VIDEO NEUROIMAGES

Clinical and imaging features of newly recognized Kelch-like protein 11 paraneoplastic syndrome

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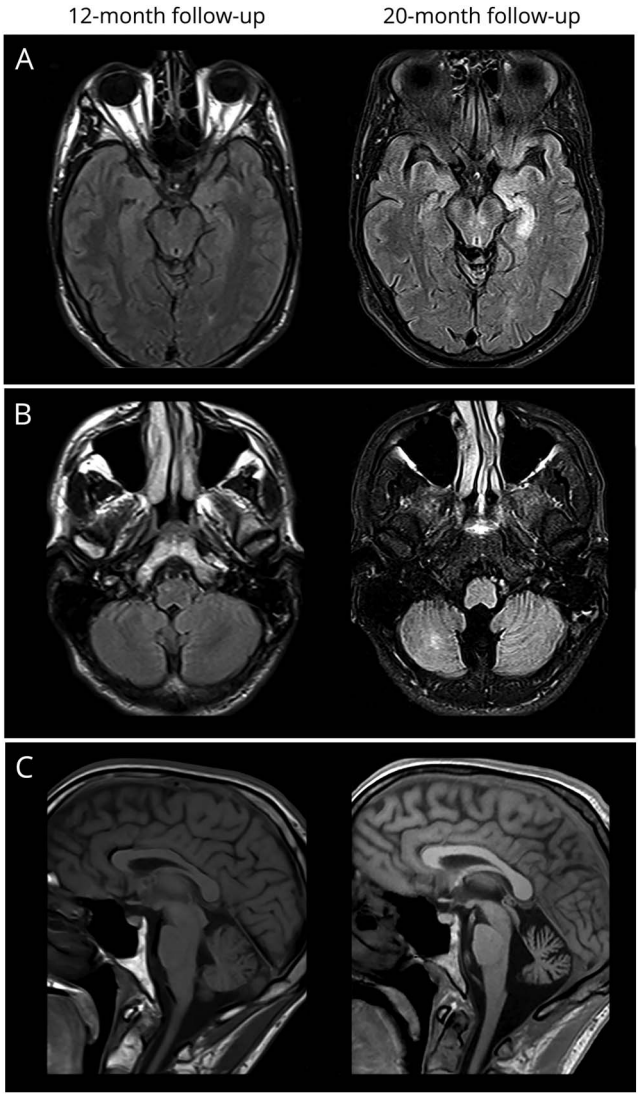
Video

Figure MRI brain at 12 and 20 months after the symptom onset



(A, B) Axial T2 fluid-attenuated inversion recovery sequences with progressively worsening left hippocampal and right cerebellar dentate nuclei hyperintensities over time. (C) Rapidly progressive cerebellar atrophy over 8 months in a sagittal T1 sequence.

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A 32-year-old man presented with hearing loss and gait difficulties. His symptoms, including double vision, dysarthria, dysphagia, neck posturing, and tremors, evolved rapidly over 9 months. Within 15 months of onset, he used a wheelchair. He experienced no improvement after trials of intravenous methylprednisolone, intravenous immunoglobulins, plasmapheresis, and rituximab. His CSF testing was unrevealing except for elevated proteins and oligoclonal bands. He underwent serial brain imaging (figure). Extensive laboratory investigations, including nutritional, metabolic, mitochondrial, infectious, autoimmune, and paraneoplastic panels, were unremarkable (video). Expanded tissue-based immunofluorescence testing revealed positive Kelch-like protein 11 immunoglobulin G in the serum, a newly recognized paraneoplastic encephalitis.[1,2]

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Appendix Authors

Name	Location	Contribution
Joshua Wong, MD	Department of Neurology, Fixel Institute for Neurological Diseases, University of Florida, Gainesville	Designed and conceptualized study, major role in the acquisition of data, drafted the manuscript for intellectual content
Jun Yu, MD	Department of Neurology, Fixel Institute for Neurological Diseases, University of Florida, Gainesville	Major role in the acquisition of data, drafted the manuscript for intellectual content
Addie Patterson, DO	Department of Neurology, Fixel Institute for Neurological Diseases, University of Florida, Gainesville	Major role in the acquisition of data
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Aparna Wagle Shukla, MD	Department of Neurology, Fixel Institute for Neurological Diseases, University of Florida, Gainesville	Major role in the acquisition of data, designed and conceptualized study, revised the manuscript for intellectual content

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