Holmes Tremor With Periolandic Demyelinating Lesions

A Distinct Clinicoradiographic Syndrome

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Figure Brain MRI in 2 Patients With Holmes Tremor and Periolandic Demyelinating Lesions

Both patients had persistent unilateral periolandic T2-fluid-attenuated inversion recovery (FLAIR) hyperintense (A.a, B.a), T1-hypointense (A.b, B.b) lesions with transient gadolinium enhancement in patient B (B.b, arrows). Patient A also had a left posterolateral mesencephalic T2-FLAIR hyperintense lesion (A.c, arrow). Patient B developed linear right cerebral peduncle T2-FLAIR hyperintensity that was not observed on initial MRI 3 months after tremor onset, suggesting Wallerian degeneration (B.c, arrow).

Patients A (38-year-old woman) and B (30-year-old man) developed unilateral, slowly progressive 4–6 Hz tremor over months to years with rest, postural, and intentional components (videos 1 and 2), indicating Holmes tremor.¹ Both had contralateral periolandic lesions (figure). Patient A had a midbrain lesion (figure) and CSF-specific oligoclonal bands; patient B had enhancement, MRI suggesting Wallerian degeneration (figure), and transient low-titer MOG immunoglobulin G positivity of doubtful clinical

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significance.¹ Both lacked spine lesions. Neither responded to symptomatic/immunosuppressive treatments. We hypothesize the perirolandic lesions caused Holmes tremor by disrupting nigrostriatal/cerebello-thalamo-cortical projections, although this requires confirmation.² We suspect progressive multiple sclerosis (MS) as the etiology, given the striking similarity to prior reports of this presentation in MS.³

Study Funding

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Disclosure

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Name	Location	Contribution
Adrian Budhram, MD	Mayo Clinic, Rochester, MN	Designed and conceptualized study, drafted manuscript, analyzed and interpreted the data, composed the figure, recorded the videos
Hugh D. Simpson, MBBCh, PhD	Mayo Clinic, Rochester, MN	Interpreted the data, revised the manuscript for intellectual content
James H. Bower, MD	Mayo Clinic, Rochester, MN	Interpreted the data, revised the manuscript for intellectual content
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Eoin P. Flanagan, MBBCh	Mayo Clinic, Rochester, MN	Designed and conceptualized study, analyzed and interpreted the data, revised the manuscript for intellectual content, study supervision

References

1. Jarius S, Paul F, Aktas O, et al. MOG encephalomyelitis: international recommendations on diagnosis and antibody testing. *J Neuroinflamm* 2018;15:134.
2. Raina GB, Cersosimo MG, Folgar SS, et al. Holmes tremor: clinical description, lesion localization, and treatment in a series of 29 cases. *Neurology* 2016;86:931–938.
3. Karmon Y, Morrow SA, Weinstock A, Hojnacki D, Weinstock-Guttman B. Limb ataxia originating from peri-central sulcus demyelinating lesion in multiple sclerosis. *J Neurol Sci* 2012;320:136–140.

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