

Two-for-one TPE: Managing Steroid-Resistant Encephalopathy in a Patient with both Hashimoto's Thyroiditis and Hepatitis C Viremia

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INTRODUCTION

Encephalopathy associated with autoimmune thyroiditis (EAAT), a.k.a. Hashimoto's encephalopathy, is an uncommon neuropsychiatric condition associated with anti-thyroid antibodies often responding to steroids. (1)

EAAT is not listed in the American Society for Apheresis 2013 Indication Categories for Therapeutic Apheresis. However, in steroid-refractory cases, therapeutic plasma exchange (TPE) may yield benefit.

The mechanism of disease remains unknown due to a lack of clinical trials. Although cases of straight-forward EAAT treated with TPE are reported, few reports of patients with EAAT and confounding diagnoses or variable presentations exist.

Case Description

Pre-admission

A 49 year old man presents with several months of progressive neurologic symptoms including dysarthria, slurred speech, ataxia, and vertigo.

His past medical history is notable for:

- Alcohol abuse with withdrawal (now sober)
- Untreated HCV
- Remote cerebellar stroke

He is given steroids (40mg/day prednisone) and his symptoms improve. Due to a steroid taper for elective shoulder surgery his neurological symptoms return.

Reinstatement of steroid therapy fails to establish lasting recovery of his symptoms and the patient soon deteriorates, loses ambulation, develops chorea, and is admitted to UC Davis Medical Center.

OBJECTIVE

Our goal was to highlight the potential benefit of therapeutic plasma exchange in cases where multiple potential causative agents of disease exist.

METHODS

We report a rare case of steroid-resistant EAAT confounded by an elevated HCV viral load and history of ethanol abuse treated by therapeutic plasma exchange (TPE).

Multiple Diagnoses and Treatment

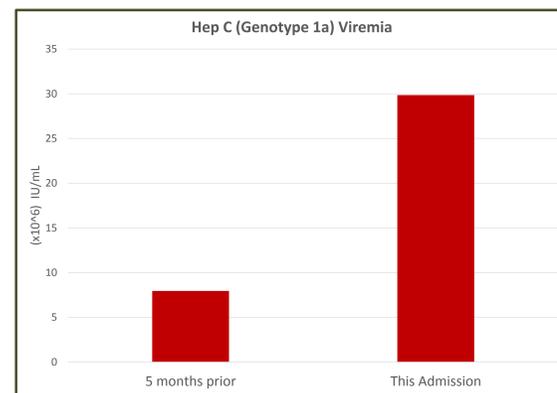


Figure 1. (Above) Hep C viremia: 275% increase or 3.75 times more than 5 months prior. Ref range 0 IU/mL.

Image 1. (Right) MRI Imaging of the brain showed cerebellar and cerebral mass atrophy by MRI

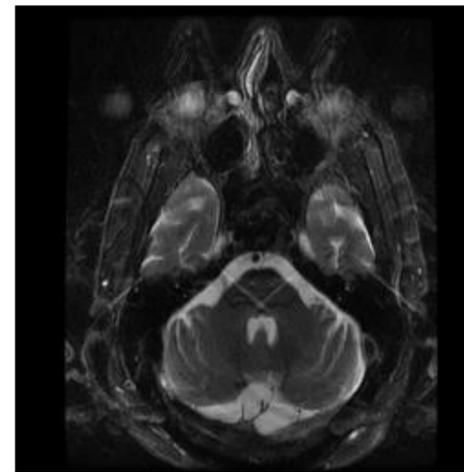


Figure 2. (Below, right) Elevated Thyroid antibodies are decreased following TPE and steroid treatment.

Hospital Course

Physical and neurological exam reveal seizure-like events with apparent loss of awareness.

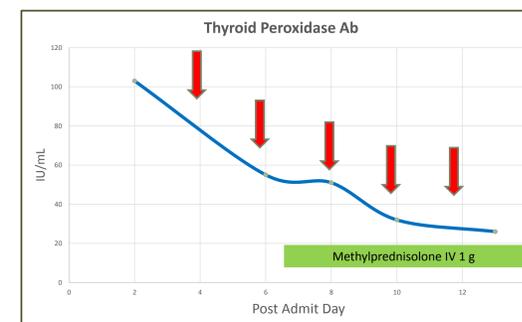
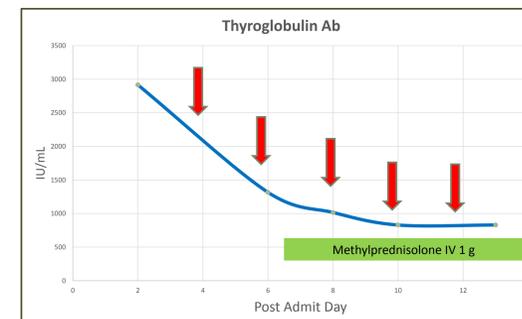
Laboratory results reveal that the patient has an elevated HCV viral load ~30 M IU/mL (Figure 1) which is 3.75 times greater (275% increase) than 5 months prior. His liver enzymes are within the normal limits.

The patient is also euthyroid with an initial elevated thyroglobulin antibody (2917 IU/mL) and thyroid peroxidase antibody (103 IU/mL) (Figure 2) with a negative paraneoplastic panel.

Imaging of the brain shows cerebellar and cerebral atrophy by MRI (Image 1).

TPE treatment with albumin 5% as total volume replacement is initiated with a 1 - 1.5 plasma volume exchange every 2 days for a total of 5 days. Adjunctive steroid treatment is started after TPE #2.

Serum antibodies decline over therapy (Figure 2).



↓ = Therapeutic Plasma Exchange

Reference ranges:

Thyroglobulin Ab ≤40 IU/mL

Thyroid Peroxidase Ab <35 IU/mL

Symptomatic Improvement

Pre-Treatment	Post-Treatment
Gross truncal ataxia: unable to sit forward in bed	Able to maintain sitting posture
Gross limb ataxia: unable to maintain contact between heel and shin during coordination exam	Able to perform finger to nose and heel to shin quickly and accurately
Marked dysarthria: having difficulty initiating speech	Able to initiate speech more reliably

Discharge

After therapy, the patient is discharged with significant improvement in his ataxia and dysarthria and regains his ambulation.

CONCLUSIONS

This case illustrates the potential use of therapeutic plasma exchange for clinical improvement of steroid resistant encephalopathy associated with autoimmune thyroiditis and elevated HCV viral load. The effect of high HCV viremia on neurologic function is still being investigated (2, 3).

Although the ultimate mechanism of disease in cases with multiple diagnoses is uncertain, the potential benefit of instigation of TPE to remove possible causative agents should be recognized; especially when standard therapies fail.

REFERENCES

- (1) Mitoma et al. Cerebellum & Ataxias (2015) 2:14
- (2) Fletcher & McKeating. Journal of Viral Hepatitis (2012)19, 301-3
- (3) Adinolfi et al. World J Gastroenterol (2015) 21(8): 2269-2280

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