Hashimoto’s encephalopathy: Response to plasma exchange

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We report a case of 52-year-old female with steroid-unresponsive Hashimoto’s encephalopathy. She underwent plasma exchange that resulted in marked clinical improvement.

**Key Words:** Hashimoto’s encephalopathy, myoclonus, plasma exchange

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**Introduction**

Hashimoto’s encephalopathy is a rare disorder. It was first described by Brain et al in 1966. Eighty-five cases have been reported in the literature so far. The clinical picture is pleomorphic and cognitive impairment is a frequently reported symptom. There are two different clinical presentations. The vasculitic type is characterized by relapsing-remitting stroke-like episodes. The diffuse-progressive type shows insidious cognitive impairment, confusion, psychosis, somnolence and comatose. Usually, these patients respond to corticosteroids, however, various immunosuppressive treatments have been used in this condition, including azathioprine, cyclophosphamide, and intravenous immunoglobulin. Plasmapheresis has been used where the response to corticosteroids has been incomplete.

We report the case of a patient with slowly progressive cognitive impairment caused by Hashimoto’s encephalopathy, which showed minimal response to steroids but a rapid reversal of deteriorating neurological dysfunction was noted after the institution of plasma exchange.

**Case Report**

A 52-year-old female presented with decline in memory and tremors of the extremities for last 6 months. On examination, there were mild extrapyramidal features in the form of mask-like facies and rest tremors of both the upper limbs with cogwheel rigidity. Her serum TSH was found raised (18.59 microIU/ml) with normal serum T3 (95 ng/dl), and serum T4 levels (4.8 microgm/dl). She received thyroid hormone replacement without any clinical response.

Her CT scan-Head (Figure 1) and MRI-Brain (Figure 2) showed cerebral atrophy. The CSF examination was within normal limits. EEG showed bitemporal slowing without any epileptiform discharges.
Randomly selected sentences from the natural text:

“Most patients of Hashimoto’s encephalopathy have normal MRI-Brain. However, abnormalities in brain MRI have been described. Cerebral atrophy is the most common non-specific neuroimaging finding as seen in our patient too. Cerebrospinal fluid is abnormal in more than 80% of the patients; with high protein levels and mononuclear pleocytosis. The EEG is helpful in evaluating and following patients with steroid-responsive encephalopathy associated with autoimmune thyroiditis in reflecting the degree of central nervous system (CNS) involvement, in determining whether their condition is better or worse, and in ruling out other causes of encephalopathy. EEG findings include mild to severe generalized slowing, triphasic waves, epileptiform abnormalities, photomyogenic response, and photoparoxysmal response. Temporal spike discharges as seen in our patient have been well described in Hashimoto’s encephalopathy.

The pathogenesis of Hashimoto’s encephalopathy remains unclear. Several theories have been proposed, including a generalized abnormality of the immune system, cerebral vasculitis, recurrent denervation, or a toxic effect of thyrotropin-releasing hormone (TRH) on the CNS. An excessive, central release of TRH was held responsible for the epileptic seizures. An abnormality of thyroid function itself cannot explain this condition, as many patients described in the literature are euthyroid either at the time of presentation or relapse. In our patient also, although the patient had subclinical thyroid abnormality initially, there was no clinical response to thyroid hormone replacement. The patient continued to worsen when the biochemical hormone levels were normal at a later stage.

An autoimmune basis is suggested by the high concentrations of antithyroid antibodies and improvement with immunosuppressive therapy. The precise role of antithyroid antibodies is also unclear: if they are to be implicated as pathogenic, then it is surprising that more cases of encephalopathy are not seen in patients with Hashimoto’s thyroiditis.

It is possible that the antithyroid antibodies in Hashimoto’s encephalopathy are a surrogate marker for other, as yet unknown, antibodies that cross the blood-brain barrier and initiate an autoimmune encephalopathy. By contrast with the Creutzfeldt-Jakob disease, which leads to death within a few months, patients with Hashimoto’s encephalitis often recover quickly when treated adequately. Corticosteroid therapy is the first choice. The successful administration of other immunosuppressants (methotrexate, azathioprine, cyclophosphamide) has been reported. Plasmapheresis should be used whenever the patient is unresponsive or poorly responsive to corticosteroid treatment. Epileptic seizures and myoclonus usually respond well to anticonvulsant drugs. Even with consistent therapy, relapsing disease courses are not rare, but the patients altogether clearly benefit from therapy.

References


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