

Case Reports

Steroid-responsive Encephalopathy Associated with Autoimmune Thyroiditis (SREAT) Presenting with Pure Cerebellar Ataxia

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Abstract

Background: Myoclonus and tremor are common movement disorder phenomenologies in steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT). Pure ataxia without encephalopathy has rarely been reported.

Case report: We report 21- and 40-year-old females who presented with subacute pure ataxia without encephalopathy. After immunotherapies, both exhibited initial improvement of ataxia, and subsequently remained in plateau phase.

Discussion: This treatable disorder should be added to the differential diagnoses of progressive cerebellar ataxia, and anti-thyroid peroxidase and anti-thyroglobulin should be considered as part of the workup. It is crucial not to misdiagnose SREAT presenting with pure cerebellar ataxia as degenerative or spinocerebellar ataxia.

Keywords: Hashimoto encephalopathy, steroid-responsive encephalopathy associated with autoimmune thyroiditis, SREAT, ataxia, movement disorders, autoimmune, autoimmune thyroiditis

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Introduction

Steroid-responsive encephalopathy with autoimmune thyroiditis (SREAT), also called Hashimoto encephalopathy, has been a controversial disorder in the literature.^{1–4} The term “non-vasculitic autoimmune meningoencephalitis” has also been less commonly used.⁵ The main clinical presentation is encephalopathy; however, patients may exhibit associated neurological features including seizures, movement disorders such as myoclonus or tremor, and psychosis.^{3,6–9} Elevation of anti-thyroid antibodies, i.e. anti-thyroid peroxidase (anti-TPO or anti-thyroid microsomal antibody) and/or anti-thyroglobulin (anti-Tg), is required to make the diagnosis.^{10,11} Patients typically show good response to immunotherapies, including steroids.^{7,12}

Uncommon movement disorder phenomenologies in SREAT include ataxia without encephalopathy,^{13–19} chorea,^{20–22} and dystonia.²² Here, we report two patients who presented with subacute onset of pure or

predominant cerebellar ataxia and negative autoimmune and paraneoplastic workups, except anti-thyroid antibodies. The diagnosis could easily have been mistaken for degenerative ataxia, such as spinocerebellar ataxia, had this etiology not been searched for.

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Patient 1

The patient was a 21-year-old female with type 1 diabetes (onset at age 15 years), treated with insulin, and hyperlipidemia. Her initial presentation was walking difficulty that gradually worsened over 3 months. There were no cognitive complaints according to the patient and her family. Examination revealed moderate-to-severe truncal ataxia. Appendicular ataxia in bilateral arms and legs was also noted. There was no evidence of overt encephalopathy on neurological examination; however, formal neuropsychological testing and electroencephalography

(EEG) were not performed. Eye movement examination revealed no square wave jerks. On pursuit examination, there was mild left beating gaze-evoked nystagmus on the extreme left gaze. No apparent saccadic pursuits were observed. Saccade examination showed no slow or hypermetric saccades. There was no limitation in range of eye movements in any direction. Deep tendon reflexes (DTRs) were all normal. There was no pes cavus.

Investigations for autoimmune ataxia revealed positive anti-TPO (1:1600, normal <1:100), and borderline positive anti-Tg (1:40, normal <1:20). Thyroid function tests were unremarkable. Serum anti-glutamic acid decarboxylase (anti-GAD) was negative. Serum anti-nuclear antibody, and anti-Ro and anti-La antibodies were negative. Serum ceruloplasmin was normal. Cerebrospinal fluid (CSF) examination revealed protein 46 mg/dL, glucose 112 mg/dL (serum glucose 196 mg/dL), white cells 1 per mm³, red cells 570 per mm³. CSF oligoclonal bands, anti-aquaporin 4 (or anti-neuromyelitis optica), and CSF paraneoplastic panel including anti-Hu (or anti-neuronal nuclear antibody type 1 [ANNA-1]), anti-Ri (ANNA-2), anti-Yo (or anti-Purkinje cell cytoplasmic antibody type 1 [PCA1]), anti-PCA2, anti-Tr, anti-MAG (anti-myelin-associated glycoprotein), anti-Ma, anti-GAD, anti-CV2/CRMP5, and anti-amphiphysin antibodies were all negative. Anti-gliadin and anti-tissue transglutaminase antibodies were not tested. Magnetic resonance imaging (MRI) of the brain was normal, and there was no evidence of cerebellar atrophy. Genetic testing for Friedreich's ataxia (FA) was not performed.

