Case Report

Hashimoto Encephalopathy with Sensory Polyneuropathy: A Case Report

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ABSTRACT

Hashimoto encephalopathy is an autoimmune disease characterized by an increase in antithyroid antibodies in the serum of patients with multiple neuropsychological symptoms. We report a case with lower level of antibody and fairly response to corticosteroid; however, clinical presentation, brain magnetic resonance imaging findings and the negative results of other diseases confirmed Hashimoto encephalopathy diagnosis. First, a relatively good corticosteroid response was seen but after one weak, the patient withdrew her drug and got back with a corticosteroid resistant progressive attack one month later. Our case had a mild increase in antithyroid antibodies and pure sensory polyneuropathy and did not show significant response to corticosteroid therapy. Can antithyroid antibodies titers be a marker of corticosteroid treatment response? this should be investigated in the future studies.

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Introduction

The term Hashimoto encephalopathy (HE) was first used to report on a disease in 1966 by Lord Brain that had various neurological symptoms with varying levels of the thyroid hormones (1). HE is an autoimmune disease characterized by an increase in antithyroid antibodies in the serum of patients suffering multiple neuropsychological symptoms such as cognition disorders, seizures, myoclonus, aphasia, and psychosis (1, 2). The last few years have witnessed a growing interest and focus on such a disease due to its being treated, the mortality rate for its being untreated, and being chosen in differential diagnosis of dementia and other neurological disease such as Creutzfeldt-Jakob disease (CJD) (1, 3). Various forms of the disease have been reported such as acute and stroke-like, subacute and chronic-progressive type (3, 4).
In this paper, we introduce a patient with

subacute onset of different neuropsychological and cerebellar signs that first respond rapidly to corticosteroid but after treatment withdrawal, she got back with new relapse that did not show good response to corticosteroid and was treated with cyclophosphamide.

Case Report

The patient was a 24-year-old woman who gradually developed behavioral and personality changes such as anxiety, paranoia, and impaired memory since 2 months before admission and she progressively showed impairment of gait and speech. When the patient was hospitalized, she had no signs of unconsciousness, but she developed cognitive impairment such as impaired orientation and memory, decreased verbal fluency, poor concentration, impaired judgment and difficulties in calculating numbers. Psychiatric symptoms of anxiety, paranoia, hallucination, and shyness were evident in the patient. Cranial nerve examination was normal except for impaired pursuit movements. There was no significant motor system involvement. Mild ataxia and impaired tandem gait was detected in cerebellar examination.

In paraclinical studies, the first brain and cervical magnetic resonance imaging (MRI) was normal (Figure 1); electroencephalography (EEG) showed diffuse slowing; and in cerebrospinal fluid (CSF) analysis, there was an increased level of protein. In abdominopelvic ultrasound and chest X-ray, no specific findings were found.

Patient's thyroid test showed a high thyroid-stimulating hormone (TSH) level of 12 mIU/l (normal range: 0.3-4.5) but other thyroid values were normal. Antithyroid peroxidase (anti-TPO) and antithyroglobulin antibodies were 275 IU/ml (normal range: less than 50) and 364 IU/ml (normal range: 5-100) which were about 4 and 6 times more than the upper limit of normal, respectively.

All collagen vascular and infectious tests including antinuclear antibody (ANA), antiphospholipid antibody, anticardiolipid antibody, antineutrophil cytoplasmic antibody (ANCA), anti-saccharomyces cerevisiae antibody (ASCA), anti-alphafodrin antibody, anti-Ro antibody, anti-La antibody, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), rheumatoid factor (RF), Wright, veneral disease research laboratory test (VDRL), human immunodeficiency virus (HIV), and anti-borreliosis antibody were negative and serum folate, vitamin B_{12}, and angiotensin converting enzyme (ACE) levels were within the normal limits.

With the probable diagnosis of HE, the intravenous injection of 100 mg hydrocortisone every eight hours was started. A rapid and fairly good treatment response was seen. The psychiatric symptoms, gait disturbance, dysarthria and EEG abnormality experienced a relatively good improvement after three days.

