Hypertrophic olivary degeneration mimics relapse in neuromyelitis optica spectrum disorder

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Figure 1 Brain MRI at onset (top row) and after 9 months (bottom row)

Axial images show T2 hyperintensity of left dentate (A), with enhancement postgadolinium (B), and area postrema T2 hyperintensity (C). Repeat MRI shows right inferior olivary nucleus T2 hyperintensity/hypertrophy on axial (D) and sagittal (E) images.

A 51-year-old woman developed subacute ataxia and intractable vomiting. MRI showed left dentate and area postrema lesions (figure 1, A–C). Serum aquaporin-4–immunoglobulin G positivity confirmed neuromyelitis optica spectrum disorder. Treatment was with steroids and plasmapheresis acutely and maintenance 6-monthly rituximab. MRI 9 months later revealed a new asymptomatic right medullary lesion (figure 1, D and E), prompting mycophenolate add-on therapy. The hypertrophy, expected location, and time course led us to diagnose hypertrophic olivary degeneration from dentato-rubro-olivary circuit (Mollaret triangle [figure 2]) interruption; no palatal tremor occurred.^{1,2} Mycophenolate was discontinued. Failure to recognize a medullary lesion as hypertrophic olivary degeneration from a prior insult along the Mollaret triangle may lead to unnecessary treatment.

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Disclosure

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Figure 2 Mollaret triangle

Interruption of dentate fibers to red nucleus after an insult results in contralateral inferior olivary nucleus deafferentation and hypertrophy. IgG = immunoglobulin G.

Appendix Authors

Name	Location	Role	Contribution
Elia Sechi	Mayo Clinic, Rochester, MN	Author	Drafted manuscript, analyzed and interpreted the data, composed the figure
Natalie E. Parks	Dalhousie University, Halifax, Canada	Author	Interpreted the data, revised the manuscript for intellectual content
Kelly K. Koeller	Mayo Clinic, Rochester, MN	Author	Interpreted the data, revised the manuscript for intellectual content
Eoin P. Flanagan	Mayo Clinic, Rochester, MN	Author	Designed and conceptualized study, analyzed and interpreted the data, revised the manuscript for intellectual content, study supervision

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