MARCHIAFAVA BIGNAMI DISEASE
CASE DISCUSSION

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MARCHIAFAVA BIGNAMI DISEASE

- first described in Italian red wine drinkers by Carducci in 1898 and by Marchiafava and Bignami in 1903.

- It is more commonly seen in middle-aged or elderly alcoholic males.

- Rarely associated with various nutritional deficiencies

PATHOLOGY

- **The hallmark is demyelination and necrosis of corpus callosum with subsequent atrophy**
- **usually affects the body of corpus callosum ,followed by the genu and splenium.**
- **Entire corpus callosum may be involved.**

PATHOLOGY

- **The CC is usually affected in the middle portion (middle lamina), its highest myelinated component**
- **Sparing of the dorsal and ventral layers producing sandwich sign on MRI**
- **The corpus callosum degenerate and splits into three layers (layered necrosis).**

PATHOLOGY

- **Lesions may also be found in the hemispheric white matter and cerebellar peduncles.**

- **White matter tracts such as the anterior and posterior commissures and cortico spinal tracts may be involved.**

CLINICAL FEATURES

- **The disease follows three clinical courses**

1. Acute

2. Sub acute and

3. Chronic

ACUTE FORM

- **Mental confusion**
- **Disorientation**
- **Seizures.**
- **Muscle rigidity and facial trismus may be severe**

- **Most of the pts presenting with acute type of MBD will go into coma and eventually die.**

SUB ACUTE FORM

- **Dementia**
- **Dysarthria**
- **Muscle hypertonia.**

- **They may survive for years.**

CHRONIC FORM

- **It is characterised by chronic dementia**
- **Pts some time may present with hyponatremia.**

Table 1: Clinical forms of MBS and their differentials [1].

Clinical form of MBS	Predominant symptoms	Clinical differentials and their MRI findings
Acute MBS	Mental confusion, disorientation, neurocognitive deficits, and seizures	(1) **Wernicke encephalopathy** Symmetric involvement of medial thalami, mammillary bodies, tectal plate, and periaqueductal grey matter; hypointense on T1WI; hyperintense on T2W and FLAIR images. Few cases with cortical involvement restricted to motor and premotor areas.
Chronic MBS	Chronic dementia	(1) **Alzheimer disease** Extreme hippocampal and medial temporal lobe atrophy, severe global atrophy in end-stage disease. (2) **Multi-infarct dementia** (i) Diffuse white matter disease with large confluent lesions (hypointense on T1WI and hyperintense on T2W and FLAIR images) affecting at least 25% of white matter, mainly in the periventricular regions. (ii) Multiple lacunar infarcts in frontal white matter, thalami, and basal ganglia. (iii) Strategic infarcts: involving Parieto-temporo-occipital association areas, angular gyrus (middle cerebral artery territory), paramedian thalamic, and inferior medial temporal lobe (posterior cerebral artery territory). (3) **Frontotemporal lobar degeneration (Pick's) disease:** Pronounced atrophy of frontal and/or temporal lobes.

