Imaging in Rare Neuroimmune Disorders

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Disclosures

- No relevant financial relationships with any commercial interest.

Goal

- To review the role of imaging in the diagnosis of neuroimmunologic disorders and their mimics.

Outline

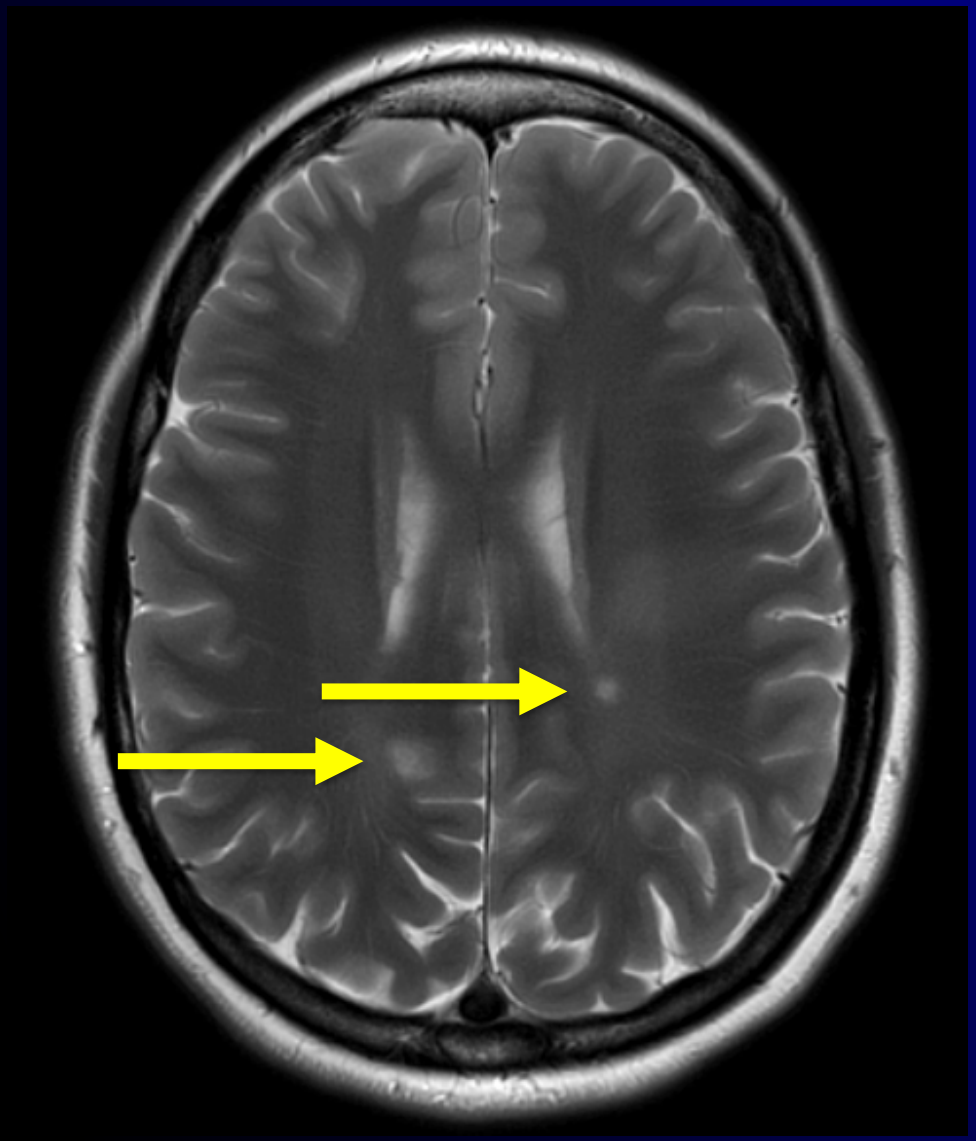
- Imaging background
  - MRI in MS
- Imaging in rare neuroimmune diseases
  - Post-radiation, NMOSD
  - AFM, spinal cord infarct
  - Neurosarcoid
  - Encephalitis (autoimmune, infectious)
  - Infection (Lyme disease, PML)
  - Vasculitis
- Discussion and Q/A

Imaging of autoimmune diseases

- Demyelination = loss of myelin sheath around axons
- Imaging = Ultrasound, X-ray, CT, MR, PET –MRI best detects demyelination
- MRI sequences detect:
  - Edema (T2, FLAIR) - bright
  - Blood brain barrier breakdown (T1 postcontrast) - bright
  - Ischemic stroke (DWI) - bright
  - Bleed (GRE) - dark

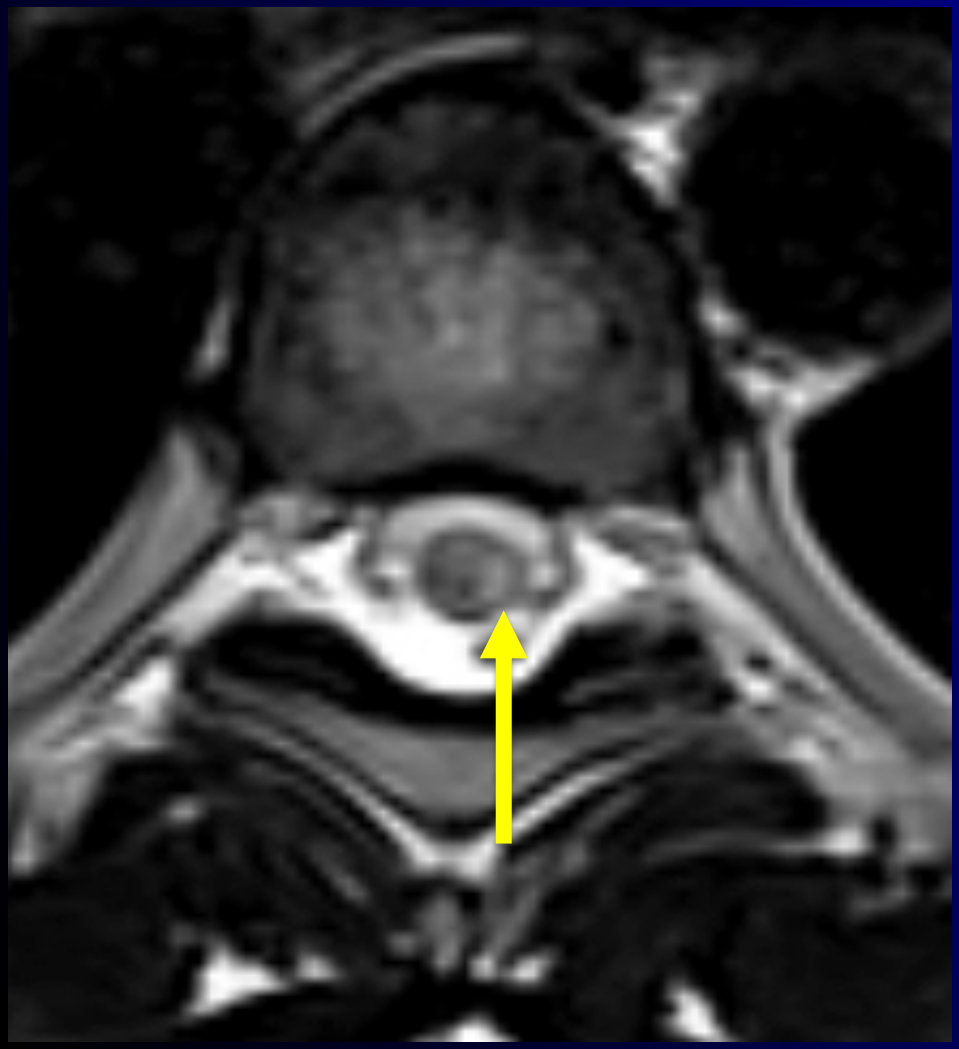
Multiple sclerosis

- Brain MRI
- Axial T2 weighted sequence



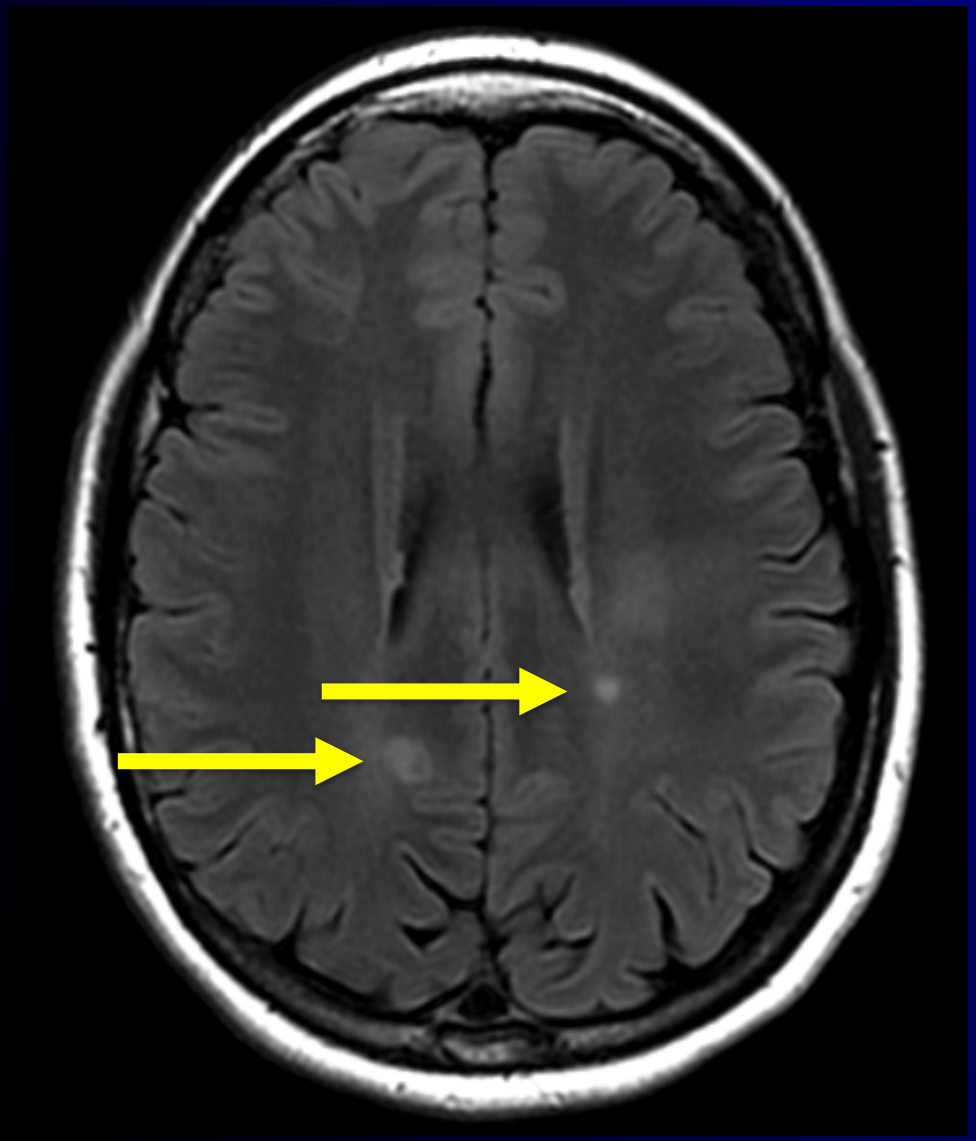
Multiple sclerosis

- Spine MRI
- Axial T2 weighted sequence



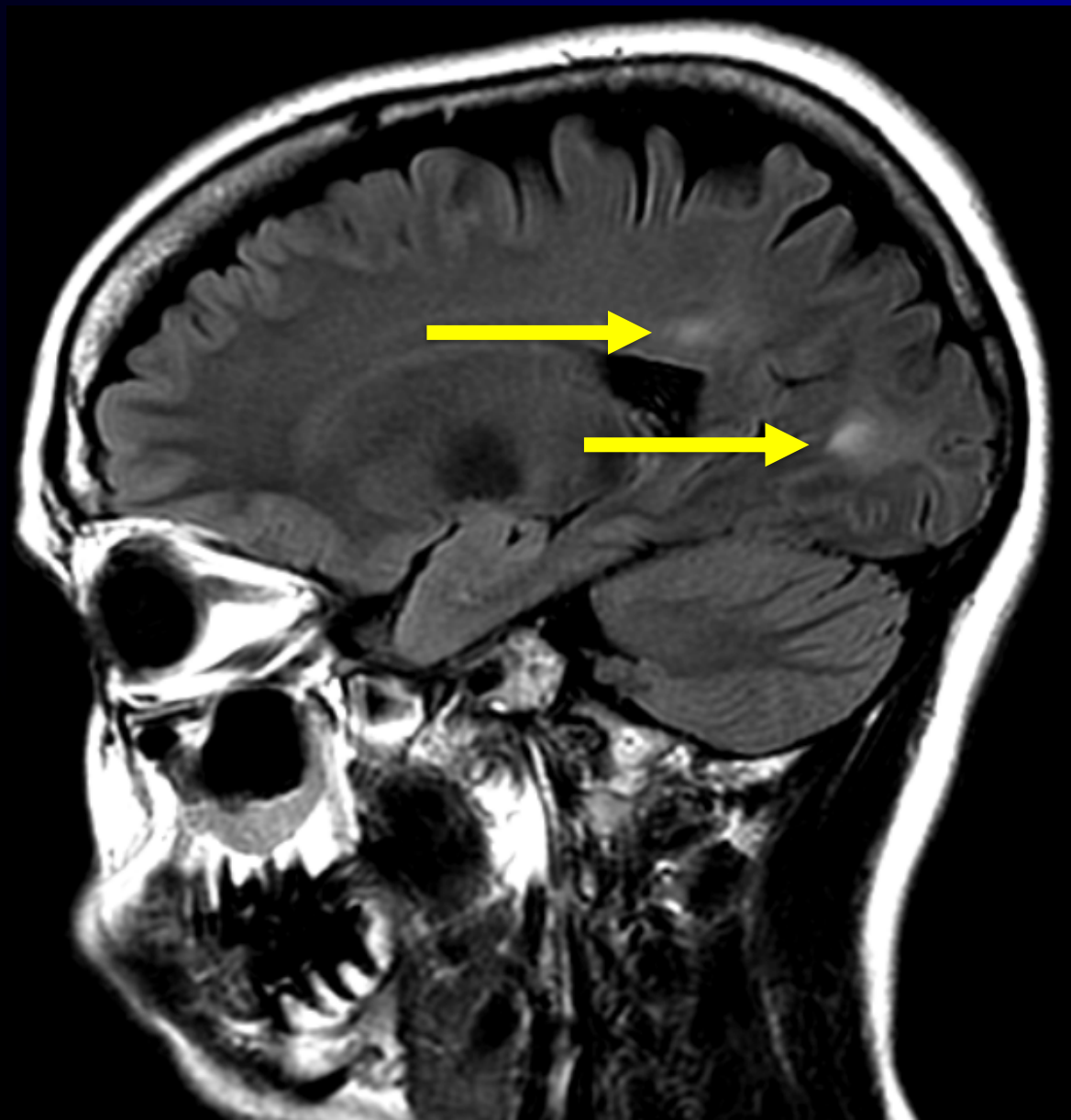
Multiple sclerosis

- Axial FLAIR sequence (T2-weighted)
- FLAIR = Fluid attenuated inversion recovery
- CSF is dark, lesions are bright and easier to see



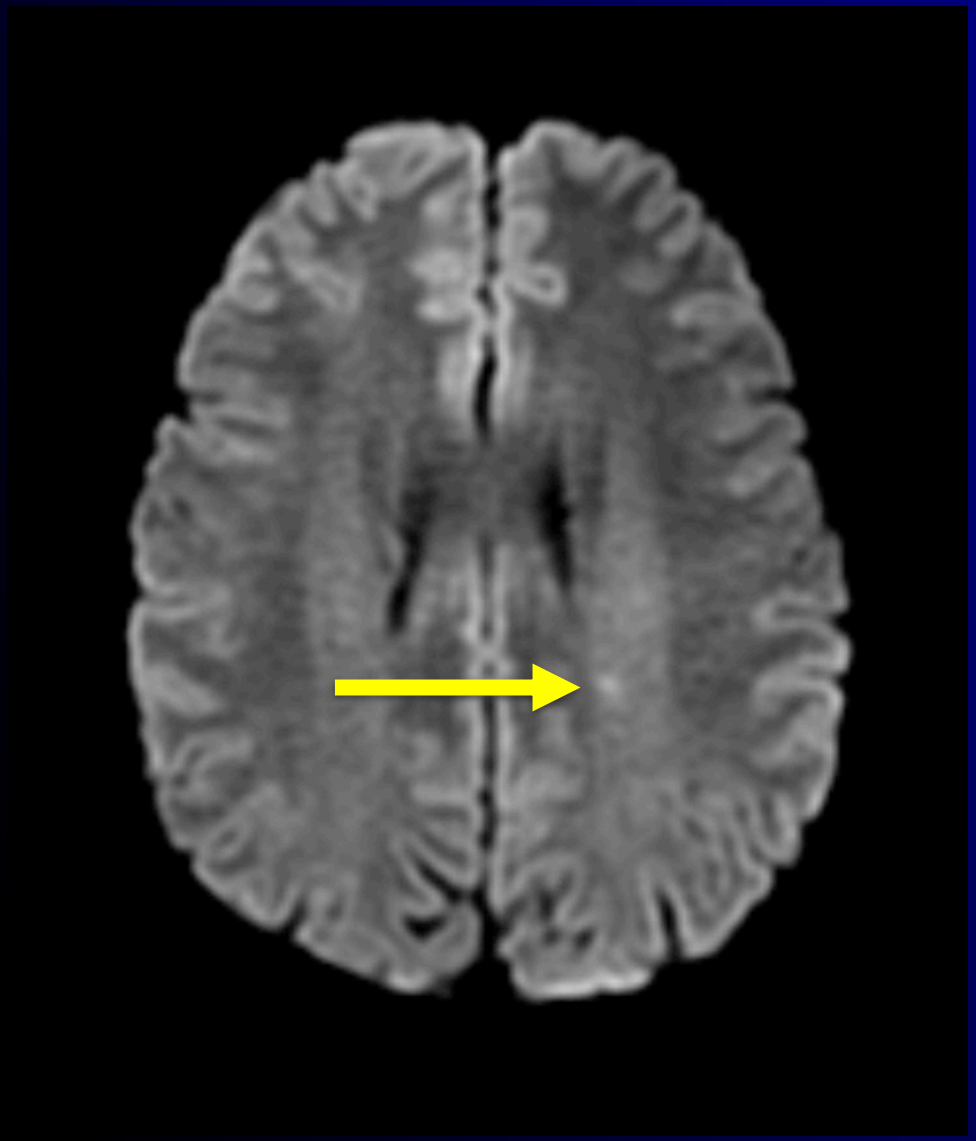
Multiple sclerosis

- Sagittal FLAIR sequence
- Detects pericallosal and callosomarginal lesions characteristic of demyelination in MS



