

AMSER Case of the Month: May 2019

New Onset Headache with Sensorineural Hearing Loss

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Patient Presentation

- 43 year old male with 2 weeks of right frontal headache, right sided hearing loss, right eye vision changes “floaters”, nausea and vomiting, and encephalopathy
 - Denies photophobia, phonophobia, and fevers
- PMH: Type 2 Diabetes Mellitus, Hypertension, Hyperlipidemia
- FH:
 - Mother: Rheumatoid Arthritis
 - Cousin: Lupus
- Physical Exam:
 - Vitals: BP 134/95 T: 98.0 HR: 114 RR: 18 O2 Sat: 96%
 - Neuro: A&Ox1, visual acuity intact, EOMI, decreased auditory acuity R>L, MOCA 5/30

Pertinent Labs

- Serum glucose: 298
- WBC: 22.5
- ESR, CRP: wnl
- Lumbar puncture:
 - Glucose: 208 (consistent with serum)
 - Protein: 95
 - WBC: 0
 - RBC: 0

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Clinical Condition: Headache

Variant 13: New headache. Focal neurologic deficit or papilledema.

Radiologic Procedure	Rating	Comments	RRL*
MRI head without and with IV contrast	8		0
MRI head without IV contrast	7		0
CT head without IV contrast	7		☢☢☢
MRA head without and with IV contrast	6		0
CT head without and with IV contrast	6		☢☢☢
MRA head without IV contrast	5		0
CT head with IV contrast	5		☢☢☢
CTA head with IV contrast	5		☢☢☢

