CNS Vasculitis: A Review

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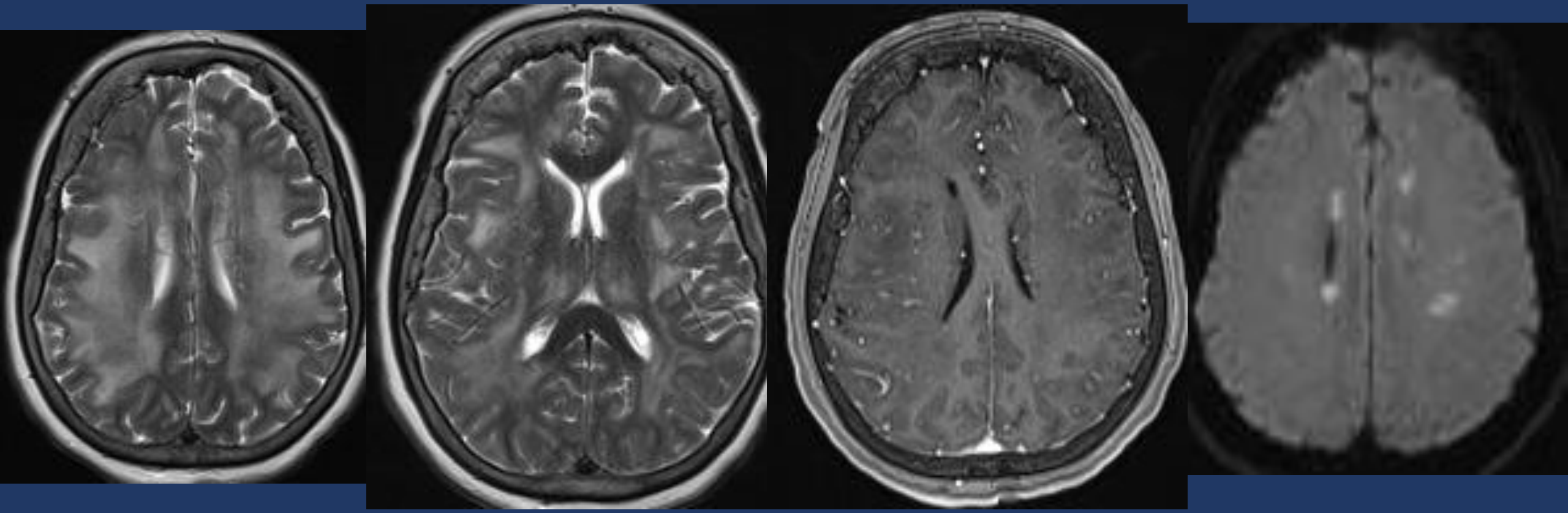
Case 1. 45 y/o man w/progressive encephalopathy & strokes



Granulomatous angiitis

Vasculitis: Definition & Classification

- Inflammation & necrosis of blood vessel wall (artery & vein)
- Secondary:
  - More common
  - In context of well-defined disorders
- Primary:
  - Very rare, idiopathic
- Disorders that mimic vasculitis (vasculopathies):
  - Atherosclerosis, RCVS
  - Both may show vessel wall enhancement

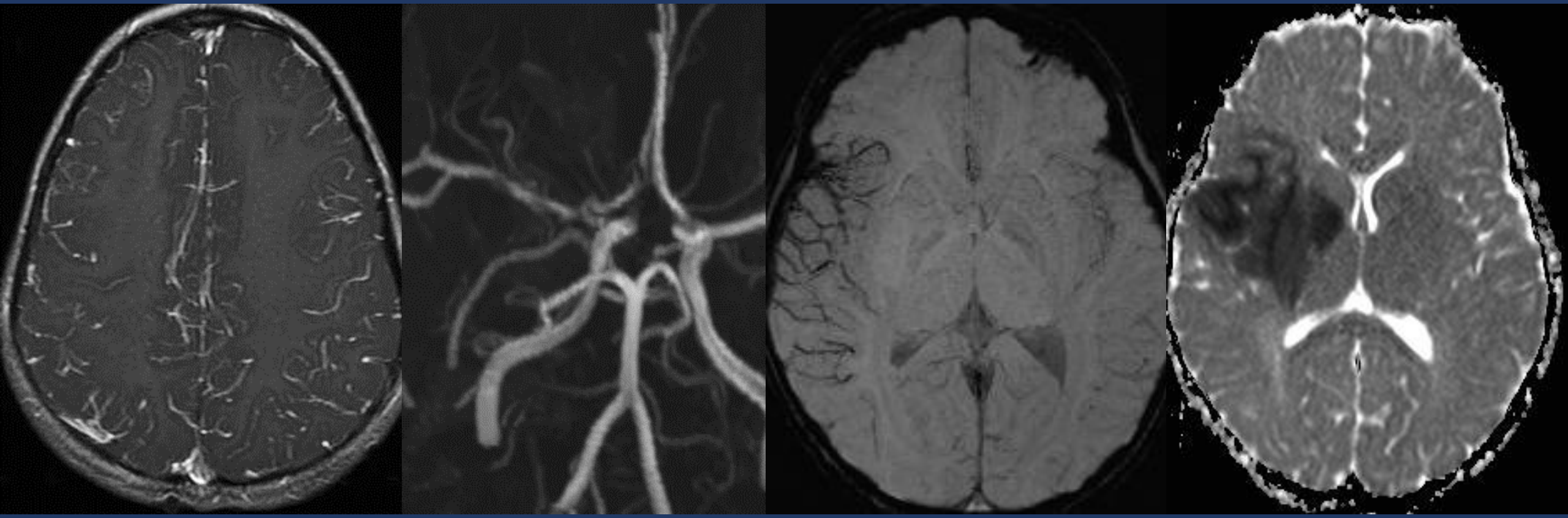
Vasculitis (autoimmune): Classification

Large vessels	Medium vessels	Small vessels
-Takayasu's -Giant cell (temporal arteritis) -Polymyalgia rheumatica	-Polyarteritis nodosa -Kawasaki's	-Non-ANCA*: Henoch Schonlein purpura (IgA vasculitis) -ANCA-associated: Wegener (granulomatosis w/polyangiitis), microscopic polyangiitis & Churg Strauss (eosinophilic)

-Most important in neuroradiology: Takayasu's, giant cell & Wegener
-Variable size vessels: Behcet & Cogan syndrome

ANCA = anti-neutrophil cytoplasmic antibody

Case 2. Child w/fever & left hemiplegia

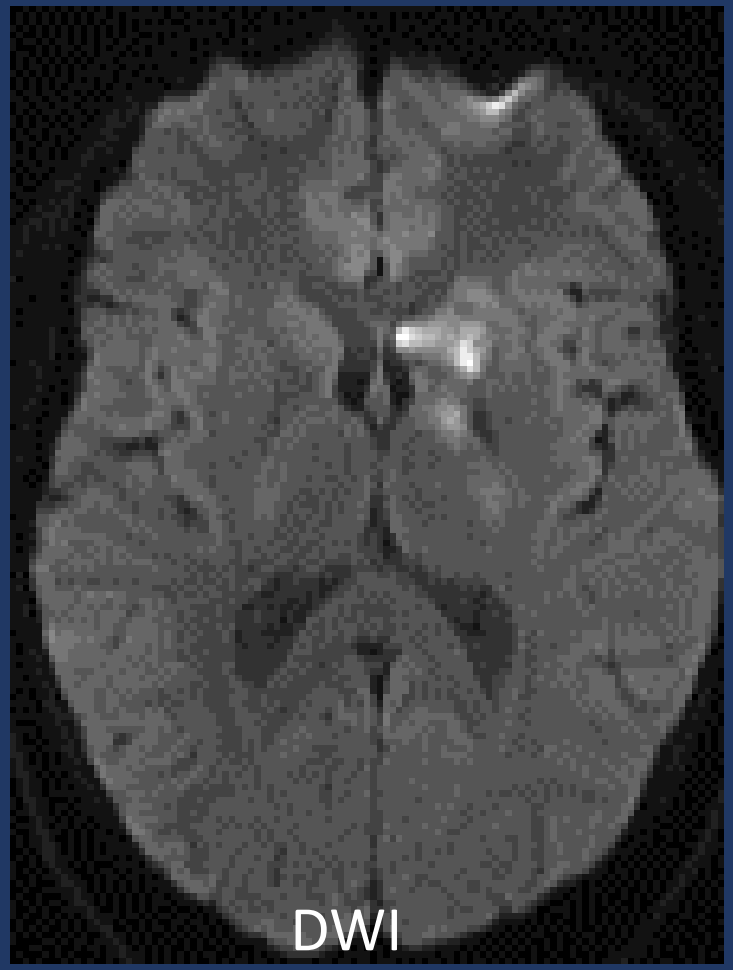


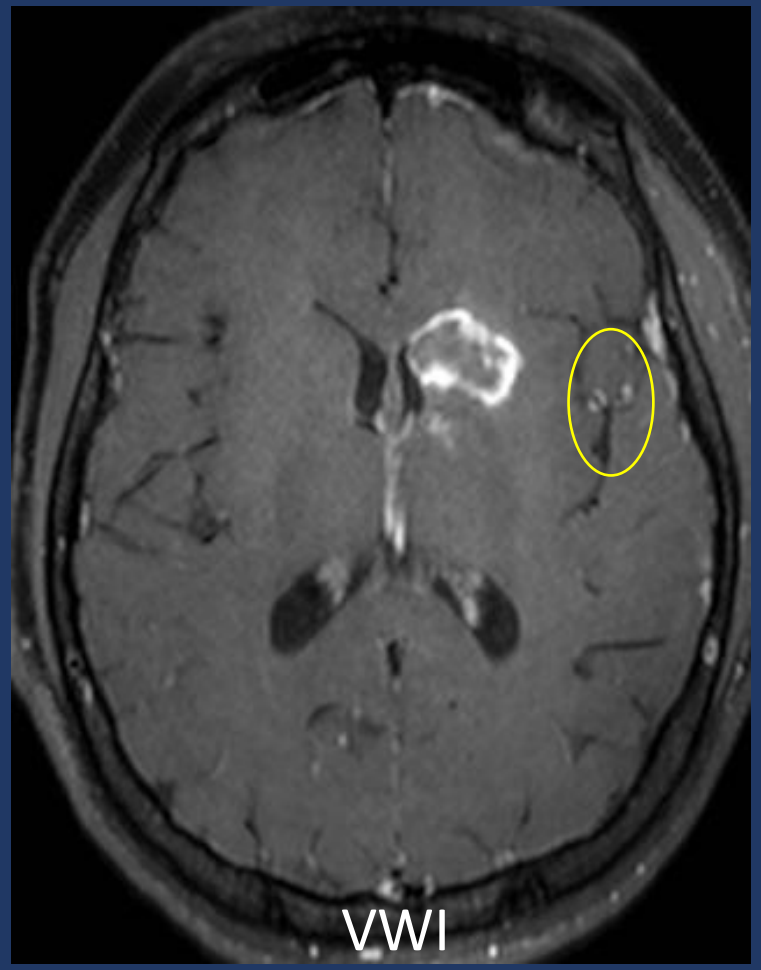
Varicella Zoster Vasculitis

Varicella Zoster Vasculitis

- Due to reactivation of virus
  - Migration through nerve fibers innervating arteries
  - Arterial involvement in 70% of pts
- Involvement related to immune status:
  - Competent: MCAs
  - Deficient: distal arteries

J Stroke Cerebrovasc Dis 2016; 25L 1561
Neurology 2013; 80: 62
J Neurol Sci 2015; 351: 168


