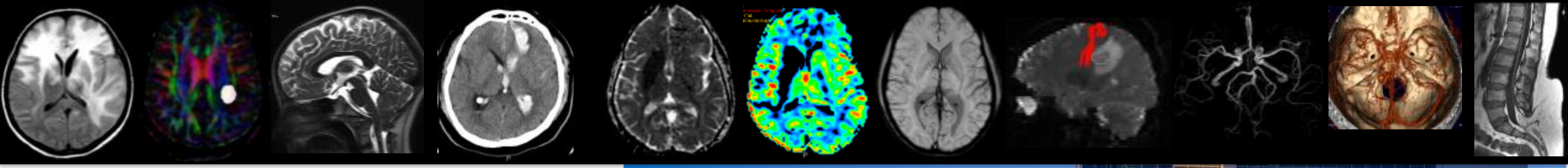
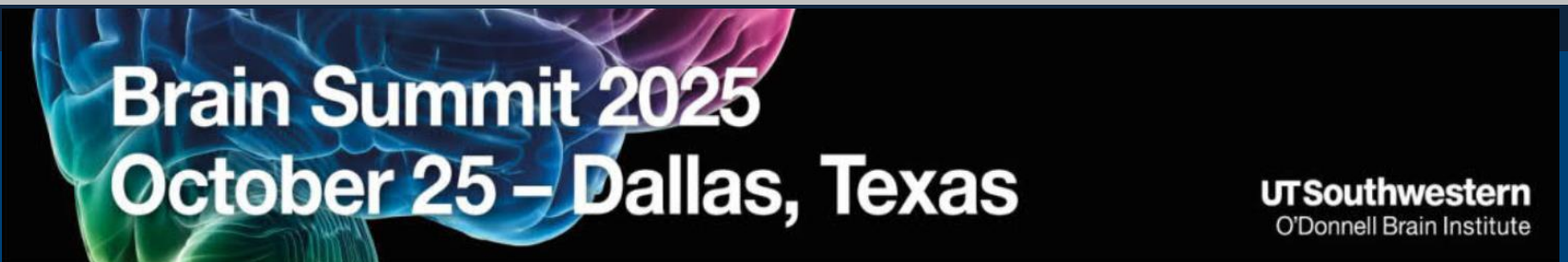


Neuroimaging Case-Based Review

Movement Disorder Edition

Marco C Pinho
Associate Professor in Radiology
UTSW Medical Center and Parkland Hospital & Health System



UTSouthwestern
Medical Center

Aims and Outline

Goals:

- review select neurological case scenarios in which a combination of clinical manifestations and imaging findings which may help in a specific diagnosis. Emphasis on movement disorders (mostly).

-

Format:

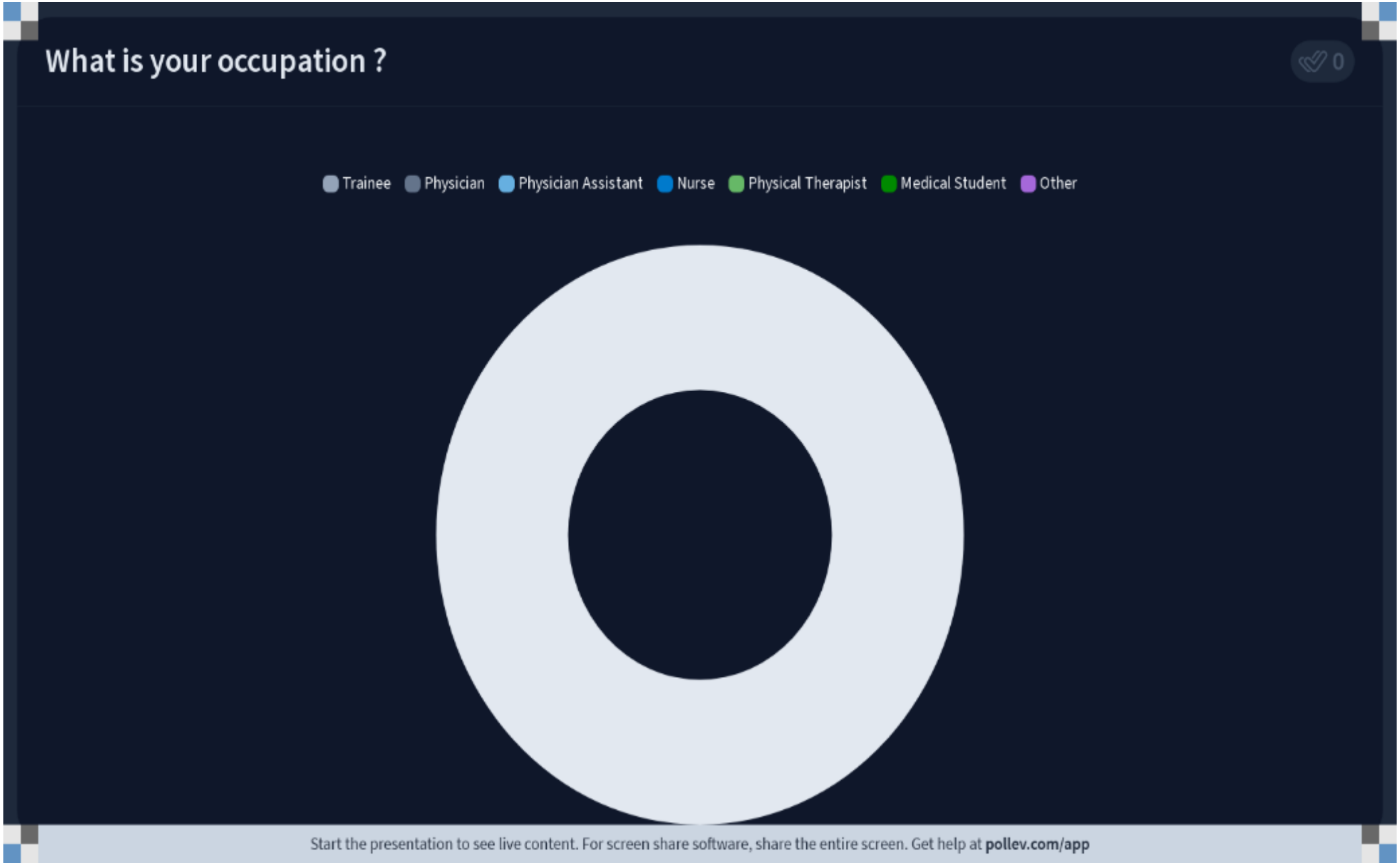
- **Cased-based discussion**
- **Clinical vignettes, selected images**
- **Interactive polling platform – engagement and self assessment**

How to Join

1. Go to **PollEv.com**
2. Enter **MARCOPINHO535**
3. Respond to activity

Disclosures

- No financial relationships to disclose
- Polling tool just for engagement, don't take it too seriously



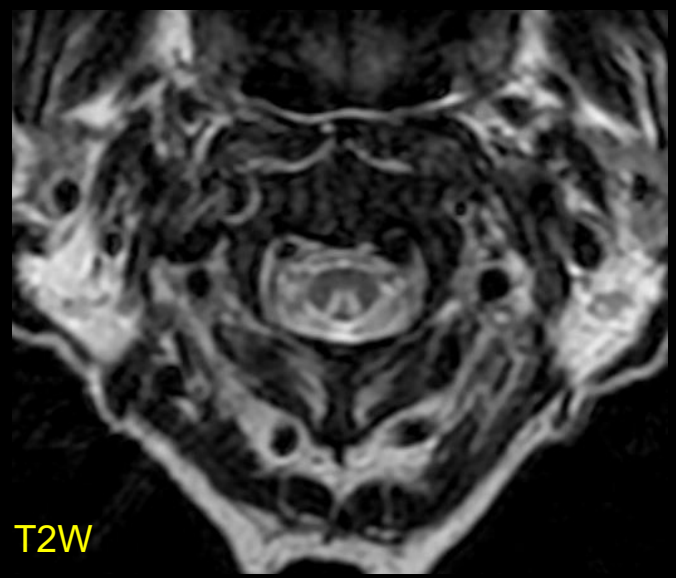
Training!

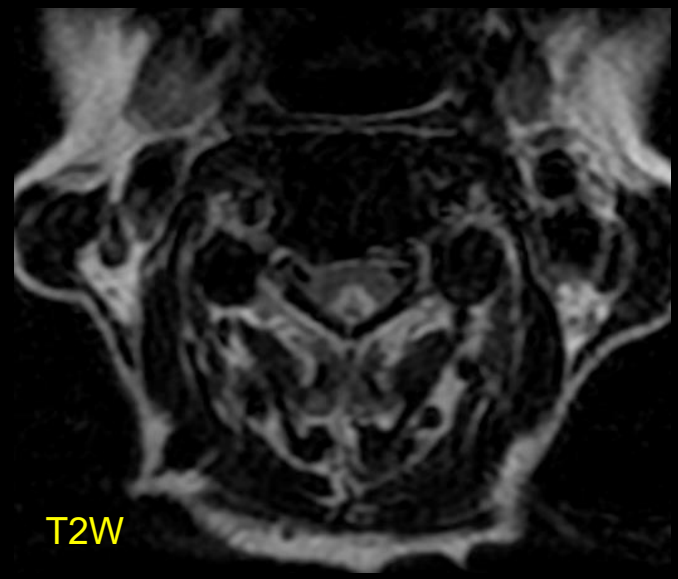
UT Southwestern Medical Center
Radiology

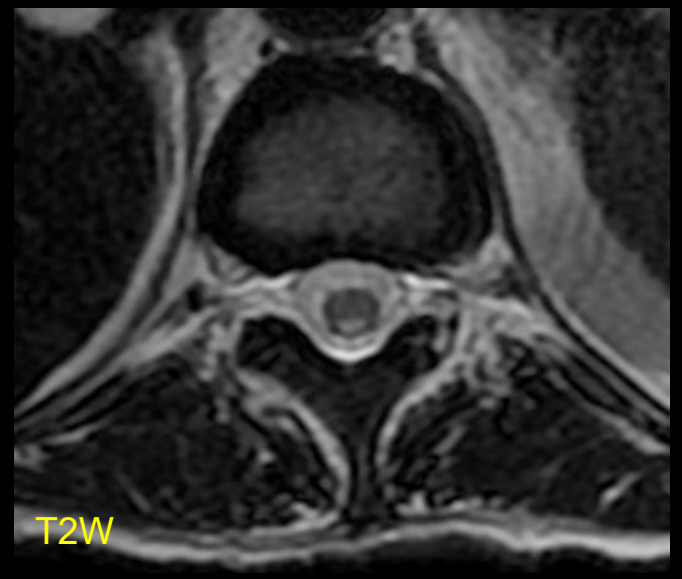
Warm up Case !!

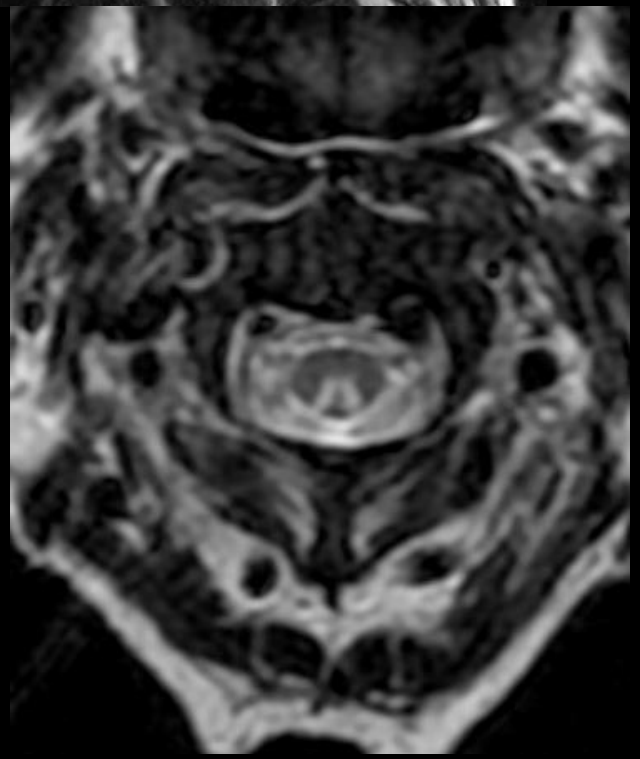
40 yo M.
Progressive paresthesia of the hands and feet, ataxia.

Hx of longstanding inflammatory bowel disease and prior surgeries.









40 yo M. Progressive paresthesias of the hands and feet, ataxia.

Hx of longstanding inflammatory bowel disease and prior surgeries.

Case #2 - What is the correct statement about this condition?

0

This is most commonly an idiopathic condition with spontaneous resolution — 0%

CSF in this patient will likely be abnormal — 0%

This disease is potentially treatable and intervention can prevent progression — 0%

Evaluation of the brain is recommended to search for additional periventricular lesions — 0%

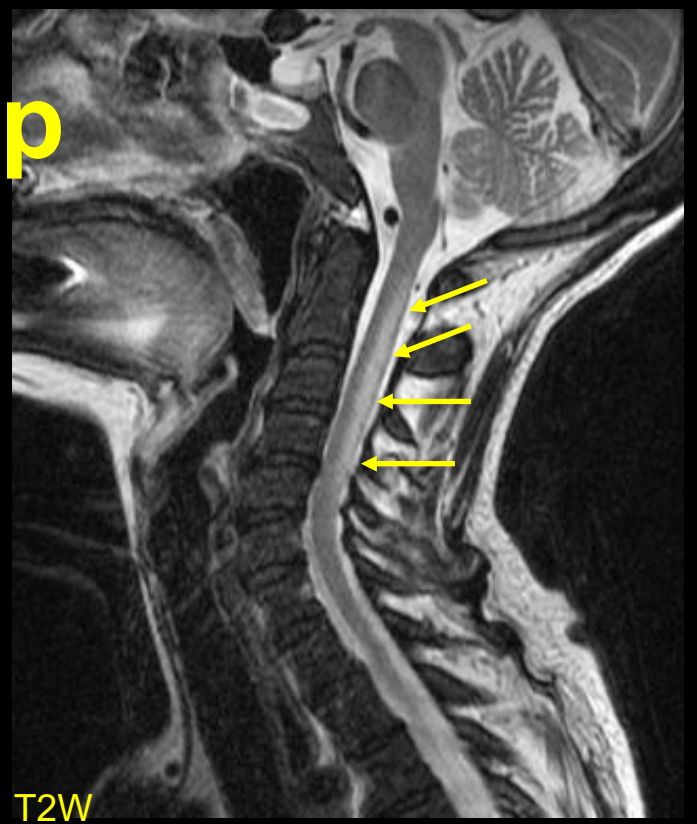
DWI would be helpful to confirm the most likely diagnosis — 0%

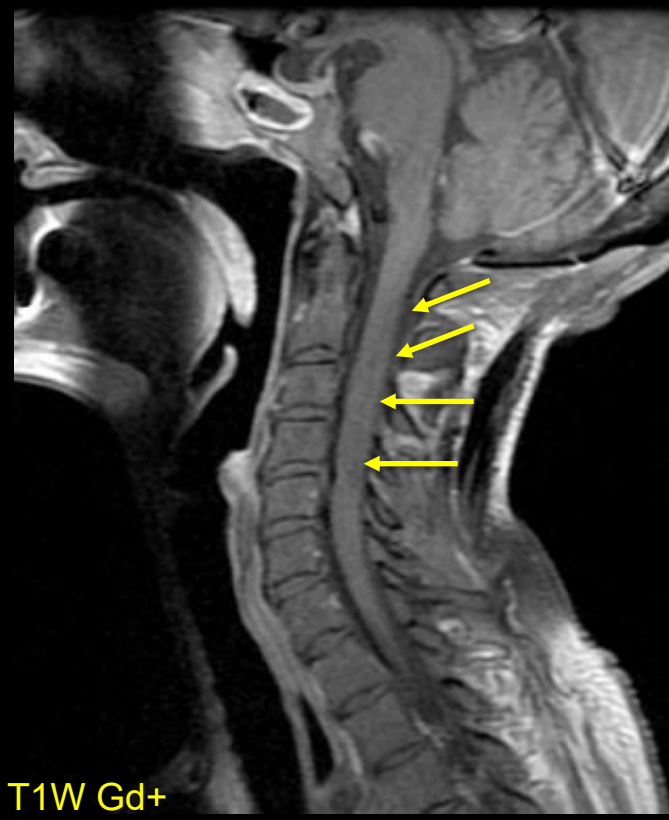
Start the presentation to see live content. For screen share software, share the entire screen. Get help at **pollev.com/app**

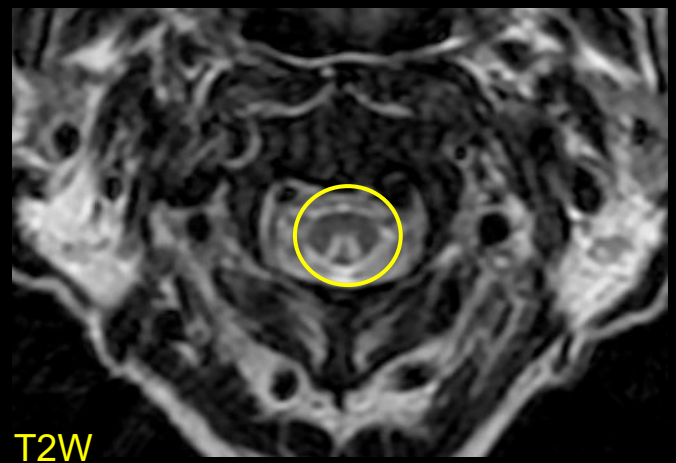
Warm Up

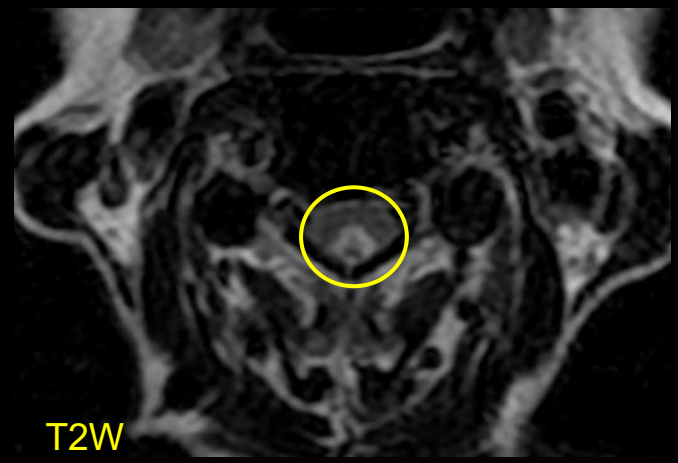
40 yo M. Progressive paresthesias of the hands and feet, ataxia.

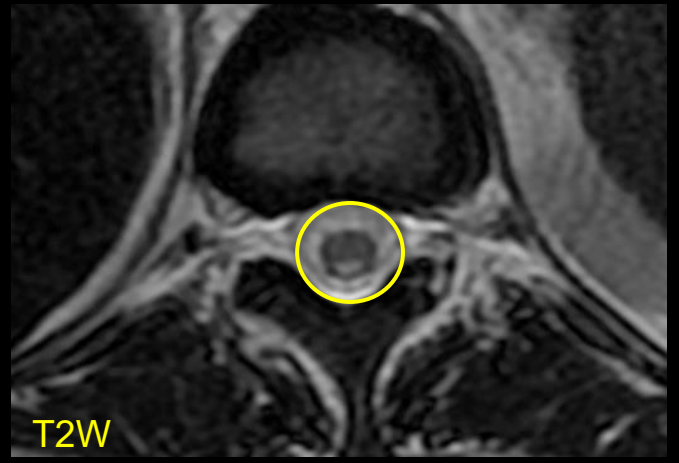
Hx of longstanding inflammatory bowel disease and prior surgeries.



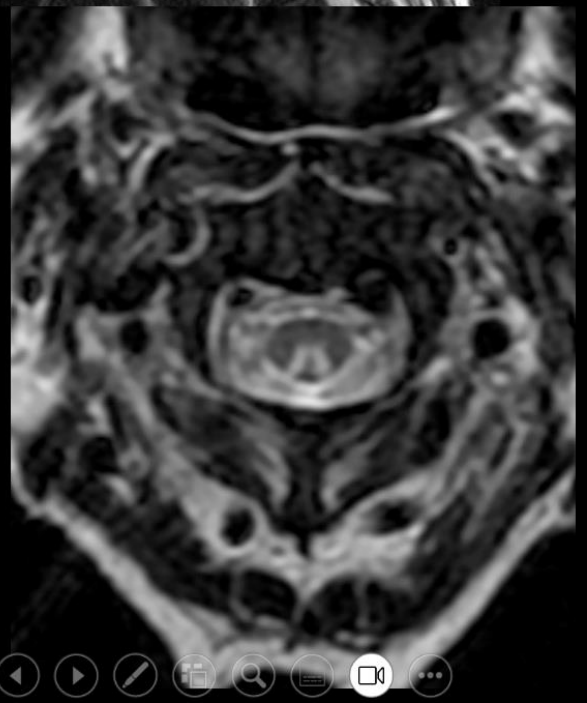








Subacute Combined Degeneration – B12 Deficiency



40 yo M. Progressive paresthesias of the hands and feet, ataxia.

Hx of longstanding inflammatory bowel disease and prior surgeries.

What is the correct statement about this condition?

