

Neurology[®]

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The Official Journal of the American Academy of Neurology



Neurology Publish Ahead of Print
DOI: 10.1212/WNL.0000000000207155

Teaching NeuroImage: Cerebellar Atrophy Due to JC Virus Granule Cell Neuronopathy: A Clinical Syndrome Distinct From Classic PML

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Alonso Quijada-Riquelme: Study concept or design; Analysis or interpretation of data
Heather Angus-Leppan: Study concept or design; Analysis or interpretation of data

Figure Count: 1

Table Count: 0

Search Terms:

[120] MRI, [142] Viral infections, [144] HIV, Cerebellar atrophy, JC virus

Acknowledgment:

Study Funding:

The authors report no targeted funding

Disclosures:

The authors report no relevant disclosures.

Preprint DOI:

Received Date:

2022-10-19

Accepted Date:

2023-01-20

Handling Editor Statement:

Submitted and externally peer reviewed. The handling editor was Resident and Fellow Section Editor Whitley Aamodt, MD, MPH.

A 34-year-old Hispanic man with HIV infection on no treatment (CD4 20/ μ l, viral load 1.9/10⁶ copies/ml) presented with four months of dizziness, ataxia and scanning speech consistent with a pancerebellar syndrome.

MRI scan (Figure) demonstrated marked cerebellar atrophy. CSF showed normal cell count, protein and glucose levels, non-reactive VDRL. CSF PCR was negative for cytomegalovirus, varicella-zoster, herpes simplex type 1 and 2, Epstein-Barr, and herpes virus 6, but positive for JC virus. JC virus granule cell neuronopathy (GCN) was diagnosed. JC virus variants may rarely infect cerebellar granule neurons instead of oligodendrocytes as seen in classic PML with white matter involvement.^{1,2}

HAART (zidovudine, lamivudine, efavirenz) commenced immediately, with slight symptomatic improvement at 12 months, MRI scan was unchanged.

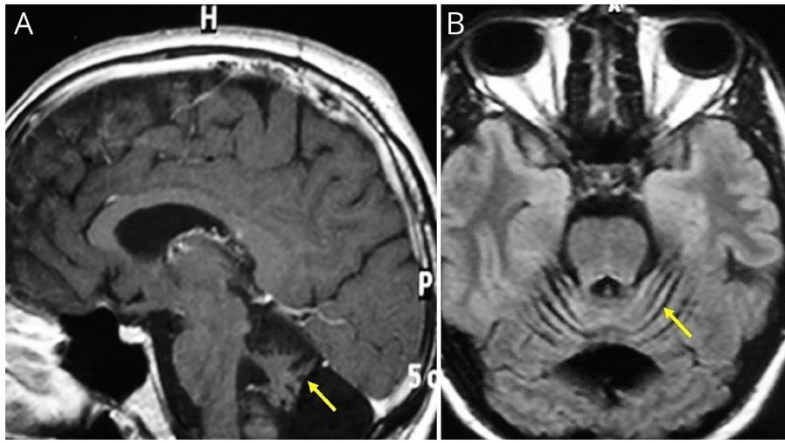
Clinicians should suspect JC virus strain infection producing GCN in AIDS patients with symptomatic cerebellar atrophy² and commence HAART promptly- IRIS is not usually a concern in cases of isolated JCV GCN.

<http://links.lww.com/WNL/C655>

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Figure (A) Sagittal T1-weighted brain MRI and (B) Axial FLAIR-weighted brain MRI show marked cerebellar atrophy (arrows)



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Neurology published online February 20, 2023

DOI 10.1212/WNL.0000000000207155

This information is current as of February 20, 2023

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