Case Report

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Quick Response Code:

Website: www.ajts.org

DOI: 10.4103/ajts.AJTS_112_17

Therapeutic plasma exchange as a life-saving therapy in a suspected case of autoimmune encephalitis: A case report from a tertiary health-care center

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Abstract:

In half of the suspected autoimmune encephalitis (AIE) patients, no antibodies are identified despite extensive investigation. Therapeutic plasma exchange (TPE) is a potential first-line therapy for various subtypes of AIE. Here, we present a case of autoantibody-negative-suspected AIE, managed successfully with TPE after patient showed no response to steroids. A total of 5 sessions of TPE was done. One standard TPE procedure session was 1.2–1.5 plasma volume exchanges using 5% albumin as a replacement fluid. After five sessions, patient's clinical condition improved significantly, and a repeated magnetic resonance imaging after 5th cycle of TPE revealed a reduction in the areas of signal alteration. This was suggestive of regression of disease. Patient was discharged on 10th day of hospital admission. With early suspicion even in the absence of detectable autoantibodies, TPE plays an important role in the management of encephalitis.

Keywords:

Autoimmune encephalitis, corticosteroid, intravenous immunoglobulins, therapeutic plasma exchange

Introduction

Autoimmune encephalitis (AIE) is a group of encephalitis syndromes.[1] Patients generally have impaired memory and cognition over a period of days or weeks. There may be clues to specific causes on history of physical examination, but often these specific signs are absent. In about 50% of suspected autoimmune encephalitis based on clinical features, no antibodies are identified despite extensive evaluation.[2] The first line of therapy consists of steroids, therapeutic plasma exchange (TPE), and intravenous immunoglobulins (IVIG). TPE is a potential first-line therapy for various subtypes of AIE. Here, we present a case of autoantibody-negative suspected AIE, managed successfully with TPE after steroid failed to improve the conditions.

Case Report

A 39-year-old diabetic male presented with altered behavior, frequent twitching of the left angle of the mouth along with clonic/dystonic movement of the left hand. He had a history of low-grade fever for 1 day followed by headache and intermittent slurring of speech along with deviation of angle of mouth on the left side. There was no present history of nausea, vomiting, rash, features suggestive of vasculitis, and no previous history of similar events. These complex partial seizures rapidly

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Submission: 22-08-2017
Accepted: 06-12-2017

How to cite this article: Pandey PK, Agarwal N, Gupta M. Therapeutic plasma exchange as a life-saving therapy in a suspected case of autoimmune encephalitis: A case report from a tertiary health-care center. Asian J Transfus Sci 2018;12:173-5.

progressed to status epilepticus in few hours. As patient's general condition was poor, he was intubated and put on ventilator. His cerebrospinal fluid (CSF) picture showed neutrophilic pleocytosis, increased protein with normal sugar. The treatment was started on the lines of autoimmune and viral encephalitis. CSF HSV type I and II reports were negative on polymerase chain reaction. Although, antivoltage-gated potassium channels (VGKC) antibody were also negative, the left focal faciobrachial seizure along with dystonic posturing of the left upper limb was more in favor of AIE. Magnetic resonance imaging (MRI) finding on the 1st day revealed few altered area of signal intensity in bilateral cerebral hemispheres showing patchy enhancement along with enhancing leptomeninges. Patient was started with IV fluids, steroids, antibiotics, and antiviral and antiepileptic medicines. MRI on the 3rd day showed gyral swelling in bilateral cerebral hemisphere with restricted diffusion of the right fronto-temporo-parietal region. When compared to the last MRI, it also showed a significant increase in site and extent of involved areas. As there was no improvement with ongoing medications, the decision to start TPE was taken. We did a total of five sessions of TPE (initially one exchange every day for 3 days followed by 2 exchanges alternate days). All the TPE procedures were done with COM.TEC (Fresenius Kabi, Germany). One standard TPE session was 1.5 plasma volume exchanges using 5% albumin as a replacement fluid. Prophylactic administration of calcium gluconate (one ampoule diluted in 100 ml of 0.9% normal saline) was done for every 1000 ml of plasma exchanged. Informed consent was obtained from family members. All the procedures were done on double-lumen femoral dialysis catheter under aseptic precautions. Compliance to TPE was excellent as all the sessions were uneventful. After five sessions, patient's clinical condition improved significantly, and a repeated MRI was done after 5th cycle of TPE which revealed reduction in the areas of signal alteration in comparison to the previous MRI. This was suggestive of regression of disease [Table 1 and Figure 1]. The patient was discharged on the 10th day of hospital admission.

