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Case Report

A case report on Hashimoto's encephalopathy: an autoimmune neuroendocrine disorder

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ABSTRACT

Hashimoto's encephalopathy (HE) is a rare neuroendocrine disorder with high titers of the thyroid antibodies. HE is more common to women than in men with a ratio of approximately 5:1. The estimated prevalence of HE was 2 per 100,000 people. We present a case of Hashimoto's encephalopathy in a 24-year-old male patient known case of hypothyroidism admitted with the complaints of unconsciousness, and further developed seizure during admission. Brain magnetic resonance imaging (MRI) was normal, electroencephalogram (EEG) revealed slow diffusion, and the serum thyroid function test showed a high concentration of anti-thyroid antibodies. The patient had a good recovery after the management with steroid therapy.

Keywords: Hashimoto's encephalopathy, Anti-thyroglobulin, Anti-TPO antibodies, Steroid-responsive

INTRODUCTION

Hashimoto's encephalopathy (HE) is a rare neuroendocrine Disorder with high titers of the thyroid antibodies.[1,2] It is also termed as "steroid-responsive encephalopathy.[3] First described by the famous British neurologist L. Brain in 1966.[4] HE is more common to women than in men with a ratio of approximately 5:1 and occurs in all age groups. The estimated prevalence of HE was 2 per 100,000 people.[5] The clinical presentation is highly variable; common clinical features include altered consciousness, tremor, seizures, sleep abnormalities, delusions, hallucinations, stroke-like episodes, and impairment of cognitive function, focal neurological deficits, presenile dementia, psychosis, and ataxia.[6] The specific mechanism of HE is not fully understood, it is believed that brain vasculitis and autoimmune anti-thyroglobulin or anti-TPO antibodies directed against common thyroid represents the most likely etiologic pathway.[7] Recently International autoimmune encephalitis Society developed clear criteria for, the diagnosis of HE, which are as follows: encephalopathy with seizures, stroke-like episodes, hallucinations, or myoclonus; subclinical or mild overt thyroid disease without severe thyroid dysfunction; normal neuroimaging data or with non-specific abnormalities; presence of serum thyroid peroxidase antibodies, thyroglobulin antibodies; absence of neuronal antibodies in serum and cerebrospinal fluid (CSF); reasonable exclusion of alternative causes. The immune pathogenic mechanisms of HE responds to steroids or other therapies such as plasmapheresis.[8]

CASE REPORT

A 24-year-old male patient presented to the neurology department. According to the patient relative's statement, on the afternoon of January 2020, being at home in complete health, patient slept around 3 pm and did not awake for a longtime and found unconscious. There was no history facial deviation, headache, and fever during this

period. According to the medical history, patient suffered 2 episodes Status Epileptics on 23.12.19 and developed hypothyroidism at the age of 22 and was on tablet eltroxin 75 mcg once daily for one year. There was no relevant family history. No history of smoking, alcohol abuse or drug addiction. On examination, he was conscious, restless, moving both upper and lower limb, pupil dilated 3 mm on eye reaction to light and no any focal abnormalities. His initial vital signs were as follows, body temperature of 36.5℃, heart rate of 100 beats per minute, respiratory rate of 20 breaths per minute, and blood pressure 130/90 mmHg. Heart auscultation revealed positive S1 and S2. The lungs were clear to auscultation bilaterally in both upper and lower airways with normal vesicular breath sound. Patient was admitted in neurology department for further examination. On the second day of admission patient had 2 episodes of generalized tonic-clonic seizure.

Routine laboratory investigation including full blood count, urea and electrolytes, erythrocyte sedimentation rate and C-reactive protein were within normal range, but vitamin D3 was 8.2 ng/ml (reference <20 ng/ml-deficient). Routine biochemical analyses of liver, renal were all within normal limits. Random glucose readings were 148 mg/dl (reference >200mg/dl). Thyroid function showed, thyroid stimulating hormone (TSH) 17.0 mIU/ml (0.5-6 mIU/ml), triiodothyronine (T3) 70 ng/dl (80-180 ng/dl), thyroxine (T4) 3 ug/dl (4.6-12 ug/dl), free triiodothyronine (FT3) 190 pg/dl (230-619 pg/dl), free thyroxine (FT4) 0.5 ng/dl (0.7-1.9 ng/dl). An autoantibody screen revealed high titre anti-TPO antibody 400 IU/ml (reference 0-50 U/L). Laboratory workup on blood culture, HIV, HBsAg, tTG, IgA were unremarkable. Magnetic resonance imaging (MRI) failed to reveal any abnormality, electroencephalogram (EEG) revealed slow diffusion with minor irregularities in waves. The positive clinical picture, thyroid studies in conjugation with anti TPO, normal MRI imaging and EEG results lead to the diagnosis HE.

