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Tremor and Other Hyperkinetic Movements

Teaching NeuroImages

Acute Cerebellar Ataxia Associated with Modest Elevation of Anti-GAD Antibodies in a Young Patient

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Abstract

Background: Anti-GAD-related cerebellar ataxia has rarely been described as an acute cause of autoimmune ataxia.

Phenomenology Shown: A young female who acutely developed anti-GAD-associated ataxia with magnetic resonance imaging (MRI) showing cerebellar edema and follow-up MRI 6 months later showing cerebellar atrophy.

Educational Value: Recognizing that anti-GAD-associated cerebellar ataxia can present in a young adult as an acute and severe cause of ataxia, with cerebellar changes evident on MRI.

Keywords: Acute ataxia, autoimmune ataxia, anti-GAD 65, glutamate decarboxylase

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A 22-year-old previously healthy Caucasian female presented with myalgia, fever, headache and cough. She tested positive for influenza A and was started on oseltamivir. Two days later, she showed mild confusion and dysarthria, followed the next day by extremities and head tremor and ataxic gait. She went to an outside hospital where cerebrospinal fluid (CSF) results were the following: red blood cells, 9,700 per mm^3; white blood cells 102 per mm^3; segmented neutrophils, 80%; lymphocyte, 14%; protein, 98.5 mg/dL; glucose 61 mg/dL. Human immunodeficiency virus and Lyme antibodies were negative. Brain magnetic resonance imaging (MRI) on admission showed cerebellar edema and hydrocephalus (Figure 1). She was treated empirically with antibiotics, acyclovir, oseltamivir, and dexamethasone 10 mg every 6 hours for 1 week. The following CSF tests were negative: bacterial and fungal cultures, venereal disease research laboratory (VDRL), Herpes polymerase chain reaction, West Nile antibodies, coccidioidomycosis antibodies, *Cryptococcus* antigen, and N-methyl-D-aspartate (NMDA) antibodies and cytology. Her speech remained severely dysarthric. She showed truncal ataxia, kinetic tremor in her four extremities, with dysdiadochokinesia and dysmetria. The lumbar puncture was not repeated because she developed a CSF leak. Chest, abdomen, and pelvis contrast tomography and pelvic ultrasound did not show evidence of malignancy.

Thyroid peroxidase antibodies were positive with a normal thyroid function. At day 19 from symptom onset she received 1 g of methylprednisolone daily for 5 days with minimal improvement of symptoms. After discharge, the send-out laboratory results came back for the central nervous system autoimmune panel showing elevated serum anti-glutamic acid decarboxylase (GAD) 65 titers: 0.07 nmol/L (normal <0.02 nmol/L). She was lost to follow-up for a few weeks. When she attended our clinic, her cognitive problems had completely resolved but her cerebellar symptoms remained