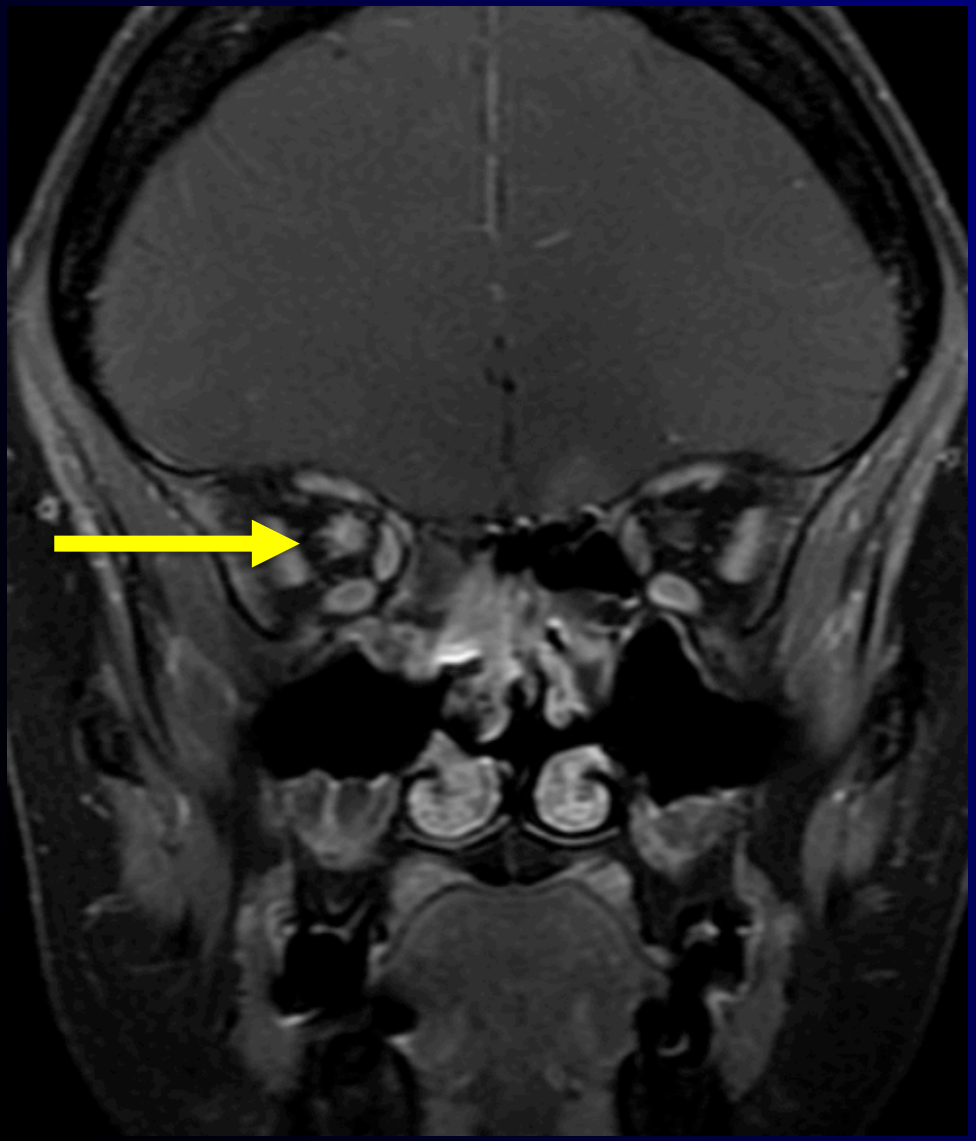
Multiple sclerosis

- Axial DWI sequence
- DWI = Diffusion weighted imaging
- DWI bright signal in cytotoxic edema from ischemic stroke, but also in active demyelination



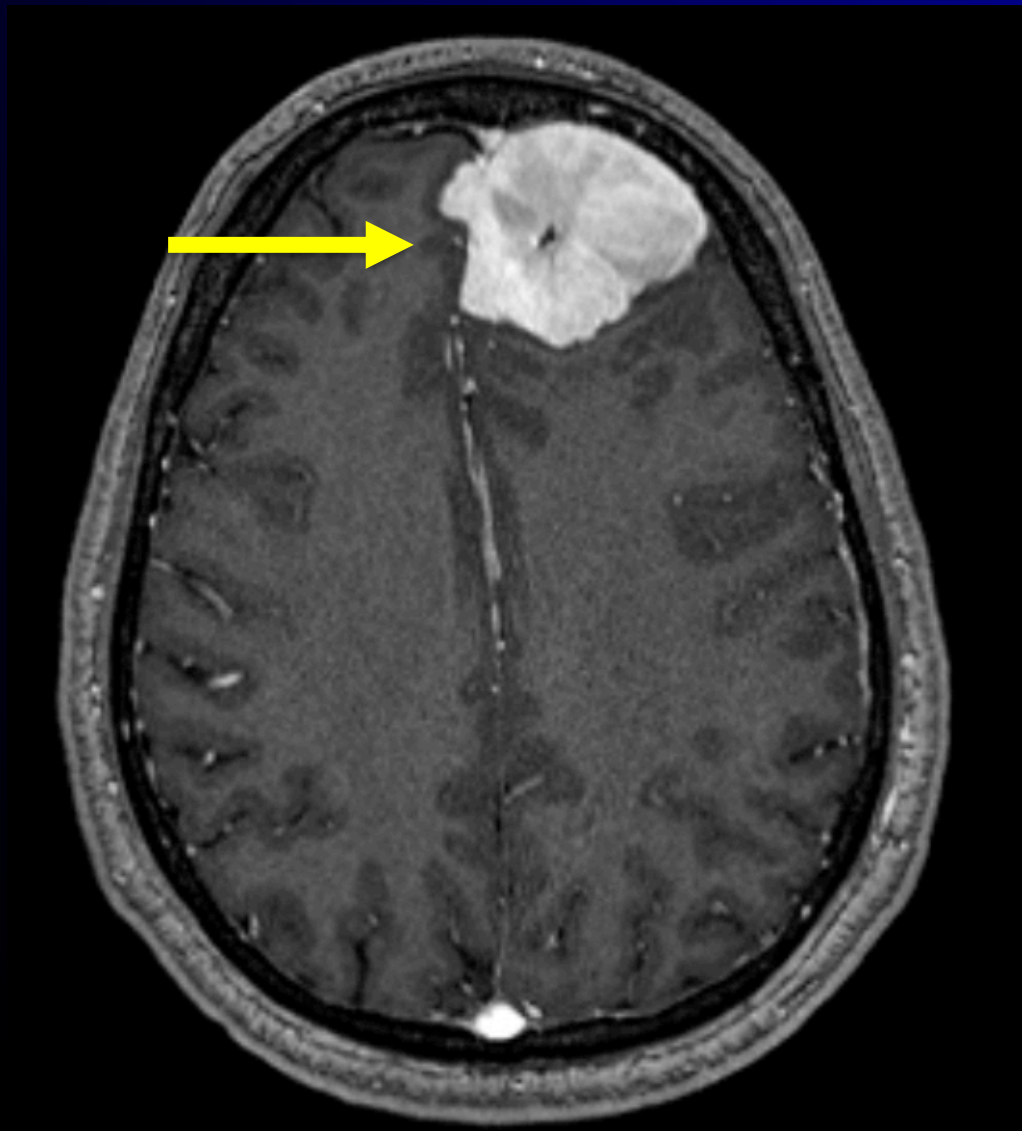
Multiple sclerosis

- Coronal T1 weighted postcontrast sequence
- Detect actively demyelinating lesions
- Can be seen in brain, spinal cord, or optic nerves
- Compare pre vs. post



Post radiation demyelination

- Patient with prior meningioma
- Resected and radiated
- 1 year later developed new symptoms



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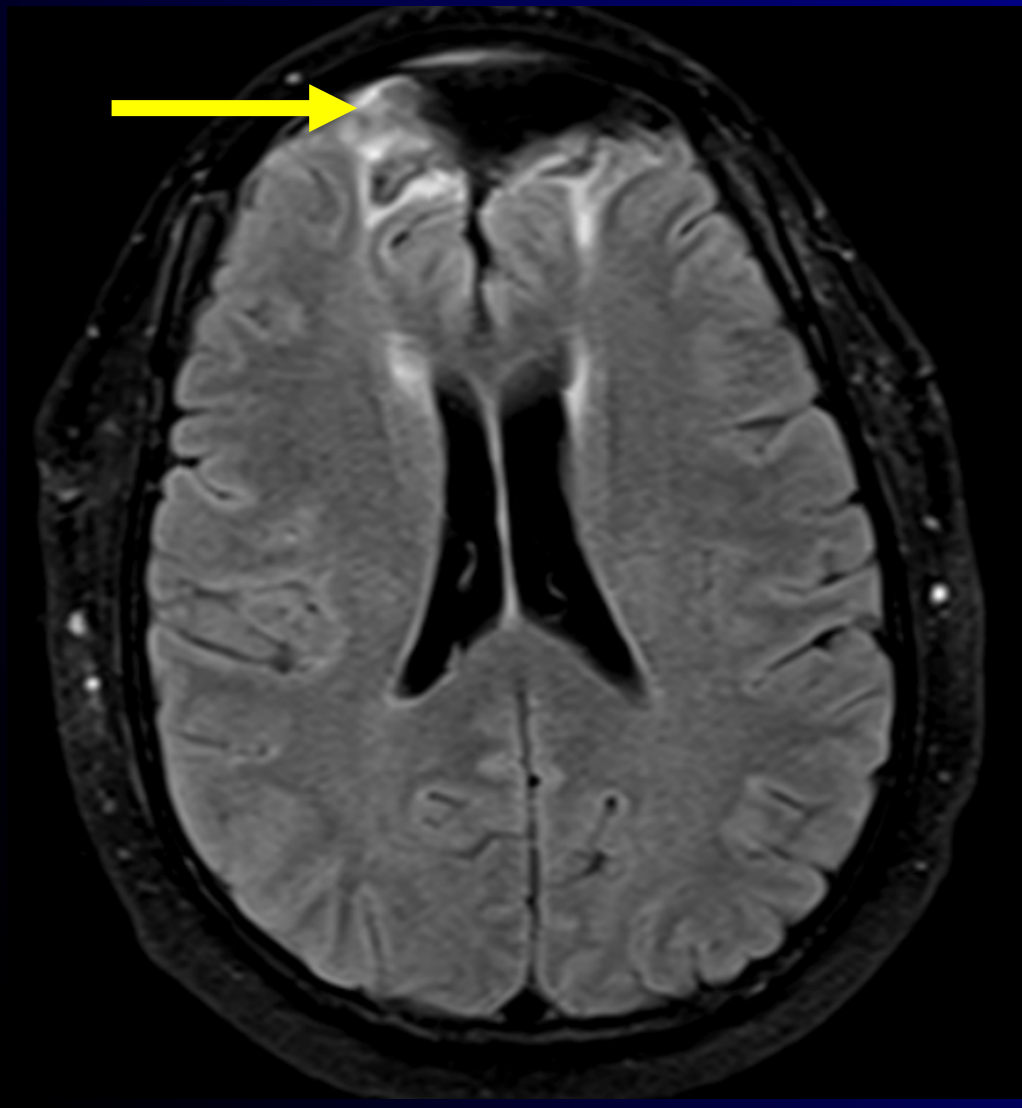

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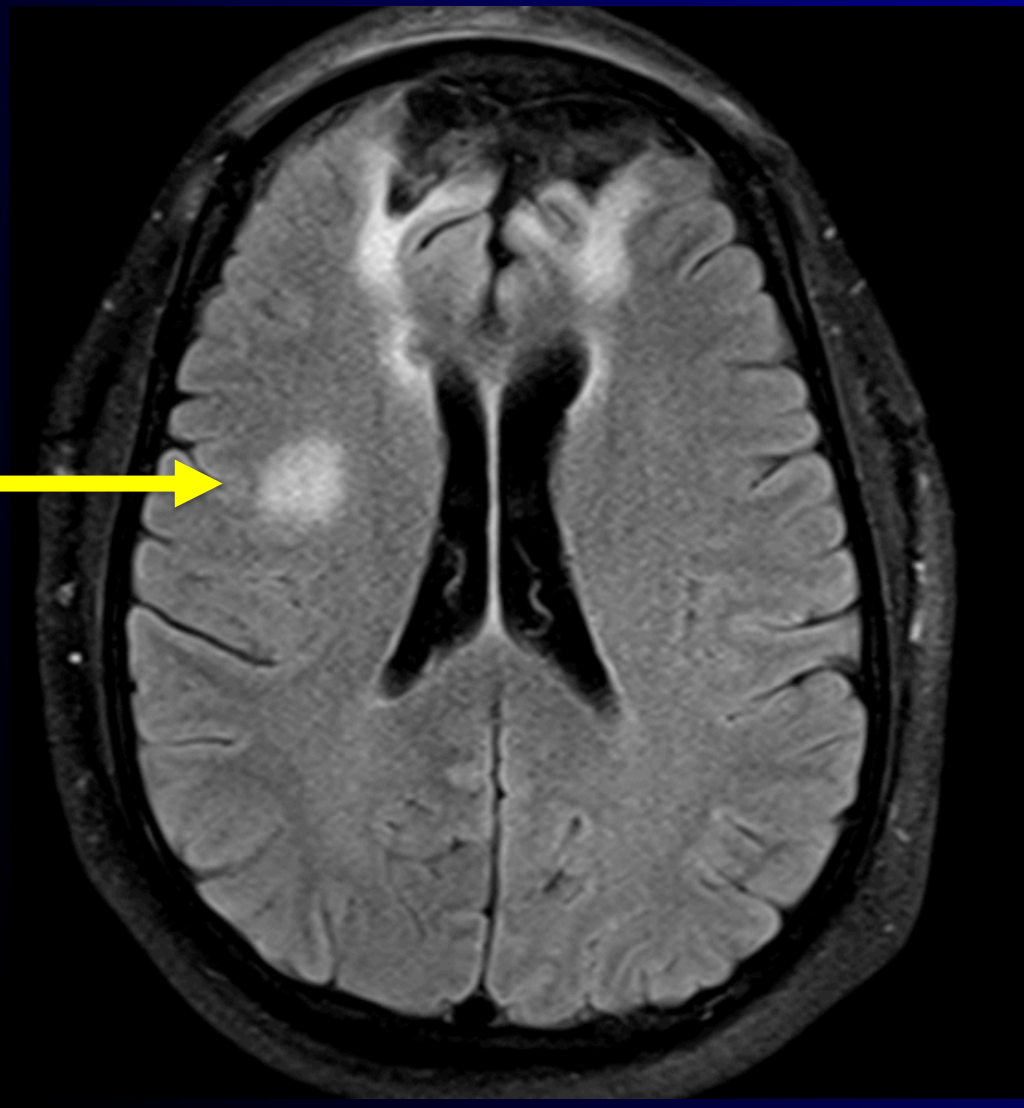

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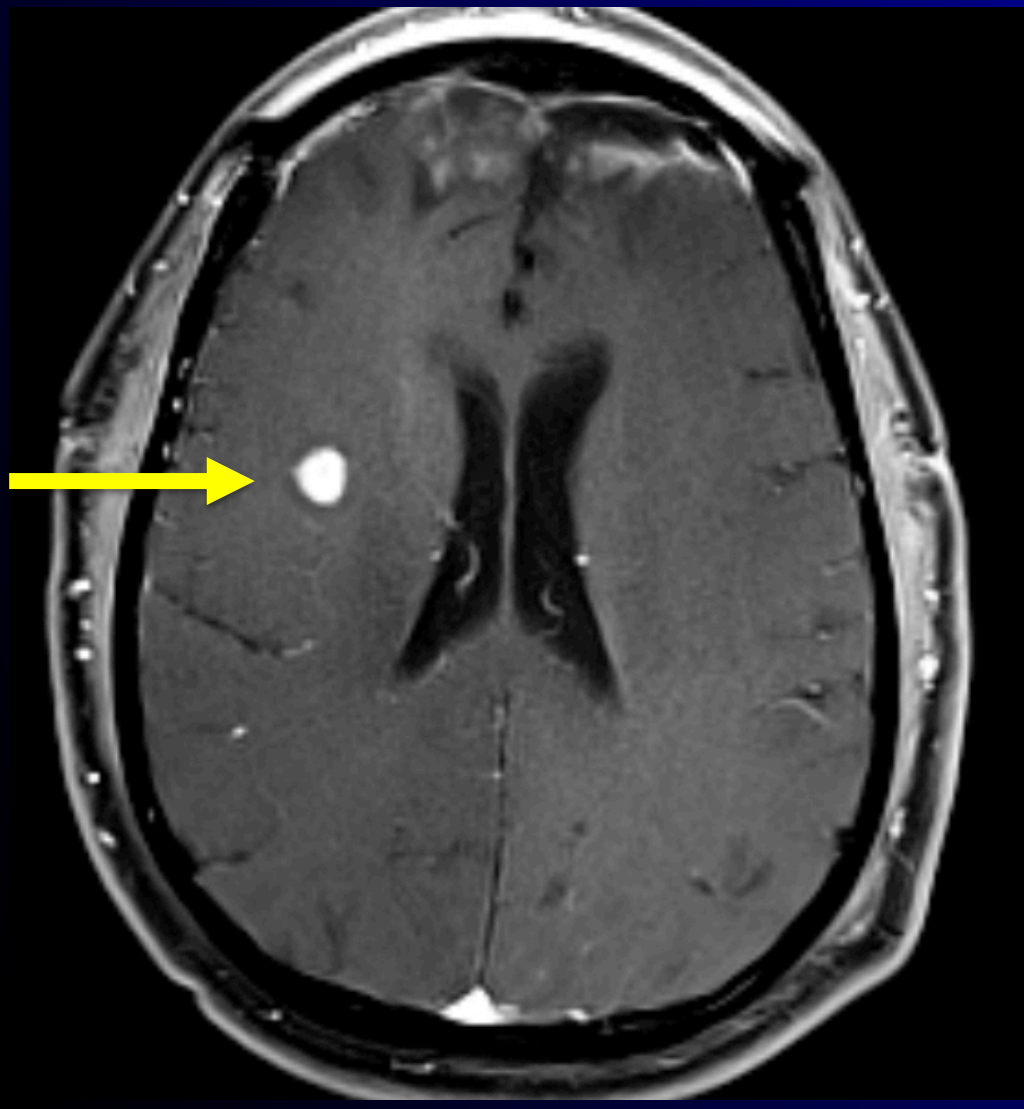