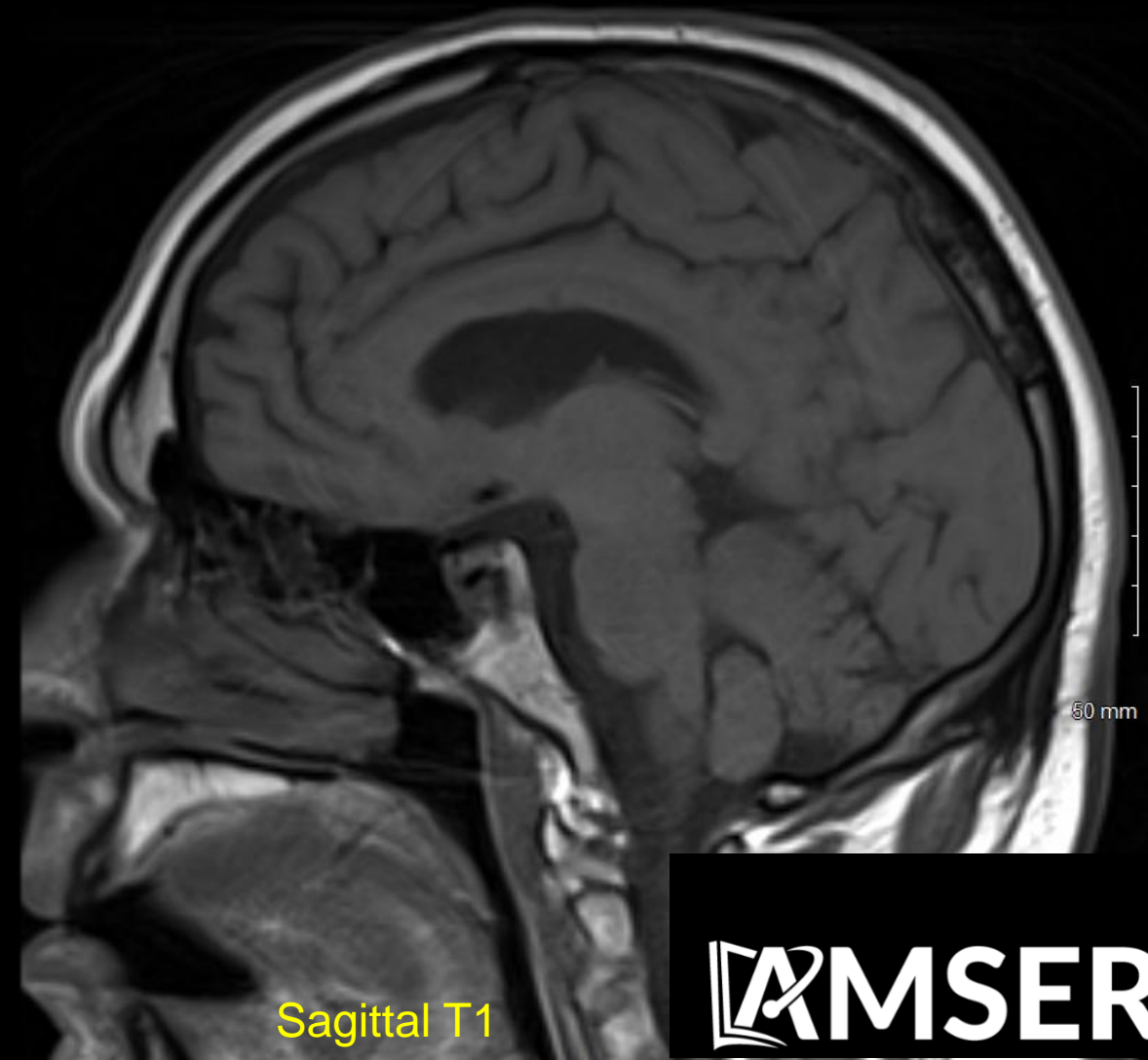
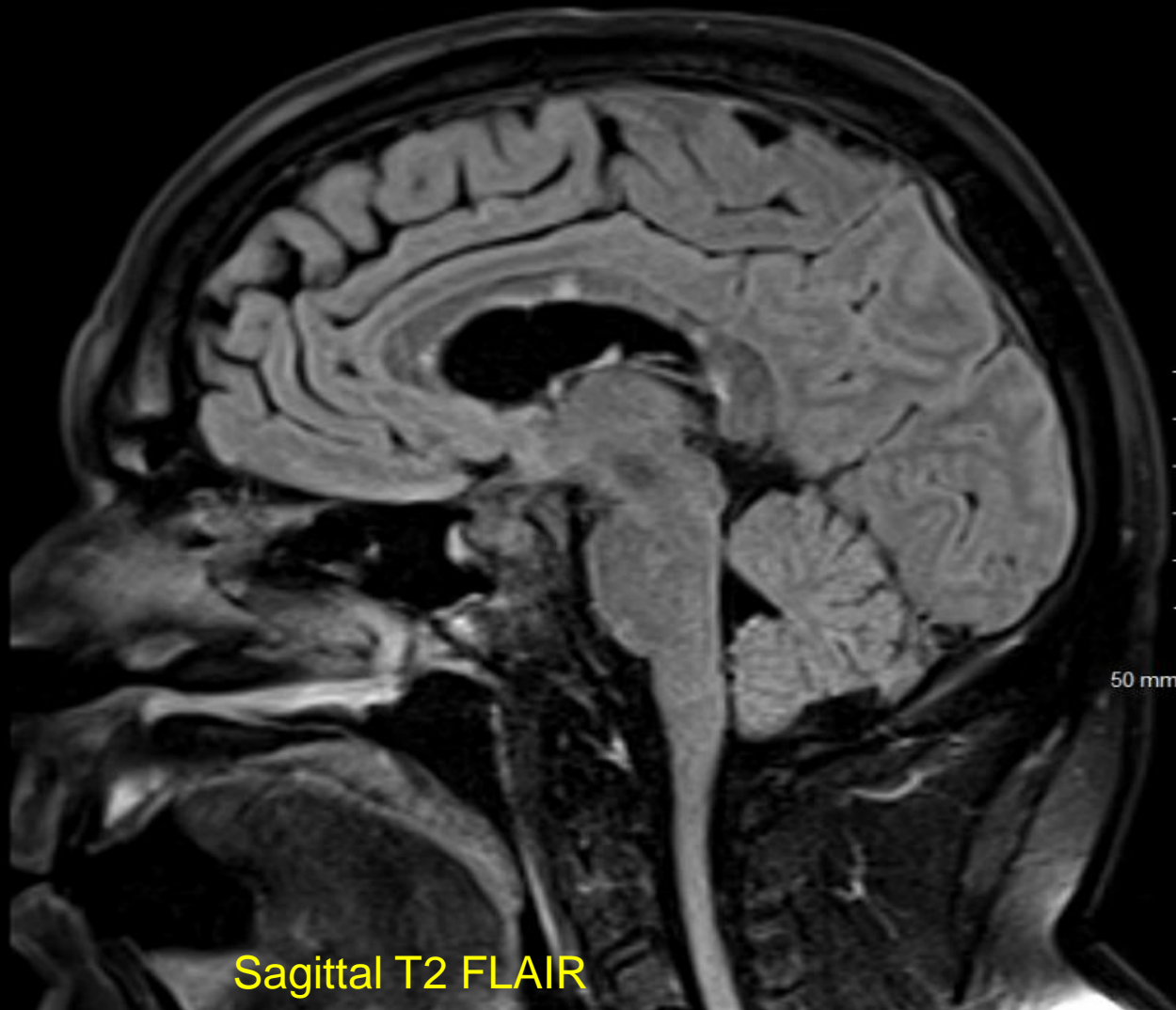
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate

