

Progressive Multifocal Leukoencephalopathy (PML)



PLAY PICMONIC

Pathophysiology

Demyelination of CNS

Unraveling-myelin on CNS-brain

PML is a disease of the central nervous system that involves demyelination and is typically caused by the reactivation of the JC virus in immunosuppressed individuals, such as HIV patients.

Reactivation of JC Virus

JC-on-a-penny

PML is affiliated with the reactivation of those previously infected with the JC virus. The immunosuppressive state (e.g., HIV) of an individual triggers the reactivation of the virus, thereby allowing for the destruction of oligodendrocytes and, thus, the demyelination of the CNS.

Clinical Features

Neurologic Deficits

Defective-neuron

PML can cause a variety of symptoms depending on which brain area is demyelinated. It can involve myriad symptoms, including neurological deficits (e.g., hemiplegia, ataxia, aphasia), seizures, vision changes, encephalopathy, and altered mental status. These symptoms can be slow in onset and progress over several weeks.

Seizure

Caesar

PML can involve various symptoms revolving around neurological deficits, seizures, vision changes, encephalopathy, and altered mental status. These symptoms can be slow in onset and progress over several weeks.

Vision Changes

Delta Eyes

Demyelination caused by PML in the brain areas responsible for vision can cause a variety of visual symptoms. The most common visual problem in PML is hemianopsia, but it can cause any kind of vision problem.

Encephalopathy

Altered Brain

Encephalopathy in PML is caused by demyelination and inflammation in the brain structures. It is relatively less common as an isolated symptom of PML but more commonly occurs in combination with other symptoms of PML.

Diagnosis

Brain MRI or CT

[Brain M-R-eyes and Cat-scanner](#)

The preferred method of diagnosis involves an MRI with and without the presence of gadolinium contrast. MRI reveals bilateral asymmetric white matter lesions with no evidence of mass, which distinguishes PML from other brain pathologies.
An MRI is more sensitive to white matter lesions, though a CT with contrast can also be ordered.

CD4 Count

[CD \(4\) Fork-Helper T Cell](#)

A CD4 count should be obtained for individuals suspected of having PML to assess their immunological and HIV status. In HIV-positive patients, PML typically occurs when the CD4 count is <200 .

CSF Sample

[Brain-Spine-Fluid Sampler](#)

The diagnosis of PML can also involve a sampling of CSF, which can detect the JC virus via PCR. CSF, in combination with clinical and radiological findings, can also be sufficient for diagnosis.

Treatment

Supportive Care

[Supportive IV Bags](#)

PML treatment revolves around supportive care to address clinical symptoms. In those with HIV, ART should be started immediately. Unfortunately, despite treatment, PML has a high mortality rate with a median survival of less than one year.

ART Therapy

[Graffiti Art](#)

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