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Research Article

HASHIMOTO'S ENCEPHALOPATHY IN A PREGNANT FEMALE: A DIAGNOSIS IN DISGUISE

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A B S T R A C T

While Hashimoto's encephalopathy (HE) is quite rare. It is also likely that there are many more undiagnosed sufferers. Because it is little known and its symptoms are primarily neurological. It is easy to misdiagnose or overlook and the symptoms frequently lead to mistaken neurological diagnoses. We report a case of a 21 year old female diagnosed with Hashimoto's encephalopathy (HE) at six months of gestation. She was successfully treated. Hashimoto Encephalopathy is a neuropsychiatric disorder of exclusion. As a good response can be obtained with corticosteroid therapy, early diagnosis and treatment is very beneficial for patients. To the best of our knowledge, there is no such report in the literature.

INTRODUCTION

Hashimoto's Encephalopathy(HE) is a very rare condition characterized by various clinical features consisting of psychiatric manifestations, seizures and focal neurologic deficits.Hashimoto encephalopathy ,a rare autoimmune disease with unknown origin, is referred to as non vasculitic autoimmune ncephalopathy/meningoencephalitis,hashimoto's thyroiditis or steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT).[1] The condition is more frequently found in females than in males, with a ratio of 5:1. It is also possible that there are many more undiagnosed cases. Because it is little known and its symptoms are primarily neurological, it is easy to misdiagnose or overlook and the symptoms frequently lead to mistaken neurological diagnoses.[2] Therefore, for all patients with unexplained acute or sub acute encephalopathy, or atypical psychiatric manifestations, especially patients who have autoimmune thyroid disease, Hashimoto's encephalopathy must be included in the differential diagnosis.[3] If treatment is delayed due to misdiagnosis, it often becomes fatal and therefore physicians should have high degree of suspicion and awareness for this condition. Here we present a case highlighting that of a 21 year old previously healthy woman diagnosed with Hashimoto's encephalopathy at six months of gestation.

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Case presentation

A 21 year old female who was carrying six months first pregnancy was admitted in our department with a history of hiccupping for 1 months, urinary retention for 3 days and gait instability for 3 weeks and weakness of limbs accompanied with dyspnoea for 2 days. Furthermore her younger brother noticed self talking behaviour, paranoid ideas and intermittent crying. She had earlier history of hiccupping, nausea and vomiting which were ameliorated with symptomatic treatment. The patient had no prior personal or family psychiatric history. She neither had any history of fetal wastage nor any past history of medical disorder. Her husband was unemployed, alcohol dependent and was hardly present at home. One day before hospitalization, the patient experienced limb weakness, dyspnoea, cough and hoarseness and discontinuous unconsciousness. The physical examination was otherwise normal except fluctuating psychomotor agitation, emotional liability, impulsivity and anxiety. The patient had incoherent speech, persecutory delusions, auditory hallucinations and was disoriented in time and space. She was not known to be taking any medications nor did she suffer from substance abuse. Neurological examination revealed dysarthria, nystagmus, weakened extremities 'muscle strength (graded 3), hyperreflexia in all limbs, positive Babinski sign on both side. Pulse 110/min regular and BP 120/78mm Hg. As a consequence, oral resperidone 2 mg daily and IV lorazepam were introduced. After 24 hours later, symptoms

worsened and she presented with permanent auditory hallucinations, as well as aggressive behaviour, stereotypic gestures and psychomotor instability.

Electroencephalography (EEG) showed widespread slowing of background activity without sharp waves suggestive of encephalopathy. Brain MRI was normal. The result of CSF examination showed the concentration of protein was 0.4g/l and cell count was 5×10^6. The concentration of glucose, sodium and chloride were normal. CSF cultures were negative. Thyroid function tests were performed (FT3 2.37pmol/L, FT4 8.04pmol/L, TSH 18.39µIU/ml, and anti thyroid peroxidise antibody (TPO) ab 686.9U/mL. The ultrasound imaging result of thyroid revealed heterogeneous gland without any sign of inflammation. No other abnormality was found from further testing including blood test for regular autoimmune disorders, serological tests for herpes simplex virus, cytomegalovirus, viral hepatitis B and C, human immunodeficiency virus, syphilis as well as blood sampling for vitamin B12, and Ceruloplasmin.

The patient was diagnosed with hashimoto encephalopathy given the association of high concentrations of antithyroid antibodies and the presence of an otherwise unexplained neuropsychiatric condition. Although medication with resperidone 2mg daily was initiated and eventually lorazepam was on going, no clinical amelioration was observed. She was treated using intravenous immunoglobulin's IV immunoglobulins (0.4g/kg/d) for 5 days. Simultaneously, she was treated using methylprednisolone (500 mg/d) for 5 days. It was quickly followed by a dramatic improvement of the clinical picture. Within 72 hours, the patient was again oriented in time and space, auditory hallucinations, as well as neurological symptoms, disappeared. A follow up EEG returned normal. Although patient had a fetal wastage .The patient continued methylprednisolone at a dose of 16mg/d after discharged and maintained treatment with gradually reduced dose and stopped within a year. There was no recurrence of symptoms.

