Case Report **IJCR (2023) 7:292**

International Journal of Case Reports (ISSN:2572-8776)

Steroid-Responsive Encephalopathy Associated with Autoimmune Thyroiditis (SREAT) Masquerading as New-Onset Bipolar Disorder in an Elderly Female

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ABSTRACT

Steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT), also known as Hashimoto's Encephalopathy (HE), is an immune-mediated condition that may present with a myriad of neuropsychiatric symptoms, making diagnosis a quandary.

We report a case of a 70-year-old female who presented with subacute onset of mania and cognitive decline, diagnosed as bipolar disorder. She was treated with valproic acid for mood stabilization with minimal improvement in symptoms and six months later presented with new-onset seizures. Interestingly, her seizures started four days after discontinuing valproic acid. Following her admission to our hospital, a diagnosis of autoimmune epilepsy was suspected. EEG revealed multiple focal onset seizures with secondary generalization originating from the right frontocentral area and/or right temporal lobe. MRI revealed asymmetric cortical thickening along the paramedian right anterior frontal region thought to be nonspecific. She was empirically treated with pulse dose steroids and intravenous immunoglobulins. She underwent an extensive work up including neuroimaging studies that were unremarkable as well as serological testing for autoimmune etiologies. A high titer of anti-thyroid peroxidase (anti-TPO) antibodies was detected. She was clinically and biochemically euthyroid. A diagnosis of SREAT was rendered and the patient had significant clinical improvement in symptoms following administration of corticosteroids.

SREAT is a diagnosis of exclusion but is supported by the presence of elevated anti-TPO and steroid responsiveness. Initial presentation may masquerade as a primary mood disorder, as in this case. Prompt diagnosis and treatment fully reverses neurological and psychiatric dysfunction in most cases.

Keywords: neuroimmunology, immunology, neuropsychiatry, geriatric neuropsychiatry, thyroid, SREAT, bipolar disorder, epilepsy, seizures, autoimmune encephalopathy

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How to cite this article:
Dawn Radford, Manish Karamchandani, McKay Hanna, Mini Singh. Steroid-Responsive Encephalopathy Associated with Autoimmune Thyroiditis (SREAT) Masquerading as New-Onset Bipolar Disorder in an Elderly Female. International Journal of Case Reports, 2023, 7:292.

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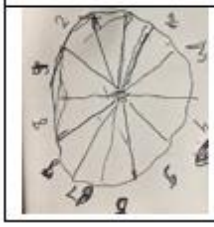
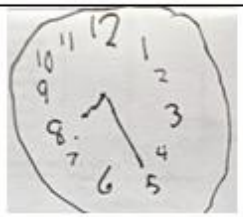
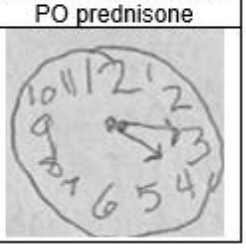
Introduction

Steroid-Responsive Encephalopathy Associated with Autoimmune Thyroiditis [SREAT], also known as Hashimoto's Encephalopathy, is an autoimmune encephalopathy associated with positive anti-thyroid antibodies, anti-thyroid peroxidase [anti-TPO] and/or anti-thyroglobulin [anti-TG] [1]. SREAT commonly presents with neuropsychiatric symptoms, such as psychosis, progressive cognitive decline, mood disturbance, and new onset seizures and has good response to steroid treatment in up to 90% of cases [2,3]. The prevalence is 2.1 per 100,000 and is more commonly seen in females at a ratio of 4 to 1 [2]. The pathogenesis is debatable, and it is unclear whether the anti-thyroid antibodies are causative or solely act as markers of autoimmunity [4]. In the absence of an alternative diagnosis, SREAT should be considered on the differential in patients presenting with new neuropsychiatric disturbance and encephalopathy so that prompt treatment with steroids can be initiated. Here we report a case of SREAT diagnosed in a 70-year-old female, who was euthyroid, presenting with subacute neuropsychiatric manifestations including mania, cognitive decline, and seizures.

Table 1: Improvement in clock drawings throughout course of IV Methylprednisolone

Day of admission prior to treatment	After 1 dose of IVMP	After 7 doses of IVMP	After 9 doses of IVMP	After 9 doses of IVMP and initiation of PO prednisone
				

Case Report

A 70-year-old female with a longstanding history of anxiety and mitral valve prolapse status post mitral valve repair presented with recurrent episodes of seizure-like events in the setting of progressive cognitive decline and psychiatric disturbances. Seven months prior to her initial presentation at our hospital, she demonstrated symptoms of mania and paranoia ultimately resulting in the patient being admitted to a psychiatric hospital. She was diagnosed with "late-onset bipolar disorder" and started on valproic acid and aripiprazole. Due to extrapyramidal side effects, valproic acid and aripiprazole were discontinued about four days prior to initial presentation. Subsequently, she was admitted to the hospital after having 3 seizure-like events within 24 hours. The episodes were described as staring, followed by full body shaking and post-ictal confusion. The patient did not have any prior history of seizures.