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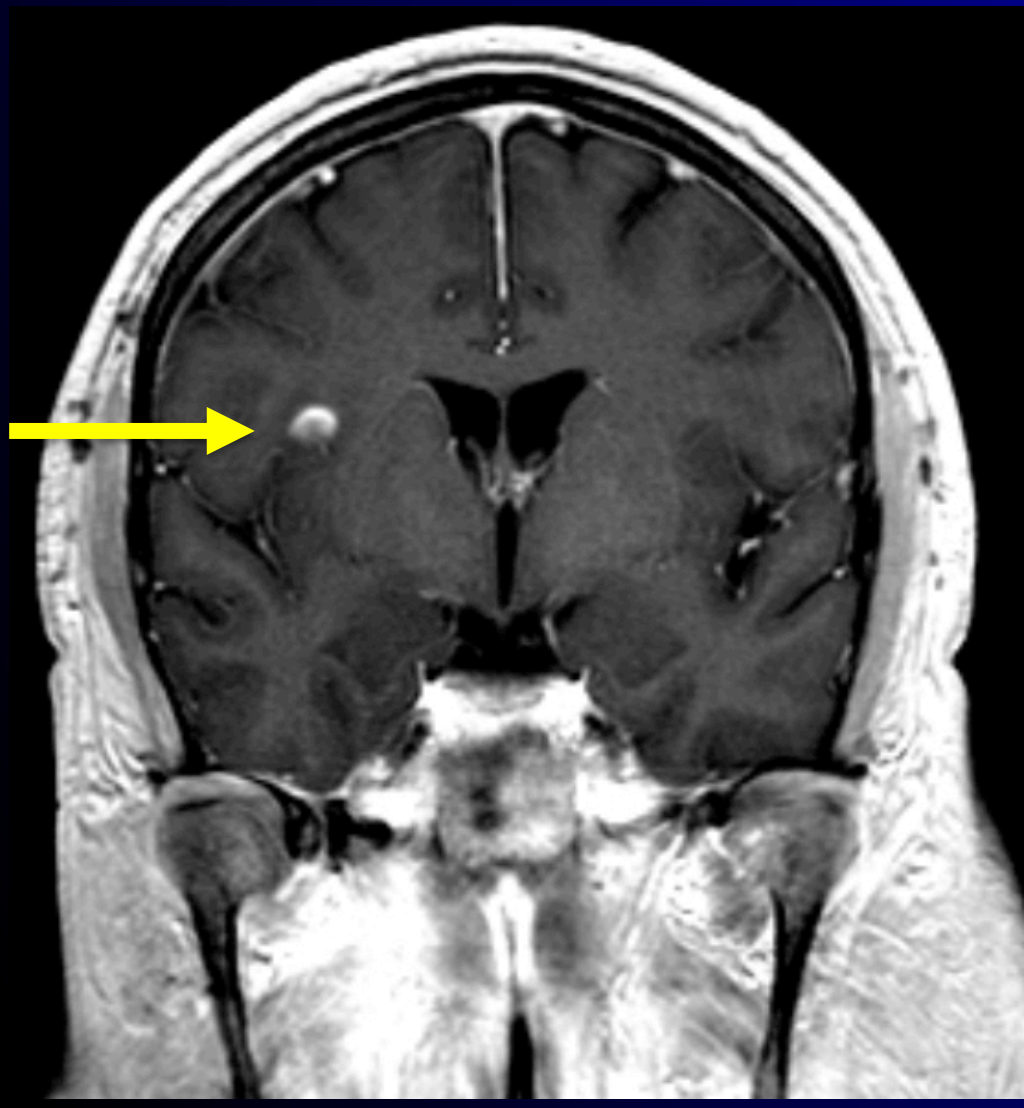
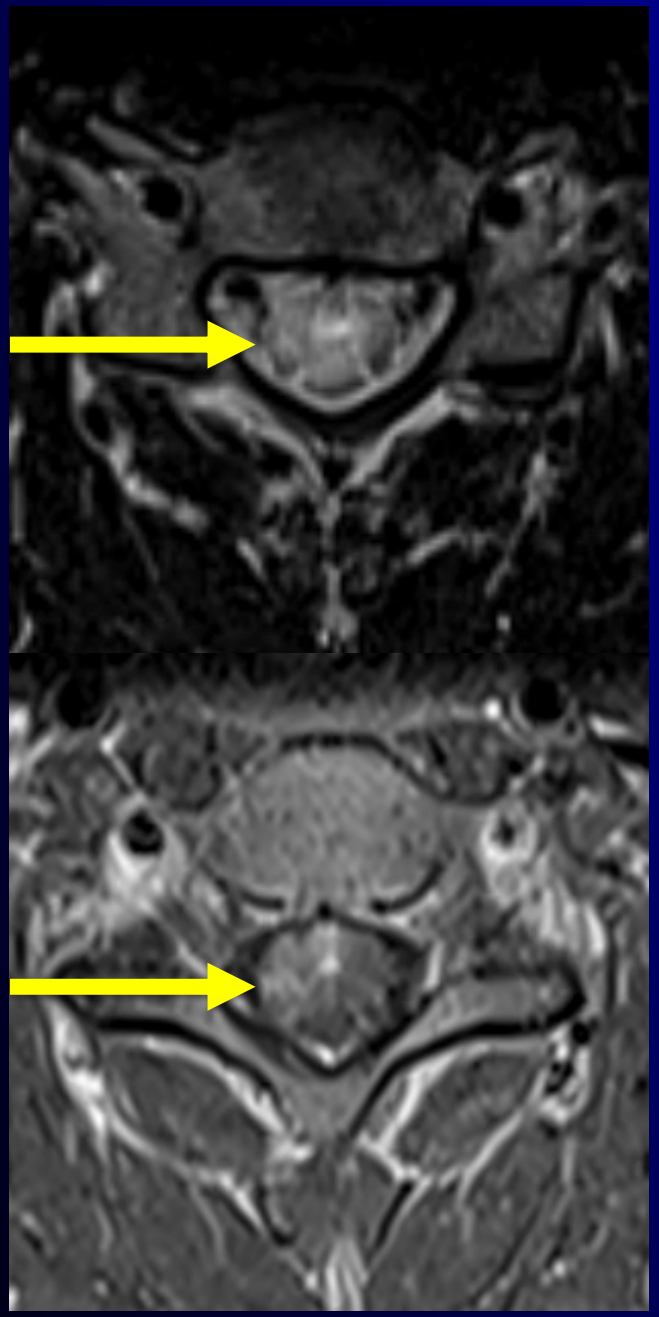


NMOSD

- NMOSD= neuromyelitis optica spectrum disease (Devic disease)
- Primarily affects optic nerves and spinal cord, with long segments of demyelination
- AQP4, MOG, or antibody negative



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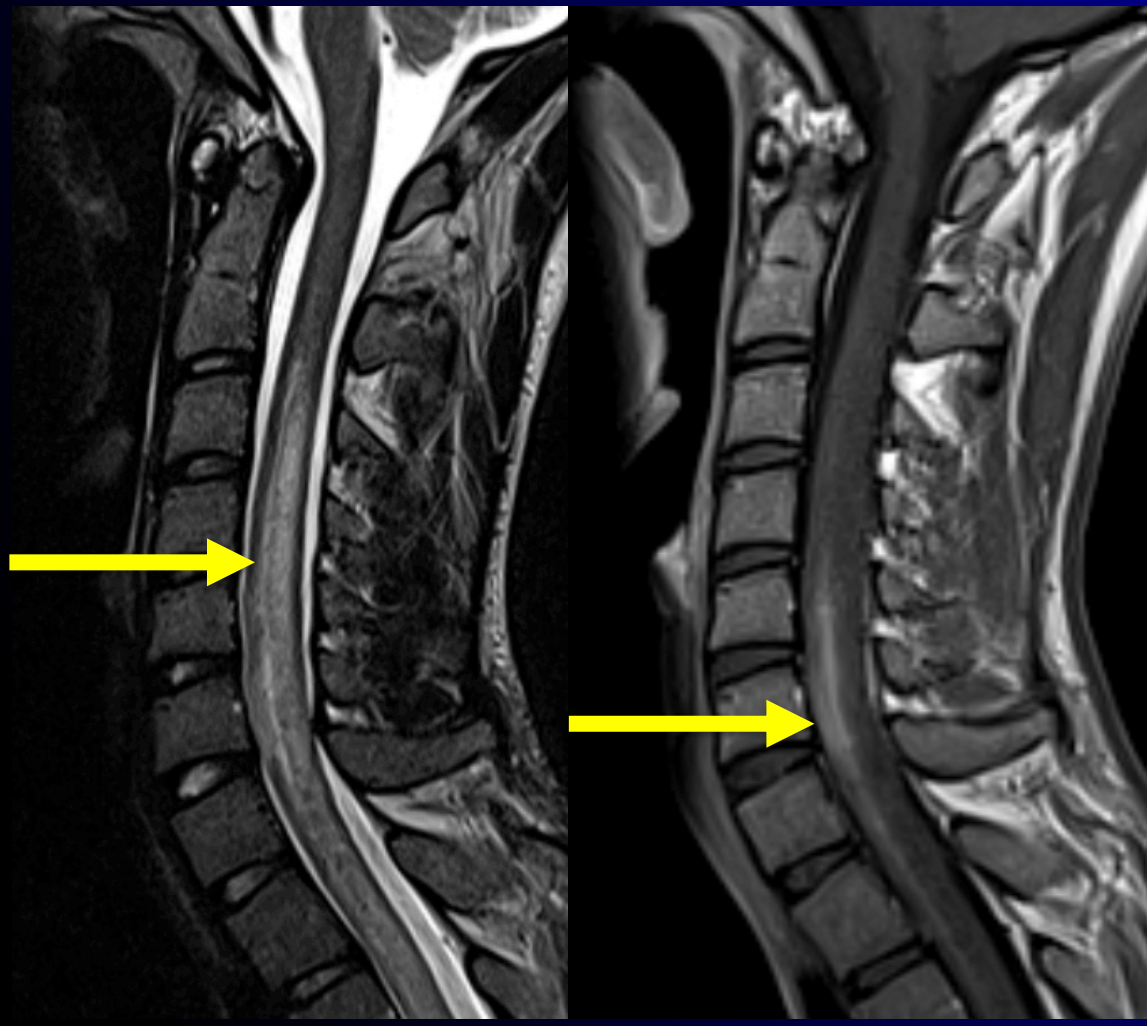

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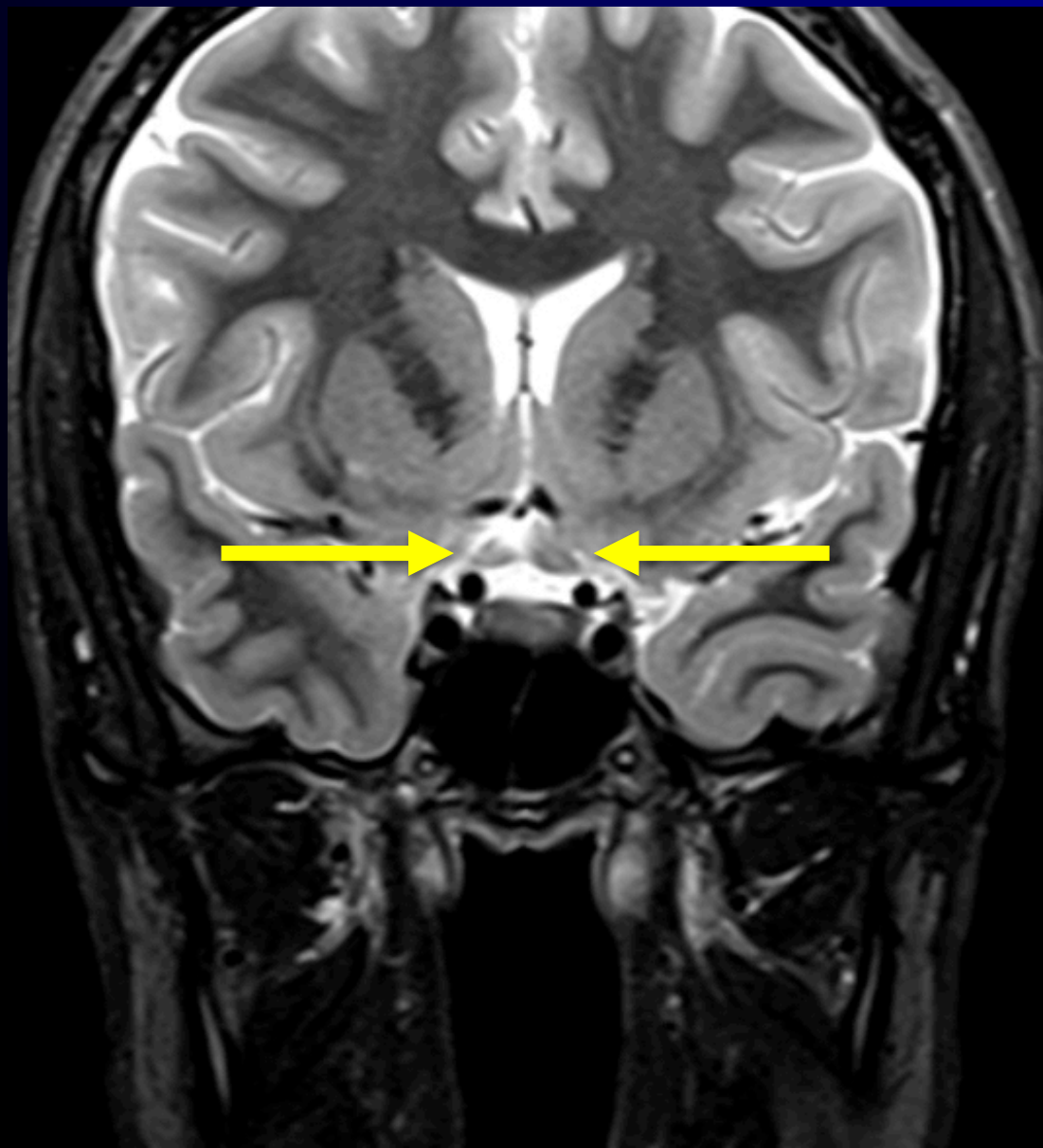
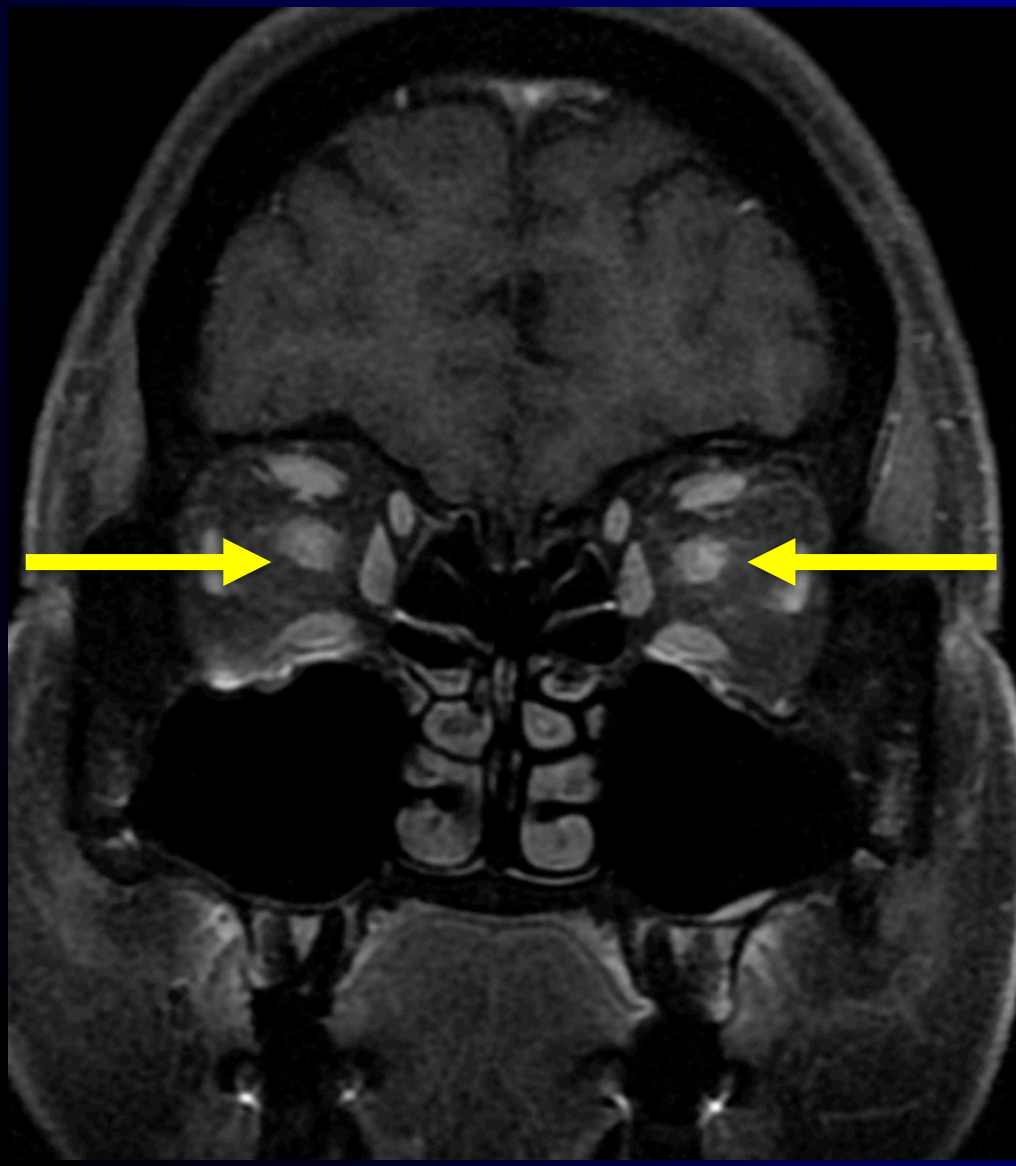


Anti-MOG NMOSD

- Anti-MOG accounts for ~25% of NMSOD
- Less % women affected compared to AQP4
- More often affects anterior optic nerves
- More often affects inferior/caudal cord



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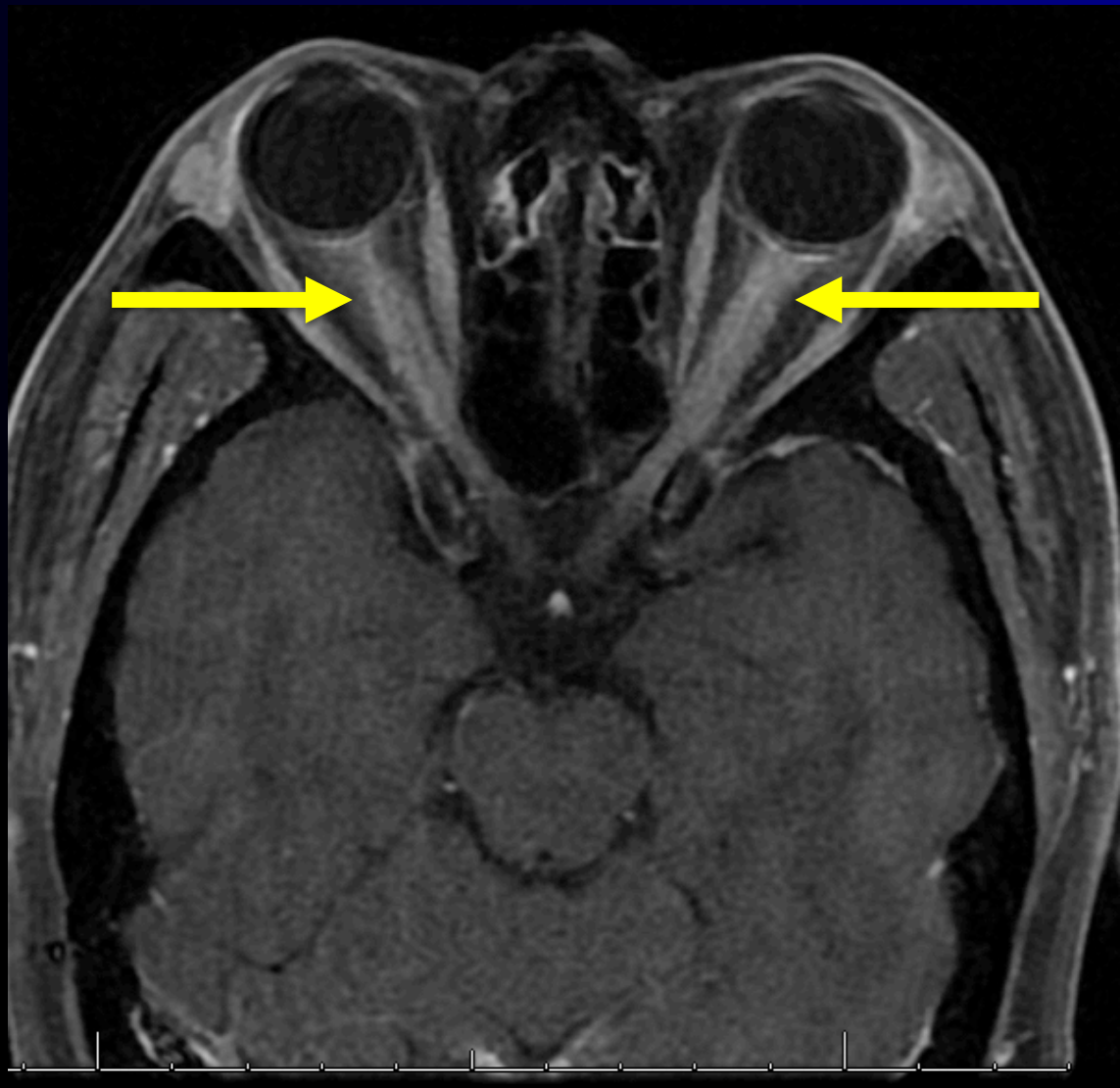

