

INTRODUCTION

Headaches are one of the most common complaints that adults present with in the emergency room. It has a wide differential diagnosis that range from benign to life-threatening causes. Progressive multifocal leukoencephalopathy is a very rare disease of the CNS that typically manifests with neurological deficits and rarely headache. The following report demonstrates the importance of a thorough evaluation which, in this case, identified a very atypical cause of severe headache.

CASE REPORT

HPI:

- A 51-year-old African American woman with no significant past medical history presented to the emergency room with a chief complaint of a progressively worsening headache for 3 days
- Headaches began as right frontal head pain that was initially dull but intensified and on presentation was a 10 out of 10 in severity
- She endorsed associated photophobia and nausea
- A family member reported that the patient had a six-week history of confusion, odd behavior, and unsteady gait

Physical Exam:

- Mentation: Alert and oriented x3, follows commands, speaks in full sentences
- Speech: No dysarthria, no aphasia
- Cranial Nerves: PERRL, EOMI, face symmetric
- MOTOR: 5/5 in bilateral upper and lower extremities
- SENSORY: Intact sensation to light touch and temperature symmetric bilaterally
- CEREBELLAR: No ataxia finger-to-nose bilaterally

Initial Laboratory Testing:

- WBC 2,200
- Minor electrolyte abnormalities

HOSPITAL COURSE

Imaging:

- CT Head without contrast revealed right frontal vasogenic edema without midline shift and without intracranial hemorrhage
- MRI Brain with contrast showed an increased signal present within the right frontal lobe on the T2 and FLAIR sequences predominantly involving the subcortical white matter although involvement of the cortex in several locations was also noted along with areas of enhancement along the periphery of the lesions

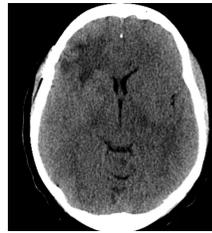


Figure 1: CT Head without contrast revealed right frontal vasogenic edema without midline shift and without intracranial hemorrhage

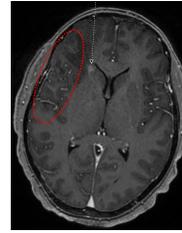


Figure 2: MRI Brain with contrast with increased signal present within the right frontal lobe and scattered areas of enhancement along the periphery of the area of concern some of which may represent draining vessels

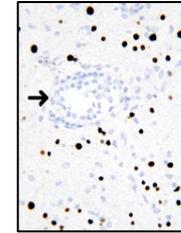


Figure 3: Presence of JC virus confirmed by BK SV40 large T antigen immunostaining

Procedures:

- Cerebral angiogram - diffuse vasculopathy of the right MCA superior division and right ACA
- Lumbar Puncture with CSF Fluid Analysis
- Craniotomy with brain biopsy

Pertinent Laboratory and Pathology Results:

- Hepatitis B Surface Antigen – **Reactive**
- HIV-1 Antibody – **Positive**
- HIV-1 Genotype - **Detected**
- HIV-1 Copies – **67,100**
- Total Helper Cells (CD4) – **40**
- JC Polyoma Virus - **Detected**
- CSF fluid presence of **oligoclonal bands** and **JC polyoma virus DNA**
- Brain tissue histopathology - cortical gray matter with **lymphocytic infiltrates**, **microglial nodules**, **perivascular chronic inflammation**, **reactive astrocytes with nuclear atypia** and **oligodendrocytes with enlarged glassy nuclei**

Discussion

Diagnosis:

With serologic testing positive for HIV-1, extensive viral cytopathic effect in glial cells with inflammatory infiltrates CD3-predominant, and the presence of JC virus confirmed by BK SV40 large T antigen immunostaining, the diagnosis of progressive multifocal leukoencephalopathy (PML) was made.

Conclusion:

PML is a severe demyelinating disease of the CNS. It is caused by reactivation of the JC polyomavirus, a virus that causes an asymptomatic primary infection acquired during childhood. In states of profound immunosuppression, JC virus can reactivate and replicate within glial cells. The virus can spread to the brain and infect oligodendrocytes causing demyelination. HIV infection, malignancy, autoimmune diseases, and immunosuppressive therapy for solid organ transplantation are the most common conditions causing reactivation of the JC virus.

Neurologic sequelae are dependent on lesion burden and location. Motor and sensory extremity deficits, ataxia, visual disturbances, aphasia, or personality changes are usually the initial chief complaint of PML. A literature search did not identify headache as a chief complaint, as did this patient case report. Although PML has no specific therapy, early identification and restoring the immune response is critical for prolonging survival.

References

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2. UpToDate.com. Koralnik, Igor (2021). Progressive multifocal leukoencephalopathy (PML): Epidemiology, clinical manifestations, and diagnosis.
3. <https://www.pathologyoutlines.com/topic/cnspml.html>