DISCUSSIONS

Hashimoto's encephalopathy is a rare autoimmune disease associated with a prevalence of 2.1/100,000 with female to male ratio of 5:1 and the mean age of onset is between 45 and 55 years.[1] It is relapsing remitting and sometimes progressive encephalopathy occurring in association with Hashimoto's thyroiditis.[2]

The pathogenesis of Hashimoto's encephalopathy is still unknown. There is no evidence that the anti TPO antibody directly causes encephalopathy, but other auto antibodies that associated with autoimmune thyroid diseases might induce encephalopathy.[3] Several mechanisms such as autoimmune vasculitis, autoantibodies against brain thyroid antigens, encephalomyelitis associated demyelination, global cerebral hypoperfusion, a direct toxic effect of thyrotropin-releasing hormone, and neuronal dysfunction due to brain oedema have been proposed for Hashimoto's encephalopathy.[4]

Hashimoto's encephalopathy is suspected whenever symptoms of acute or sub acute encephalopathy are associated with high serum levels of antithyroid antibodies. Because it lacks specific markers and is a clinically heterogeneous syndrome, Hashimoto's encephalopathy remains an elusive nosologic entity.[5] Clinical manifestations include confusion, coma, stroke like episodes, seizures, psychosis, dementia, myoclonus and myelopathy. As in our case, the pregnant female came with intractable hiccoughing which gradually progressed to stumbling and urinary incontinence along with predominance of psychiatric symptoms and the presence of multiple risk factors. Our initial diagnosis was psychosis. However treatment resistance made us suspect an organic aetiology. Hashimoto's encephalopathy was very probable in our case after giving high titres of antithyroid antibodies and rapid normalization of her clinical symptoms and EEG once treatment was introduced. It must be noted that neither abnormal brain imaging nor the presence of antithyroid antibodies in the CSF are necessarily required for Hashimoto's encephalopathy diagnosis. Additionally, the thyroid status may vary from normal to pathological among patient with Hashimoto's encephalopathy. Our patient had a fetal wastage at six months which could be due to thyroid auto antibody positive and such women are likely to have increase miscarriage rate which could lead to fourfold increase in the incidence of placental abruption.[6]A number of aetiologies have been hypothesized as the cause of relationship between spontaneous termination of pregnancy and autoimmune thyroid antibodies .These include (1) the existence of a subtle degree of hypothyroidism (2) thyroid antibodies reflecting an autoimmune imbalance in the pregnant female, (3) thyroid auto antibodies acting directly on the placenta. One study showed a marked reduction in miscarriages when thyroid antibody women were treated.[7]

Due to its variety of clinical symptomlogy, this condition may be difficult to diagnose at initial presentation and this may mimic stroke, rapidly progressive dementia, CJD, paraneoplastic or viral encephalitis. This poses a diagnostic challenge and requires extensive workup to rule out toxic, metabolic, vascular and infective causes. It is often called investigation negative encephalopathy.[2]

EEG studies, while always abnormal (98%) are usually nondiagnostic.The most common findings are diffuse or generalised slowing or frontal intermittent rhythmic delta activity. Prominent triphasic waves, focal slowing, epileptiform abnormalities, photoparoxysmal and photo myoclonic responses may also be seen. These finding are nonspecific and may be seen in toxic, metabolic and post anoxic encephalopathy.[6,7]

Most patients responds to high dose steroid therapy and therefore it is called SREAT. Initial treatment is with oral prednisolone or high dose IV solumedrol for 7 days along with thyroid hormone treatment for concurrent thyroid disorder. Alternative therapies including immunosuppressants may be effective along with immunomodulation with periodic IV immunoglobulins and plasma exchange.[8] None of the treatment has been studied in controlled trials and therefore there are no clear guidelines for their use.

CONCLUSIONS

As psychiatric symptoms may be prominent in encephalopathy, they could potentially be misleading and point to psychiatric disorder. However, an organic aetiology should always be considered, especially in absence of any mental disorder history. Hashimoto's encephalopathy is a difficult diagnosis, especially when thyroid hormone status is normal. This case is unusual as it the first report of a patient

with first time pregnancy presenting with Hashimoto's encephalopathy. The present case also highlight the importance of early establishment of multidisciplinary diagnosis approach with psychiatrist, obstetrician and neurologist, prompt initiation of steroid therapy in successful management of Hashimoto's encephalopathy.

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