0

This is most commonly an idiopathic condition with spontaneous resolution **(A)**

CSF in this patient will likely be abnormal **(B)**

✓ This disease is potentially treatable and intervention can prevent progression **(C)**

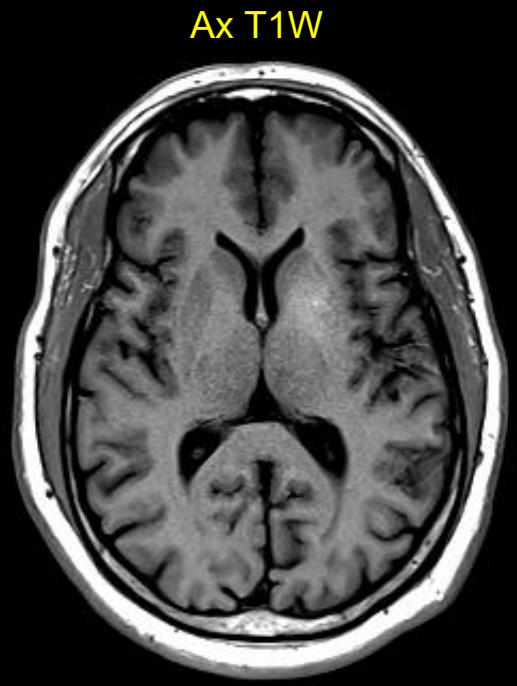
Evaluation of the brain is recommended to search for additional periventricular le… **(D)**

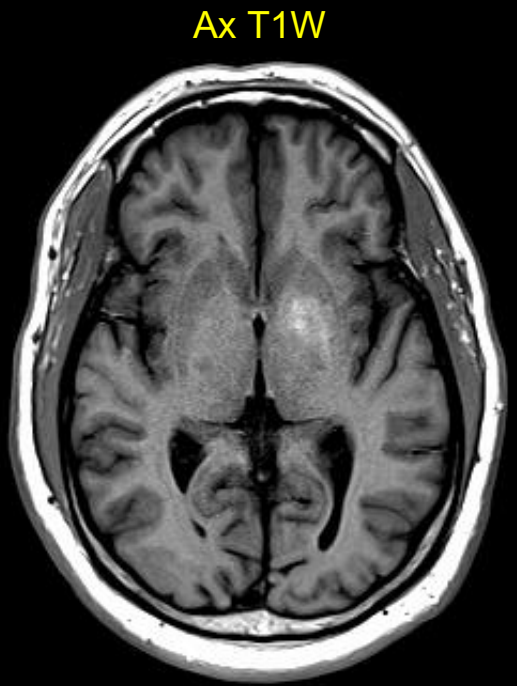
DWI would be helpful to confirm the most likely diagnosis **(E)**

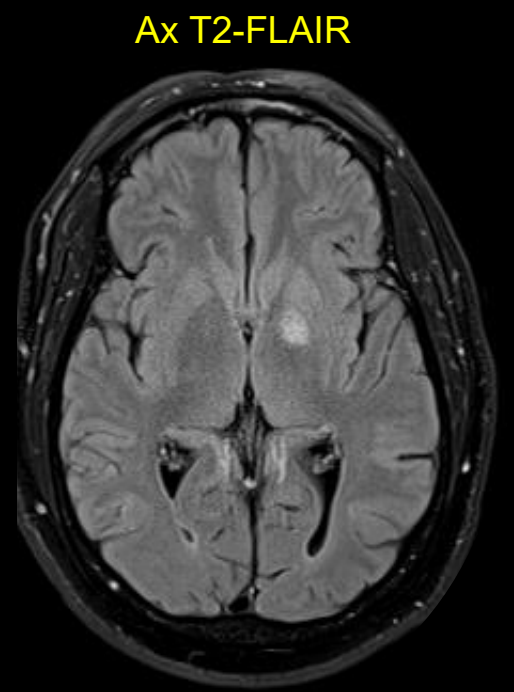
Case #1 – Sudden Abnormal Arm Movements

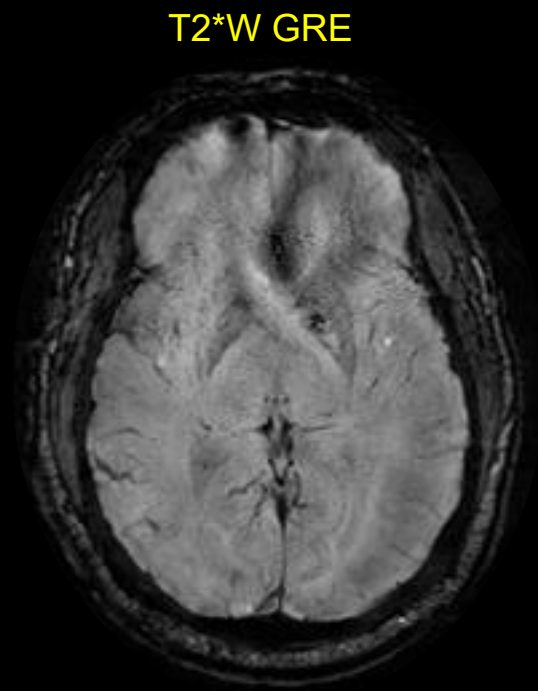
- 56 yo right male, presents to ED with 2 weeks of abnormal movements on RUE.
- Patient in usual state of health until woke up with burning pain on the RUE nd abnormal movements.
- Initial mild weakness, now resolved, followed by uncontrollable movements of shoulder, elbow wrist and fingers. Outside MRI saw "spot on the brain". Referred for stroke eval
- **Physical:** Irregular, fluid involuntary movements of RUE: trap elevation/ twitching, occasionally with flexion R elbow and writhing of wrists and fingers.
- **PMHx:** hypertension, T2DM, hyperlipidemia
- **Labs:** glucose (random): 246 mg/dL, Hgb A1c 10.2%

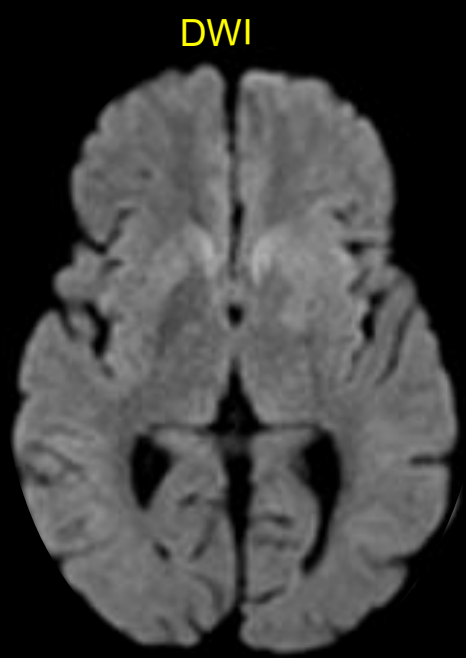
Case #1

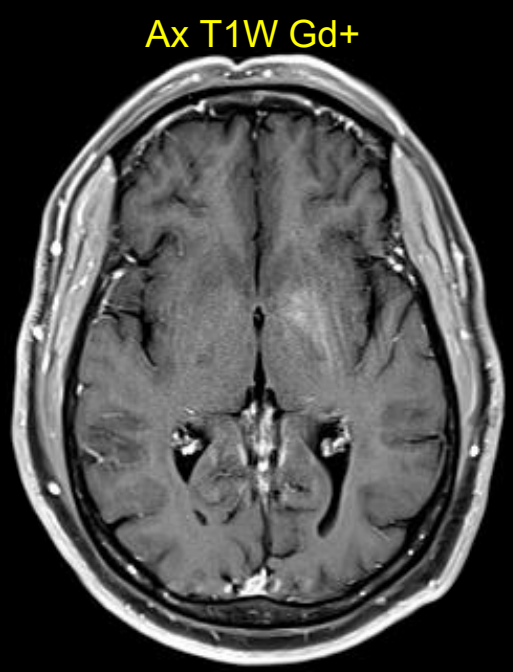


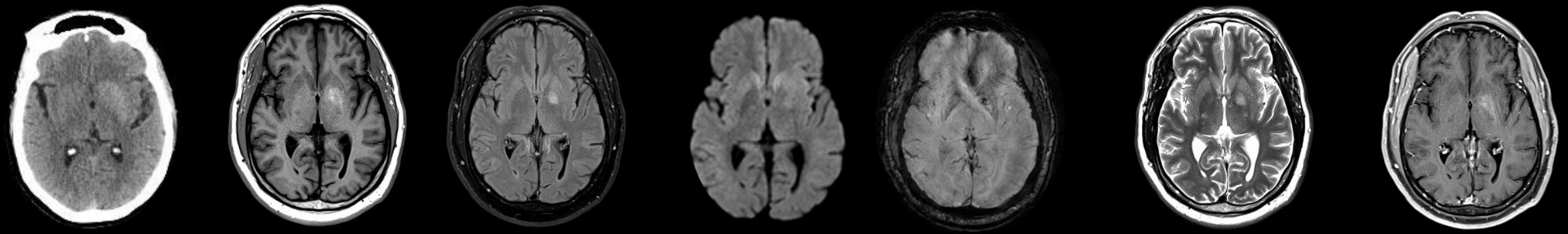












Case #1

Sudden Abnormal Arm Movements

Case #1 - What is the most likely etiology for this type of injury ?

0

Metabolic 0%

Vascular 0%

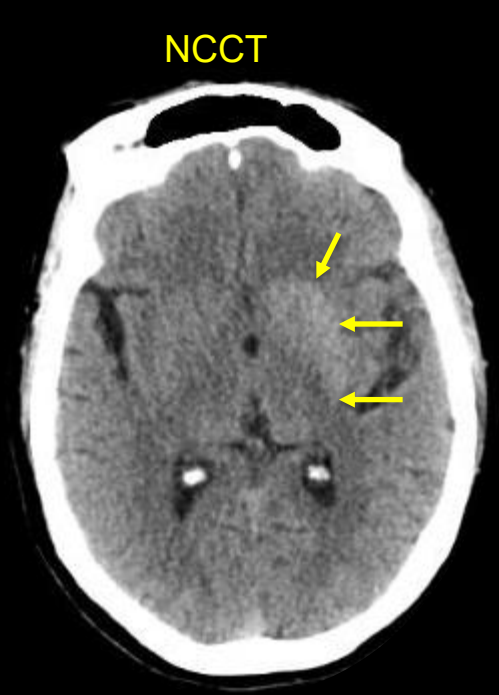
Neoplastic 0%

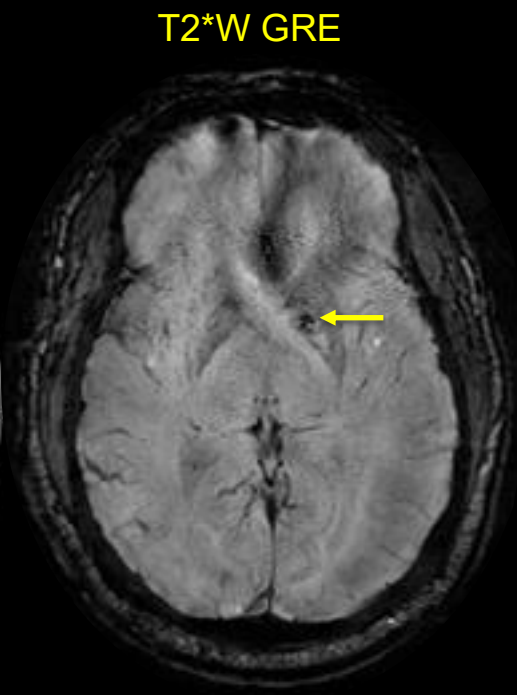
Immune-mediated 0%

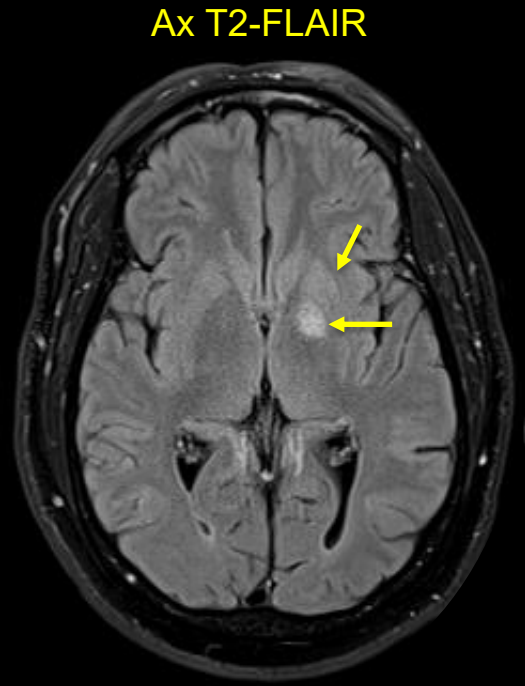
Infectious 0%

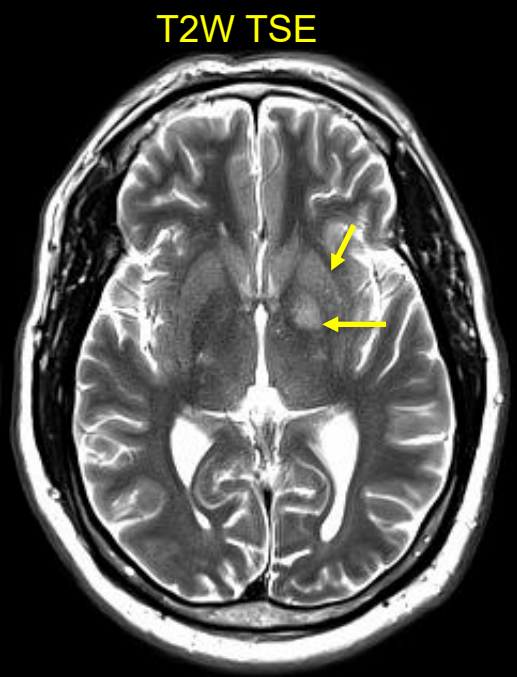
Start the presentation to see live content. For screen share software, share the entire screen. Get help at **pollev.com/app**

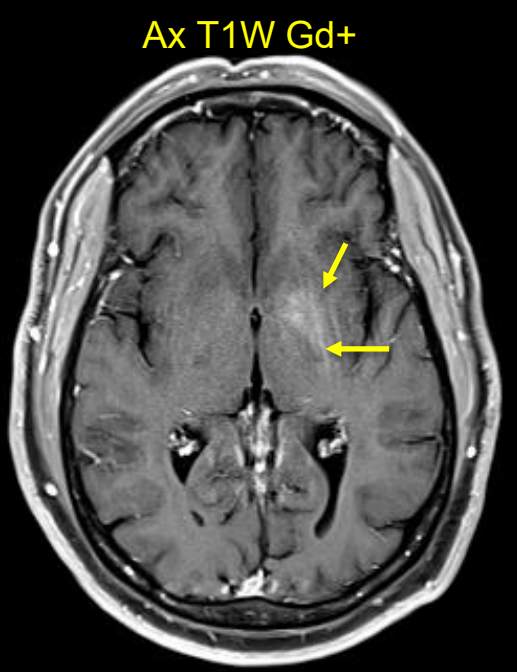
Case #1



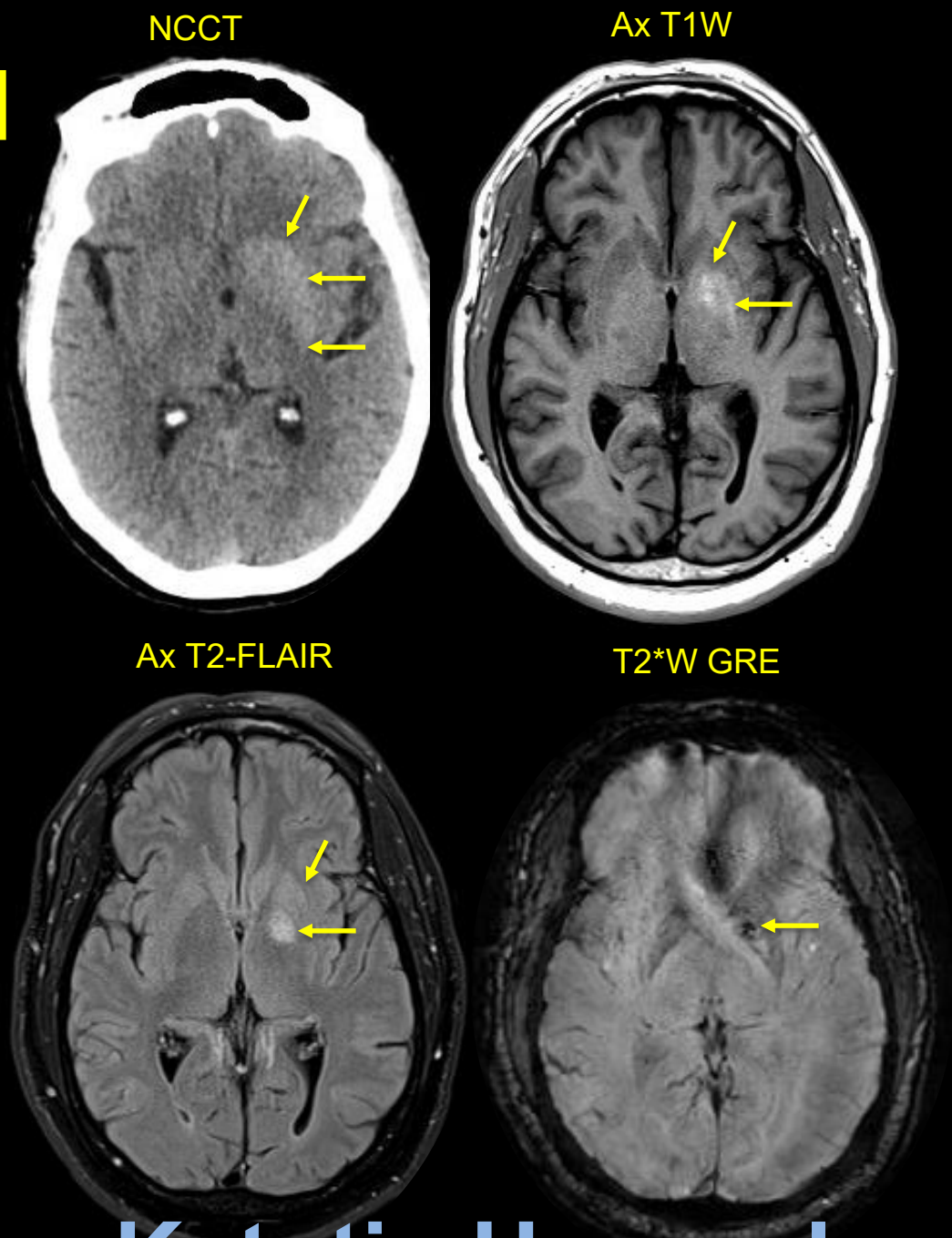








Case #1



Case #1 Diagnostic Checklist

- 56 yo M, sudden onset unilateral arm involuntary movements
- previously asymptomatic
- DW2, Hgb A1c 10.2%
- Imaging
  - unilateral BG signal abnormality
  - Elevated attenuation on CT
  - T1 shortening on MRI

Non-Ketotic Hyperglycemic Hemichorea

Nonketotic Hyperglycemic Hemichorea

- aka Diabetic Striatopathy or Chorea, Hyperglycemia Basal Ganglia Syndrome (C-H-BG)
- Rare, under recognized neurological complication of nonketotic hyperglycemia
- # non-ketotic hyperglycemic seizures and non-ketotic hyperglycemic coma

Typical Clinical Presentation:

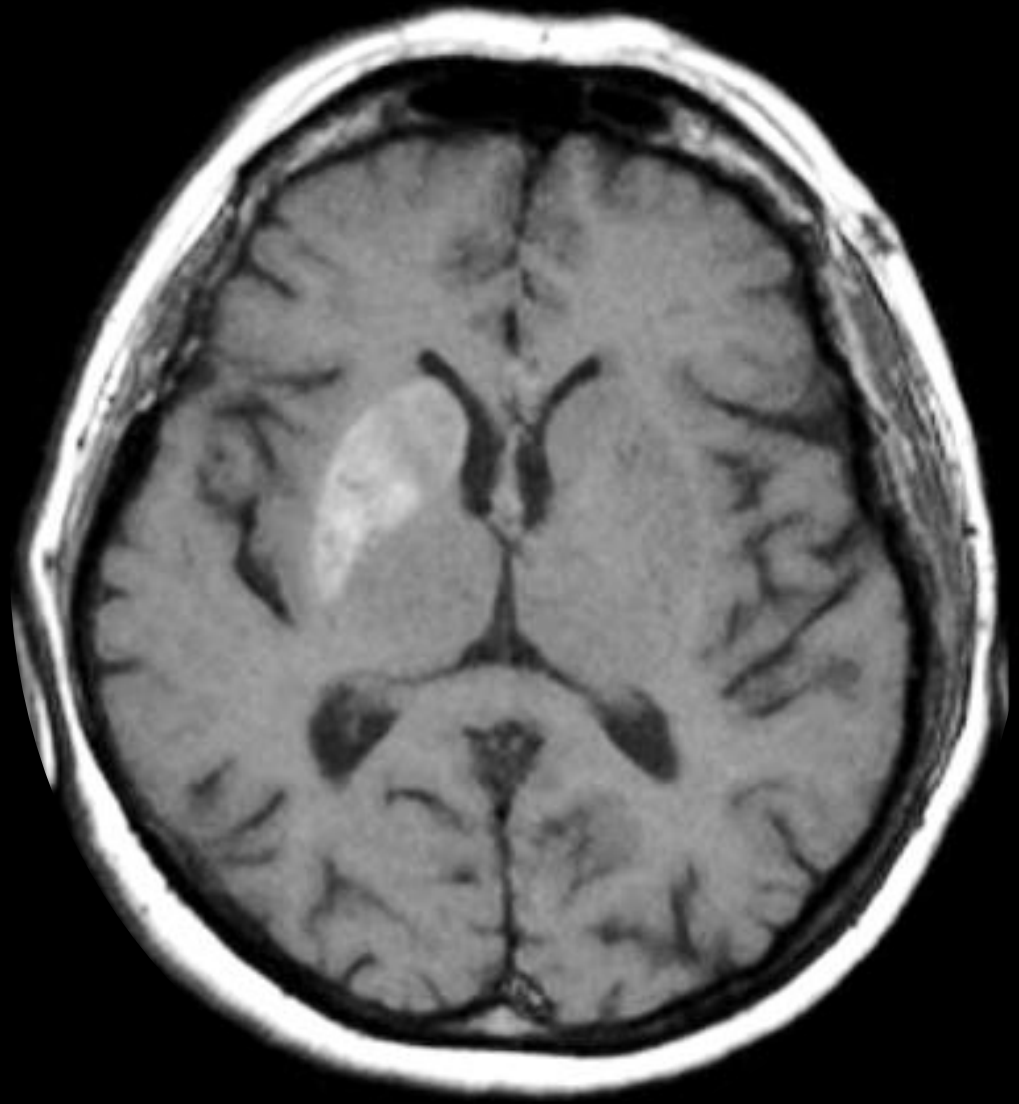
- Elderly patients with poorly controlled DM2
- Chorea and ballismus (usually unilateral) with sudden onset
- Often resolves completely or partially after correction of hyperglycemia
- Some patients may require chronic management of their movement disorder (haloperidol)

- Contentious theories for pathophysiology
  - GABA depletion
  - Oxidative stress
  - Hyperviscosity
  - Neurovascular unit dysfunction (cerebral autoregulation and microglial activation)