A 3-day course of intravenous methylprednisolone 1 g/day was initiated, and led to marked improvement in her ataxia, which then plateaued after a few weeks. The patient exhibited residual mild ataxia. She continued on oral prednisone 1 mg/kg/day. With high-dose prednisone, she developed steroid-induced hyperglycemia with blood sugar levels in the range between 200 and 300 mg/dL. This was also co-managed by her endocrinologist. Prednisone was successfully tapered very gradually, without exacerbation or worsening of ataxia. She remained on 5 mg/day of oral prednisone. Her ataxia had plateaued over the period of the 4-year follow-up without further improvement or worsening (Video 1).

Patient 2

The second patient was a 40-year-old female with no known underlying disease. She presented with progressive impairment of gait and fine motor coordination of both hands over the time course of 6 months. There was no family history of ataxia or other movement disorders. Her mental status was normal without any evidence of encephalopathy. On examination, she was cognitively normal. She could respond to all questions and commands appropriately. However, formal neuropsychological testing and EEG were not performed. Her speech was moderately slurred and had scanning quality. There was moderate left dysmetria that was slightly greater than right dysmetria. She walked with moderately wide-based ataxic gait. In addition to ataxia, she also had mild dystonic posturing of both hands, the right slightly greater than the left. Eye movement examination revealed mild square wave jerks. Pursuits were normal. There was no limitation in



Video 1. This Video Shows Patient 1 after the Treatment. The examination was performed at age 25, 4 years after the onset of ataxia. At that time, she was on prednisone 5 mg every other day. She had only very mildly slurred and scanning speech. Eye movement examination revealed no square wave jerks. There was no limitation of eye movements in any direction on both smooth pursuit and saccade examination. There were no apparent saccadic or broken pursuits. There was mild left beating gaze-evoked nystagmus on the extreme left gaze. Saccades were normal. There was neither slowing of saccades nor hypermetric saccade. Mild dysmetria and overshoot dysmetria, left greater than right, were demonstrated on finger-to-nose-to-finger and finger following tests, respectively. There was mild dysdiadochokinesia, which was more prominent on the left. She was able to walk independently, but her gait was wide-based. She had imbalance when standing with both feet together and eyes closed. She required assistance when standing on only one foot, either left or right.

range of eye movements in any direction. Saccades were hypermetric and more prominent in horizontal directions. There was no slowing of saccades. DTRs were all normal. No pes cavus was observed.

Extensive investigations, including serum ceruloplasmin, urine copper, and a serum paraneoplastic panel, were negative except for positive serum anti-TPO (238.87 IU/mL, normal 0–5.61; note the different laboratory from patient 1) and anti-Tg (57.35 IU/mL, normal 0–4.11) antibodies. A thyroid function test including thyroid stimulating hormone (TSH), free thyroxine (FT4), and triiodothyronine (T3) was normal. CSF examination revealed protein 40.8 mg/dL, glucose 63 mg/dL, white cells 0 per mm³, and red cells 50 per mm³. CSF paraneoplastic panel including anti-Hu (ANNA1), anti-Ri (ANNA-2), ANNA-3, anti-GAD, anti-amphiphysin, anti-CV2/CRMP5, anti-NMDA receptor (anti-N-methyl-D-aspartate receptor), anti-AMPA receptor, anti-GABA(B) receptor and anti-VGKCC (anti-voltage-gated potassium channel complex) were all negative. Anti-gliadin and anti-tissue transglutaminase antibodies were not tested. MRI of the brain revealed no cerebellar atrophy or other abnormalities. Genetic testing for common types of spinocerebellar ataxia (SCA) in Thailand including SCAs 1, 2, and 3 were negative. Genetic testing for FA was not performed.

