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Case Report

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Hormone responsive hashimoto encephalopathy: a case of hashimoto encephalopathy treated with only oral thyroid medication and symptomatic therapy for seizures and psychosis, but without steroids

Abstract

Hashimoto encephalopathy is a rare disease associated with autoimmune thyroiditis. Symptoms may include delirium, encephalopathy, seizures, and psychosis. Pathophysiology of this disease is not completely understood. This condition is often under-diagnosed if not suspected and should be considered in patients with diverse neuropsychiatric manifestations in whom no obvious etiology of encephalopathy can be determined. Anti-thyroid antibodies and thyroid peroxidase antibodies are crucial for diagnosis. Along with supportive management, empiric steroid treatment is usually administered as soon as infectious and other causes are ruled out. We report a case of Hashimoto thyroiditis which presented with agitation, psychosis, hallucinations, complex partial seizures. In this case the disease was controlled with only oral thyroid therapy and supportive management with antiepileptic drugs and antipsychotic drugs, but without the use of steroids.

Keywords: Hashimoto encephalopathy; Autoimmune thyroiditis; Seizures; Delirium

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Introduction

Hashimoto encephalopathy was first described in 1966 by Brain et al.,[1] in a 49-year-old man with progressive impairment of consciousness and cognition interspersed with episodes of confusion or coma, agitation, hallucinations, tremor and recurrent stroke-like episodes, who had histologically confirmed Hashimoto thyroiditis [1]. Several cases of Hashimoto encephalopathy have been reported since then.[2-5] There are however controversies regarding its establishment as a specific disorder.

Case history

A retired 71-year-old man living independently presented to the emergency department with new onset complex partial seizures and psychotic behavior. Laboratory work up revealed normal complete blood count, serum electrolytes, kidney function and liver function, however his thyroid stimulating hormone level was high with low free thyroxine levels. Magnetic resonance imaging of brain was normal. A preliminary diagnosis of secondary seizure was made and the patient was treated with antiepileptic drugs and was discharged home on oral thyroid supplement with plans to obtain an outpatient electroencephalogram and neurology follow up in clinic. The seizures recurred and his psychosis worsened over next two days evolving to partial complex status epilepticus and encephalopathy with delirium. He presented again to the emergency department and was admitted for further workup and management. Electroencephalogram was severely abnormal with diffuse polymorphic slowing and sharp wave activity suggesting epileptic encephalopathy. His clinical course at that time was associated with delirium and acute psychosis manifested as visual hallucinations, agitation, manic episodes and insomnia.

Laboratory work up revealed normal complete blood count, serum electrolytes, kidney function and liver function, his thyroid stimulating hormone was elevated to 77.4 and free thyroxine levels was very low, 0.8. Cerebrospinal fluid analysis was normal, no evidence of infection was found in blood, urine and cerebrospinal fluid and patient did not have fever or any other signs of infection. MRI brain was unremarkable (Figure 1).

He was managed with oral thyroid supplementation, antipsychotics and antiepileptic medications (levetiracetam). He improved over next 2 days with cessation of seizures and improvement of delirium. He continued to improve over next 2 weeks and was discharged to extended care facility from which he was discharged to home after 10 days. Electroencephalogram at discharge and two months later was normal. He did not receive any steroid therapy at any time during or after hospitalization.

Laboratory data received after discharge showed that elevated thyroid peroxidase antibodies and anti-thyroid antibodies were present on admission. He was followed up in clinic 2 months later and by telephone 10 months later. He was still on the same medications and was cognitively intact and living independently. No seizure or psychosis has occurred in the interim.

Discussion

Hashimoto encephalopathy is a syndrome of variety of neuropsychiatric manifestations in the setting of elevated anti-thyroid antibodies in absence of any obvious etiology. The clinical manifestations can range from cognitive impairment, alteration of consciousness, myoclonus, seizures, chorea, myelopathy, stroke like symptoms and agitation, psychosis, hallucinations etc. Two different forms have been suggested although there is considerable overlap between the clinical features in these patient [6,7]. Autoimmune vasculitic encephalopathy characterized by recurrent, acute to subacute episodes of focal neurologic deficits with a variable degree of cognitive dysfunction and alteration of consciousness. Generalized non-vasculitic meningioencephalitis is characterized by diffuse, progressive pattern, characterized by slowly progressive cognitive impairment with dementia, confusion, hallucinations, with or without seizures. Our patient had symptoms suggestive of the generalized

form with confusion, hallucinations and seizures, though his seizures were complex partial in semiology and not generalized.