Nonketotic Hyperglycemic Chorea

Imaging Presentation

- MRI modality of choice
  - hallmark is BG T1 shortening (hyperintensity)
  - predominantly on putamen, but also caudate and GP
  - contralateral to abnormal movements
- T2W/FLAIR – variable hyperintensity
- T2*/GRE – hypointensity
- DWI – variable



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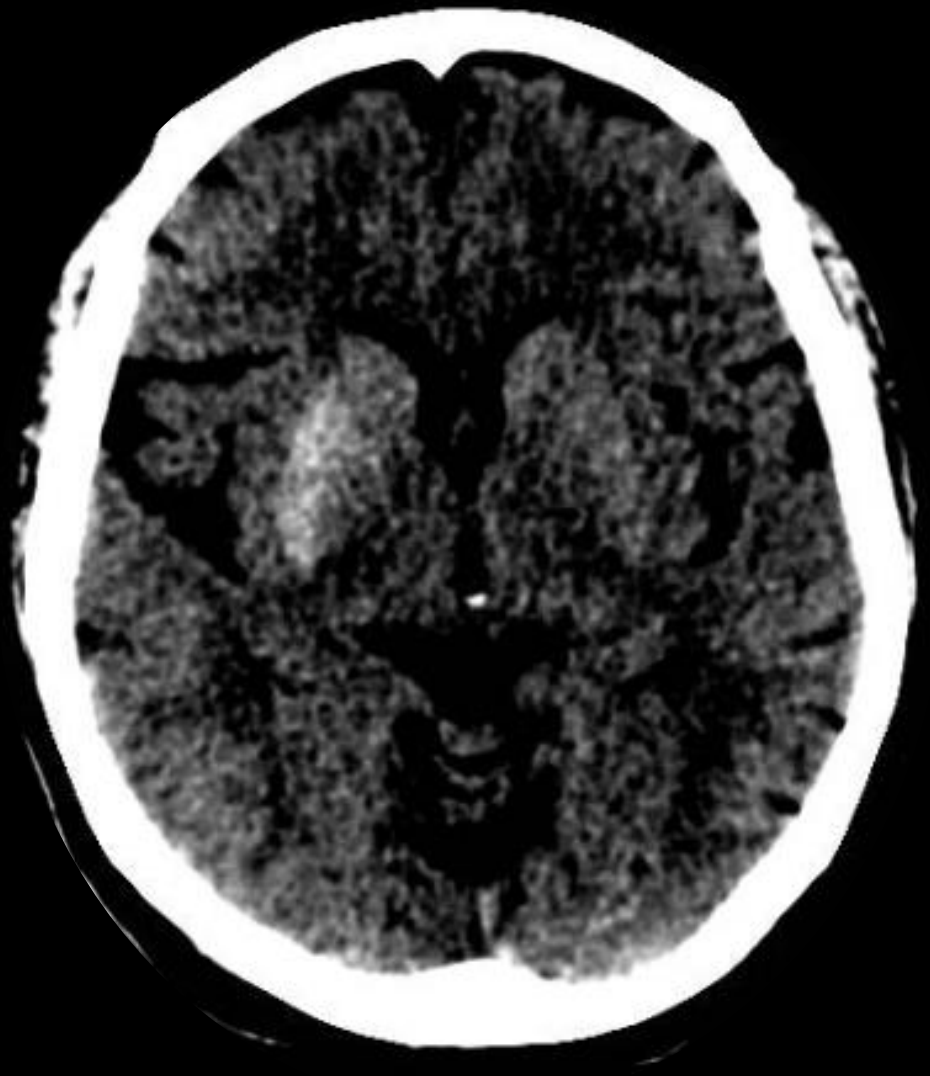


Nonketotic Hyperglycemic Chorea

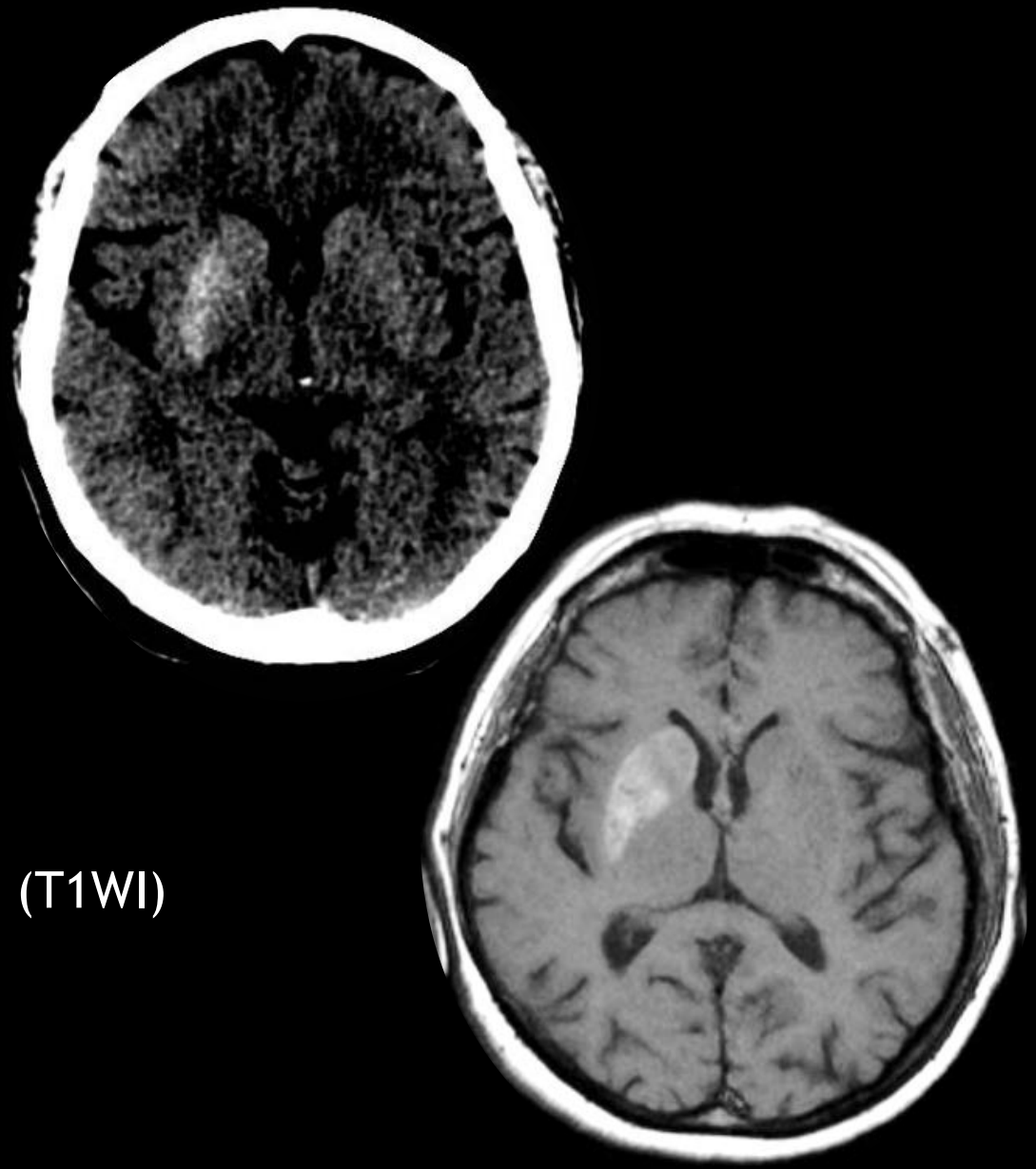
Imaging Differential Diagnosis

Unilateral Involvement

- Basal ganglia acute hemorrhage
  - hypertensive
  - hemorrhagic transformation lenticulostriate infarct
- Basal ganglia calcifications

Bilateral Involvement

- Wilson's Disease (T1WI)
- Manganese Accumulation – Chronic Liver Disease (T1WI)
- Basal ganglia calcifications (T1WI and CT)



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Clinical Research Article

Case Series of 46 Patients With Nonketotic Hyperglycemia-associated Chorea: A Retrospective Follow-up Study

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*All authors contributed equally to the study.

Take-home Points:

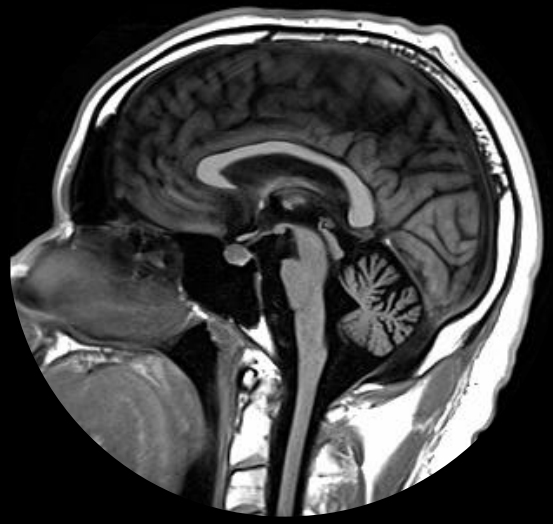
- Retrospective cohort of 46 patients – largest to date
- Mean age 68.6 years (48-89), equal sex distribution
- Hyperglycemia (mean Hgb A1c 11.4% +- 3.8%) and T1 shortening on contralateral BG universal
- 25% patients presented with normoglycemia at admission yet typical chorea and MRI with T1-weighted changes
- 2 cases had worsening symptoms despite MRI signal resolution – paradoxical imaging changes

Case #2 – Progressive Incoordination + Speech Changes

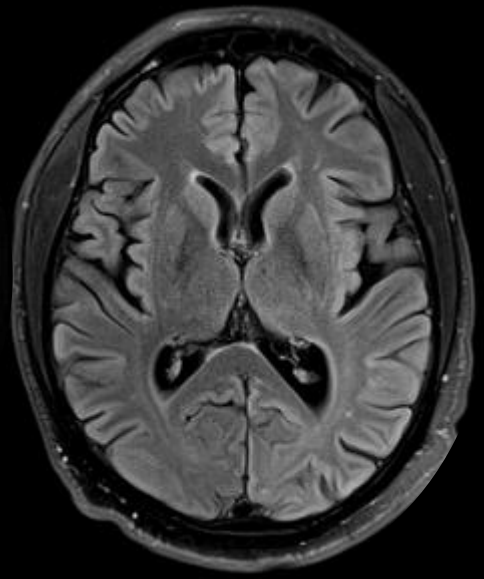
- 36 yo right handled male
- Initial symptoms (6 yr prior): trouble playing video games, then trouble walking on uneven surfaces (i.e. walking or jumping on a trampoline)
- Speech difficulties started 3 years ago (others mentioned his voice was different)
- No sensory disturbances, dysphagia, vision changes, cognition changes, bowel or bladder changes, REM sleep behavior disorder, orthostatic complaints, or weakness that he complains of. No family history of similar neurologic changes or early deaths. No significant alcohol use.
- Physical: Dysmetria, L > R, slurred speech. No motor or sensory deficit. Intact cognition. Orthostatic hypotension.

Case #2

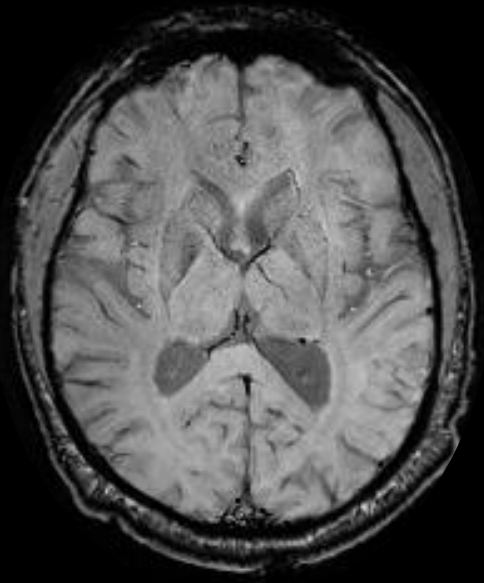
Sag T1W



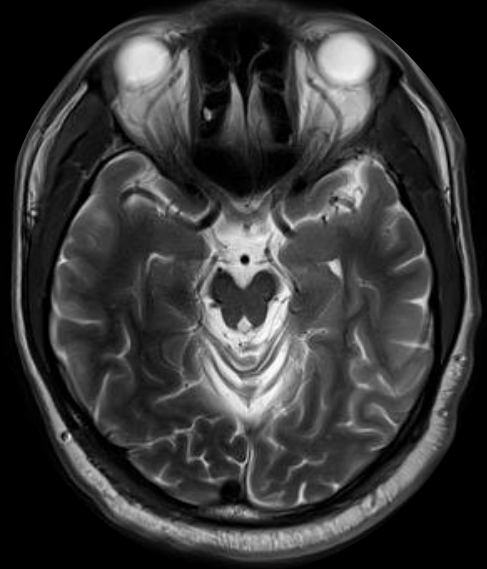
T2W FLAIR



T2*W GRE



T2W TSE

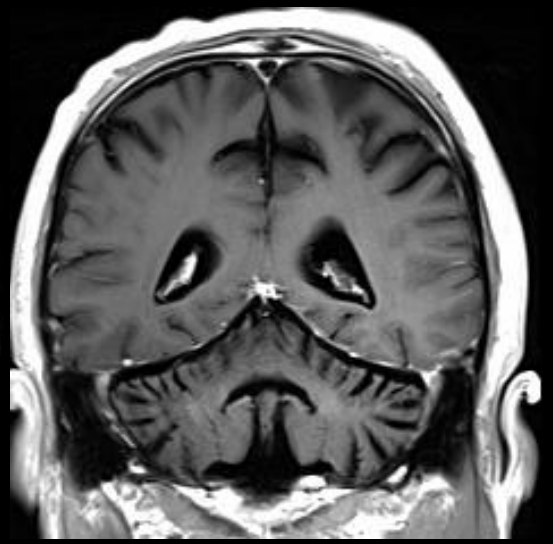


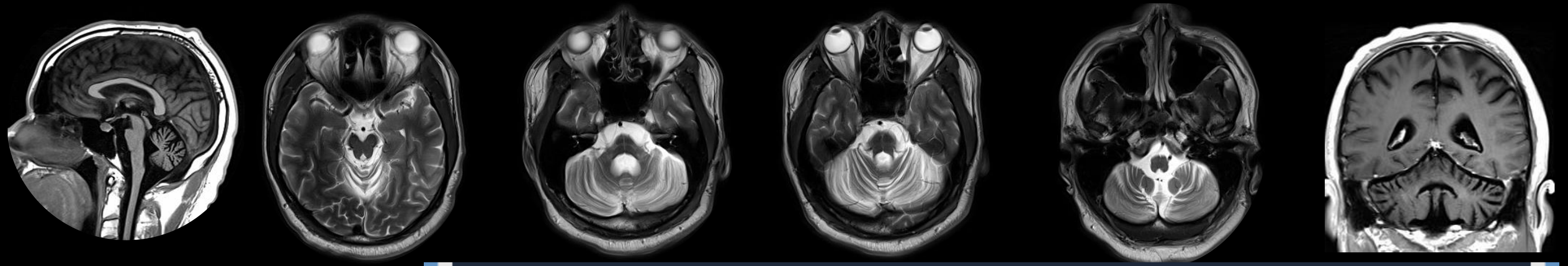
T2W TSE

T2W TSE

T2W TSE

Cor T1W Gd+





Case #2 – Progressive Incoordination + Speech Changes

Case #2 - Regarding the disease mechanism for this process, which is correct :

0

The combination of clinical and imaging findings suggest a tauopathy 0%

The combination of clinical and imaging findings suggests a synucleinopathy 0%

This patient likely has an immune mediated disorder which will likely benefit from steroids 0%

Patient workup should prioritize exclusion of a systemic malignancy 0%

Genetic testing will likely yield a specific diagnosis 0%

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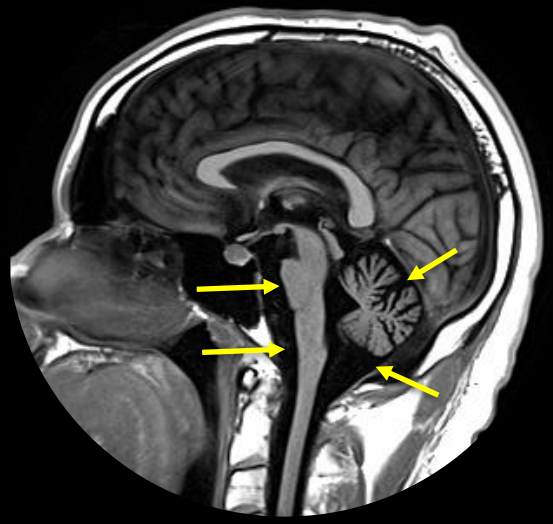
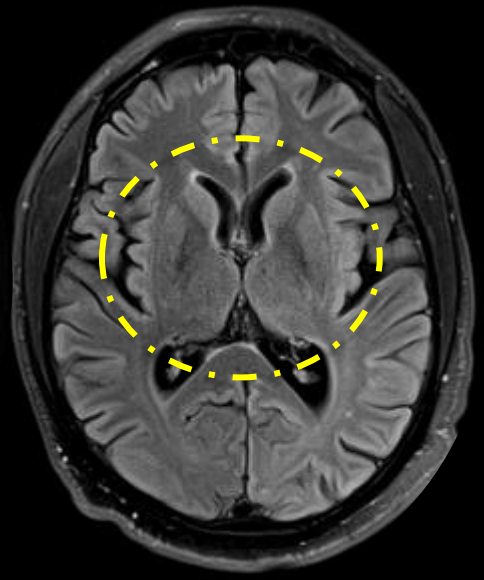
Case #2

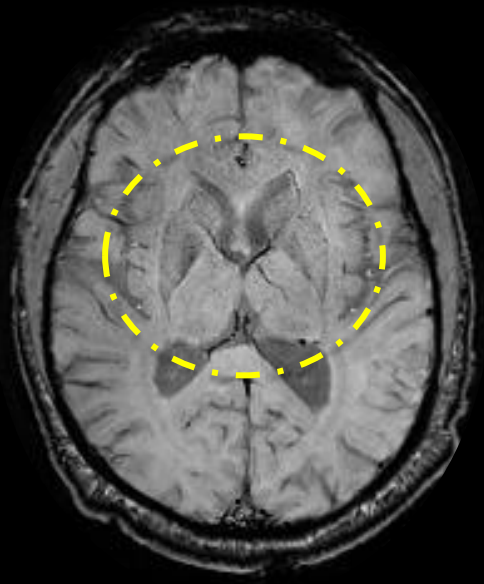
Sag T1W



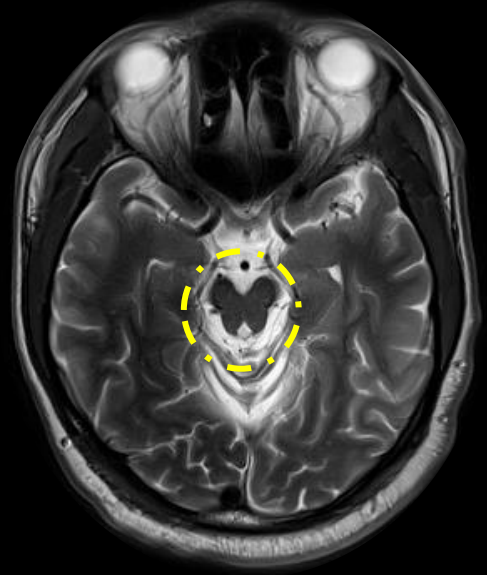
T2W FLAIR



T2*W GRE



T2W TSE

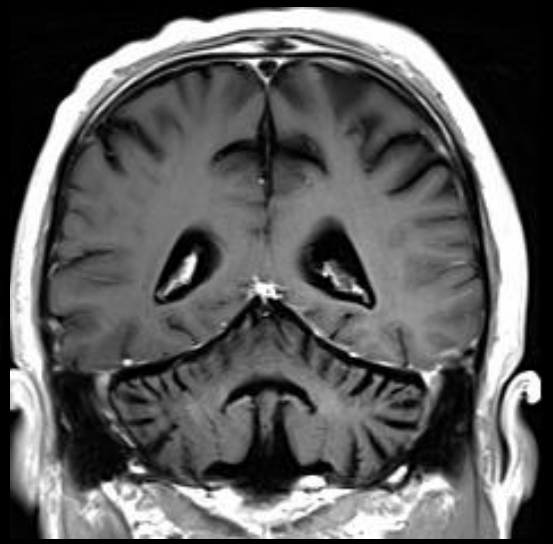


T2W TSE

T2W TSE

T2W TSE

Cor T1W Gd+



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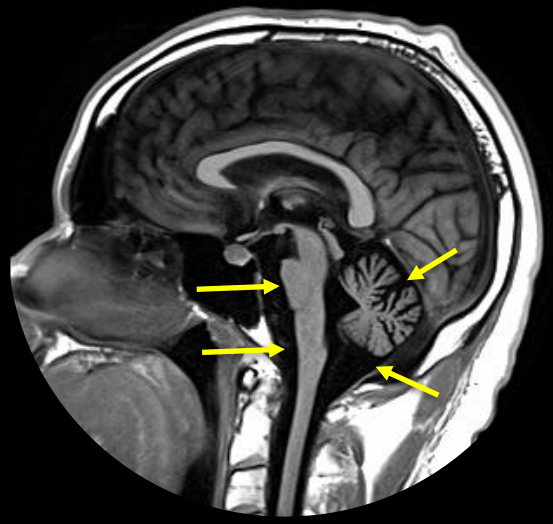
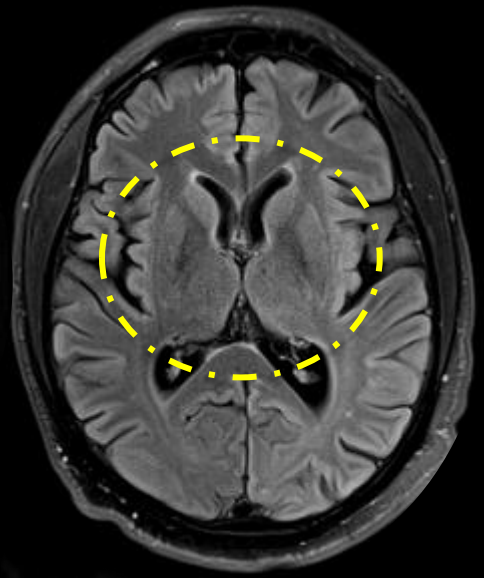
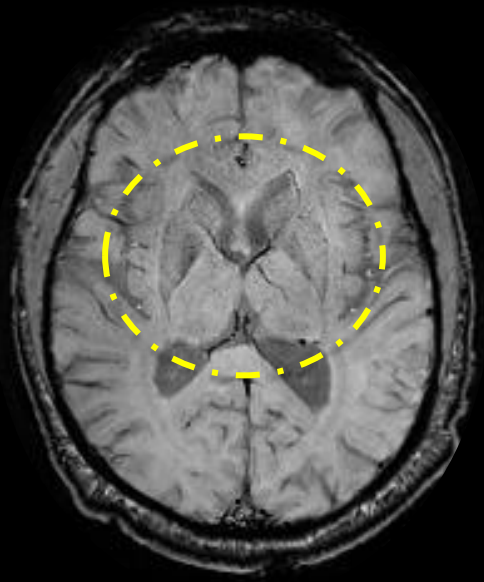
Sag T1W