On examination, the patient was hypoactive and minimally responsive to external stimuli. She was largely non-verbal but was intermittently able to speak in slow monosyllabic phrases with frequent perseveration. Her writing was disorganized and nonsensical, and her initial clock drawing [Table 1] had incorrect numerical sequence, missing numbers, and inaccurate insertion of lines indicating deficits in executive function, visual-spatial ability, motor programming, and attention. Due to concern for a possible catatonic state, psychiatry was consulted who performed a Bush-Francis Catatonia Rating Scale [BFCRS]. The patient's BFCRS was 7 which is not indicative of catatonia.

Her initial presentation was that of status epilepticus and the seizures lateralized to the

right hemisphere with electrographic suggestion of right frontocentral onset and/or right temporal propagation on continuous video EEG [Figure 1]. The clinical phenomenology consisted of left-sided clonic jerking, eye deviation, and head version prior to secondary generalization with tonic clonic movement of all extremities. The patient was treated with benzodiazepines followed by Levetiracetam. However, the patient subsequently had multiple right hemispheric onset seizures associated with decreased responsiveness and staring on day six and eight of admission which resolved with the addition of Lacosamide. MRI brain with and without contrast was obtained which revealed a suggestion of non-specific, mild asymmetric cortical thickening along the paramedian right anterior inferior frontal region on axial T2 imaging. CT chest, abdomen, and pelvis were obtained which were unremarkable. Pelvic ultrasound revealed a hypoechoic mass that was further characterized as a uterine fibroma when further evaluated with a pelvic MRI.

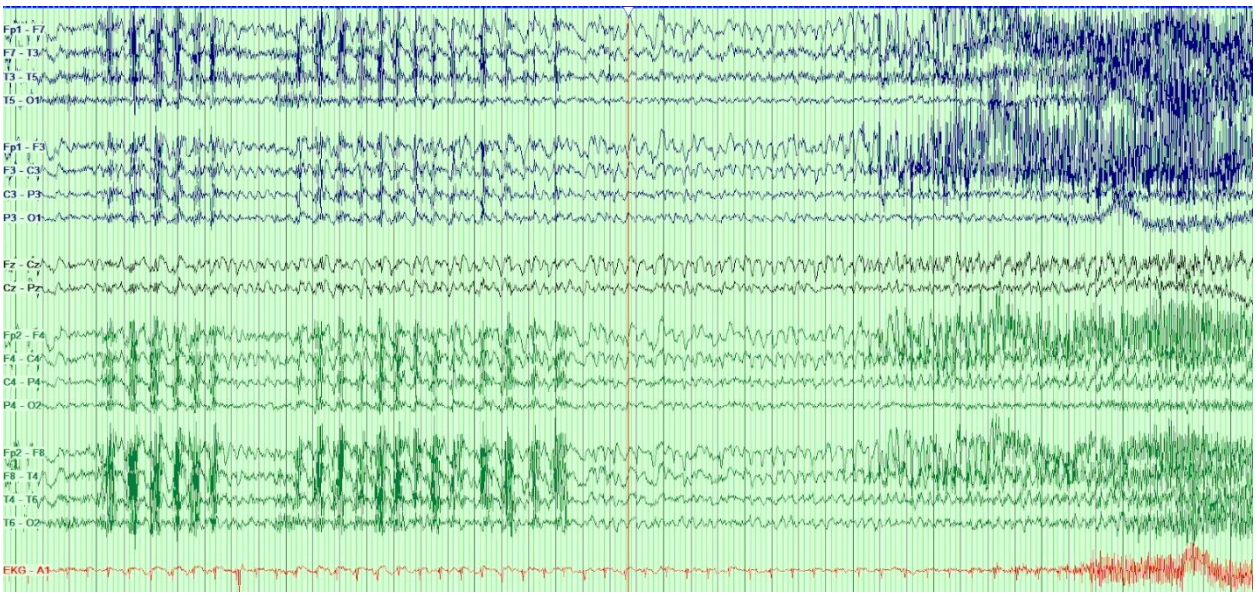


Figure 1: Secondary generalized electroconvulsive seizure on continuous video EEG