*Relative Radiation Level

This imaging modality was ordered by the Neurology Physician

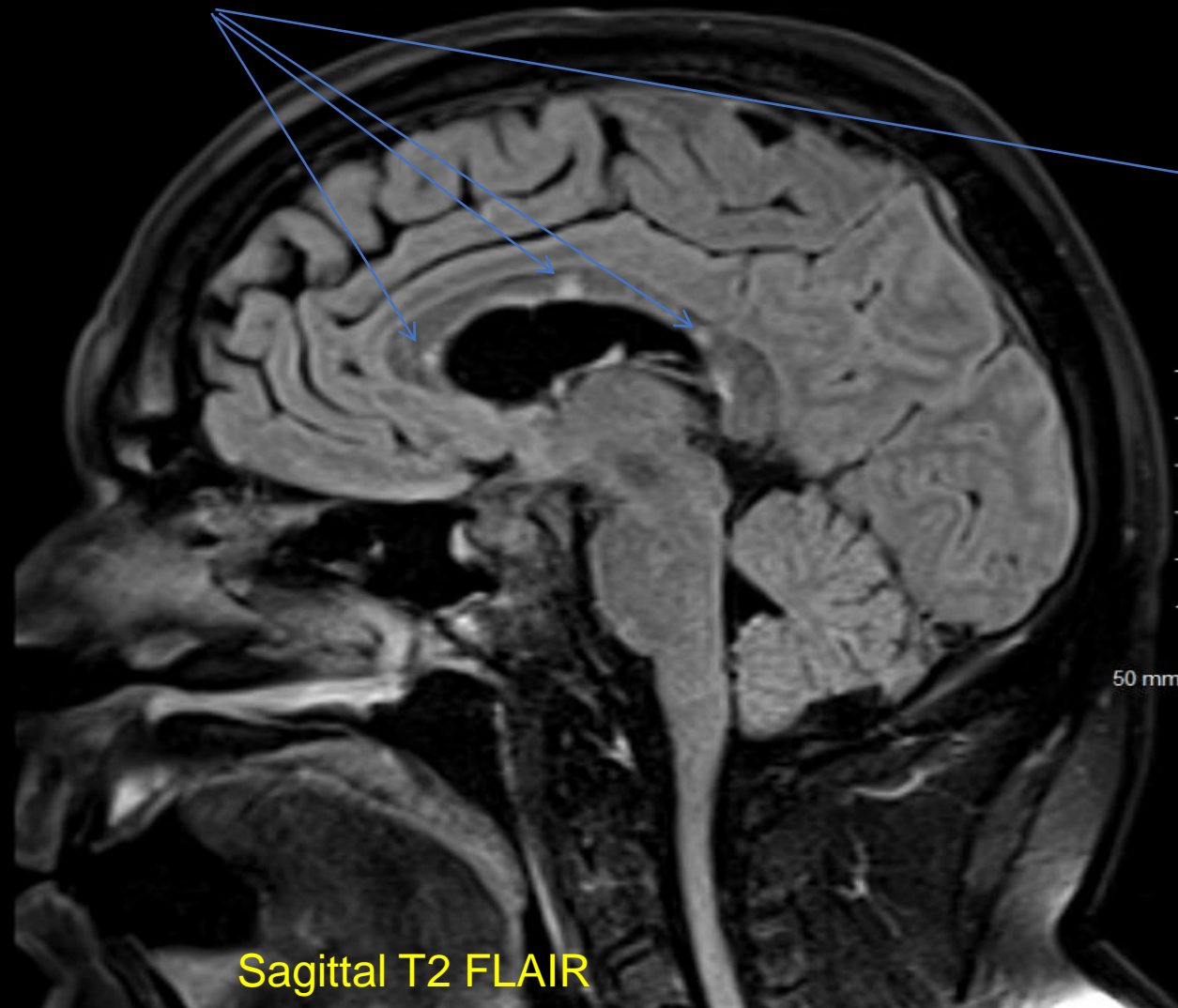


Findings (unlabeled)

