

CASE REPORT

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A Patient with Hashimoto's Encephalopathy Presenting with Convulsive Seizure Alone as the Initial Symptom

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Abstract: A 71-year-old Japanese woman with Sjögren syndrome, Hashimoto's disease and a 6-month history of cognitive impairment was admitted to our hospital because of consciousness disturbance and convulsion. Her convulsive seizure disappeared by intravenous administration of diazepam following carbamazepine, and conscious level became alert the next day. But, her cognitive function was persistently deteriorated, and a score of mini-mental state examination (MMSE) was 17/30 points. Magnetic resonance imaging (MRI) and single photon emission computed tomography (SPECT) of the brain did not show any abnormal findings. The electroencephalogram showed increased slow waves in bilateral parieto-occipital regions. Serum anti-thyroglobulin antibodies were elevated (1780 U/ml), but thyroid function was within the normal range. In addition, anti-NAE (NH₂-terminal of α -enolase) antibodies were positive. We diagnosed Hashimoto's encephalopathy, and started steroid therapy. Her cognitive function gradually improved after steroid therapy, and convulsive seizure did not recur until 3 months later.

We emphasize that Hashimoto's encephalopathy should be considered even in patients with convulsive seizure of adult onset without thyroid dysfunction.

Keywords: encephalopathy, Hashimoto's thyroiditis, epilepsy, anti-thyroglobulin antibodies

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Hashimoto's encephalopathy (HE) was first described by Brain et al as autoimmune encephalopathy associated with Hashimoto's disease (HD).¹ Steroid therapy is effective for HE,² but diagnosis can be difficult because of few specific neurological and neuroradiological features.³ Here, we report a patient with HE who presented with convulsion alone as the initial symptom.

A 71-year-old Japanese woman was admitted to our hospital with a disturbance in consciousness and convulsions. She had previously been diagnosed with HD and Sjögren syndrome (SjS) at the Tokai University's School of Medicine (Department of Rheumatology). She was found at home standing among scattered clothes and food and suffering from a convulsive seizure affecting mainly the left side (including her face), which continued for several minutes in the ambulance. Neurological examination showed no abnormality except for frequent convulsive seizures localized on the left side of the face. Convulsion disappeared following intravenous administration of diazepam (5 mg). She became alert on day 2, but cognitive dysfunction remained. Her mini-mental state examination (MMSE) score was 17/30 points.

Laboratory examinations revealed anti SS-A antibodies ($\times 16$). Anti-thyroglobulin antibodies (ATGA) were 1780 U/ml (normal range, below 40 U/ml), whereas TSH, T3 and T4 levels were within normal ranges. Anti-NAE (NH₂-terminal of α -enolase) antibody was positive by measuring in immunoblotting. Cerebrospinal fluid (CSF) showed elevated total protein (58 mg/dl; (normal range, 15–55 mg/dl)), no pleocytosis and negative oligoclonal bands. The cytodagnosis was class I. Brain magnetic resonance imaging and single photon emission computed tomography were normal. The electroencephalogram (EEG) showed excess slow waves in parieto-occipital regions.

Carbamazepine (400 mg/day) was started to prevent convulsions. We diagnosed HE and high-dose glucocorticoid therapy (methylprednisolone 1 g/day intravenous for 3 days) was initiated, followed by oral prednisolone therapy (30 mg/day). These therapies resulted in improved cognitive function, improved MMSE (25/30 points, Fig. 1) and reduction of slow waves on EEG.

