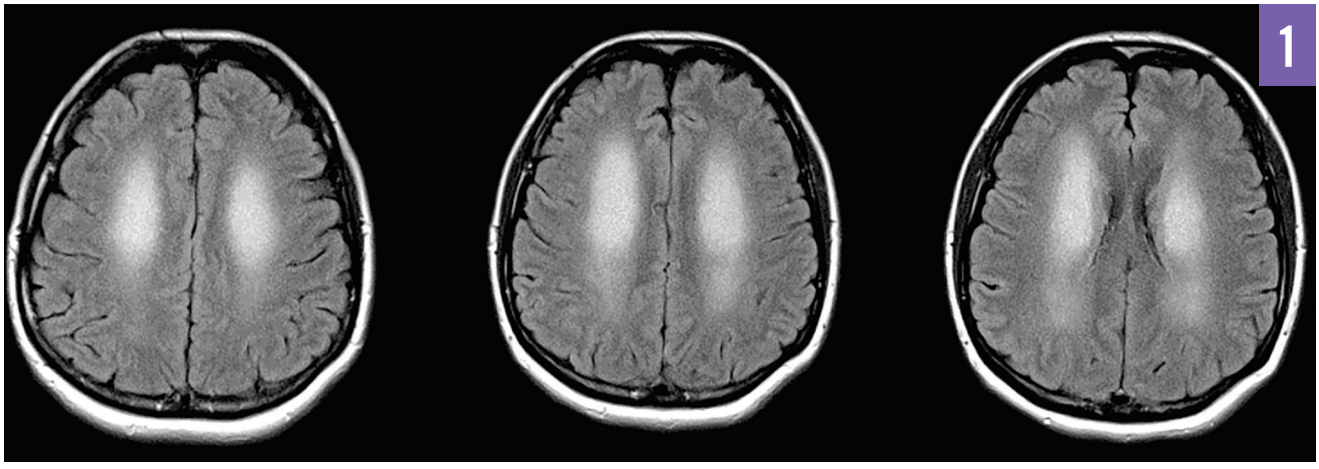
Foresee Your Next Patient



Pretreatment MRI, sagittal FLAIR images, taken at the patient's initial presentation in October 2012.

A Case of Hashimoto Encephalopathy With Long-Term Follow-Up

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A 43-year-old woman with recently diagnosed hypothyroidism presented in October 2012 with a 3-month history of declining cognition, confusion, disorientation, and an inability to communicate or ambulate. She had become incontinent and had signs of gait ataxia and poor motor control. Over this time period, she had become wheelchair- and bed-bound. Family members described the woman as being increasingly lethargic and less engaged.

Results of laboratory testing at that time revealed an elevated antithyroglobulin (anti-Tg) antibody level of 681 IU/mL (reference value, ≤40 IU/mL) and an elevated antithyroid peroxidase (anti-TPO) antibody level of 550 IU/mL (reference value, ≤9.0 IU/mL).

Results of magnetic resonance imaging (MRI) of the brain showed diffuse hyperintense signal abnormalities on fluid-attenuated inversion recovery (FLAIR) (**Figure 1**) and T2-weighted (**Figure 2**) sequences affecting the subcortical white matter. There was no enhancement with GAD on T1 images.

Bedside electroencephalography findings were normal, with no evidence of focal slowing, epileptiform discharges, or electrographic seizure. Results of laboratory workup for infectious and metabolic etiologies—including herpes simplex virus, Lyme disease, cryptococcosis, West Nile virus, anti-Smith antibodies, anti-Ro/anti-La antibodies, and antiphospholipid antibodies—were negative.

Given the subacute encephalopathy, the laboratory test results, and the imaging results, along with a lack of another discernible cause, a diagnosis of Hashimoto encephalopathy was entertained. The patient was initially treated with intravenous immunoglobulin (IVIG) therapy (400 mg/kg/day for 5 days) and oral prednisone. The family reported that this led to a slight improvement in the woman's mentation.

