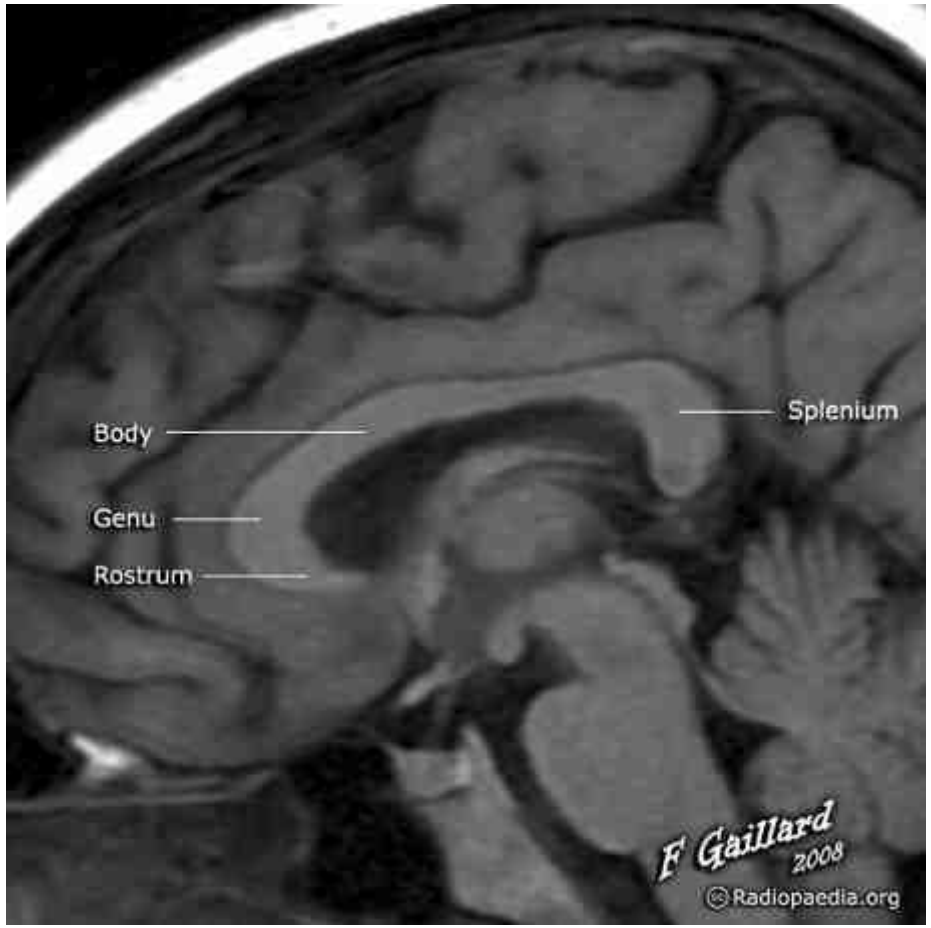
Marchiafava – Bignami Disease (MBD) and Diffusion Tensor Image (DTI) Tractography

Priscilla Chukwueke, MD, MPH

INTRODUCTION

- Definition: A rare CNS disease characterized by demyelination of the Corpus Callosum.
- Discovered by two Italian pathologists Marchiafava and Bignami in 1903.
- It affects the middle 2/3rd of the Corpus Callosum
- Common age of onset: 45 y/o men
- Deficiency of Vitamin B complex has been implicated.
- It has a high mortality rate (Of the 250 cases reported in 2001, 200 died (80%), 20 had a favorable outcome and 30 were severely disabled).

