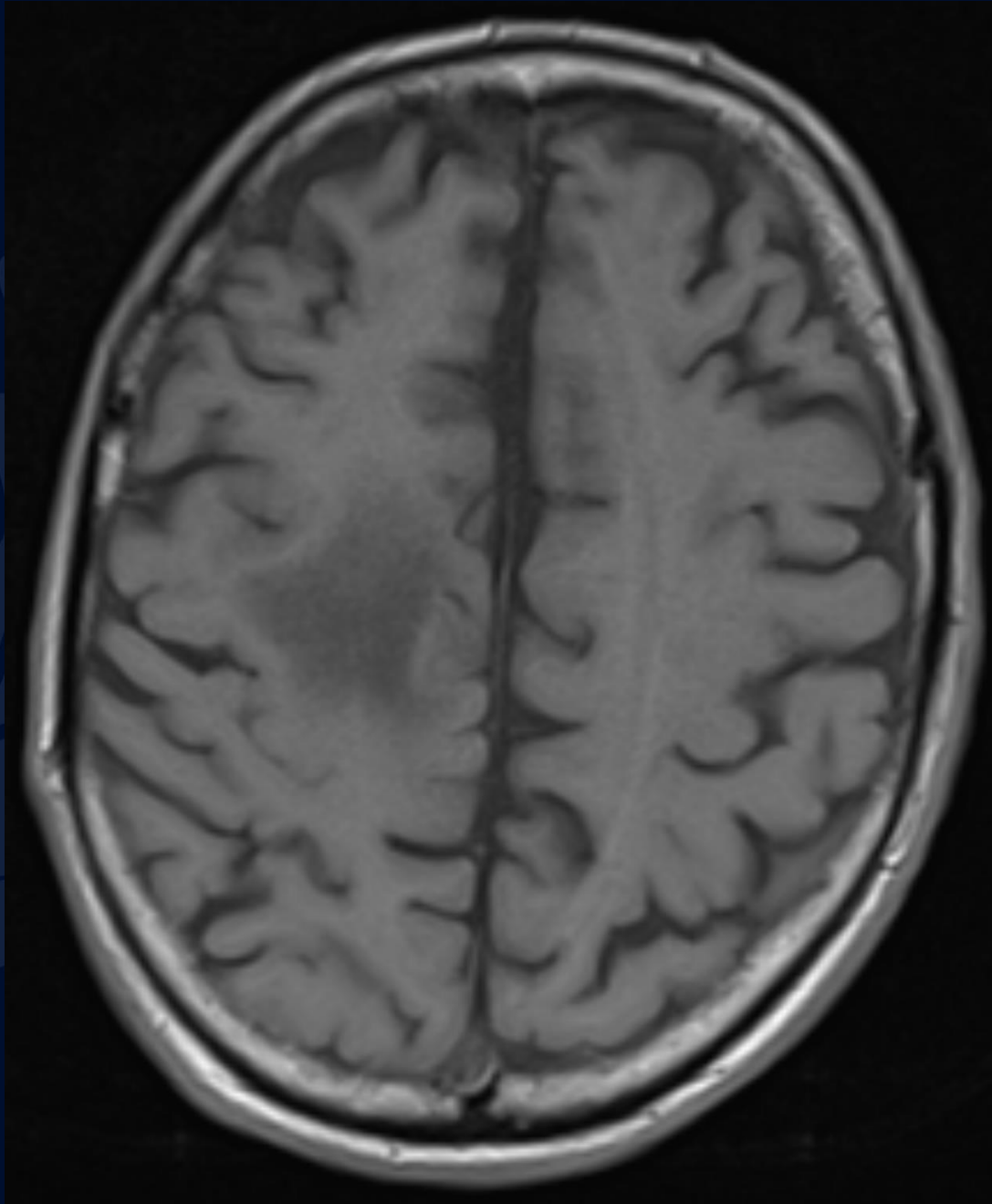


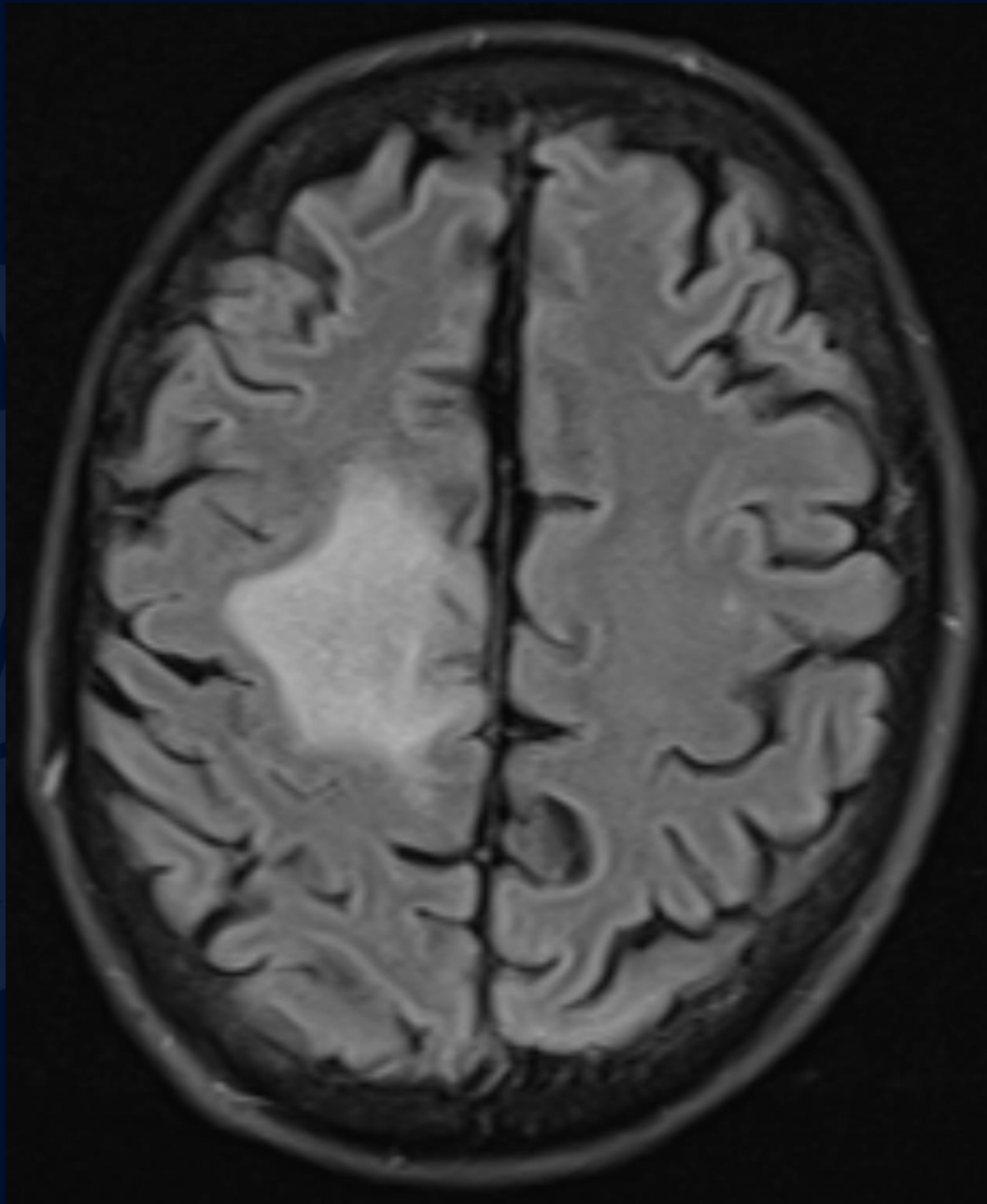
64-year-old male with new
onset seizure, history of
untreated HIV, serologies
pending

