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Case Report

Hashimoto Encephalopathy with an Unusual Presentation of Status Epilepticus Seizures: A Case Report

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Abstract

A 33 yr old man, previously diagnosed with hypothyroidism, presented with decreased level of consciousness and generalized tonic-clonic (GTC) seizure to Namazi hospital, Shiraz, Iran, during April 2015. The patient later referred with another episode of seizure like attack for which he received phenytoin, carbamazepine and levothyroxine and was discharged. During his last admission, the patient was admitted with chief complaints of decreased consciousness and four GTC attacks. On admission, the patients had aphasia, ataxia, loss of verbal communication, eye contact and complete loss of obedience. Thyroid function tests showed low levels of T3 and T4 with high levels of thyroid stimulating hormone. Other blood tests were all either normal or slightly abnormal. Lumbar puncture and CSF analysis had a high titer of Anti-TPO antibodies. With high suspicion of Hashimoto encephalopathy, pulsed methyl prednisolone (10 mg) was administered, however the patient showed little improvement. Therefore, plasmaphresis was started, to which the patient showed dramatic response.

Keywords: Hashimoto encephalopathy, Epilepsy, Iran

Introduction

Hashimoto Encephalopathy (HE) is defined as a steroid responsive encephalopathy associated with autoimmune thyroiditis (1). Symptoms are non-specific and include a wide spectrum of neurologic deficits. Among all neurologic symptoms, status epilepticus is a rare presentation (2). Herein, we report a case of HE with an unusual presentation of drug resistant status epilepticus.

Case Report

A 33 yr old man, previously diagnosed with hypothyroidism, presented with decreased level of consciousness and generalized tonic-clonic (GTC) seizure to Namazi hospital, Shiraz, Iran, during April, 2015. The seizure lasted for about 1–2 min and the patient recovered completely after that episode, so the patient did not refer for any medical care. After a few days, the patient experienced another seizure-like attack and developed urinary and defecation incontinency, aphasia, total body weakness and disorientation. At that time, the patient was admitted to a hospital and in 7 days of his admission, his signs and symptoms improved and he was discharged with

phenytoin, carbamazepine and levothyroxine. After these events, the patient had three more hospital admissions due to similar symptoms. After a one-month symptom free period, he experienced another GTC seizure attack. The patient experienced four episodes of GTC seizures during the 24 h prior to his last admission, during which the patient did not regain consciousness after the second episode of GTC seizure.

On admission, the patient was aphasic, did not obey completely and had impaired gait. The GTC seizures were controlled by administering intra venous phenytoin. During the hospital admission, the patient developed myoclonic jerks, followed by mental disorders including aphasia, ataxia, loss of verbal communication, eye contact and complete loss of obedience. The patient also developed urinary tract infection for which he received ceftriaxone and was resolved.

His laboratory tests for viral markers including the human immunodeficiency and hepatitis C viral antibodies and hepatitis B virus antigen were all negative. The Wright plus test was negative. The Venereal Disease Research Laboratory test (VDRL) was negative. His serum Anti-thyroid peroxidase antibody (Anti-TPO Ab) and blood ammonia levels were high (1095.8 IU/ml and 189.3 μmol/L, respectively). His CPK, lactate dehydrogenase (LDH) and CRP levels were 137 mg/L, 437 U/L and 58 mg/l, respectively. Although the CRP level decreased (28 mg/l) after receiving ceftriaxone.

Thyroid function tests (TFT) showed low levels of T_3 and T_4 with high levels of thyroid stimulating hormone (TSH). Other blood tests, including complete blood count, liver function test, kidney function tests and electrolytes, erythrocyte sedimentation rate, homocysteine levels and the peripheral blood smear were all normal.

Brain magnetic resonance Imaging (MRI) and brain computed tomography (CT) scan were normal. Electroencephalogram (EEG) showed paroxysmal generalized sharp waves.

Various anticonvulsant and antipsychotic medications were prescribed for the patient after he developed myoclonic jerk movements and other mental disorders. The treatment included a combination of sodium valproate, clonazepam and levetiracetam. The combined treatment controlled the myoclonic jerks but did not improve the mental problems.