DWI


VWI

Cor>Tra
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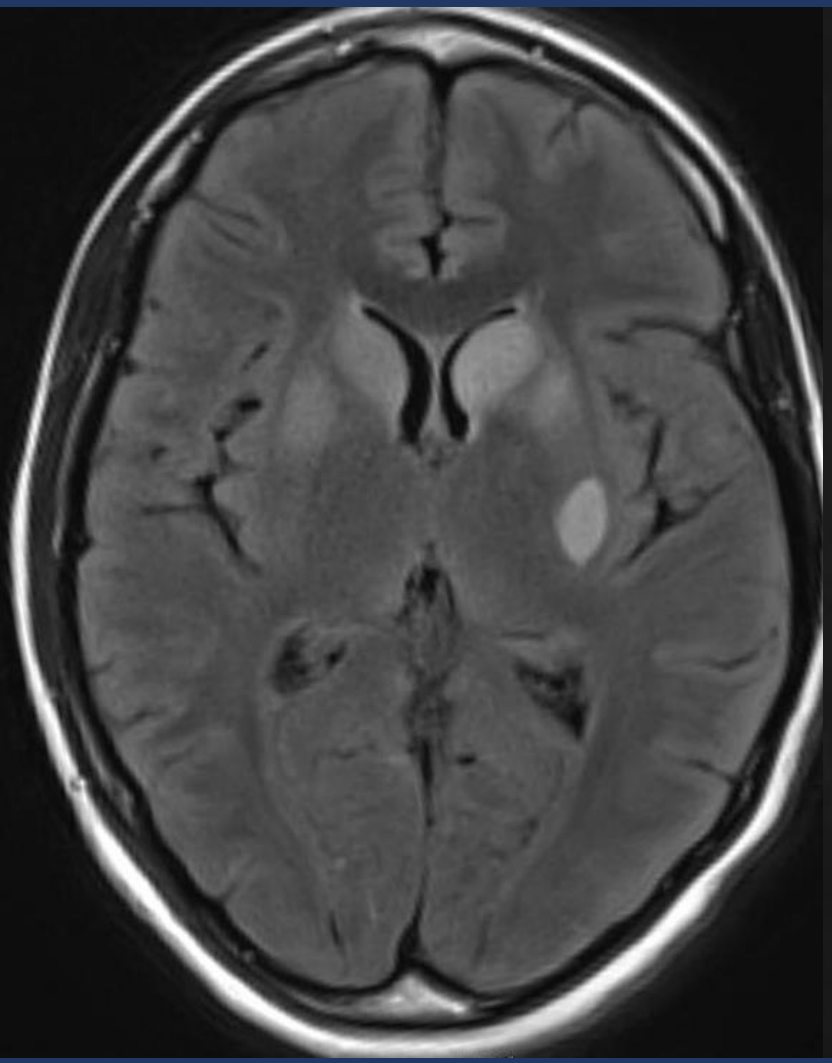
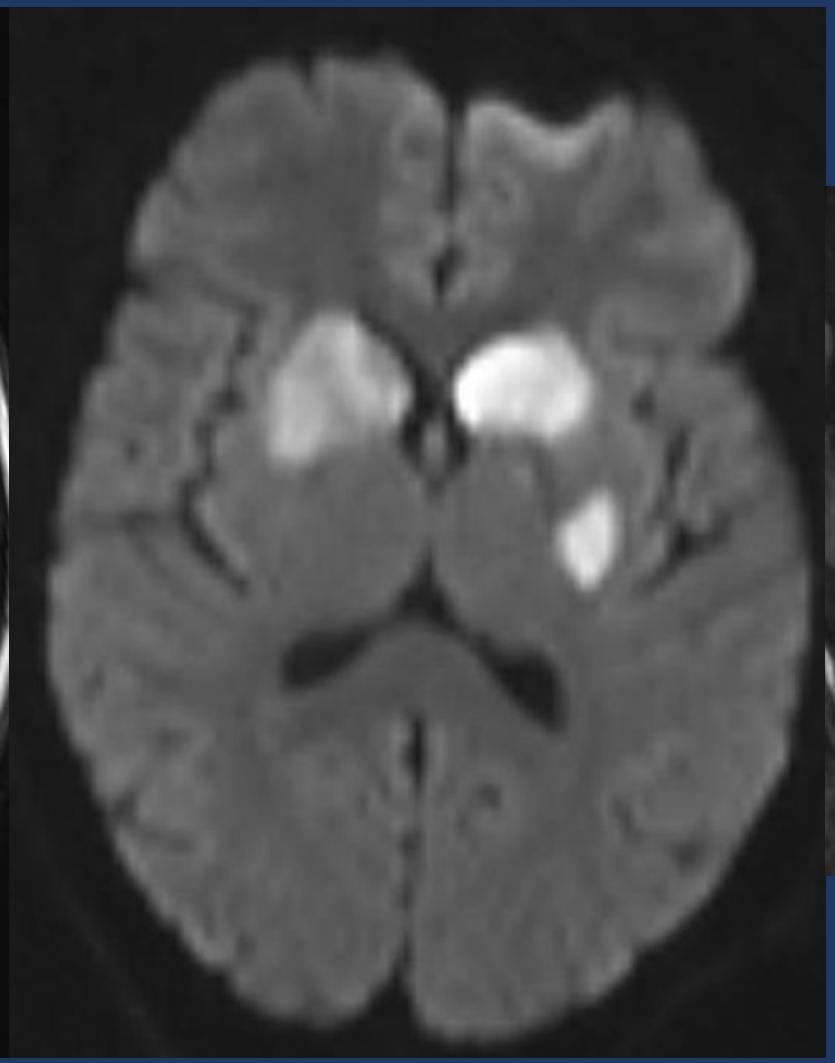
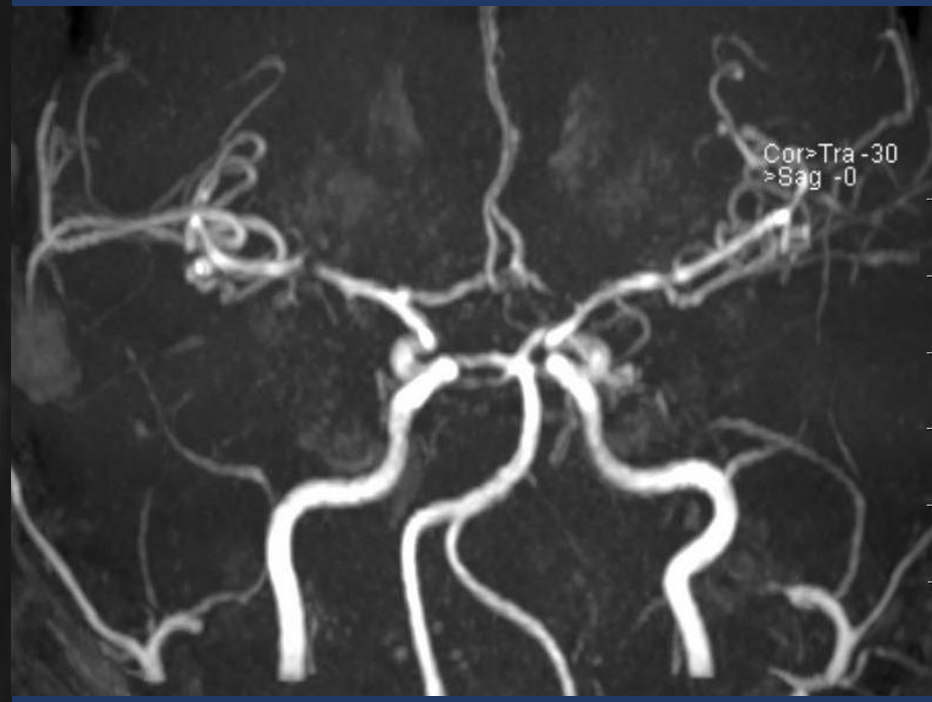
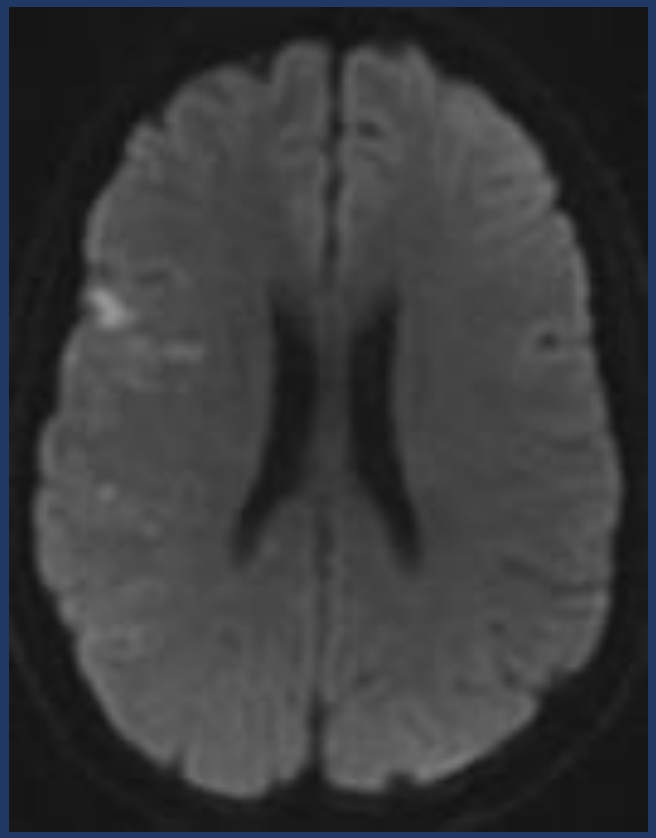