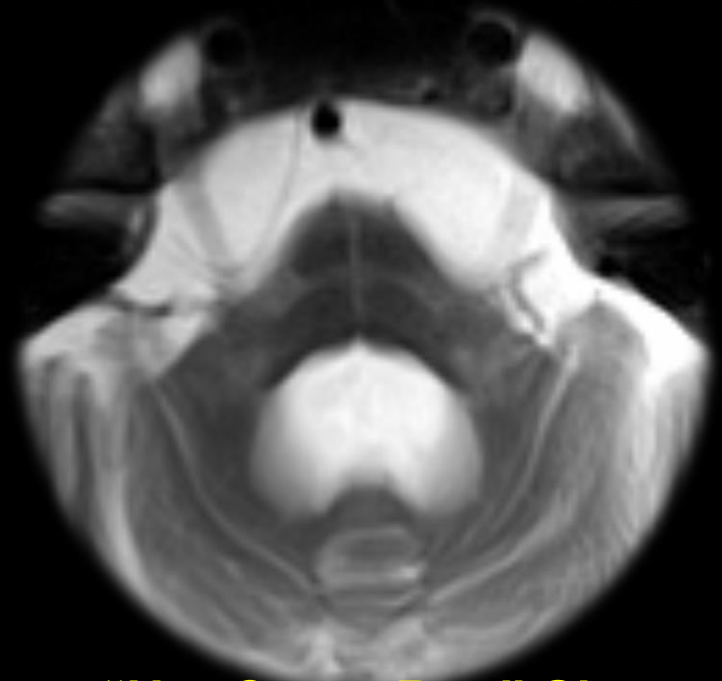
T2W FLAIR



T2*W GRE

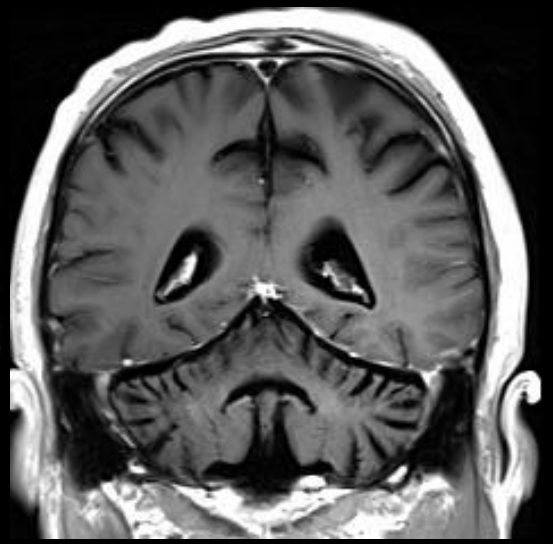


T2W TSE



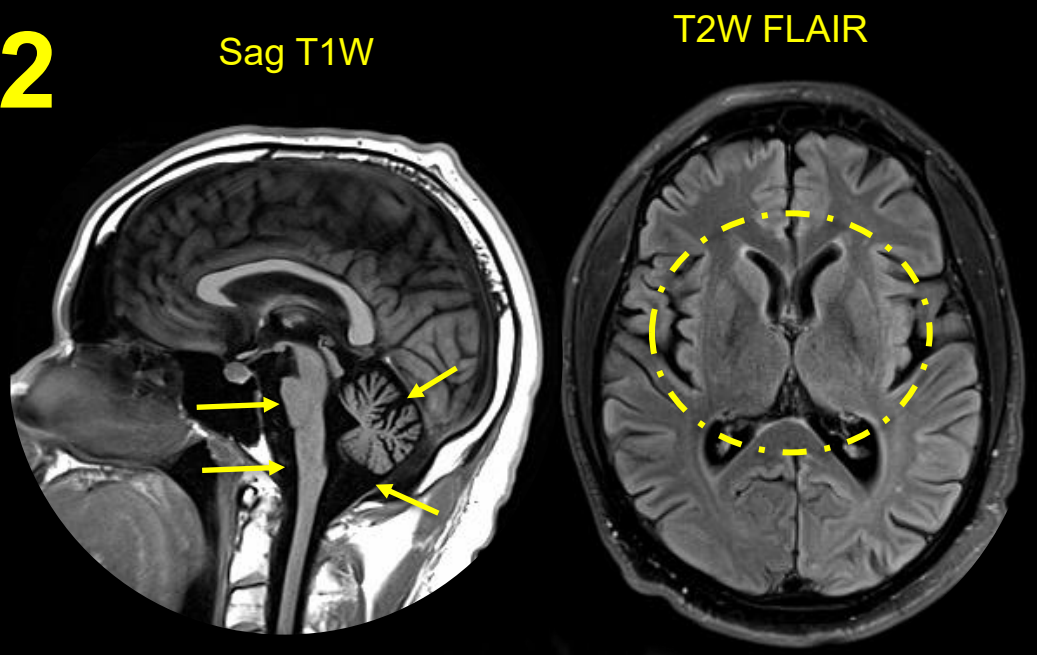
T2W TSE

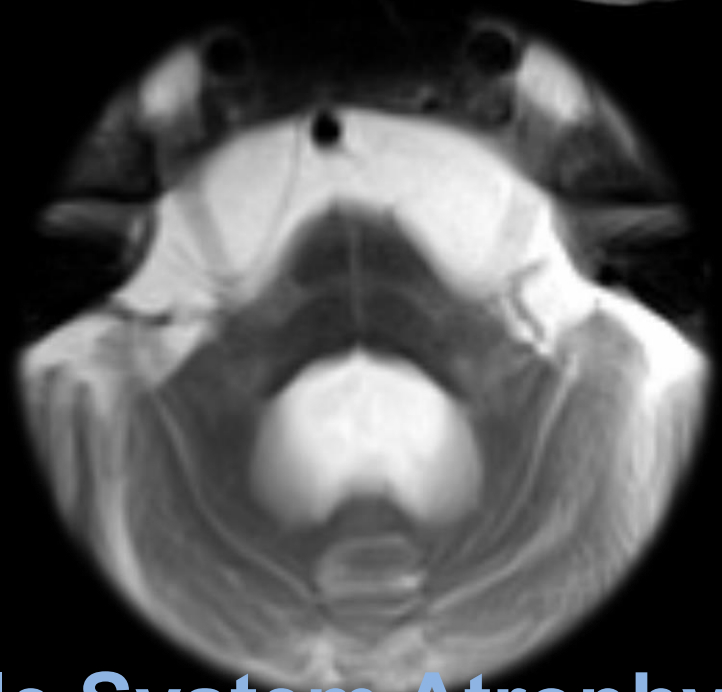
Cor T1W Gd+



“Hot Cross Bun” Sign

Case #2





Case #2 Diagnostic Checklist

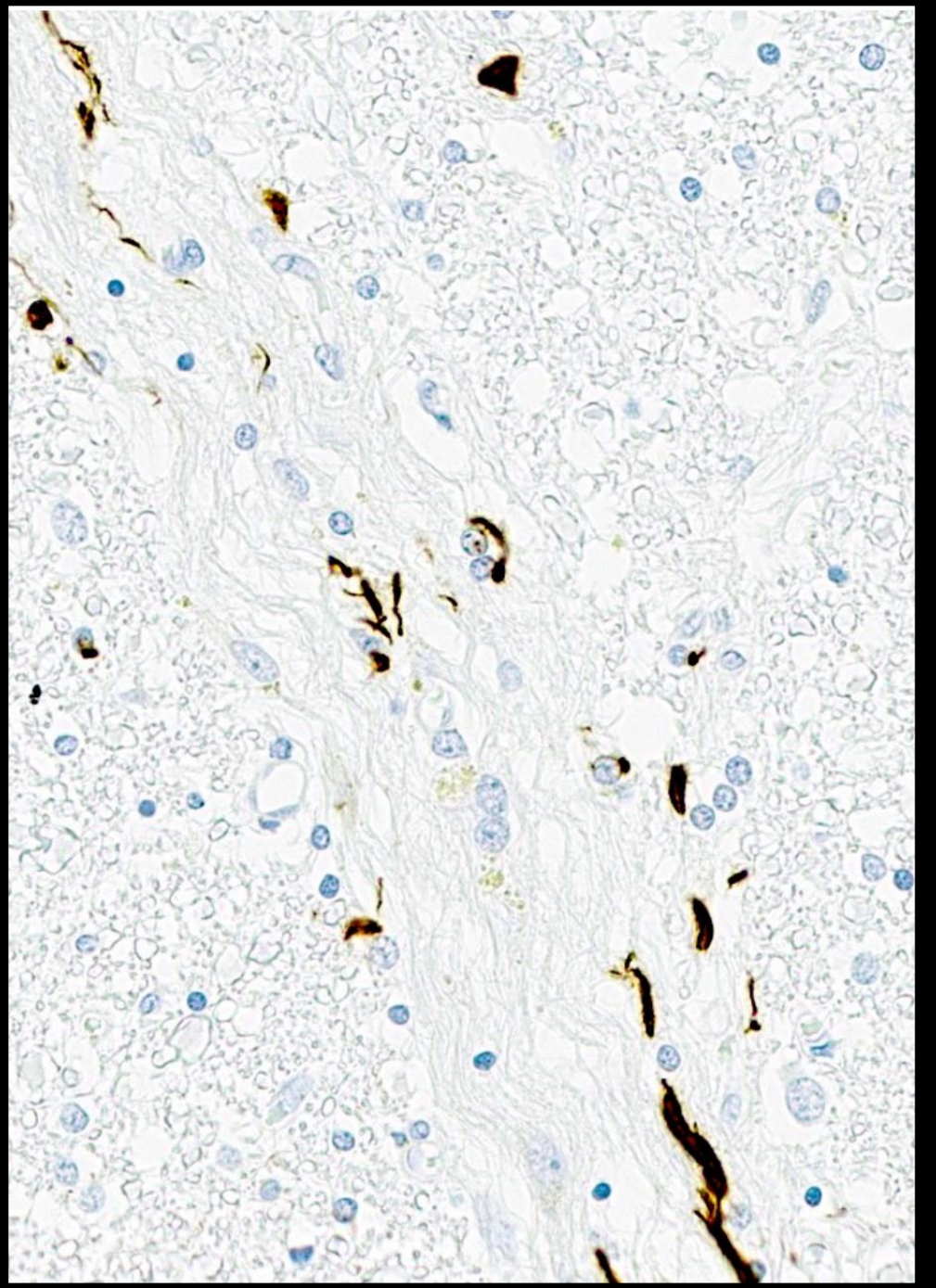
- Young male
- Slowly progressive incoordination
- Speech changes
- Cerebellar Symptoms
- Autonomic dysfunction
- Imaging
  - cerebellar + brainstem atrophy
  - “hot cross bun” sign

Multiple System Atrophy – Cerebellar Type (MSA-c)

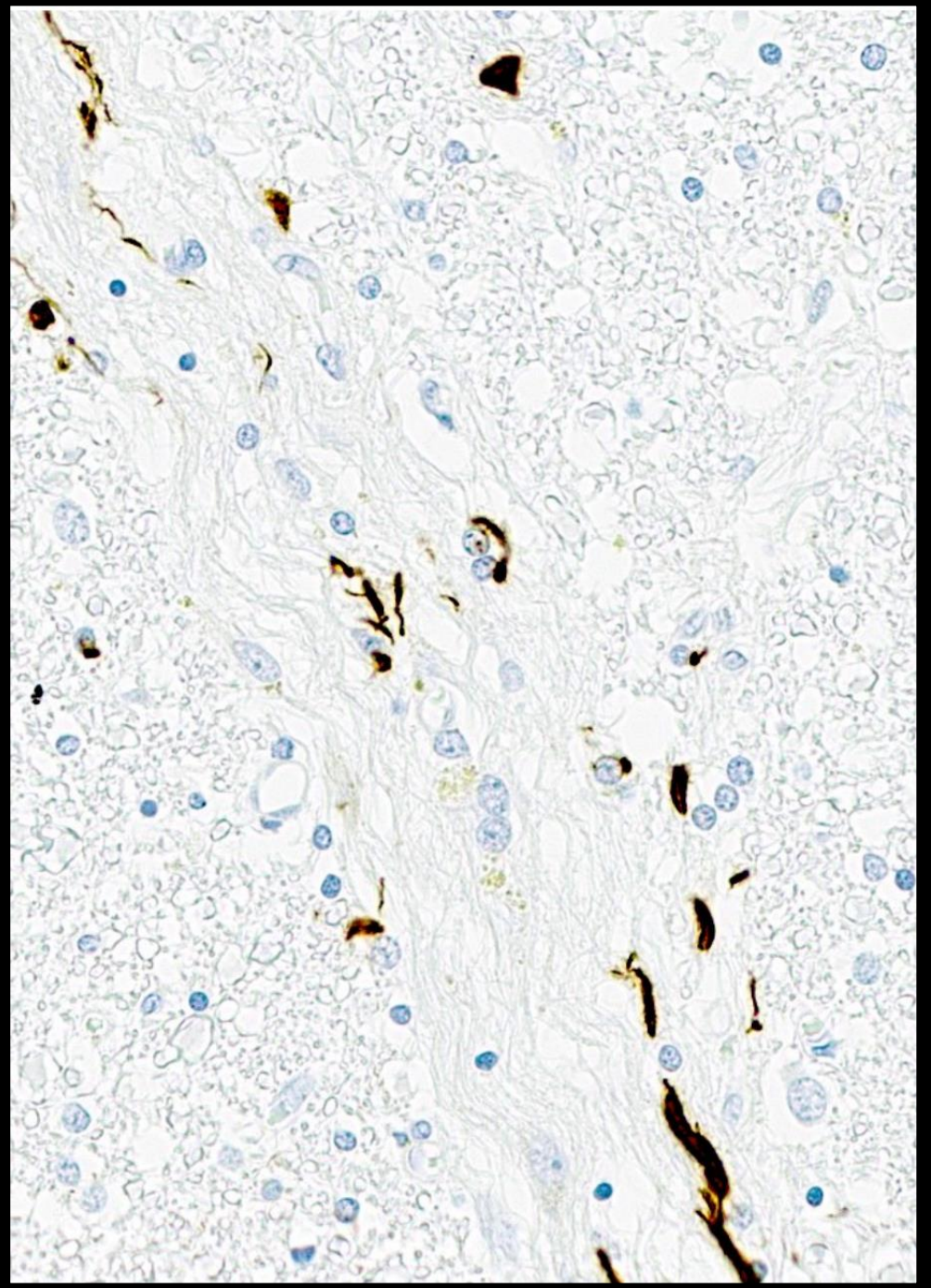
Multiple System Atrophy

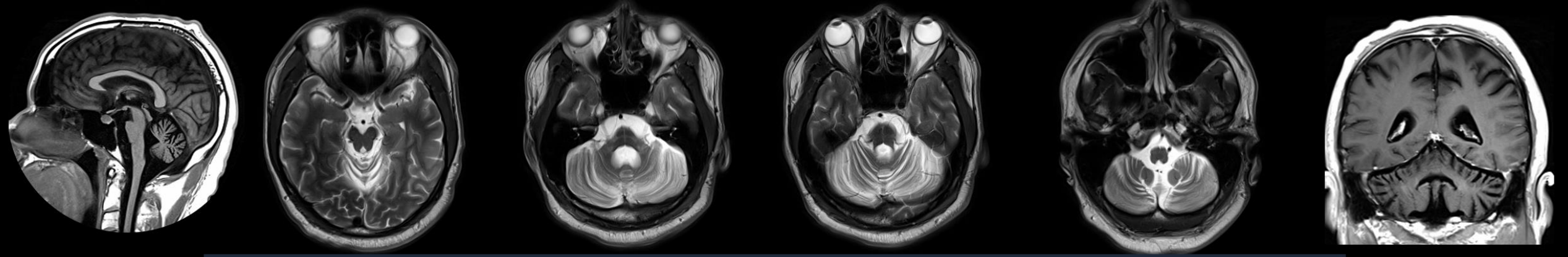
- Rare, adult-onset, sporadic and progressive neurodegenerative disease
- Estimated incidence of 0.6 / 100.000 person-years
- Great geographic variability  

- Clinical Presentation and Diagnosis
  - Challenging diagnosis
    - rarity
    - overlap other movement disorders
    - lack of specific in vivo diagnostic tool
  - Three main syndromes:
    - parkinsonisn
    - cerebellar ataxia
    - autonomic failure
  - Two subtypes:
    - Parkinsonian predominant (MSA-p)
    - Cerebellar predominant (MSA-c)



Multiple System Atrophy





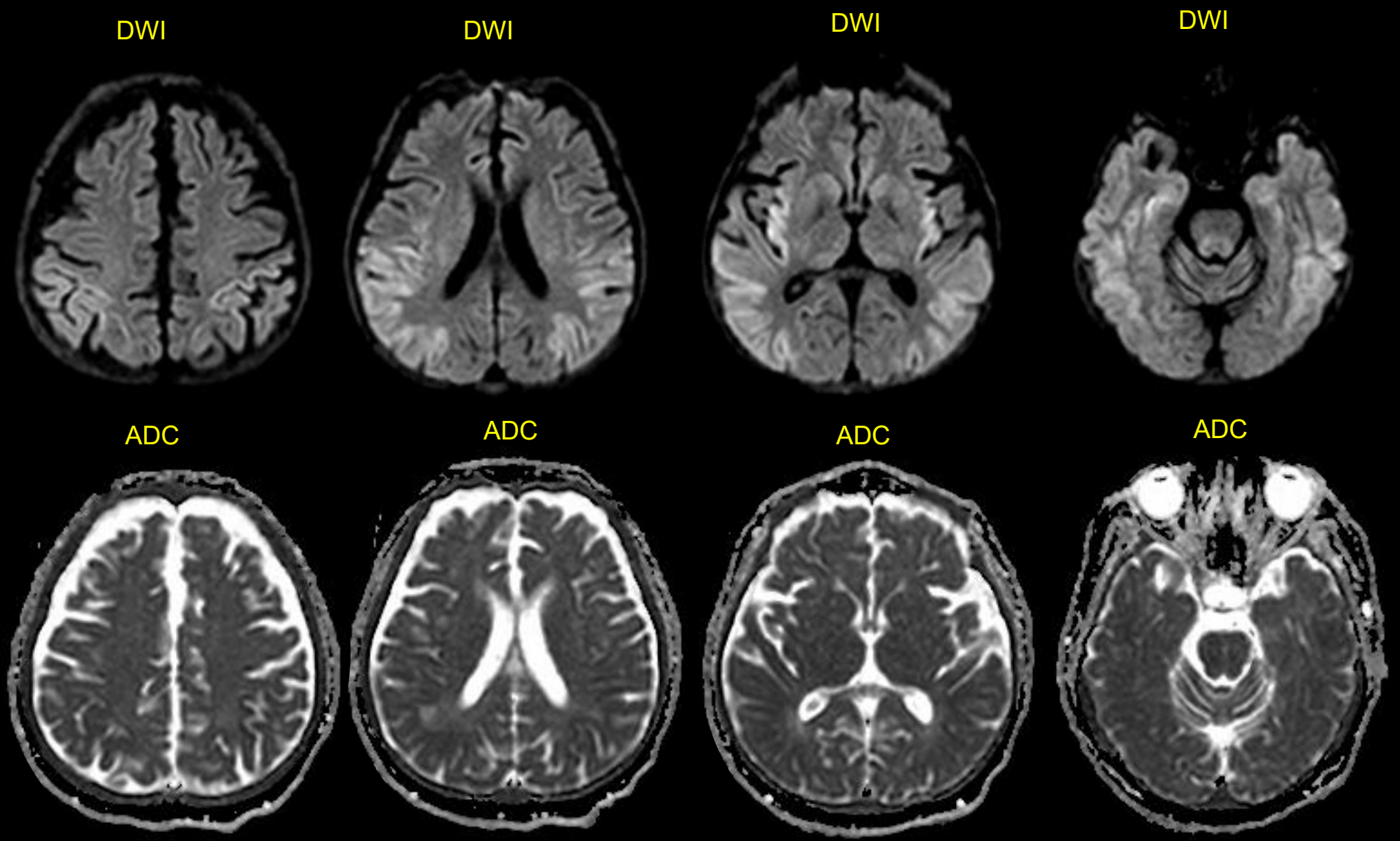
Regarding the disease mechanism for this process, which is correct :

0

- The combination of clinical and imaging findings suggest a tauopathy **(A)**
- ✓ The combination of clinical and imaging findings suggests a synucleinopathy **(B)**
- This patient likely has an immune mediated disorder which will likely benefit from steroids **(C)**
- Patient workup should prioritize exclusion of a systemic malignancy **(D)**
- Genetic testing will likely yield a specific diagnosis **(E)**

Case #3 – Acute Altered Mental Status

- 60 yo female
- Altered mental status and visual hallucinations. Rapid worsening over 2 days
- Recent GI Bleed and icterus
- PMHx: alcohol abuse and cirrhosis.
- Last drink 24hs ago



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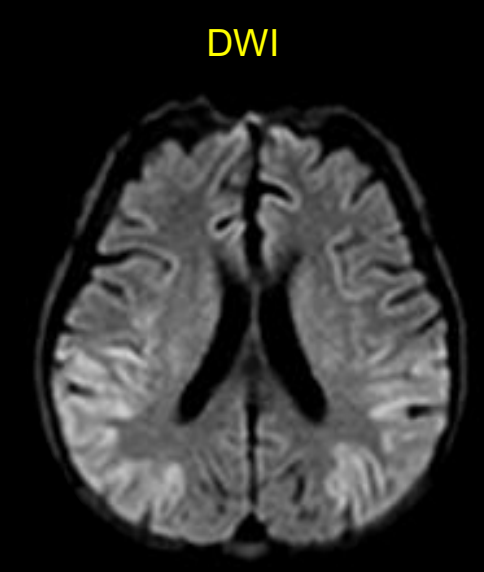