Corpus Callosum



Common Presenting Symptoms

- Commonly found in middle aged men with alcohol use disorder and malnutrition
- The onset is sudden
- Stupor
- Muscle rigidity, Trismus, coma or seizures
- Some may present with gait abnormality (spasticity) hemiparesis, aphasia, apraxia and incontinence
- Psychiatric symptoms such as depression, agitation and impaired mental status and confusion

SUBTYPES OF MBD

- Type A: Is a more severe form
- radiologic findings include involvement of the entire corpus callosum,
- major impairment of consciousness and poor outcome
- Type B – Less severe than type A
- Characterized by normal or mildly impaired mental status
- radiologic features are partial or focal callosal lesions

Differential Diagnosis

- Wernicke Encephalopathy: Ataxia, opthalmoplegia, nystagmus and confusion
- Stroke
- Cytomegalovirus (CMV)
- Progressive multifocal leukoencephalopathy is a rare infection of the brain that is caused by the JC virus.
- Herpes Simplex Virus (HSV) encephalitis
- HIV related encephalopathy (as in PML).
- Multiple Sclerosis

Differential Diagnosis Contd.

- Meningitis
- Toxoplasmosis
- Post ictal
- Korsakoffs dementia with polyneuropathy and confabulation
- Multiinfarct dementia
- Alzheimers disease in the chronic form

Case Presentation

- Patient is a 30 year old left handed African-American with history of hypertension, diabetes type I, hypothyroidism, alcohol use disorder.
- He was found on the floor by his uncle, incontinent of urine, had slurred speech and left-sided weakness.
- On presentation, he had altered mental status,
- followed a few commands,
- speech was slurred,
- left facial asymmetry,
- left hemiparesis,
- cerebellar gait
- Aggressive, agitated and verbalized suicidal ideation

Investigations and consultations

- EEG showed abnormal findings consistent with structural abnormality
- Lab results were significant for anemia Hb/Hct 12.5/38.4, Albumin was low 2.4
- HSV PCR was negative
- VDRL: negative
- CMV & JCV: negative
- HIV: negative
- Toxoplasmosis negative

Imaging

- CT brain w/o contrast: No acute ischemic stroke or hemorrhage
- CT angio: Not diagnostic due motion artifact
- MRA head: No gross aneurysm or vessel wall irregularity
- MRA neck: No stenosis
- MRI W/WO contrast: showed diffusion abnormality with associated restriction involving: splenium symmetrically
  - subcortical white matter of the centrum semiovale, right greater than the left side with no associated abnormal enhancement
  - no mass effect
  - At this point, the differential diagnosis was MBD versus diffuse encephalitis/encephalopathy with reversible splenial lesions versus osmotic demyelinating disease.

Imaging Contd.

- Fluid attenuated inversion recovery (FLAIR) showed three lesions:
- 1. Right centrum semiovale dominant lesion, 2. Left centrum semiovale small lesion and 3. Large lesion in splenium of corpus callosum.
- DTI Tractography: Showed significantly diminished commissural fibers extending to the right central semiovale lesion. Near absent or significantly diminished commissural fibers extending through the splenium of corpus callosum indicating demyelination.
- Fractional Anisotropy (FA) on DTI. Absence of transverse (red-color coded fibers in splenium of corpus callosum).