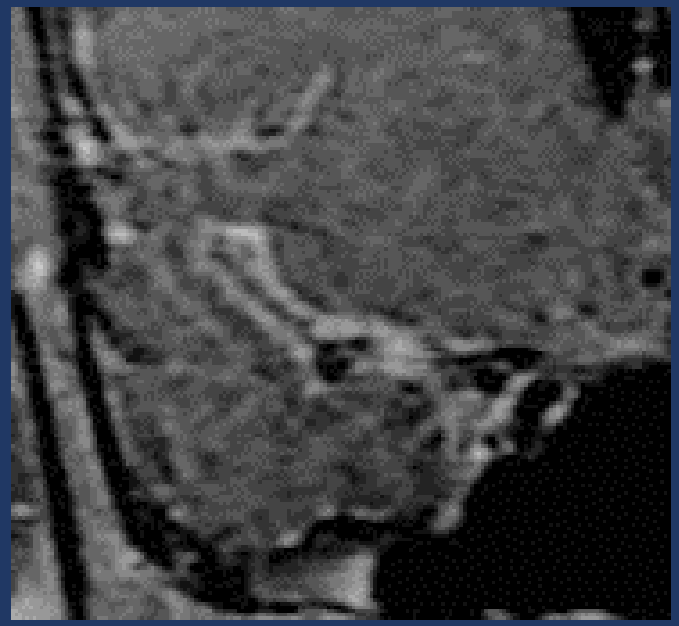



Cor>Tra -30
>Sag -0

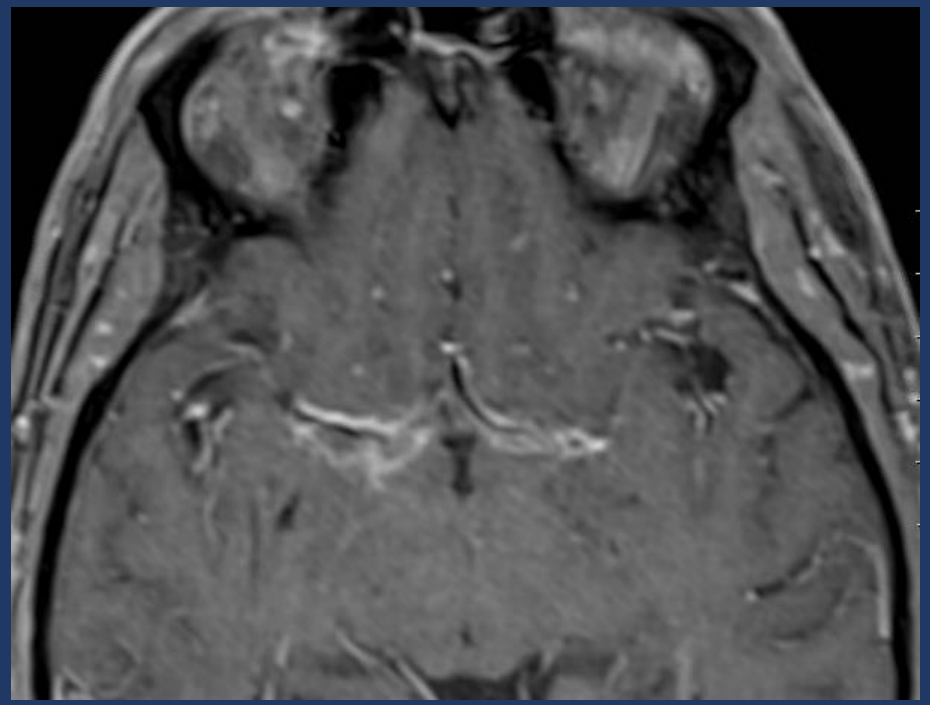
Infection-related Vasculitis

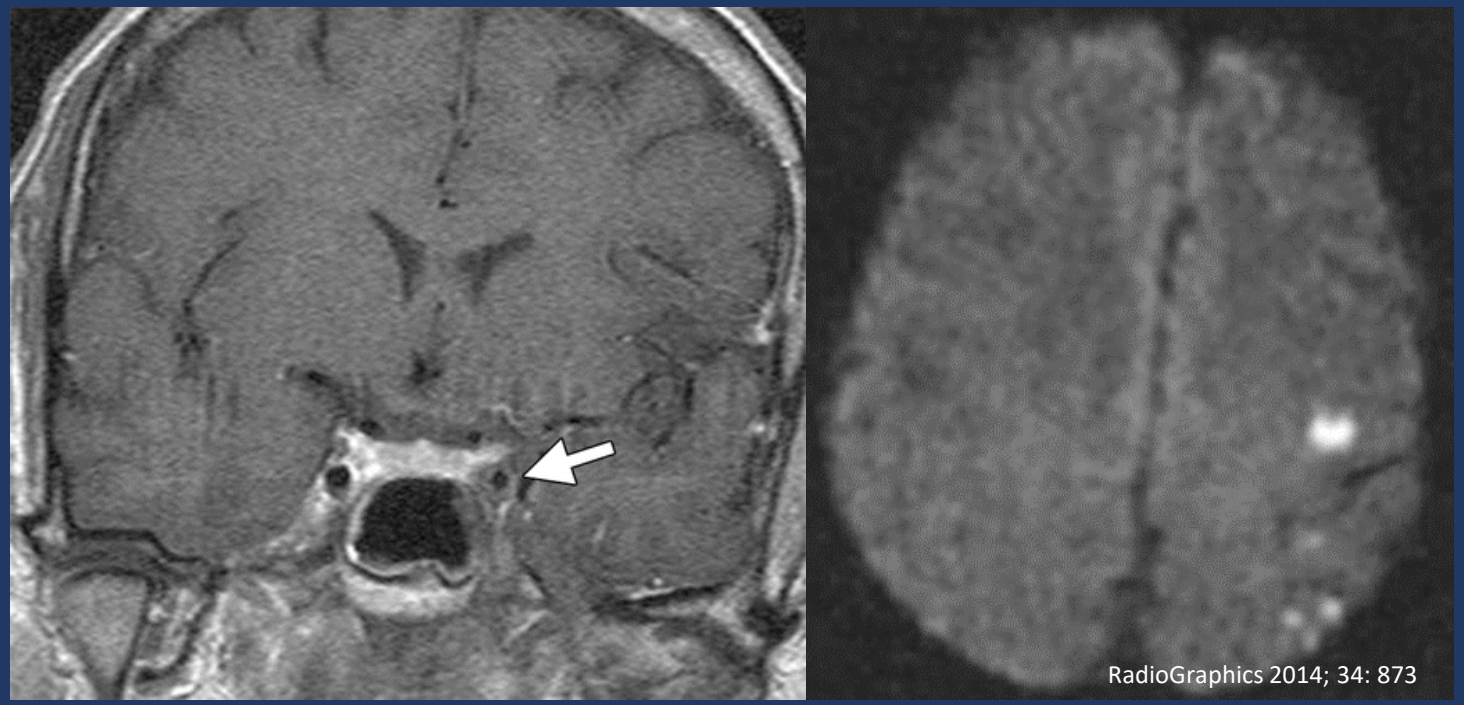
- Other causes:
  - TB
  - Angiophillic fungi (aspergillus, mucormycosis)
  - Staph aureus
  - Syphilis



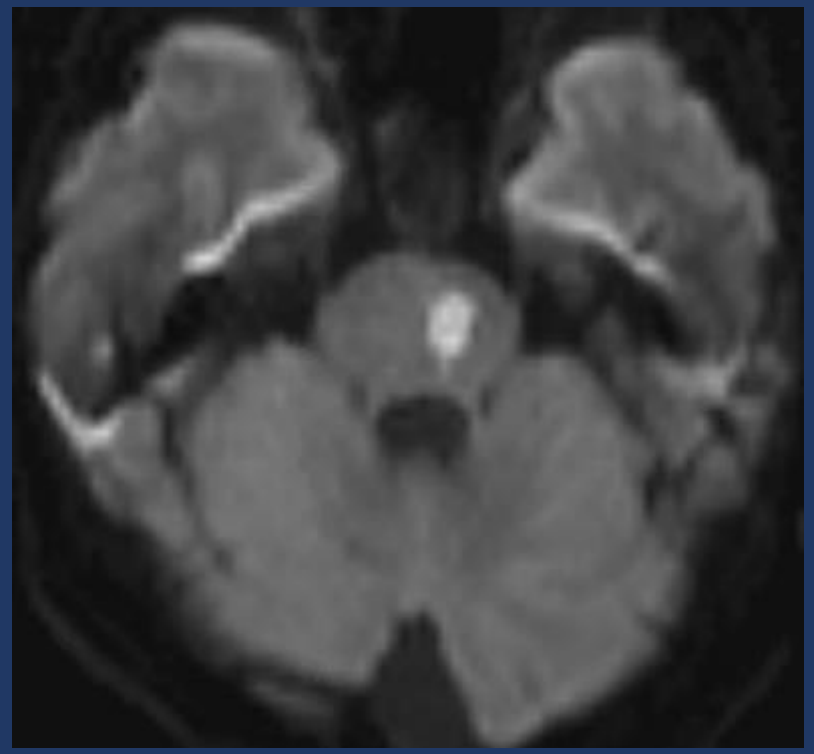


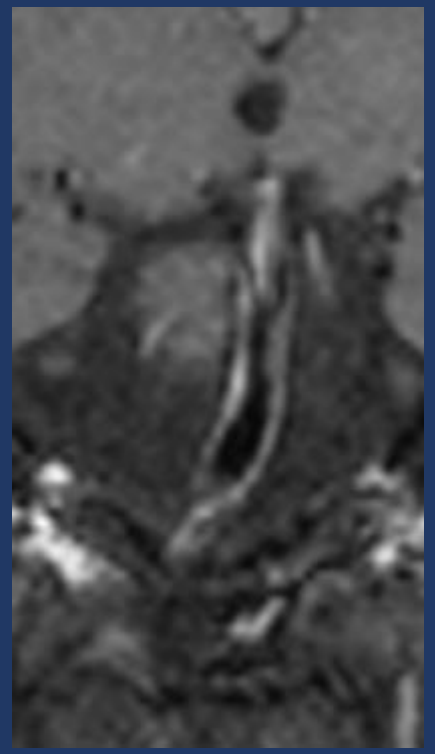
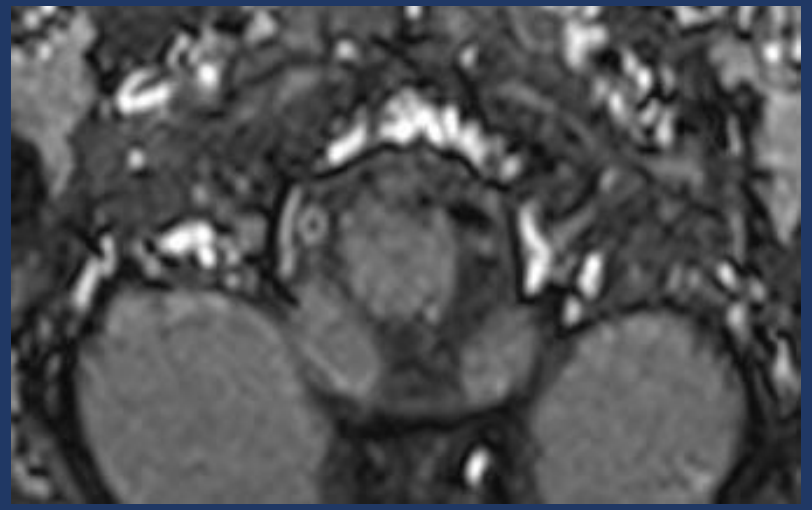
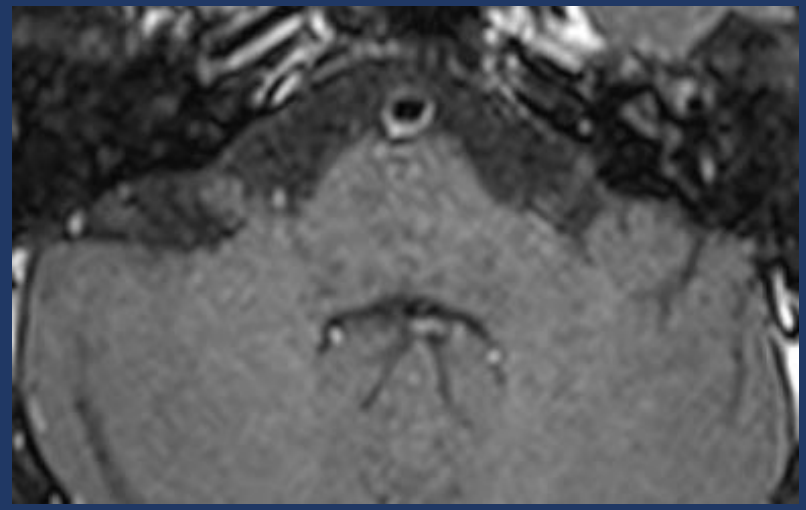
Tuberculosis