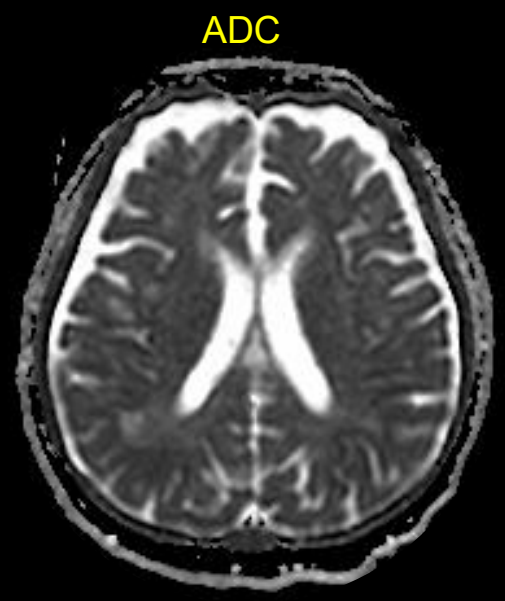

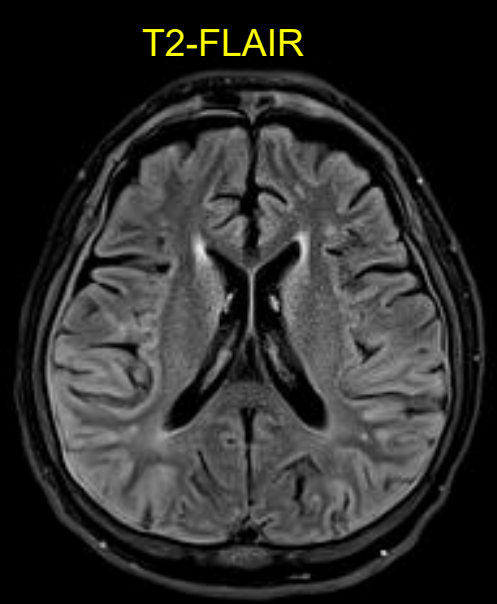

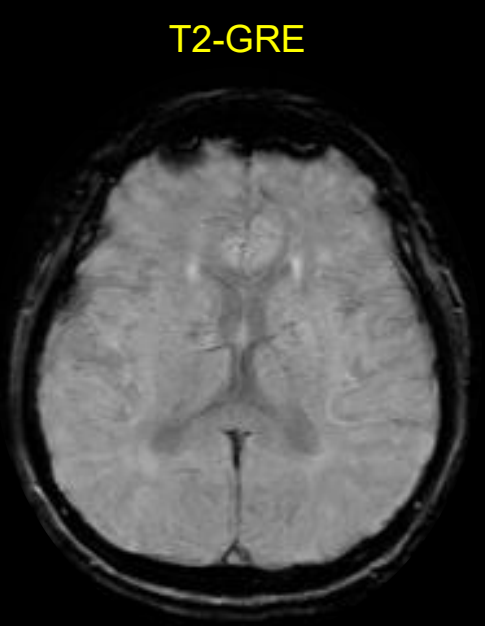

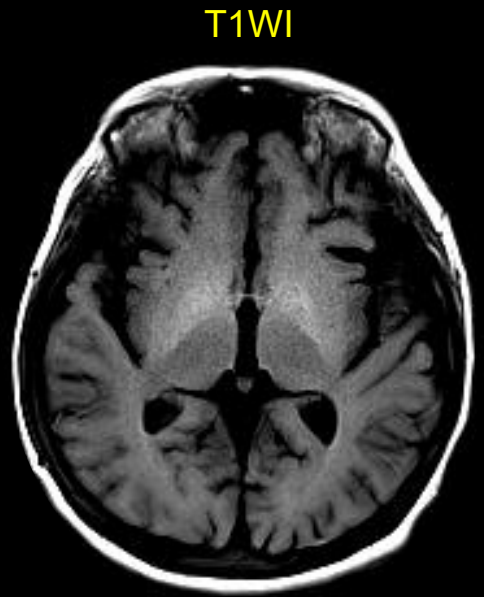

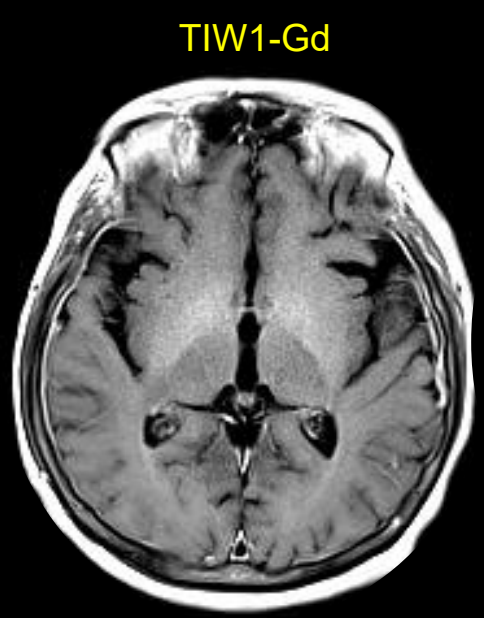

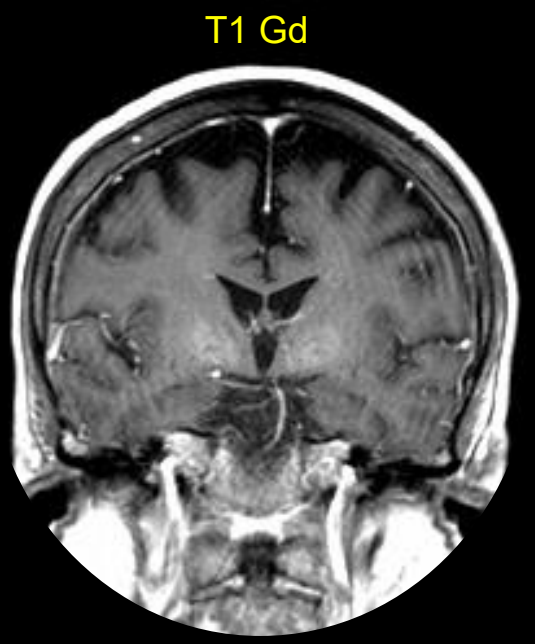

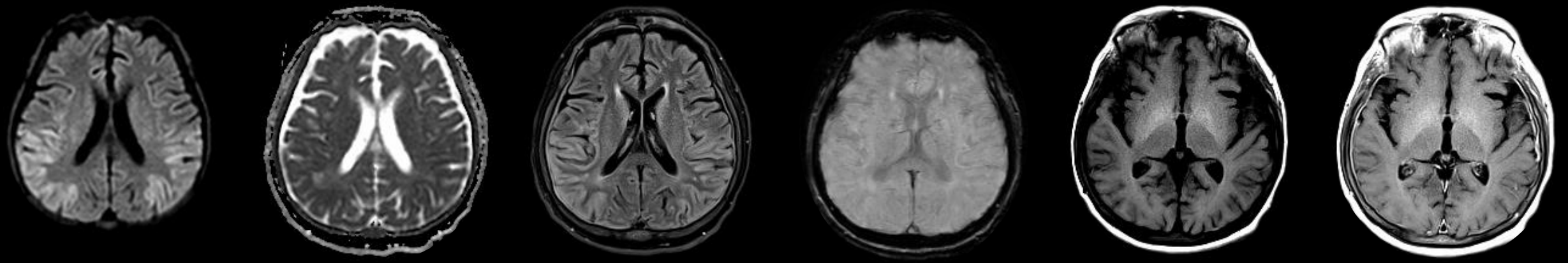

Case #3 – Acute Altered Mental Status

Case #3 - Choose the correct statement regarding this condition:

Treatment is mainly supportive and may include optimizing cerebral perfusion pressure and targeted temperature management 0%

Lumbar tap and EEG may create additional strong evidence of the diagnosis 0%

Antiviral use is the most impactful strategy for this condition 0%

Treatment of precipitating causes and correction of metabolic abnormalities can improve outcomes 0%

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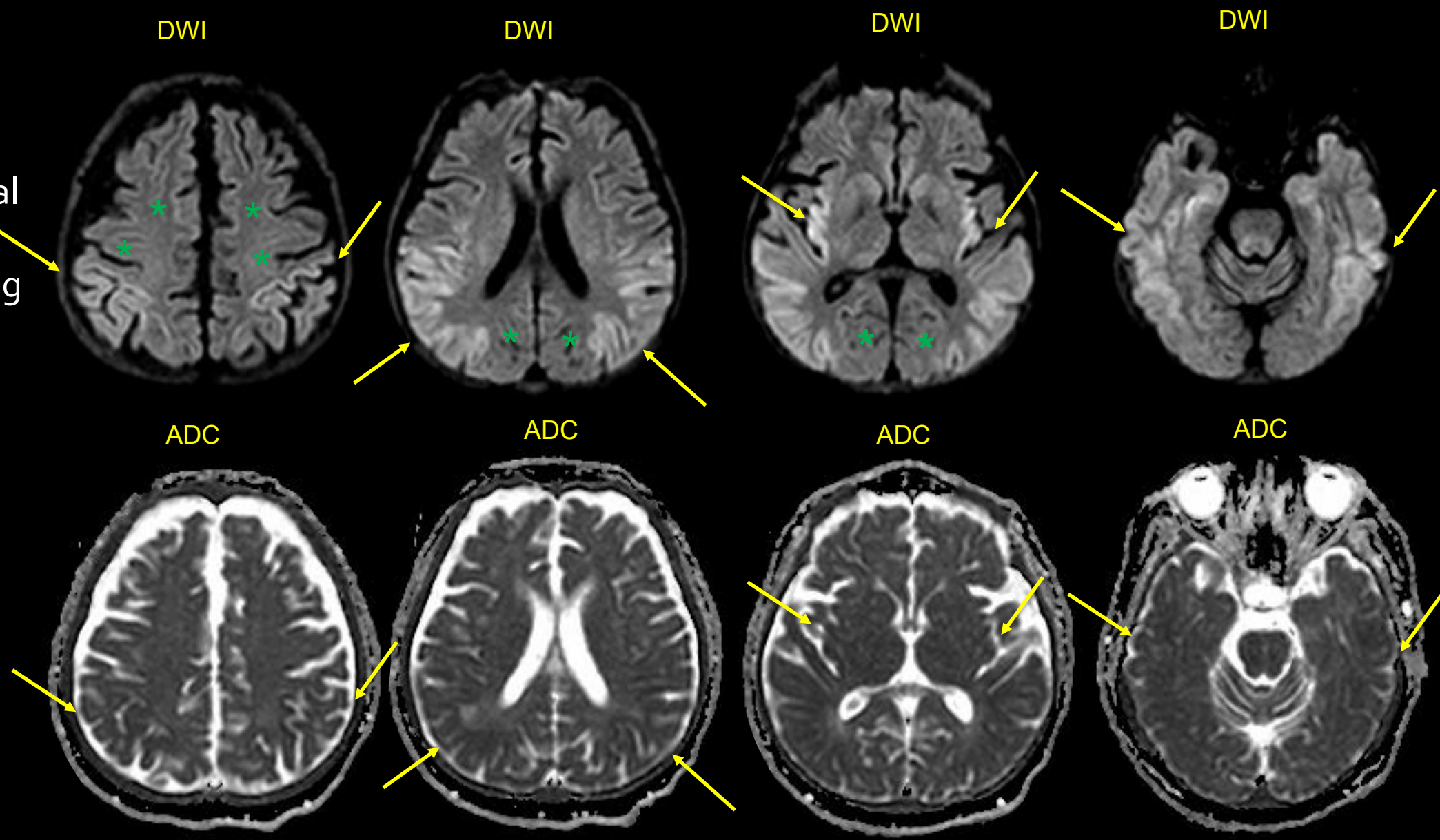
Case #3 – Acute Altered Mental Status

- 60 yo female
- Altered mental status and visual hallucinations. Rapid worsening over 2 days
- GI Bleed and icterus
- PMHx: alcohol abuse and cirrhosis. Last drink 24hs ago



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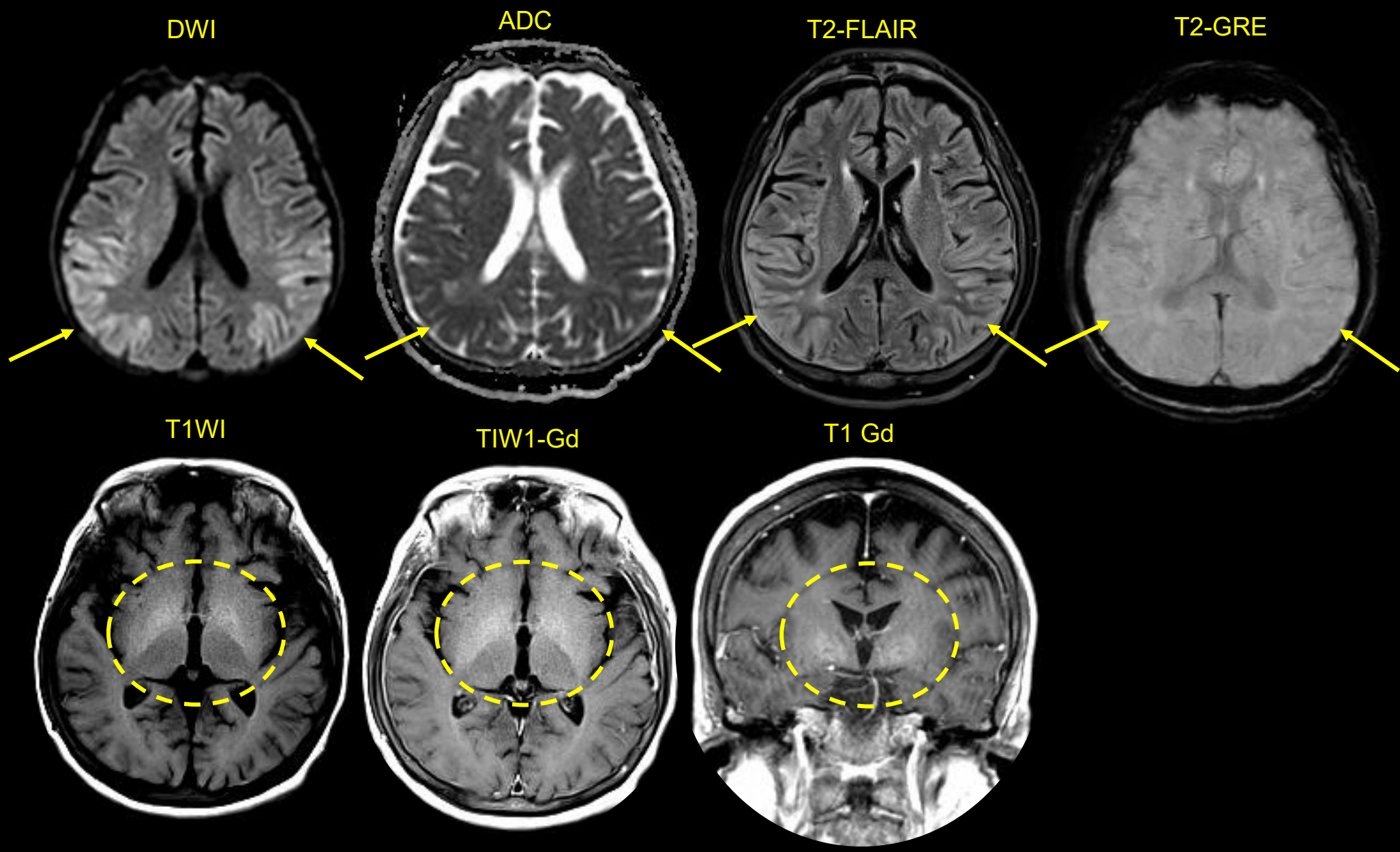
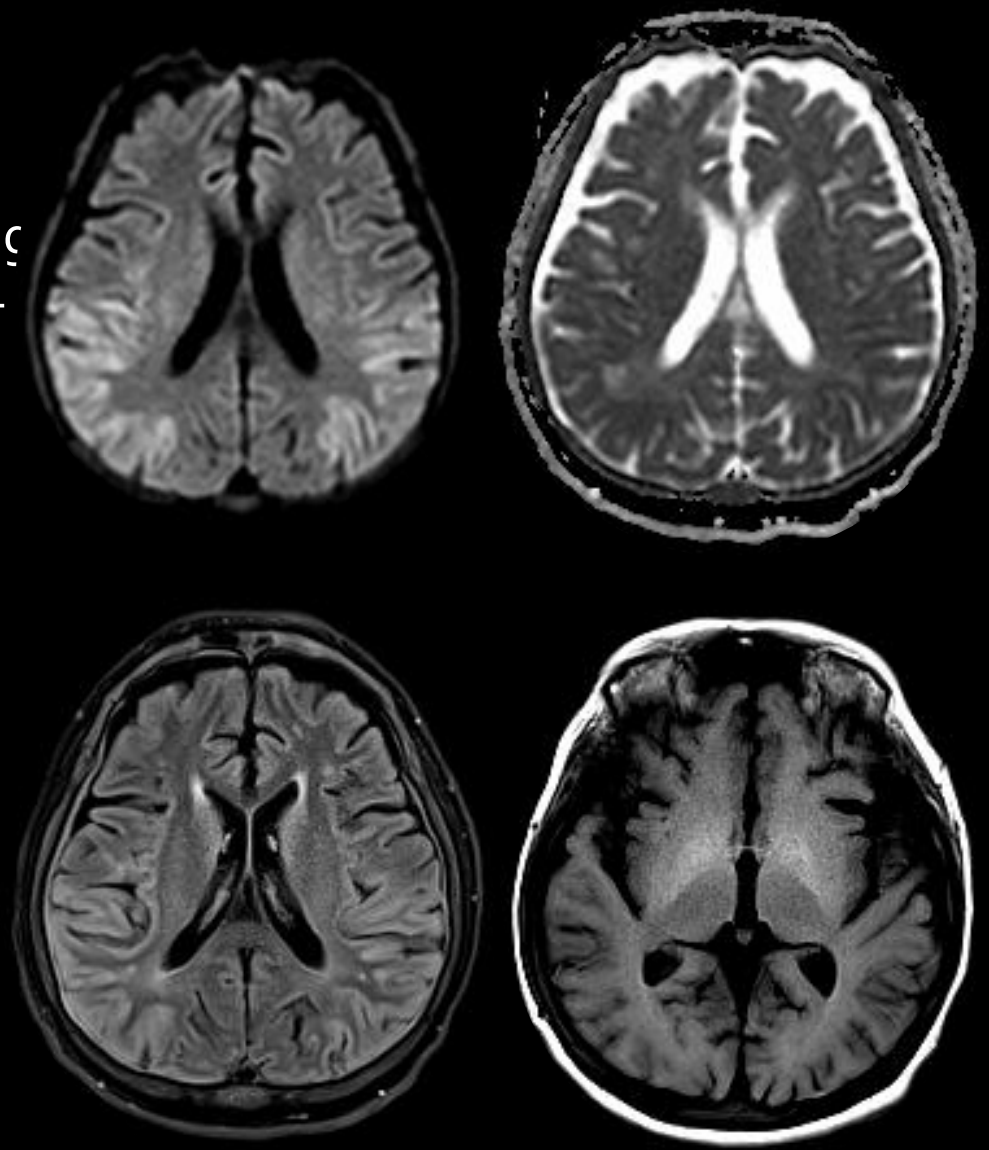


Case #3 – Acute Altered Mental Status

- 60 yo female
- Brought to ED: rapidly worsening mental status + visual hallucinations.
- Recent GI Bleed and icterus
- PMHx: alcohol abuse and cirrhosis.
- Last drink 24hs ago

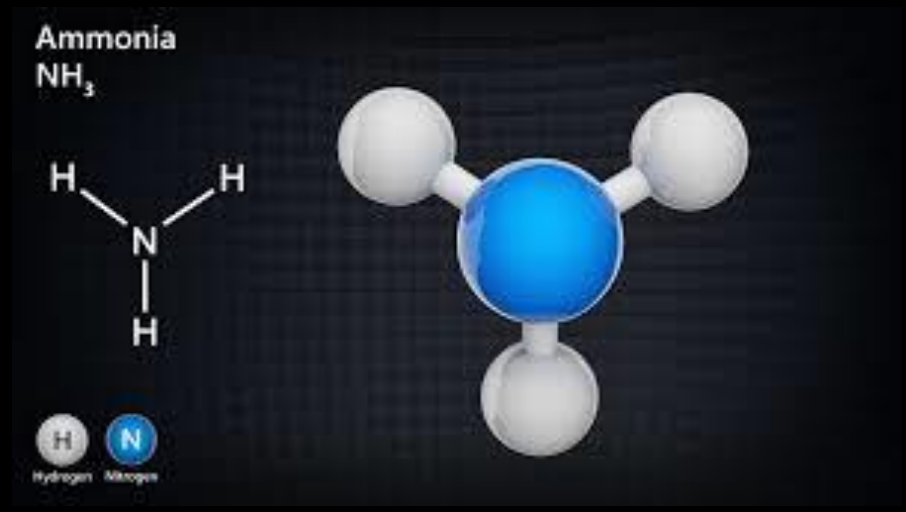


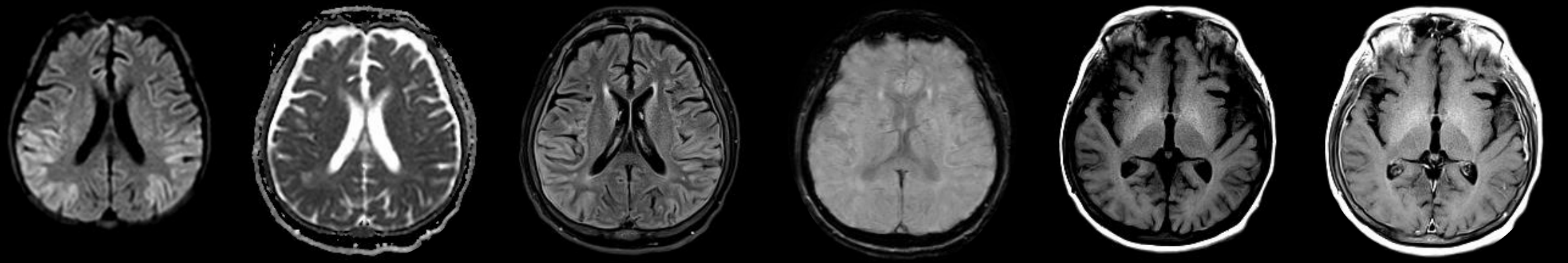
Case #3 Diagnostic Checklist

- Elderly female
- Acute change mental status
- Imaging
  - bilateral symmetrical DWI abnormalities non vascular distribution
  - Symmetrical faint T1 elevation GP
- Trigger: Upper GI bleed and icterus
- PMHx: alcohol abuse and cirrhosis

