

Case report

Intra venous immunoglobulin responsive relapsing Hashimoto's encephalopathy

R A D R Lasika¹, S Senanayake², W K M G Amarawardena³

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Introduction

Hashimoto's encephalopathy is an immune mediated disorder, which presents with a broad range of neurological symptoms. It is characterized by associated elevated anti thyroid antibody levels and responsiveness to steroids. Only a few cases have been published in the literature of patients with poor response to steroid therapy. We report a case of a young girl who presented with a relapse of Hashimoto's encephalopathy while being on a steroids maintenance dose, responding to IV immunoglobulin.

Case report

A 19-year-old previously healthy girl was brought to hospital in December 2015 with agitated behavior, irritability and insomnia. Patient has initially complained that she was unable to concentrate on studies because of some suicidal ideations and visual hallucinations. She had no fever or history of drug intoxication.

She was afebrile and pulse rate was 104 beats per minute. Neurological examination revealed occasional myoclonic jerks of right upper limb. There was normal muscle power and tone of limbs with generalized brisk reflexes. But plantar response was flexor. Cranial nerve examination was normal including fundal examination. There were no signs of meningism. Cognitive status could not be assessed since she was agitated and non-cooperative.

Basic investigations including full blood count, inflammatory markers, renal function tests, liver function tests and electrolytes were normal. EEG revealed generalized slowing with right focal spikes. Generalized cerebral atrophy was noted on MRI (Figure 1).

During hospital stay, she developed generalized tonic clonic seizures proceeding to status epilepticus requiring intubation and ventilation. Her cerebral fluid analysis was normal. Japanese encephalitis IgM antibodies, HSV 1 & 2 IgM antibodies, measles IgM and IgG antibodies were negative in CSF. Thick and thin blood smears did not show malarial parasites. Serum NMDA

antibodies, anti-nuclear antibodies, anti ds-DNA antibodies were negative. Serum ceruloplasmin and serum copper were normal. The thyroid function tests revealed normal T3/T4 and thyroid stimulation hormone levels. Anti-thyroid peroxidase antibody (TPO-Ab) level was 465 IU/ml (< 30) and anti-thyroglobulin antibody (TGA) was 69 IU/ml (< 40). Further evaluation was done with FNAC of thyroid gland, which revealed diffuse lymphocytic infiltration suggestive of chronic autoimmune thyroiditis (Figure 2). Depending on the clinical presentation and investigation findings, the diagnosis of Hashimoto's encephalopathy was made. Patient was treated with intravenous methyl prednisolone 1g daily followed by oral prednisolone 1mg/kg/day with combination of antiepileptic drugs for seizures. After two months of treatment, with the clinical evidence of complete recovery, tapering off of steroids was started to a maintenance dose. Anti-thyroid antibody levels were found to be normal when starting tapering off steroids.

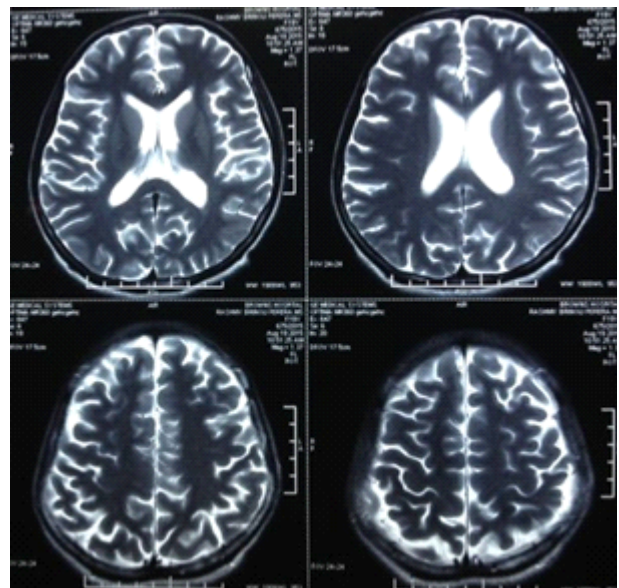


Figure 1. MRI brain showing diffuse cerebral atrophy.