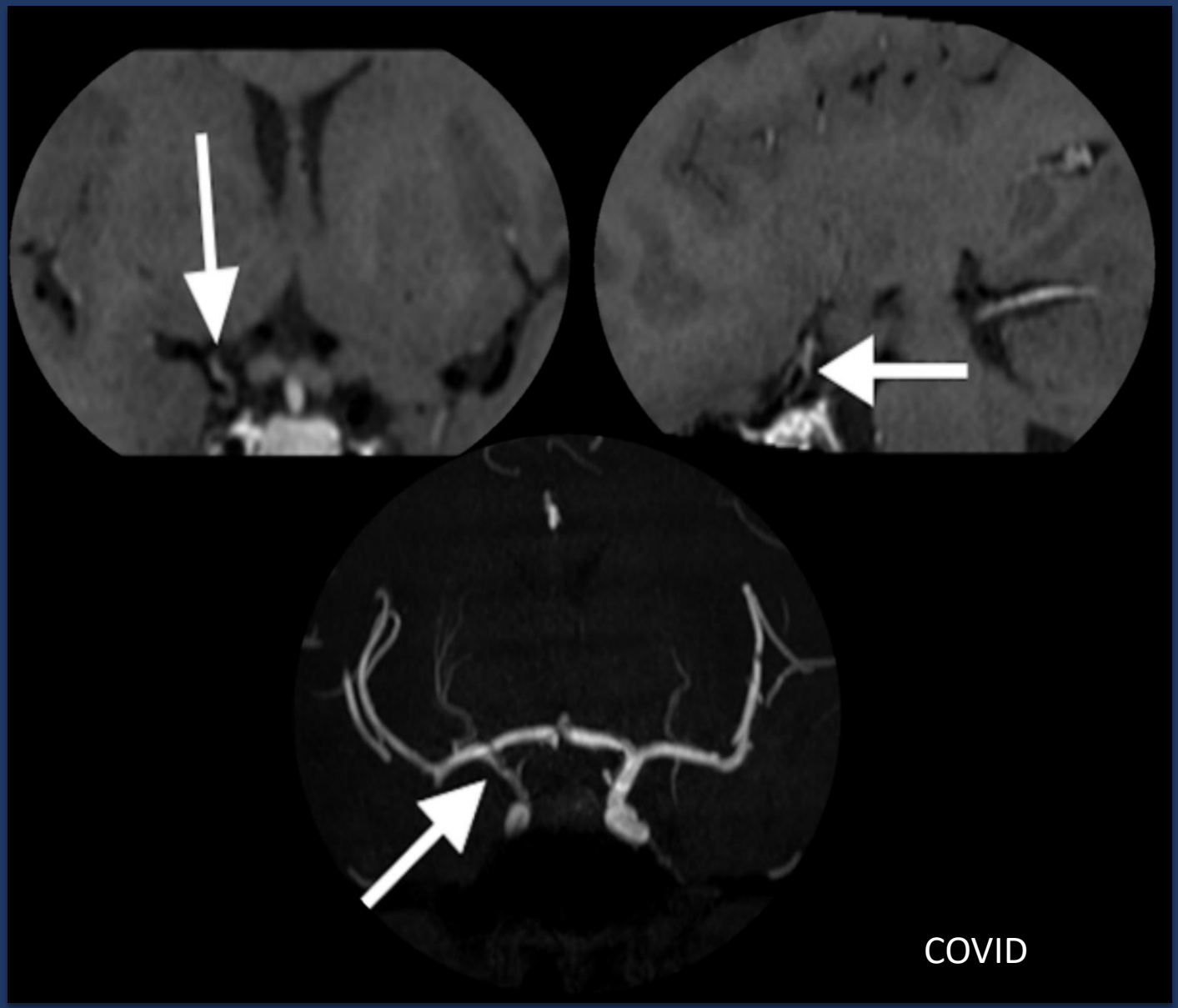


Mucormycosis

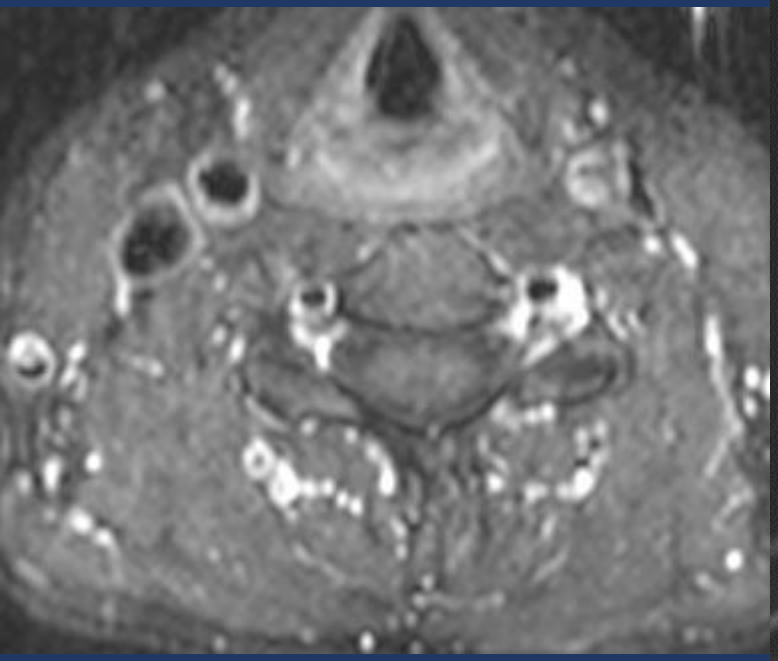


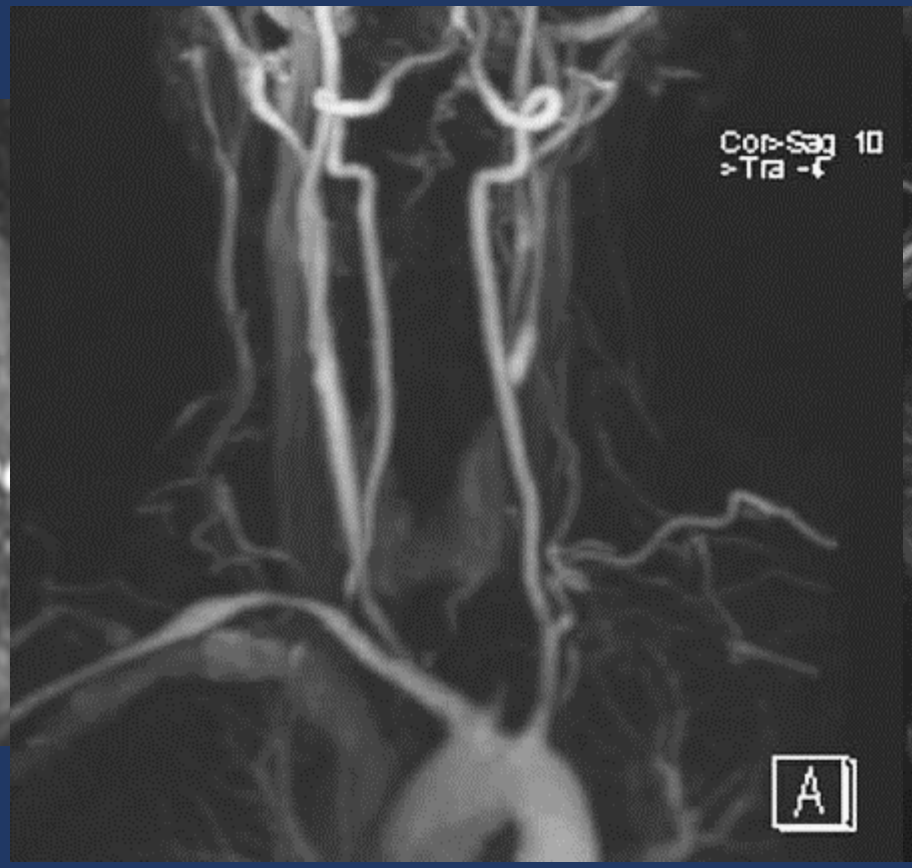




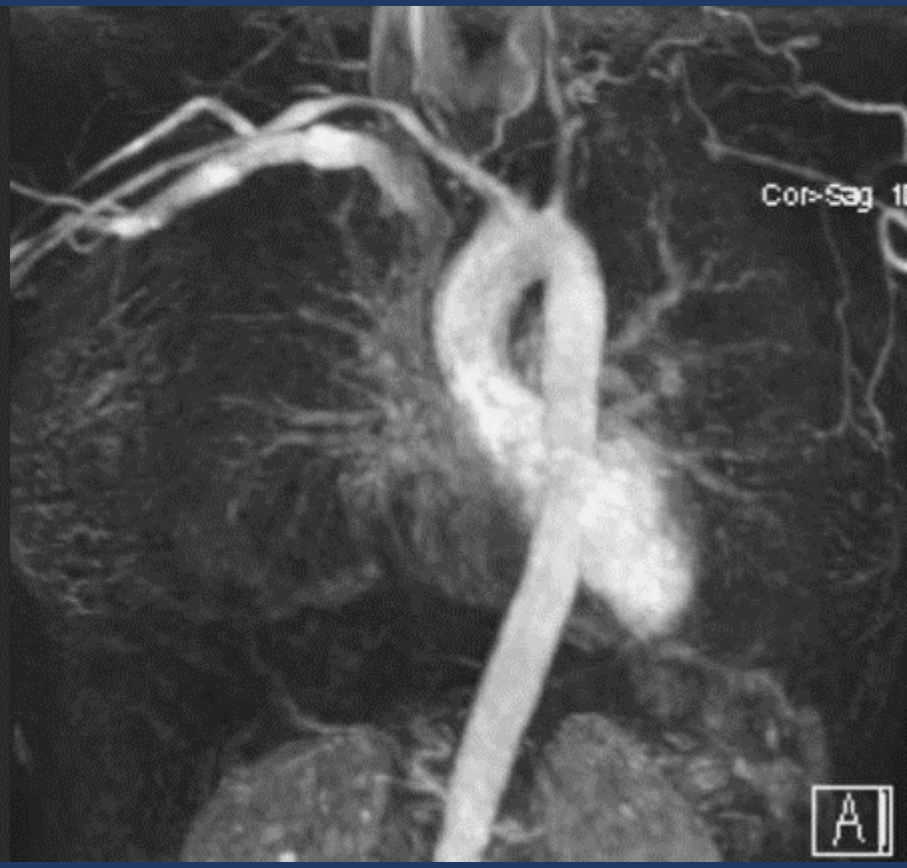
Syphilis


COVID

Case 3. Young female w/decreased UE pulses







Takayasu’s arteritis

Takayasu's Arteritis

- Incidence: 1: 3,000,000 (x100 in Asia)
- Predominantly women, 15-30 years of age
- Affects aorta & major arteries (including renal ones)
- Clinical:
  - Pre-pulseless phase: systemic symptoms, MRI: arterial wall enhancement, arterial dilatation
  - Occlusive phase, MRI: wall enhancement + narrowing

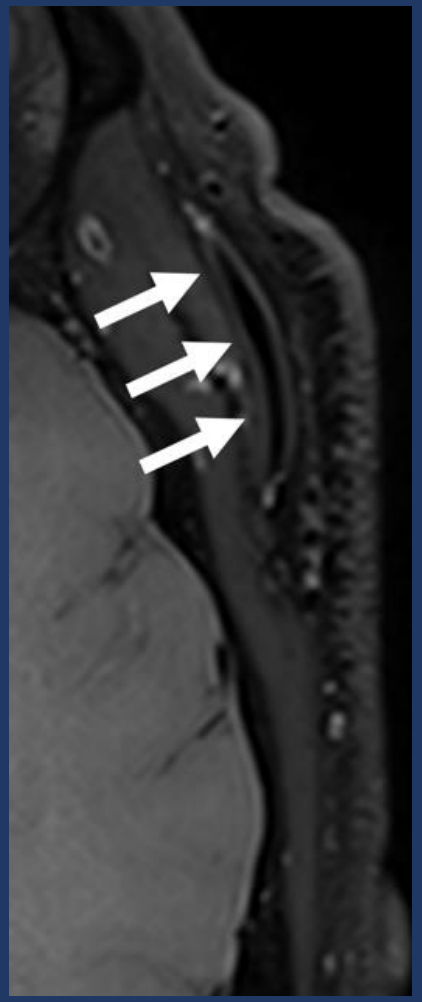
Takayasu's Arteritis

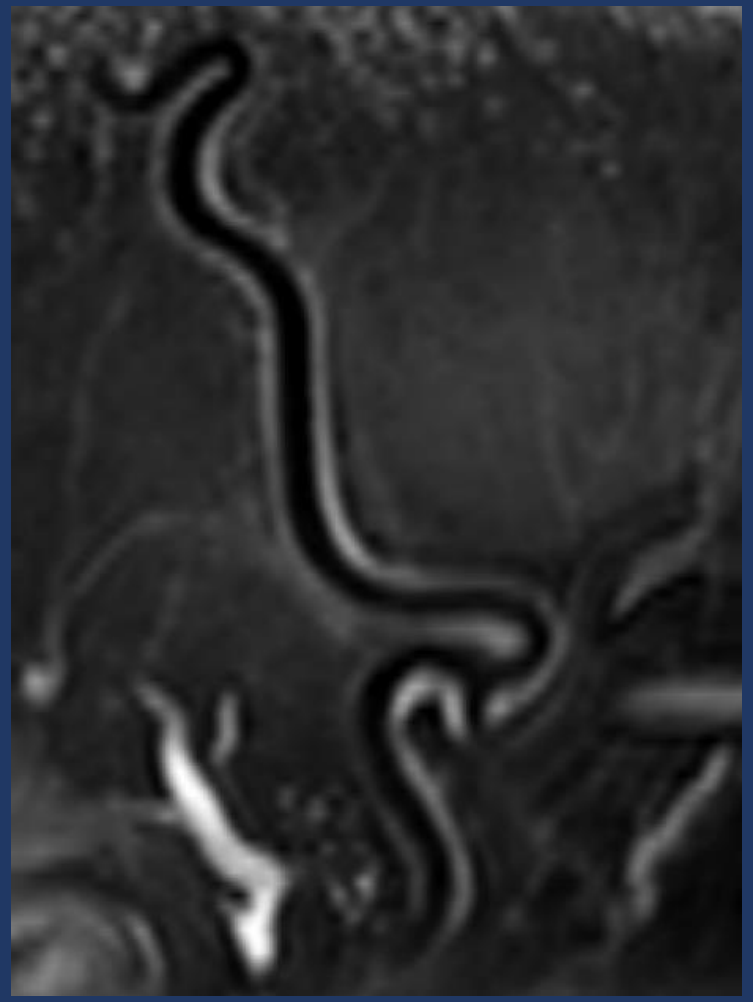
ARTERY	ABNORMALITIES %	CLINICAL MANIFESTATIONS
Subclavian	93%	UE claudication, Raynaud phenomenon
Common carotid	58%	Visual changes, stroke
Abdominal aorta	47%	Pain
Renal	38%	Hypertension, renal failure
Aortic arch	35%	Aortic insufficiency
Vertebral	35%	Visual changes, stroke
Pulmonary	20%	Chest pain, dyspnea
Celiac axis	18%	Abdominal pain

