KARNATAKA RADIOLOGY EDUCATION PROGRAM

DEPARTMENT OF RADIO-DIAGNOSIS
JJMMC, DAVANGERE
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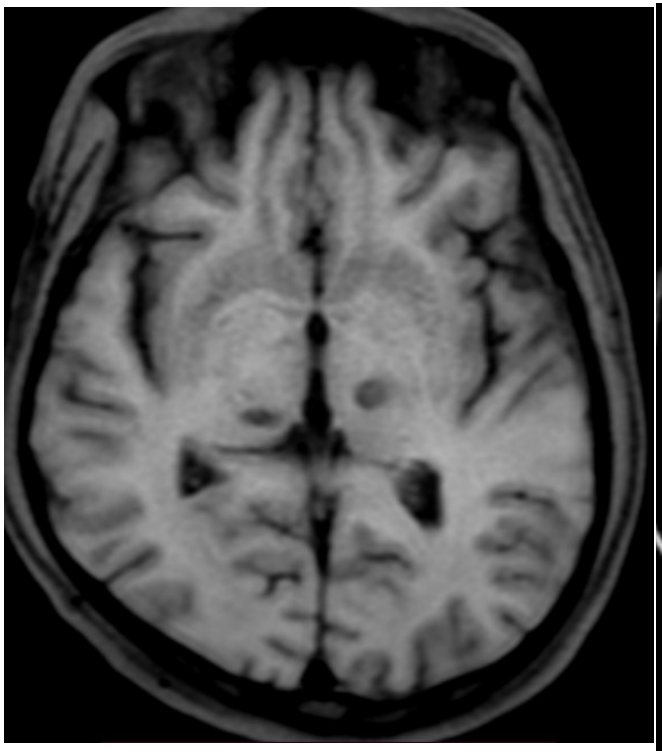
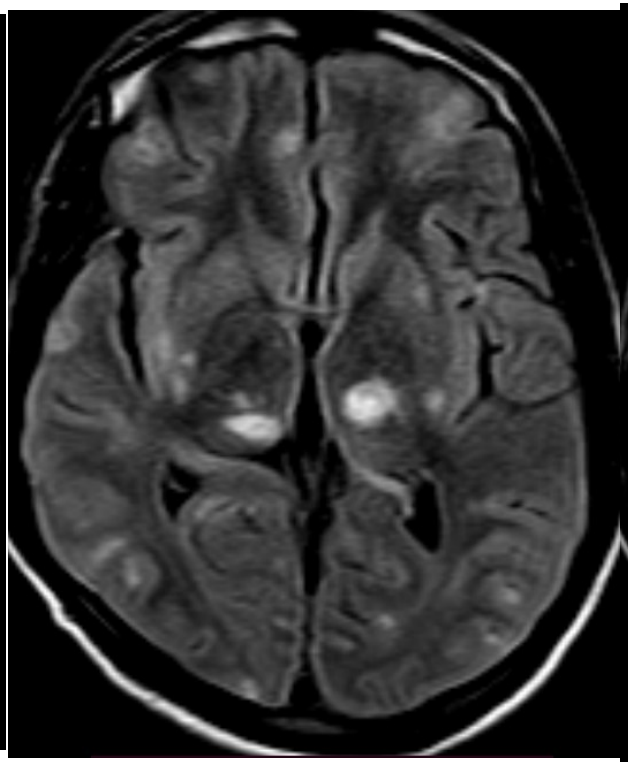
History:

- 33 year old male with headache and complaints of multiple episodes of seizures and altered sensorium since 6months
- Seizures associated with LOC and post- ictal confusion.
- Not a k/c/o seizure disorder
- K/C/O RVD positive status on irregular treatment since 6 years
- K/C/O old PTB (Completed treatment) and Alcoholic since 15years.
- No h/o fever / vomiting / trauma.

