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# Progressive Multifocal Leukoencephalopathy: Opportunistic Infection in ART Era

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# PML: Still Relevant in the ART Era

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## Introduction

PML is a demyelinating disease caused by reactivation of the polyomavirus JC

Asymptomatically acquired in childhood with positive antibodies in 86% adults.

Lies dormant in the kidney and lymphoid tissue.

With profound IS, reactivation occurs with spread to CNS and demyelinating disease of oligodendrocytes.

Since the widespread use of combined ART, the incidence of PML in patients with HIV infection has decreased. In a population-based study from Denmark involving a nationwide cohort of patients aged 16 and older with HIV infection, the incidence of PML declined from 3.3 cases/1000 patient years at risk to 1.1 cases/1000 patient years at risk.<sup>1</sup>

## History of Present Illness

A 33 year old gentleman presented to Henry Ford Hospital in November 2018 with lower extremity weakness, fatigue, and progressive difficulty with activities of daily living over the last 2-3 weeks with multiple falls at home.

Associated with 20 lb. weight loss over the same period

Denied any other infectious symptoms of fever, chills, nausea, vomiting, diarrhea, rash, joint pain, URI symptoms, dysuria, genital lesions or other problems. He denied sick contacts or recent travel or occupational exposure.

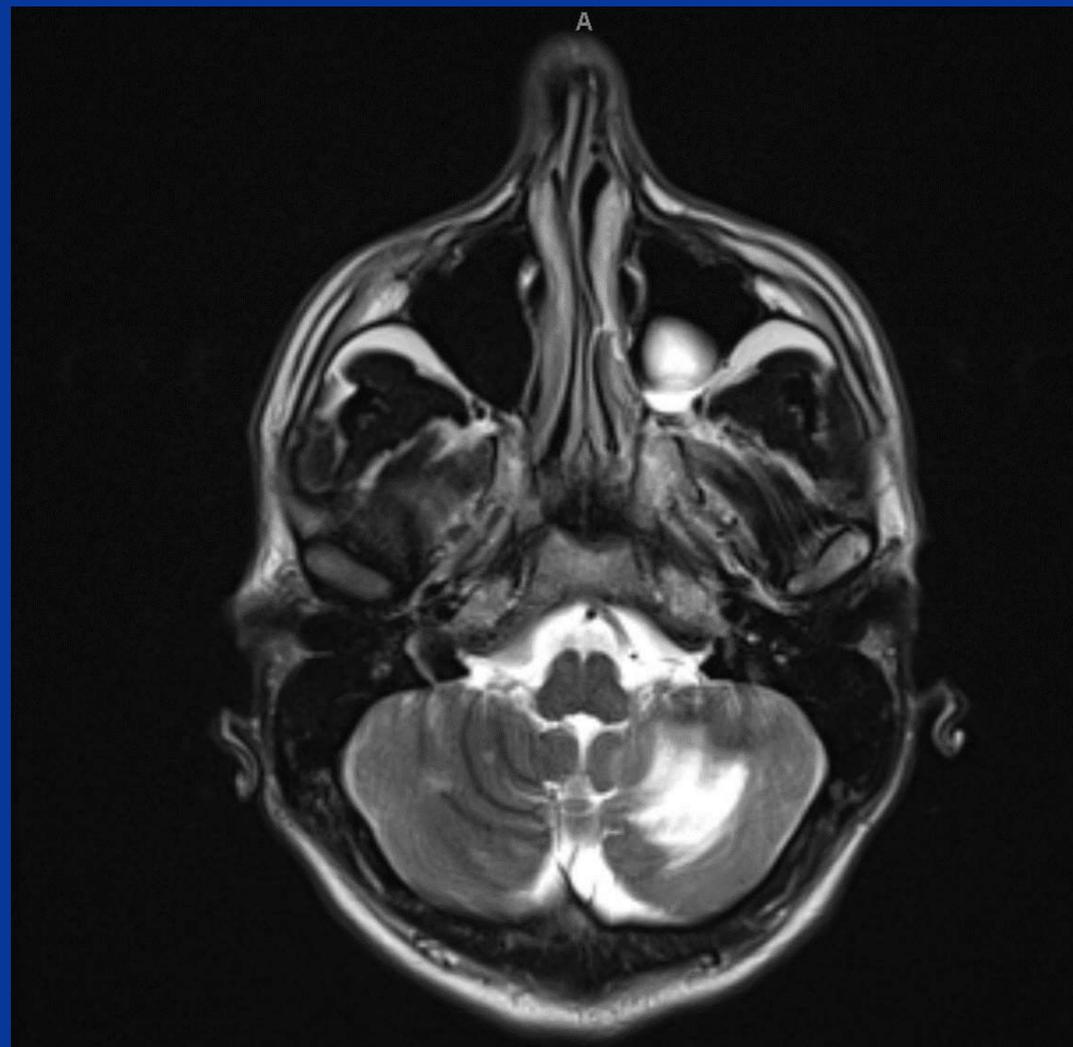
Admitted to sexual promiscuity with high risk female partners in years past

## Physical Examination

- BP 129/93, RR 18, Pulse 77, Temp 36.7
- BMI 17.9
- Diminished muscle bulk
- Dysdiadokokinesia and dysmetria on right hand
- Preserved mental status
- Slowed speech and movement.

Progressive Multifocal Leukoencephalopathy is a rare, often fatal demyelinating neurological disease seen in profoundly immunocompromised patients. It is caused by reactivation of JC Virus.

The diagnosis is made by CSF PCR for Polyomavirus JC. The test is 72-92% sensitive and 92-100% specific for the disease.<sup>2</sup> Treatment is aimed toward correcting the patient's underlying immunodeficiency.



MRI Brain: Demonstrates a large lesion in the left cerebellar hemisphere. The lesion demonstrates low signal intensity on T1, high signal intensity on T2/FLAIR, without diffusion restriction or enhancement. Companion lesions in the right parietal and left frontal lobes have predilection for the cerebral cortex white matter.



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## Lab Findings

- HIV positive 4<sup>th</sup> generation testing.
- CD4 count 35
- HIV Viral load 500,000
- CSF JC Virus PCR: Positive

## Types of PML

### Classical

- Variable neurological deficits patient to patient. Ataxia, hemianopia, weakness
- T cell count less than 200

### Inflammatory

- Paradoxical unmasking of PML due to IRIS
- Better prognosis than classical

### JC virus cerebellar granule cell neuronopathy

- Ataxia and cerebellar atrophy
- Mutations in the structure of the JC virus VP1 capsid

**JC virus meningitis:** More typical headache, photophobia, nuchal rigidity presentation.

**JC virus encephalopathy:** Rarely described, but rapidly fatal

## Treatment

- Correct the underlying immunodeficiency by starting ART in AIDS patients.
- Remove offending drugs for those patients exposed to notorious immunomodulator therapies like natalizumab in Multiple Sclerosis.
- Plasma exchange and steroids have been used to varied but unreliable success.
- Investigational therapies including cytarabine, cidofovir, topotecan, mirtazapine, mefloquine, maraviroc have been tried unsuccessfully.
- Outcome is often fatal and if not, leaves patients with significant neurological deficits. Our patient succumbed to his illness 1 month after presentation.

### References:

- 1) Weber et al, CID 1997
- 2) Cinque P, AIDS 1997
- 3) Marzocchetti A, J Clinical Microbio 2005