Pediatr Nephrol 2010; 25: 1037

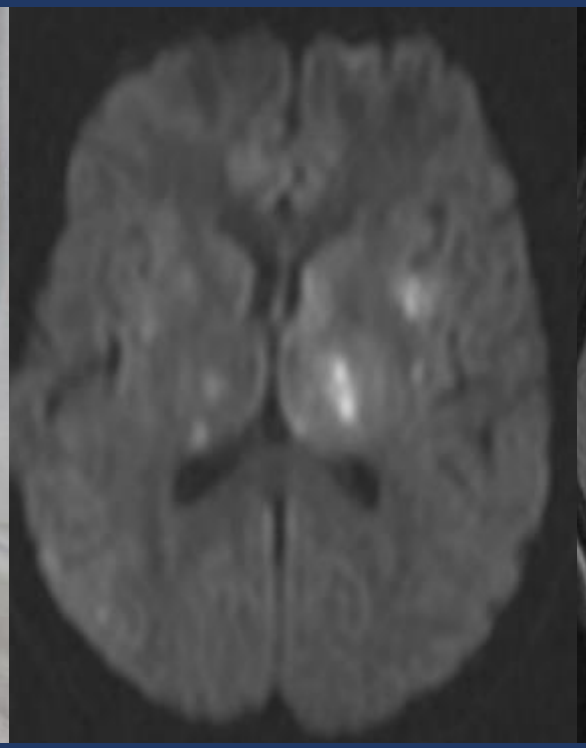
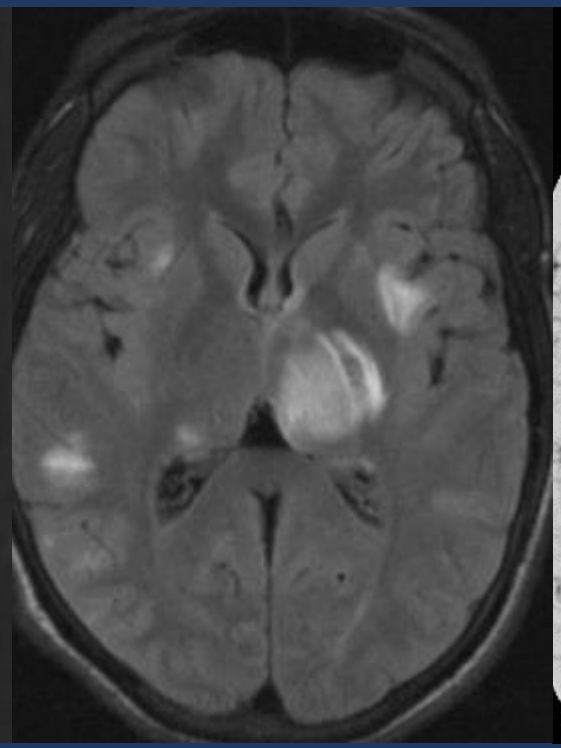
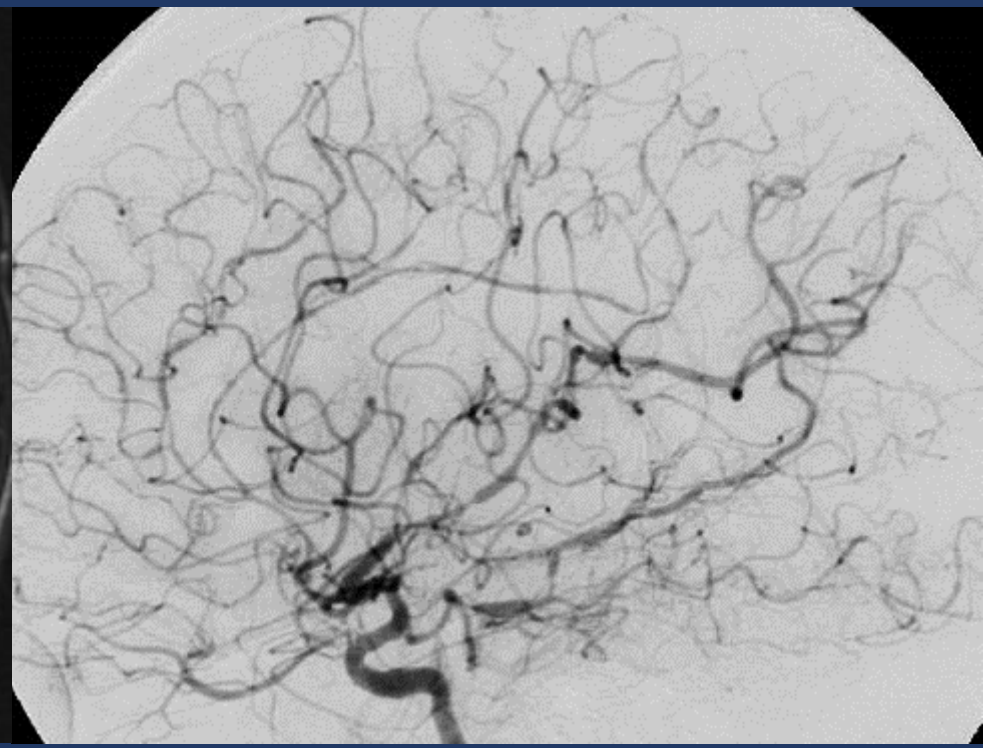
Pre-pulseless & occlusive phase

Giant Cell Arteritis





Case 4. Young female w/malaise, fever & strokes

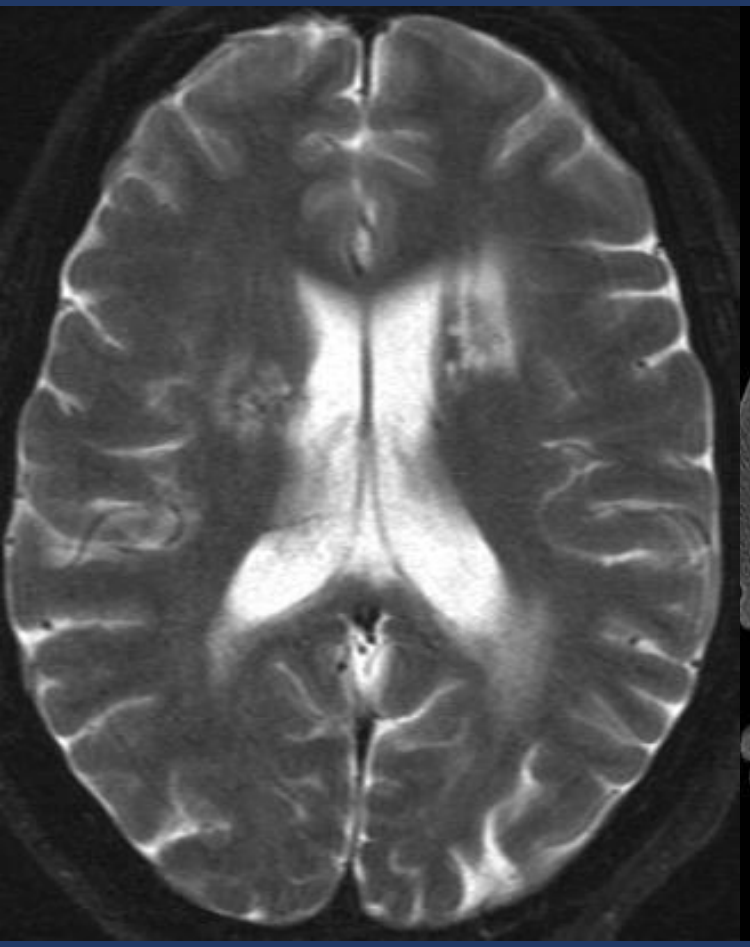




Polyarteritis nodosa

Polyarteritis Nodosa, Medium-size Vessels

- Incidence: 2-9/1,000,000
- Rare in children
- Diagnosis: histological evidence of necrotizing vasculitis + skin lesions *or* myalgia *or* hypertension *or* neuropathy *or* renal involvement
- Imaging: vasculitis, microaneurysms are classic but not identified on imaging

Case 5. Clinical history withheld



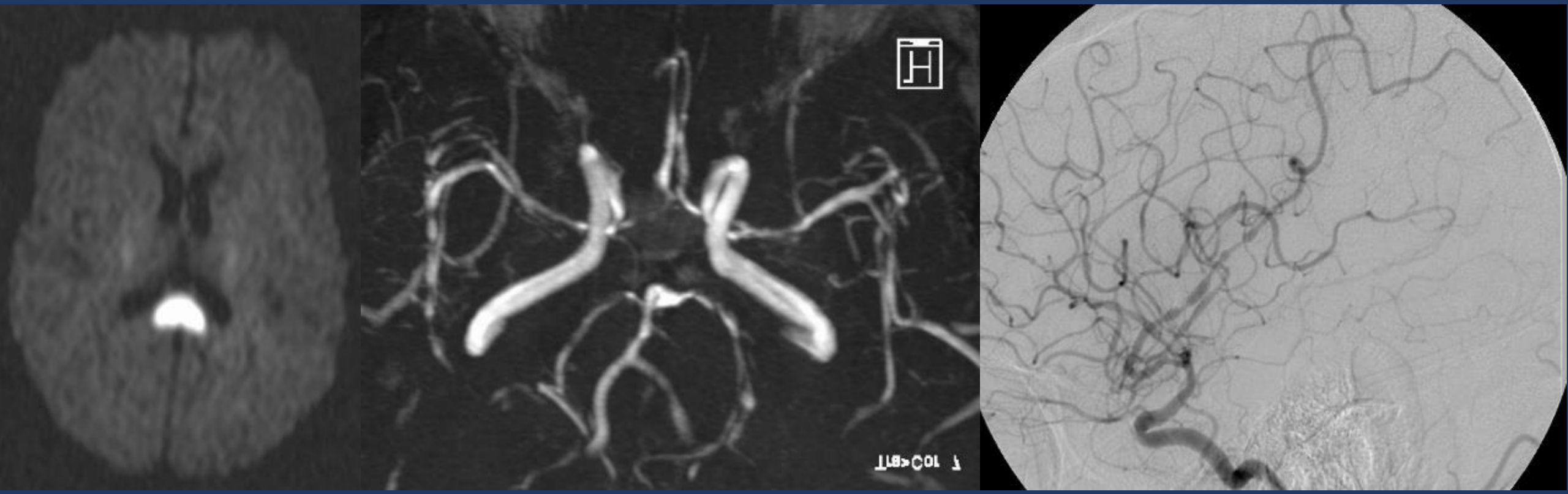
Granulomatosis with polyangiitis A.K.A. Wegener

Granulomatosis w/polyangiitis, Mostly Small Vessels, cANCA +

- Incidence: 30/1,000,000, both genders equal, 30-50 yr of age
- 10-45% affect head & neck, 5% affect brain
- Brain: infarcts, demyelinating-like lesions, large pituitary, pachymeningitis
- Chapel Hill criteria (3): positive histology, renal/pulmonary/laryngotracheal *and/or* +cANCA

AJR 2015; 205:619

Case 6. Severe post partum headaches.