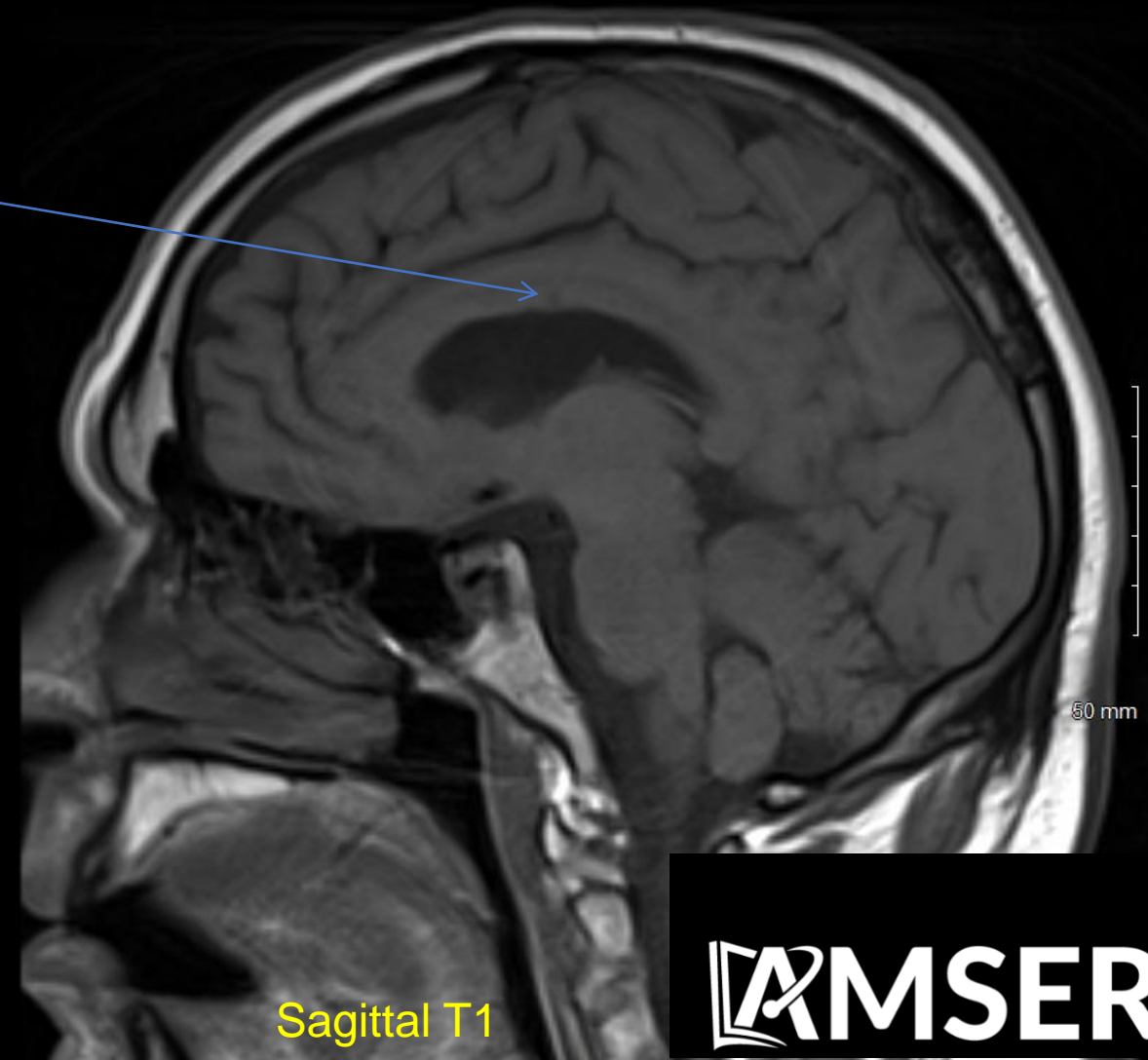


Findings (Labeled)

