Faciobrachial dystonic seizures in an Lgi1 VGKC-complex antibody-mediated encephalitis

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Video

Neurology.org/cp

A 71-year-old woman presented with a 6-week history of progressive involuntary right face and arm jerking movements associated with prominent memory loss and frequent falls. She subsequently developed similar movements involving her left face and arm. Montreal Cognitive Assessment (MoCA) score was 19/30 and physical examination was otherwise normal. There was no alteration of consciousness. A video EEG recorded bilateral clinical activity with no alteration in consciousness and no electrographic epileptiform correlate (see video at Neurology.org/cp). MRI demonstrated nonenhancing diffuse T2 hyperintensity of the hippocampal region of the left medial temporal lobe (figure). Serum and CSF were positive for voltage-gated potassium channel (VGKC) antibodies (6,534 pmol/L and 160 pmol/L, respectively). Anti-Lgi1 antibodies were strongly positive, and anti-Caspr2 antibodies were negative. Onconeural antibodies and paraneoplastic workup were negative; serum sodium nadir was 130 mmol/L, coinciding with cognitive nadir. She received pulsed methylprednisolone and IV immunoglobulin and responded rapidly, with involuntary movements abolished within 2 weeks. Memory improved over 2 months and function returned to baseline, although amnesia for the period of illness and retrograde amnesia extending 2 months prior persists. Her MoCA score plateaued at 27/30. IV immunoglobulin was discontinued after 2 months, and the steroid taper continues gradually. Follow-up MRI at 4 months demonstrated resolution of the left hippocampal T2 hyperintensity.

Although classification has been debated, faciobrachial dystonic “seizures” are characteristic of the usually nonparaneoplastic Lgi1 VGKC-complex antibody-mediated limbic encephalitis and often precede associated cognitive impairment.^{1,2} Early recognition allows for accurate antibody diagnosis and timely immunotherapy initiation for these frequently anticonvulsant-refractory seizures, thereby potentially preventing cognitive impairment and cerebral atrophy.^{1,2}

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Figure T2 MRI demonstrating medial temporal lobe hyperintensity

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AUTHOR CONTRIBUTIONS

Eloise Watson: drafting/revising the manuscript, analysis or interpretation of data, acquisition of data; Ian Rosemergy: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, study supervision; Jennifer Taylor: study concept or design, analysis or interpretation of data; David Abernethy: drafting/revising the manuscript, editing video; Jeremy Lanford: study concept or design, analysis or interpretation of data.

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