Imaging: Ax FLAIR

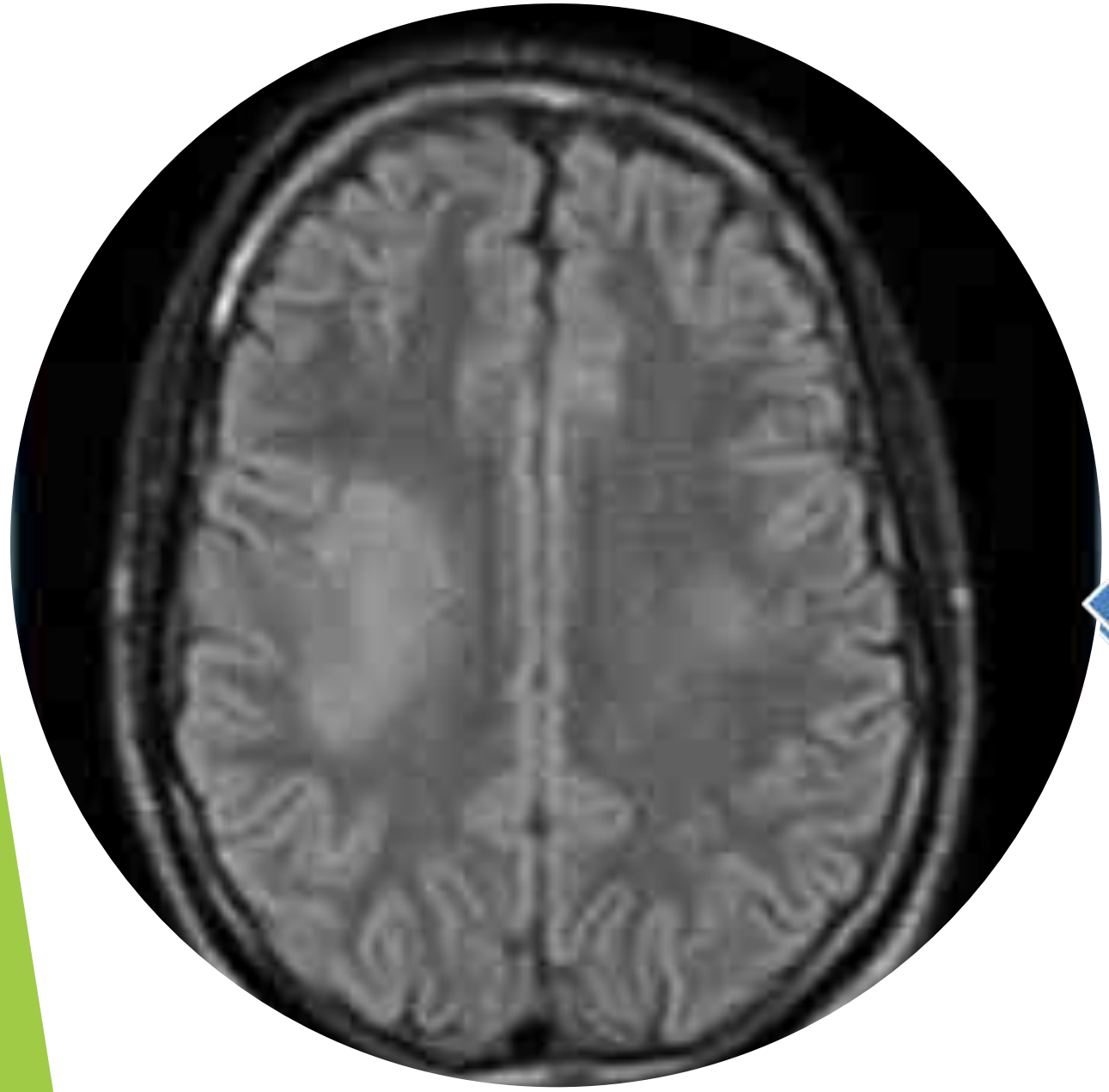


Figure 1. Ax. FLAIR: Hyperintense lesions in bilateral centrum semiovale, Rt > Lt