Discussion

AIE is considered to be a rare disease. In a multicentric prospective study in the UK, it was shown that in one-third of the patients' diagnosis of AIE was not conclusive and almost 50% of the patients were found negative for the common autoimmune antibodies suspected.[3,4] "California encephalitis project" also demonstrated the same finding. In this cohort of patients (a cohort of 1500 adults as well as children), confirmed suspected etiologies were not found in almost two-third of the study populations.[5]

The imaging and laboratory findings of many forms of autoimmune and infectious encephalitis are similar to AIE making it difficult to diagnose clinically.[6] For the diagnosis of definite autoimmune limbic encephalitis, all four of the following criteria have to be met: subacute onset, bilateral brain abnormalities on T2-weighted fluid-attenuated inversion recovery MRI highly restricted to the medial temporal lobes, CSF pleocytosis or electroencephalogram with epileptic or slow-wave activity on the temporal lobe, and reasonable exclusion of alternative causes.[1] Detection of autoantibodies is a definite diagnostic tool; however, there are case reports of patients presenting with features of AIE without any detectable antibodies but had full response with treatment with IVIG, steroids, and plasma exchange.[7] Early clinical suspicion is very important in these cases, as early treatment based on immunotherapy or TPE is essential. Our patient though was screened for anti N-methyl D-aspartate (NMDA) and anti-VGKC antibodies but not for other possible antibodies that may cause AIE. There are studies that have reported patients presenting with features of encephalitis, but negative for anti-LGI1 and anti-NMDA

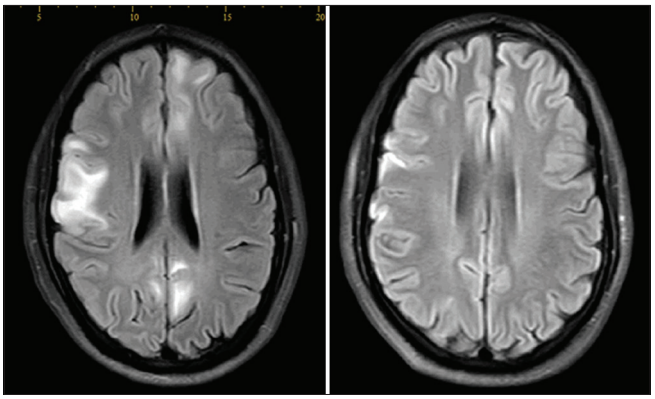

Figure 1: Contrast magnetic resonance imaging pre- and post-therapeutic plasma exchange

Table 1: Relevant findings on cerebrospinal fluid

Laboratory parameters	Results/Values
Total leukocyte count (mm^3)	80 cells/mm^3
DLC	Lymphocytes-10%, neutrophils-90%
Protein	79 mg/dl
Glucose	94/156 mg/dl
ADA	2.64 U/L
Indian ink, AFB staining, gram staining, cryptococcal antigen	Negative
Pan neurotropic viral panel (PCR)	Negative
HSV I and II (PCR)	Negative
Anti-NMDA and VGKC antibody (immunofluorescent assay)	Negative

VGKC = Voltage-gated potassium channels, ADA = Adenosine deaminase, HSV = Herpes simplex virus, PCR = Polymerase chain reaction, DLC = Differential leukocyte count, NMDA = N-methyl-D-aspartate, AFB = Acid-fast bacillus

antibodies had full response to immunotherapy with IVIG, steroids, and TPE.[8] All the three therapies have been termed as first-line therapies, and it is difficult to compare these therapies statistically. In our study, patient did not respond initially to steroids and IVIG but significantly improved after initiation of plasma exchange. Compliance to TPE was excellent in our patient. Patient did not have any adverse event during any of the five TPE sessions. Compliance to TPE is excellent if apheresis physician has good exposure of TPE procedure and management of its complications and careful monitoring of patient during procedure. Limited use of FFP in modern clinical practice has reduced significantly the complications related to TPE as most of the complications during plasma exchange procedures were associated with FFP infusion as replacement fluid. As a routine practice, we use 5% albumin as replacement fluid in all TPE procedures except in TTP and in chronic liver disease patients having deficiency of coagulation factors.

Conclusion

With early suspicion based on clinical features and rapid treatment initialization, even in the absence of detectable autoantibodies (due to lack of diagnostic kits and facilities) TPE plays an important role in the management of AIE, as it can even remove some hitherto undetected autoantibodies. Compliance to TPE was excellent in our case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

None.

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