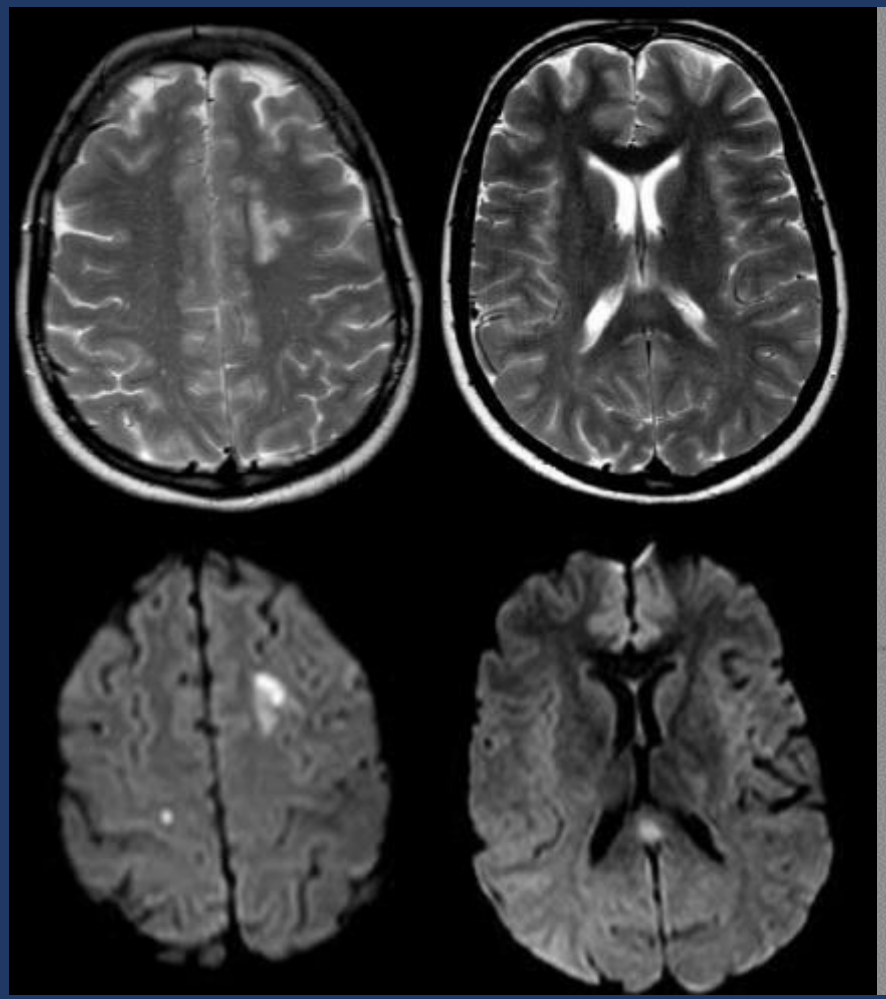
Reversible vasoconstriction syndrome

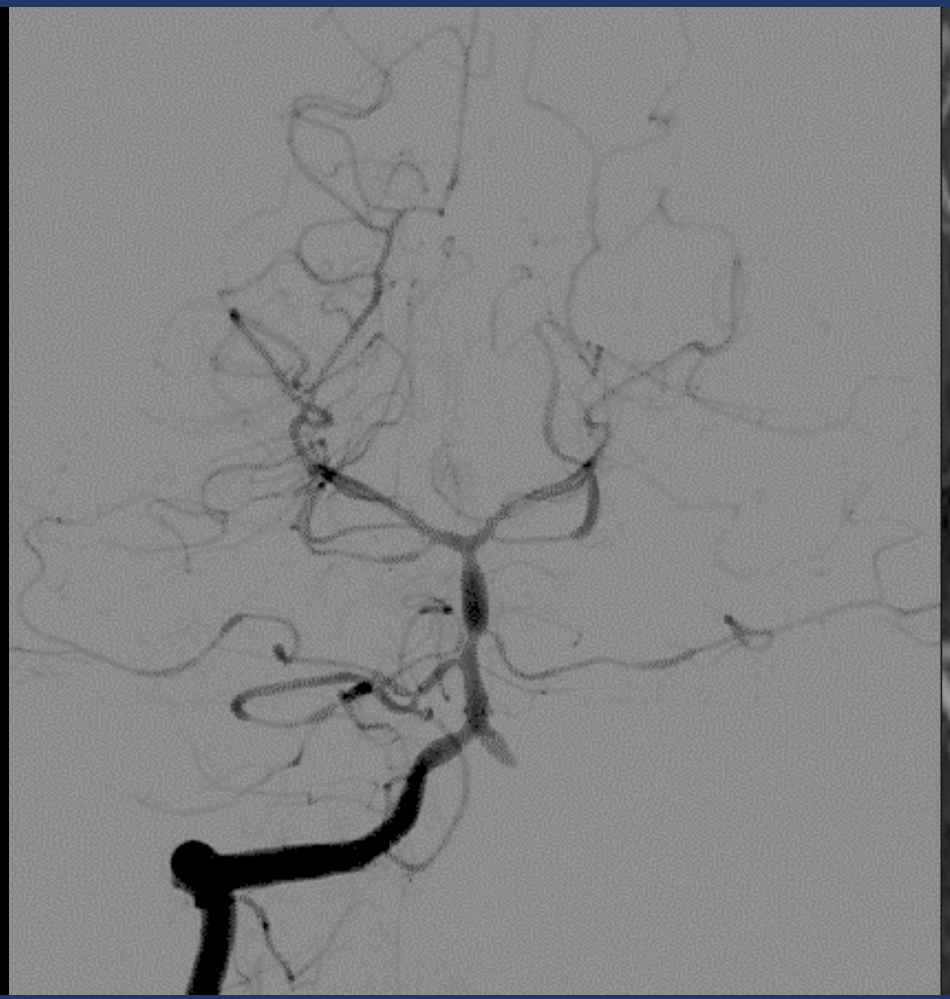
Post Partum Angiopathy (RVCS)

- Not “vasculitis”
- Rare, 1-2 weeks post partum (5 days)
- Symptoms: severe headaches, encephalopathy, seizures
- Predisposing factor: ergot in 30%
- MRI: normal in 70%, hemorrhage vasogenic edema (PRES-like) & bleeds in 30%
- DSA: abnormal in all