CT SCAN IN MBD

- Hypodensities in corpus callosum

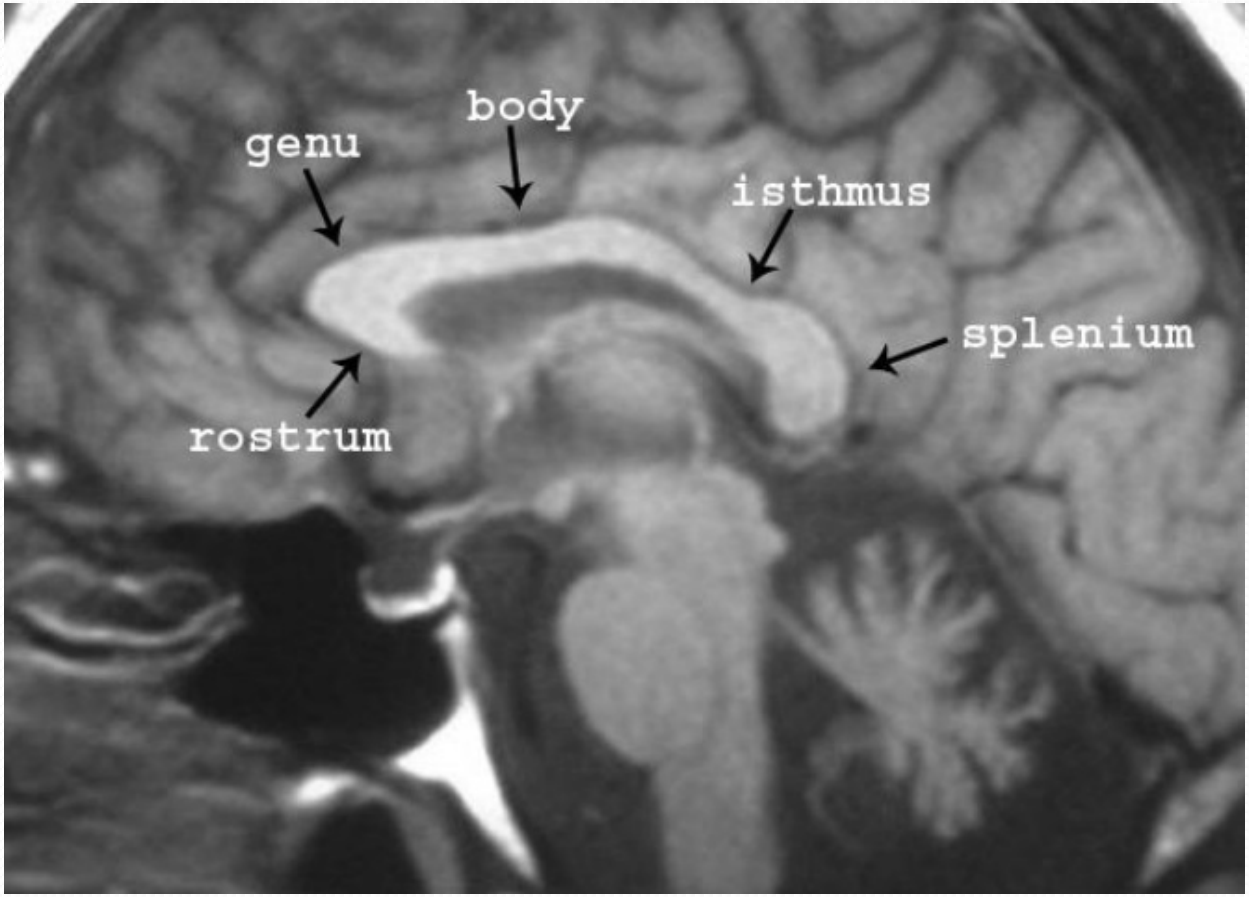
MRI FINDINGS

- Acute form : thickening and edema of corpus callosum

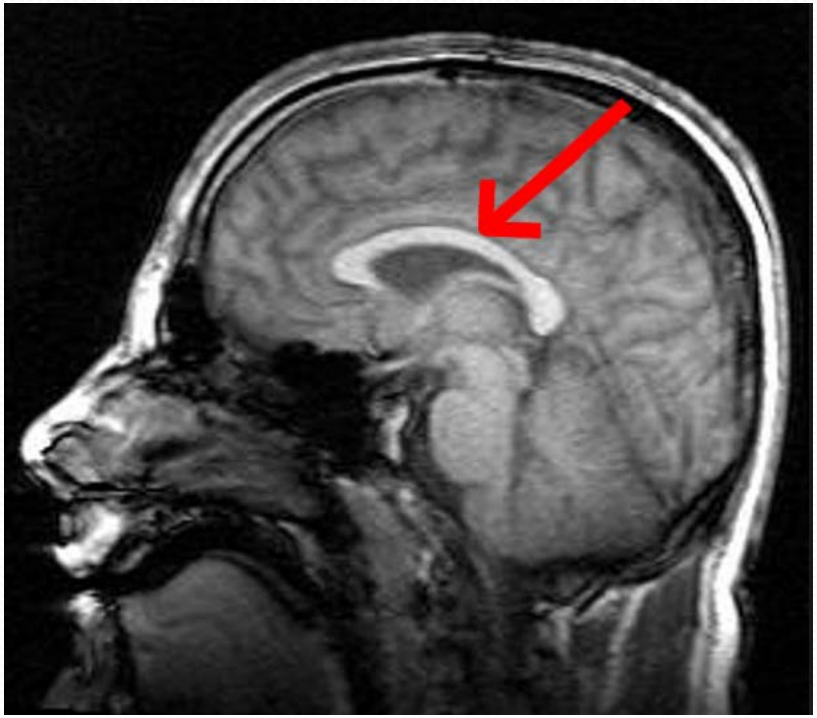
- Subacute and chronic forms :
  Hyperintensities
  Cyst formation
  Thinning and atrophy