She was treated with a 3-day course of intravenous methylprednisolone 1 g/day, which led to slight improvement in her ataxia initially. She was discharged with oral prednisone 1 mg/kg/day. After a gradual moderate improvement over several weeks, her ataxia plateaued and was stable at a mild-to-moderate degree. Tapering of the oral prednisone was trialed but led to worsening of her ataxia. Despite treatment with oral prednisone for 1.5 years, there was no further improvement and the decision was made to start azathioprine as a



Video 2. This Video Shows Patient 2 after the Treatment. The examination was performed at age 40, 2 years after the onset of ataxia. She was on azathioprine 150 mg/day and a tapering dose of oral prednisone. The cerebellar features in this patient were overall more prominent than in patient 1. Her speech was moderately slurred and scanning. Eye movement examination demonstrated mild square wave jerks. Pursuits were normal. There were hypermetric saccades, more prominent in horizontal than vertical directions. There was no slowing of saccades or limitation of eye movements in any direction. Mild dystonic posturing of both hands, right slightly greater than left, was demonstrated. Finger-to-nose-to-finger test revealed mild dysmetria, left greater than right. Finger following test demonstrated mild-to-moderate overshoot dysmetria, left greater than right. Mild-to-moderate dysidiadochokinesia, left greater than right, was also demonstrated. She was able to walk independently, but had wide-based gait. She was unable to stand with both feet together without assistance, and had marked difficulty standing on only one foot.

steroid-sparing agent. During the last follow-up, she had been on azathioprine 150 mg/day and a taper dose of prednisone. Her ataxia had remained stable during the transitional period (Video 2).

Discussion

Our patients presented with subacute onset of cerebellar ataxia with speech, trunk, and appendicular involvement. Movement disorders can be manifestations of SREAT, and the common phenomenologies are myoclonus, tremor, and ataxia.^{6,8} Dystonia and chorea have much less commonly been reported in this rare entity.²⁰⁻²² Although ataxia

(in combination with encephalopathy and other features) is not an uncommon phenomenology in SREAT, there have been a limited number of reports of patients with pure or predominant ataxia *without* encephalopathy (Table 1). When ataxia is present as a sole or predominant feature, the diagnosis can easily be missed or mistaken for degenerative ataxia.

Although this disorder has long been described, there remain several controversial issues, including heterogeneity and the wide spectrum of its clinical features (e.g. the absence of encephalopathy in some cases), unclear association with thyroid disorders (since most patients are euthyroid), appropriate nomenclature, non-specificity, and pathogenic roles of anti-TPO and/or anti-Tg, as well as their correlations with clinical severity.¹⁻⁴ Anti-TPO and anti-Tg are common in the general population and in patients with benign thyroid disorders without any neurologic features.²³ This can be a confounding factor in the diagnosis of SREAT; therefore, the positivity of these antibodies should be interpreted in the appropriate clinical context. In spite of these controversies, clinical recognition of SREAT should not be dismissed due to its treatable nature and generally good outcomes.

Our patients demonstrated improvement with steroids, as did several cases previously reported in the literature.^{7,12} Patients may be misdiagnosed with degenerative ataxia, especially spinocerebellar ataxia, given an all negative extensive workup, as has occurred in some reported cases with clinical features mimicking spinocerebellar ataxia.^{16,17} Since this is a treatable entity, we propose the inclusion of investigations for SREAT as a first or second tier in patients presenting with subacute or even chronic progressive cerebellar ataxia, so that an opportunity for treatment and improvement is not missed. Although there are no systematic studies on treatment outcome in this rare entity, amelioration of ataxia can potentially improve ambulatory function and reduce disability in these patients.

While encephalopathy and various neurologic deficits such as seizures, myoclonus, or psychosis are typical clinical features, many patients without encephalopathy have been reported. Recently, Miranda et al.²² reported two cases who presented with movement disorders (one with myoclonus, dystonia, and ataxia, and the other with choreoathetosis and dystonia) without encephalopathy, and proposed the name, “non-encephalopathic Hashimoto’s thyroiditis”. Given the broadening spectrum of this entity and unclear/controversial relationship with Hashimoto thyroiditis, we propose the name “anti-TPO/Tg antibody-related neurologic disorders responsive to steroids (ATANDS)” to cover the entire spectrum of neurologic disorders associated with these two antibodies. This should not only avoid the confusion about encephalopathy and the association with thyroid disorders, but also highlight the presence of anti-thyroid antibodies and typically good response to steroids as its core features.