IMAGING

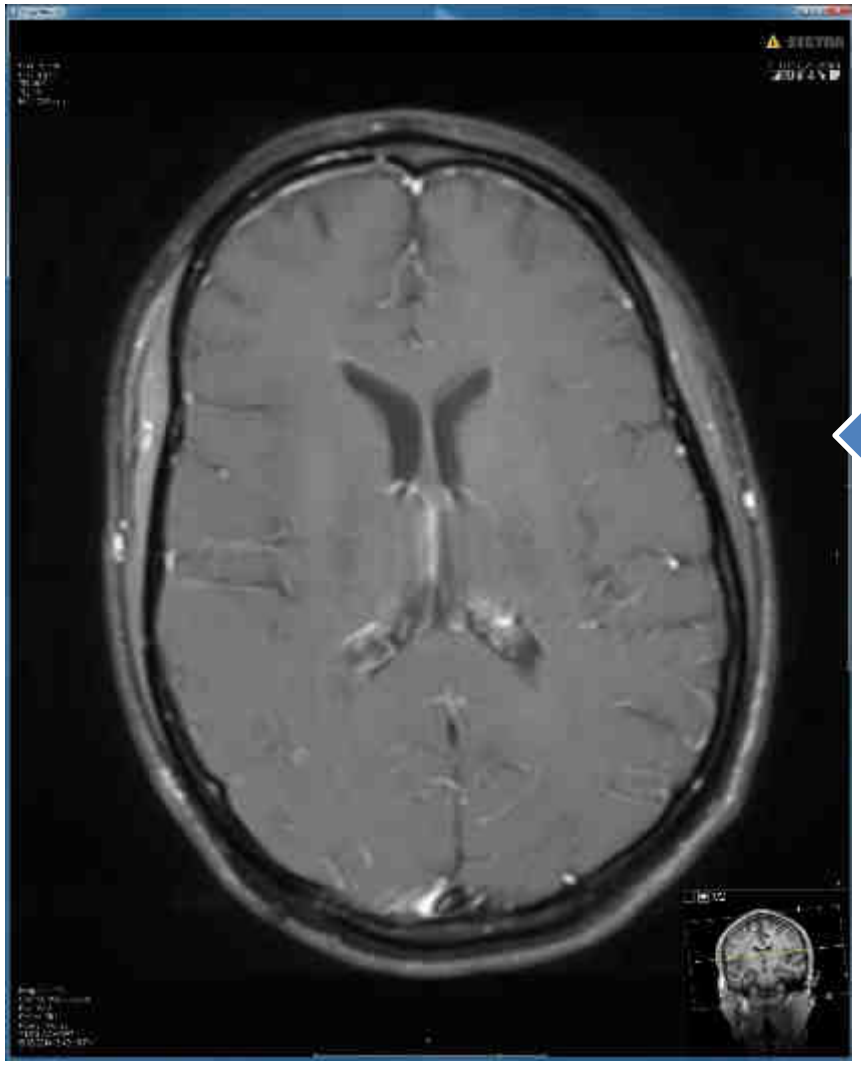


Figure 4. Ax T1 contrast enhanced: absence of abnormal enhancement with splenium of corpus callosum