Acute Hyperammonemic Encephalopathy ↑**Ammonia: 211µg/dL**

Hyperammonemic Encephalopathy





Case #3 – Acute Altered Mental Status

Choose the correct statement regarding this condition:

(A) Treatment is mainly supportive and may include optimizing cerebral perfusion pressure and targeted temperature management — 0%

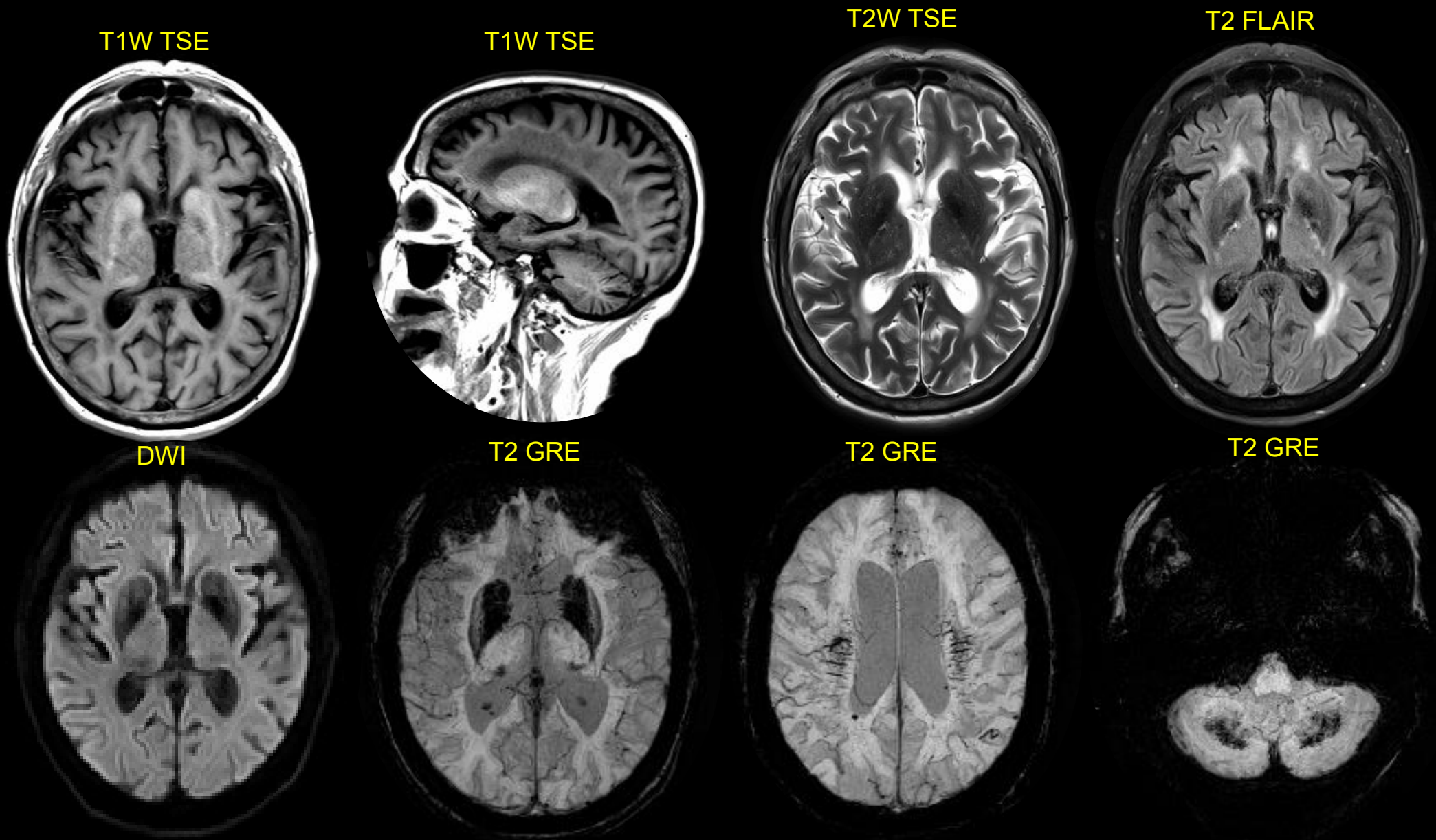
(B) Lumbar tap and EEG may create additional strong evidence of the diagnosis — 0%

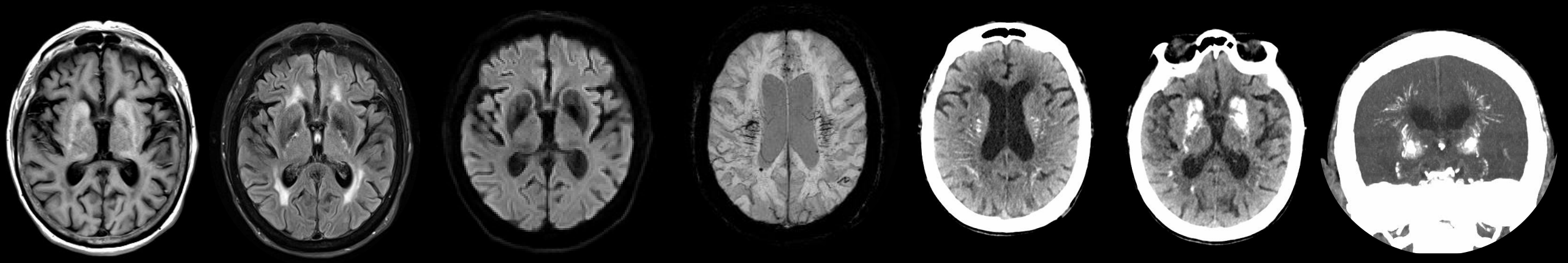
(C) Antiviral use is the most impactful strategy for this condition — 0%

✓ (D) Treatment of precipitating causes and correction of metabolic abnormalities can improve outcomes — 0%

Case #4 - Progressively declining gait and speech

- 82 yo F, slowly progressive unsteady gait, slurred speech, cognitive decline
- PMHx: DM2, HTN, prior thyroidectomy for toxic multinodular goiter
- Recurrent hypocalcemia





Case #4 - Progressively declining gait and speech

Case #4 - Choose the correct statement:

This condition has several possible etiologies, both inherited and acquired — 0%

Imaging findings are likely physiological and unrelated to the patient's symptoms — 0%

There is no treatment for this condition — 0%

This condition can be made worse by anticoagulants — 0%

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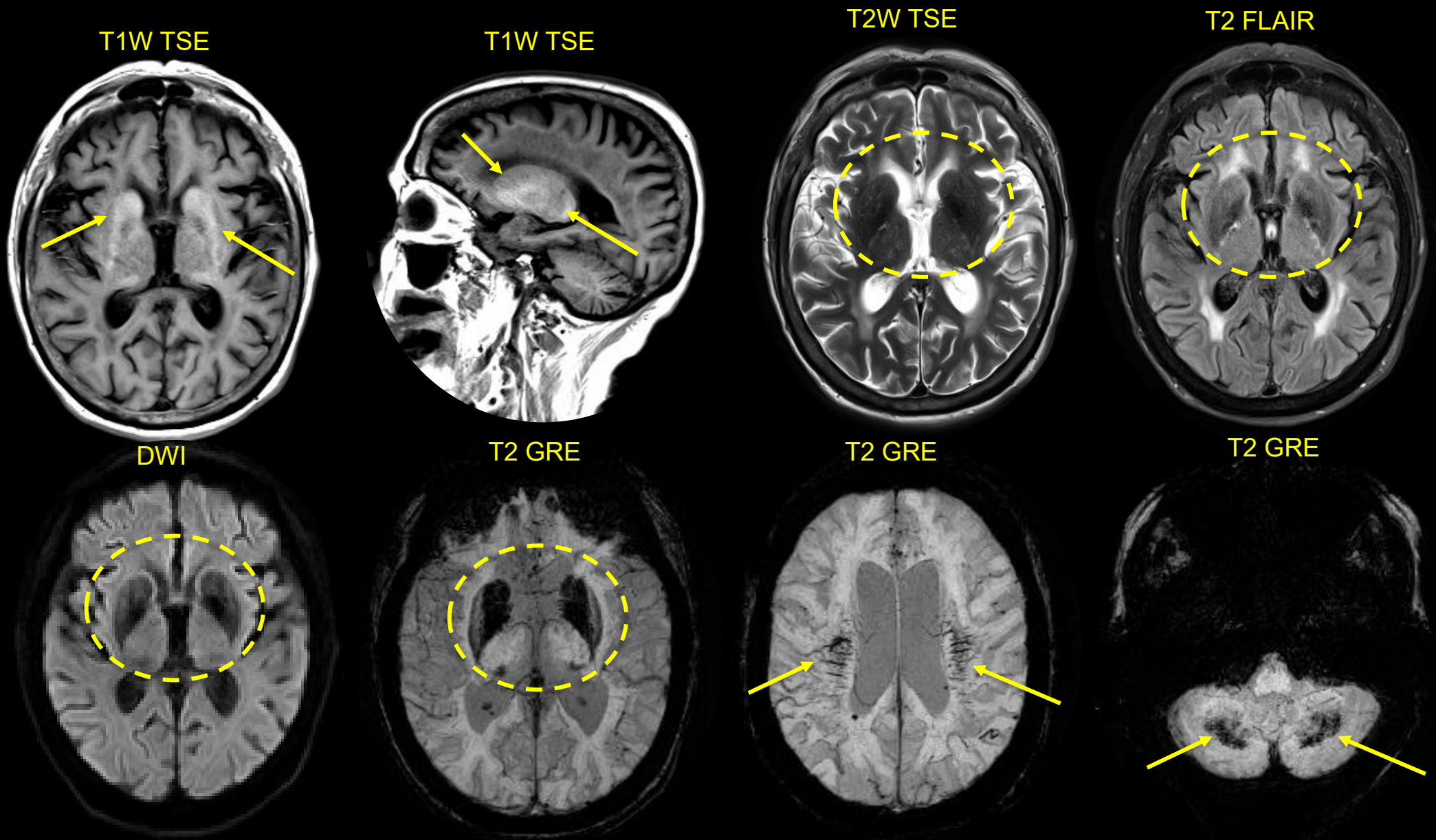

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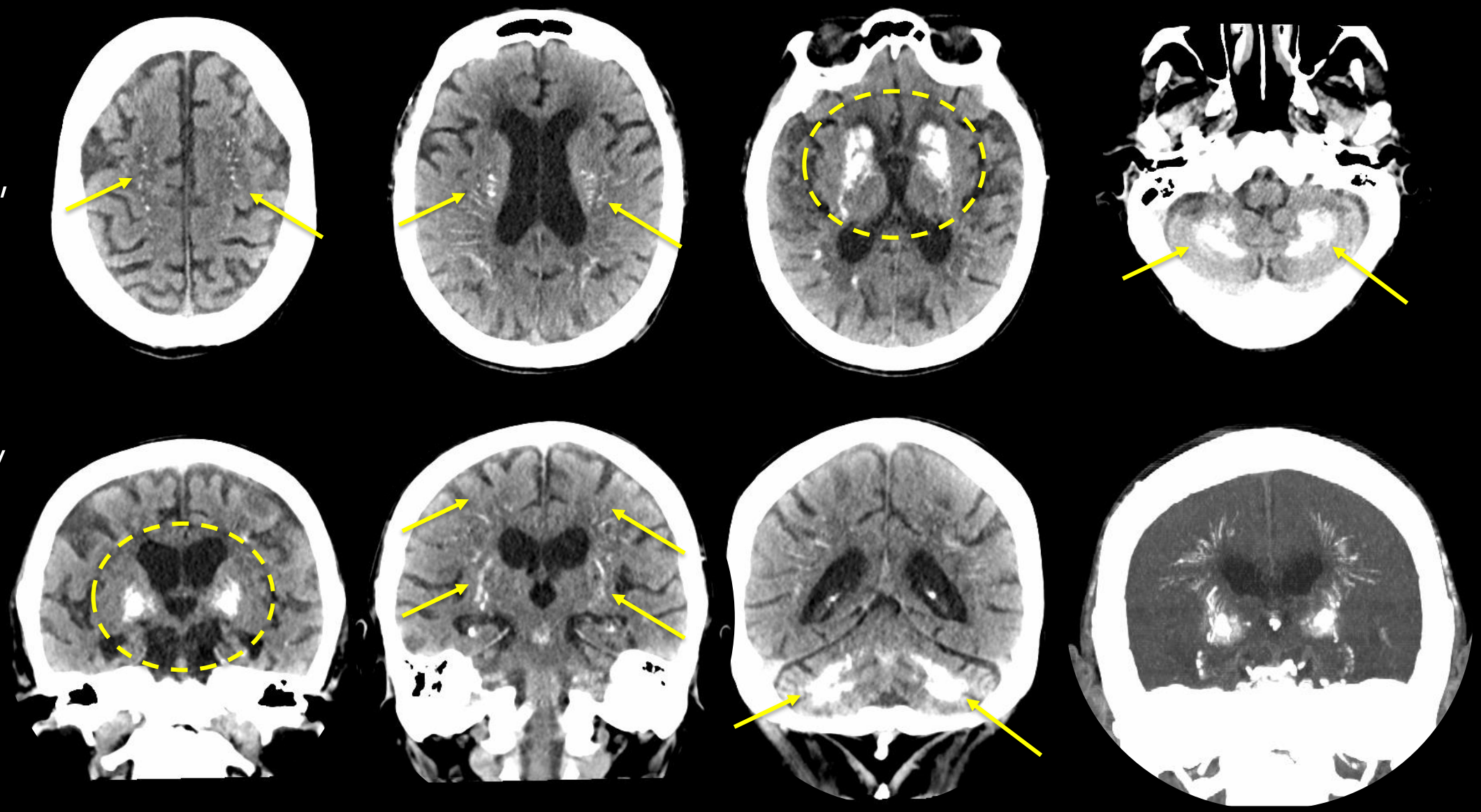

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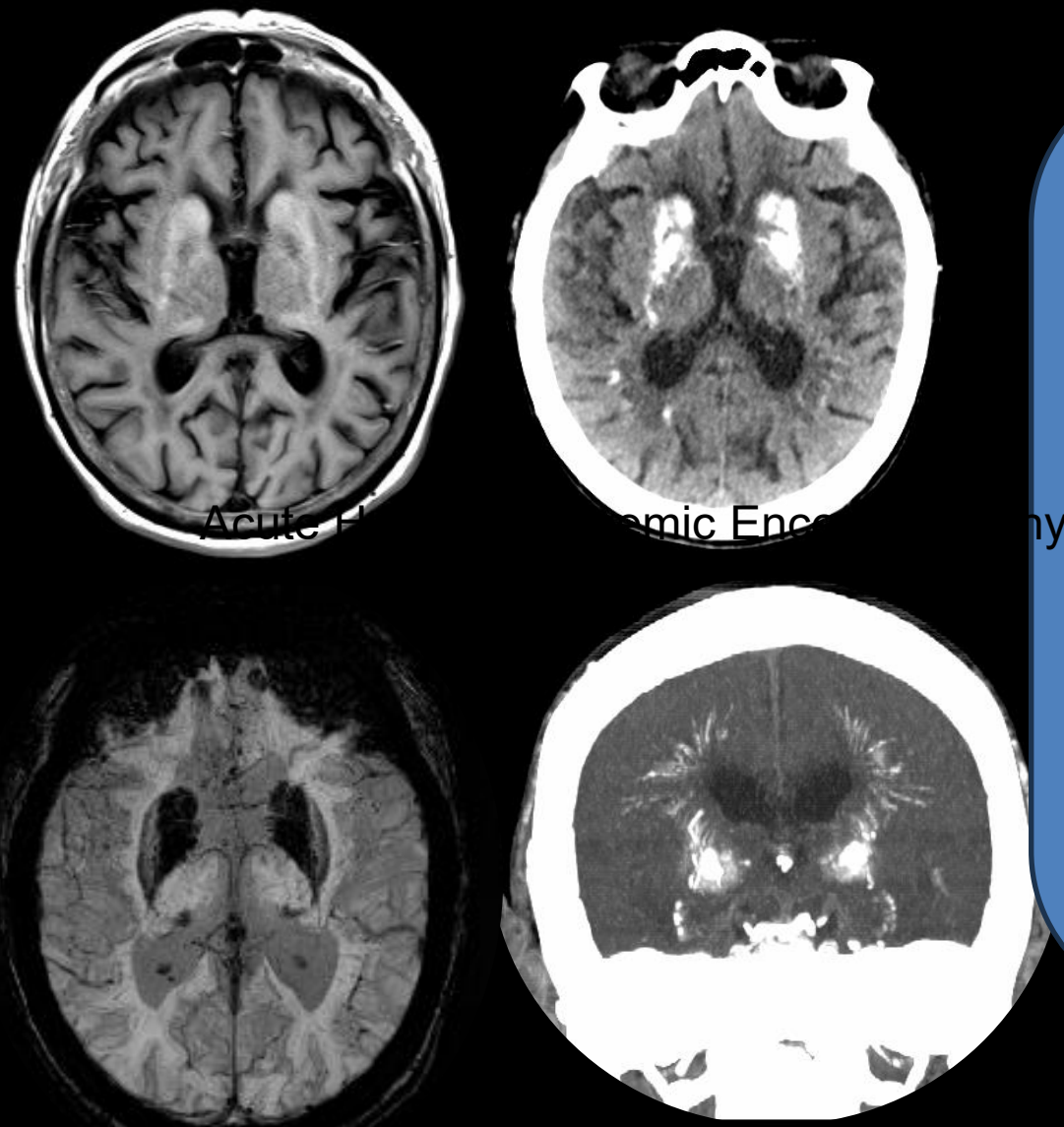


Case #4 Diagnostic Checklist

- Elderly female
- Slowly progressive deterioration
  - gait, speech, cognition
- Imaging
  - Extensive abnormal bilateral calcifications - deep grey nuclei and perivascular WM
  - Atrophy
- PMHx: thyroidectomy and recurrent hypocalcemia (hypoparathyroidism)

Fahr Syndrome (Hypoparathyroidism)

Fahr Syndrome (Bilateral Striatopallidodentate Calcinosis)

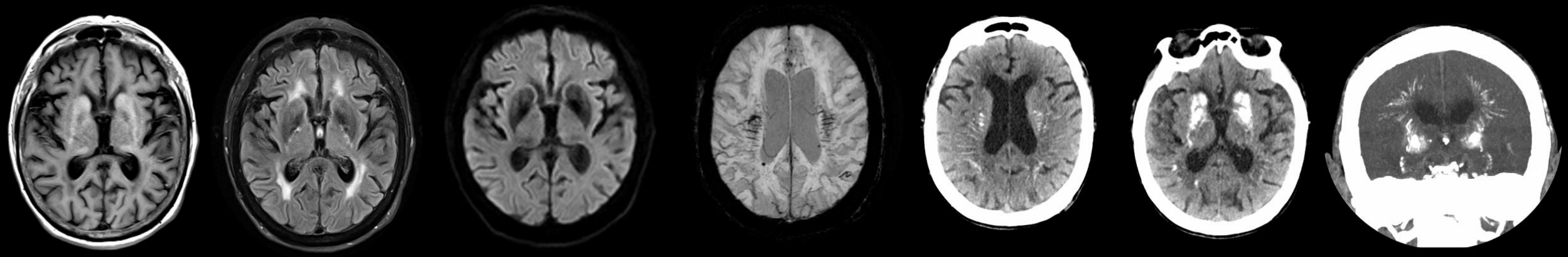
- Abnormal vascular calcium deposition (BG, dentate nuclei, WM)
- Atrophy
- Primary (autosomal dominant) or secondary
  - Fahr Disease (Primary Familial Brain Calcification)
- Fahr Syndrome (secondary to endocrinopathies, mitochondrial disorders, infections, radiation therapy, others)

Clinical Presentation and Diagnosis

- Asymptomatic – Incidental Imaging Finding
- Parkinsonism, other movement disorders (chorea, dystonia)
- Psychosis, depression, progressive cognitive impairment

Diagnostic Evaluation and Management

- Investigation of underlying (treatable) causes
- Family history, genetic testing
- Treatment of underlying conditions, otherwise supportive and symptomatic



Case #4 - Progressively declining gait and speech

Choose the correct statement:

✓ (A) This condition has several possible etiologies, both inherited and acquired 0%

