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# Steroid Responsive Encephalopathy Associated with Autoimmune Thyroiditis (Hashimoto Encephalopathy)

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**Abstract:** ***Steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT), also known as Hashimoto encephalopathy, is a rare and well recognised neurological complication of autoimmune thyroid disease and occurs independently of the thyroid status. Patients exhibit neurological symptoms in the presence of serologic evidence of autoimmune thyroid disease, usually, in the majority of cases, with a normal MRI scan. It is a rare condition with a few case reports in the medical literature. In this case report, we discuss a case having steroid responsive encephalopathy***.

**Keywords:** encephalopathy, steroid responsive, Hashimoto, Jharkhand

## 1. Introduction

Hashimoto's encephalopathy is a rare disorder that causes relapsing-remitting or progressive confusion, impaired consciousness, seizure, ataxia, psychosis and myoclonus. The term "Hashimoto encephalopathy" (HE) was used perhaps for the first time in 1991 by Shaw, who collected five cases with similar symptoms such as seizures, disorientation, frequent episodes of alternating hemiparesis, high protein levels in the cerebrospinal fluid and electrocardiographic abnormalities. It is thought to be immune mediated, though its exact pathophysiology remains speculative. In many patients, an increased level of thyroid antibodies has been found and in some patients, even such an occasional finding became a decisive input that led to a final diagnosis. We recently treated a patient with Hashimoto's encephalopathy who responded well to glucocorticoid therapy.

## 2. Case Report

A 27 year old Hindu male, Mr. Parmeshwar Prasad, was admitted in Medicine Department of RIMS, Ranchi for seizures, fever, & unconscious state for last 1 day. He was labourer by occupation with no prior history of such episode. He had no H/O trauma, DM, HTN, TB, seizure disorder, heart disease, or thyroid disorder. He was an occasional alcoholic with no significant drug history. Vitals were reported normal. His unconsciousness at admission & between seizures was reflective of NCSE or post-ictal state.

On CNS examination, pupils were reactive, plantar reflex was bilateral extensor and jerks were normal to brisk. There was no neck rigidity or Kernig's sign and Dolls eye movements were intact. Routine work up was done. Blood reports were mostly normal except for elevated liver enzymes & alkaline phosphatase.

**Table showing laboratory results of the patient**

| | |
|---|---|
| **CBC**<br>Hemoglobin/ TLC/ DLC | 10.6 gm%<br>7500 cells/$mm^3$<br>($N_{61}L_{34}M_3E_2B_0$) |
| **LFT**<br>AST/ ALT/ ALP | 344/ 144/ 493 IU/ml |
| **RFT**<br>B.urea/ S.creatinine | 21/ 0.9 mg/dl |
| **RBS/ELECTROLYTES**<br>$Na^+$/ $K^+$/ $Ca^{2+}$/ RBS | 140/ 3.9/ 8.9/ 90 mg/dl |
| **ANA** | Negative |
| **CSF**<br>Protein/ Glucose/ WBC/ Bacteria | 110/ 49/ 02/ Absent |
| **Thyroid Function**<br>fT3/ fT4/ TSH | 1.71/ 0.93/2.49 mIU/L |
| Optimal for Malaria/ Leptospiral Ab/ HIV & HbsAg/ Dengue profile/ Japanese Encephalitis/ HSV1&2 PCR/ | Negative |

MRI Brain & EEG were done on subsequent days revealing normal study and generalised high amplitude, frontally dominant slow waves suggestive of encephalopathy respectively.

Even with empirical antibiotic & antiepileptic coverage, patient's condition did not improve in first 7 days. IV benzodiazepines (lorazepam) were used for GTCS in between, ruling out NCSE as post-ictal unconsciousness persisted. Fever used to be post-ictal only & not continuous. Acyclovir was also started in view of possible encephalitis.

Meanwhile on 8$^{th}$ post admission day, Inj. dexamethasone was added to combat possible CIRCI (Critical Illness Related Corticosteroid Insufficiency), we were astonished to see improvement on GCS next day as patient started to produce incomprehensible sounds the next day. With 3 doses of steroids, he became conscious & seizure free. This led us to check for Anti-TPO Antibodies in serum which was 265.9IU/ ml (<34 IU/ml). Acyclovir was subsequently stopped, and patient was discharged on tapering steroids & antiepileptic drugs. He is on constant follow up in order to observe any recurrence.


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## 3. Discussion

The clinical, laboratory, and radiologic findings associated with SREAT are more varied than previously reported. Misdiagnosis at presentation is common. Until the pathophysiologic mechanism of this and other autoimmune encephalopathies is better characterized, we believe that descriptive terms that reflect an association rather than causation are most appropriate for this syndrome.
The diagnosis of HE should be considered in patients presenting with the characteristic neuropsychiatric manifestations excluding other causes of encephalopathy. They should have-

- Presence of high levels of antithyroid antibodies in serum or CSF
- No alteration in the CSF and/or imaging tests compatible with infectious, vascular, or neoplastic etiology and
- A good response to immunosuppressive therapy.

The differential diagnosis for encephalopathy is wide, but the clinical features and findings on blood, CSF, electroencephalography (EEG) and neuroimaging studies often lead to an accurate diagnosis. Once infectious causes are excluded, an autoimmune or inflammatory process may be suspected on the basis of inflammatory and autoimmune markers in the serum and CSF and meningeal and parenchymal abnormalities on magnetic resonance imaging (MRI) of the brain.

Autoimmune encephalopathy may take many forms, including paraneoplastic or idiopathic limbic encephalitis that is defined by characteristic serologic and neuroimaging abnormalities.[1,2] Idiopathic autoimmune encephalopathy is also often defined on the basis of a clinical response to steroids. Steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT), often referred to as *Hashimoto encephalopathy*, was initially described by Brain et al.[3] Although high serum levels of thyroperoxidase (TPO) antibody, formerly known as antimicrosomal antibody, are often found in patients with SREAT, this organ-specific autoantibody is a serologic marker for the common condition of autoimmune (Hashimoto) thyroiditis and a common marker of autoimmunity in most autoimmune neurologic disorders.[4,5] A direct causal relationship between thyroid antibodies and encephalopathy is unlikely.[6] Nevertheless, many uncertainties regarding the condition persist, including the spectrum of clinical findings, the associated laboratory and radiologic findings, the clinical significance of the quantitative level of TPO antibody, the criteria required for diagnosis, the appropriate terms for the condition, and the typical outcome of steroid treatment. The literature thus far has not provided sufficient data on these challenging issues to aid the diagnosis of this disorder and guide its management.

The disease is responsive to immunosuppressive therapy and intravenous (IV) methylprednisolone (500-1000mg/day) for 3-5days, followed by an oral dose of prednisone (1-2mg/kg/day), and followed by gradual tapering based on the clinical response being the commonly followed protocol. Around 25% of patients may not respond to steroids, in which azathioprine, IV immunoglobulins or plasmapheresis can be used with good results.

Earlier reports suggested that 90% of cases stay in remission after discontinuation of treatment; however, this is at odds with more recent studies which suggest that relapse commonly occurs after initial high dose steroid treatment[7,8]. Left untreated, this condition can result in coma and death.
This case report emphasizes that the incidence of Hashimoto Encephalopathy is probably underestimated because of the low overall awareness about the disease. HE may be found in cases of unexplained encephalopathy, particularly together with the presence of high thyroid antibody levels, especially against thyroperoxidase (Anti TPO). Because of the autoimmune origin of the disease, corticosteroid treatment usually provides a dramatic recovery.

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