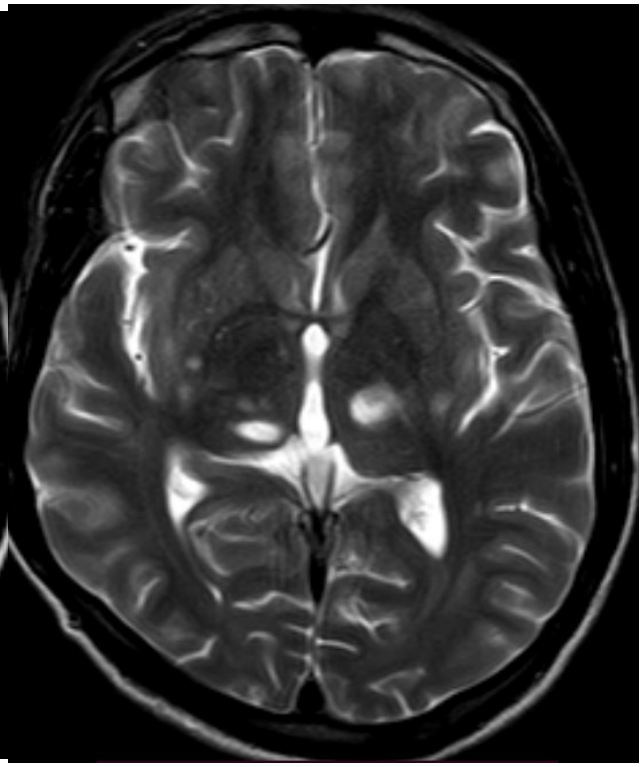
T1



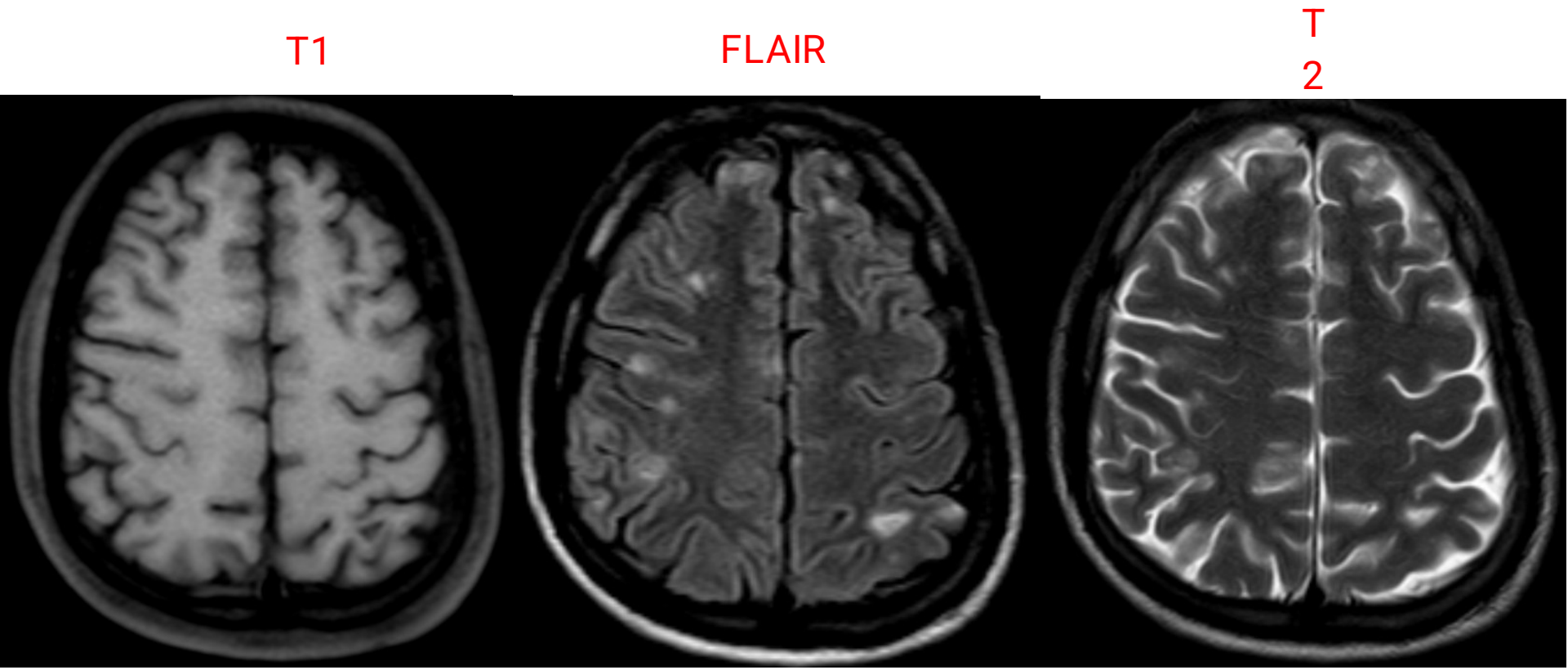
FLAIR



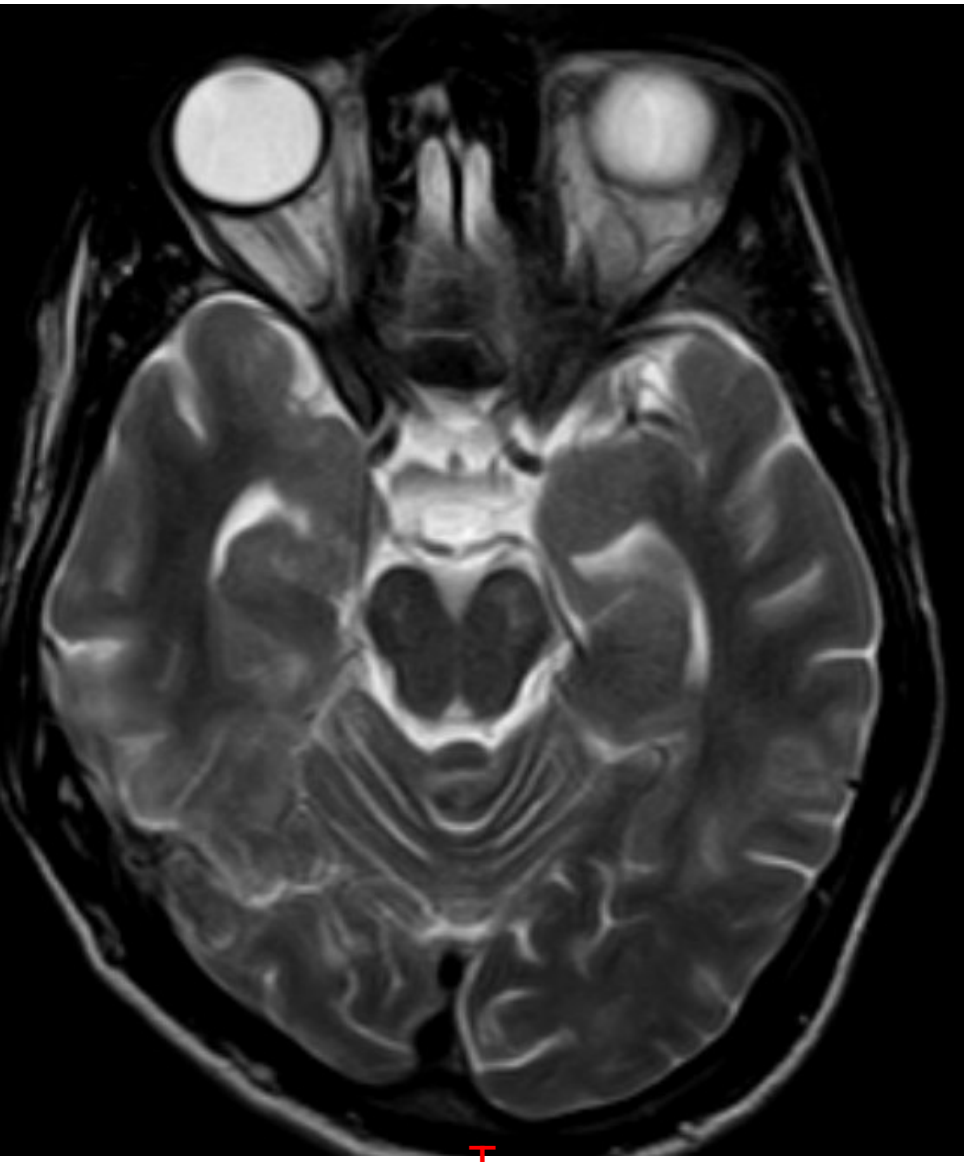
T2



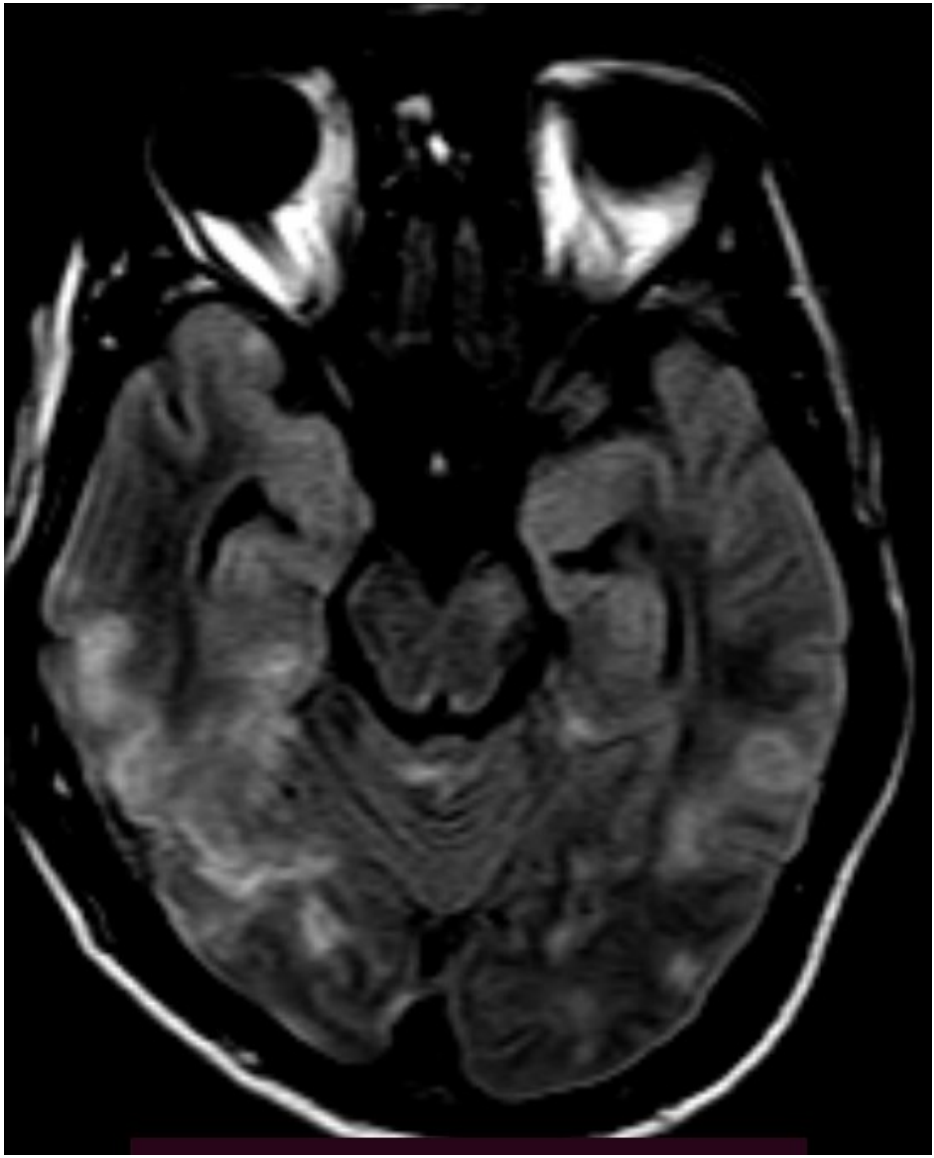
Bilateral diffuse, asymmetrical, non- enhancing, multifocal areas of T2/FLAIR hyperintensities with corresponding T1 hypointensity, noted in cortical and grey white matter junction of bilateral thalamus, bilateral lentiform nucleus



Bilateral diffuse, asymmetrical, non- enhancing, multifocal areas of T2/ FLAIR hyperintensities with corresponding T1 hypointensity, noted in cortical and grey white matter junction of bilateral cerebral hemispheres.