Imaging: Ax DTI Fractional Anisotropy

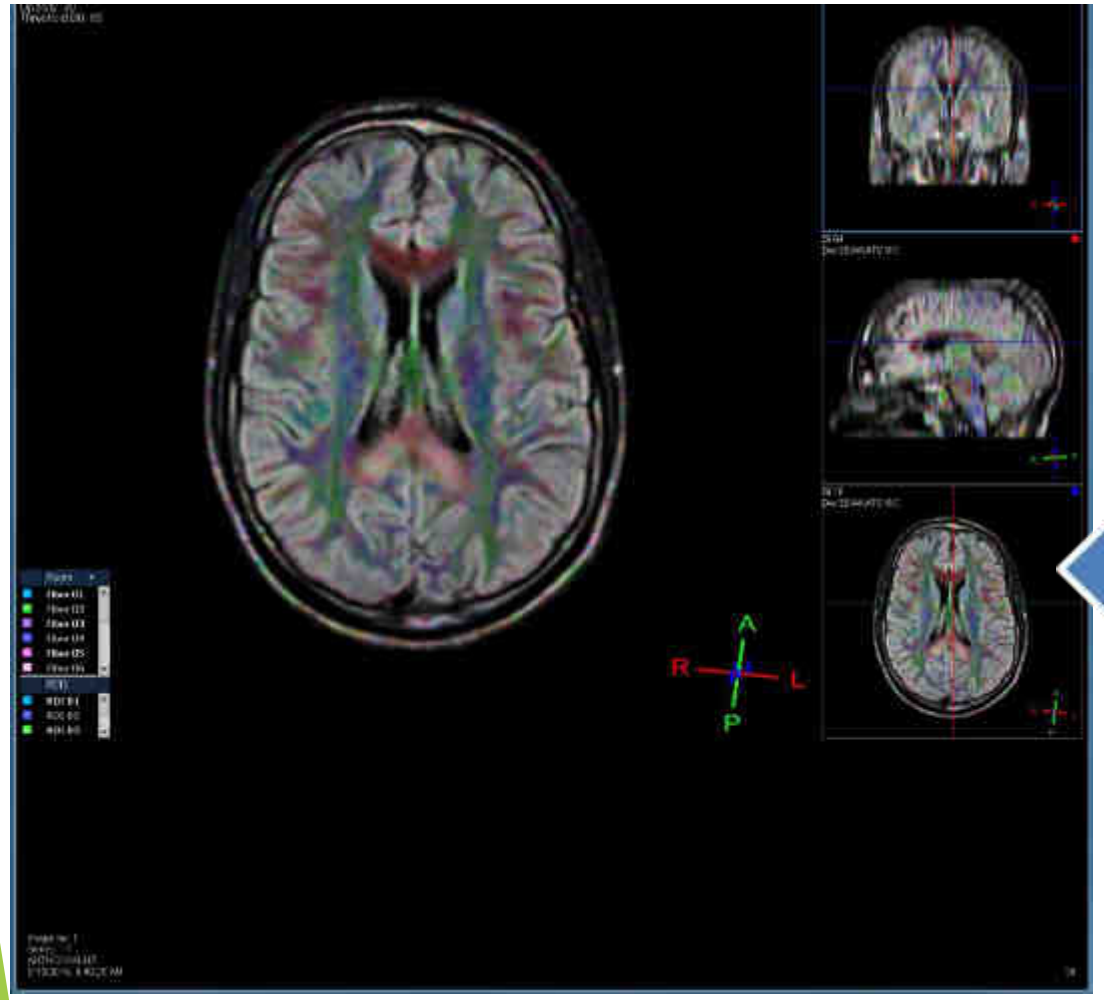


Figure 2. Ax Diffusion tensor imaging (DTI) fractional anisotropy (FA) map: absence / abnormality of red color coded horizontal fibers within splenium of corpus callosum.