Multiple T2 hyperintense and T1 hypointense foci within the corpus callosum



Sagittal T2 FLAIR



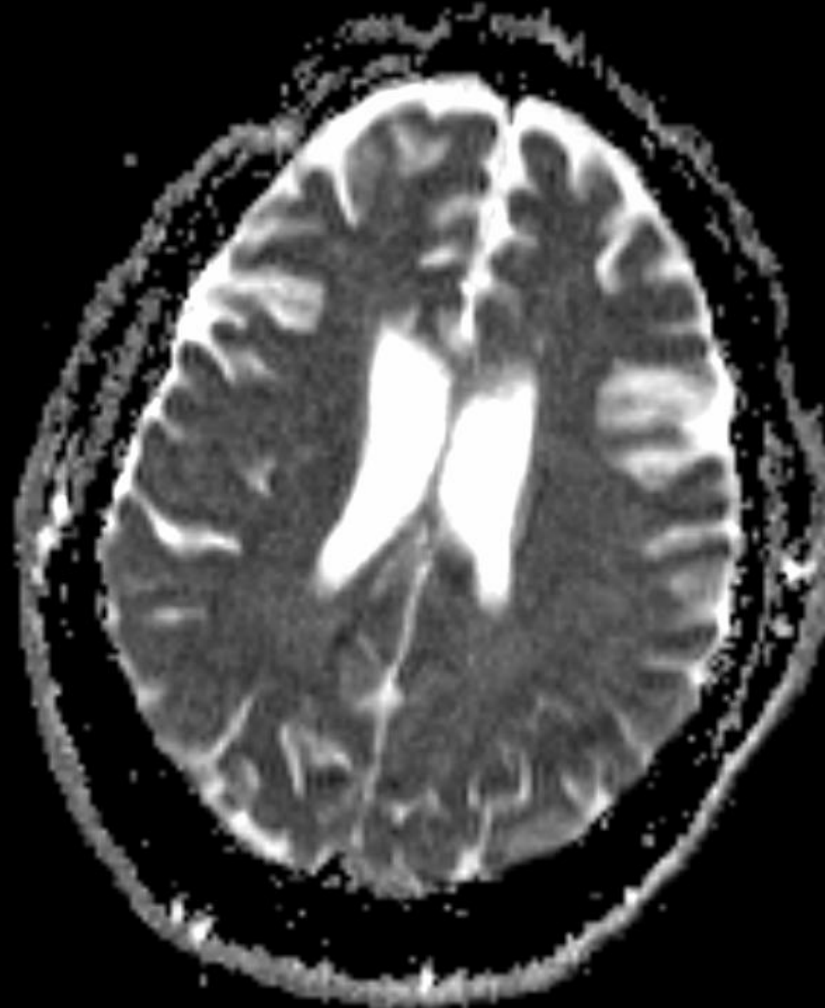
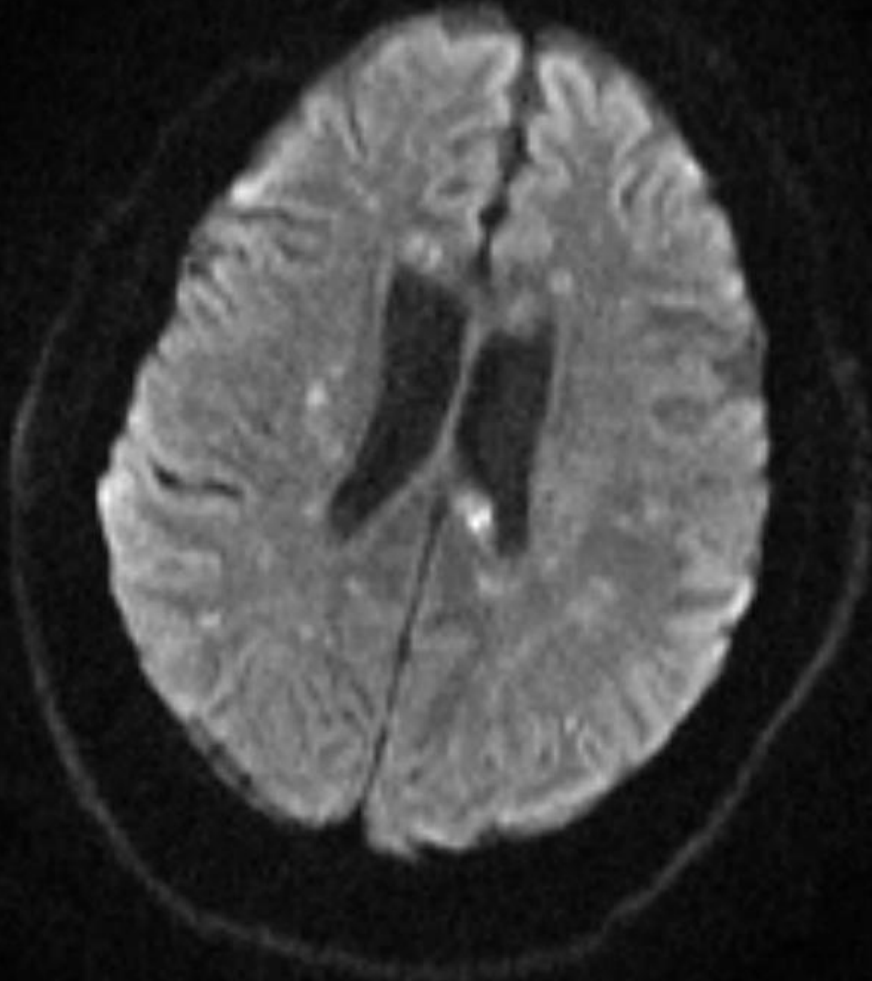
Sagittal T1



Findings (unlabeled)