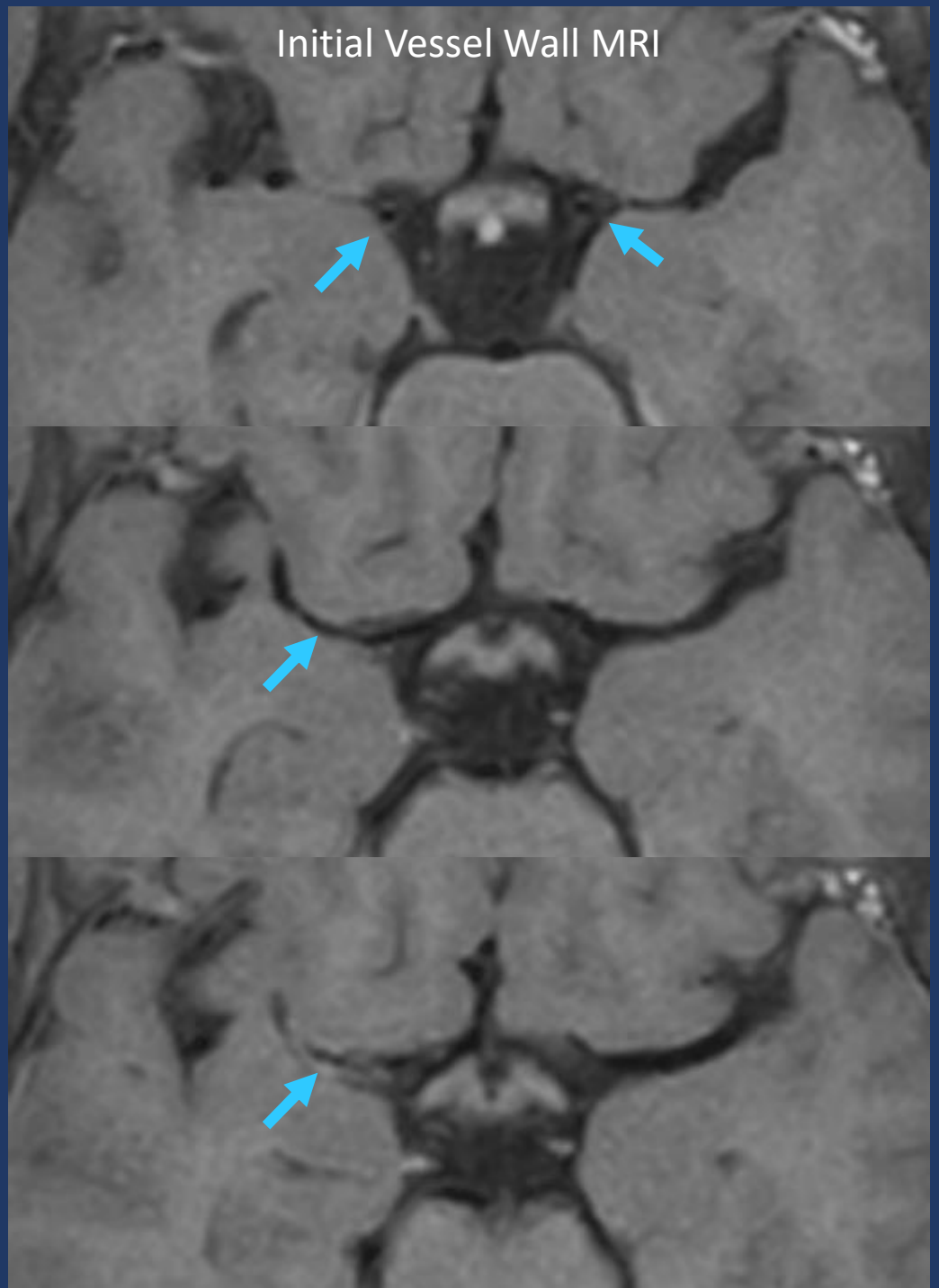
Stroke 2015; 43: 670

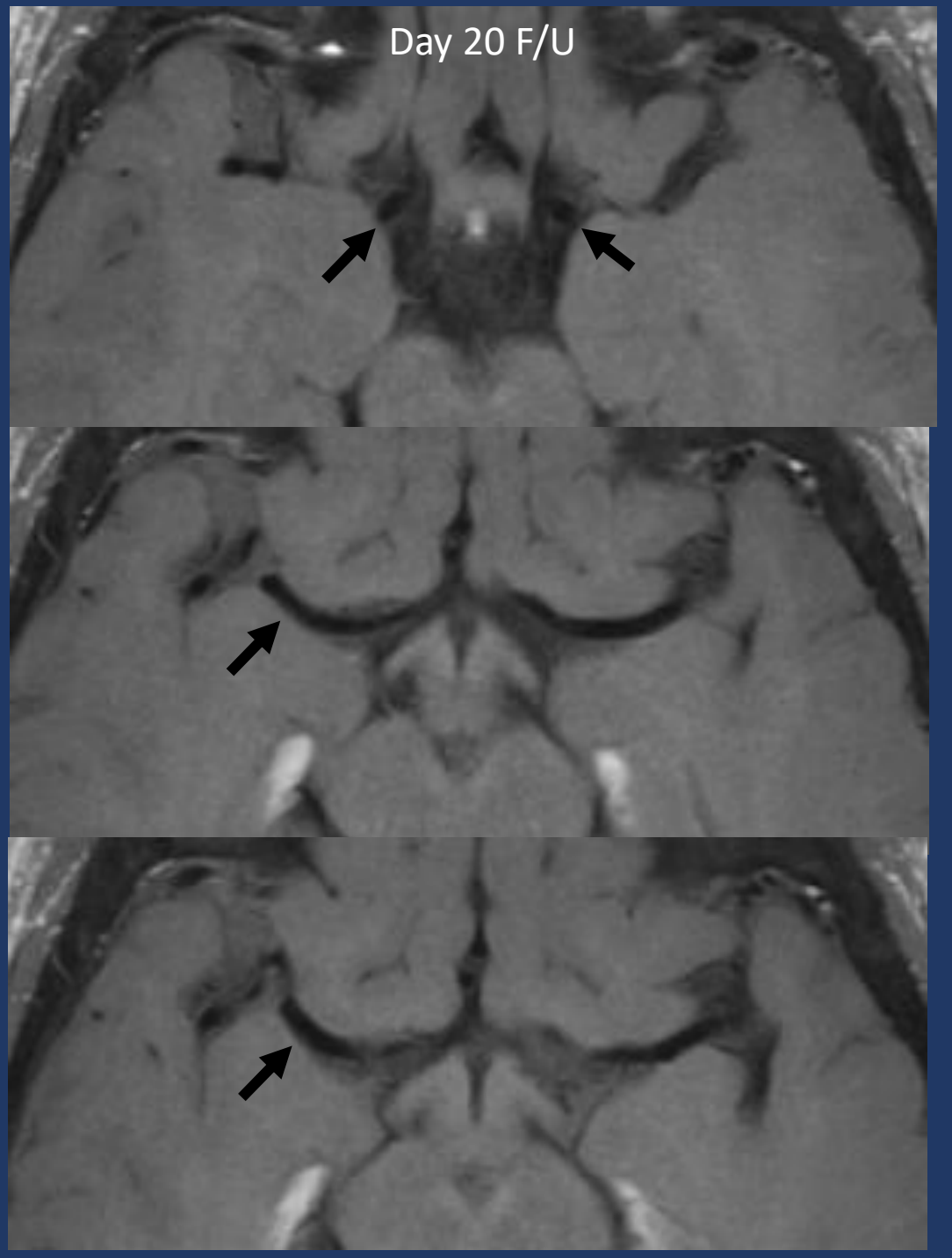
RVCS





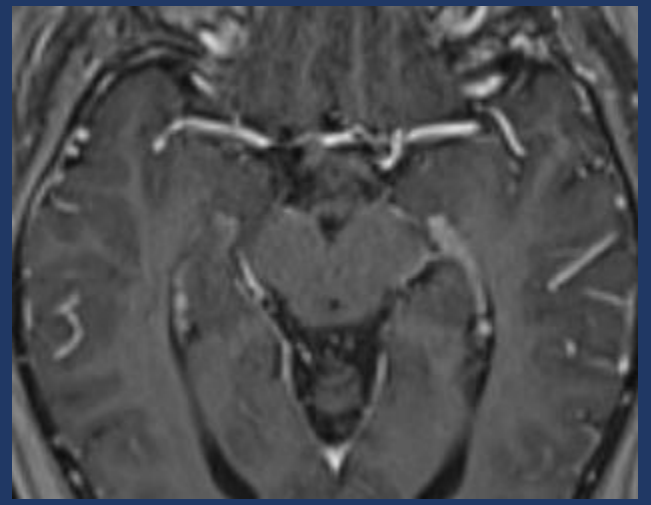
C/o H Sonwalkar, UK


Initial Vessel Wall MRI

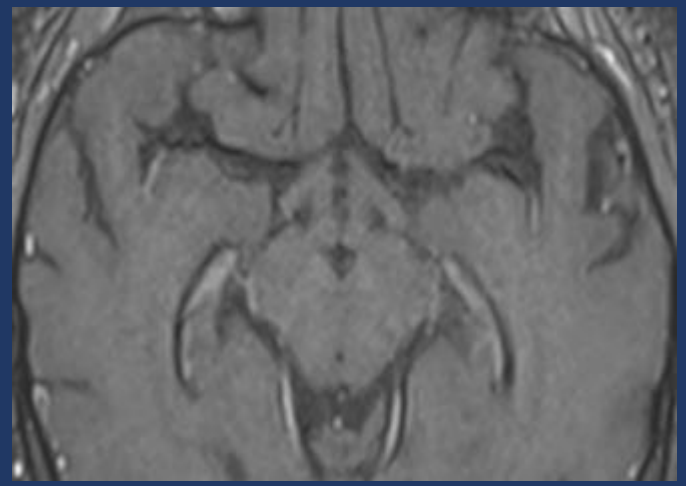

Day 20 F/U

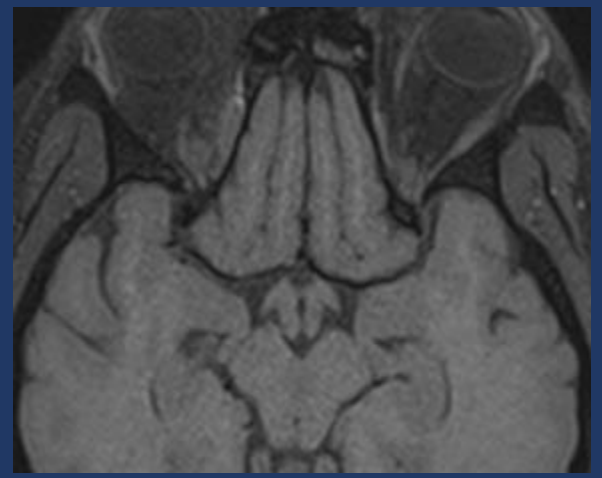
Vessel Wall MR Imaging

High res (MCA wall is 0.2-0.3 mm), 3D acquisition, low signal from CSF & flowing blood (T1 or proton density)



Better at 3T, isotropic voxel (0.4-0.7 mm)

