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

Gregory S. Day, MD,
MSc, FRCPC
Peter Tai, MD, FRCPC
Mahendranath Moharir,
MD, FRACP
David F. Tang-Wai, MD,
FRCPC

Correspondence to
Dr. Day:
dayg@abraxas.wustl.edu

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.

STUDY FUNDING

No targeted funding reported.

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

1. Anlar B, Yalaz K, Oktem F, Kose G. Long-term follow-up of patients with subacute sclerosing panencephalitis treated with intraventricular alpha-interferon. *Neurology* 1997;48: 526–528.

Download teaching slides:
Neurology.org

From the University of Toronto (G.S.D., P.T., M.M., D.F.T.-W.); Toronto Western Hospital (G.S.D., P.T., D.F.T.-W.); and Hospital for Sick Children (M.M.), Toronto, Canada.

2. Manners DN, Parchi P, Tonon C, et al. Pathologic correlates of diffusion MRI changes in Creutzfeldt-Jakob disease. *Neurology* 2009;72:1425–1431.
3. Oksuzler YF, Cakmakci H, Kurul S, Oksuzler M, Dirik E. Diagnostic value of diffusion-weighted magnetic resonance imaging in pediatric cerebral diseases. *Pediatr Neurol* 2005;32:325–333.

Neurology®

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

Gregory S. Day, Peter Tai, Mahendranath Moharir, et al.

Neurology 2015;85:e141-e142

DOI 10.1212/WNL.0000000000002096

This information is current as of November 9, 2015

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/85/19/e141.full
Supplementary Material	Supplementary material can be found at: http://n.neurology.org/content/suppl/2015/11/07/WNL.0000000000002096.DC1
References	This article cites 3 articles, 2 of which you can access for free at: http://n.neurology.org/content/85/19/e141.full#ref-list-1
Citations	This article has been cited by 1 HighWire-hosted articles: http://n.neurology.org/content/85/19/e141.full##otherarticles
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Agnosia http://n.neurology.org/cgi/collection/agnosia All Infections http://n.neurology.org/cgi/collection/all_infections MRI http://n.neurology.org/cgi/collection/mri Viral infections http://n.neurology.org/cgi/collection/viral_infections
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2015 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.