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A Rare Case Of Hashimoto's Encephalopathy Responding To Plasma Exchange

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ABSTRACT

Hashimoto's encephalopathy is one of the rare causes of encephalopathy of unknown etiology. It manifests with neuropsychiatric symptoms. The estimated prevalence is 2/100,000[1]. Hashimoto's encephalopathy is characterized by elevated antithyroid antibodies in absence of infections, tumors or stroke of central nervous system[2-4]. The disease manifests with wide array of clinical features which can be cognitive fluctuations, ataxia, myoclonus, seizures[5-10]. Psychitric manifestations like depression, mania, psychosis, and hallucinations have also been reported[11-13]. Anti thyroid antibodies are considered to be the biomarkers of the disorder but the titers of the antibodies do not correlate with the severity of the disease presentation [14-15].

Keywords: Hashimoto's encephalopathy, Hashimoto's encephalopathy

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INTRODUCTION

Steroids remain the mainstay and the first line treatment for this disease and rapid clinical improvement is seen in majority of the patients[16]. But a subset of patients does not improve with steroids and need other modalities like plasma exchange. It is postulated theoretically that plasma exchange will result in removal of the bad antibodies anti-TPO and would eventually result in improvement of the symptoms and disease activity. In this case, we report the practical application of plasma exchange in Hashimoto's encephalopathy in Indian scenario and Indian patient. In 2016, the American Society of Apheresis (ASFA) Guidelines recognized therapeutic plasma exchange as adjunct therapy (category II indication) in the treatment of Hashimoto's encephalopathy[17].

CASE REPORT:

A 60 year old female without a past medical history of any chronic medical illness presented to us with a complaint of changes in mental status since 2 days. As per the history obtained from the patient's son, the patient was conscious and oriented to time, place and person before 2 days. There was a gradual decline in the mental status of the patient over 2 days. The patient had global aphasia and delirium at the time of presentation to the hospital. She also had 1 episode of GTCS prior to the admission to the hospital.

The patient lived with her son and her son denied any recent history of fever, vomiting, history of trauma to head or ingestion of any substance. On examining the patient, the patient was vitally stable and on CNS examination, the patient was in delirium. MMSE could not be calculated as the patient was in delirium.

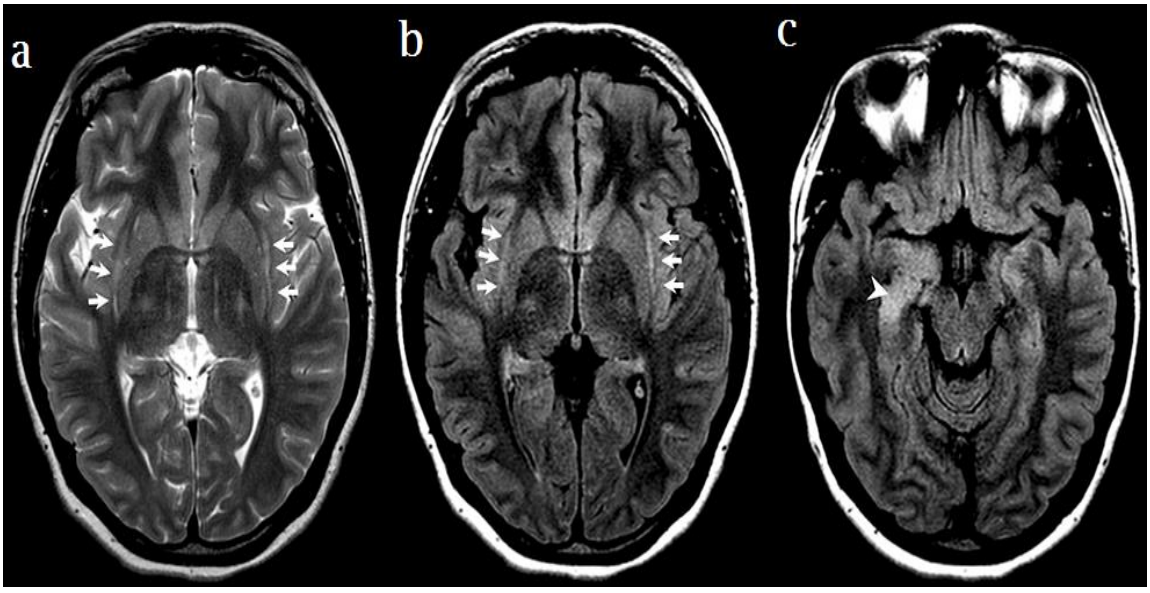
Muscle tone was normal in all the group of muscles in upper and lower limb. Muscle reflexes were +1 in all joints of 4 limbs. Muscle power could not be elicited due to the inability of patient to follow verbal commands. There was no neck rigidity and Kernig's and Brudzinski's sign were negative.