During hospitalization, patient started with anticonvulsant intravenous infusion therapy of leviracetam 1.5 g BD, fosophenytoin 150 mg TDS for 5 days followed by oral leviracetam 500 mg BD and phenytoin 100 mg BD for 4 days. The steroid, methyl prednisolone 1g OD was given intravenously for 5 days, followed by methyl prednisolone 60 mg OD for 4 days to treat HE. Oral levothyroxine 200 mcg OD was given for 9 days to maintain the normal thyroid levels. Tablet Shelcal (caco3+vitamin D3) 500 mg TDS was administered to treat vitamin D deficiency. Patient showed improved clinical picture significantly.

At discharge patient was prescribed with oral leviracetam 500 mg and phenytoin 100 mg BD, Oral levothyroxine 200 mcg, methyl prednisolone 60 mg, tablet Shelcal (caco3+vitaminD3) 500 mg OD. Steroids were gradually tapered over a period of 3 months. The patient remained healthy during his follow-up visit on 7th month without ant recurrences.

DISCUSSION

Hashimoto's encephalopathy (HE) is a rare steroid responsive neuroendocrine disorder.[5] HE occurs predominantly in females, with a male to female ratio 1:5, and between the age of 45 to 55 years.[9] However, it can affect any age group including children and the noted prevalence is high in girls than boys.[2,9] Recent studies revealed thyroid status does not correlate with development of HE, almost 40% were euthyroid, around 30% of patients had subclinical hypothyroidism, 20% had hypothyroidism, less than 8% had hyperthyroidism at presentation. Our patient was hypothyroid.[2,6,15] The pathogenesis of Hashimoto's encephalopathy is still controversial.[9] Autoimmune vasculitis, autoantibodies against brain-thyroid represent the most probable etiologic pathway.[6,9] Recently, Blanchin et al reported that TPOAb from HE patients could bind cerebellar astrocytes in HE patients but not in Hashimoto thyroiditis patients.[11,12] This may support the role of TPOAb in the pathogenesis of HE. In addition, the titre of thyroidantibodies does not correlate with severity or improvement neurological illness.[2] The clinical presentation is characterized by numerous neurological and neuropsychiatric symptoms, it include seizure and altered consciousness (51%), myoclonus, altered sensorium (38%), hallucinations and psychosis (30%), stroke-like symptoms (21%), cognitive impairment and memory loss (48%), tremor and involuntary movements (12%), and ataxia (6%).[9,11,13] Therefore, presenting symptoms of HE may be quite variable. Diagnosis of HE should be considered in patients presenting with the neuropsychiatric symptoms and high levels of thyroid antibodies in serum or CSF, in particular thyroperoxidase antibodies excluding other causes of encephalopathy, such as the central nervous system involvement of vasculitic syndromes, metabolic disease, electrolyte imbalance, intracranial infection, poisoning or toxins, and neoplasm.[1,3,4,13,14] EEG abnormalities presented were nonspecific in majority of patients with slow background diffusion.[2,3,10,14] Brain MRI may show abnormalities such as cerebral atrophy, focal cortical abnormality, diffuse subcortical abnormality and non-specific subcortical focal white matter abnormality.[3,12,14] In our patient the diagnosis of HE was made by the noted high serum thyroid peroxidase antibodies, slow background diffusion of waves shown by EEG, positive neurological symptoms, normal MRI, after excluding all other potential causes. Steroid is the first line therapeutic choice, proved to be the most successful and effective treatment method for HE.[8,12] Because of this fact HE is also termed as "steroid-responsive encephalopathy associated with Hashimoto's thyroiditis".[2] High-dose intravenous methylprednisolone therapy is the preferred choice for acute encephalopathy of HE, followed by prednisone (1–2 mg/kg/day, max 60 mg/day) for 6–8 weeks, followed by gradual tapering.[13] Patients symptoms improvement may be seen in a few days and significant clinical improvement occurs over 3–6 months after initiation of corticosteroid treatment. Accordingly, our patient started on Methylprednisolone with marked

improvement in his condition during the short term follow up. Some authors suggested steroid treatment for at least 6 months, but duration of therapy should be designed as per the EEG, neuropsychological testing and clinical response. Immunomodulatory treatments, such as azathioprine, cyclophosphamide, mycophenolate mofetil, methotrexate, intravenous immunoglobulin, and plasmapheresis, alone or in combination, have been used in some cases non-responsive to corticosteroids.

CONCLUSION

HE is a rare neuroendocrine disease related to autoimmune thyroid diseases. The incidence is probably underestimated because of low overall awareness about the disease. We suggest that HE be considered in any patient with the presence of high thyroid antibody levels and unexplained encephalopathy presenting with uncontrolled seizures and cognitive dysfunction. HE can be excluded as a diagnosis for patients with normal serum levels of the thyroid antibody. HE is being an autoimmune origin; corticosteroid treatment usually provides a dramatic recovery.

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