

# Hashimoto's Encephalopathy Presenting with Acute Cognitive Dysfunction and Convulsion

## Case Report

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Hashimoto's encephalopathy is an immune-mediated disorder characterized by acute or subacute encephalopathy related to increased anti-thyroid antibodies. Clinical manifestations of Hashimoto's encephalopathy may include stroke-like episodes, altered consciousness, psychosis, myoclonus, abnormal movements, seizures, and cognitive dysfunction. Acute cognitive dysfunction with convulsion as initial clinical manifestations of Hashimoto's encephalopathy is very rare. We report a 65-year-old man who developed acute onset of cognitive decline and convulsion due to Hashimoto's encephalopathy. (2013;3:70-73)

**Key words:** Hashimoto's encephalopathy, Seizures, Cognition

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## Introduction

Hashimoto's encephalopathy (HE) is an acute or subacute encephalopathy with elevated anti-thyroid antibodies in patients with Hashimoto's thyroiditis. The age of onset varies, but most frequently occurs to people in their 40s. Approximately 85% of the patients are women. Its clinical features are as follows; decreased cognitive function, abnormal behaviors, myoclonus gait apraxia language disorders, confusion and psychosis.<sup>1,2</sup>

The diagnosis should be considered in patients with encephalopathy with elevated thyroid autoantibody level and without any other etiologies. It is known to show a good response to steroids and other immune therapies.<sup>2,3</sup> The cognitive dysfunction of HE is often observed and it usually has progressive course. If cognitive dysfunction lasts for a prolonged period of time, it is related with poor response to treatment. It is very rare that it occurs all of a sudden.<sup>4</sup>

We report a case of HE presenting acutely developed cognitive decline and convulsion.

## Case

A 65-year-old man visited the hospital due to a sudden onset of cognitive decline. Two weeks prior to visiting to the hospital, the patient caught a cold for one week. He couldn't remember where the razor was and was unable to find the bathroom. He confirmed that he indeed brushed his teeth when asked in case certainly he didn't

brush his teeth and didn't use toothpaste when brushing his teeth. At work, he was unable to find the office keys nor able to recognize his co-workers. As well, he was unable to recall the road he routinely walked along, where he parked his car and if he owned a car. He didn't give the hospital staff proper attitude while saying there was something when he saw the electric light turned on. Besides, he showed the nervousness as serious as being restless and after that he presented urinary incontinence. He collapsed turning his body around to the left as neurological examination were performed and lost consciousness with the convulsion to move his head to the right for about 20 seconds when he visited the hospital. His medical records reported hypertension without thyroid disease or dementia.

When he visited to the hospital, his vital signs showed a blood pressure of 135/80 mmHg, respiration rate of 22 times per minute, pulse of 80 times per minute and a body temperature of 36.5°C. His physical check-up showed no hair loss, edema, goiter, and etc. Neurological examination showed a clear consciousness with disorientation to time and place. Korean version of Mini-Mental State Examination (K-MMSE) showed a 2/5 point in time orientation, 4/5 in place orientation, 0/3 in the three word recall and 4/5 point in serial seven subtraction test. The total score was 22. There were no abnormal results among complete blood count, serum chemistry and electrolytes, serum tumor markers, paraneoplastic syndrome antibodies, and autoimmune disease and connective tissue disease test. Erythrocyte sedimentation rate and C-reactive protein level were also normal. Thyroid function test confirmed the asymptomatic hypo-

thyroidism; the level of triiodothyronine was 62.98 ng/dL (reference value: 65-150), that of thyroxine 0.88 ng/dL (reference value: 0.78-1.54), that of thyroid stimulating hormone 5.85  $\mu$ IU/mL (reference value: 0.55-4.78  $\mu$ IU/mL). The level of thyrotropin-releasing hormone was normal. According to the thyroid autoantibody tests, the level of anti-thyroglobulin antibody was 77 IU/mL (reference value: 0-60) and that of anti-thyroid microsomal antibody was >1,300 IU/mL (reference value: 0-60). The cerebrospinal fluid (CSF) showed a slightly increased level of protein (58 mg/dL) and the normal level of white blood cells. And there were no abnormalities in virus including herpes simplex, bacteria and Mycobacterium tuberculosis tests. CSF 14-3-3 protein was negative.