Urgent MRI Brain with contrast was done which showed Axial T2-weighted (A) and FLAIR (B, C) images demonstrate bilateral claustral hyper intensities and asymmetric hyper intensity of the right mesial temporal lobe suggesting a diagnosis of Hashimoto's encephalopathy.

CSF routine and micro was sent which showed 5 cells with protein and glucose in normal range. A diagnosis of autoimmune encephalitis was thought and CSF for autoimmune and paraneoplastic panel were sent. The patient had one episode of GTCS after hospital admission and was started on anti convulsants. CSF was negative for NMDA, AMPA-GluR1, AMPA-GluR2, GABA-B, LGI, CASPR2 antibodies by cell based essay. CSF was negative for antibodies of paraneoplastic panel of Amphiphysin, CV-2, PNMA2, Ri, Yo, Hu by immunoblot method. CSF was negative for HSV-1 HSV-2 IgG and IgM. Urine for

toxicology was also done which was negative. Thyroid function test was normal. Anti-TPO antibodies were sent which was 297.90 IU/ml and Anti Thyroglobulin antibody was sent which was 921.97 IU/ml. Thyroid function test was done twice at a gap of 7 days and TSH, T3 and T4 were normal in both the results. Serology for HIV, Syphilis (VDRL), Lyme disease was negative. ANA, RF, ACE enzyme levels (marker for Neurosarcoid), serum Calcium levels were normal. A diagnosis of Hashimoto's encephalitis with euthyroid state was made and the patient was started on pulse Methyl Prednisolone at 1gm/day for 5 days. There was subtle improvement in the patient's sensorium. The patient was drowsy but could be aroused easily and would follow verbal commands occasionally. The patient did not have any more episodes of convulsion. The patient was

There was no further improvement in the patient's sensorium after 2 days. It was decided to start Plasma exchange. Plasma exchange was started at 250ml/day. A total of 5 cycles of plasma exchange were done. After 3 cycles of plasma exchange, the patient's mental status improved and the patient became conscious to time and place but not person. After 5 cycles, the patient was conscious and oriented to time, place and person. The patient would follow all the verbal commands. MMSE was calculated which was 23/30. Muscle power was 5/5 in all major muscle groups of 4 limbs. Deep tendon reflexes were +2 in upper and lower limbs. Supportive treatment was continued and the patient was subsequently discharged. Anti-TPO levels at the time of discharge was 269.78 IU/ml. The patient further improved and at the time of discharge the patient's MMSE was 28/30.



Axial T2-weighted (A) and FLAIR (B, C) images demonstrate bilateral claustral hyperintensities and asymmetric hyperintensity of the right mesial temporal lobe.

DISCUSSION:

This case describes a patient who presented with delirium, acute seizures, aphasia and neuropsychiatric symptoms. Absence of concurrent infections, tumors or stroke along side

MRI findings of encephalitis and raised anti –TPO antibodies prompted a working diagnosis of Hashimoto's encephopathy. There was a modest improvement with steroids but the improvement remained static after a few days. Hence, it was decided to start her on therapeutic plasma exchange. Her clinical status improves significantly after 5 cycles of plasma exchange.

Hashimoto's encephalopathy is a rare disease and is considered to be a diagnosis of exclusion. According to a publication in 2004, it is estimated that the prevalence of the disease is 2 in 100,000(1). The syndrome was first described in 1966 when a patient previously diagnosed with Hashimoto's thyroiditis developed focal neurologic deficits and went into coma[18]. Hashimoto's encephalopathy is defined as documented evidence of encephalopathy with elevated antithyroid antibodies in absence of an alternate diagnosis [19-21]. Epidemiologically, it is more common in females as compared to males (4:1) and the mean age of onset is about 40-50 years of age [20]. HE is labelled by many physicians as "steroid responsive encephalopathy" as it is commonly responsive to the treatment with iv steroids[22]. There is a wide spectrum of the presentation of the disease but typically there are 2 distinct presentations.