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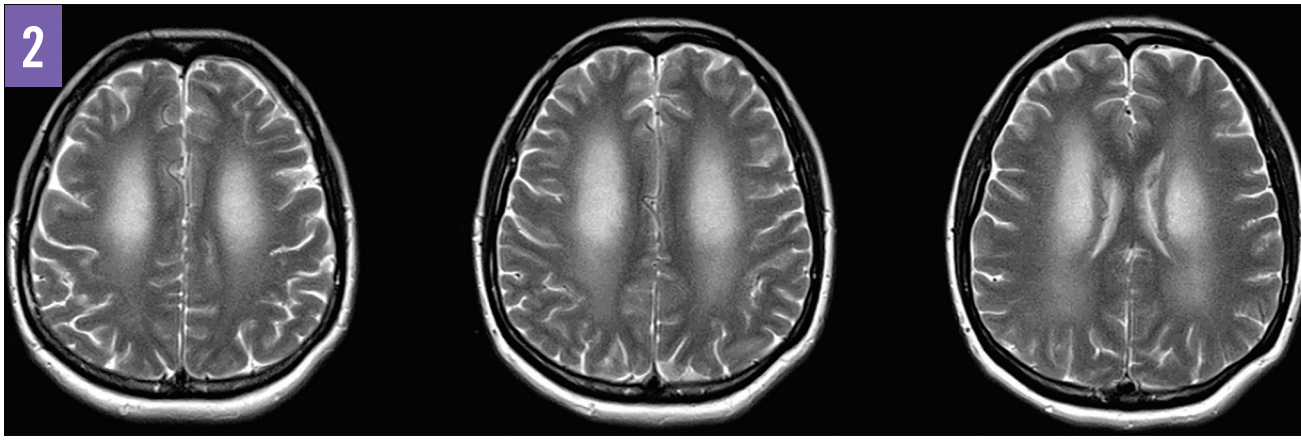
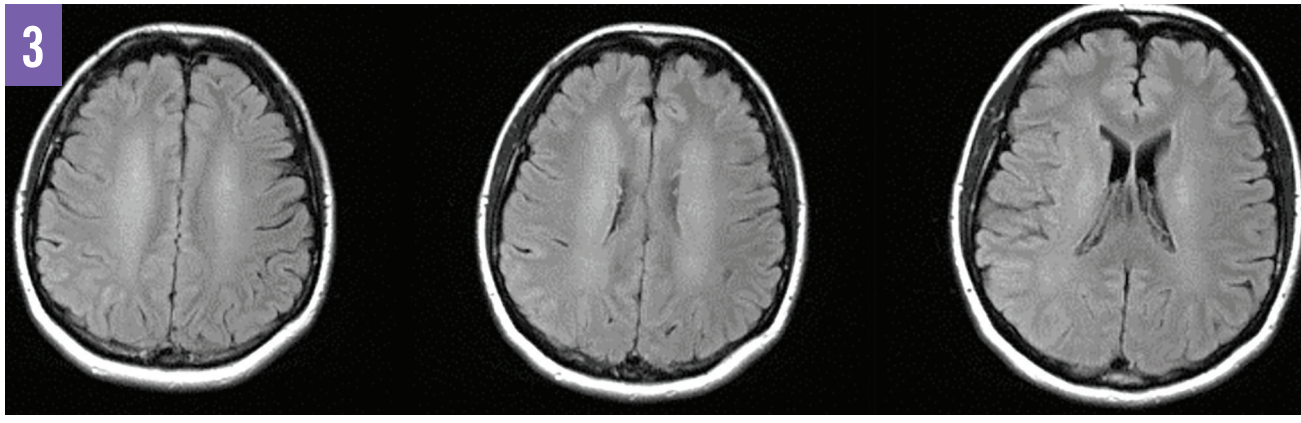
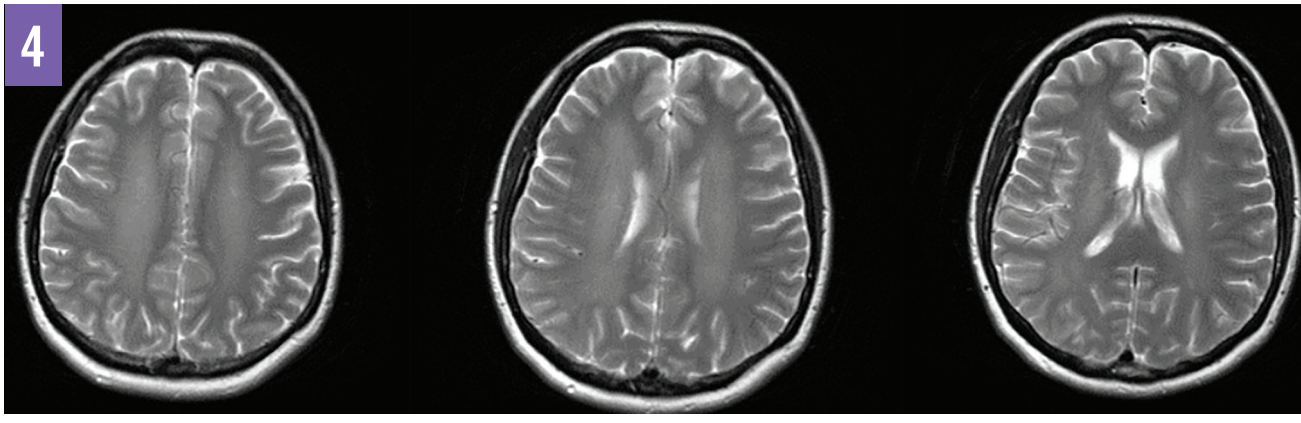


