

Teaching NeuroImage: Cerebellar Atrophy Due to JC Virus Granule Cell Neuronopathy

A Clinical Syndrome Distinct From Classic PML

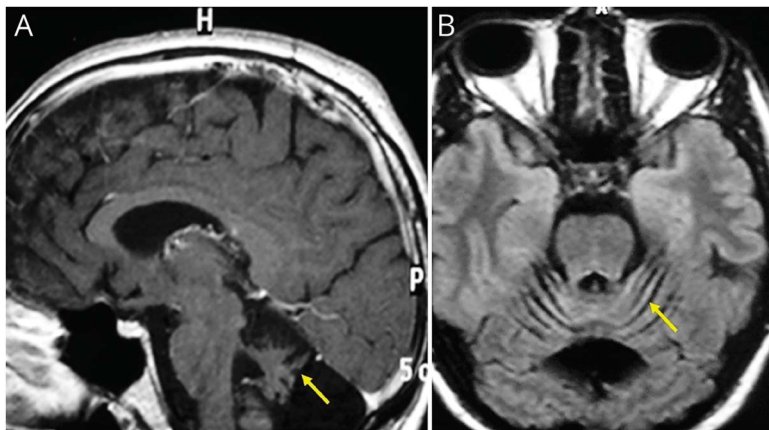
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Figure (A) Sagittal T1-Weighted Brain MRI and (B) Axial Fluid-Attenuated Inversion Recovery-Weighted Brain MRI Show Marked Cerebellar Atrophy (Arrows)



A 34-year-old Hispanic man with HIV infection on no treatment (CD4 20/ μ L, viral load 1.9/ 10^6 copies/mL) presented with 4 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, nonreactive venereal disease research laboratory test. CSF PCR was negative for cytomegalovirus, varicella-zoster, herpes simplex type 1 and 2, Epstein-Barr, and herpes virus 6, but positive for John Cunningham (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 progressive multifocal leukoencephalopathy with white matter involvement.^{1,2} HAART (zidovudine, lamivudine, and 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—immune reconstitution inflammatory syndrome is not usually a concern in cases of isolated JC virus GCN.

Author Contributions

C. Silva-Rosas: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; analysis or interpretation of data. G. Abudinén: study concept or design; analysis or interpretation of data. A. Quijada-Riquelme: study concept or design; analysis or interpretation of data. H. Angus-Leppan: study concept or design; analysis or interpretation of data.

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Appendix (continued)

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