Acute flaccid myelitis (AFM)

- AFM = acute flaccid myelitis
- ‘Polio-like’ postviral myelitis affecting motor cells
- Associated with recent viral infection (respiratory viruses, enterovirus, etc)

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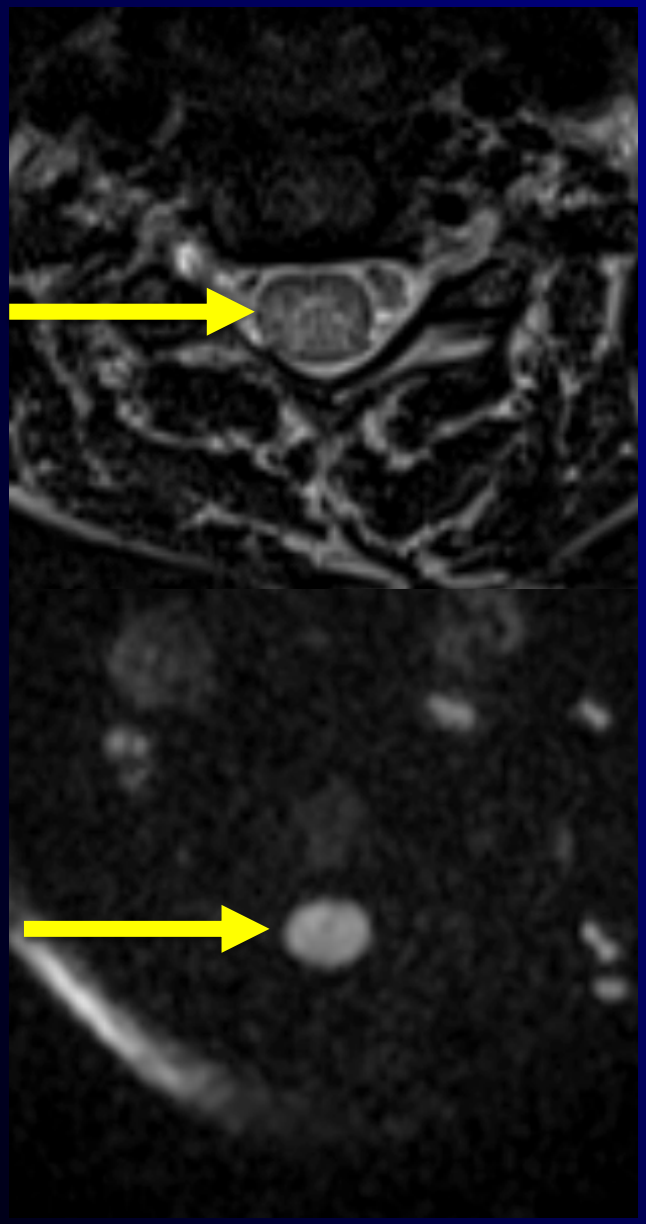
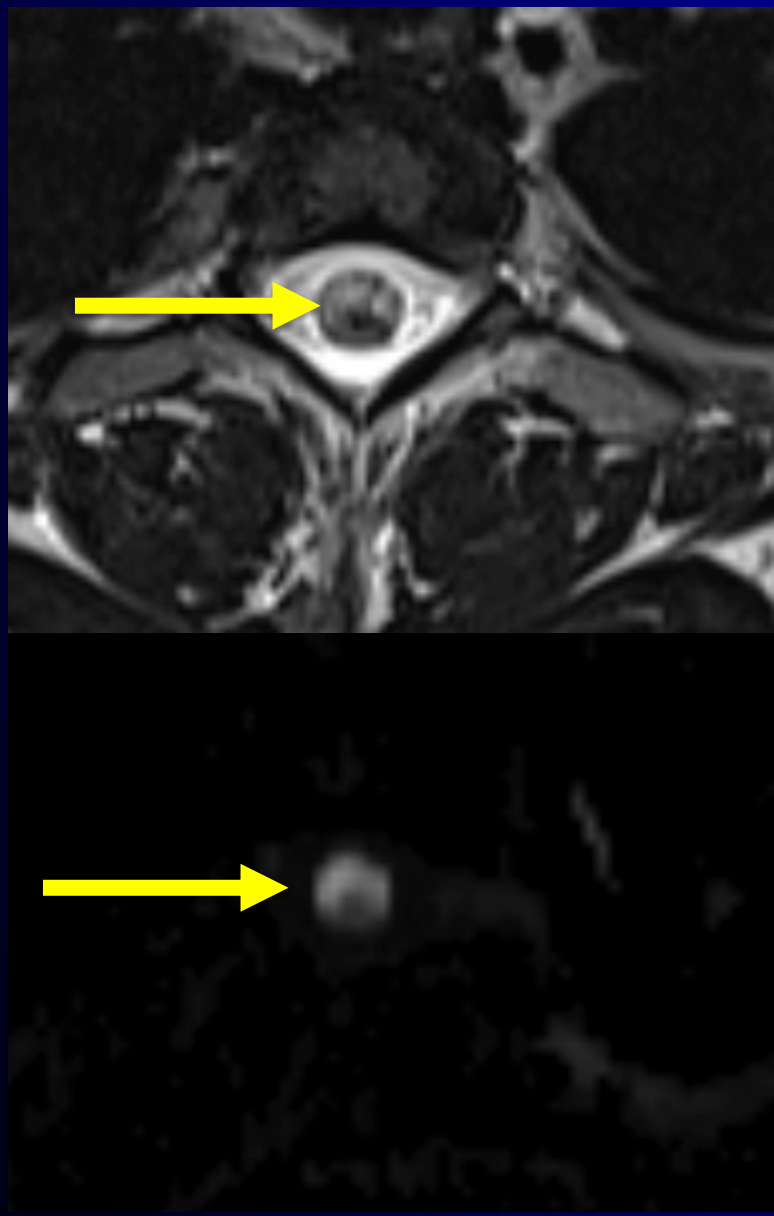


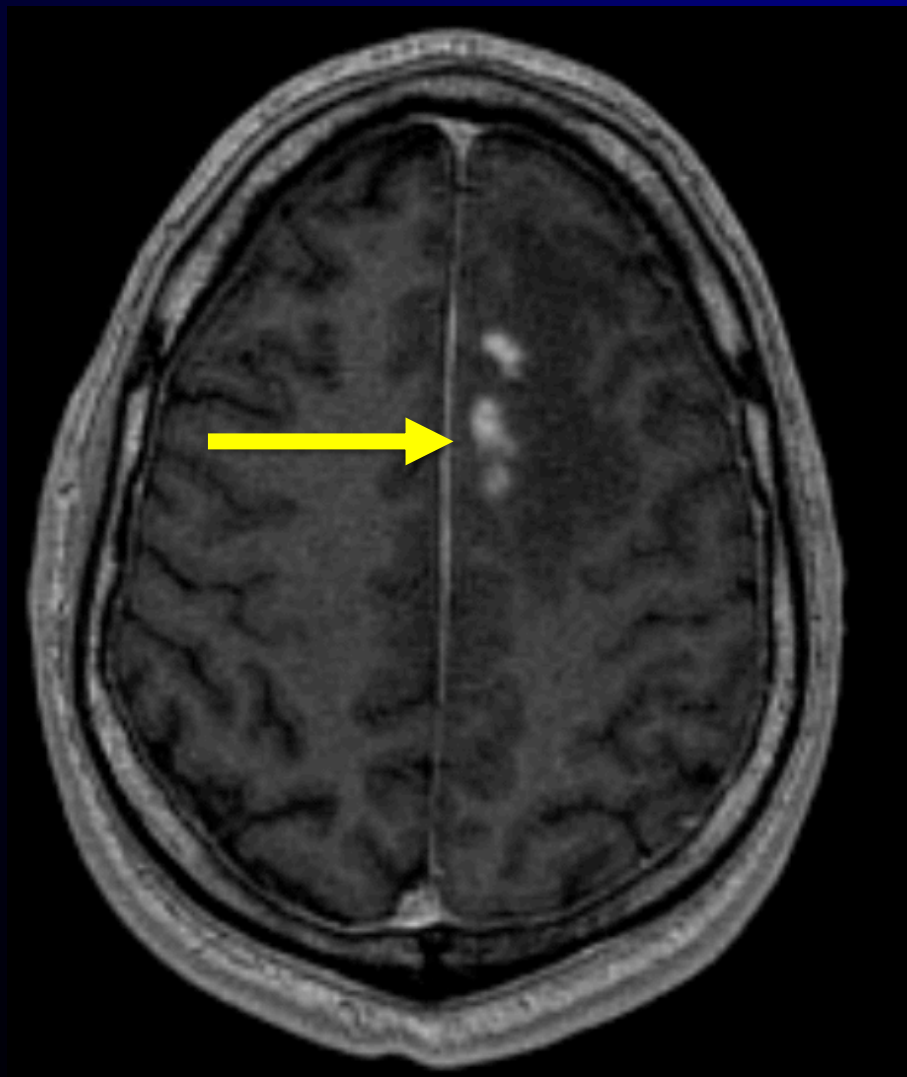
Cord infarct

- Spinal cord ischemic stroke
- Abrupt onset, no preceding viral illness
- Primarily motor neuron loss, similar to AFM
- Key is diffusion restriction on DWI MRI



Neurosarcoid

- Usually manifestation of systemic sarcoidosis, a granulomatous disease
- Variety of manifestations
- Lesions in brain, optic nerves, leptomeninges, cord, cranial nerves



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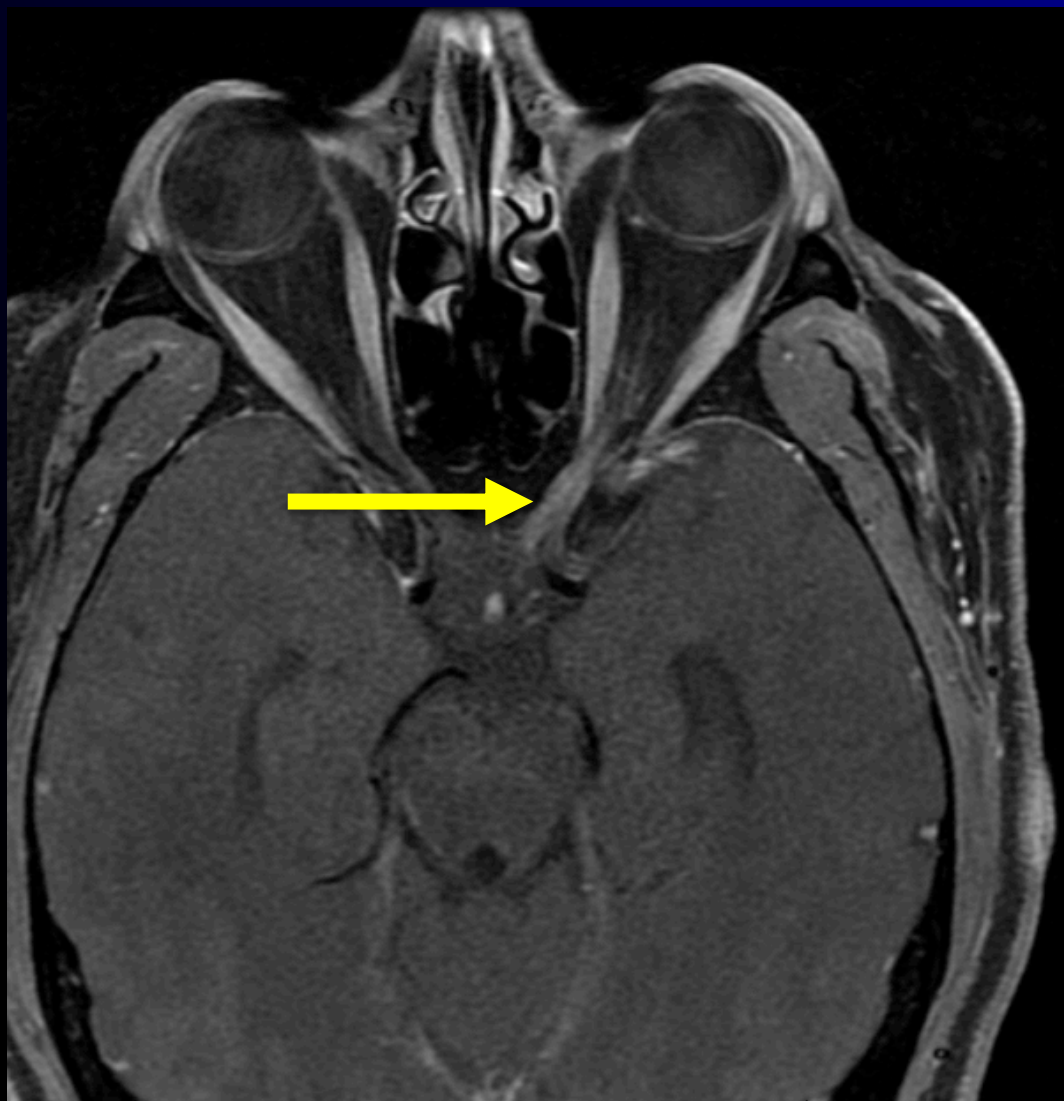

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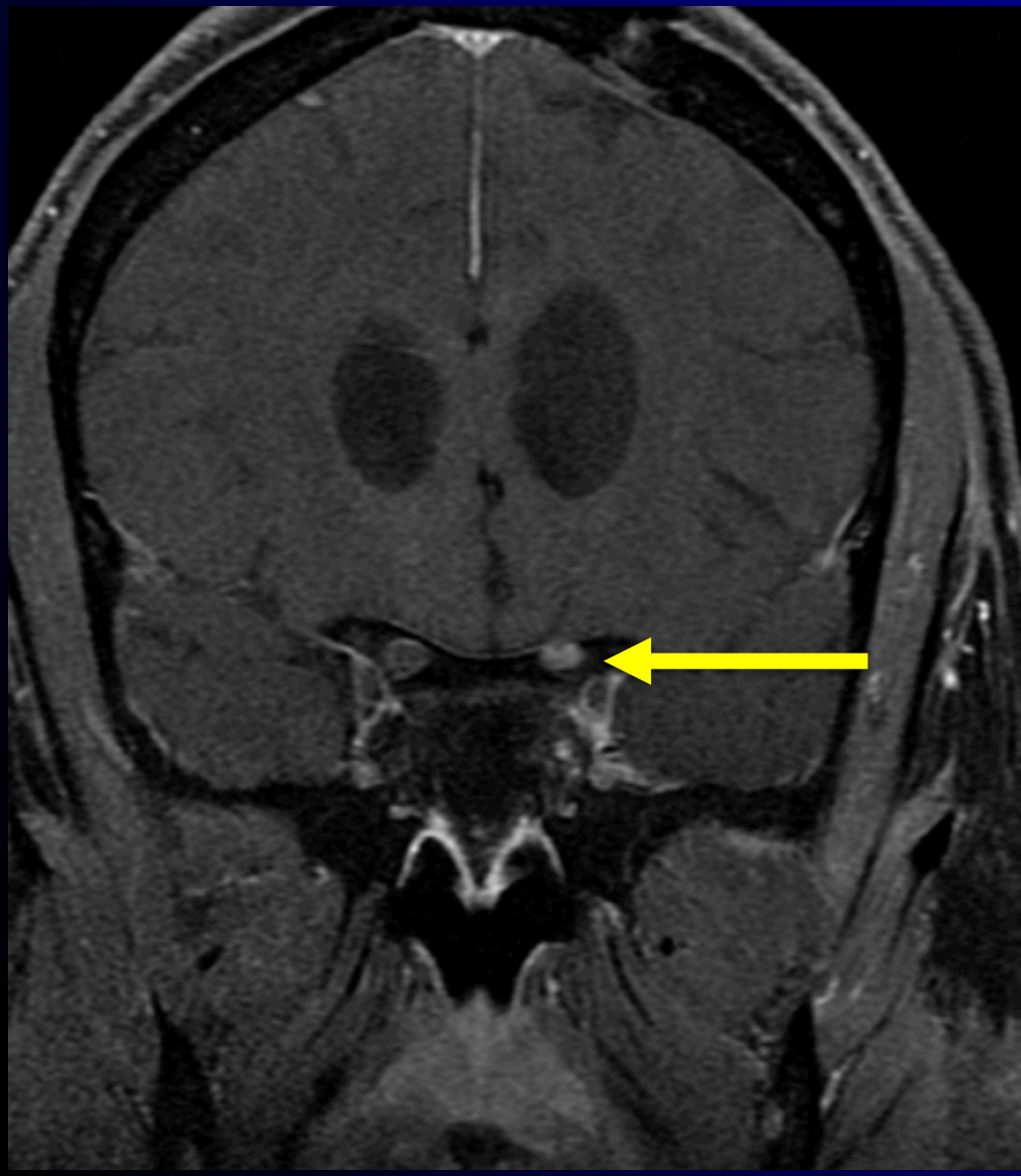