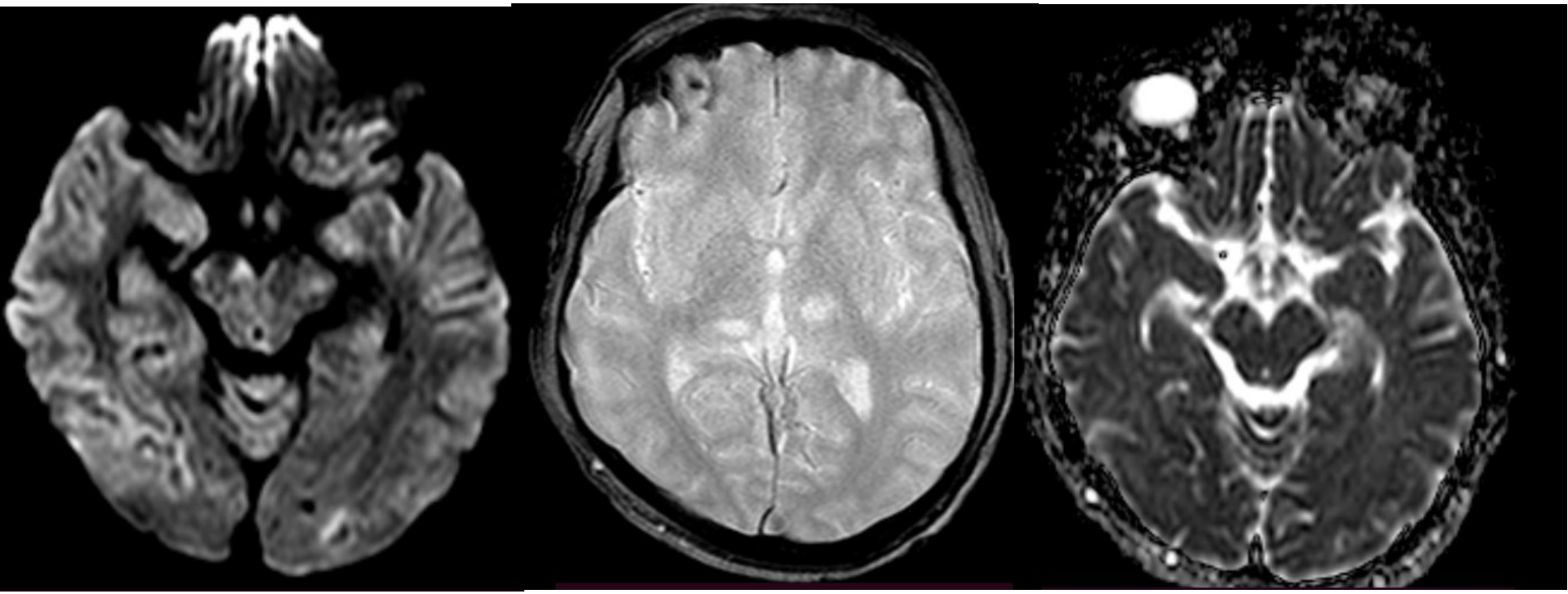
T2



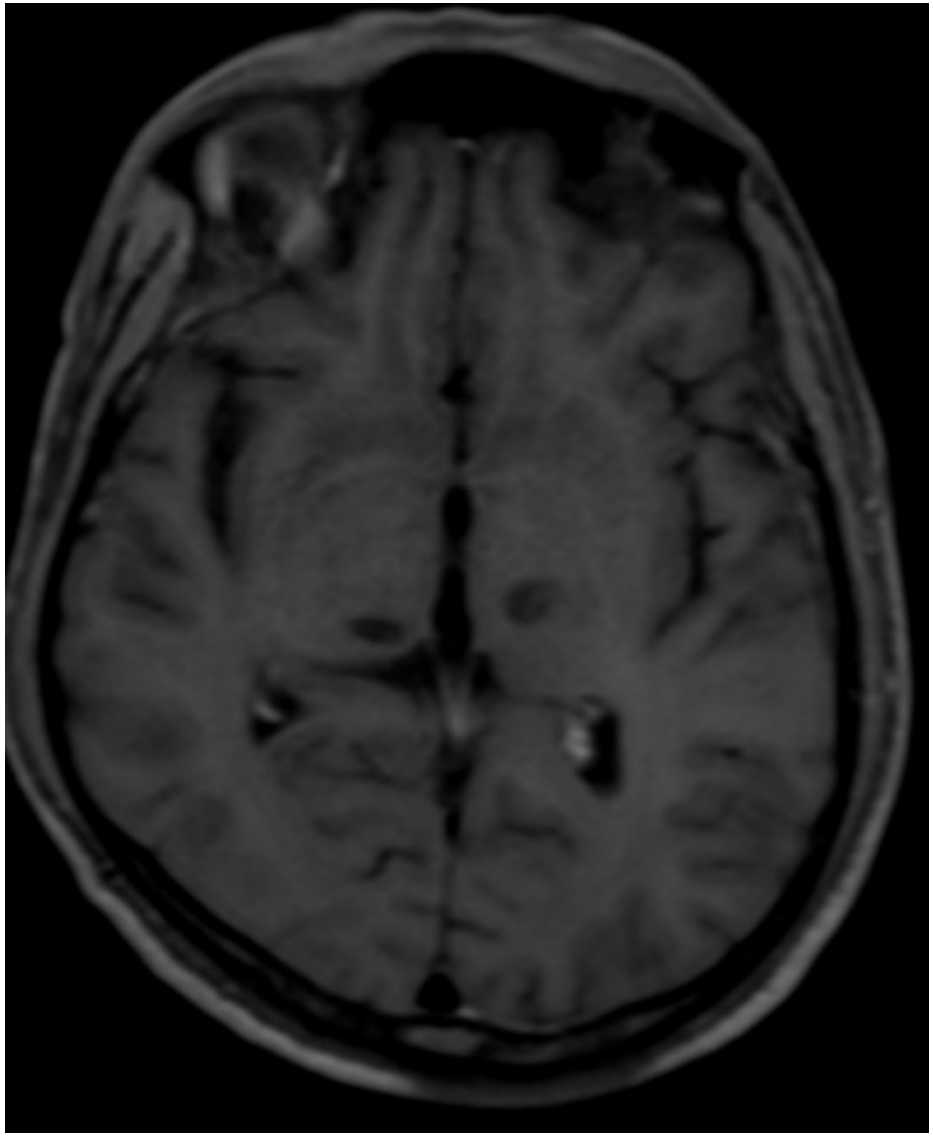
FLAIR

Other similar focal lesion right hemi mid brain

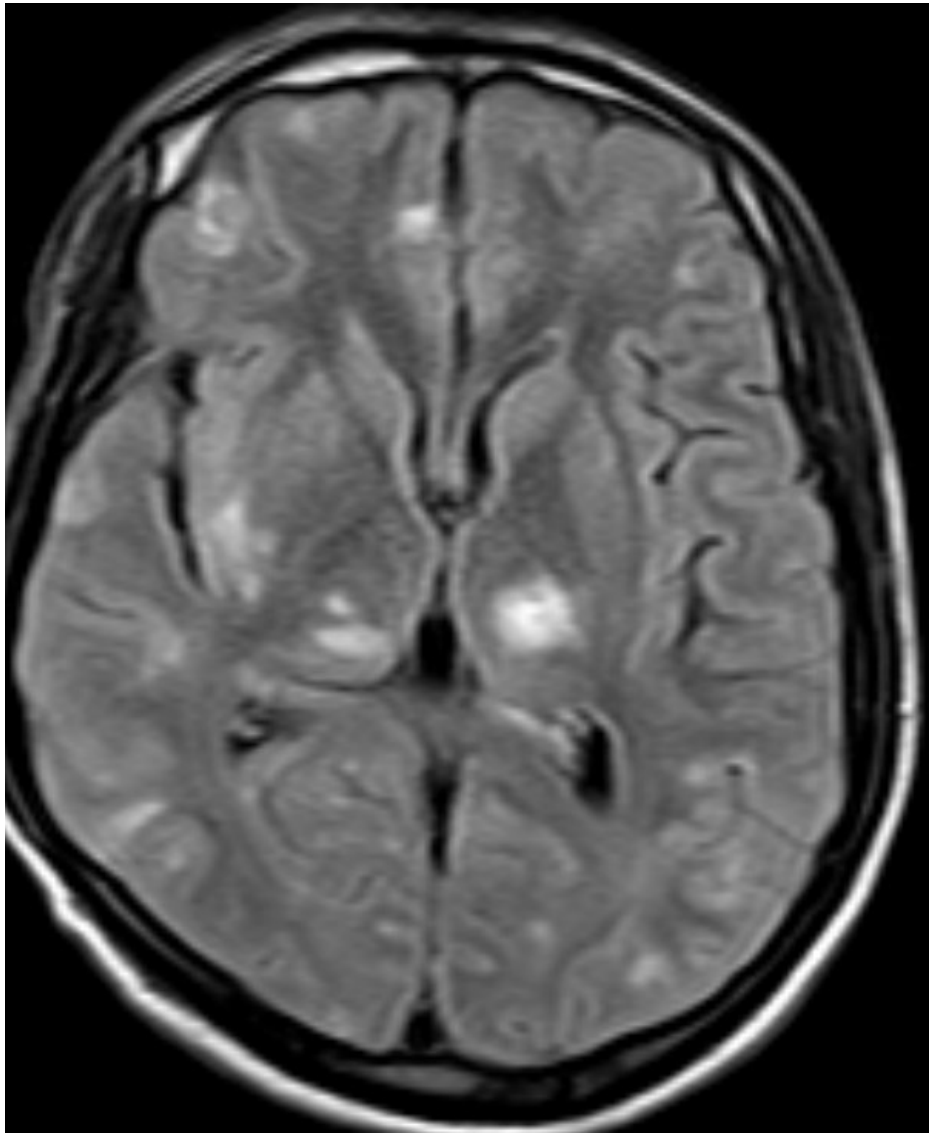
DWI FFE ADC



No diffusion restriction or blooming noted in GRE sequences



Post contrast T1



Post contrast FLAIR

Post contrast T1 FLAIR – no ehnancement

No ring enhancing lesions

Findings:

- Bilateral diffuse, asymmetrical, non- enhancing, multifocal areas of T2/FLAIR hyperintensities with corresponding T1 hypointensity, noted in cortical and subcortical white matter and grey white matter junction of bilateral cerebral hemispheres
- Similar focal lesions noted in bilateral thalamus, bilateral lentiform nucleus, right cerebral peduncle, cerebellar vermis, right cerebellar hemispheres, right hemi mid brain , left hemi pons and right hemi medulla.
- In a k/c/o HIV positive

Findings consistent with Progressive Multifocal Leukoencephalopathy

Progressive Multifocal Leukoencephalopathy

- Progressive multifocal leukoencephalopathy (PML) is a demyelinating disease which results from the reactivation of JC virus infecting oligodendrocytes in patients with compromised immune systems.
- Pre-disposing factors include:
  1. HIV-AIDS (PML is an AIDS defining illness and HIV infection accounts for almost 85% of the total cases)
  2. Post-transplant
  3. Leukaemia and solid organ malignancies
  4. immunosuppressive monoclonal antibody therapy
  5. it is also seen in patients whose immune function is recovering, PML-IRIS
- Patients typically experience insidious onset and steady progression of focal symptoms that include behavioral, speech, cognitive, motor and visual impairment

Imaging Features

- PML can appear as solitary or multifocal widespread lesions. Any area of the brain can be affected, although the supratentorial lobar white matter is the most commonly affected site.
- The posterior fossa white matter—especially the middle cerebellar peduncles—is the second most common location. In occasional cases, a solitary lesion in the subcortical U-fibers is present.
- Multifocal, bilateral but asymmetric, irregularly shaped lesion which are heterogeneously hyperintense on T2WI and hypointense on T1WI.
- Lesions typically extend into the subcortical U-fibers all the way to the undersurface of the cortex
- Appearance on DWI varies according to disease stage.
  - In newly active lesions, DWI restricts strongly.
  - Slightly older lesions show a central core with low signal intensity surrounded by a rim of higher signal intensity.
  - Chronic, "burned-out" lesions show increased diffusion due to disorganized cellular architecture
- "Classic" PML generally does not enhance on T1C+ scans
- Enhancement can be seen in PML-IRIS, AIDS with ART, and in patients on

Differential diagnosis of PML

- HIV encephalitis (HIVE). HIVE demonstrates more symmetric white matter disease while sparing the subcortical U-fibers.
- IRIS is usually more acute and demonstrates strong but irregular ring-like enhancement.

Treatment and Prognosis

- There is no specific treatment for progressive multifocal leukoencephalopathy (PML). Therefore, the main approach is restoring the host adaptive immune response, a strategy that appears to prolong survival.
- In patients with PML and HIV infection, initiation and optimization of effective antiretroviral therapy (ART) therapy is crucial.
- For patients with multiple sclerosis who develop PML during natalizumab treatment, natalizumab should be immediately discontinued. In addition, a course of plasma exchange is suggested In these patients.
- For patients who develop PML-IRIS and have both neurologic deterioration and clinical or radiologic evidence of brain swelling, high-dose glucocorticoid therapy is suggested.

THANK YOU