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

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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, ophthalmoplegia, 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
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.
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 of abnormality of red color coded horizontal fibers within splenium of corpus callosum.
Imaging: Ax FLAIR

Figure 3. Ax Flair: Hyperintense 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

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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