

STEROID-RESPONSIVE ENCEPHALOPATHY ASSOCIATED WITH AUTOIMMUNE THYROIDITIS (HASHIMOTO'S ENCEPHALOPATHY) COMPLICATED BY STATUS EPILEPTICUS SUCCESSFULLY TREATED WITH THERAPEUTIC PLASMA EXCHANGE

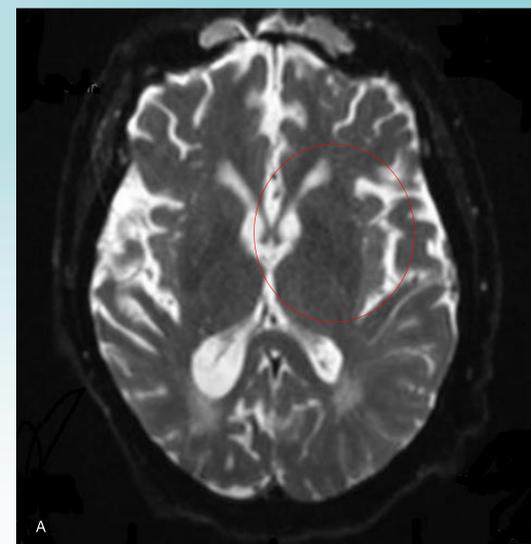
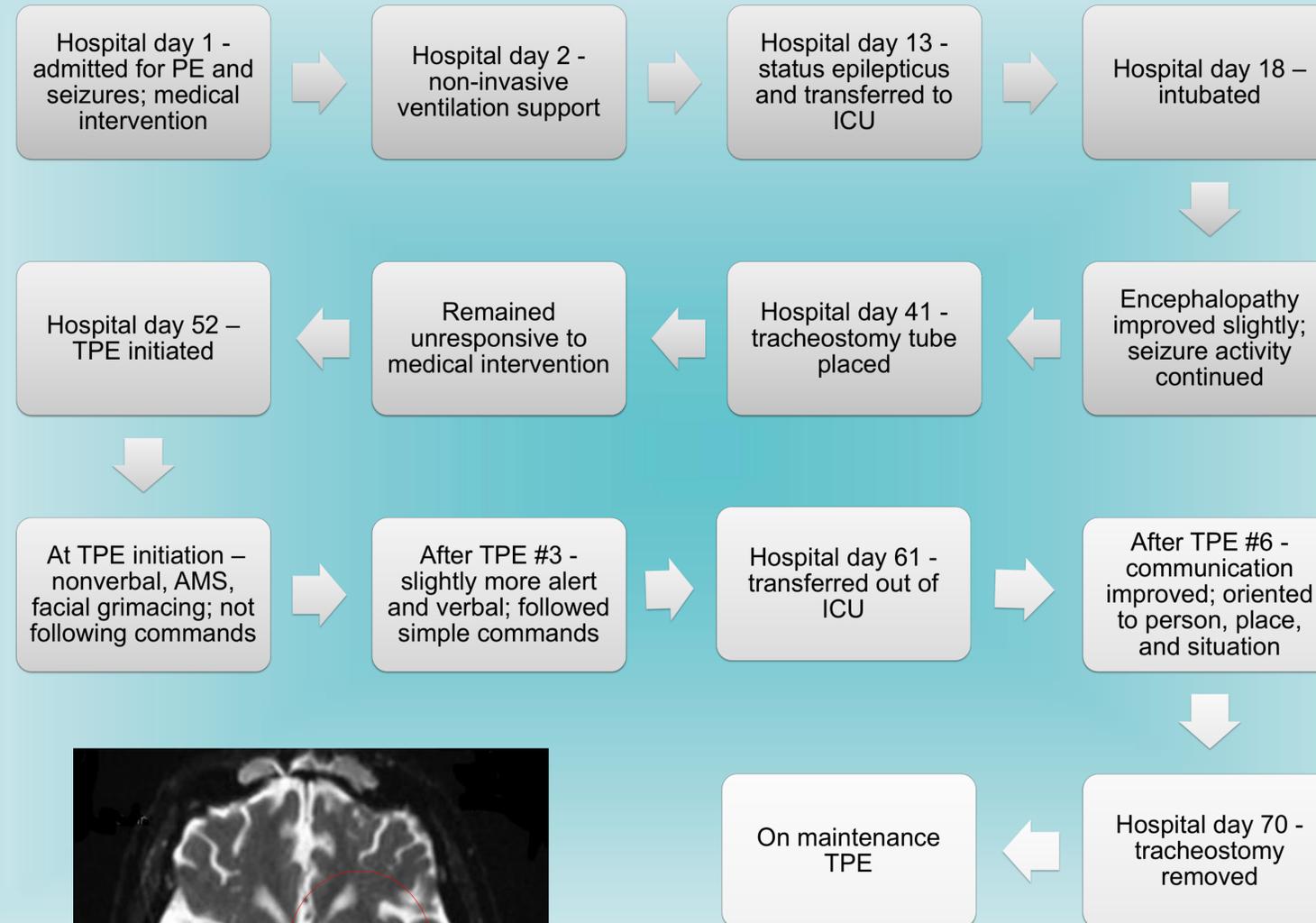
Background

- Steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) is a rare syndrome. It is characterized by encephalopathy of unknown etiology in the absence of alternative diagnoses and elevated anti-thyroid antibody titers.
- The mean onset of age is 40-50 years with a female predilection.
- The clinical presentation is variable, ranging from acute stroke-like episodes to the more common indolent course, which usually presents with cognitive impairments and/or psychomotor symptoms.
- First-line treatment includes high dose corticosteroids. There is increasing acceptance of an autoimmune etiology, with reported cases to date showing TPE to be efficacious.
- TPE is a category II indication with grade 2c recommendation for SREAT.

Case History

- The patient is a 70-year-old female with multiple comorbidities including COPD, focal seizures complicated by epilepsy partialis continua, recurrent encephalopathy, hypothyroidism, bipolar disorder, DMII, and SREAT associated with focal seizures responsive to high-dose corticosteroids.
- She was discharged from hospital one month prior to present admission for SREAT-associated seizures on outpatient corticosteroid therapy.

Case Description



A. MRI Brain (Hospital Day #35) showing mild diffusion edema involving the left insula and temporal and parietal cortex; mild ventricular enlargement secondary to brain atrophy. There was no significant interval worsening in subsequent imaging studies of the brain/head.

Discussion

- In this case, we describe a patient with a complex medical history including SREAT who was successfully treated with TPE, despite not having elevated antithyroid antibodies.
- The patient showed marked clinical improvement after three treatments of TPE, and ultimately no longer required ventilatory support.
- TPE was deemed to be an effective therapy for this patient with SREAT. The use of TPE as a long-term treatment for this patient may provide further insights into the impacts of apheresis therapy in this syndrome.
- In summary, SREAT is a rare neuropsychiatric syndrome with non-specific clinical presentations, and it is often associated with the high titers of antithyroid antibodies.
- While thyroid antibodies are considered a biomarker of the disorder, the antibody titers may not correlate with disease severity. Additionally, their role in the underlying disease pathogenesis remains uncertain; however, there is increasing acceptance of an autoimmune etiology in SREAT, supporting the efficacy of TPE and potentially long-term therapy.

References

1. Padmanabhan A 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 Eighth Special Issue. J Clin Apher. 2019; 34:171–354
2. Chong JY et al. Hashimoto encephalopathy: syndrome or myth? Arch Neurol. 2003; 60(2):164–171.
3. Olmez I et al. Diagnostic and therapeutic aspects of Hashimoto's encephalopathy. J Neurol Sci. 2013; 331(1–2):67–71.
4. Brain L et al. Hashimoto's disease and encephalopathy. Lancet. 1966; 2(7462):512–514.

	Previous Admission Value #1	Previous Admission Value #2	Hospital Day #1	Hospital Day #21
Anti-Thyroid Antibody <20 Intl_Unit/mL	<20	<20	---	<20
Thyroid Stimulating Hormone 0.358-3.74 UIU/mL	0.802	6,600	10,500	---