Metabolic Demyelinations

Last updated: September 5, 2017

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Demyelination of Corpus Callosum (Marchiafava-Bignami disease)

- primary degeneration of corpus callosum.

* first described by Marchiafava and Bignami in 1903.
* > 100 cases have been reported.
* frequent reports in Italian men (genetic predisposition?).

Etiology

- not known; possible causes / risk factors:

* 1. longstanding **alcoholism** (may have common pathogenesis with central pontine myelinolysis or Wernicke encephalopathy)
  2. nutritional deficiencies
  3. toxic factors

Pathophysiology

**Noninflammatory demyelination** → **necrosis** of middle lamina of corpus callosum (dorsal and ventral rims are spared!).

Constant bilateral symmetry!

* + - necrosis varies from softening & discoloration to cavitation & cyst formation.
    - rostral position of corpus callosum is affected first.
    - small symmetric lesions extend and become confluent.
* other CNS areas may be involved: *anterior commissure*, posterior commissure, centrum semiovale, subcortical white matter, long association bundles, middle cerebellar peduncles.
* spared structures: internal capsule, corona radiata, subgyral arcuate fibers, gray matter.
* **microscopy** - sharply defined necrotic process with myelin loss; relative preservation of axis cylinders in periphery of lesions;
* no inflammation!
* fat-filled phagocytes are common.
* gliosis is not well advanced.

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| Medial necrosis of corpus callosum and anterior commissure with sparing of margins: | D:\Viktoro\Neuroscience\Dem. Demyelinating disorders\00. Pictures\Marchiafava-Bignami disease (myelin stain).jpg |

Clinical Features

* onset - middle age or elderly.
* symptoms are insidious & nonspecific (only scarcely explained by callosal lesions) - ***multifocal & diffuse neurologic signs***:

1. transient focal neurological deficits (frontal release signs)
2. cognitive and behavioral (progressive dementia, depression and extreme apathy, confusion, manic, paranoid, or delusional states).
3. seizures
4. altered mental status (stupor → coma → death).

* slowly progressive → death within 3-6 years.

Diagnosis

**CT / MRI** - typical symmetric demyelinating callosal lesions.

Treatment

- no known therapy.

Central Pontine Myelinolysis

Pathophysiology

- acute symmetric ***noninflammatory demyelination*** in central basis pontis.

* demyelination and associated reduction in oligodendroglia; relative preservation of axons and surrounding neurons (lesions resemble Marchiafava-Bignami disease).
* in 10% cases, demyelination also occurs in extrapontine regions (midbrain, thalamus, basal nuclei, cerebellum; never below pontomedullary junction; rarely supratentorially).
* hypothesis - in regions of compact interdigitation of white and gray matter, **cellular edema** (caused by fluctuating osmotic forces) **compresses fiber tracts** → demyelination.
  + during prolonged hyponatremia, concentration of intracellular charged protein moieties is altered; reversal cannot parallel rapid correction of electrolyte status.

Etiology

Predisposing conditions:

1. alcoholism
2. liver disease, orthotopic liver transplantation surgery
3. malnutrition (esp. after burns)

Cause - ***too rapidly corrected*** severe and prolonged (< 120 mEq/L for > 48 hours) ***hyponatremia*** (osmotic myelinolysis).

Clinical Features

* 1. **Locked-in** (horizontal gaze paralysis + pseudobulbar palsy + spastic quadriplegia)
  2. **Preserved functions**: sensory modalities, vertical eye movements, blinking, breathing, alertness.
  + if demyelination extends through midbrain → **vertical ophthalmoparesis**.
  + if demyelination extends to pontine tegmentum and/or thalamus → **delirium, coma**.

Typical scenario:

* severe hyponatremia is diagnosed in person with delirium.
* IV fluid therapy is administered, and serum [Na+] is normal by next day.
* mental status improves, but is followed by neurologic deterioration 48-72 hours later.
* maximum recovery may require several months; full recovery has been reported.
* *death* is common within days or weeks.

Diagnosis

* **CSF** - increased opening pressure, protein↑, mononuclear pleocytosis.
* **EEG** - diffuse bihemispheric slowing.

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| * **T2-MRI** (imaging modality of choice) - ***hyperintense bright areas*** (water content↑) in central pons ***sparing peripheral rim***; later central lesion diminishes in size and signal, and mild pontine atrophy may result. D:\Viktoro\Neuroscience\Dem. Demyelinating disorders\00. Pictures\Central pontine myelinolysis (MRI2).jpg | D:\Viktoro\Neuroscience\Dem. Demyelinating disorders\00. Pictures\Central pontine myelinolysis (MRI).jpg |

Treatment

- supportive only.

* correct hyponatremia at 10 mmol/L/24 h + free water restriction.
* vitamin supplementation for alcoholic patients.

Bibliography for ch. “Demyelinating Disorders” → follow this [link >>](http://www.neurosurgeryresident.net/Dem.%20Demyelinating%20disorders\Dem.%20Bibliography.pdf)

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