With no significant improvement in his condition, he underwent two lumbar punctures (LP). The first LP was to rule out herpes encephalopathy and the second was done to check the possibility of HE. The first Cerebrospinal Fluid (CSF) analysis showed an elevated cell count (19500 /mm^3) with two WBCs (two segments), elevated protein levels (56 mg/dL) and normal Glucose and LDH levels. The second CSF analysis had a total cell count of 1600 /mm^3 with a total WBC count of 10 (five lymphocytes, 5 segments) and a high titer of Anti-TPO antibodies. With high suspicion of HE, treatment was started. As the first line of treatment, pulsed methyl prednisolone (10 mg) was administered, however the patient showed little improvement. Therefore, plasmaphresis was started, to which the patient showed dramatic response.

Discussion

With a prevalence of 2.1 in 100000 (3), HE is considered a rare condition. Due to its vague clinical manifestations (2) and uncertain diagnostic tests, it is mostly diagnosed by excluding other conditions (3). The diagnosis is made when high anti-thyroid antibodies accompany encephalopathy symptoms (4), and no other infectious, metabolic or paraneoplastic processes are responsible for a patient's condition (3).

Clinical Manifestations

Patients' clinical symptoms can be categorized into two main types. First the vasculitic type and second the indolent progressive type, with the former including acute stroke-like episodes (27%), seizures (66%) and psychosis (38%) and the latter including dementia, myoclonus and altered consciousness (3). Recently a third set of symptoms has been introduced, as the relapsing-remitting type, with patients showing cognitive dysfunction and psychiatric symptoms (5).

Our patient developed status epilepticus along with other non-specific neurological symptoms such as aphasia, incontinency and disorientation. In Hashimoto encephalopathy, patients most commonly present with seizures, but status epilepticus is a rare symptom to present, manifesting in an estimated 12% of patients (2). To the best of our knowledge, very few cases of HE have been reported in literature with such a presentation.

Diagnosis and work up

As stated earlier, HE presents with a wide variety of symptoms and serological tests and imaging studies usually reveal non-specific results. However, they are necessary to rule out other possible causes of encephalopathy (2, 3).

At the time of the disease onset, thyroid function may vary. As reported, patients are mainly hypothyroid or euthyroid, as in our patient who was a known case of hypothyroidism and presented with hypothyroidism on admission and during his disease course. Rarely patients may be hyperthyroid (2).

Brain imaging findings are non-specific, showing ischemia, granuloma or degenerative process (5). Although the changes are non-specific, more than 50% of patients have an abnormal brain imaging from the onset of their disease course, with the most common finding being cerebral atrophy (1). In our case both brain MRI and CT scan were normal.

EEG is abnormal in almost 100% of the patients, mostly showing transient epileptic activity (1, 3). Our patient was no exception showing paroxysmal sharp waves.

CSF findings mostly document increased cerebral fluid protein and pleocytosis (3, 5). After our patient displayed resistance to antiepileptic medication, LP was performed and CSF analysis showed elevated protein and pleocytosis along with high Anti-TPO Ab titer (6).

Treatment

Initially to control the symptoms, administration of antiepileptic and thyroid modulatory medications is common (7). Then to control the disease from its basic pathogenesis, the key treatment is immunosuppression. First line immunosuppressive medications are corticosteroids, to which most patients show dramatic response, for which the disease is also termed "Steroid Responsive Encephalopathy". Only 10% of the patients do not respond to steroids, for whom alternative treatments such as IVIG, plasma exchange should be started (3).

Ethical consideration

Informed and written consent was obtained from the patient before reporting the case.

Conclusion

If diagnosed and treated in time, HE patients recover with minimal complications. On the other hand, delay in correct treatment can be fatal (8). Due to the non-specific symptoms and laboratory findings, HE is easily missed. Based on our patient we recommend thyroid function tests, serum, and CSF anti-thyroid antibodies be evaluated in all patients with encephalopathy or anti-epileptic-resistant seizures including status epilepticus.

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The authors declare that there is no conflict of interest.

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