Seoul Neuropsychological Screening Battery (SNSB) carried out 4 days after he was hospitalized revealed a decrease in language and visual memory and language fluency, but there were no abnormalities in the concentration and time and place orientation.

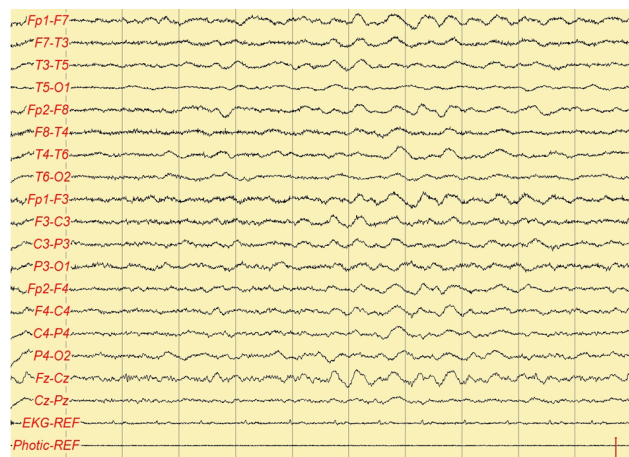


Figure 1. Electroencephalography revealed diffuse slow waves.

Electroencephalography (EEG) during awake revealed intermittent diffuse slow waves (Fig. 1). There was no abnormal signal in brain MRI (Fig. 2). Single photon emission computed tomography (SPECT) showed decreased in perfusion in the bilateral temporal lobes (Fig. 3). Ultrasonography demonstrated an increased vascularity and irregular echoing in the bilateral thyroid lobes. Chest and abdomen CT showed no abnormality. It was diagnosed as Hashimoto's encephalopathy based on the clinical features and test results. After oral steroids (prednisolone 60 mg/day) and anti-convulsant (Levetiracetam 1,000 mg/day) administration, he showed a gradual recovery of cognitive function. Therefore, on the 14th day after he was hospitalized, the score of K-MMSE became normal to 30/30. Since then, the patient's cognitive dysfunction gradually improved and showed no suffering from any convulsions.

## Discussion

We presented a patient with Hashimoto's encephalopathy who had a sudden cognitive deterioration and convulsion. The sudden decline of cognitive function is a rare clinical feature and it is unusual for the HE to affect the old man. The common cold he caught before the onset of the disease is considered to affect the symptoms' beginning.

Its pathophysiological mechanism is not clearly known, but the factors considered to cause it are as follows; vasculitis by autoimmune mechanism, antineuronal antibody mediated reaction, and the direct toxicity of thyroid stimulating hormone releasing hormone to the central nervous system.<sup>2,3</sup> The elevated serum level of anti-thyroid antibody is essential in diagnosing HE and it means the autoimmune mechanism of the thyroid gland.<sup>2</sup>