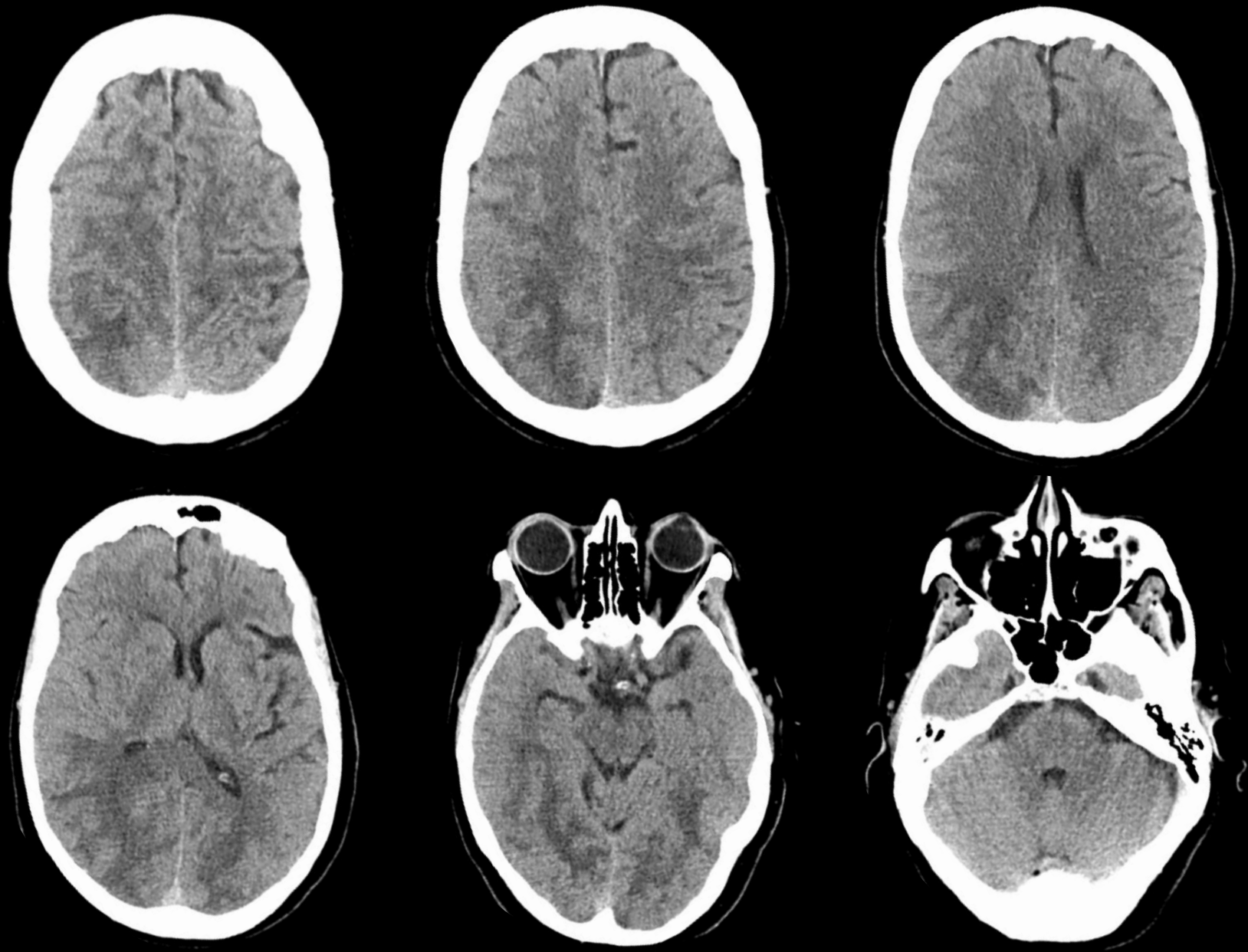
(B) Imaging findings are likely physiological and unrelated to the patient's symptoms 0%

(C) There is no treatment for this condition 0%

(D) This condition can be made worse by anticoagulants 0%

Case #5 - Rapidly Progressive Cognitive Decline

- 75 yo female. Bumping into thing while walking, dizziness, headaches, cognitive decline for a week
- PMHx: Anxiety, hypertension, AD year prior, stable. Donanemab infusions 2 month ago.
- Physical: left visual field defect. Normal vitals and basic labs
- Outside window for thrombolytics and thrombectomy



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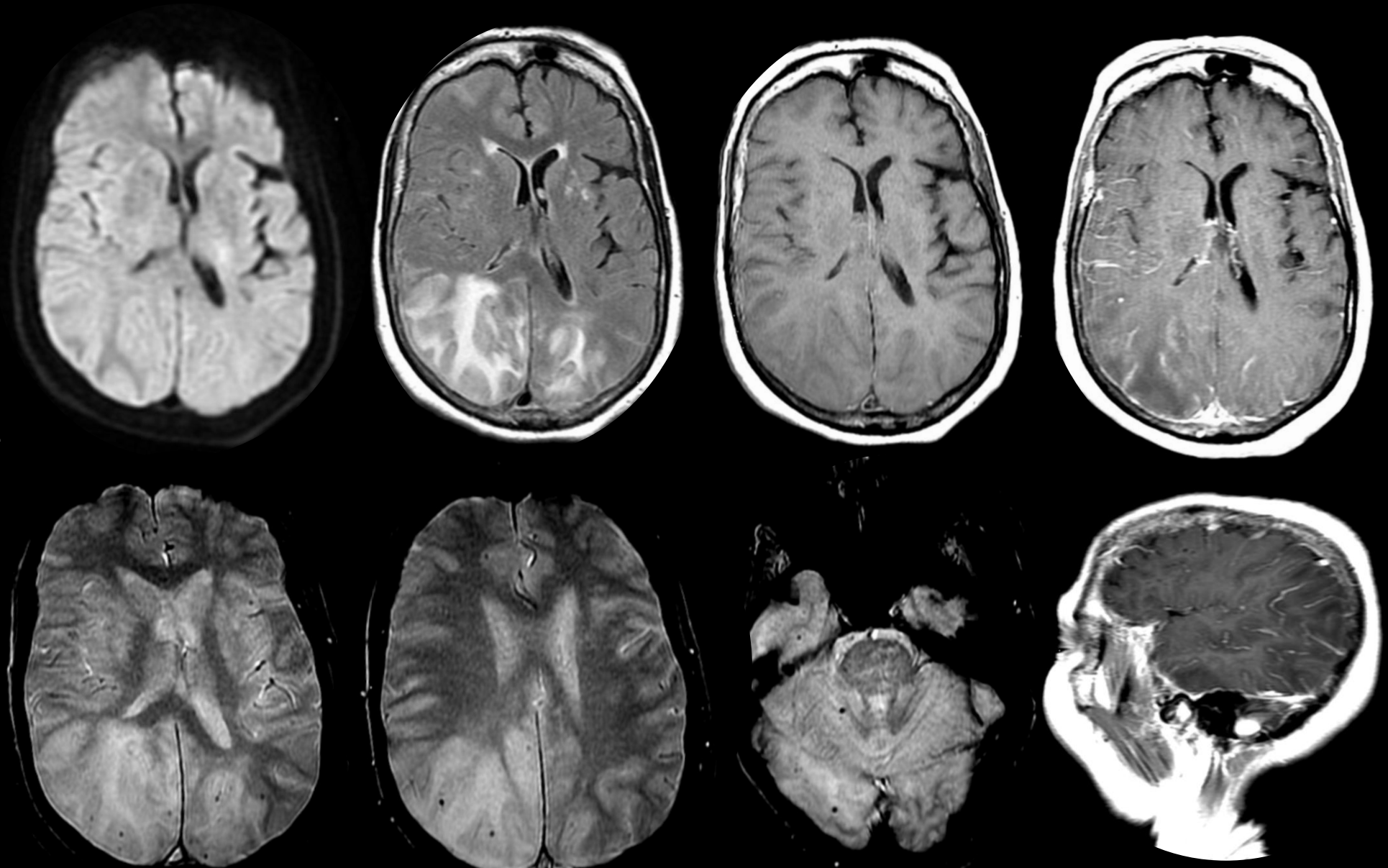

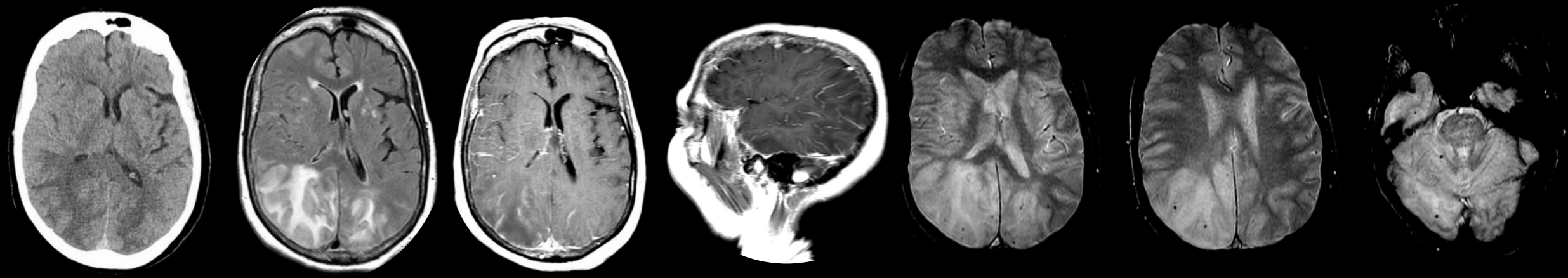

Case #5 - What would be the best initial management ?

(A) CTA + CT Perfusion for evaluation of vascular etiology 0%

(B) Broad spectrum antibiotics 0%

(C) Lumbar puncture and prompt use of acyclovir 0%

(D) Brain Biopsy 0%

(E) Discontinuation of all medications, steroids 0%

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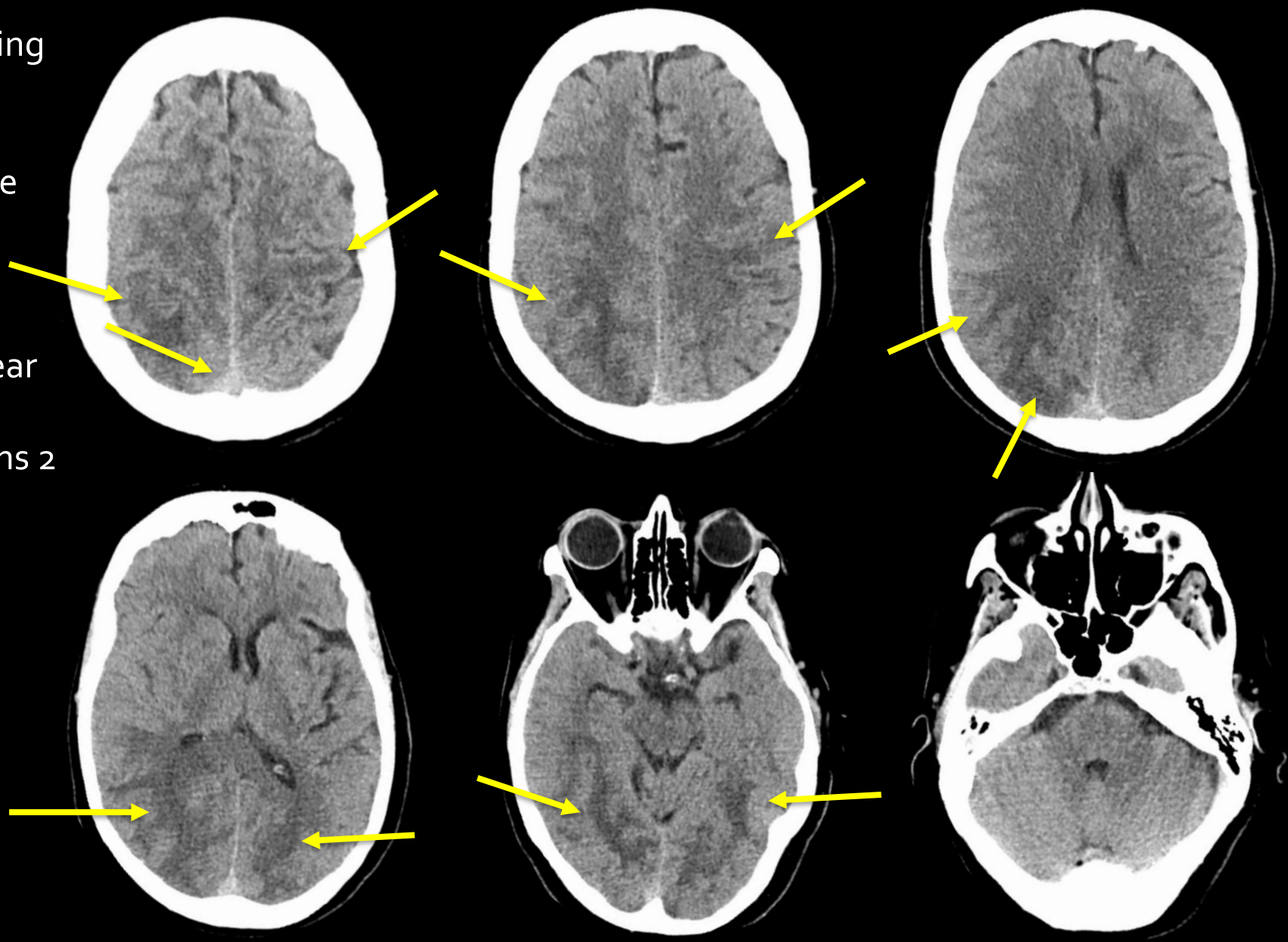

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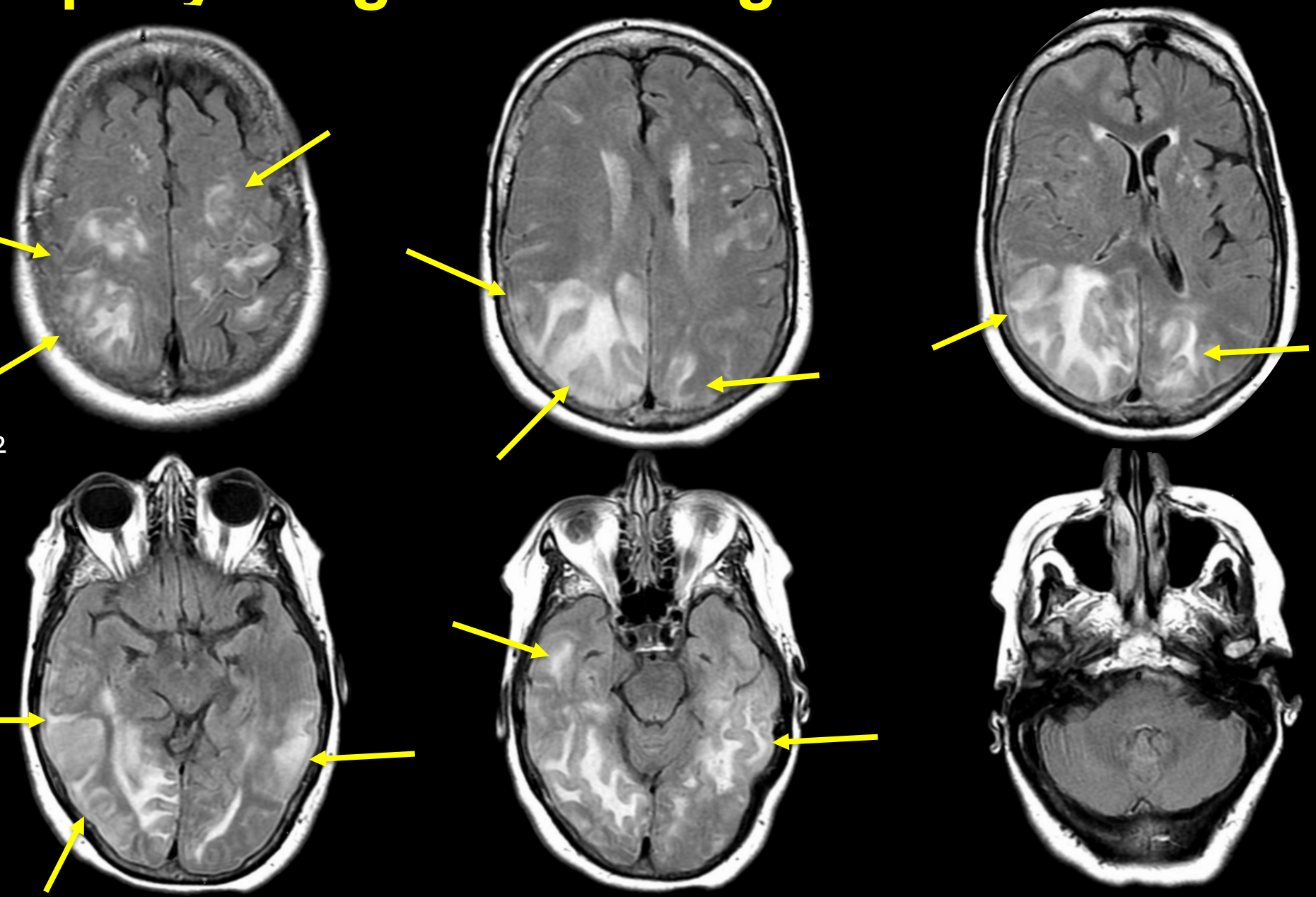

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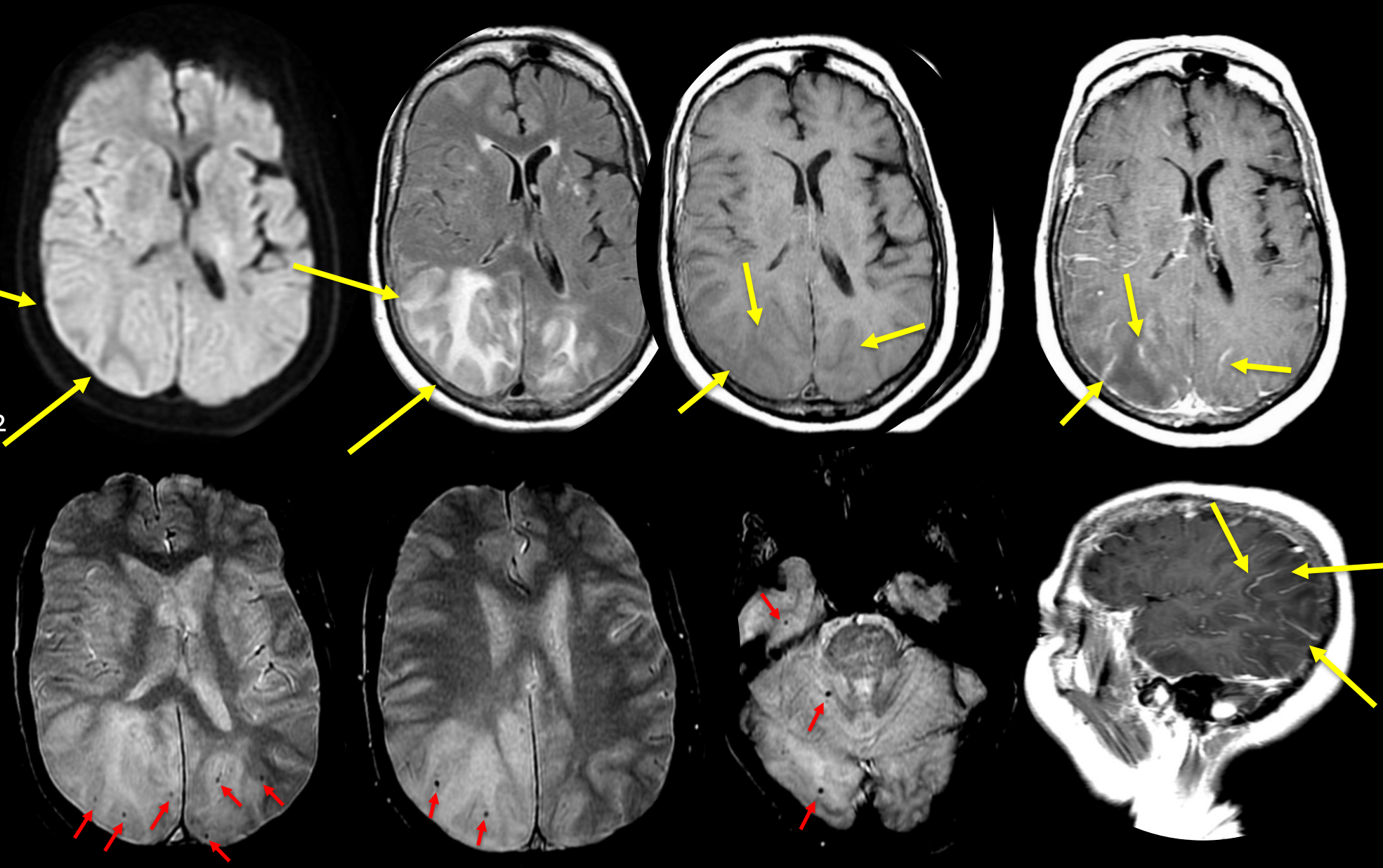

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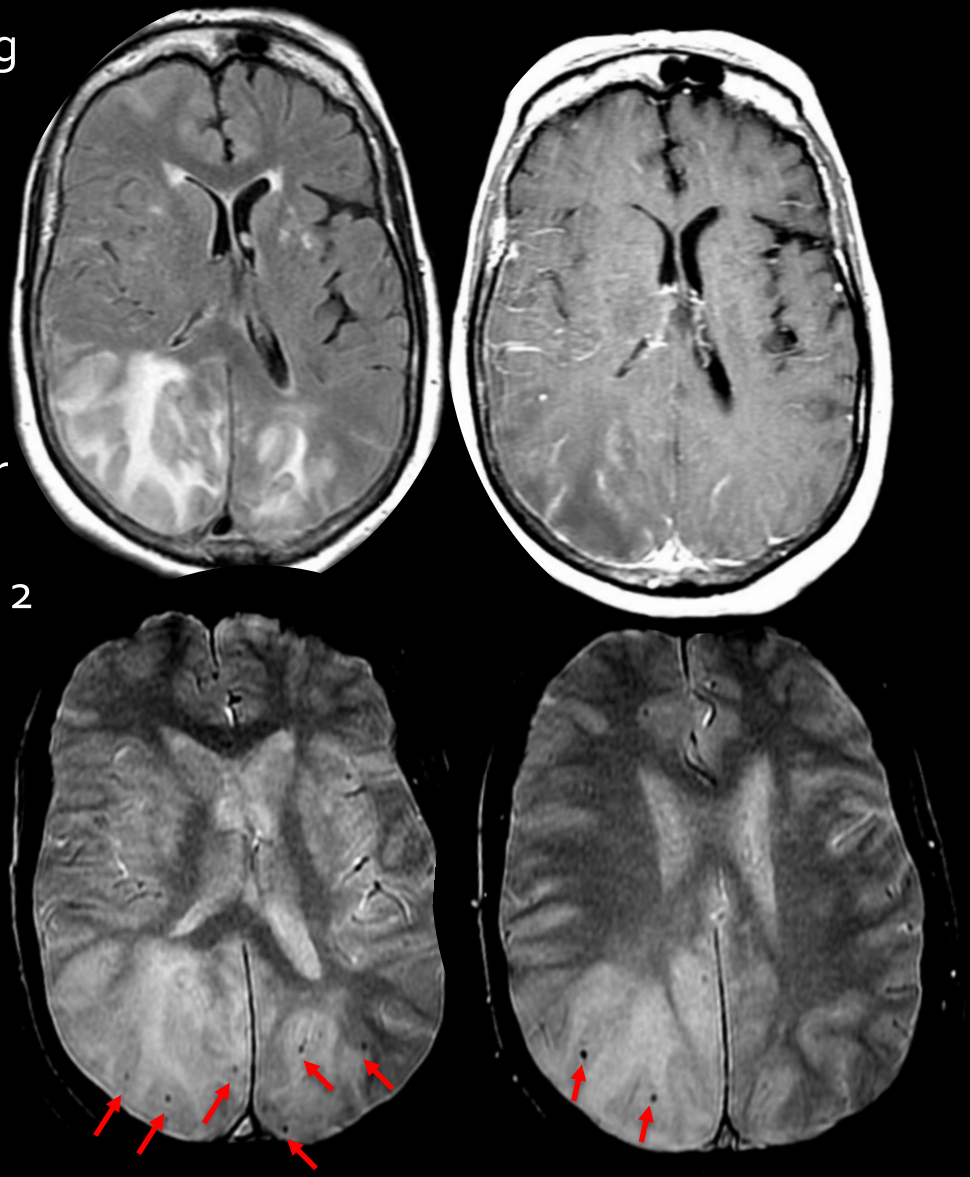