We performed a literature review of SREAT cases presenting with pure ataxia or ataxia as a predominant feature, without encephalopathy (Table 1). Similar to classic SREAT, patients with predominant ataxia were mostly females (16/22 patients, female/male 8:3), and euthyroid (20/22 patients, the other two had subclinical hypothyroidism). Interestingly, two patients with predominant ataxia also had saccadic intrusions (one with opsoclonus and one with ocular flutter).

Table 1. Our Cases and Previously Reported Cases of Steroid-responsive Encephalopathy Associated with Autoimmune Thyroiditis (SREAT) with Pure or Predominant Cerebellar ataxia.

Authors/ Year	Age (yr)	Sex	Phenomenology of Ataxia	Other Neurologic Features	Time to Dx (mo)	Thyroid status	Anti-TPO	Ant-Tg	Treatment	Outcome of Ataxia
Patient 1	21	F	Limb, trunk, gait	None	3	Eu	1:1600 (1:100)	1:40 (1:20)	IVMP → PO Pred	Improved markedly with residual plateaued mild ataxia
Patient 2	40	F	Limb, gait	Dystonia – b/1 hands	6	Eu	238.87 IU/mL (0–5.61)	57.35 IU/mL (0–4.11)	IVMP → AZA	Moderately improved, then plateaued over 2-yr follow-up period
Alvarez Bravo et al. ¹³	47	F	Limb, trunk, gait	Opsoclonus	0.5	Eu	765 IU/mL (<34)	nl	IVMP → PO Pred	Excellent
Alzubi et al. ¹⁴	17	F	Limb, gait, nystagmus, intention tremor	Vestibular - vertigo, nausea, vomiting; Optic neuritis, L>R	2	Hypo (subclinical)	423 IU/mL (at the first f/u)	62.9 IU/mL	IVMP → MMF	Markedly improved → 2 relapses (4 and 20 mo later); also had similar symptoms 1 yr prior to this Dx
Lee et al. ¹⁵	30	M	Limb	Ocular flutter	0.13	Eu	nl	398 U/mL (<115)	IVMP → PO Pred	Excellent (full recovery 3 mo after)
Matsunaga et al. ¹⁶ (Pt 4)	63	F	Limb, trunk	"Tremor"	1	Eu	28.1 U/mL (<0.3)	316 U/mL (<0.3)	IVMP	Excellent (full recovery)
Matsunaga et al. ¹⁶ (Pt 5)	66	F	Limb, trunk, dysarthria	"Tremor"	2	Eu	105 U/mL (<0.3)	255 U/mL (<0.3)	IVMP	Good
Matsunaga et al. ¹⁶ (Pt 7)	84	F	Limb, trunk, dysarthria	None	72	Eu	14.8 U/mL (<0.3)	2.7 U/mL (<0.3)	IVMP	Fair
Matsunaga et al. ¹⁶ (Pt 8)	55	M	Limb, trunk, dysarthria	None	4	Eu	7.7 U/mL (<0.3)	<0.3 U/mL (<0.3)	IVMP	Fair
Matsunaga et al. ¹⁶ (Pt 10)	54	M	Limb, trunk, dysarthria	None	120	Eu	0.5 U/mL (<0.3)	10 U/mL (<0.3)	IVMP	Good
Matsunaga et al. ¹⁶ (Pt 11)	61	M	Limb, trunk, dysarthria	None	12	Eu	59.7 U/mL (<0.3)	5.1 U/mL (<0.3)	IVMP, IVIG	Good
Matsunaga et al. ¹⁶ (Pt 12)	57	F	Limb, trunk	None	12	Eu	109 U/mL (<0.3)	1.6 U/mL (<0.3)	IVMP	Fair
Matsunaga et al. ¹⁶ (Pt 13)	46	F	Limb, trunk	"Tremor"	6	Eu	1.9 U/mL (<0.3)	<0.3 U/mL (<0.3)	IVMP	Fair
Tang et al. ¹⁹	39	M	Limb, gait, dysarthria	Facial, b/1 arm weakness (gr 4/5)	17	Eu	>1,300 U/mL (0–60)	83.2 U/mL (0–60)	IVMP → PO Pred	Able to walk 1,000 m along but speech was unchanged