Initial laboratory workup was largely unremarkable. Serum labs revealed elevated liver enzymes, AST 53 and ALT 116, and thyroid function was normal [TSH 2.2, free T4 1.16]. Lumbar puncture was performed with normal cell counts, glucose, and protein [49.4]. Viral CSF studies were negative, including herpes simplex virus [HSV], cytomegalovirus [CMV], enterovirus, and West Nile Virus. CSF bacterial culture was negative, as well as negative oligoclonal bands [OCB], Angiotensin Converting Enzyme [ACE], and VDRL. EKG revealed sinus bradycardia but was otherwise unremarkable. Due to concern for autoimmune encephalopathy with an Antibody Prevalence in Epilepsy and Encephalopathy [APE2] score of 6, the patient was treated with a four-day course of intravenous immune globulin [IVIG] 0.5 mg/kg and a five-day course of IV methylprednisolone [IVMP] 1 gram daily. During treatment with IVIG and IVMP, the patient had a marked improvement in neurological symptoms. She became alert, oriented, and interactive with increased mobility. She was able to converse with normal speech and was able to follow commands with accuracy. The patient's clock drawings improved throughout the treatment course [table 1]. However, following completion of the IVMP, the patient had an acute decline in neurological status associated with immobility, staring, mutism, rigidity, and withdrawal. Additional labs were obtained including serum thyroid peroxidase [TPO] antibody and anti-thyroglobulin antibody which were elevated at

62.7 and 80.7, respectively. TSH receptor antibody and thyroglobulin mass spectroscopy were within normal limits. Serum paraneoplastic antibody panel was negative. Serum and CSF autoimmune encephalopathy panel was negative. Due to the combination of subacute onset encephalopathy, elevated thyroid antibodies [TPO and thyroglobulin antibodies], absence of specific neuronal antibodies, and marked immunotherapy responsiveness [5], the presumed diagnosis was Steroid Responsive Encephalopathy associated with Autoimmune Thyroiditis [SREAT]. Therefore, the decision was made to treat with an additional four days of IV methylprednisolone [IVMP] 1 gram daily, and the patient again showed marked improvement. The patient was transitioned to prednisone 60 mg daily, however the patient again had a clinical deterioration about three days later, manifesting as an acute psychotic episode of mutism and disorganized thoughts. Prednisone was increased to 80 mg daily with clinical improvement, and the patient was discharged from the hospital.

Discussion

SREAT represents a rare and underdiagnosed clinical condition which may be missed on initial evaluation. The diagnosis of SREAT is based on the following criteria: relapsing-remitting episodes of neurological and/or psychiatric symptoms in the absence of other etiologies of encephalopathy, an abnormal elevation of anti-TPO antibodies, and symptomatic improvement or remission following treatment with corticosteroids [5]. The initial presentation of SREAT is diverse and includes convulsions, confusion, speech disorders, memory impairment, gait disturbance, persecutory delusions, myoclonus, headaches, and coma [6]**.** In addition, cases of acute onset of psychosis, depression, or mania have also been reported as initial manifestations [5, 7, 8, 9]**.** Castillo et al. [10] found psychiatric presentation [psychosis or paranoia] in 25% and Laurent et al. [6] found isolated psychiatric disorder in 10% of SREAT patients. Our patient presented with new-onset seizures, progressive cognitive decline, and mania and was given an initial diagnosis of Late-Onset Bipolar Disorder.

The differential diagnosis of SREAT is broad, and misdiagnosis is common. Per Castillo et al [10], the most frequent misdiagnoses were viral encephalitis, Creutzfeldt-Jakob disease, and degenerative dementia. 5% of the patients in this study had an initial misdiagnosis of psychosis. Our patient had an initial misdiagnosis of Late-Onset Bipolar Disorder [LOBD] and was started on valproic acid for mood stabilization. LOBD is less likely in our patient as Yassa et al [11] proposed an age of 50 as a cut off for the late onset bipolar disorder, and our patient was 70 years old at time of diagnosis. About 5% of patients are reported to have onset of illness after age 60 [12]. In addition, the patient had minimal improvement in manic symptoms and six months later developed new-onset seizures, not typical of LOBD. Interestingly, the seizures started four days after discontinuation of valproic acid which theorizes a hypothesis that the misdiagnosis and treatment of mania with valproic acid kept the natural course of SREAT at bay. Other findings that favor a diagnosis of SREAT over LOBD include cognitive impairment, sleep disturbance, tremor, elevated anti-TPO and antithyroglobulin antibodies, findings of both focal and generalized seizure activity on EEG, and non-specific findings on MRI brain. Most importantly, the patient had a marked improvement with corticosteroids which would not be seen with LOBD.

In conclusion, SREAT represents a rare condition that is likely under-diagnosed. SREAT is a diagnosis of exclusion, however, clinicians should have a high degree of suspicion due to its heterogeneous clinical presentation. The diagnosis is supported by the presence of elevated anti-TPO antibodies and steroid responsiveness. Marked symptom improvement and clinical remission after corticosteroid treatment emphasizes the importance of considering SREAT in the differential diagnosis among patients with acute neuropsychiatric

symptoms. The reported case is a prototypical example of how common psychiatric diagnoses, such as Bipolar Disorder, can mask an underlying diagnosis of SREAT and treatment with steroids should not be delayed if the diagnosis is considered.

Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this article.

Authors Contributions

All the authors were involved in patient management and care. The article was written by Dawn Radford, Manish Karamchandani, McKay Hanna, and Mini Singh. The authors have made contributions to the analysis and interpretation of data and revising the manuscript. All authors have given final approval of the version to be published.

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