After one week, the patient discontinued treatment because of intolerability and one month later, she returned with paraparesis and bilateral Babinski sign. CSF analysis and all mentioned lab tests repeated again. All of them were within normal limit except for CSF and antithyroid antibodies levels that showed similar abnormality of the past exams. Nerve conduction study (NCS) showed axonal sensory polyneuropathy more obvious in the lower limbs.

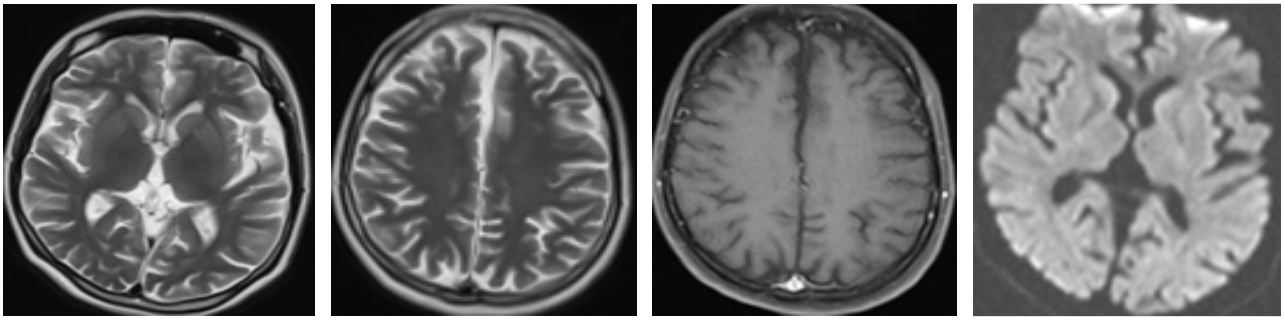

Figure 1. The first magnetic resonance imaging (MRI); No obvious abnormality was seen

Brain MRI showed bilateral symmetric oval-shape hyperintensities in centrom semioval with restricted diffusion and without enhancement (Figure 2-A and B).

Intravenous methylprednisolone (1 g daily) was started for three days and then, maintenance therapy with 50 mg prednisolone per day was continued. Since no significant response and no decrease in serum antithyroid antibody level were seen after 7 weeks, cyclophosphamid pulse was started. Partial therapeutic response occurred in the patient forces but not in neuropathy. Antithyroid antibodies levels got back to the normal limit. The patient's follow-up images in the 4 months after the treatment showed no significant radiological changes except mild brain atrophy (Figure 3).

Brain MRI findings, increased antithyroid antibodies, neuropsychiatric manifestations and the negative results of other differential diagnosis confirmed the diagnosis of Hashimoto encephalopathy.

Discussion

Hashimoto encephalopathy was previously determined by encephalopathy, elevated thyroid antibodies, and often responded well to corticosteroid treatment (5, 6). However, for the time being, it is a diagnosis of exclusion (5).

The neurological symptoms can be classified as acute, sub-acute or chronic-progressive types (2, 3). In various pathological conditions, the disease is described as different conditions including cerebral vasculitis, cerebral edema and demyelinating lesions which can justify various modes of clinical disease such as strokes-like attacks, seizures and psychosis (1). Ataxia and myoclonus are other representations of the disease (7).

