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Episodic ataxia in CASPR2 autoimmunity

A. Sebastian López Chiriboga, MD, and Sean Pittock, MD

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Correspondence
Dr. López
lopez.alfonso@mayo.edu

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Video

A 64-year-old man developed cognitive dysfunction and generalized seizures, followed by episodes of transient speech and gait disturbances lasting several minutes, occurring multiple times per day; the events were not associated with any triggers. Neurologic examination during one of his episodic symptoms (video 1) showed transient ataxic dysarthria. Testing for neural autoantibodies revealed evidence of voltage-gated potassium channel complex antibodies by radioimmunoprecipitation assay; CASPR2-IgG was positive in serum and CSF by cell-based assay. Brain MRI was normal; CSF analysis showed an elevated protein level at 101 mg/dL, but it was otherwise normal. Oncologic evaluation was negative. The cognitive symptoms improved; seizures and episodic ataxia resolved after 12 weeks of treatment with IV methylprednisolone. Episodic ataxia is a manifestation of CASPR2 autoimmunity;[1] patients can have normal MRI, and immunotherapy is beneficial.

Author contributions

A.S. Lopez-Chiriboga and S. Pittock contributed to the conception and design of the study; collection, analysis, and interpretation of the data; drafting and critical revision of the manuscript; and generation/collection of the figures. Both authors gave final approval of the manuscript.

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Disclosure

A.S. Lopez-Chiriboga reports no disclosures. S. Pittock has a patent pending for GFAP, Septin-5 and MAP1B autoantibodies as biomarkers of neurological autoimmumity; received research support from Grifols, MedImmune, Alexion, AEA, and NIH; had compensation for consulting activities paid to Mayo Clinic from Alexion and MedImmune; and has a financial interest with Mayo Clinic in patents that relate to functional AQP4/NMO-IgG assays and NMO-IgG as a cancer marker. Full disclosure form information provided by the authors is available with the full text of this article at Neurology.org/NN.

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Reference

1. Joubert B, Gobert F, Thomas L, et al. Autoimmune episodic ataxia in patients with anti-CASPR2 antibody-associated encephalitis. Neurol Neuroimmunol Neuroinflamm 2017;4:e371. doi: 10.1212/NXI.0000000000000371.

From the Department of Neurology (A.S.L.C.) and Laboratory Medicine and Pathology (S.P.), Mayo Clinic, Rochester, MN.

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