¹Senior Registrar in Neurology, ²Consultant in Neurology, ³Senior Registrar in Endocrinology, National Hospital of Sri Lanka.

Correspondence: RADR Lasika E-mail: rukshanilasika@yahoo.com

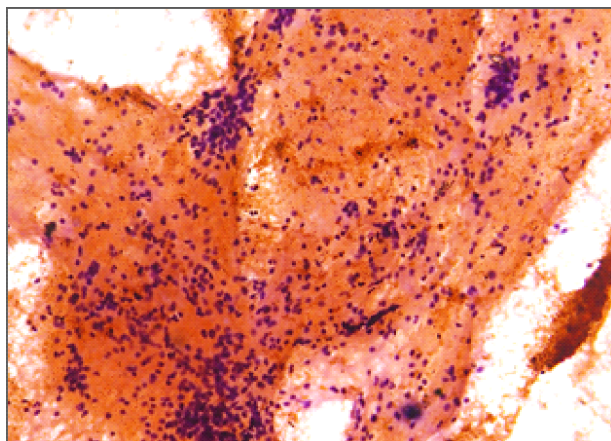


Figure 2. FNAC of the thyroid showing diffuse lymphocytic infiltration

While being on maintenance dose, in May 2016, the patient was readmitted with recurrence of seizures associated with altered behaviour manifested as visual hallucination and aggressiveness. Examination didn't reveal any focal neurological signs or features of meningism, but there was generalized hyperreflexia. Repeat anti thyroid antibody levels were found to be elevated suggesting relapse of Hashimoto's encephalopathy. Rest of the investigations were normal. The treatment with methyl prednisolone 1 g/d was started. Although she responded to steroids in her first presentation, this time she remained encephalopathic despite treatment. A week later, she was given IVIG 0.4 g/kg for 5 days. Patient showed a dramatic improvement with IVIG with return of normal mental status within 72 hours. She returned to her cognitive baseline and was discharged home.

Discussion

Hashimoto's encephalopathy is an immune mediated disorder, associated with Hashimoto thyroiditis. Pathophysiology of the disease is unknown. Goitre and hypothyroidism can be associated with the disorder, but the majority of patients are euthyroid¹. Patients usually present with subacute onset of confusion, altered conscious level and focal or generalized seizures². Other clinical presentations include myoclonus, stroke-like episodes, psychosis, visual hallucinations, generalized hyperreflexia and pyramidal tract signs³. The long-term course of illness may be self-limited, relapsing-remitting, or progressive^{4,5}. Because of its dramatic response to treatment with corticosteroids, it is also described as a corticosteroid-responsive encephalopathy associated with thyroiditis⁶. Our patient showed a prompt response to steroids in her first presentation. But she developed a relapse while being maintained on low dose steroids.

The relapse was resistant to steroids. Interestingly, she recovered rapidly with IV immunoglobulin. Relapsing and remitting Hashimoto's encephalopathy has been documented. According to one of the large systematic reviews of the literature, out of 82 follow up patients with Hashimoto encephalopathy, in 48 cases (58.5%), there was no recurrence of disease during follow-up, and in 22 cases (26.8%), relapse was associated with the withdrawal of corticosteroid; 12 patients (14.7%) relapsed, requiring further treatment, and 4 patients (4.8%) died⁷. There are few cases reported in the literature of patients with steroid resistant Hashimoto's encephalopathy but responsive to IVIG^{1,8}. This is the first case reported from Sri Lanka of this rare disease entity.

In conclusion, although by definition Hashimoto's encephalopathy was described as a corticosteroid responsive disease, cases with poor response to steroids exist. IV immunoglobulin can be considered as a convenient and effective alternative in treatment for such cases.

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