Axial DWI

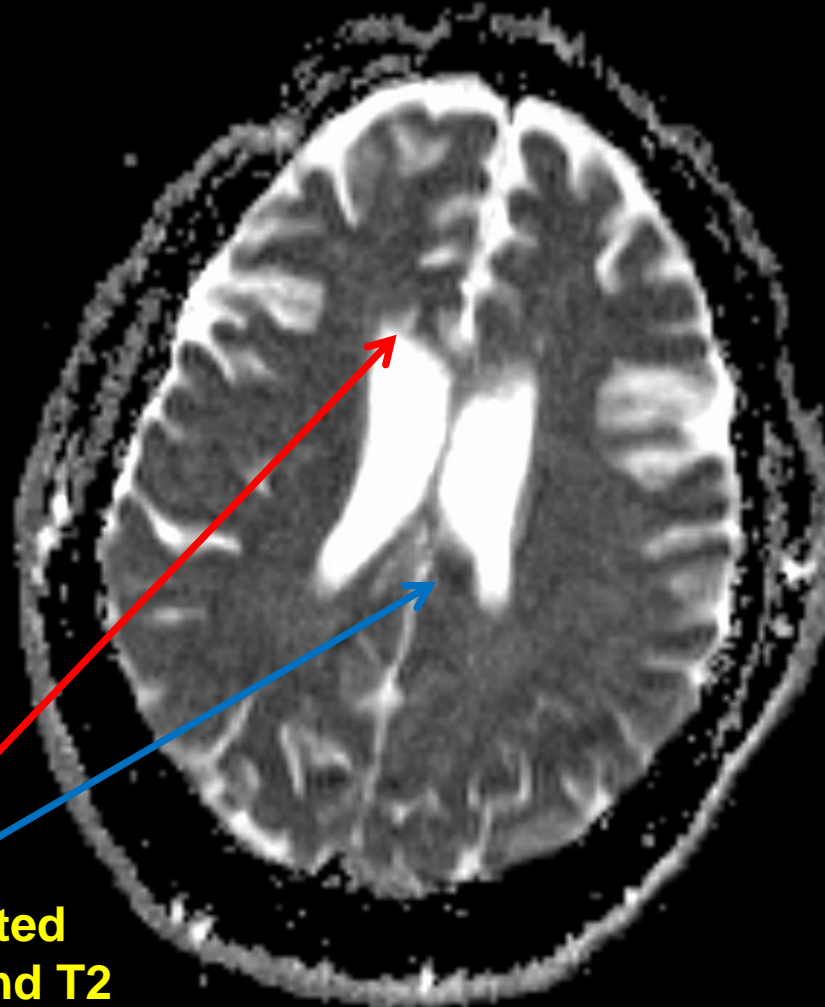
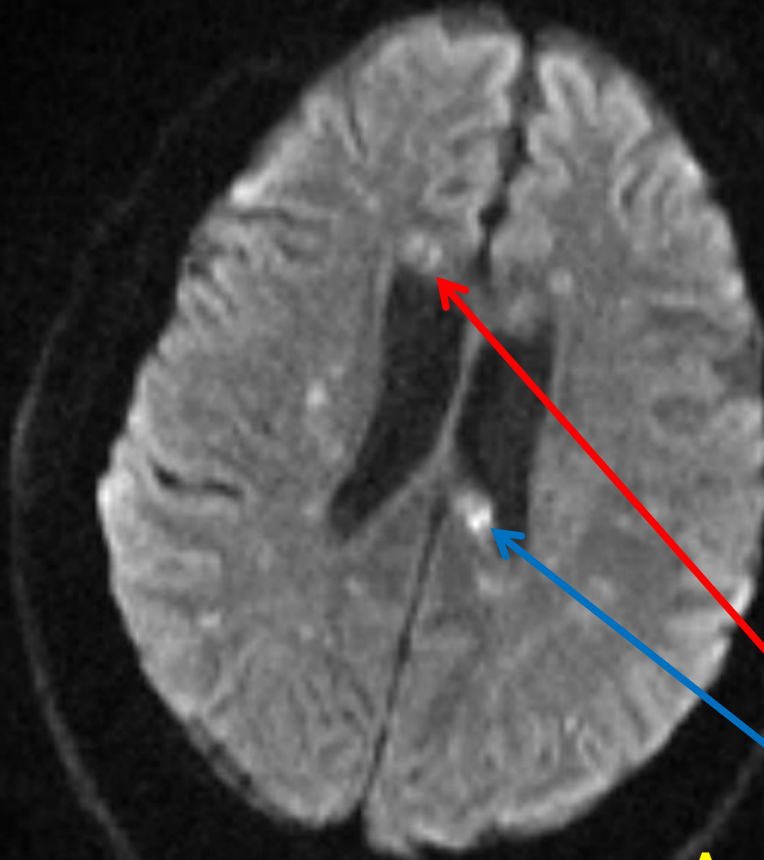
Axial ADC Map