- The central layers of the corpus callosum are affected, with sparing of the dorsal and ventral layers producing sandwich sign

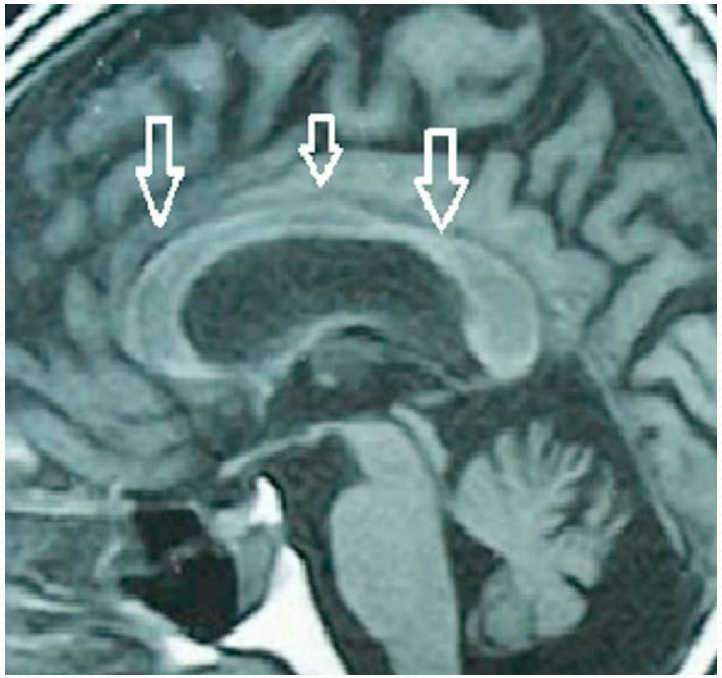
NORMAL CORPUS CALLOSUM



NORMAL CC



MBD