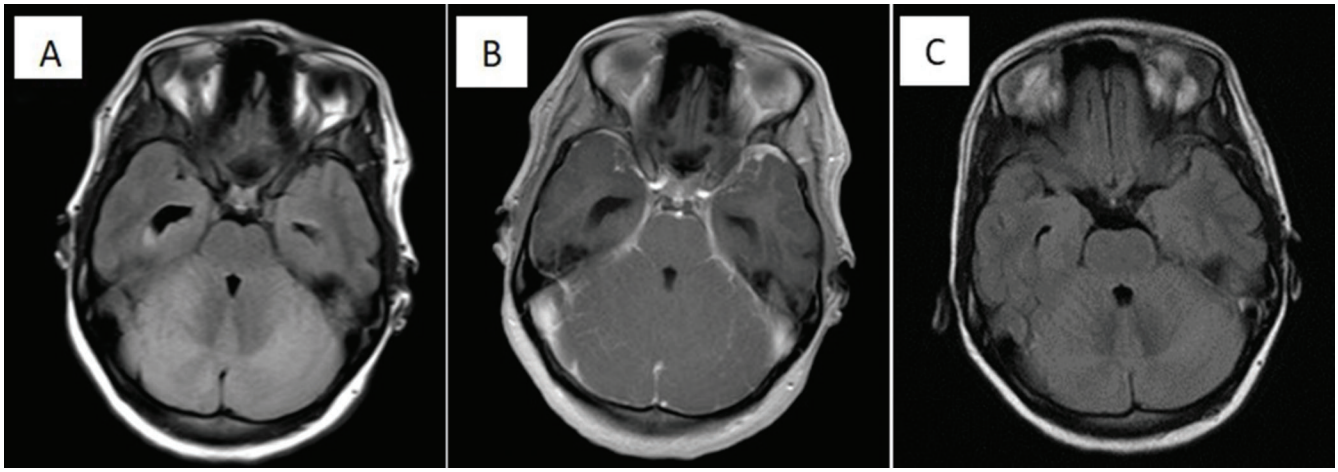

Figure 1. **Brain MRI Demonstrating Cerebellar Inflammation.** (A) Brain magnetic resonance imaging (MRI), axial view, fluid-attenuated inversion recovery sequence showing high signal intensity in the cerebellar hemisphere with dilatation of temporal horns of the lateral ventricles due to hydrocephalus. (B) MRI brain, axial view, post-contrast T1-weighted sequence demonstrating prominence of perforating vessels in the cerebellar hemispheres but no apparent parenchymal enhancement. A and B scans were obtained at the initial presentation. (C) Brain MRI, axial view, FLAIR sequence. This scan was done 20 days from disease onset. It shows subtle increased T2 signal within the bilateral cerebellar hemispheres, improved, in comparison with previous studies. It also shows improvement of the hydrocephalus.

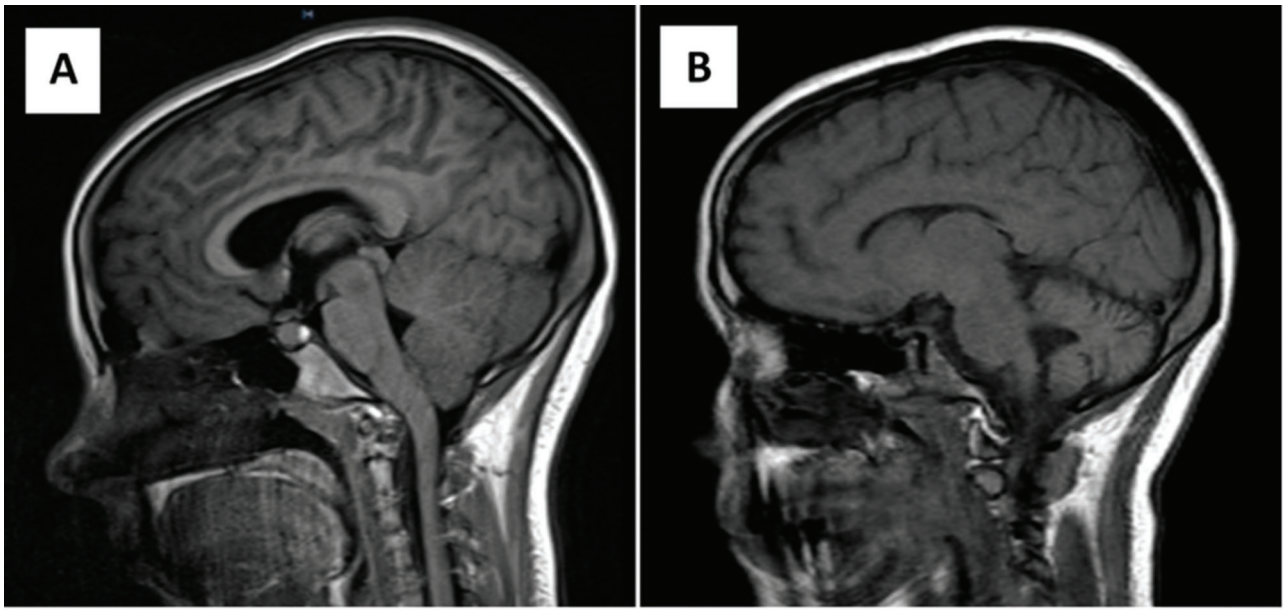

Figure 2. **Brain MRI Demonstrating Cerebellar Atrophy.** (A) Brain magnetic resonance imaging (MRI), sagittal view, T1-weighted image at the level of midline cerebellum, this image was obtained at the initial presentation. (B) Brain MRI, sagittal view, T1-weighted image. This is a follow-up MRI at 6 months after disease onset showing significant cerebellar atrophy.

similar. She was started on monthly intravenous immunoglobulin therapy (0.4 g/kg/day for 5 days, monthly); she has received it for several months with partial improvement of the ataxia. Brain MRI 6 months from disease onset showed significant cerebellar atrophy (Figure 2).

Anti-GAD-associated cerebellar ataxia affects mostly women in about 80–90% of cases, occurring mainly in their sixth decade of life. Most patients develop a chronic presentation, whereas around one-third of cases present with subacute symptoms;[1] very rarely has it been reported as an acute condition.[2]

Imaging studies of anti-GAD 65-related cerebellar syndrome were reported to be either normal or to indicate cerebellar atrophy.[3] This case, with rapid onset of cerebellar symptoms and imaging evidence of active changes in the cerebellum, implies that it can present as an acute cerebellar inflammatory process than can affect young patients.

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