Table 1. Continued

Authors/ Year	Age (yr)	Sex	Phenomenology of Ataxia	Other Neurologic Features	Time to Dx (mo)	Thyroid status	Anti-TPO	Ant-Tg	Treatment	Outcome of Ataxia
Nakagawa et al. ¹⁷	41	F	Gait, limb, mild dysarthria	None	9	Eu	50 IU/mL (<0.3)	4.9 IU/mL (<0.3)	IVMP → PO Pred	Ataxia almost disappeared at 3-mo follow up
Avila et al. ²⁸	72	F	Gait	Myoclonus at b/l arms; impaired verbal memory, attention and mild frontal syndrome on cognitive testing	18	Hypo (subclinical)	>453 IU/mL (<18)	>4,262 IU/mL (<28)	Steroids	Dramatic but temporary improvement; Died 6 wk later
Selim and Drachman ¹⁸ (Pt 1)	57	F	Gait, hands, dysarthria, nystagmus	“Dizziness”, diplopia peripheral neuropathy	“Long- standing”	Eu	>30 IU/mL (0.3–1)	2.9 IU/mL (0.3–1)	L-thyroxine	Continued to deteriorate; able to ambulate with only a walker 6 yr after
Selim and Drachman ¹⁸ (Pt 2)	35	F	Gait, hands, dysarthria	None	60	Eu	13.2 IU/mL (0.3–1)	1.7 IU/mL (0.3–1)	L-thyroxine	No improvement; worsening gait disability 3 yr later
Selim and Drachman ¹⁸ (Pt 3)	56	F	Gait, hands, dysarthria	Vertigo	72	Eu	22.1 IU/mL (0.3–1)	16.9 IU/mL (0.3–1)	PO Pred (then stopped due to hypertension)	Continued to deteriorate; became wheelchair bound
Selim and Drachman ¹⁸ (Pt 4)	69	F	Gait, hands, dysarthria	Diplopia, extrapyramidal features (at onset); lapses in concentration on cognitive testing	N/A	Eu	3.7 IU/mL (0.3–1)	1.8 IU/mL (0.3–1)	L-dopa	Continued to deteriorate
Selim and Drachman ¹⁸ (Pt 5)	46	F	Gait, hands, dysarthria, nystagmus	Diplopia, vertigo, hearing impairment, peripheral neuropathy	3	Eu	2 IU/mL (0.3–1)	1.5 IU/mL (0.3–1)	IVIG (given 8 mo after the initial evaluation)	Able to ambulate with walker 2 mo after IVIG but other features are unchanged
Selim and Drachman ¹⁸ (Pt 6)	61	M	Gait, hands, dysarthria	Hearing impairment, extrapyramidal features	60	Eu	82.7 IU/mL (<35)	<20 IU/mL (<35)	N/A	N/A

Abbreviations: Anti-Tg, Anti-thyroglobulin; Anti-TPO, Anti-thyroid peroxidase; AZA, Azathioprine; b/l, Bilateral; Dx, Diagnosis; Eu, Euthyroid; F, Female; gr, Grade; Hypo, Hypothyroidism; IU/mL, International Units per Milliliter; IVIG, Intravenous Immunoglobulin; IVMP, Intravenous Methylprednisolone; M, Male; m, Meter; MMF, Mycophenolate mofetil; mo, month(s); N/A, Not Applicable; nl, Normal; PO Pred, Oral Prednisone; Pt, Patient; U/mL, Units per Milliliter; wk, Week(s); yr, Year(s). The patients were reported in chronological order of year of publication. Of note, the numbers of patients in the original reports are indicated in parentheses. Age at the time of diagnosis, sex, phenomenology of ataxia, other neurologic features, time to the diagnosis of SREAT (in months), serum anti-TPO, anti-Tg with normal values in parentheses, treatment and outcomes of ataxia after treatment are demonstrated. Note that normal values of anti-TPO and anti-Tg antibodies are different between laboratories.