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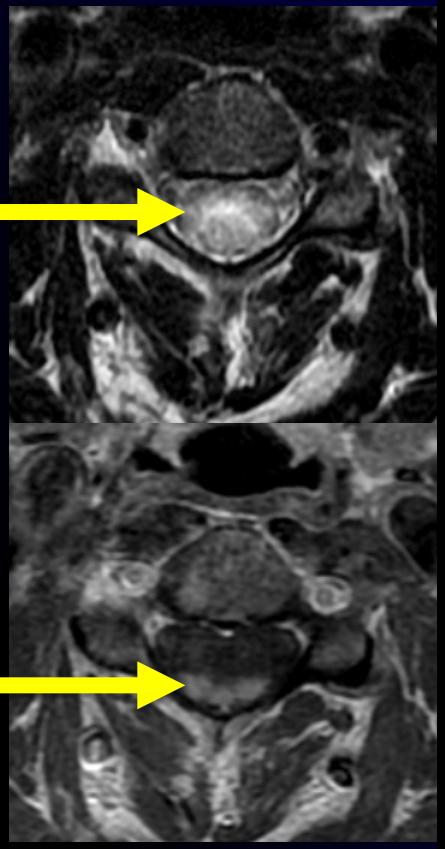
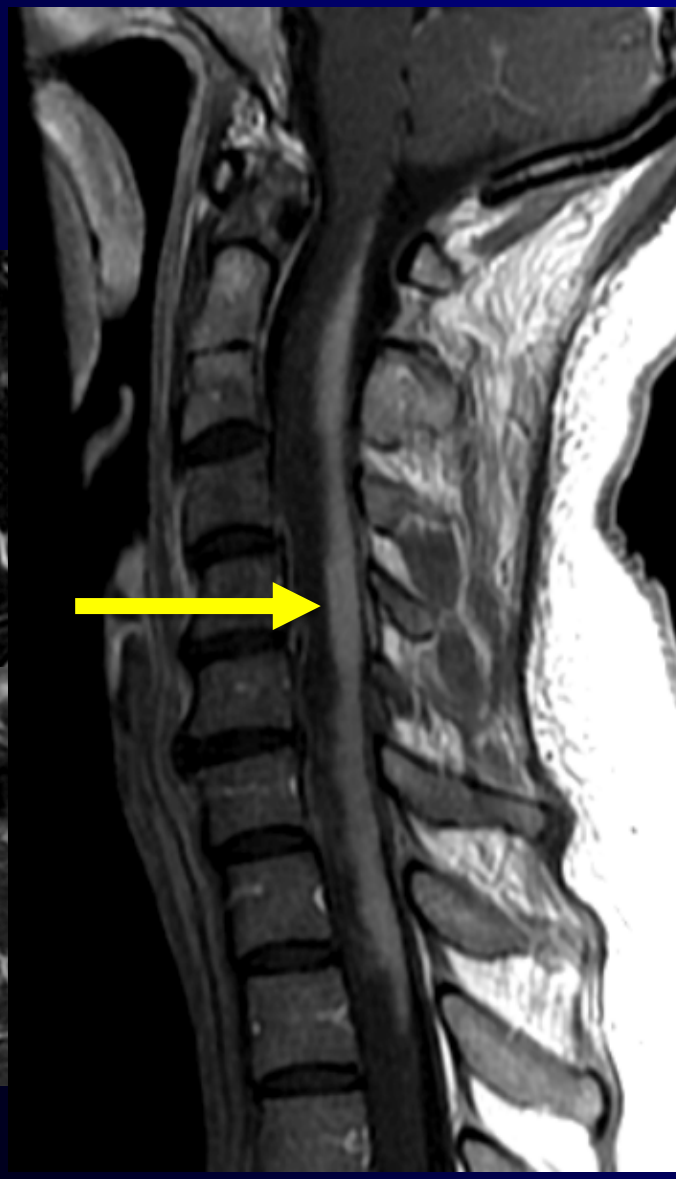
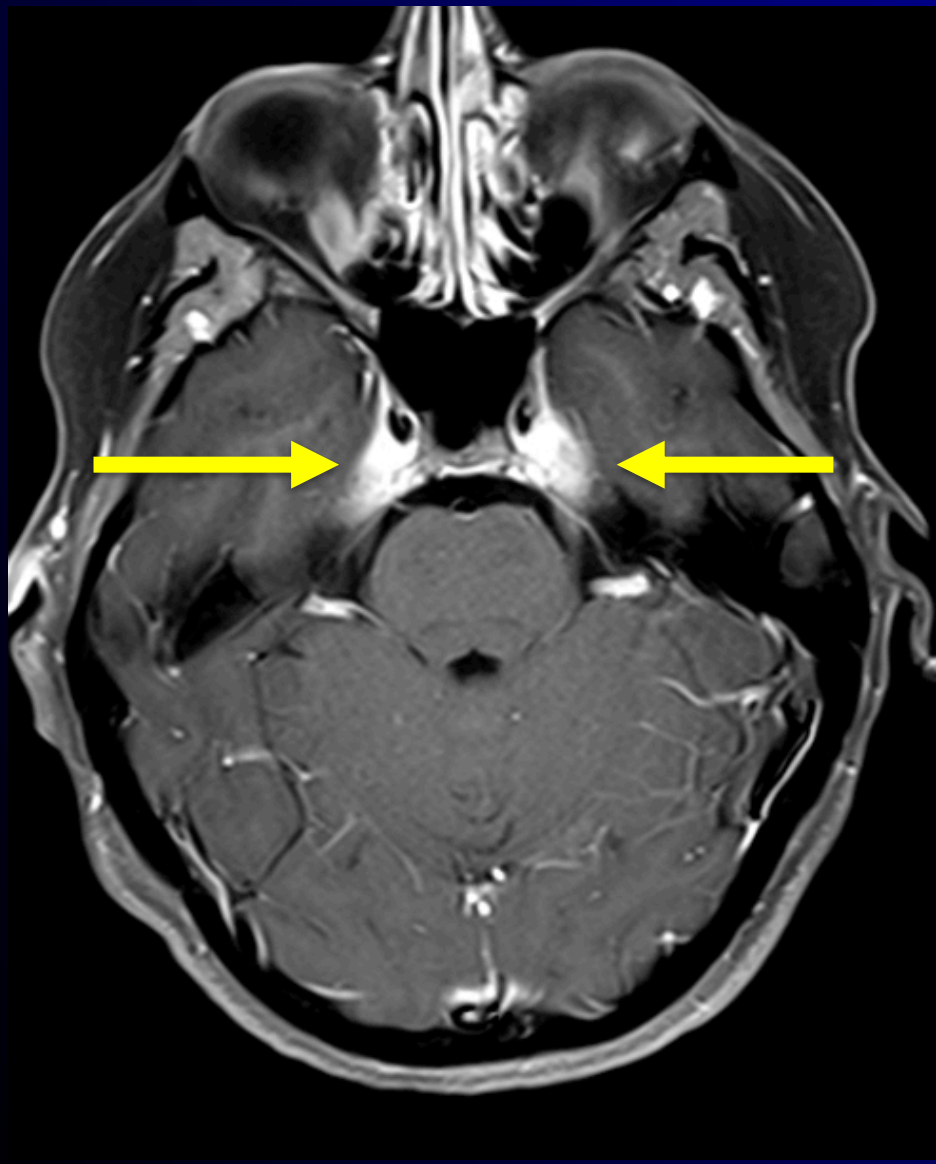




Lyme disease

- Can mimic many neuroimmune diseases
- Caused by Borrelia burgdorferi infection
- Affects white matter, nerve roots, leptomeninges



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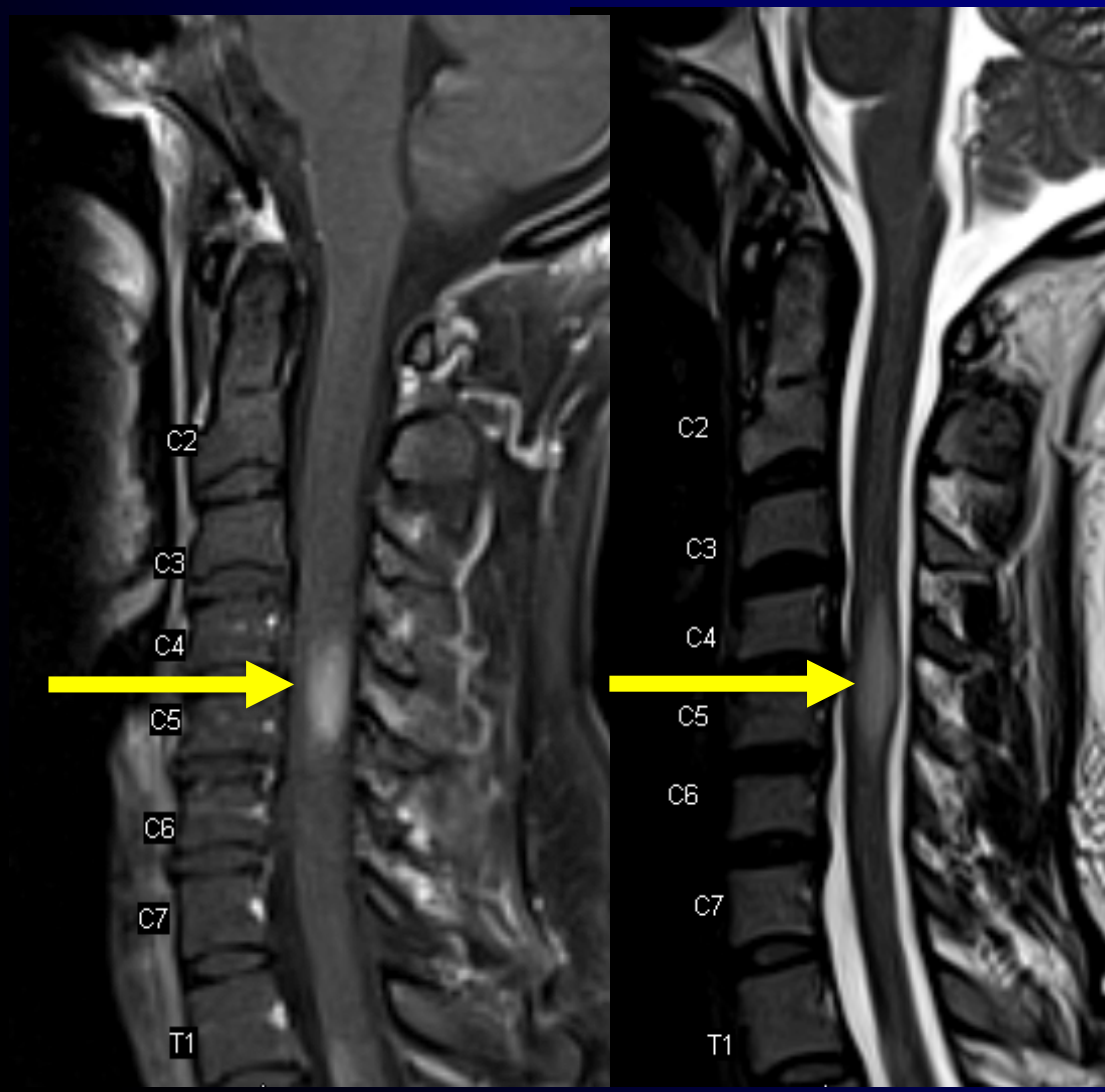

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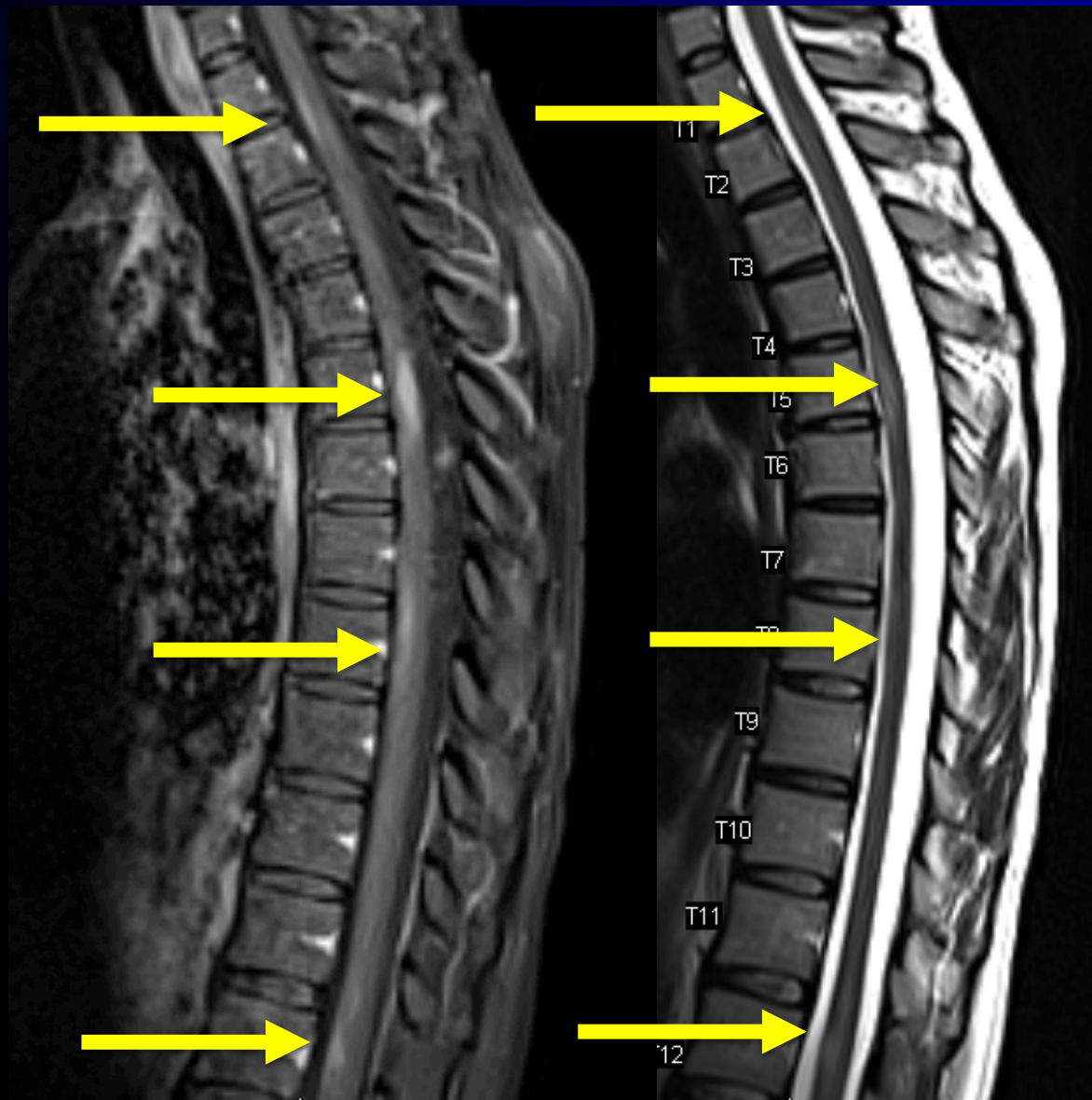

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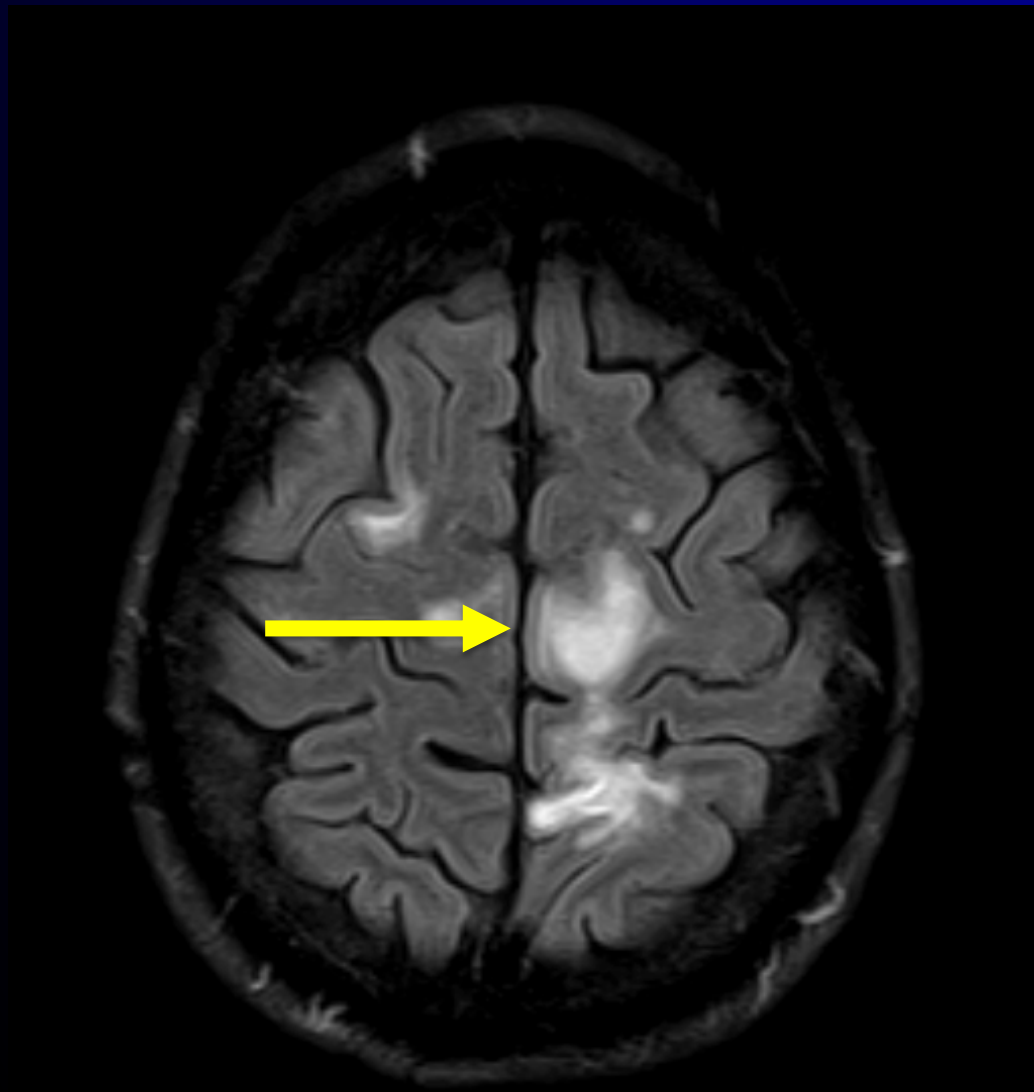
PML

- Progressive multifocal leukoencephalopathy
- Caused by the JC virus
- Infectious demyelination can occur with severe immunosuppression
- Rituximab (Rituxan) and natalizumab (Tysabri)



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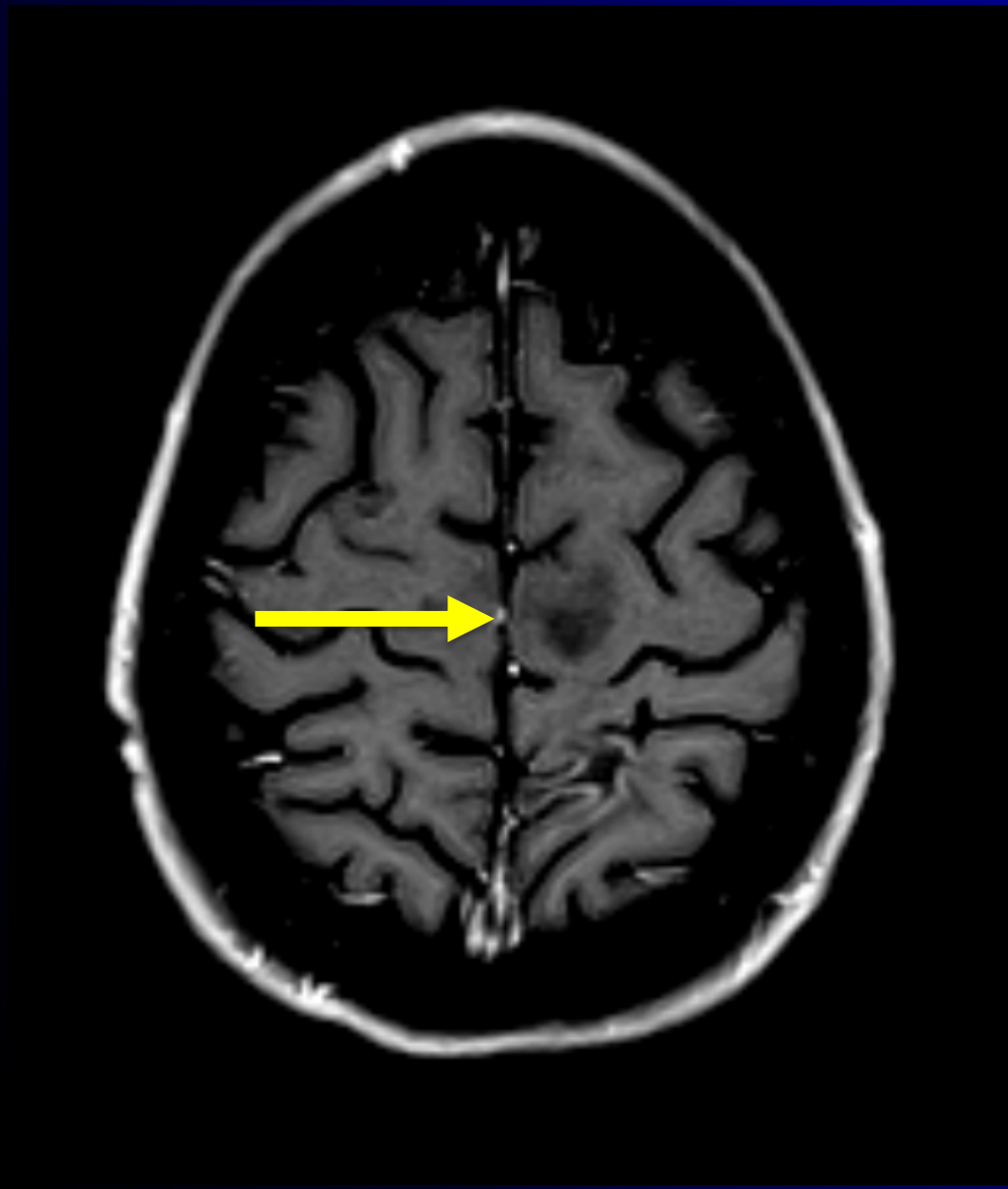