MRI t2 weighted sagittal image

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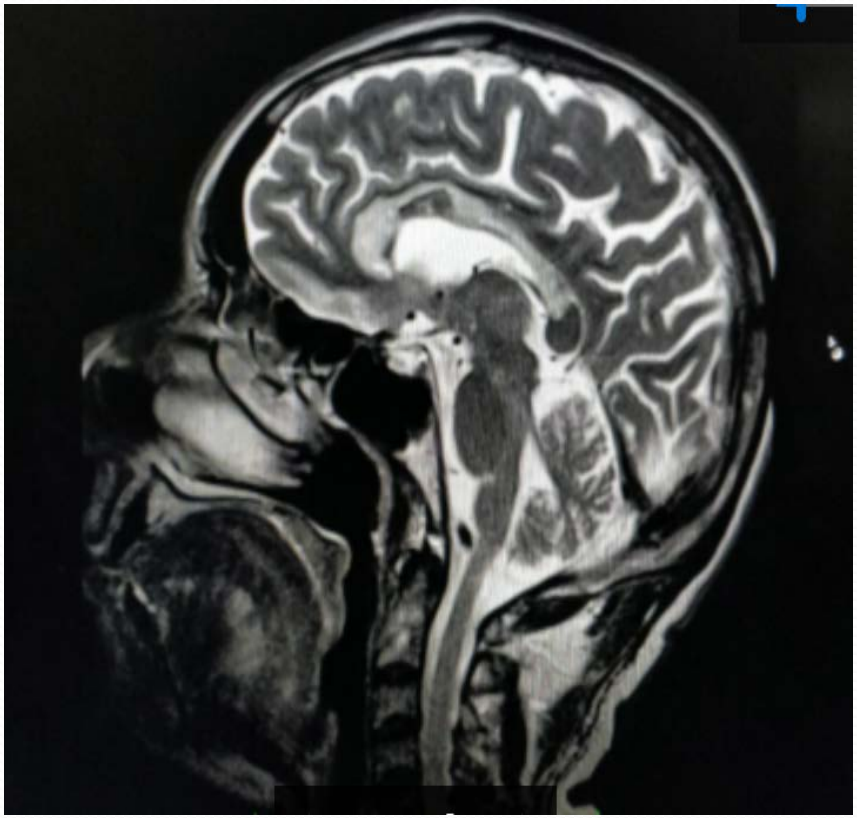
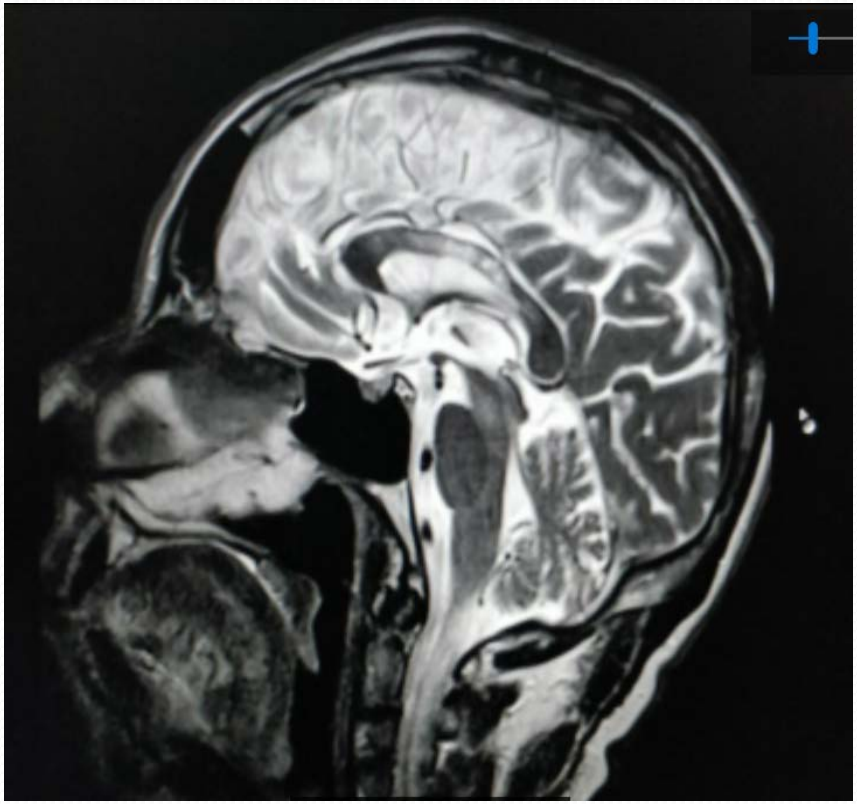
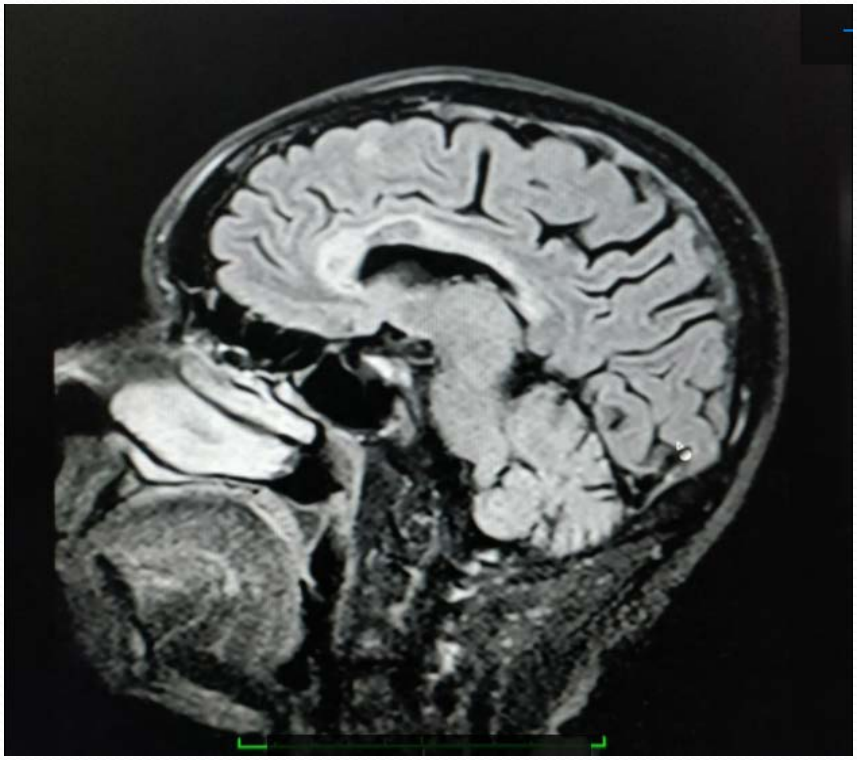