Pretreatment MRI, sagittal T2-weighted images, taken at the patient's initial presentation in October 2012.



Posttreatment MRI, sagittal FLAIR images, taken at follow-up in March 2013



Posttreatment MRI, sagittal T2-weighted images, taken at follow-up in March 2013.

The patient was discharged with ongoing physical therapy and occupational therapy and a plan for repeated cycles of long-term IVIG therapy. She was continued on cycles of IVIG, 400 mg/kg/day for 3 days, every 4 weeks. The oral prednisone was tapered and eventually discontinued.

At a follow-up visit in March 2013, the patient demonstrated drastic improvement in mental status and near-resolution of ataxia. She was able to return to full-time work. Repeated MRI of the brain at that time revealed significant improvement in subcortical white matter hyperintensities on FLAIR (**Figure 3**) and T2-weighted (**Figure 4**) sequences. Her improvement correlated with a gradual decline in anti-TPO antibodies. She was continued on IVIG therapy every 4 weeks.

The patient was followed up with repeated antibody testing and imaging studies. The final IVIG cycle was completed in January 2014. She was started on azathioprine, 150 mg/day, which was discontinued in 2016. Anti-TPO antibody levels were interval-monitored; test results in August 2019 showed a value of 7.60 IU/mL (reference value, <9.0 IU/mL). The patient continues to do well with complete clinical, serological, and radiological remission.

Discussion. Hashimoto encephalopathy is a rare type of autoimmune encephalopathy. The presentation is heterogenous, including but not limited to cognitive decline, ataxia, seizures, myoclonus, hallucinations, and stroke-like episodes.[1] Positive test results for anti-TPO antibodies, anti-Tg antibodies, and antibodies to the NH_2-terminal of α-enolase can be seen. MRI findings are usually normal or exhibit nonspecific changes.[2] A clear link has not been established between thyroid autoantibody titers and disease severity.[3]

This patient's case shows that symptoms of Hashimoto encephalopathy can be associated with dramatic changes on MRI and elevations in thyroid autoantibody levels. Reversal of these changes with immune therapy led to clinical remission. Anti-TPO antibodies may serve as a biomarker correlating with clinical severity and may be useful for guiding the treatment duration.[3] Further research is needed to establish this correlation and to define the optimal treatment protocol. ■

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