Imaging: Ax FLAIR

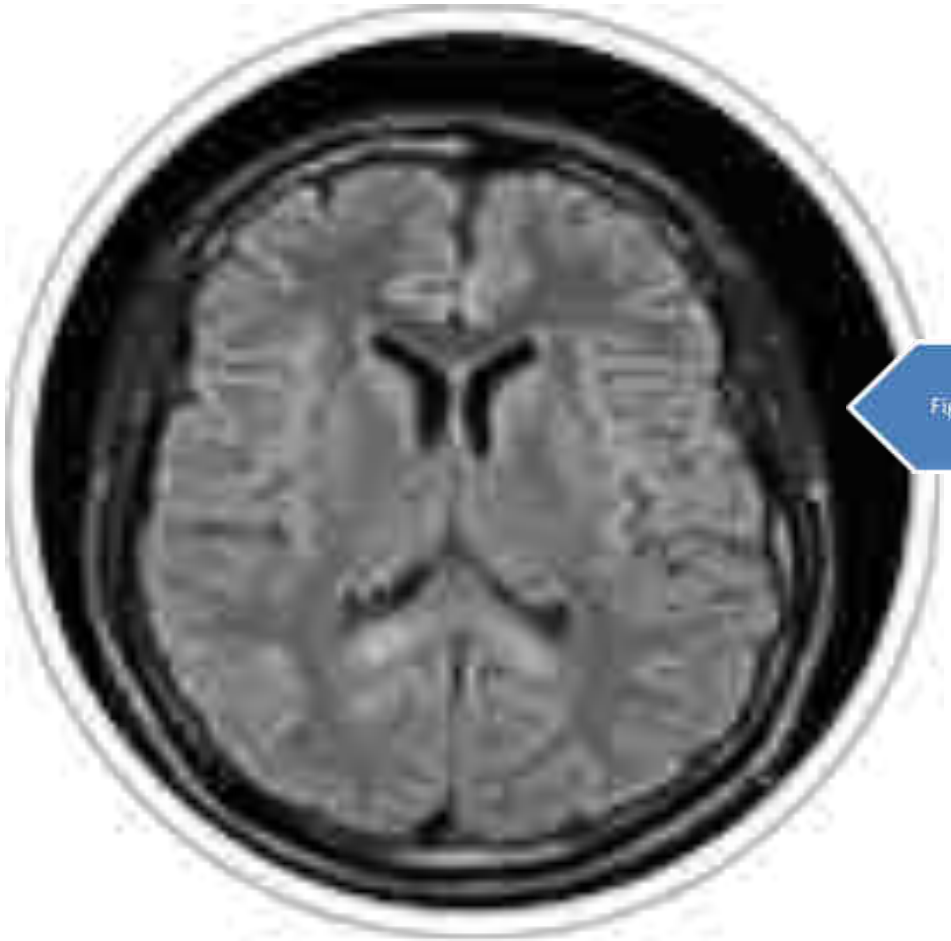
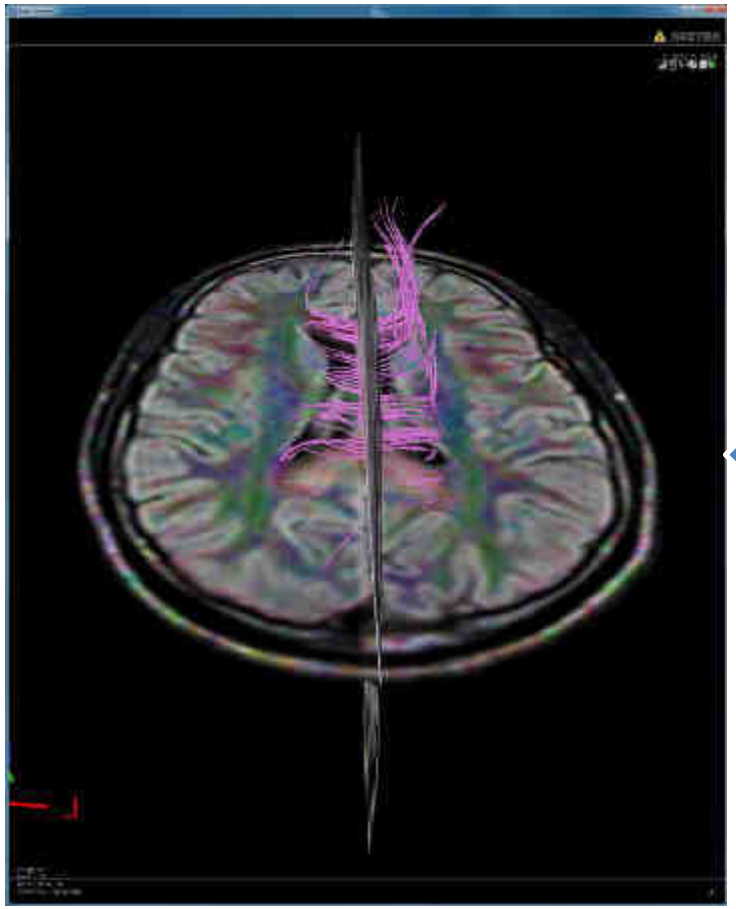


Figure 3. Ax Flair: Hyper intense lesion involving splenium of the corpus callosum.

Diffusion Tensor Image (DTI) Tractography



Figure[5]. DTI Tractography: Paucity of commissural fibers in the rt. centrum of semiovale and splenium of corpus callosum.