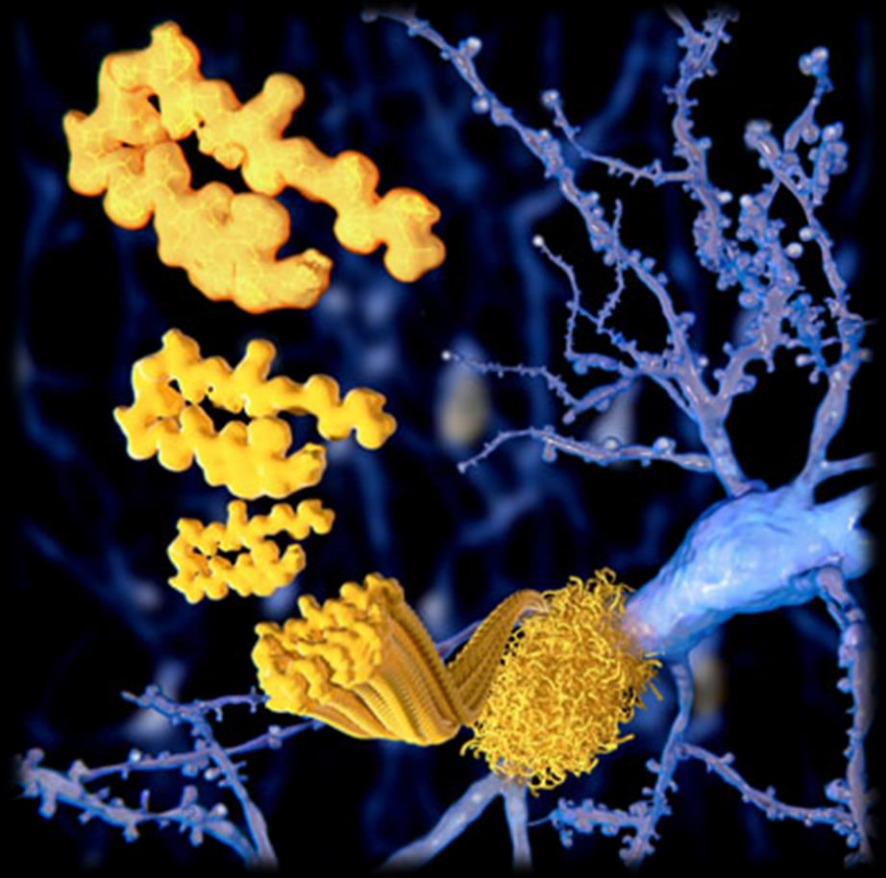
Case #5 Diagnostic Checklist

- Elderly female
- Sudden onset neurological decline
  - gait
  - cognition
  - dizziness and headaches
- Imaging:
  - multifocal brain edema/swelling
  - sulcal contrast enhancement
  - parenchymal microhemorrhages
- PMHx: AD post second infusion Donanemab

Amyloid-Related Imaging Abnormalities (ARIA)

Amyloid-Related Imaging Abnormalities

- Imaging findings during AD treatment
- Novel amyloid lowering monoclonal antibodies
    - lecanemab, donanemab, aducanumab
- Usually incidental, but occasionally symptomatic
    - cerebral edema and irritation
    - headaches, vomiting, confusion, gait abnormalities



Mechanism and Pathology

- not completely understood
- breakdown of BBB due to successful mobilization of vascular amyloid deposition
- Risk Factors:
    - drug dosing
    - number of APOE ε4 alleles
    - use of antithrombic agents

Amyloid-Related Imaging Abnormalities

ARIA-E (edema/effusions):

- **Parenchymal edema:**
  - high T2-FLAIR signal in subcortical white matter and cortex
  - no restricted diffusion
  - subtle leptomeningeal enhancement
- **Sulcal effusions**
  - high FLAIR signal (non-attenuating) in sulci often overlying an area of parenchymal edema

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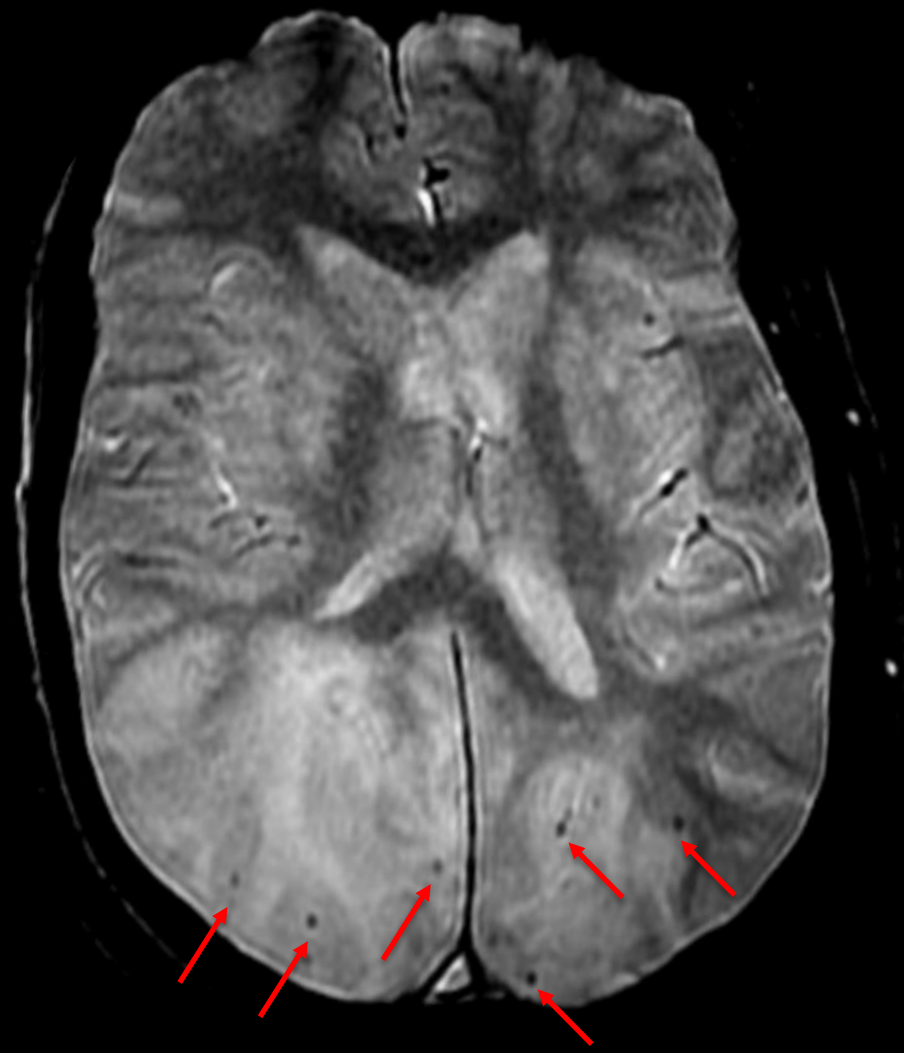
ARIA-H (hemorrhage):

- parenchymal microhemorrhages (most common)
- sulcal hemorrhage (superficial siderosis)



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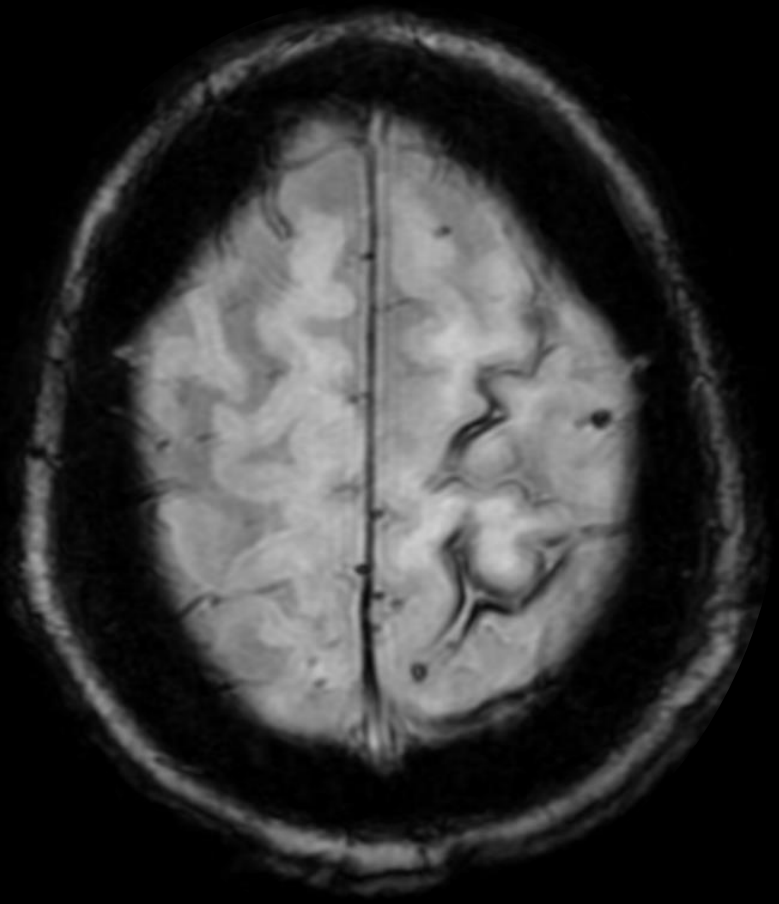

Amyloid-Related Imaging Abnormalities

Table 2: ARIA Grading Criteria

ARIA Type	Mild	Moderate	Severe
ARIA-E	FLAIR hyperintensity confined to sulcus and cortex/subcortical white matter in one location <5 cm	FLAIR hyperintensity 5–10 cm, or more than one site of involvement each measuring <10 cm	FLAIR hyperintensity >10 cm, often with sulcal involvement, may involve one or more sites
ARIA-H microhemorrhage	Four or more new microhemorrhages	Five to nine new microhemorrhages	10 or more new microhemorrhages
ARIA-H superficial siderosis	One focal area of superficial siderosis	Two focal areas of superficial siderosis	More than two focal areas of superficial siderosis

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Amyloid-Related Imaging Abnormalities

Table 3: Management of ARIA-E

	ARIA-E Severity at MRI		
Clinical Severity of ARIA-E	Mild	Moderate	Severe
Asymptomatic	Continue dosing at current dose and schedule	Suspend dosing; once imaging findings resolve, resume dose	Suspend dosing; once imaging findings resolve, resume dose
Mild, moderate, severe, serious ("other medically important event" only)	Suspend dosing; once ARIA-E resolves, same dose treatment can resume	Suspend dosing; once ARIA-E resolves, same dose treatment can resume	Suspend dosing; once ARIA-E resolves, same dose treatment can resume
Serious, except for "other medically important event"	Discontinue dosing	Discontinue dosing	Discontinue dosing

Source.—Reference 48.

Table 4: Management of ARIA-H

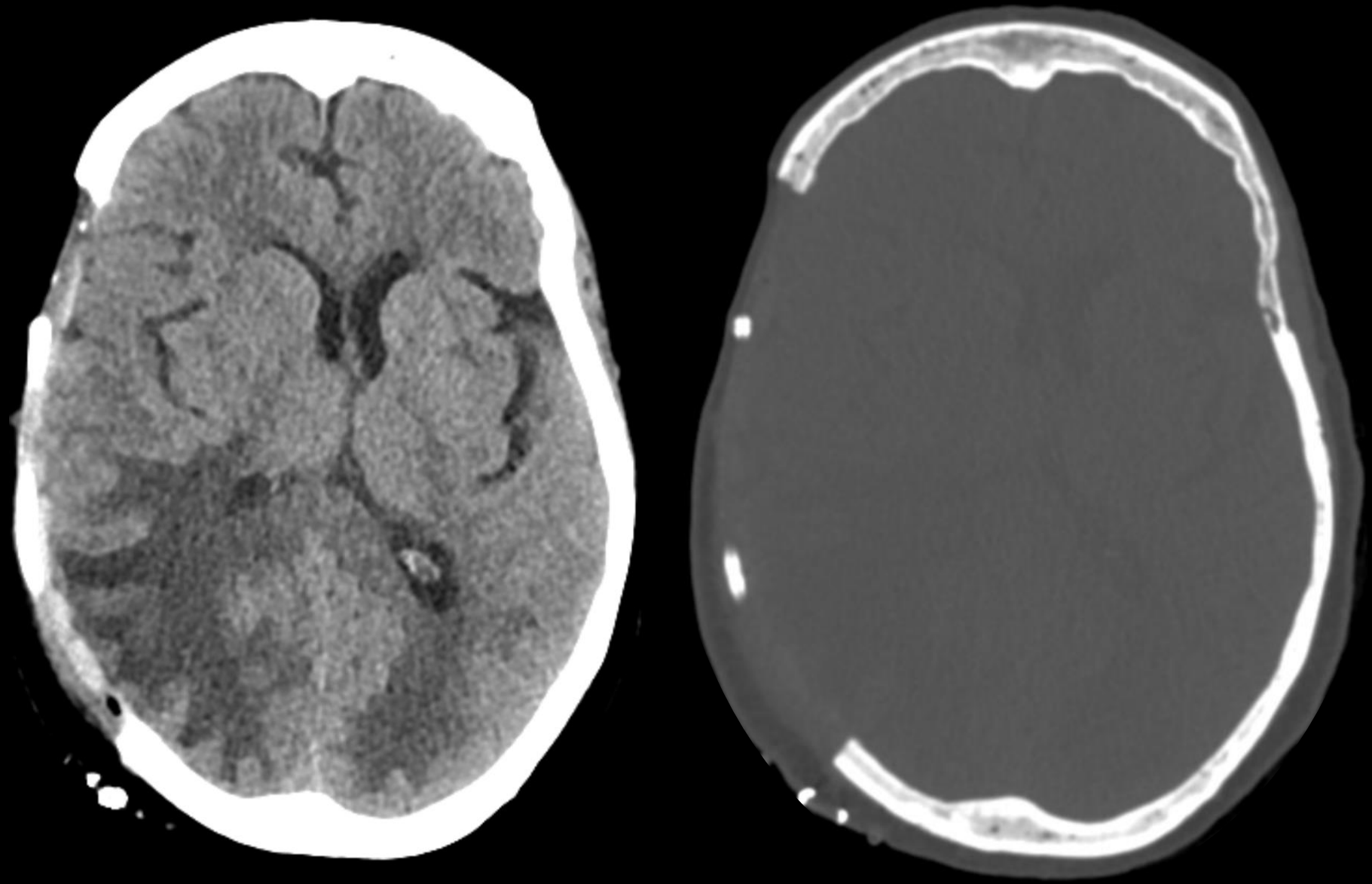
	ARIA-H Severity at MRI		
Clinical Severity of ARIA-H	Mild	Moderate	Severe
Asymptomatic	Continue dosing at current dose and schedule	Suspend dosing; once imaging findings resolve, resume dose	Discontinue
Mild, moderate, severe, serious ("other medically important event" only)	Suspend dosing; once ARIA-H resolves, same dose treatment can resume	Suspend dosing; once ARIA-H resolves, same dose treatment can resume	Suspend dosing; once ARIA-H resolves, same dose treatment can resume
Serious, except for "other medically important event"	Discontinue dosing	Discontinue dosing	Discontinue dosing

Source.—Reference 48.

Agarwal et al Radiographics Sep 2023

Case #5 - Rapidly Progressive Cognitive Decline

- 75 yo female. Bumping into thing while walking, dizziness, headaches, cognitive decline for a week
- PMHx: Anxiety, hypertension, AD year prior, stable. Donanemab infusions 2 month ago.
- Physical: left visual field defect. Normal vitals and basic labs
- Outside window for thrombolytics and thrombectomy



Take Home Points

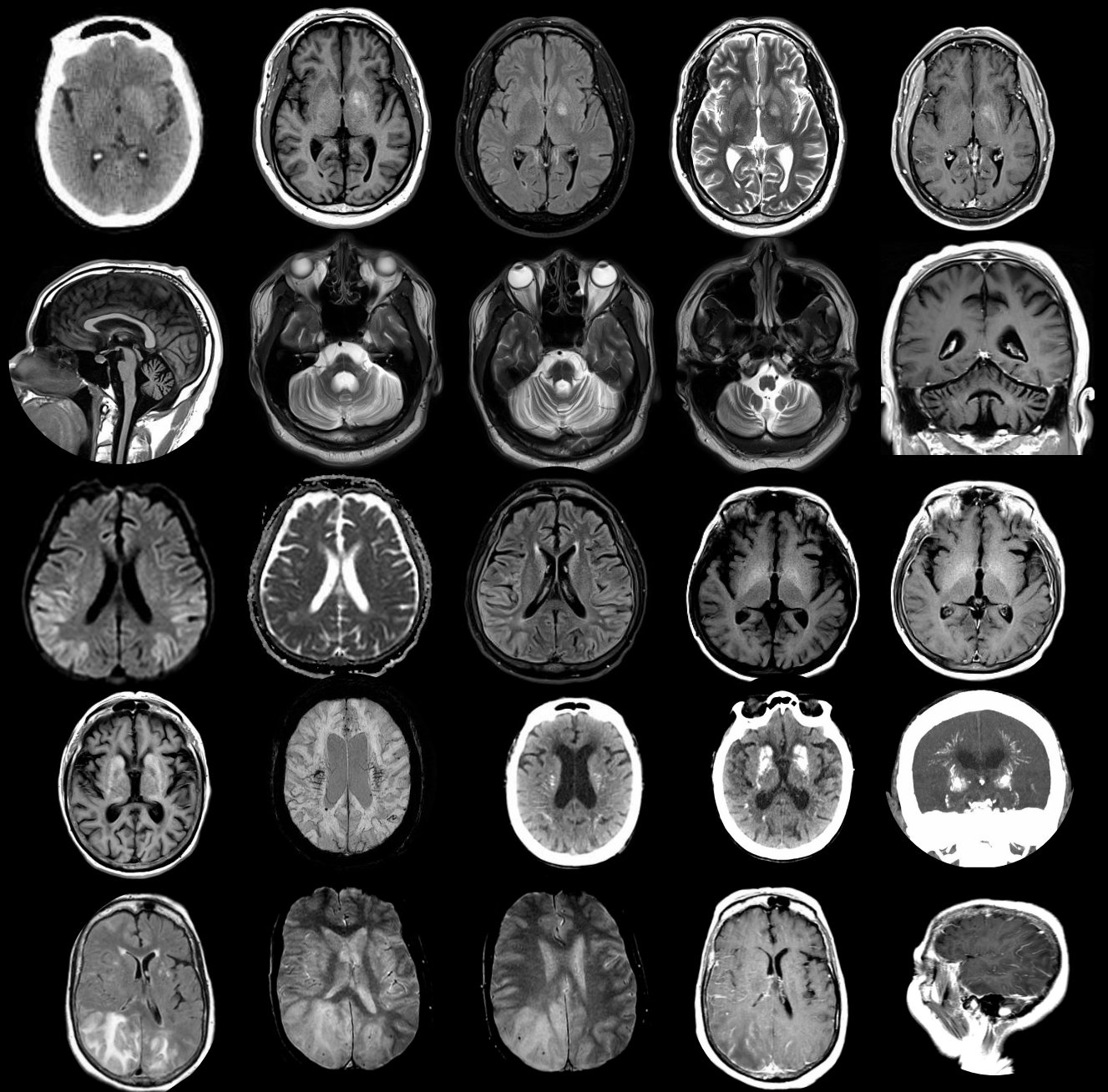
Non-Ketotic Hyperglycemic Hemichorea

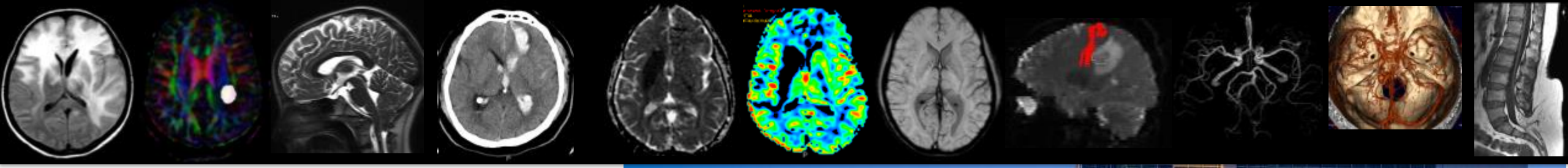
Multiple System Atrophy - Cerebellar

Acute Hyperammomenic Encephalopathy

Autoimmune GFAP Astrocytopathy

Amyloid Related Imaging Abnormalities





Thank you for participating !

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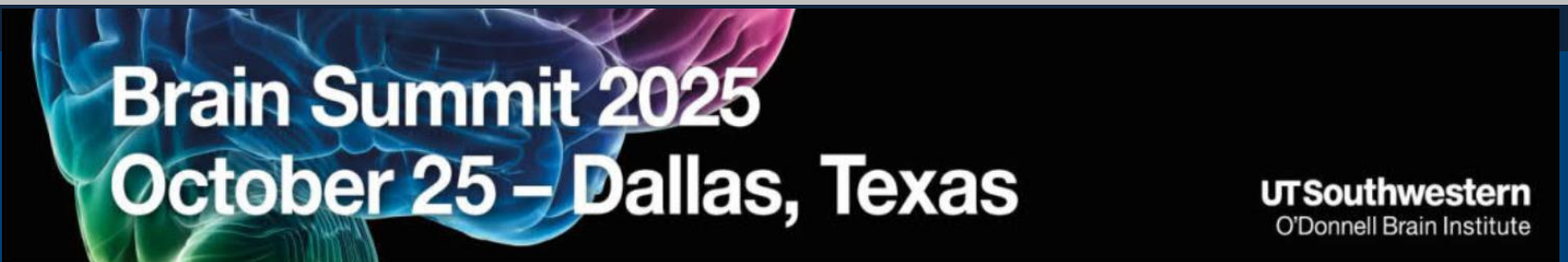

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