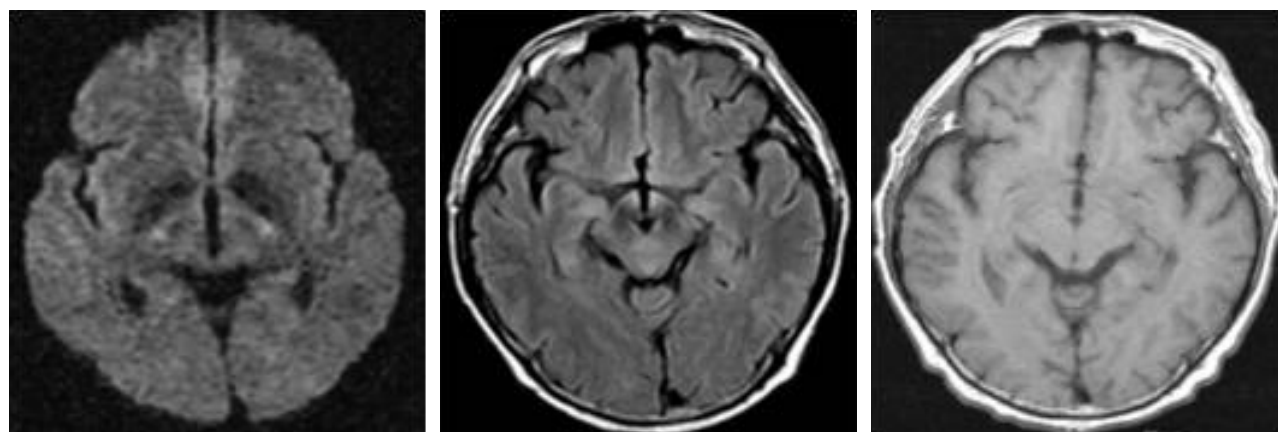
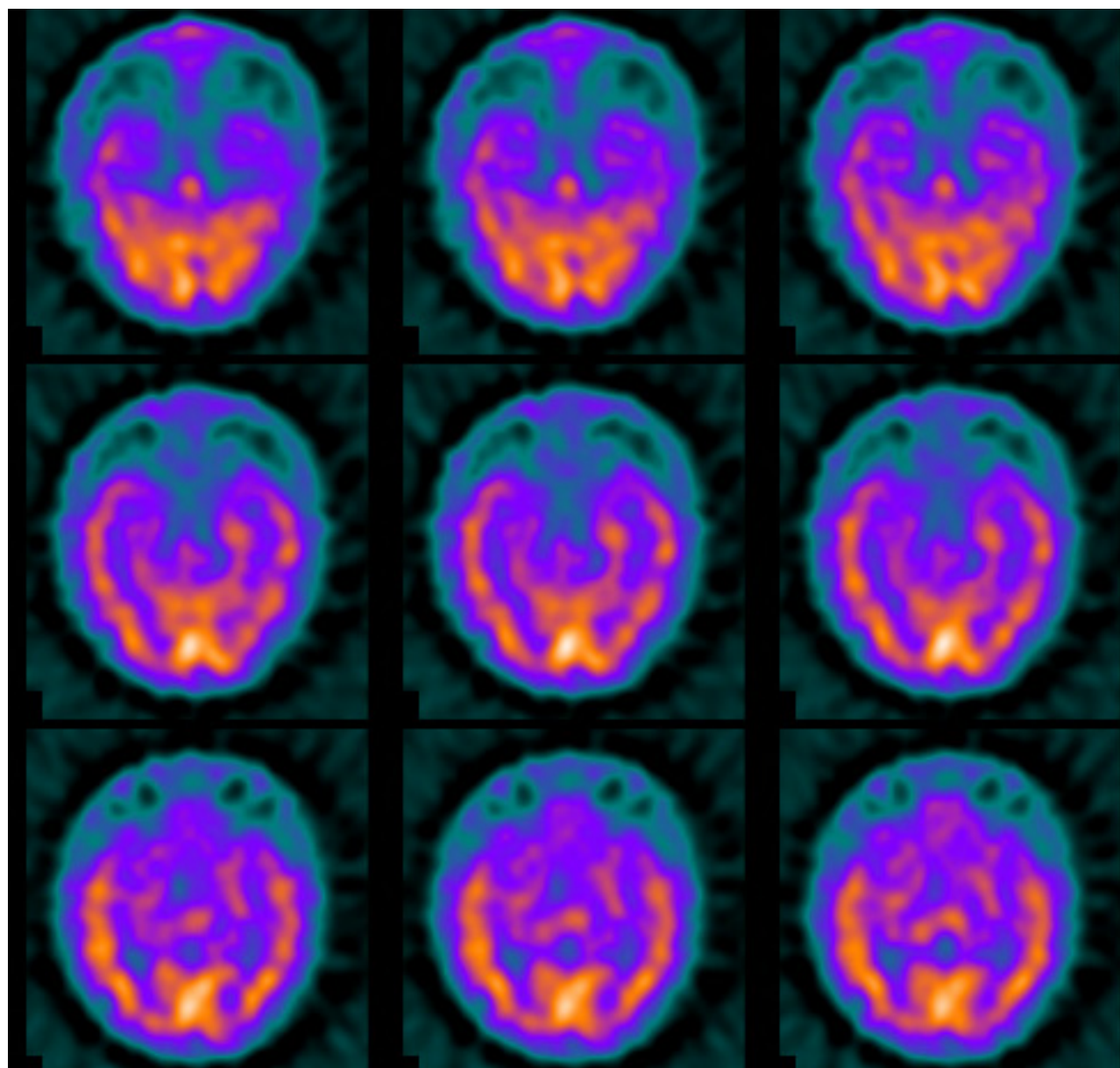


