Recurrent Hashimoto's encephalopathy: a case report of reversible coma and status epilepticus

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Background:

Steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) also known as Hashimoto's encephalopathy (HE), is a rare immune-mediated complication independent of functional status of thyroid, which leads to either stroke-like symptoms, or presents as diffuse progressive symptoms of altered mental status, seizures, and cognitive dysfunction. Here we present a case of SREAT in a female with recurrent episodes.

Case History:

A 54-year-old female was brought in by ambulance after a prolonged tonic-clonic generalised seizure, requiring intubation with a Glasgow coma scale (GCS) of 6/15. Her past medical history was significant for autoimmune hyperthyroidism, epilepsy, smoker and alcohol excess. She did have previous ITU admissions with a similar episodes and after ruling out all possible causes of encephalitis; was treated as SREAT (based on EEG, thyroid history and cerebrospinal fluid (CSF) raised protein); with a 5 day course of i.v. methylprednisolone with good response. Her medications included propylthiouracil (PTU) 50mg twice-a-day and levetiracetam 1250 mg twice-a-day.

Investigations on Admission:

Her thyroid functions showed TSH 0.29, T4 23, T3 3.2. Her CSF showed normal biochemistry, microbiology and virology (except CSF protein raised at 0.7 g/l). Her CT head was Normal. Antibodies for other causes of encephalitis were negative. Her Anti-TPO antibodies and TSH-receptor antibodies were both positive. Her EEG was normal. Her all other blood workup was normal. See table for details

Treatment:

While sedated she continued to be in non-convulsive status. With the background of autoimmune thyroiditis/possible SREAT and above mentioned investigations, a trial of methylprednisolone 1 g i.v. was given for 5 days with dramatic improvement in her mental status and GCS within 24 hours. She was discharged on tapering dose of steroids.

Hb	**143**	**Na**	**140**	**Anti Hu abs**	**Negative**
WCC	16.02	K	4.8	NMDA receptor abs	**Negative**
Neutrophils	13.48	Urea	9	AMPA1 & 2 receptor abs	**Negative**
Platelet	300	Creatinine	90	LGI1 receptor abs	**Negative**
INR	1.0	eGFR	84	CASPR2 receptor abs	**Negative**
Anti Yo abs	**Negative**	**Anti Ri abs**	**Negative**	ENA abs screen	**Negative**
CSF Protein	**0.7**	**CSF glucose**	**6.1**	CSF WCC	1
CSF Red cell	**64**	**CSF organism**	**Nil**	CSF virology	Nil

Table 1: Laboratory data of the patient in Intensive Treatment Unit prior to initiation of methyl Prednisolone therapy (All units in SI NHS standards)

Conclusion:

Steroid-responsive encephalopathy associated with autoimmune thyroiditis or Hashimoto's encephalopathy is a rare diagnosis but the recognition of this uncommon condition is essential for early treatment.

References:

1. Castillo P, Woodruff B, Caselli R, et al. Steroid-responsive encephalopathy associated with autoimmune thyroiditis. Arch Neurol 2006;63:197–202
2. Sawka AM, Fatourechi V, Boeve BF, et al. Rarity of encephalopathy associated with autoimmune thyroiditis: a case series from Mayo Clinic from 1950 to 1996. Thyroid 2003;13:227–28