HE is thought to be autoimmune encephalopathy associated with anti-thyroid antibodies.² However, the antibody itself is considered a bystander because of the lack of relation between the titer and sever-

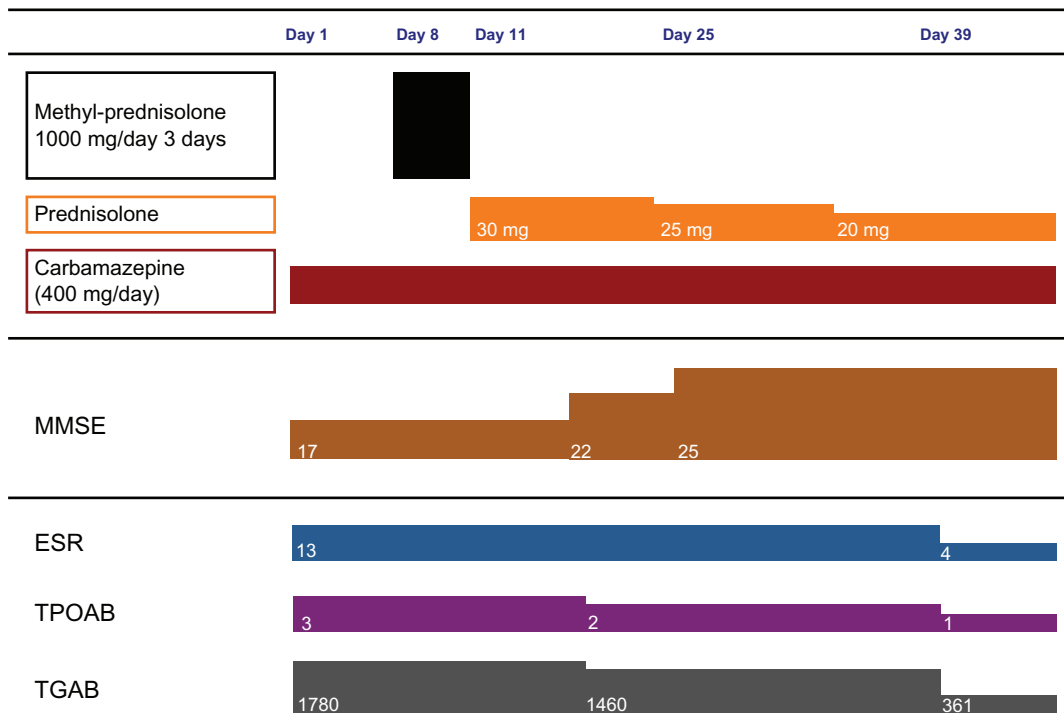


Figure 1. Clinical course.

Abbreviations: MMSE, mini-mental state examination; ESR, erythrocyte sedimentation rate; TPOAB, anti-thyroid peroxidase antibodies; TGAB, anti-thyroglobulin antibodies.



ity of disease.³ In contrast to myxedema, thyroid function is not clinically predictive for this disorder.² HE most frequently shows the following signs and symptoms: seizures, myoclonus, hallucinations or paranoid ideations, and stroke-like signs.⁴ Because of the non-specific clinical features, differential diagnosis of HE from other encephalitis (collagen disease, paraneoplastic syndrome, infection), metabolic derangements, and psychiatric illness is difficult.^{5–7} Anti-NAE antibody is reported to have a high degree of specificity⁸ and to be useful for differential diagnosis.

We first considered SjS-related autoimmune encephalopathy, but there was no laboratory evidence. We reached the final diagnosis of HE based on the past history of HD, the positive reaction for anti-thyroid antibodies, anti-NAE antibody, and good responsiveness to steroid therapy.

In conclusion, convulsive seizure is frequently seen as a presenting symptom of HE. However, the importance of convulsive seizure as initial symptom was not emphasized on previous reports. Therefore, HE may often be overlooked in patients with unidentified convulsive attacks. We suggest that regardless of thyroid dysfunction, HE should be considered in such patients and anti-thyroid antibodies should be examined.

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Author Contributions

Conceived and designed the concept: AM. Analyzed the data: AM. Wrote the first draft of the manuscript: AM. Contributed to the writing of the manuscript: AM. Agree with manuscript results and conclusions: WT and STakizawa. Jointly developed the structure and arguments for the paper: STakizawa. Made critical revisions and approved final version: STakizawa. All authors reviewed and approved of the final manuscript.

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