Findings (labeled)

Axial DWI

Axial ADC Map



Areas of restricted diffusion (blue) and T2 shine through (red) within the corpus callosum

Differential Diagnosis:

- Multiple Sclerosis
- Thromboembolic Stroke (CADASIL)
- ADEM
- Susac's Syndrome
- SLE
- Migraine
- Meniere's Disease
- Schizophrenia

Final Dx:

Susac's Syndrome

Susac's Syndrome

- Triad of encephalopathy, sensorineural hearing loss, and branch retinal artery occlusions
- This patient's symptoms of encephalopathy, sensorineural hearing loss, and right sided visual changes with imaging findings favor diagnosis
- MRI is the modality of choice to diagnose and follow the disease process
- Classic imaging features:
 - T2/FLAIR:
 - "Snowballs" T2 hyperintense lesions in the body and splenium of the corpus callosum attributed to microinfarction
 - Callosal undersurface is typically spared as compared to multiple sclerosis
 - T2 lesions commonly located in the basal ganglia, which is unusual for multiple sclerosis
 - T1:
 - Chronic lesions can appear as "black holes"
 - Active lesions can have enhancement if acute
 - DWI/ADC
 - Commonly will present with restricted diffusion and/or T2 shine-through

Susac's Syndrome: Diagnostics

- Microangiopathy affecting the arterioles of the brain, cochlea, and retina
 - Antiendothelial antibodies may play a role
 - **Full triad only in 13% of patients on onset**
- Need fluorescein angiography to investigate branch retinal artery occlusions
- Audiogram
 - Typical low to mid tone hearing loss
 - Our patient has had multiple audiograms showing moderate, bilateral, low frequency sensorineural hearing loss
- Lumbar Puncture
 - Typically mild elevation in protein count and nucleated cell count¹
 - Rarely oligoclonal bands
- Treatment
 - Acute: High dose corticosteroids often with immunomodulators
 - Intratympanic dexamethasone may help with acute sensorineural hearing loss
 - Maintenance: Corticosteroids w/ immunomodulators
 - Mycophenolate, IV Ig, Methotrexate, Azathioprine, Cyclophosphamide, Rituximab

References:

1. Dörr J, Krautwald S, Wildemann B, et al. Characteristics of Susac syndrome: a review of all reported cases. *Nature reviews. Neurology* 2013;9: 307–16.
2. Kleffner I, Dörr J, Ringelstein M for the European Susac Consortium (EuSaC), et al Diagnostic criteria for Susac syndrome *J Neurol Neurosurg Psychiatry* 2016;87:1287-1295.
3. Vodopivec, I. & Prasad, S. *Curr Treat Options Neurol* (2016) 18: 3.
4. Case 142: Susac Syndrome. Mustafa Kemal Demir. *Radiology* 2009 250:2, 598-602
5. Crawley BK, Close A, Canto C et-al. Susac's syndrome: intratympanic therapy for hearing loss and a review of the literature. *Laryngoscope*. 2009;119 (1): 141-4.