

# Steroid-responsive Encephalopathy Associated with Autoimmune Thyroiditis: An Underdiagnosed Cause of Rapidly Progressive Dementia—A Case Report

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## ABSTRACT

**Background:** Steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) is a rare autoimmune disease associated with varied manifestations like rapidly progressive dementia (RPD) and other neuropsychiatric manifestations.

**Case description:** A 65-year-old female presented with subacute onset rapidly progressive memory loss and behavioral disturbances along with extrapyramidal symptoms for 3 months. Her MRI brain and EEG showed no significant findings. On further evaluation, her anti-TPO levels and antithyroglobulin levels were found to be significantly elevated. Other causes of RPD were ruled out. The patient showed a dramatic improvement after administering steroids.

**Conclusion:** Given the reversibility of clinical manifestations, SREAT should be considered in the differential diagnosis when evaluating such a clinical picture, and treatment with steroids should not be delayed if the diagnosis is considered. This case report highlights the importance of early identification and treatment with steroids for autoimmune causes of RPD.

**Keywords:** Case report, Hashimoto's encephalopathy, Rapidly progressive dementia, Steroids, Steroid-responsive encephalopathy associated with autoimmune thyroiditis.

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## INTRODUCTION

Hashimoto's encephalopathy otherwise known as Steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) is rarely associated with autoimmune thyroid disorder.<sup>1</sup> This is a rare condition, with a worldwide prevalence of about 2.1 per 1,00,000.<sup>2</sup> Rapidly progressive dementia (RPD) is one of the many presentations of the disease which is highly responsive to steroids.<sup>3</sup> Steroid-responsive encephalopathy associated with autoimmune thyroiditis bears a favorable prognosis, many patients return to their baseline or close to their baseline within weeks.

## CASE DESCRIPTION

This is a case history of a 65 years old hypertensive lady who presented with subacute onset rapidly progressive memory loss in the form of misplacing objects, not being to recall names of spouse, daughter, and behavioral disturbances in the form of aggressive behavior with insomnia for 3 months. There is nil significant family history.

On examination, apart from very poor comprehension, she also had extrapyramidal features like the rigidity of upper limbs and lower limbs with mask-like facies, non-accommodative glabellar tap, and short stepping gait.

Investigations showed a high ESR of 60 mm/hr. Her CSF analysis was found to be normal. EEG showed diffuse background slowing. The MRI brain showed diffuse cortical atrophy. Her free T4 was 0.9 ng/dL and her thyroid stimulating hormone (TSH) level was 20 mIU/L. Her anti-TPO levels were as high as 1000 IU/mL and anti-thyroglobulin antibodies were also grossly elevated.

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After intravenous steroid therapy, she had a drastic improvement. Whether steroid responsiveness can be used as a diagnostic criterion of SREAT is still a matter of debate, but the pathogenesis of SREAT is characterized by a good responsiveness to steroids.<sup>4</sup>

## DISCUSSION

Steroid-responsive encephalopathy associated with autoimmune thyroiditis, is a rare immunological disorder that characterized by encephalopathy which is subacute in onset with elevated thyroid antibodies and responsiveness to steroids in the absence of other autoantibodies.<sup>5</sup> It is characterized by various neuropsychiatric

manifestations like psychosis, and seizure, or can present with cognitive decline, delirium, depression, myoclonus, and tremors.<sup>6</sup> Women with elevated antithyroid antibodies usually manifest with such a condition and is a diagnosis of exclusion.<sup>7</sup> The conditions that considered before making a diagnosis of Hashimoto's encephalopathy include neuroinfections, inflammatory conditions such as SLE, and neurodegenerative diseases such as CJD, limbic encephalitis, and vascular etiologies.<sup>8</sup> We ruled out other causes like paraneoplastic disorders, infectious causes, connective tissue disorders, other autoimmune causes and vascular etiologies. The proposed hypothesis for this encephalopathy include a disordered immune system, vasculitis, recurrent demyelination, and thyrotropin-releasing hormone and its toxic effects.<sup>9</sup> Fatal complications can be avoided by fast initiation of treatment. It is fully reversible if treated on time.

## CONCLUSION

When there are high titres of anti-TPO and/or antithyroglobulin antibodies, after ruling out other etiologies such as neoplastic, inflammatory and infective etiologies, a diagnosis of SREAT is made.<sup>4</sup> As SREAT is fully reversible, this diagnosis is to be kept in mind when evaluating such a clinical picture and prompt treatment with steroids should be initiated.<sup>4</sup>

## REFERENCES

1. Carlone C, Todini L, Marini I, et al. Acute psychiatric presentation of steroid-responsive encephalopathy: The underrecognized side of autoimmune thyroiditis. *Rivista di Psichiatria* 2013;48(2):169–173. DOI:10.1708/1272.14042.
2. Ferracci F, Moretto G, Candeago RM, et al. Antithyroid antibodies in the CSF: Their role in the pathogenesis of Hashimoto's encephalopathy. *Neurology* 2003;60(4):712–714. DOI: 10.1212/01.wnl.0000048660.71390.c6.
3. Anand KS, Garg J, Verma R, et al. Hashimoto's encephalitis: Unusual cause of reversible dementia. *J Family Med Prim Care* 2014;3(3):284–286. DOI: 10.4103/2249-4863.141650.
4. Liyanage CK, Munasinghe TM, Paramanatham A. Steroid-responsive encephalopathy associated with autoimmune thyroiditis presenting with fever and confusion. *Case Rep Neurol Med* 2017;2017:3790741. DOI: 10.1155/2017/3790741.
5. Stern JM, Salamon N, Stern JM, et al. Hashimoto encephalopathy also known as steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT). *Imaging of epilepsy: A Clinical Atlas*. 2022:169–173. DOI: 10.1007/978-3-030-86672-3\_40.
6. John R, Datta A, Ovallath S. A case of euthyroid steroid-responsive encephalopathy with subacute dementia. *Cureus* 2021;13(9):e17689. DOI: 10.7759/cureus.17689.
7. Chong JY, Rowland LP, Utiger RD. Hashimoto encephalopathy: syndrome or myth? *Arch Neurol* 2003;60(2):164–171. DOI: 10.1001/archneur.60.2.164.
8. Lee SW, Donlon S, Caplan JP. Steroid responsive encephalopathy associated with autoimmune thyroiditis (SREAT) or Hashimoto's encephalopathy: A case and review. *Psychosomatics* 2011;52(2):99–108. DOI: 10.1016/j.psym.2010.12.010.
9. Aladdin Y, Shirah B. Hashimoto's encephalopathy masquerading as rapidly progressive dementia and extrapyramidal failure. *J Neurosci Rural Pract* 2022;13(1):101–104. DOI: 10.1055/s-0041-1741487.