

Case Report of Steroid Resistant Hashimoto Encephalopathy Presenting with Non-Convulsive Status Epilepticus

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Abstract

Hashimoto's encephalopathy is considered as an autoimmune encephalopathy characterized by sub-acute altered level of consciousness associated with high titers of anti-thyroid peroxidase antibodies. In steroid resistant cases, intravenous immunoglobulin, plasmapheresis and other immunotherapy may be an effective treatment. Herein, we describe a case report of 62 years old lady presented for acute onset of altered level of consciousness. All labs, lumbar puncture and imaging including MRI of brain were not significant. Thyroid function tests were normal, but anti-TPO was more than 1000. Excluding all other common causes of altered level of consciousness, elevated level of Anti-TPO lead us to think of Hashimoto encephalopathy. We treated the patient with steroid bolus therapy for 3 days and completing with dose of 1 mg/kg for a total of seven days, with no improvement. Then, we decided to do several sessions of plasmapheresis after which the patient started gaining her consciousness. This case further supports plasma exchange as a potentially beneficial treatment modality, in severe cases of Hashimoto's encephalopathy which are completely or partially resistant to steroids.

Keywords: Hashimoto Encephalopathy (HE); Plasmapheresis; Steroid-resistant

Introduction

Hashimoto's encephalopathy is generally a steroid responsive disorder characterized by high titers of anti-thyroid antibodies, where patient commonly present with sub-acute onset of confusion, episodes of myoclonus, seizures, and stroke like episodes [1]. In steroid resistant cases, other treatments have been described in case reports including plasmapheresis [2,3] and intravenous immunoglobulin [1,4]. In this case report we describe a plasmapheresis responsive Hashimoto Encephalopathy. We found that the use of plasma exchange maybe an important choice of treatment in patients who fail to respond to steroids.

Case Presentation

We report a case of 62-year-old woman know to have mild hypertension controlled by perindopril presented to the emergency department for disorientation, ataxia of 1 day duration followed by a generalized tonic seizure with complete loss of consciousness. As by family, the patient's symptoms started 2 months prior to this episode when the patient had several episodes of confusion of few seconds duration.

Urgent CT brain was conducted and was reported normal. Lumbar puncture showed no significant abnormalities. Patient's routine labs were not relevant. Anti-NMDA in cerebral spinal fluid was negative and also ANA in blood was negative.

On the second day of presentation, her altered level of consciousness was complicated by respiratory arrest so she got intubated directly and put on mechanical ventilation. TSH level was done and was normal but Anti-TPO level was more than 1000, so we suggested that she is having HE and begun pulse steroid therapy 1 g IV

for 3 days in addition to Levetiracetam 500 twice daily and Valproic acid 400 mg three times per day. Patient's level of consciousness didn't improve after one week of treatment. EEG showed sustained non-convulsive seizures. MRI brain done and showed typical picture of PRES syndrome.

Although, EEG done showed no seizure activity, patient's level of consciousness didn't improve despite anti-epileptics and steroid therapy. The level of anti-TPO decreased till 500 after pulse steroid therapy, but then re-increased again to 700. Plasmapheresis for five consecutive days was decided. She started to move her right upper limb just after the third session and even to open her eyes spontaneously then we added two more sessions. After seven sessions, the patient started to move her head and eyes toward voice and to localize pain. One month later the patient is totally conscious, able to talk, to eat and to walk with help.

Discussion

Hashimoto encephalopathy is a rare syndrome with a variety of neuropsychiatric manifestations associated with elevated thyroid antibodies and usually responds well to steroid therapy. It was first described in the literature in 1966 [5]. The incidence of HE is estimated to be 2 per 100.000, and is more common in females with a female male ratio 4:1 and a mean age of 50 to 60 years old [6].

The pathogenesis of HE is not well established. Despite that the elevated anti TPO is one of the diagnostic criteria, its effect is unclear. Ferracci et al., declares that there is no direct pathogenic due to absence of correlation between anti-TPO level and clinical status [7]. Other reports suggest autoimmunity against specific CNS antigen as alpha enolase which was found to be a specific CNS antigen in HE [8].

Hashimoto encephalopathy has a wide and variable presentation. In our review of literature, rare cases of status epilepticus were reported as a clinical picture of HE. Where most was found to be refractory to the

antiepileptic drugs as our patient who did not respond to dual antiepileptic drugs and generalized sedation was used to induce remission of status epilepticus [9].

Diagnosis of HE depends on association of neurological and psychiatric manifestations plus high levels of anti-TPO [7]. In relevant diagnosis, there is a good response to steroid therapy. Thyroid function had no influence on the disease with almost most cases have been euthyroid (18% to 45%), hypothyroidism (clinical in 25%-35% and subclinical in 17%-20%) and rarely hyperthyroidism (7%) [9]. CSF analysis may show mild lymphocytic pleocytosis and increased protein. EEG will give generalized slowing activity, epileptiform abnormalities, and focal slowing triphasic waves. CSF and EEG findings were return to normal after response to treatment.

Our patient admitted complaining of decrease level of consciousness and episodes of seizures complicated by non-convulsive status epilepticus. This is the first case in literature that presents a case of Hashimoto disease with a clinical picture of non-convulsive status epilepticus. She didn't respond to steroid which was reported in some case reports who suggest treating such cases using immunotherapy and plasmapheresis [10,11].

Conclusion

In conclusion, this case supports the literature with a new steroid resistant HE with marked clinically improvement after plasmapheresis. It also highlights on the wide variety of clinical presentations of HE by which we are reporting the first case of HE presenting by non-convulsive status epilepticus.

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