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CASE REPORT

Hashimoto Encephalopathy in Case of Progressive Cognitive Impairment; a Case Report

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Abstract

Hashimoto's encephalopathy (HE) is a rare condition characterized by atypical psychiatric and heterogeneous neurological manifestations such as acute cerebral ischemia, seizure, tremors, myoclonus, psychosis, depression, cognitive disorders, and fluctuating loss of consciousness. Here, a case of 28 year-old man was reported who referred to the emergency department (ED) with different acute neurologic disorders and final diagnose of HE.

Key words: Encephalopathy; unconsciousness; cognition disorders; immunoglobulins, thyroid-stimulating; neurologic Manifestations

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Introduction:

Hashimoto's encephalopathy (HE) is a rare condition characterized by atypical psychiatric and heterogeneous neurological manifestations such as acute cerebral ischemia, seizure, tremors, myoclonus, psychosis, depression, cognitive disorders, and fluctuating loss of consciousness (1-3). Despite a wealth of studies identifying the etiology of HE, its exact pathogenesis is not still completely understood (4, 5). Due to non-specific findings it is often considered to be a diagnosis of exclusion. Currently, HE is considered to be a treatable dementia but there is no consensus on the duration or drug of choice for treatment. Here, a case of 28 year-old man was reported who referred to the emergency department (ED) with different acute neurologic disorders and final diagnose of HE.

Case report:

A previously healthy 28 year-old man was referred to the ED with a history of ophthalmoplegia and ataxia followed by progressive cognitive impairment. Two months prior to this episode, mild neck pain and sore throat were the only findings in his past medical history. He had no history of alcohol consumption, substance abuse, medication use, congenital disease, syncope, ischemic or hemorrhagic cerebrovascular attract, seizure, trauma, or any other known medical problems. The patients' on-arrival vital signs were as follow: systolic blood pressure (SBP): 120 mmHg, pulse rate (PR): 90/minute, respiratory rate (RR): 14/minute, oral temperature: 37.5◦C, oxygen saturation 96% with nasal cannula and 100% oxygen, and Glasgow coma scale (GCS) 15/15. Physical examination revealed jerky movements in limbs, normal size and reactive pupils, fluctuating disorientation, severe cerebral ataxia, bilateral sixth nerve and upward gaze palsy, and increased deep tendon reflexes. A mini mental state examination resulted in 10 of 30 points. The patients' head and neck examination, lung and heart sounds, four limbs pulses, and capillary refile were normal. After assessment of airway, breathing, and circulation (ABC) patients were checked in terms of coma cocktail. Blood sugar was measured 100 mg/dl with glucometer. Electrocardiography (ECG) revealed normal sinus rhythm with normal axis. Brain computed tomography had not any abnormal findings, but bilateral hyper-signal white matter lesions were detected on fluid attenuation inversion recovery (FLAIR) and T2 weighted magnetic resonance imaging (MRI) (Figure 1). Cerebrospinal fluid (CSF) analysis results were as follows: protein: 170 mg/dl, glucose: 67 mg/dl, cell count: 80/mm3 with lymphocyte dominancy. All of laboratory parameters as cell blood count, coagulation profile, kidney and liver function tests, venous blood gas parameters, and electrolyte were in normal range. Neurology consult was taken and patient admitted in neurology ward for further evaluations. Electroencephalography showed abnormal nonspecific sharp and spike waves in a slow background. Neural antibodies such as anti–N-methyl-D-aspartate receptor (NMDAR) were negative. Thyroid statues demonstrated subclinical hypothyroidism. Anti-thyroglobulin and anti-peroxidase antibod-

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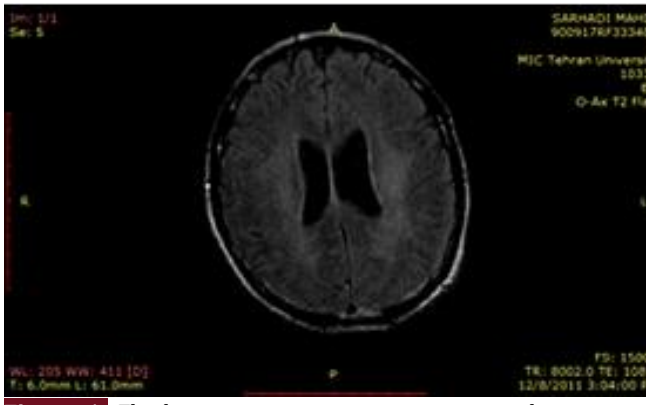

Figure 1: Fluid attenuation inversion recovery shows symmetric white matter lesion before treatment ⇧

Figure 2: Fluid attenuation inversion recovery shows improvement in white matter lesion after treatment ⇧

ies were 220 IU/ml and 740 IU/ml, respectively, which were significantly higher than normal ranges. All other assessments were normal. Taking the above data into consideration and excluding other differential diagnosis, HE was presumed. The patient underwent ten days of Methyl Prednisolone pulse regime (one gram per day) with tapering Prednisolone (120 mg orally per day) for two weeks. All signs and symptoms especially cognition and ophthalmoplegia dramatically improved and white matter lesions disappeared after two weeks of treatment (Figure 2).

Discussion:

Heterogeneous clinical manifestations and absence of non-specific tests have made the HE diagnosis as a challenging problem for physicians. Susan lee et al. in a review concluded that psychiatrists should be aware of this often unrecognized entity to ensure accurate diagnosis and timely treatment (6). HE has different MRI manifestations from normal appearance to demyelination, ischemic lesions, edema, and atrophy (7). Usually, HE is diagnosed by high levels of anti-TPO antibodies, normal T4, and thyroid stimulating hormone (TSH) titers in the presence of above mentioned heterogeneous clinical manifestations (8). Recent studies have suggested cerebrospinal fluid titer of anti-thyroid antibodies as a pathognomonic test for HE (1, 9). According to the published case series, steroid is the only accepted treatment for this disease and among 81 adults and children, about 50% of patients recovered completely and the other cases relapsed or improved with residual deficits (10, 11). Previous studies have recommended three to five days corticosteroids (12). The present study is the first one that give the patient 10 days of Methyl Prednisolone. The patient responded completely to the corticosteroid. It seems that a favorable prognosis may depend upon rapid recognition of HE and aggressive steroid treatment. However, further studies are needed to evaluate the advantage of this type of treatment for disease.

Conclusion:

In the case of unknown psychiatric and neurologic manifestations, measuring serum level of thyroid hormones and CSF titer of anti-thyroid antibodies could be helpful in limitation of differential diagnosis and timely initiation of proper treatment.

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Conflict of interest:

Authors declared no conflict of interest.

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Authors' contributions:

All authors passed four criteria for authorship contribution based on recommendations of the International Committee of Medical Journal Editors.

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