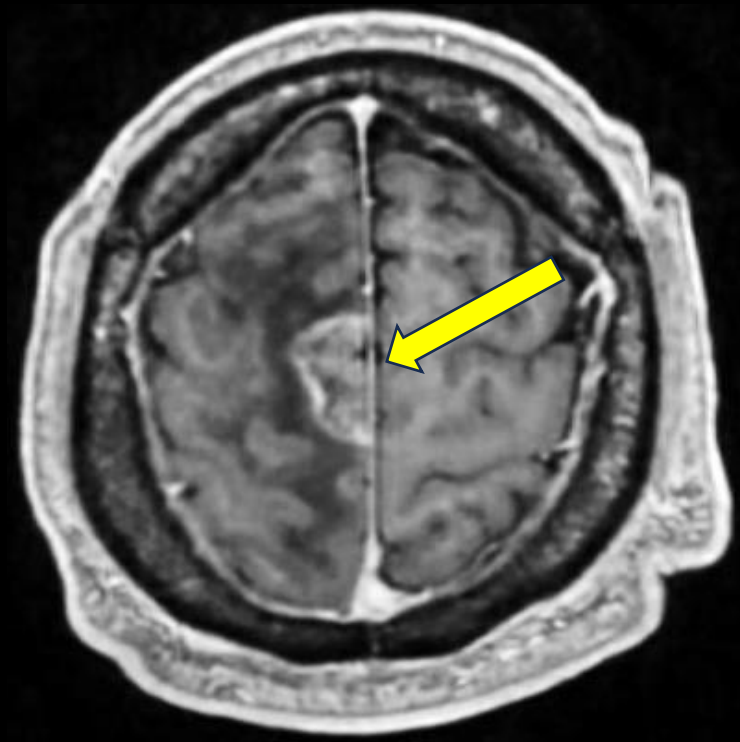
Findings (unlabeled)



T1-Pre



T1-Post

T1-pre and T1-post contrast images demonstrate heterogenous enhancement within the area of restricted diffusion

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Follow-up biopsy and genetic finding

- Biopsy was negative for malignancy but showed:
  - Occlusive vasculopathy and microangiopathy
    - Narrowing/occluding and microangiopathic changes of medium-sized leptomeningeal blood vessels and smaller vessels in deeper brain parenchyma
  - Acute/subacute ischemic necrosis of brain
    - Acute ischemic neuronal necrosis and cavitating subacute necrosis with mixed populations of macrophages and neutrophils surrounded by brain tissue rich in reactive fibrous astrocytes
- Based on the above, a diagnosis of CNS vasculitis was made.
- Patient was later found to be heterozygous for RAG2 mutation.

Final Dx:

Primary CNS vasculitis (Primary angiitis of the central nervous system)

Case Discussion

- Vasculitis is a known major complication in patients with partial RAG deficiency (including heterozygous for RAG2 mutation).
- Primary CNS vasculitis:
  - Rare (2.4 cases/million person-year) inflammatory disorder of the blood vessels of brain the spinal cord without evidence of systemic vasculitis
  - Onset is typically insidious, but can present acutely
  - Etiology and pathogenesis are unknown but there are proposed triggers including infection (bacterial, viral), connective tissue disorders and systemic vasculitides, and amyloid angiopathy (in animal models)
  - 3 common types have been described: granulomatous (numerous granulomas with multinucleated cells), necrotizing (fibrinoid necrosis) and lymphocytic (extensive lymphocytic inflammation with plasma cells)

Case Discussion (Continued)

- Diagnosis for primary CNS vasculitis can be challenging because:
  - Symptoms are non-specific (headache, stroke, cognitive dysfunction, seizure, TIA, aphasia, visual change, ataxia, papilledema, intracranial bleeding)
  - Biomarkers (ESR, CRP, RF, ANCA) are not sensitive or specific
  - Ischemia is common, CSF shows abnormality ~ 90% of cases
- Diagnostic criteria: meeting all three of the following:
  - Presence of unexplained acquired neurologic deficit (after ruling out other causes)
  - Presence of an inflammatory arteritic process within the CNS
  - No evidence of secondary cause of systemic vasculitis or other disorders

Case Discussion (continued)

- MRI without and with contrast is important imaging modality to rule out other etiologies such as tumor, infection or demyelination when suspecting CNS vasculitis.
- Although lymphocytic is the most common subtype, our case presents the necrotizing subtype evidenced by fibrinoid necrosis and infiltration of mixed macrophage and neutrophils on biopsy.
- Our patient improved on steroids and continued to do well at 1-month outpatient follow-up.

References:

Geier CB, Farmer JR, Foldvari Z, et al. Vasculitis as a Major Morbidity Factor in Patients With Partial RAG Deficiency. *Frontiers in Immunology*. 2020;11. doi:https://doi.org/10.3389/fimmu.2020.574738

Godasi R, Bollu PC. Primary Central Nervous System Vasculitis. PubMed. Published 2020. https://www.ncbi.nlm.nih.gov/books/NBK482476/

Agarwal A, Sharma J, Srivastava MVP, et al. Primary CNS vasculitis (PCNSV): a cohort study. *Scientific Reports*. 2022;12(1). doi:https://doi.org/10.1038/s41598-022-17869-7

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