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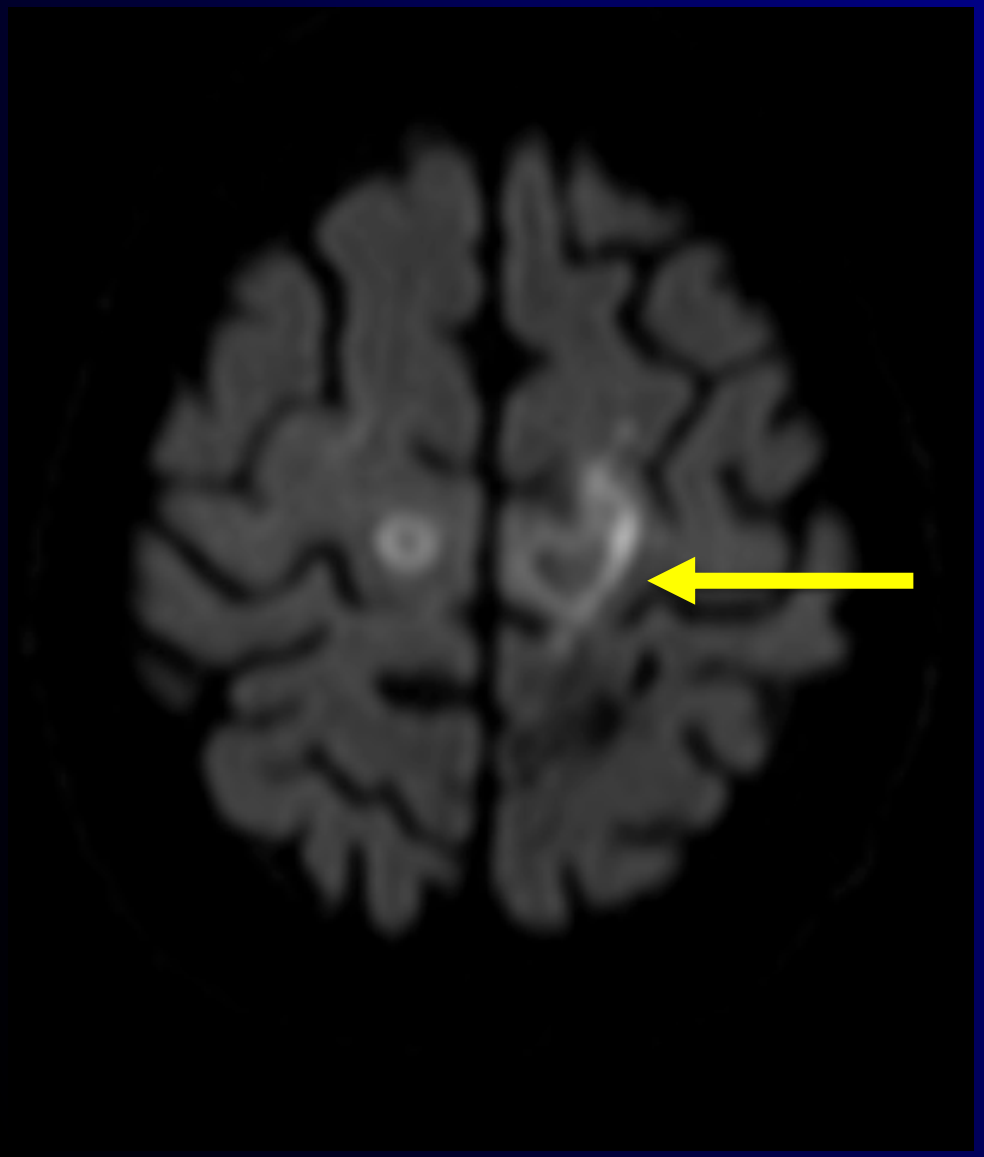

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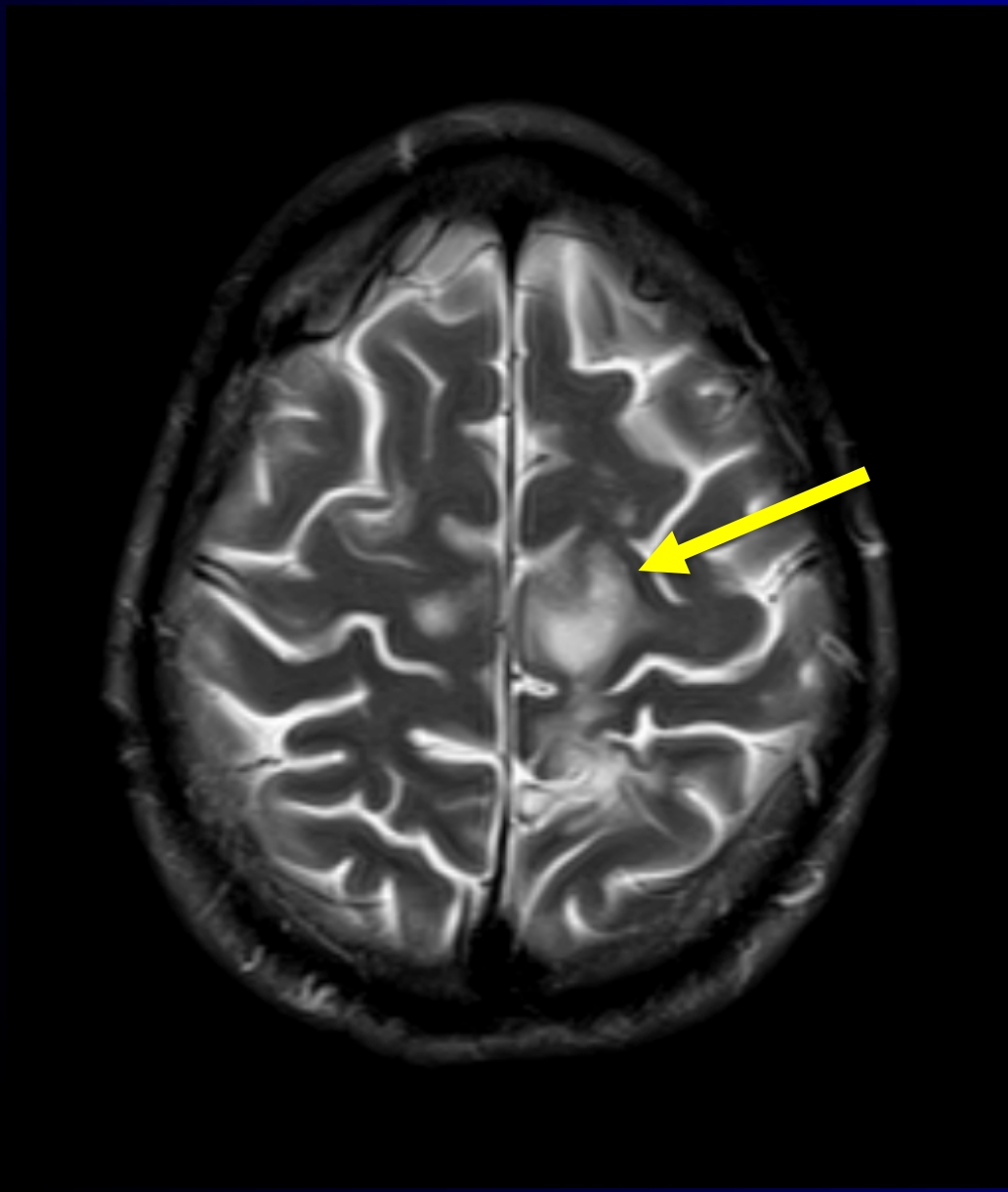
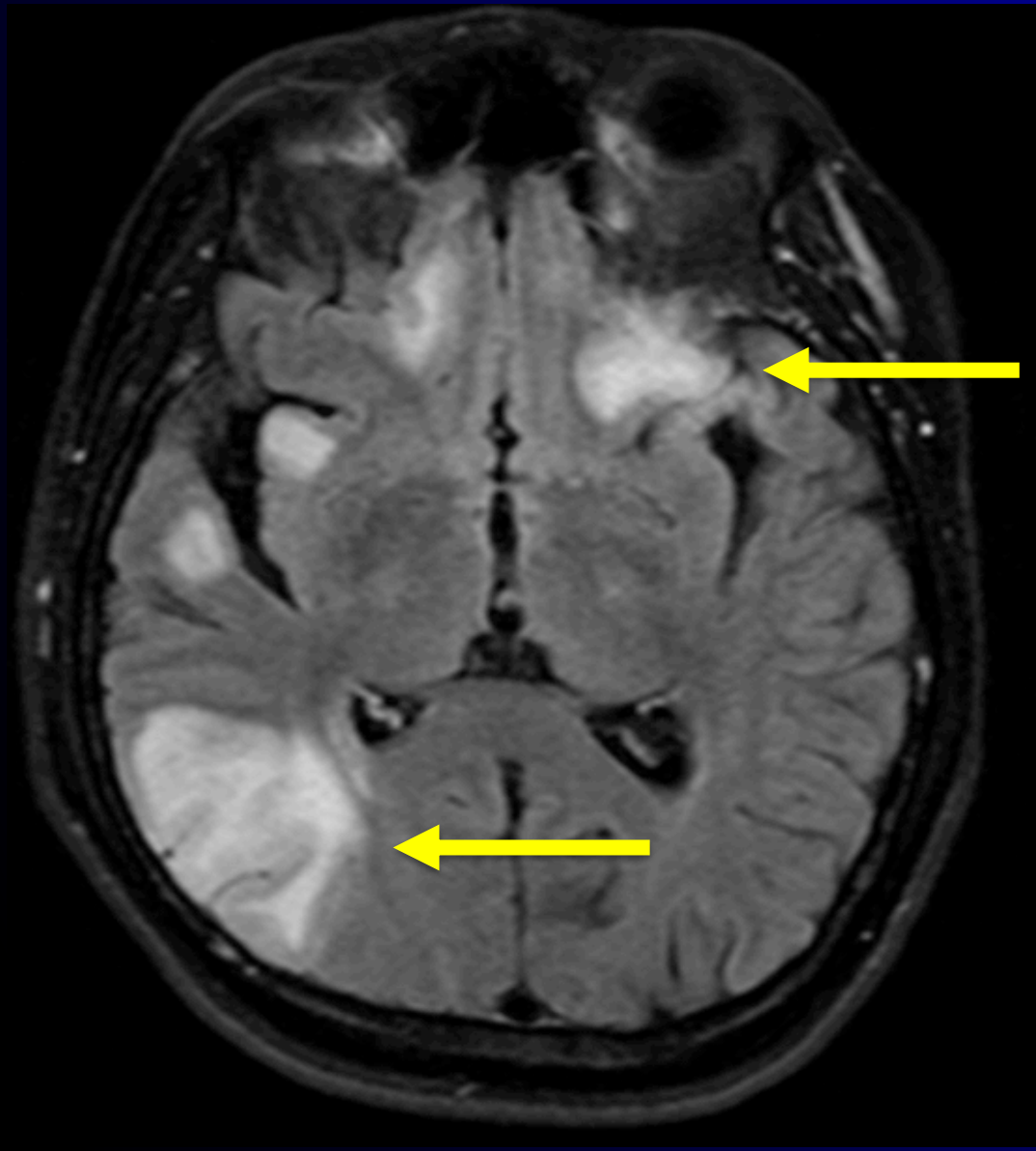


Encephalitis

- Encephalitis is inflammation of the brain
- Can be autoimmune (e.g. Anti-Gaba A Receptor)
- Can be infectious (e.g. HSV – herpes)



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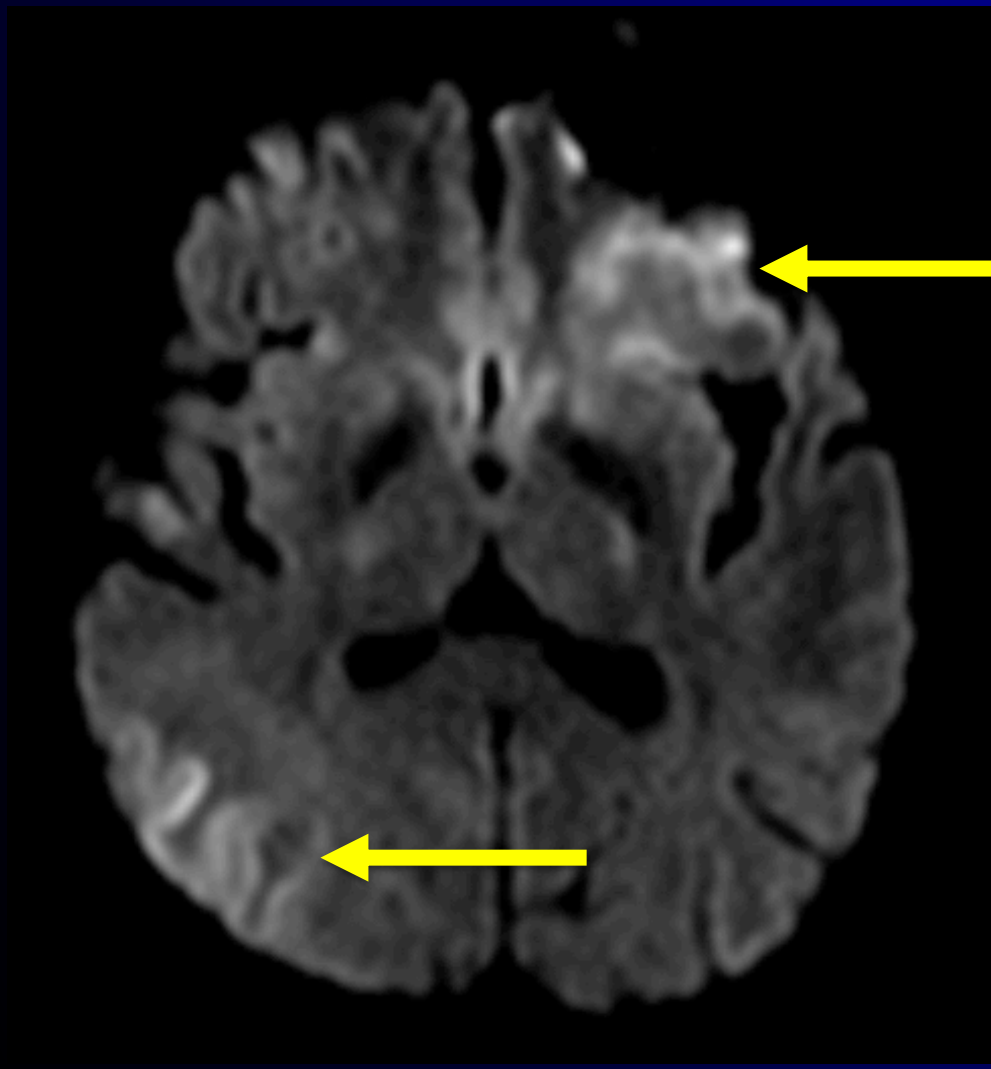

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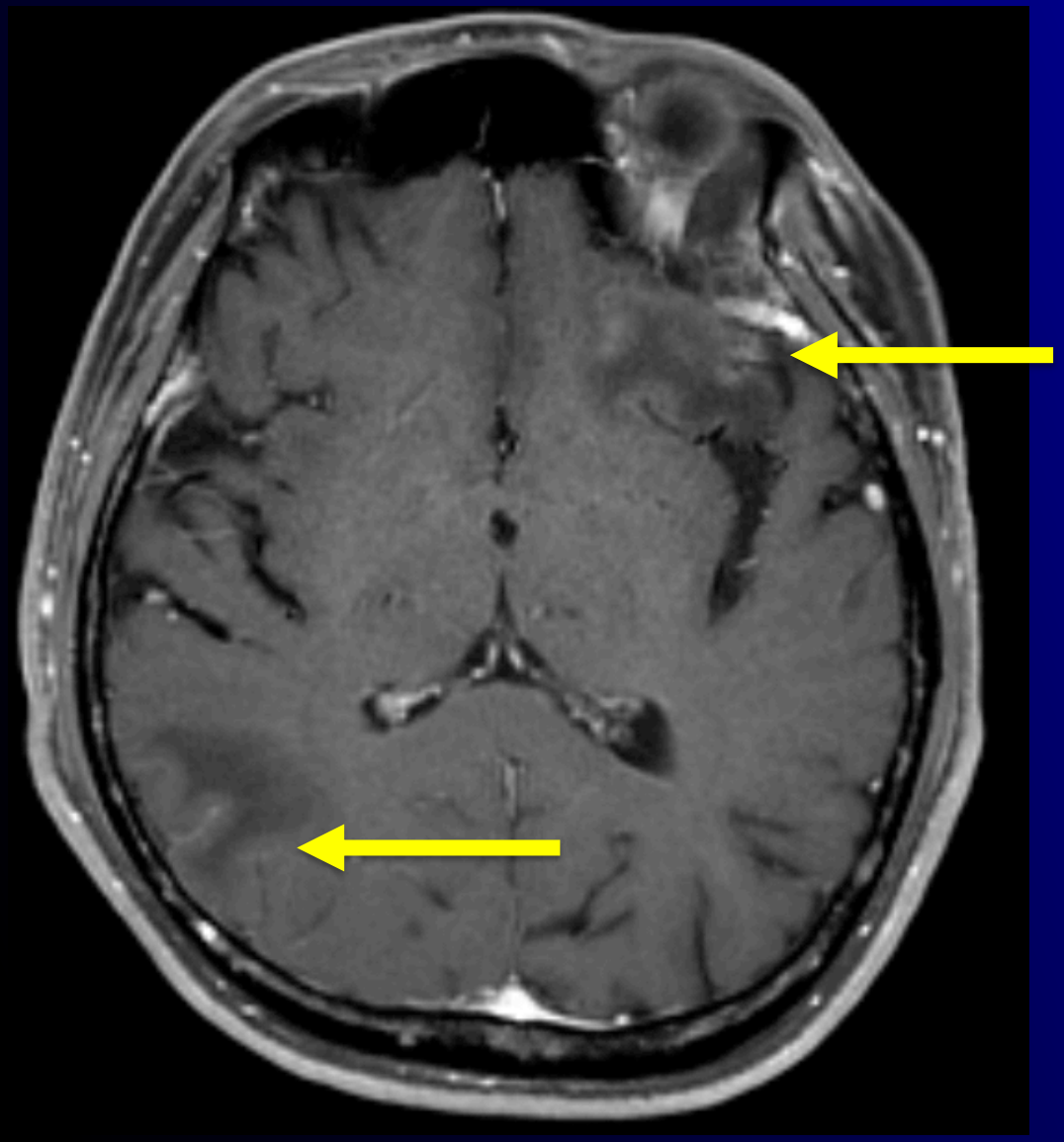