Imaging: DTI Tractography

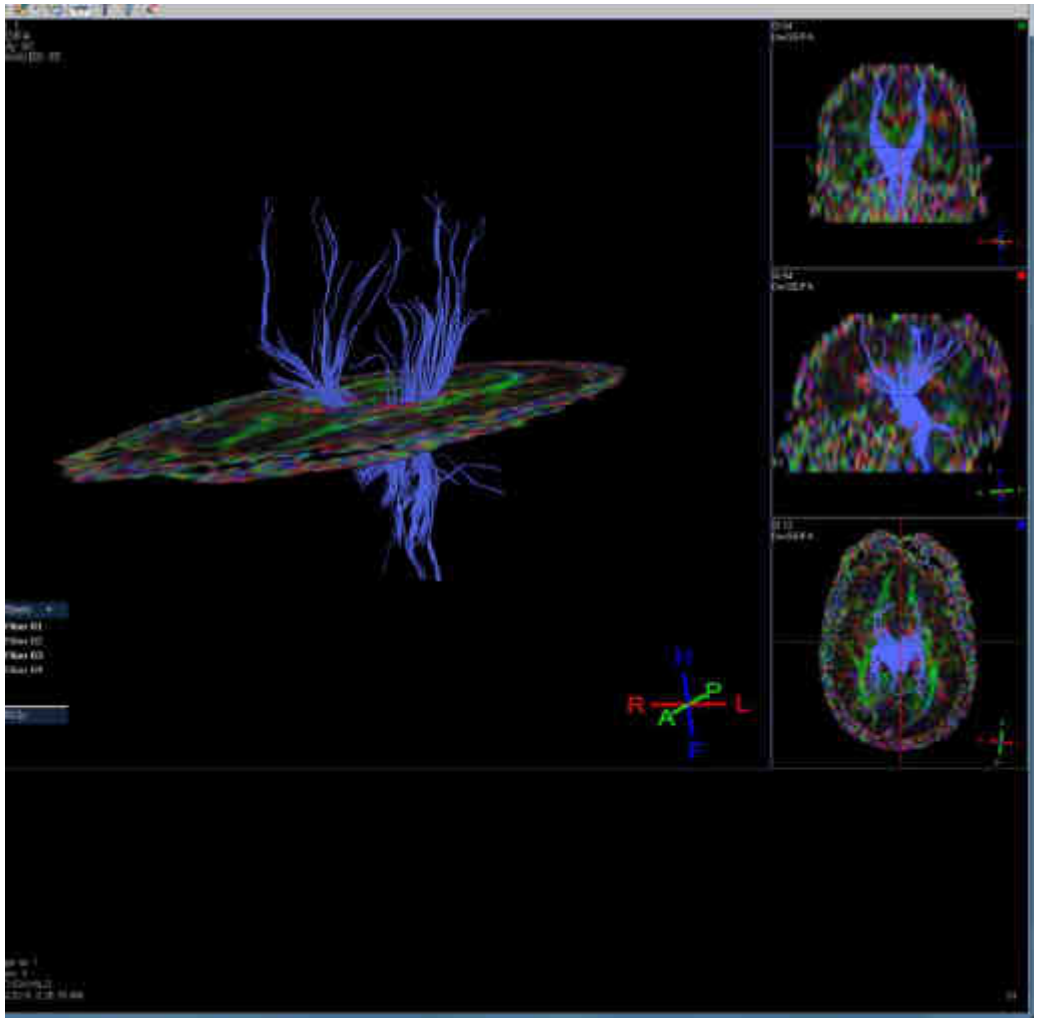
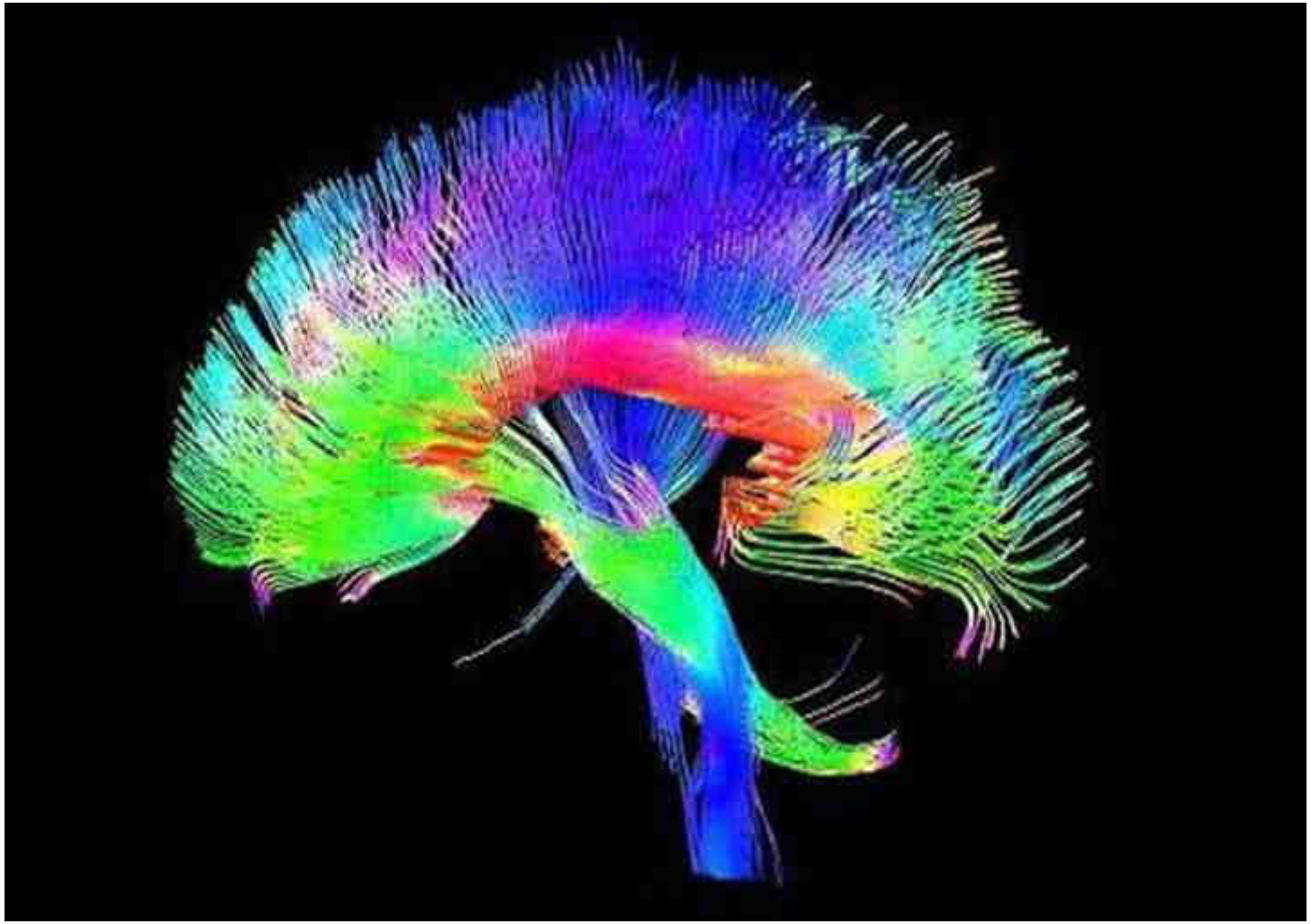