Joseph Ryan, MD, PhD
Leo Wolansky, MD

T1



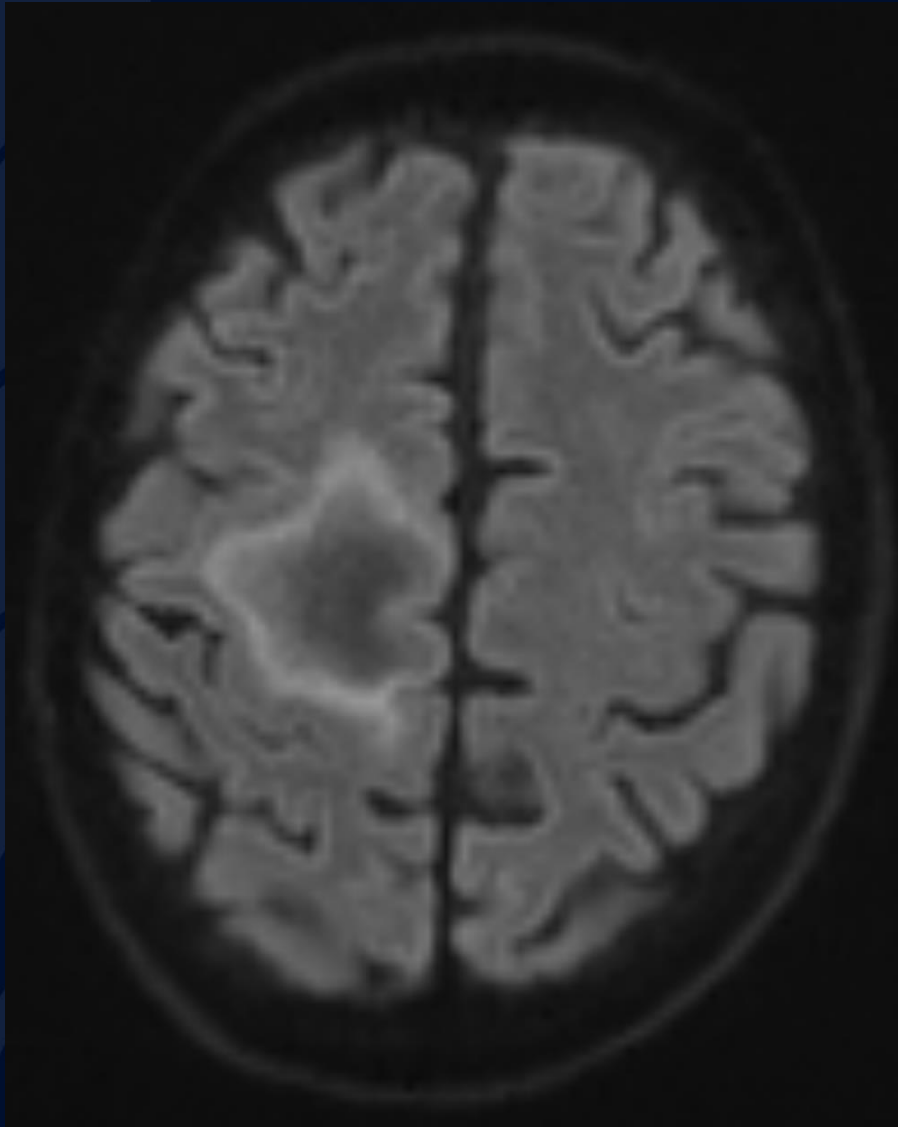
T2 FLAIR



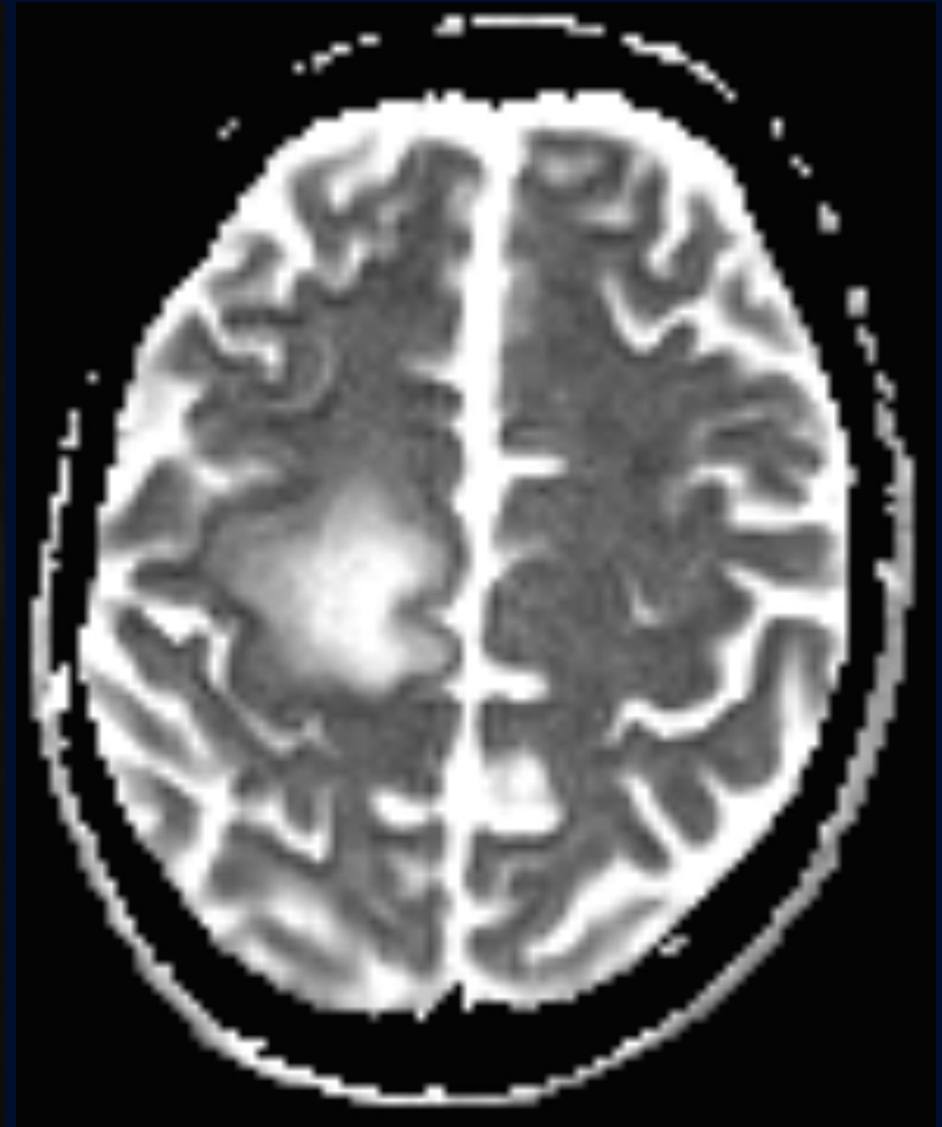
UConn
HEALTH

RADIOLOGY

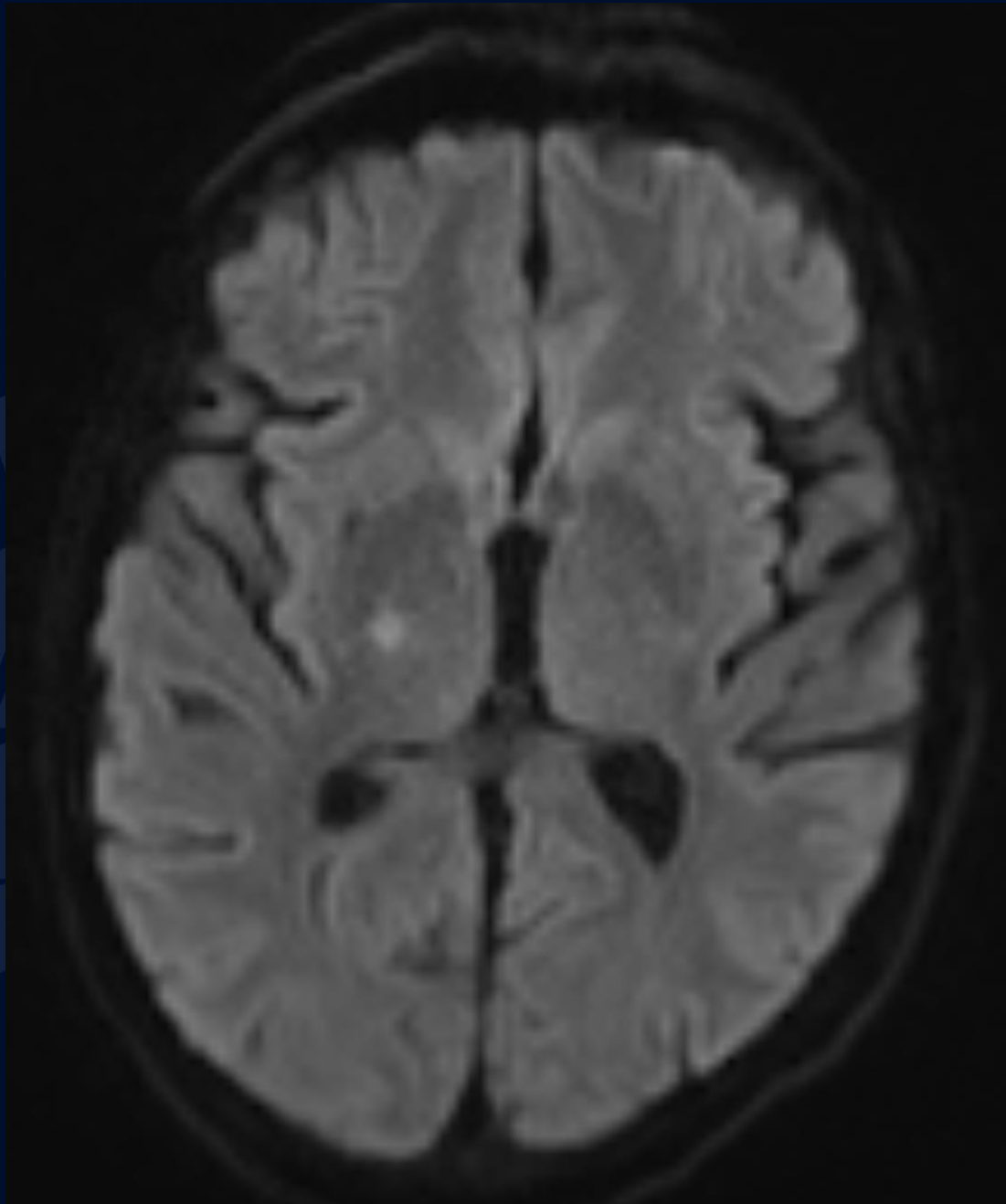
DWI



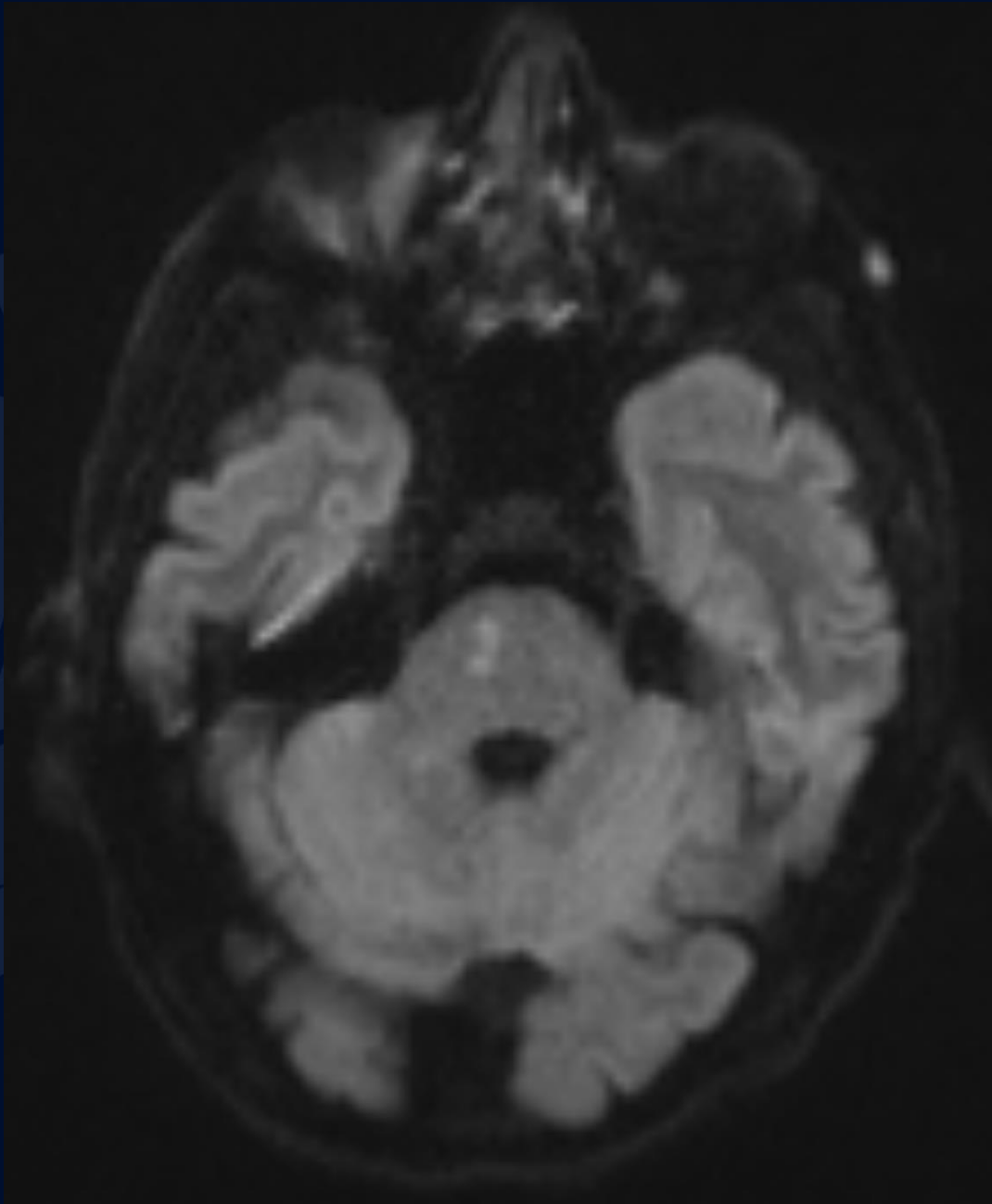
ADC



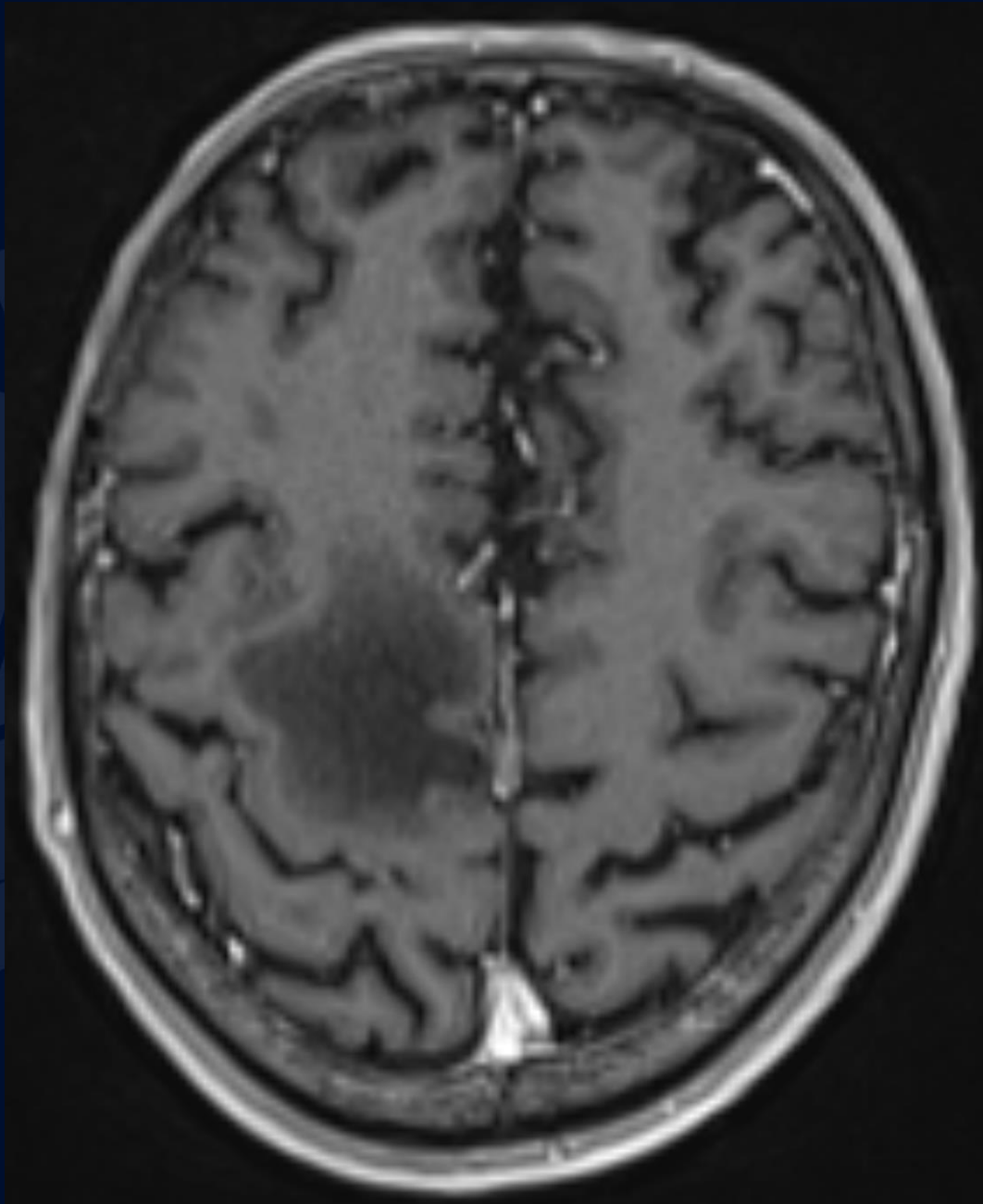
DWI



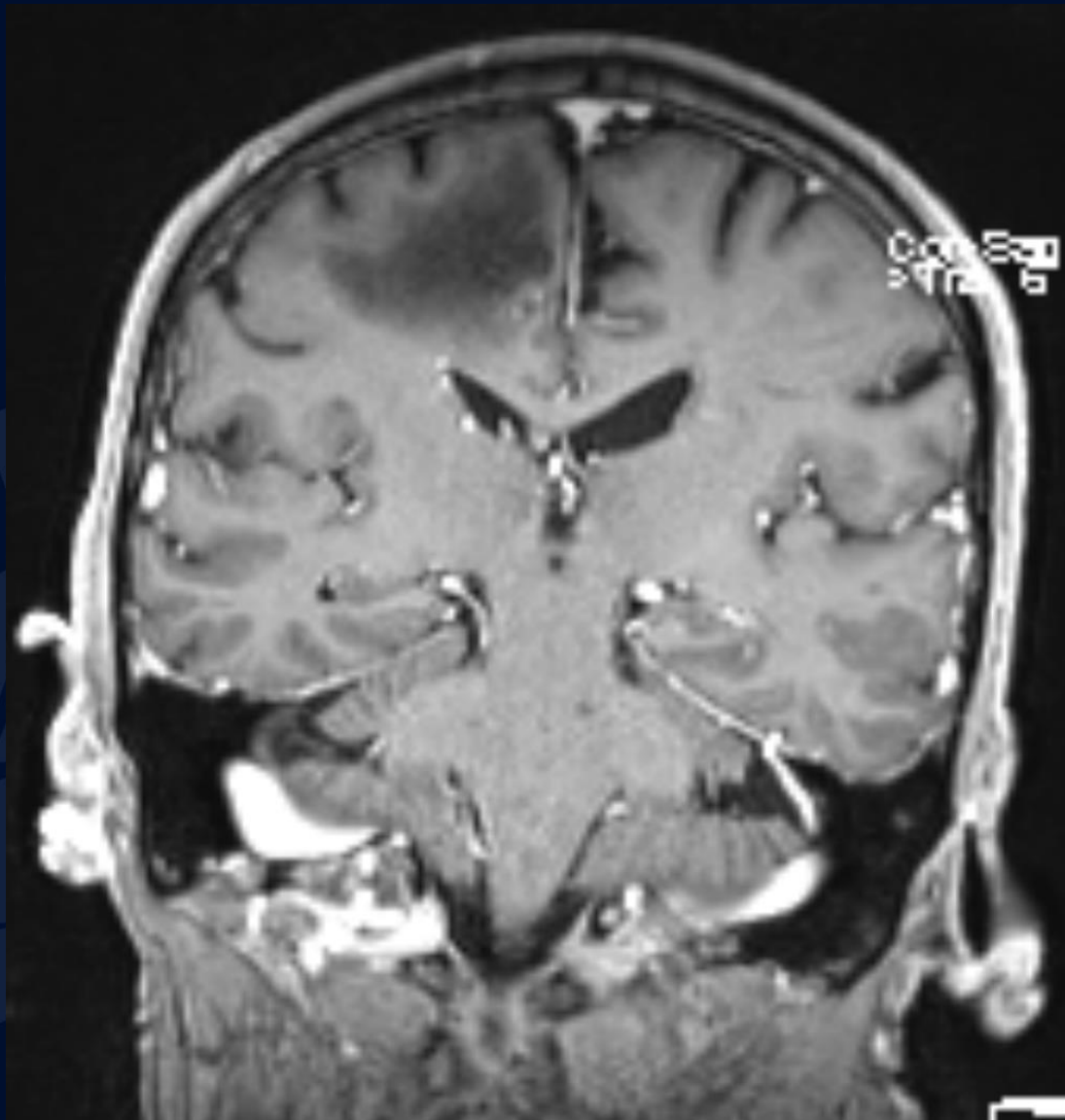
DWI



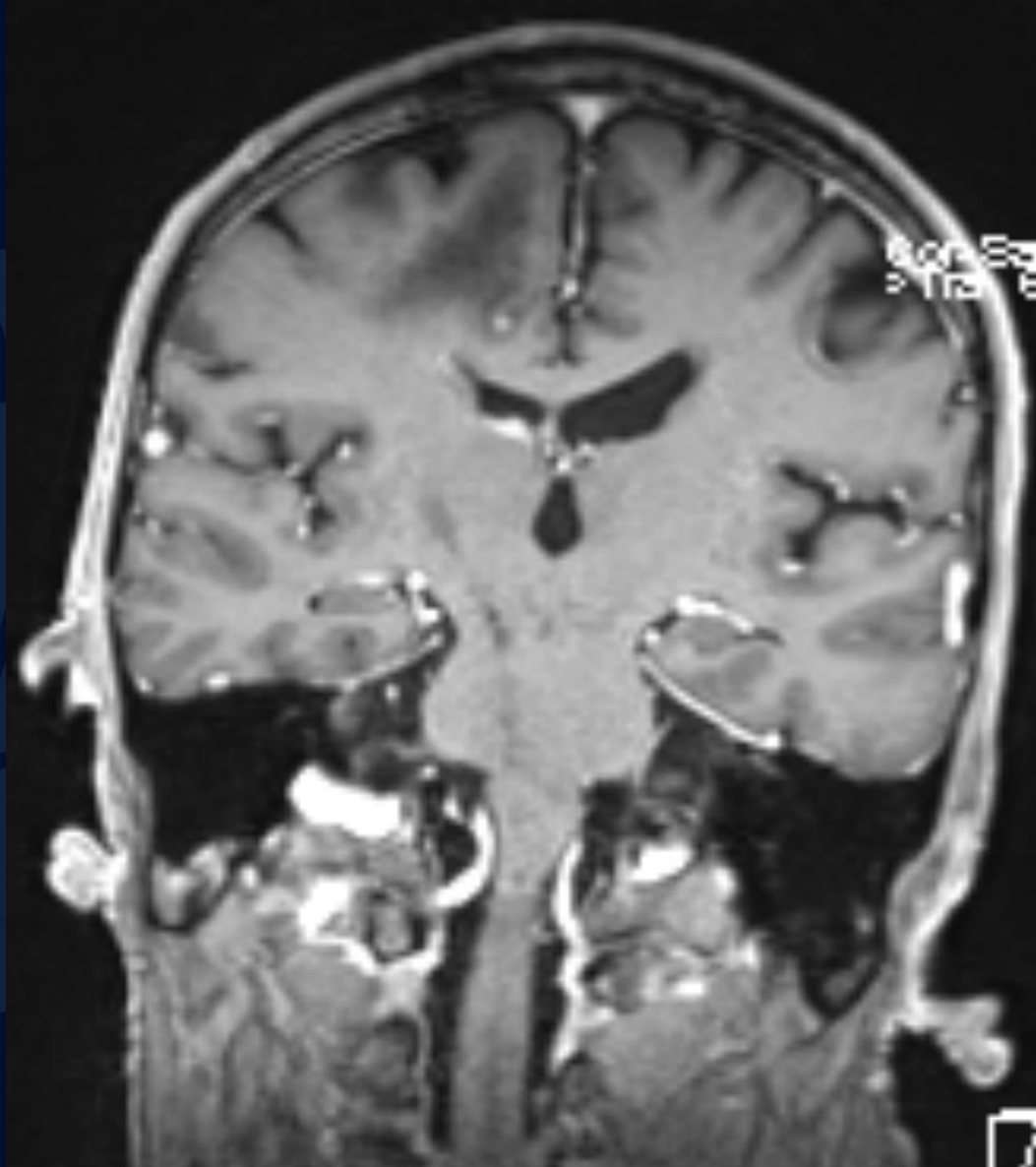
T1 post gadolinium



T1 post gadolinium



T1 post gadolinium





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Progressive Multifocal Leukoencephalopathy (PML)

Background