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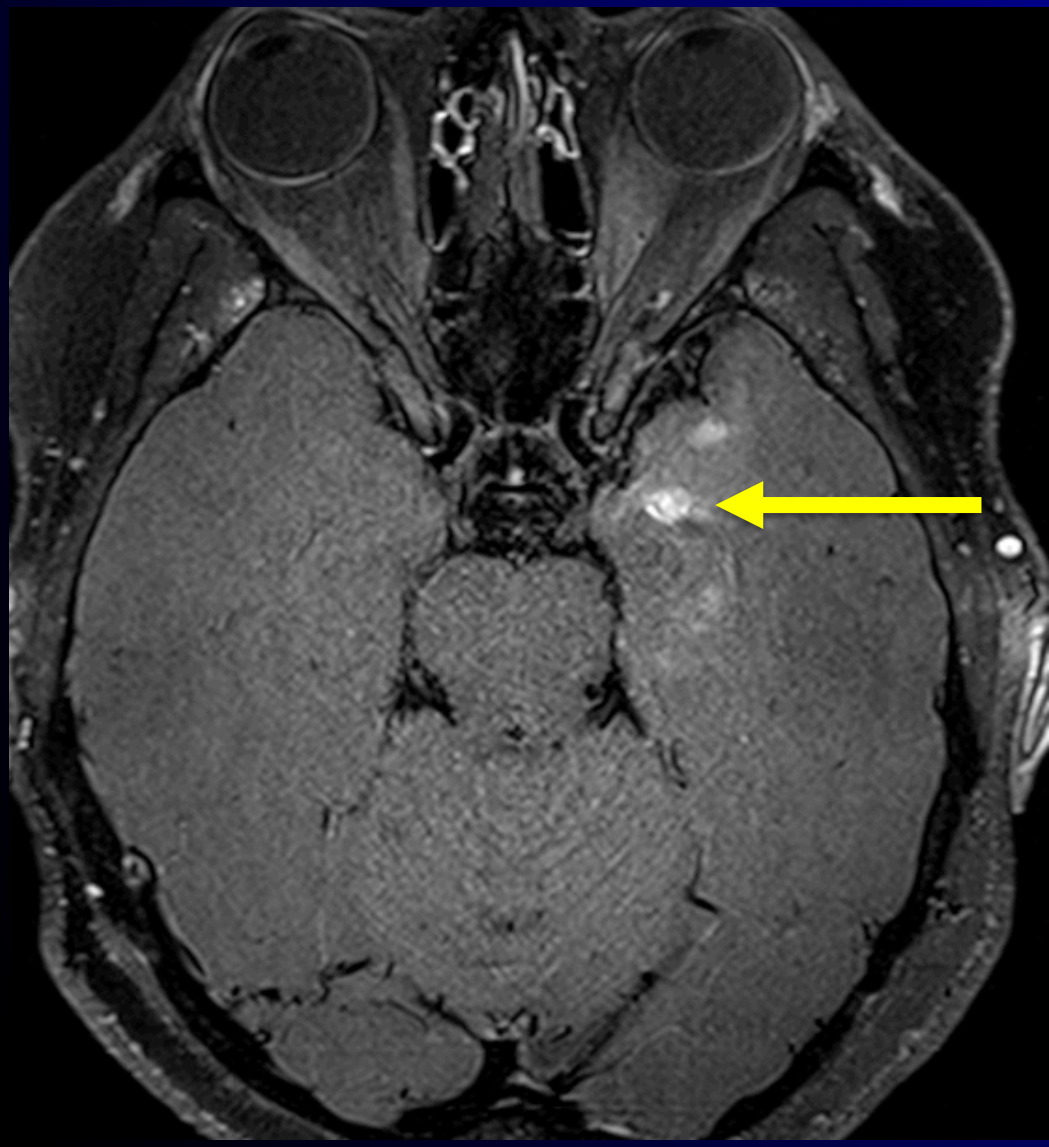

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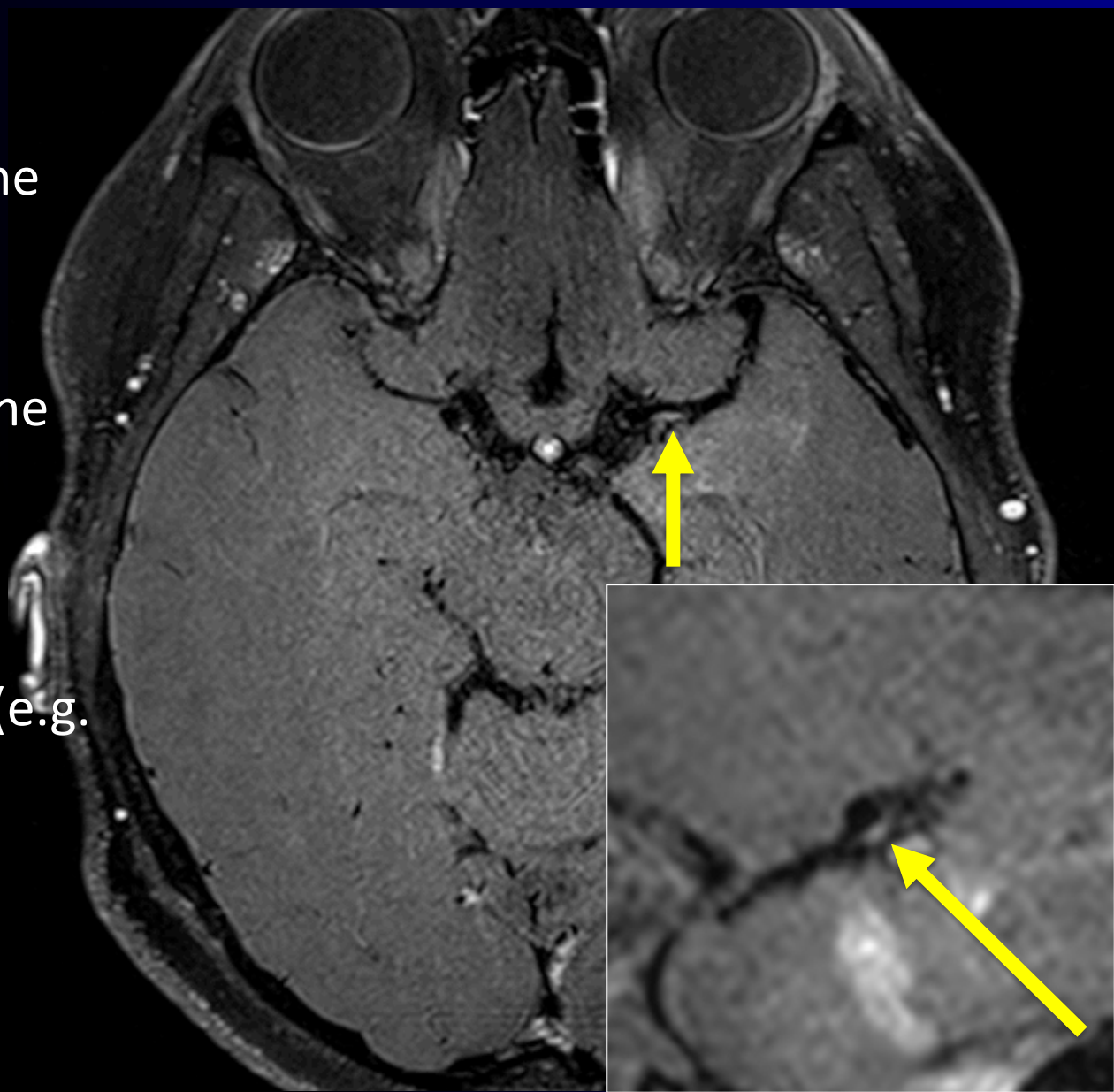

Susac syndrome

- Autoimmune endotheliopathy causing small infarctions of cochlea, retina and corpus callosum

- Acute encephalopathy, bilateral hearing loss and branch retinal artery occlusions

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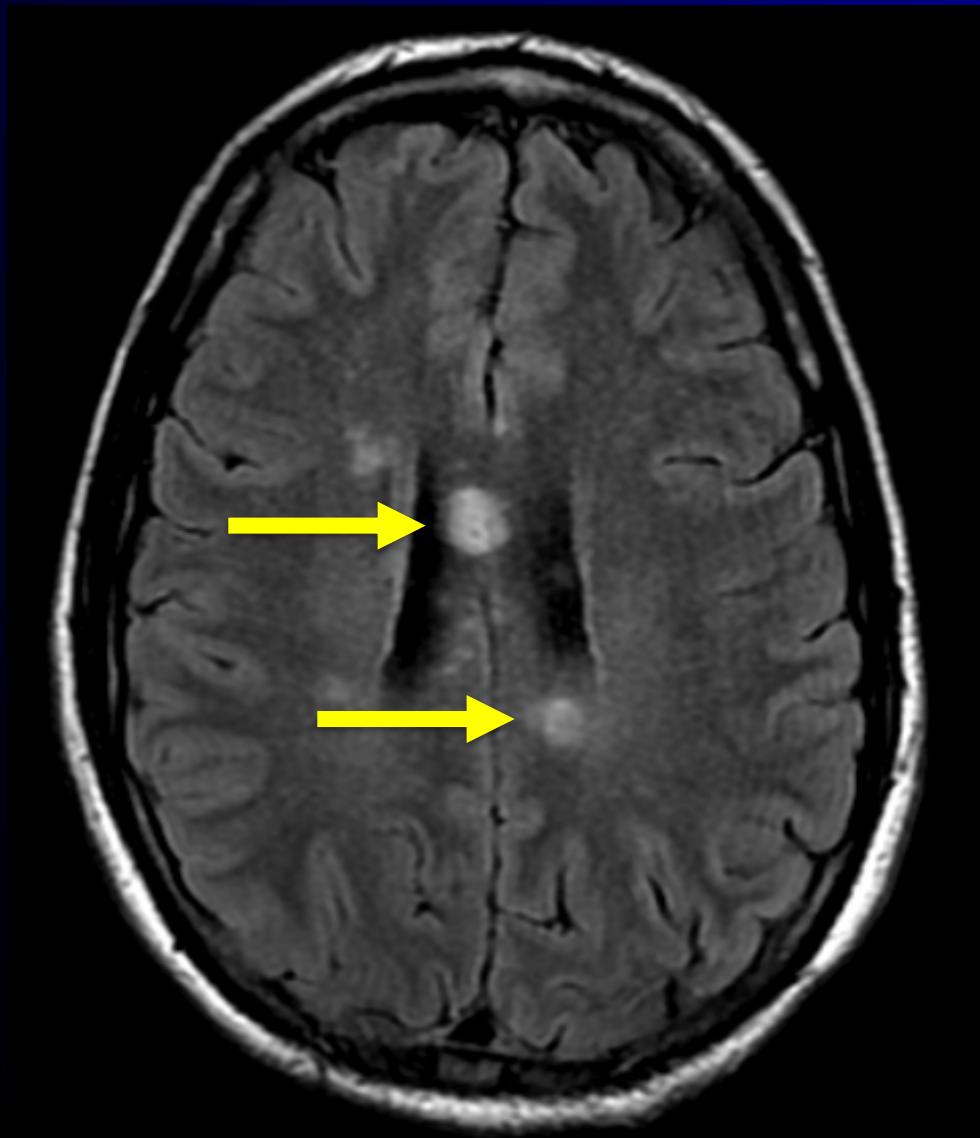

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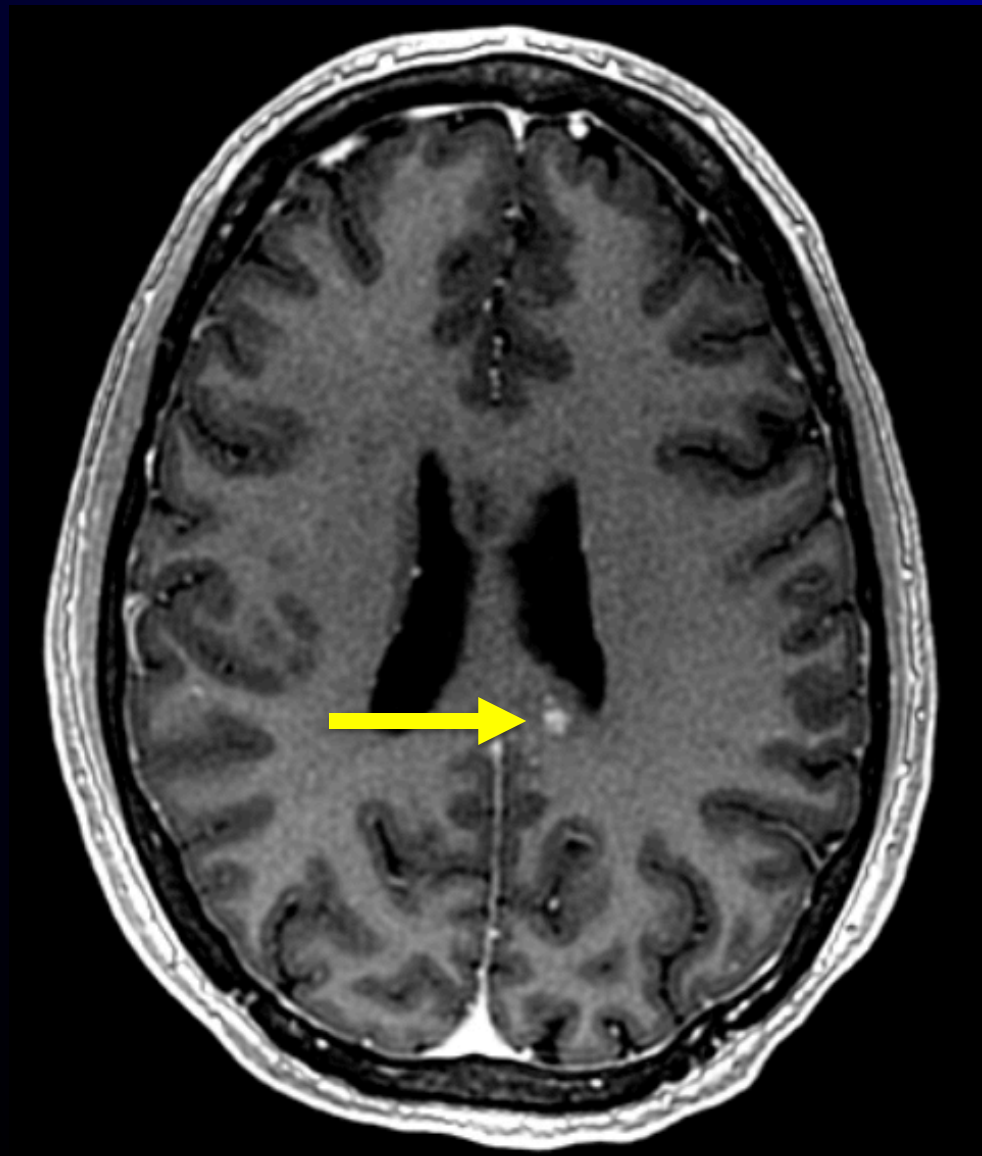

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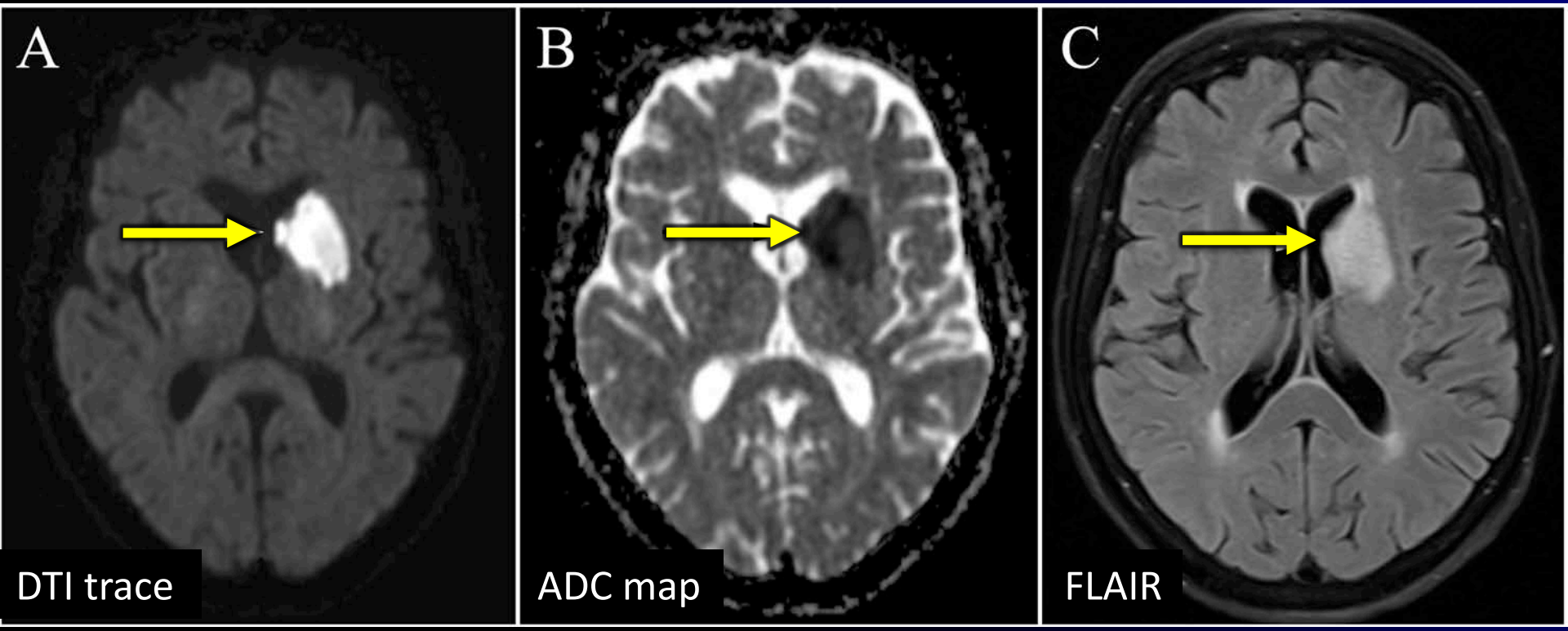
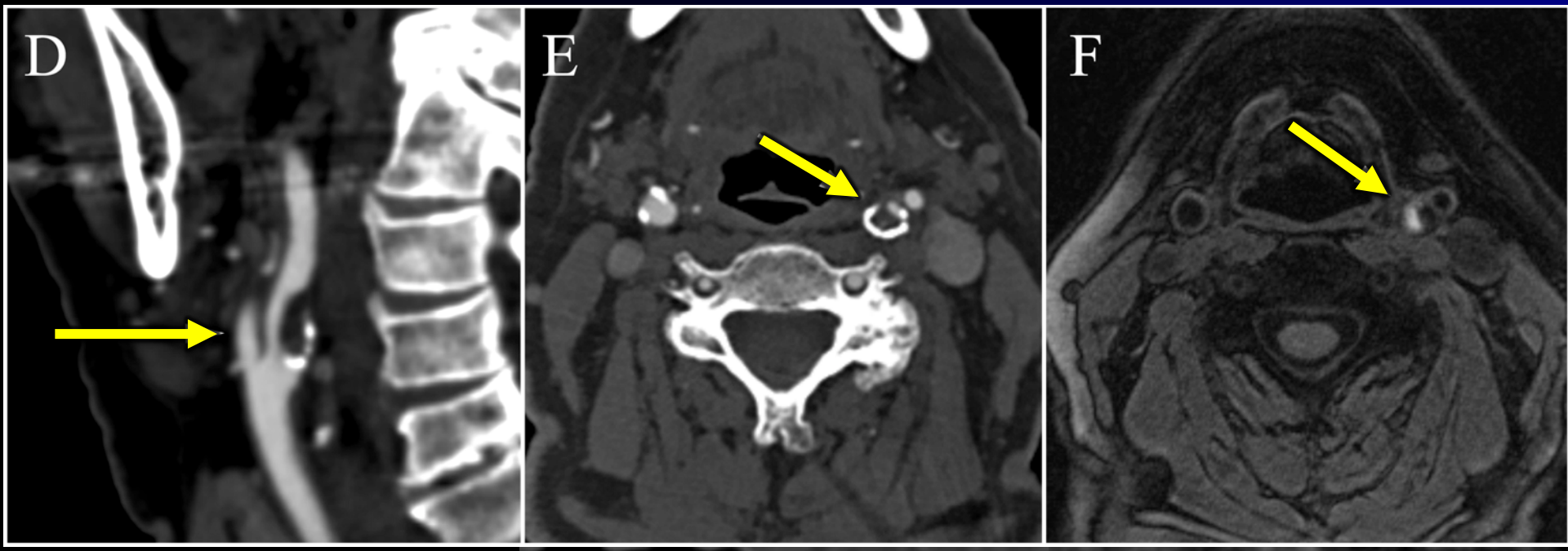
Ischemic stroke

- Patient presented with headache and right sided weakness.