Figure 6. DTI Tractography: Paucity of fibers in the bilateral corticospinal tracts, Rt>Lt.

Normal DTI Image



Retrieved from https://www.stgeorges.nhs.uk/education-and-research/research/neurosurgery-research/current-

Management

- Multivitamin
- Thiamine
- Folic acid
- Initially was empirically started on Valproate for seizures
- He was empirically started on Ceftriaxone, Ampicillin, and Acyclovir, as meningitis was being ruled out.
- Followed by Neurology
- Followed by Psychiatry
- Followed by Dietary
- Followed by Rehab and occupational medicine
- By day ten he was cleared by all services and discharged to home.

Key Points

- Why Is this a Clinical Novelty?
  - MBD is a rare case and often an incidental diagnosis with high morbidity and mortality.
  - Earlier onset (age 30) as opposed to onset around age 45
  - Rapid recovery and minimal disability as he could work independently before discharge from hospital.
  - Added benefit of diffusion tensor image (DTI) Tractography in the confirmatory diagnosis of MBD
  - His presentation matches the type A which has high disability and mortality rate.
- Early diagnosis may have played a role in the prognosis of this patient as he responded to treatment and was commenced early on rehabilitation.
- Keep MBD in mind when dealing with patients with AUD who present with MBD symptoms.

References

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