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Teaching NeuroImages: Recurrent SSPE presenting as Anton syndrome with cortical ribboning

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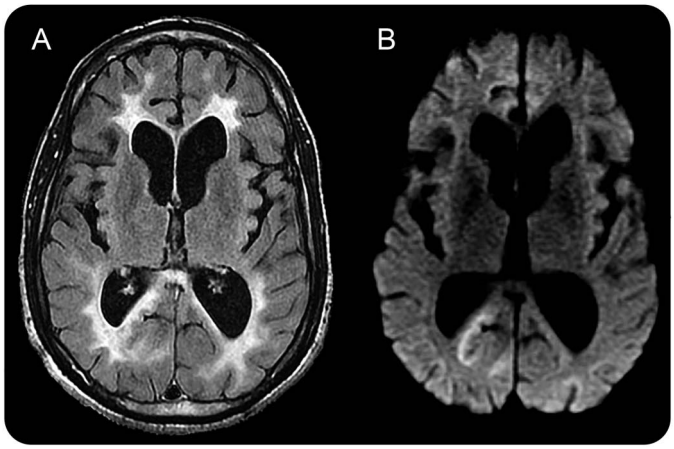
Figure Axial MRI at the level of the thalami



(A) Fluid-attenuated inversion recovery sequences demonstrate stable atrophy and periventricular white matter changes. (B) Increased diffusion-weighted signal is observed within the cortical ribbons of both occipital lobes (right > left), concurrent with decreased acute diffusion coefficient signal (not shown).

A 21-year-old man, an immigrant from India who had not been vaccinated against measles in childhood, presented with new visual loss, on the background of a 4-year history of cognitive impairment due to subacute sclerosing panencephalitis (SSPE). SSPE was previously managed with 3 times weekly intrathecal α-interferon, which was stopped 18 months previously given stable disease. Examination confirmed baseline cognitive impairment. Vision was reduced to light perception, although the patient denied visual loss. Brain MRI showed occipital cortex diffusion restriction (figure). CSF measles titers were elevated. The patient was diagnosed with Anton syndrome due to SSPE. Off-label intrathecal α-interferon was restarted, recognizing prior effect in this patient and reports of improvement or stabilization in treated patients.[1] Restricted diffusion of the cortical ribbon may be seen in SSPE in addition to Creutzfeldt-Jakob disease,[2] reflecting cortical dysfunction and neuronal loss.[3]

AUTHOR CONTRIBUTIONS

G.S. Day participated in the conception and design of the study; participated in the acquisition, analysis, and interpretation of data; and helped to draft, revise, and finalize the manuscript. P. Tai participated in the acquisition, analysis, and interpretation of data and helped to revise and finalize the manuscript. M. Moharir participated in the acquisition, analysis, and interpretation of data and helped to revise and finalize the manuscript. D.F. Tang-Wai participated in the acquisition, analysis, and interpretation of data and helped to revise and finalize the manuscript. D.F. Tang-Wai had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of neuroradiologic interpretation.

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DISCLOSURE

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