MRI FLAIR images

- Hyperintensities noted in the genu and body of corpus callosum



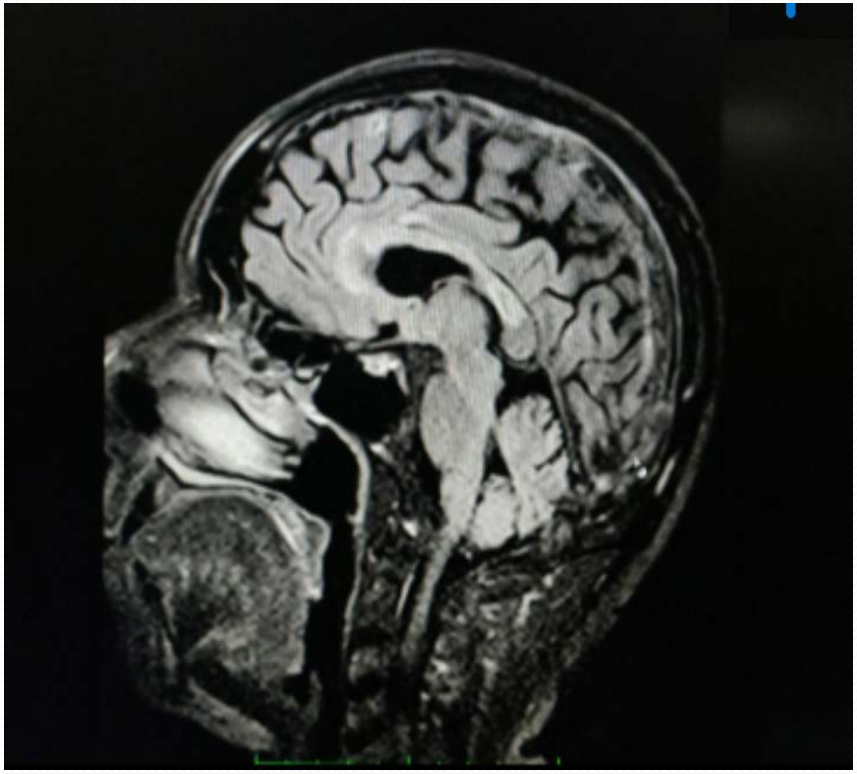


Table 2: Correlation of imaging findings in MBS with pathophysiology [1, 6].

	MRI findings	Underlying pathophysiology
1	Hyperintensity on T2 weighted images	Edema and myelin damage
2	Hypointensity on T1 weighted images	Total loss of myelin with replacement of the region by a cyst
3	Hyperintense rims and hypointense cores on FLAIR images	Damage to the myelin at the rim with a central necrotic area
4	Uniformly hyperintense lesions on FLAIR	Mixture of demyelination and edema
5	Areas of restricted diffusion on DWI (acute phase)	Cytotoxic edema

DIAGNOSIS

Diagnosis is made on the basis of clinical findings in combination with imaging features.

- **Increased T2 signal intensity involving the corpus callosum, in the correct clinical setting, the diagnosis of Marchiafava- Bignami disease can be made.**

MANAGEMENT

- Neurologist - For seizure control
- Critical care specialist - For coma management
- Neuropsychologist - For workup of the dementia
- Neurorehabilitation specialist
- Psychiatrist or psychologist - For treatment of alcoholism

TREATMENT

- The most common treatments are thiamine and other B vitamins
- No specific proven treatment is available

CONCLUSIONS

MARCHIAFAVA –BIGNAMI DISEASE is a rare demyelinating disorder predominatly affecting corpus callosum.

It is mostly seen in alcoholics.

The disease can occur in Acute , Sub acute and Chronic forms

MRI is a valuable tool in diagnosis of MARCHIAFAVA BIGNAMI DISEASE

THANK YOU

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