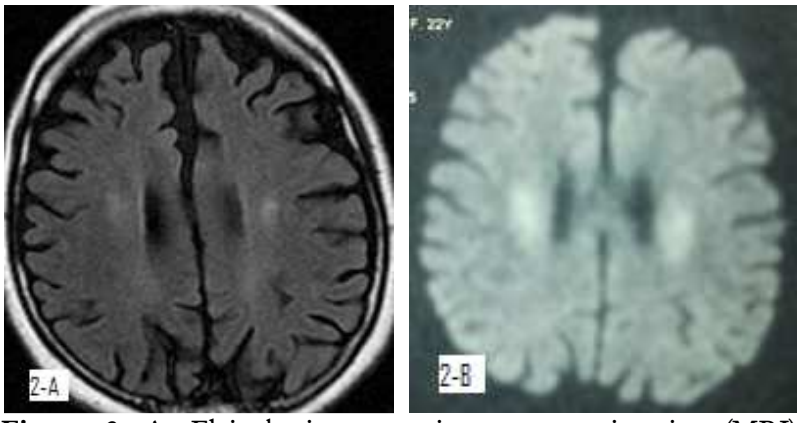

Figure 2. A. Flair brain magnetic resonance imaging (MRI) showed two hyperintense periventricular lesions; B: Diffusion weighted imaging recovered restriction

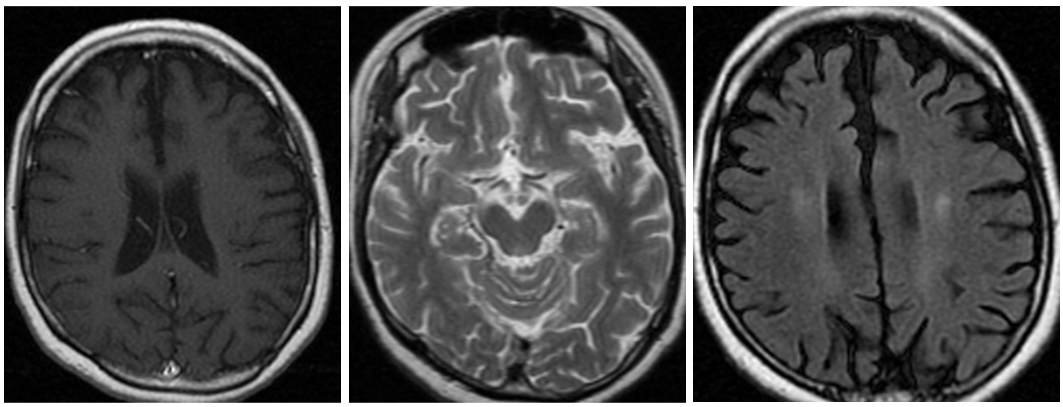

Figure 3. T1 brain magnetic resonance imaging (MRI) (left image), mild brain atrophy in T2 (middle image), and two hyperintense periventricular lesion in fluid-attenuated inversion recovery MRI (FLAIR) (right image)

Dementia and progressive cognitive impairments may also be seen in these patients (8, 9). About 35% of patients have simultaneous subclinical hypothyroidism, 30% of them have euthyroid and approximately 20% are hypothyroid (10).

But in all cases, the level of one or both antithyroid antibodies, anti-TPO and antithyroglobulin level are increased (10). The EEG is abnormal in 98% of patients and can be seen as diffuse slowing or epileptiform discharges (5).

Brain MRI can be normal in about half of the cases and non-specific hyperintense lesions in the ventricular zone can be seen as strokes-like pathology (5). Diffusion weighted (DWI) and apparent diffusion coefficient (ADC) imaging are helpful in earlier lesions detection (2).

In the CSF fluid analysis, increased protein level may also be seen (10). Concomitant encephalopathy and peripheral neuropathy are rarely reported. Different types of polyneuropathy are reported in literature including motor neuropathy (11), demyelinating neuropathy (12) and sensory polyneuropathy (13, 14). HIV infection or vitamin B_{12} deficiency should be considered in differential diagnosis. Corticosteroid is the most important medicine for treatment of HE (3, 6, 8). However, in some cases, no response has been seen, especially in patients with chronic progressive types (3). In patients who are non-responder to corticosteroid treatment with cyclophosphamide, intravenous immunoglobulin (IVIG) and plasma exchange can be prescribed (15).

Increased antithyroid antibodies, brain MRI findings, neuropsychiatric manifestations and rule out of other possible causes are compatible with the diagnosis of HE. Sensory neuropathy, which was mentioned in our case, is a rare complication of HE. In our case, there was a mild to moderate increase of antithyroid antibody and little response to corticosteroid therapy.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

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