Hashimoto's Encephalopathy Presenting with Status Epilepticus and Mental Disorder as the Main Manifestations

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ABSTRACT

Hashimoto's encephalopathy (HE) is an uncommon disease. It is easily misdiagnosed as many clinicians have an insufficient understanding of it. The clinical presentation, auxiliary examination, diagnosis, and treatment of a patient with HE were retrospectively analysed, and the relevant literature was reviewed and summarised. The patient, a middle-aged woman, presented with status epilepticus, persecutive delusions, elevated serum thyroid antibodies, and a negative autoantibody profile test for autoimmune encephalitis and responded well to steroid pulse therapy and plasma exchange. Elevated serum thyroid antibodies are the most important investigations for the diagnosis of HE. Most patients have a good prognosis but early investigation of thyroid function and antibodies is imperative for an early and accurate diagnosis. An early treatment can greatly improve patient prognosis.

Key Words: *Status epilepticus, Hashimoto's encephalopathy, Autoimmune disease, Mental disorder.*

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INTRODUCTION

Hashimoto's encephalopathy (HE) is an autoimmune encephalopathy associated with thyroid antibodies, which lacks specific clinical manifestations.[1] The disease often has a relapsing and remitting course, and its clinical manifestations are complex and diverse including neuropsychiatric symptoms, cognitive impairment, epilepsy, stroke-like episodes, and ataxia.[2] Since there is no unified standard for clinical diagnosis, it is easily misdiagnosed. In this case, the patient had an acute onset of status epilepticus and was promptly diagnosed with HE after physical examination and relevant investigations. She responded well to appropriate treatment.

CASE REPORT

A 32-year female presented with recurrent spasms of the extremities without obvious triggers with intermittent periods of persistent unconsciousness. The patient was admitted to the emergency department, and a physical examination revealed confusion, cyanosis of the lips, staring at the left side, disappearance of bilateral pupillary light reflexes, and high tension in the limb muscles.

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The convulsions could not be controlled with diazepam and phenobarbital injection, so the patient was transferred to the intensive care unit after tracheal intubation. During this time, the patient had delusions of victimisation, yelled and screamed at night, and claimed that someone was trying to harm her.

The laboratory tests revealed decreased thyroid function, a serum thyroglobulin antibody (TG Ab) concentration of 40.31 IU/ml (Normal, <4.11 IU/ml), and an anti-thyroid peroxidase antibody (TPO Ab) concentration of 44.1 IU/ml (Normal, <5.61 IU/ml). The patient was also negative for 12 autoimmune encephalitis autoantibodies in the serum and cerebrospinal fluid (CSF). No abnormalities were found in the acid-fast staining of CSF smear, cryptococcus, syphilis, and bacterial culture. However, there were elevated white blood cells in the CSF.

HE was diagnosed after the exclusion of common causes such as infection, poisoning, metabolism, vascular disease, a tumour, *etc.* and combined with thyroid function examination and head MRI (Figure 1). After mechanical ventilation, midazolam and propofol sedation, sodium valproate, acyclovir, high--dose steroid pulse therapy, and plasma exchange, the patient's seizures and psychotic symptoms were controlled. The thyroid antibody titer returned to normal and there were no symptoms at the three-month follow-up.

DISCUSSION

HE also known as autoimmune alpha-related steroid-responsive encephalopathy, is a relatively rare disease.[3] Most patients are middle-aged women with thyroid dysfunction. The increase of

TPO Abs has a certain specificity, and the treatment effect is significant after the use of hormones.[4] The pathogenesis of HE is still unclear but it is generally believed to be related to the increase of TPO Ab and TG Ab levels.[3] The clinical manifestations of HE are diverse with seizures and mental disorders being the most common. There is no unified standard for HE diagnosis and it should be differentiated from viral encephalitis, autoimmune encephalitis, a tumour, poisoning and stroke with epileptic seizures.[5]

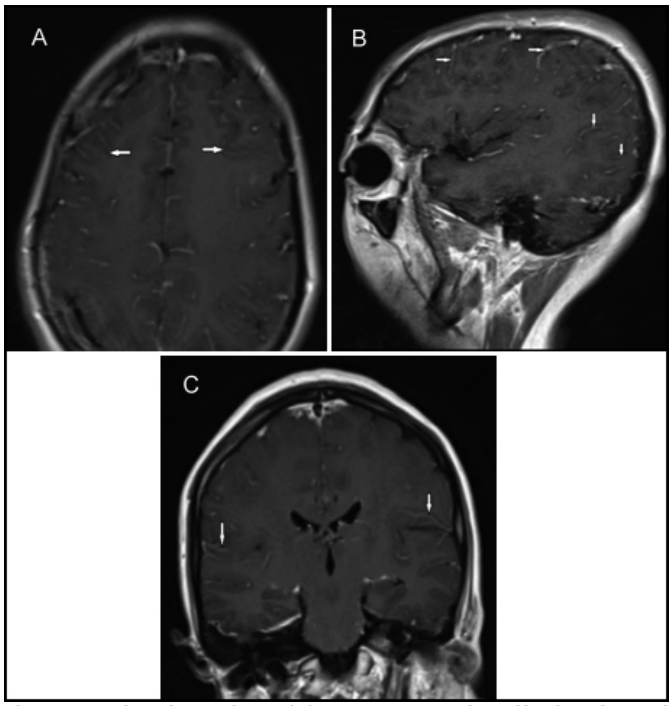

Figure 1: Brain enhanced MRI. (A) Transverse section of brain enhanced MRI showing mild leptomeningeal enhancement (arrows); (B) Sagital section of brain enhanced MRI showing diffuse leptomeningeal enhancement (arrows); (C) Coronal section of brain enhanced MRI showing bilateral leptomeningeals fine linear enhancement (arrows).

In this case, the main manifestations were status epilepticus and mental and behavioural abnormalities.[6,7] There were many slow wave discharges in the electroencephalogram and bilateral leptomeninges showed high signal intensity on DWIon MRI of the brain. The patient's serum TG Ab and TPO Ab levels were significantly increased but her symptoms were relieved after corticosteroid and plasmapheresis treatments, which supported the HE diagnosis.[4,8,9]

In summary, HE is rare in clinical practice and may be misdiagnosed due to atypical clinical manifestations and insufficient understanding of the disease. Conventional treatment is ineffective for any acute or chronic encephalopathy with unknown causes. If the above causes are excluded, the possibility of this disease should be considered, and thyroid functions and related antibodies should be examined as soon as possible. Regardless of whether the patient has a history of thyroid disease, the diagnosis can be made when the patient is sensitive to hormone therapy. Most patients have a good prognosis but it should be recognised and treated as soon as possible to reduce nerve damage and improve patient prognosis.

COMPETING INTEREST:

The authors declared no competing interest.

PATIENT'S CONSENT:

Patient's explicit consent has been obtained to publish the case.

AUTHOR'S CONTRIBUTION:

LP: Responsible for collecting patient data, writing, and final review of the manuscript.

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