Recurrent Hashimoto’s Encephalopathy, Showing Spontaneous Remission: A Case Report

Lin LI, Fen-ping Zheng, Guoxing Wang and Hong Li

Abstract

Hashimoto’s encephalopathy (HE) is a rare condition associated with Hashimoto’s thyroiditis (HT). It is characterized by neurological-psychiatric symptoms, high levels of anti-thyroid antibodies, non-specific radiological examinations or electroencephalogram abnormalities, and responsiveness to corticosteroid treatment. We describe the case of a man with HE who showed decreased mentality, cognitive impairment, dysarthria, and gait disturbance. The initial attack was improved rapidly by corticosteroid treatment. When the symptoms recurred in 7 months, the patient achieved spontaneous remission without corticosteroid treatment. The recognition of the condition was essential for the prognosis and treatment of this rare disease.

Key words: Hashimoto’s encephalopathy, Hashimoto’s thyroiditis, hypothyroidism, treatment

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Introduction

Acute encephalopathy can result from various causes, including stroke, infection, toxic material, seizure, and metabolic disturbance. Hashimoto’s encephalopathy (HE) is also one of the causes of treatable encephalopathy. Since the first report of Brain et al (1), there have been more than 200 patients reported with HE (2). HE is known to be associated with autoimmune thyroid disease (3). Almost all HE patients are affected by Hashimoto’s thyroiditis (HT), although some patients with Graves’ disease or subacute thyroiditis have been also reported (4, 5). HE is characterized by neurological-psychiatric symptoms, high levels of anti-thyroid antibodies, non-specific radiological examinations or electroencephalogram abnormalities, and responsiveness to corticosteroid treatment. The clinical manifestations can be fluctuating or persistent, and reversible (6). Here we report a case of recurrent encephalopathy that manifested spontaneous remission.

Case Report

A 69-year-old married Asian man, with a university education, from Hangzhou, was brought to hospital in November 2009 due to decreased mentality, cognitive impairment, dysarthria, and gait disturbance. His wife could not understand his speech and he could not dress himself well. He had no previous psychiatric history; he was being treated for chronic thyroiditis with L-thyroxine at a dose of 62.5 μg/day. On admission his temperature was 36.4 °C and pulse rate was 62 beats/minute. Physical examination showed no abnormal findings in either the chest or the abdomen. The neurologic examination revealed memory impairment, dysarthria, myoclonus with no paresis in the extremities, gait disturbance and no signs of meningeal irritation. No involvement was seen in either cranial nerves or sensations. All deep tendon reflexes were normal with negative Babinski’s sign on both sides. There were no abnormal findings in routine laboratory data, including hematology, urinalysis, and blood chemistry. The thyroid function showed free T3 3.06 pmol/L (3.50-6.50 pmol/L); free T4 15.70 pmol/L (8.90-20.60 pmol/L); thyroid-stimulating hormone (TSH) 7.60 mIU/L (0.35-4.60 mIU/L). Anti-thyroid peroxidase antibody (TPO-Ab) and anti-thyroglobulin antibody (TGAb) were markedly elevated (>1,000 IU/mL). No abnormal findings were detected on radiological examinations, such as MRI including diffusion-weighted imaging of the brain, or on the electroencephalogram (EEG). Magnetic resonance angiography demonstrated no significant stenosis in any branch of...
the main cerebral arteries. Thyroid ultrasound revealed diffuse glandular enlargement with a homogeneous but coarse parenchymal echo texture, multiple discrete hypoechoic micronodules were seen. The diagnosis hypothesized was Hashimoto’s encephalopathy. The treatment was methylprednisolone 500 mg/day for three consecutive days, followed by prednisone with initial dose of 60 mg/day. In the first week, a considerable improvement in the level of consciousness and myoclonus was observed. The prednisone dosage was gradually tapered in the subsequent two months to 5 mg/day. In this period there was a regression of all signs and symptoms.

Unfortunately, seven months after the initial admission, he had decreased mentality, cognitive impairment, dysarthria, and gait disturbance again. He could not walk without assistance and it became difficult for his wife to understand his speech. After admission to our hospital, there were no abnormal findings in routine laboratory data, radiological examinations (Fig. 1) and EEG (Fig. 2). A repeated thyroid function test demonstrated free T3 2.39 pg/mL (1.71-3.71 pg/mL); free T4 0.87 ng/dL (0.7-1.48 ng/dL); TSH 24.17 mIU/L (0.35-4.94 mIU/L). The TPO-Ab and TGAb were markedly elevated (>1,000 IU/mL). Serology, cultures and diagnostic imaging did not indicate infection, collagen disease, or vitamin deficiency. ESR (4 mm) and CRP (0.2 mg/L) were normal. ANCA, anti-DNA and ANA were negative. Biochemical results showed aminotransferases, blood glucose, serum creatinine, serum electrolytes and vitamin B12 were normal. Mini-mental evaluation scale (MMSE) examination revealed that this patient had mild cognitive dysfunction (score 25). Both cognitive impairment and involuntary movement gradually improved without any treatment other

Figure 1. Brain MRI demonstrates no abnormal findings in T1 diffusion-weighted image (A), T2 diffusion-weighted image (B), contrast-enhanced T1 diffusion-weighted image (C) and fluid-attenuated inversion recovery (D).
than successive administration of L-thyroxine (75 μg/day). The patient completely recovered with no obvious symptoms, and was discharged from our hospital 1 week after admission. He has been in good neurological condition without any relapse of neuropsychological symptoms to date.

Discussion

The first case of HE was reported in 1966. Criteria for the diagnosis of HE included unexplained occurrence of relapsing myoclonus, generalized seizures, psychiatric disorders or focal neurological deficits and three conditions among which are: abnormal EEG, elevated thyroid antibodies, elevated CSF protein, excellent response to steroids and unrevealing cerebral MRI (7). After this early proposal the clinical range of symptoms and signs was greatly expanded. The present patient was diagnosed as HE, after we excluded vascular, toxic, metabolic, and infectious disease. Various clinical manifestations of HE have been reported. Decreased mentality, cognitive impairment, dysarthria, gait disturbance and high serum concentrations of thyroid autoantibodies have been described as common clinical features and accepted as diagnostic criteria. The present case showed dramatic neurological improvement following steroid treatment, which is one of the typical clinical characteristics of HE.

Hashimoto’s encephalopathy has a wide variety of clinical manifestations, and it also has many ways of remission. According to the analysis of published cases, 42 patients on steroid therapy recovered without relapses, 38 relapsed or had no effect, 11 improved with residual deficits and 19 had spontaneous improvement of which 14 relapsed and 5 remained stable (7). The initial attack of our patient was improved rapidly by corticosteroid treatment. When the symptoms recurrence, the patient achieved spontaneous remission without corticosteroid treatment. Therefore even in the same patient, the means of remission may be different.

In the present patient, the TPO-Ab and TGAb were markedly elevated. TPO-Ab is present in 95-100% and TGAb in 73% of patients with HE (8). Elevated titers of antithyroid antibodies are the most relevant paraclinical finding, which could be considered to be a mark of HE. Elevated serum level of TPO-Ab may be with vasculitic type HE and elevated serum levels of TPO-Ab and TG-Ab may be with diffuse progressive type of HE (7). However, the role of those antibodies and their pathophysiology are unknown.

Cerebrospinal fluid (CSF) analysis is abnormal in approximately 80 percent of patients, usually revealing an elevated CSF protein level (9). Unfortunately, the present patient refused the examination, so we could not analyze the patient’s CSF.

The authors state that they have no Conflict of Interest (COI).

References

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