- Progressive multifocal leukoencephalopathy (PML) is a demyelinating disease which can occur in patient with compromised immune systems.
- Caused by reactivation of the John Cunningham (JC) virus
- JC virus infects oligodendrocytes
- Classically occurs in AIDS patients with CD4 counts of 50-100 cells/uL
- Non-HIV-related PML can be seen in transplant patients, leukemia, other malignancies, inflammatory/autoimmune diseases including SLE and sarcoidosis, isolated CD4 lymphocytopenia, and immunosuppressive monoclonal antibody therapy (natalizumab used in MS, efalizumab, and rituximab)
- Can also occur in patients with recovering immunity, such as in the setting of immune reconstitution inflammatory syndrome (IRIS)

Presentation & Diagnosis

- Patients present with neurological symptoms, generally sparing the optic nerve and the spinal cord
- Specific symptoms can include AMS, motor deficits, limb/gait ataxia, visual symptoms, seizure (2/2 subcortical U-fiber and eventually gray matter involvement in more advanced stages)
- Brain lesions are generally confluent, asymmetrically bilateral, mostly involving supratentorial white matter and thalamus. Basal ganglia, brainstem and cerebellum can also be involved.
- Confirmatory diagnosis made at autopsy

Imaging Features

- CT imaging shows asymmetric focal areas of low attenuation in periventricular and subcortical white matter (vs. HIV encephalopathy, which is more symmetric)
- MR imaging shows similar pattern/distribution of lesions, with little to no mass effect or enhancement, subcortical U-fiber involvement especially in the parieto-occipital region
 - Subcortical U-fiber lesions often have sharply marginated peripheral border and a hazy, ill-defined inner margin
 - T1-hypointense lesions
 - Generally little or no enhancement on T1-postcontrast
 - T2-hyperintense lesions
 - Multiple punctate (Milky Way sign)
 - Parieto-occipital lesion crossing the splenium (Barbell sign)
 - Cerebellar white matter sparing the dentate nucleus (Shrimp sign)
 - Peripheral patchy diffusion restriction can be seen
- Reduced NAA, lactate; increased choline and lipids on MR spectroscopy
- Increased perfusion at leading edges can be seen on MR perfusion

Differential diagnosis

- DDX includes:
 - Multiple sclerosis patient with new demyelinating lesions (periventricular or well-defined inner margins favors MS)
 - HIV/AIDS encephalopathy (more diffuse, symmetric white matter, atrophy, spares U-fibers)
 - Posterior reversible encephalopathy syndrome (PRES) – concurrent hypertension
 - Acute disseminated encephalomyelitis (ADEM) – recent infection or vaccine, lesions enhance
 - Cerebral toxoplasmosis (lesions enhance)
 - Primary CNS lymphoma (lesions enhance)

Prognosis

- Generally poor prognosis with neurological decline, coma, death
- Usually fatal within 1 year, often within 2-6 months, if untreated
- Some patients may benefit from antiretroviral therapy (ART) or high-dose steroids

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