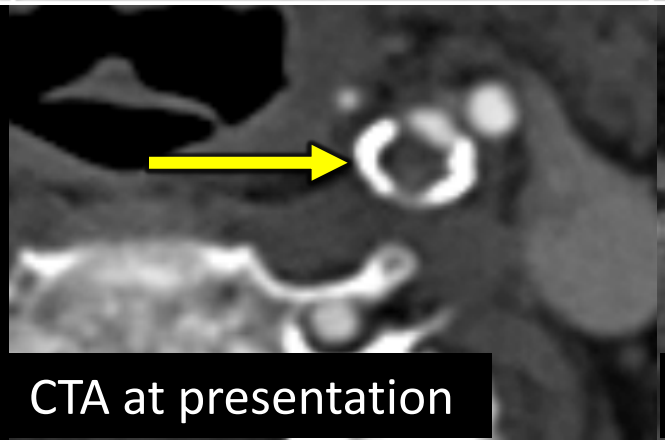


Ischemic stroke

- Initially thought to be secondary to carotid plaque



CTA at presentation



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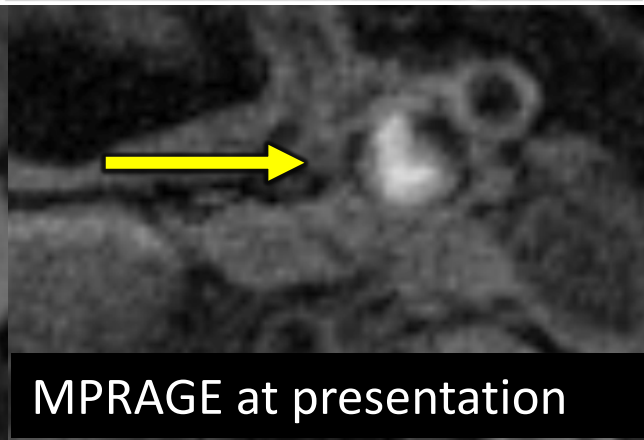
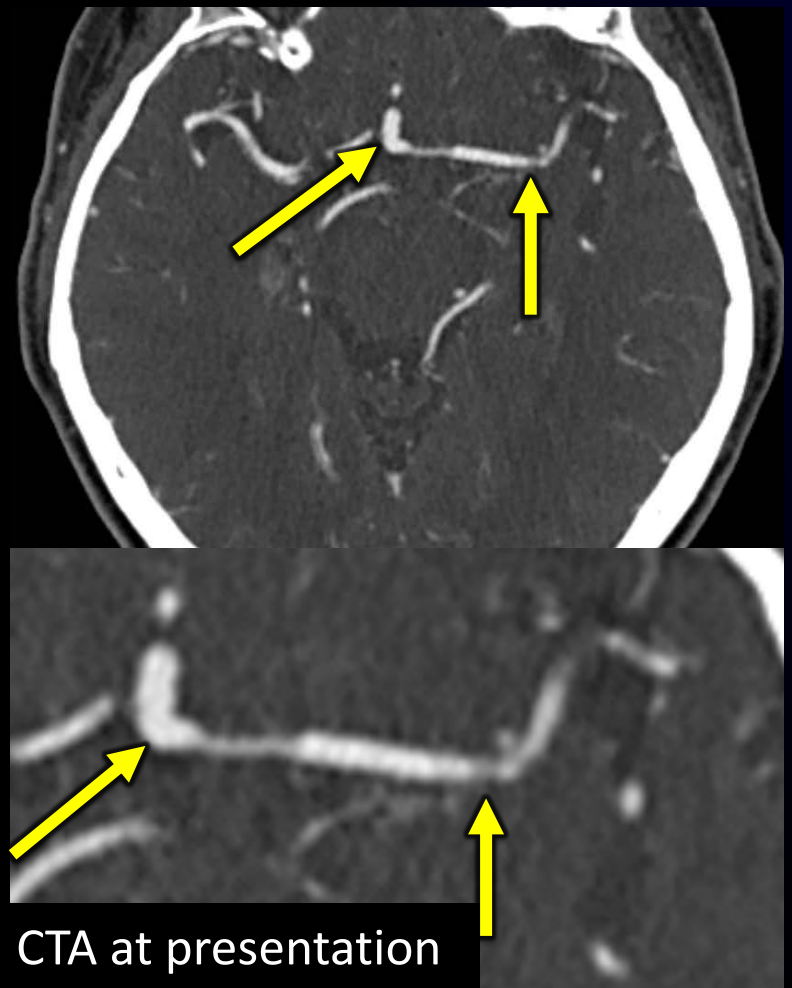


MPRAGE at presentation

Ischemic stroke

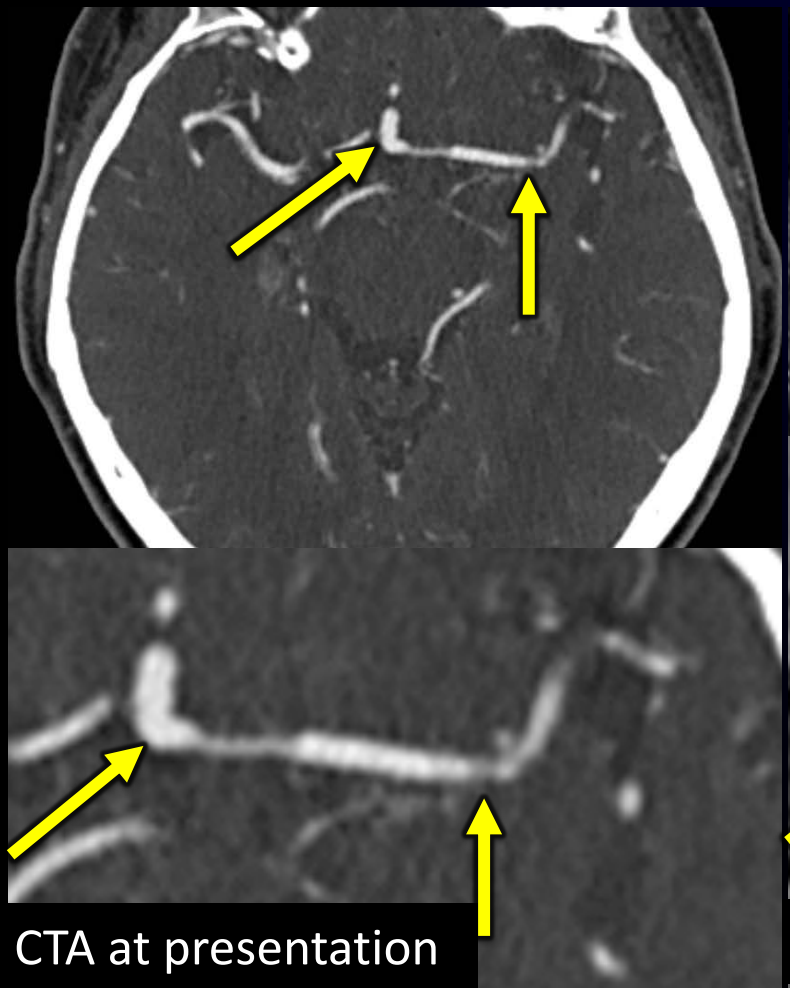
- Brain CTA showed left MCA/ACA lumen irregularity



CTA at presentation

Ischemic stroke

- vwMRI showed avid MCA/ACA wall enhancement
- Hx of facial rash 3mo ago, VZV positive LP



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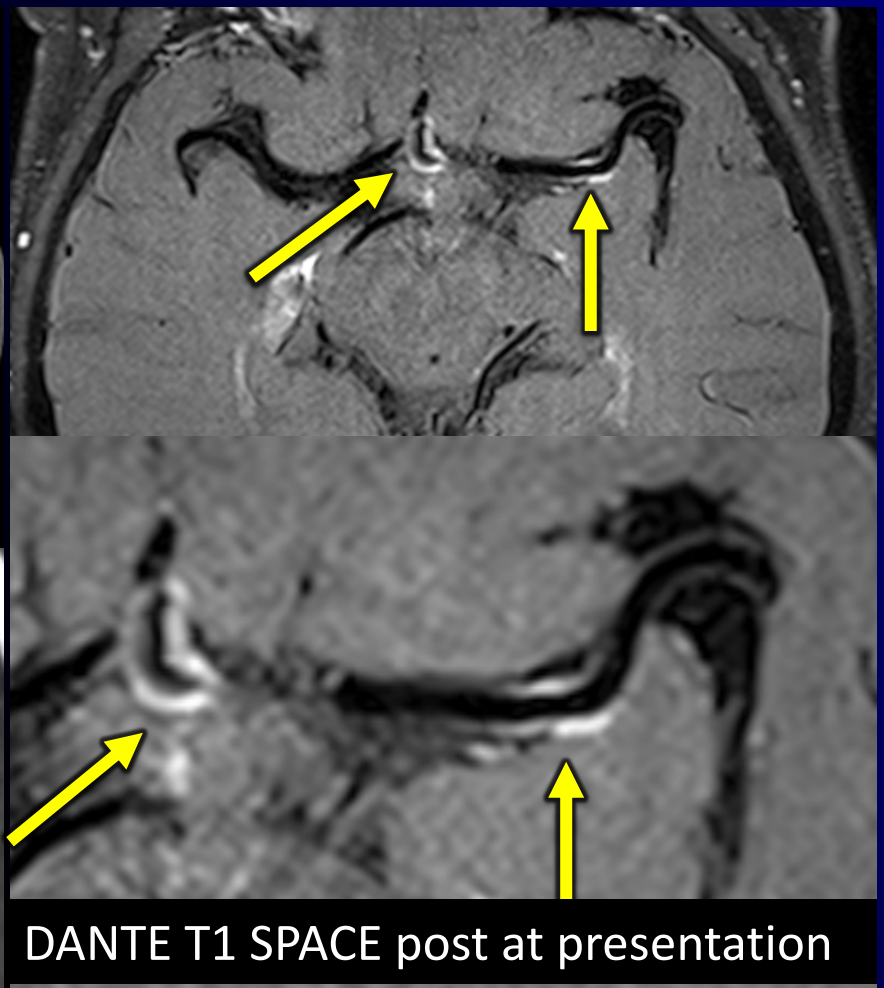
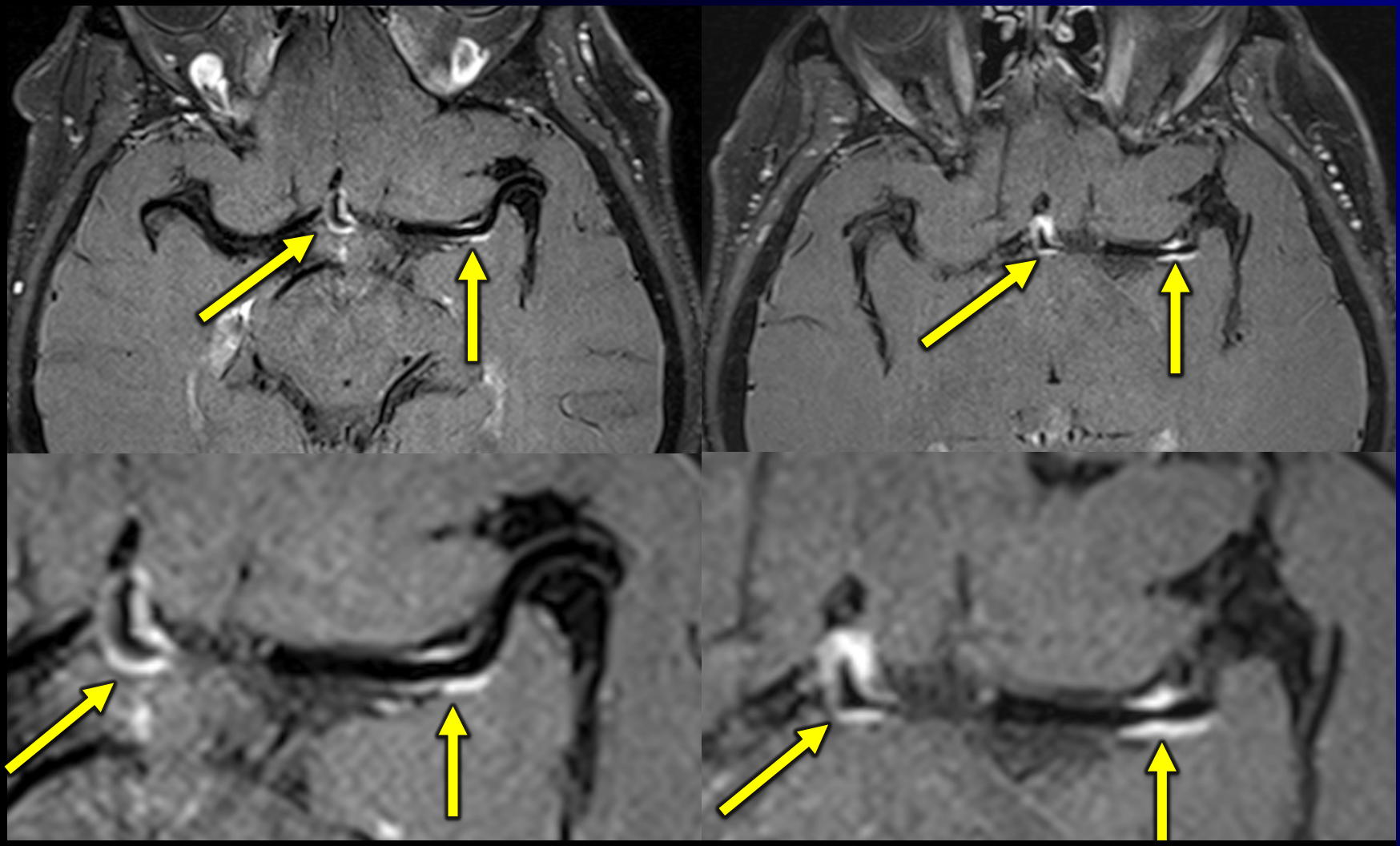


DANTE T1 SPACE post at presentation

Postviral vasculitis

- Despite clearing VZV, enhancement progressed



DANTE T1 SPACE post at presentation

DANTE T1 SPACE post at 1mo

Postviral vasculitis

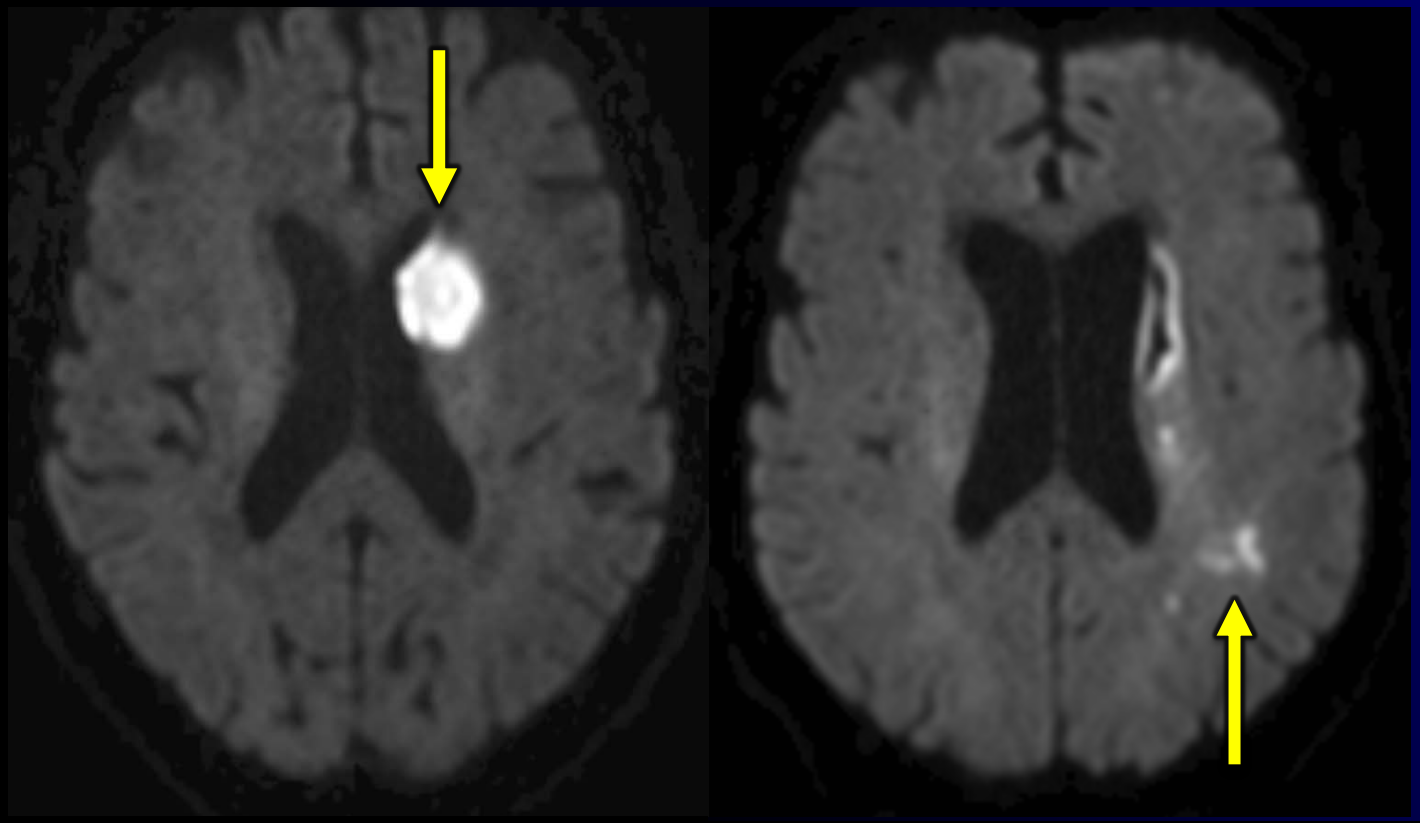
- Narrowing worsened

DSA 1mo after presentation

DSA 2mo after presentation

Postviral vasculitis

- The patient also developed new left MCA strokes after clearing VZV



DTI trace at presentation

DTI trace 2mo after presentation

Summary

- Rare autoimmune diseases can affect the brain, spinal cord, leptomeninges, nerves and blood vessels

- Imaging helps detect and diagnose autoimmune diseases

- Imaging allows monitoring of treatment response and detection of disease progression

Thank you

The University of Utah

Department of Radiology and Imaging Sciences

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