The critical test in establishing the diagnosis of Hashimoto encephalopathy is presence of elevated anti-thyroid peroxidase antibodies and/or anti-thyroglobulin antibodies in serum or cerebrospinal fluid and no evidence of bacterial or viral meningitis or encephalitis. Anti-thyroid peroxidase antibodies are present in almost 100% of patients[8] and anti-thyroglobulin antibodies are present in 70% of cases.[9] However, there is no clear relationship between the severity of neurologic symptoms and the type or concentration of antibodies. Thyroid hormone levels range anywhere between overt hypothyroidism to overt hyperthyroidism. The variability in the level of thyroxin may be related to the extent of stimulation of the thyroid gland and possible "burnout" of thyroid production. In this respect, the term "Hashimoto encephalopathy" is misleading as it could suggest primary thyroid hypofunction instead of overstimulation. Cerebrospinal fluid may show elevated protein concentration, lymphocytic pleocytosis, oligoclonal bands or may be totally normal. Our patient had normal cerebrospinal fluid analysis, with positive serum anti-thyroid peroxidase antibodies and anti-thyroglobulin antibodies in addition to elevated thyroid stimulating hormone and low free thyroxine levels consistent with hypothyroid state.

Magnetic resonance imaging of the brain is usually normal but may show signs of vasculitis or cerebral atrophy. Our patient had normal neuroimaging (Figure 1-3). EEG may show various degrees of slowing and/or triphasic waves.[10] Focal slowing, focal epileptiform discharges, periodic lateralized epileptiform discharges have been seen in isolated cases.[7,11,12] Our patient had severely abnormal electroencephalogram with diffuse polymorphic slowing and sharp wave activity suggesting epileptic encephalopathy.

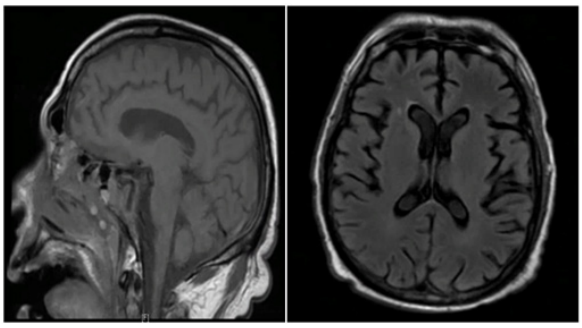

Figure 1 MRI brain on admission.

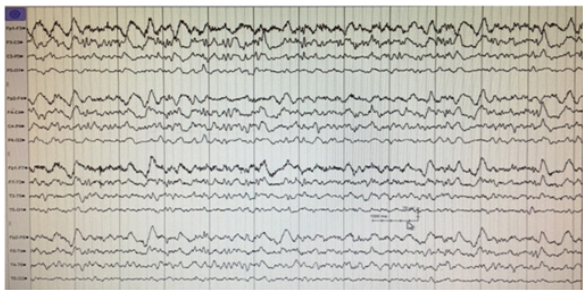

Figure 2 EEG on admission showing polymorphic disorganization and triphasic waves.

Hashimoto encephalopathy is usually treated with prolonged oral steroid therapy over 1-2 years. High dose i.v. steroids are used in the acute setting followed by administration of oral steroids over longer period of time. Patients are also given oral thyroid supplementation depending on their thyroid hormone levels. There have been a few cases of treatment with IVIG and / or plasmapheresis in which benefit was reported[13,14]. However, our patient improved without steroids and was spared the potential complications of long term steroid administration.

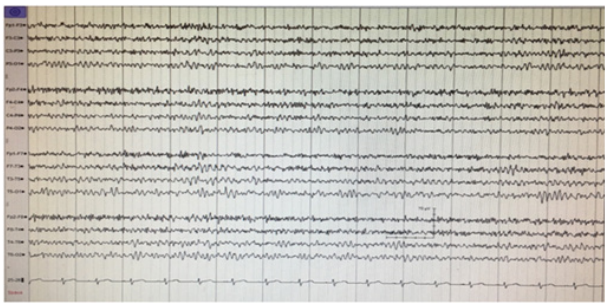

Figure 3 EEG on discharge at 3 weeks after admission.

Future Prospects

Our patient's recovery with thyroxine administration without any steroid or immunomodulating agents suggests that there might be a toxic etiology of Hashimoto encephalitis. There may also be metabolic components involved in the picture of Hashimoto encephalopathy. In few selected cases of Hashimoto encephalopathy with very low free thyroxine levels, early thyroxine therapy may be initiated with symptomatic management of seizures and psychosis, if patient shows early improvement, the use of steroids and their side effects may be avoided. In our patient, control of seizures with levetiracetam along with thyroxin supplementation provided effective recovery beginning within 2 days. Hence, we suggest the hypothesis that Hashimoto encephalopathy may have a toxic metabolic etiology besides an autoimmune component. Further investigation is needed.

Acknowledgments

None.

Conflicts of interest

None.

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