Time to diagnosis varied from 4 days to 120 months. Two patients with relatively early (<1 month) diagnosis were the ones with saccadic intrusions. When ataxia was more pure or with fewer prominent additional features, the time to diagnosis tended to be delayed into the subacute and chronic periods. All but one patient (21/22, 95%) had positive anti-TPO antibody, while anti-Tg was elevated in 18 out of 22 patients (82%). Eighteen patients (82%) were treated with immunotherapies including steroids (almost all intravenous) and/or intravenous immunoglobulin (IVIG). Two patients who initially received steroids were switched to steroid-sparing agents: azathioprine and mycophenolate mofetil in each patient. Seventeen of 18 patients who received immunotherapies had improvement of variable degrees (marked or excellent improvement in seven patients [34% of patients treated with steroid]). Of note, the other one who did not improve with immunotherapies received only oral steroids without any intravenous treatment. Interestingly, two patients with delayed diagnosis (72 and 120 months) still had “good” response to immunotherapies. While the analyses from previously reported cases provide some insights into the clinical features and treatment outcome of pure or predominant ataxic form of this disorder, these may be prone to biases. One limitation of the present and previous retrospective studies was the lack of objective assessment of ataxia, such as using the Scale for the Assessment and Rating of Ataxia (SARA) or International Cooperative Ataxia Rating Scale (ICARS) to evaluate treatment outcome. Future prospective studies of this subtype of SREAT with incorporation of these standard rating scales can be helpful, but may not be feasible given its rarity. Furthermore, the true prevalence of this subtype is unknown. It is possible that this subtype of SREAT is under-recognized, as it can easily be hidden among a group of patients with “undiagnosed degenerative ataxia”.

While SREAT can be misdiagnosed as degenerative ataxia, routine tests for anti-TPO and anti-Tg in *all* patients with suspected degenerative ataxia may lead to false-positive results and overdiagnosis of SREAT. Nevertheless, misdiagnosis of SREAT as degenerative ataxia may have more clinical impact on patients, especially as no effective treatment for degenerative ataxia is currently available. SREAT patients who are misdiagnosed as having degenerative ataxia will miss an opportunity for treatment and improvement; however, mistaking degenerative ataxia for SREAT may lead to a trial of steroid, with no improvement and subsequent reconsideration of the diagnosis. Currently, there are no specific clinical features or biomarkers other than anti-TPO, anti-Tg antibodies, and steroid/immunotherapy responsiveness to differentiate SREAT from other degenerative ataxias. We suggest searching for autoimmune etiologies including SREAT in patients with subacute temporal profile of ataxia. However, in patients with chronic ataxia, while we do not advocate routine tests for anti-TPO and anti-Tg, SREAT should be kept when considering differential diagnoses, especially if extensive investigations do not reveal any degenerative etiologies.

FA may be included in the differential diagnoses for the present patients, especially patient 1, who also had co-existing diabetes. However, our patients did not show other clinical features of FA including macrosaccadic oscillations, hypo- (in classic FA) or hyperreflexia

(in late-onset FA). In addition, with regard to the natural history of FA, diabetes usually develops many years after the onset of ataxia. In one large series of 90 FA families, the mean duration of ataxia at the onset of diabetes was 15.5 ± 6.82 years.²⁴ One case report described a FA patient who presented with type 1 diabetes.²⁵ However, this patient subsequently developed ataxia a year later. Our patient had diabetes over 6 years before she developed ataxia. In addition, the subacute time course of ataxia in both of our patients (3 and 6 months) initially prompted workup for autoimmune etiologies (over degenerative causes). Steroid responsiveness in both patients would also argue against the diagnosis of FA. While it may not be totally unreasonable to include FA genetic testing in our patients, this is unfortunately not available.

In summary, identification of SREAT by anti-TPO and anti-Tg testing, which may not be included in conventional “autoimmune panels”, should be considered as part of the workup of subacute and chronic progressive cerebellar ataxia. This disorder is treatable with immunotherapies, and therefore care must be taken not to misdiagnose this entity as degenerative ataxia. While some patients with long disease duration may still benefit from immunotherapies, diagnosis and treatment should not be delayed, as in other autoimmune neurologic disorders.^{26,27}

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