Figure 2. Brain MRI at admission showed no definite abnormalities.



**Figure 3.** Single photon emission computed tomography demonstrated decreased perfusion in the bilateral temporal regions.

HE may not be related to the functional status of thyroid gland and it can appear in various ways. It shows asymptomatic hypothyroidism 23-35%, hypothyroidism 17-20%, hyperthyroidism 7% and normal thyroid 18-45%.<sup>2</sup> It often shows the normal thyroid function, a little decreased function of thyroid gland and a high level of thyroid gland autoantibody.<sup>2,3</sup> We presented a case with asymptomatic hypothyroidism and the high titer of autoantibody. Clinical manifestations of HE are various and non-specific, but it is generally classified into two types. One consists of about 25% presenting focal neurological deficits repeatedly such as stroke

and the other consists of about 75% showing diffuse progressive course such as dementia, confusion, and hallucination.<sup>1,2,5</sup>

Two clinical manifestations can occur simultaneously in the course of HE.<sup>5</sup> Here, it occurred in the way that convulsion is added while the sudden decline of cognitive function is the main symptom. It is considered to happen in the way the focal neurological abnormality is combined to the diffuse clinical pattern of the disease.

The two thirds of patients with HE may have focal or generalized seizure and the generalized one is more common.<sup>1,2</sup> It rarely happens to have status epilepticus.<sup>2,6</sup> The EEG characteristics are various

and mostly show non-specific features.<sup>1,2</sup> The slow wave is the most common and appears generally or restricted to forehead or temporal area. Focal spike, sharp wave and transient epileptic discharges were rare.<sup>1,6</sup> These EEGs are related to the severity of the disease and may show improvement according to the response to medical treatment.<sup>6</sup>

The cognitive decline is often observed in HE and it has relatively gradual clinical course. The neuropsychological studies which were carried out before and after treatment are rare. As the cognitive decline starts, all domains of cognition may be affected. With progression, there are high probability of decreased executive function and procedural memory.<sup>7</sup> With the worsening of cognition, frontal lobe dysfunction become more common.<sup>7</sup> Compared with mild Alzheimer's disease, HE shows impairment in episodic memory concentration, visuospatial function, and executive function but showed normal in naming test.<sup>8</sup> In this patient, there were some problems in orientation, recall, concentration, and calculation. on the day the patient was hospitalized. And in the SNSB carried out 4 days after visiting a hospital, it showed mainly the impairment in memory and partial frontal lobe dysfunction.

Brain imaging studies frequently reveal non-specific findings.<sup>4,9</sup> The brain MRI is sensitive to find the lesion and it is likely that signal changes can appear in subcortex, basal ganglia, cerebellum, and medial temporal lobe.<sup>1,2,4,9</sup> SPECT may show focal, multifocal, or global hypoperfusion.<sup>10</sup> The brain imaging may show reversible improvement after treatment for HE.<sup>9,10</sup>

This case handles HE with sudden onset of cognitive deterioration and convulsion, which abruptly appeared after the patient caught a common cold without the clinical evidences of hypothyroidism.

When the sudden cognitive function decrease or convulsion appears, HE should be considered regardless of age and gender.

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