REFERENCES:

1. Ferracci F, Bertiato G. Hashimoto's encephalopathy: epidemiologic data and pathogenetic considerations. J Neurol Sci 2004; 217:165.
2. de Holanda NC, de Lima DD. Hashimoto's Encephalopathy: systematic review of the literature and an additional case. J Neuropsych Clin Neurosci 2011;23:384–390.
3. Chong JY, Rowland LP, Utiger RD. Hashimoto encephalopathy: syndrome or myth? Arch Neurol 2003;60:164–171.
4. Marshall GA, Doyle JJ. Long term treatment of Hashimoto's encephalopathy. J Neuropsychiatry Clin Neurosci 2006;18:14–20.
5. Matsunaga A, Ikawa M, Fujii A, Nakamoto Y, Kuriyama M, Yoneda M. Hashimoto's encephalopathy as a treatable adult onset cerebellar ataxia mimicking spinocerebellar degeneration. Eur Neurol 2012;69:14–20.
6. Nakagawa H, Yoneda M, Fujii A, Kinomoto K, Kuriyama M. Hashimoto's encephalopathy presenting with progressive cerebellar ataxia. J Neurol Neurosurg Psychiatry 2007;78:196–197.
7. Nakagawa H, Yoneda M, Fujii A, Kinomoto K, Kuriyama M. Hashimoto encephalopathy presenting with progressive cerebellar ataxia. BMJ Case Rep 2009.
8. Tang Y, Chu C, Lin MT, Wei G, Zhang X, Da Y, Huang H, Jia J. Hashimoto's encephalopathy mimicking spinocerebellar ataxia. J Neurol 2011;258:1705–1707.
9. Arya R, Anand V, Chansoria M. Hashimoto encephalopathy presenting as progressive myoclonus epilepsy syndrome. Eur J Paediatr Neurol 2013;17:102–104.

10. McGinley J, McCabe DJ, Fraser A, Casey E, Ryan T, Murphy R. Hashimoto's encephalopathy: an unusual cause of status epilepticus. Ir Med J 2000;93:118.
11. Gomez-Bernal GJ, Reboreda A, Romero F, Bernal MM, Gomez F. A case of Hashimoto's encephalopathy manifesting as psychosis. Prim Care Companion J Clin Psychiatry 2007;9:318–319.
12. Wilcox RA, To T, Koukourou A, Frasca J. Hashimoto's encephalopathy masquerading as acute psychosis. J Clin Neurosci 2008; 15:1301–1304.
13. Mahmud FH, Lteif AN, Renaud DL, Reed AM, Brands CK. Steroid- responsive encephalopathy associated with Hashimoto's thyroiditis in an adolescent with chronic hallucinations and depression: case report and review. Pediatrics 2003; 112:686–690.
14. Chong JY, Rowland LP, Utiger RD. Hashimoto encephalopathy: syndrome or myth? Arch Neurol. 2003;60(2):164–171.
15. Olmez I, Moses H, Sriram S, Kirshner H, Lagrange AH, Pawate S. Diagnostic and therapeutic aspects of Hashimoto's encephalopathy. J Neurol Sci. 2013;331(1–2):67–71.
16. Mocellin R, Walterfang M, Velakoulis D. Hashimoto's encephalopathy: epidemiology, pathogenesis and management. CNS Drugs 2007;21:799–811.
17. Schwartz J, Padmanabhan A, Aqui N, et al. Guidelines on the use of therapeutic apheresis in clinical practice-evidence-based approach from the Writing Committee of the American Society for Apheresis: The Seventh Special Issue. J Clin Apher. 2016;31(3):149–162.
18. Brain L, Jellinek EH, Ball K. Hashimoto's disease and encephalopathy. Lancet 1966;2:512–514.
19. De Holanda NC, de Lima DD. Hashimoto's Encephalopathy: systematic review of the literature and an additional case. J Neuropsych Clin Neurosci 2011;23:384–390.
20. Chong JY, Rowland LP, Utiger RD. Hashimoto encephalopathy: syndrome or myth? Arch Neurol 2003;60:164–171.
21. Marshall GA, Doyle JJ. Long term treatment of Hashimoto's encephalopathy. J Neuropsychiatry Clin Neurosci 2006;18:14–
22. Huang W, Xia C. Infectious disease or Hashimoto's encephalopathy flares: a case report. Seizure 2011; 20:717–719.