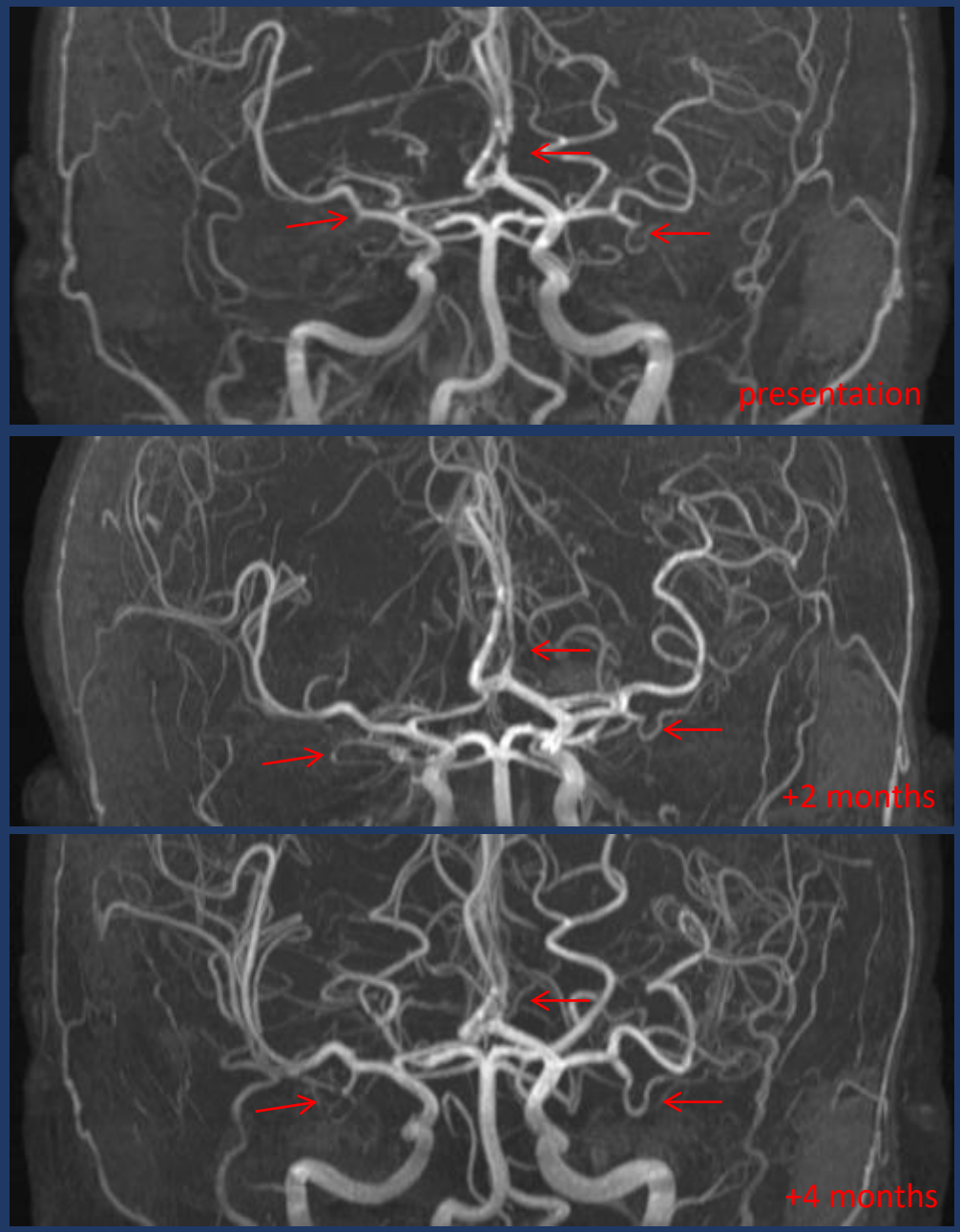


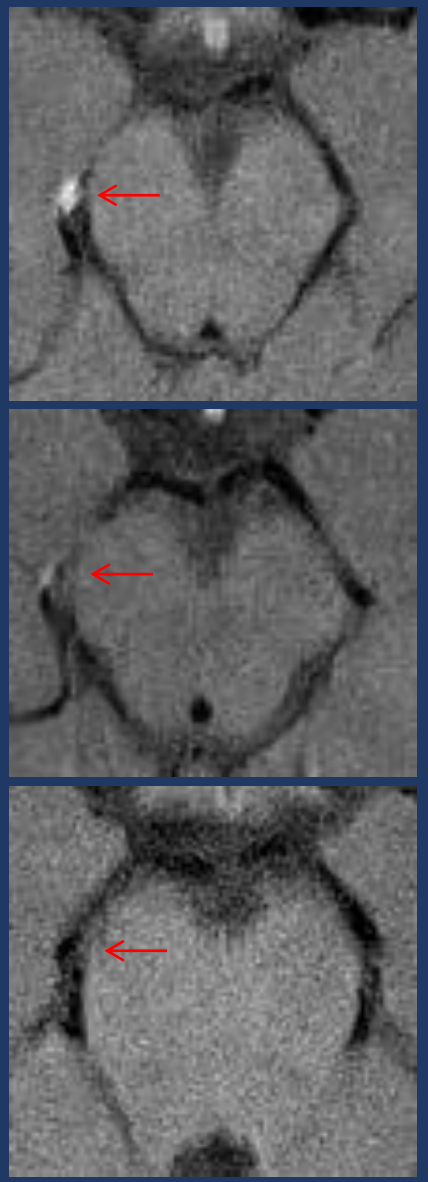


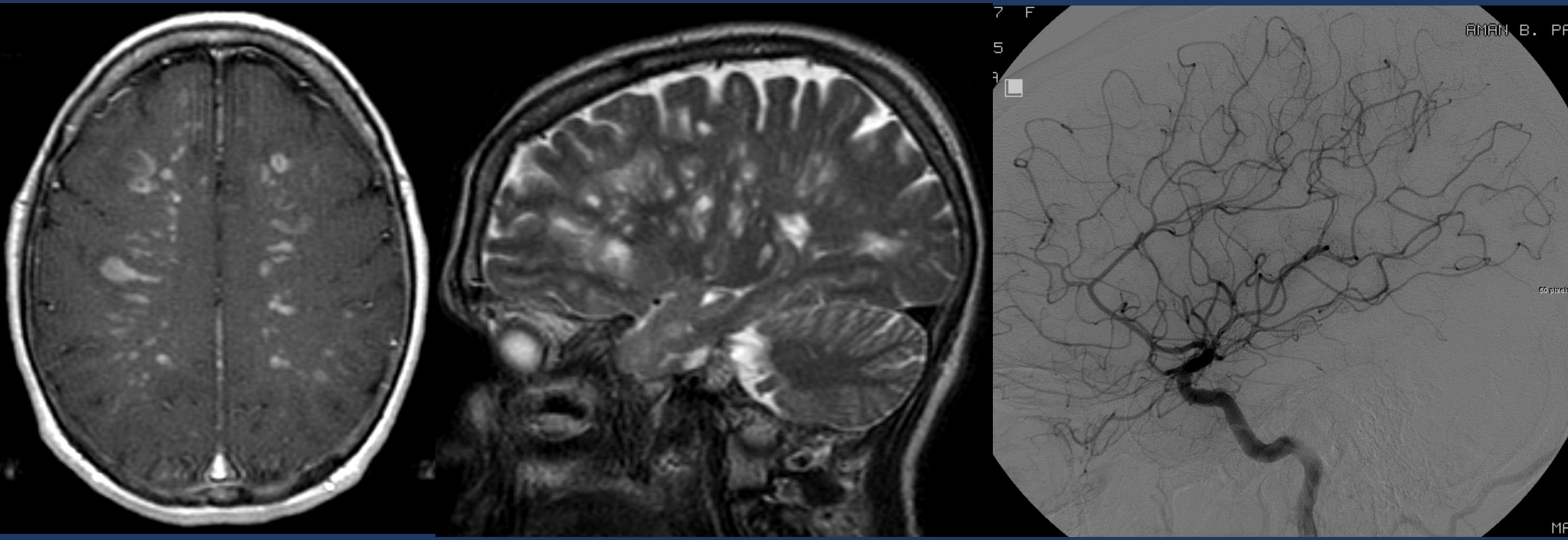
Longitudinal Findings

Clinical & angiographic features correlate

With treatment, vascular wall enhancement diminishes on VWMRI

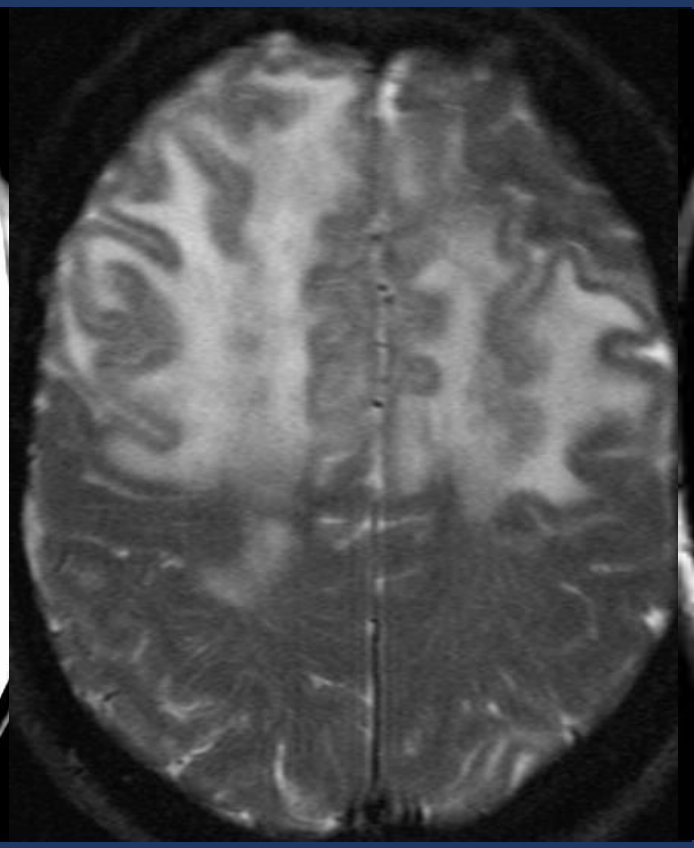




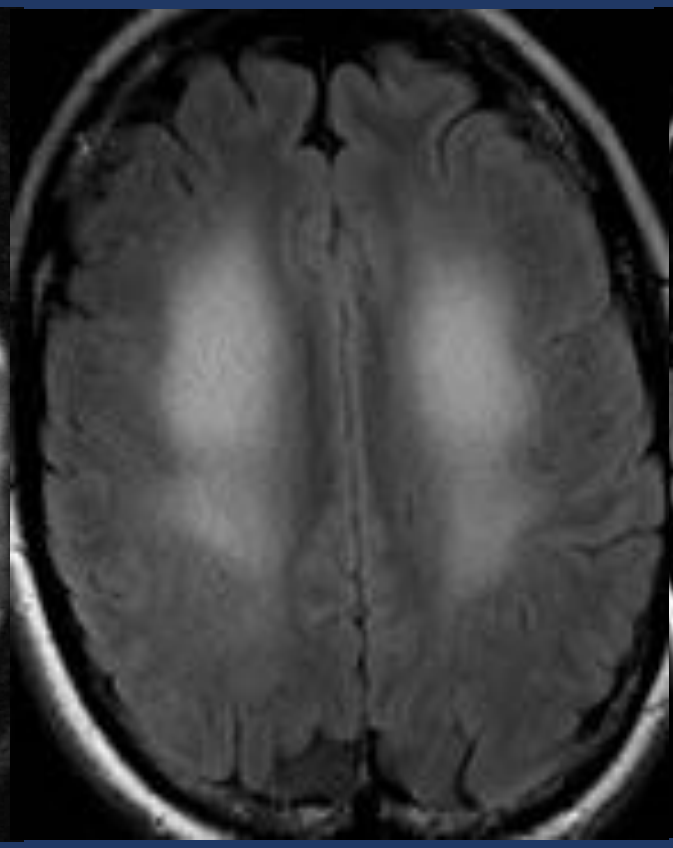


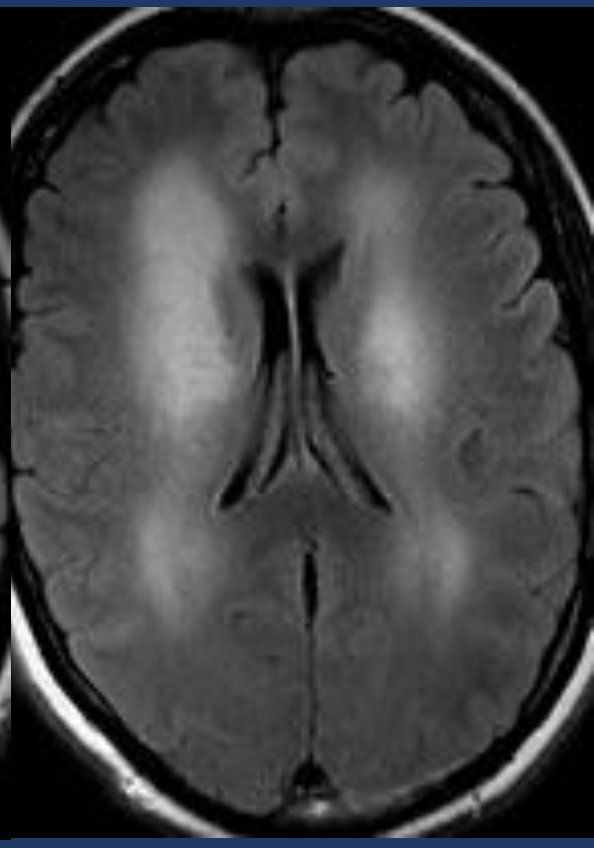
Smaller size vessels: DSA may be negative. PACNS

Again, diagnosis is difficult…

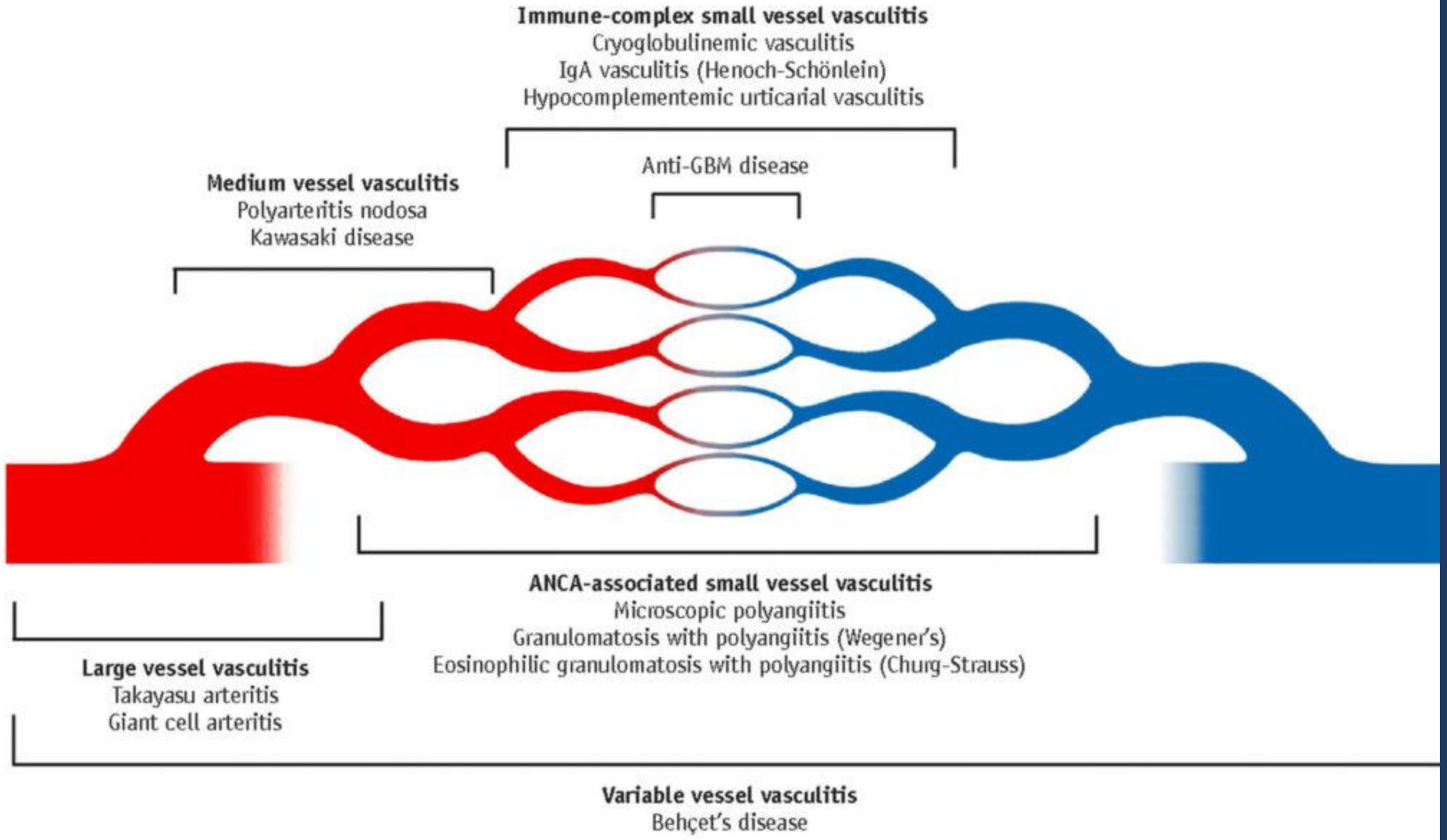


Granulamatous angiitis





SLE-related vasculitis


Immune-complex small vessel vasculitis
Cryoglobulinemic vasculitis
IgA vasculitis (Henoch-Schönlein)
Hypocomplementemic urticarial vasculitis
Anti-GBM disease
Medium vessel vasculitis
Polyarteritis nodosa
Kawasaki disease
ANCA-associated small vessel vasculitis
Microscopic polyangiitis
Granulomatosis with polyangiitis (Wegener's)
Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)
Large vessel vasculitis
Takayasu arteritis
Giant cell arteritis
Variable vessel vasculitis
Behçet's disease

Conclusions

- DX of vasculitis is very difficult
- Clinical suspicion & inflammatory markers, predisposing factors
- MRI (DWI, T2, FLAIR, perfusion) is a good to start
  - High resolution vessel wall imaging is best
- MRA is not optimal, need DSA
- Most patients will need biopsy for DX (100% diagnostic)

- Thank you: M Mossa Basha, R Nunez Hoffman, S Petcharunpaisan