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

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. Restricted diffusion of the cortical ribbon may be seen in SSPE in addition to Creutzfeldt-Jakob disease, reflecting cortical dysfunction and neuronal loss.

(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).

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DISCLOSURE
G. Day is the Clinical Director of the Anti-NMDA Receptor Encephalitis Foundation (Canada) and is involved in the development and implementation of projects to support the Foundation’s goals of improving outcomes in patients with autoimmune-mediated encephalopathy through education and research. The Foundation is supported by private donations. G.S. Day is the recipient of a Future Leaders in Dementia award, including support for travel (Pfizer Canada). P. Tai and M. Moharir report no disclosures relevant to the manuscript. D.F. Tang-Wai holds a grant with the Weston Foundation, and is a collaborator on grants from the Canadian Institutes of Health Research, Alzheimer Society of Canada, Parkinson Society of Canada, and the Michael J. Fox Foundation. Off-label use of medications: interferon-α is not approved by the US Food and Drug Association for the treatment of chronic subacute sclerosing panencephalitis. Go to Neurology.org for full disclosures.

REFERENCES

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