Case Report

DOI: 10.7860/JCDR/2020/45701.14115

Role of Plasma Exchange in Steroid Resistant Neuromyelitis Optica with Loss of Vision

Transfusion Medicine Section

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ABSTRACT

Neuromyelitis Optica (NMO) is a demyelinating inflammatory disorder of the spinal cord and optic nerve. As per American Society For Apheresis (ASFA) guidelines, Therapeutic Plasma Exchange (TPE) is considered as a second-line treatment in patients with weak or no response to steroid therapy. A patient of NMO presented to the tertiary care institute with a sudden loss of vision in the right eye. The patient was resistant to steroid treatment and improved significantly with TPE. Hence, TPE may be an effective treatment modality in steroid-resistant NMO with vision loss.

Keywords: Optic neuritis, Therapeutic plasma exchange, Vision loss

CASE REPORT

A 38-year-old female patient presented with complaints of a sudden loss of vision in the right eye for one week. Difficulty in vision started with the blurring of vision, which gradually progressed over a few hours to complete blackout. She had similar two episodes in the past. The first episode was 10 years back and was treated with injectable steroids, after which she regained 70-80% of her vision. There was residual peripheral blurring in the right eye. The second episode occurred five years ago and was treated with methylprednisolone pulse therapy, which resulted in 60% of vision improvement. In the current episode, only light perception was present in the right eye. Magnetic Resonance Imaging (MRI) findings and the presence of anti-aquaporin 4 (AQP4) were suggestive of NMO.

She was started with an injectable steroid (methylprednisolone 1 g for five days) and oral steroid (Omnacortil 50 mg for five days). No improvement in the vision was observed with steroids alone and hence, TPE was planned. Informed consent was obtained from the patient and her attendant for TPE procedure and central line placement. A triple lumen jugular dialysis catheter was inserted under aseptic precautions. A total of five sessions were performed on an alternate day basis using intermittent cell separator (Haemonetics Mobile Collection System plus LN9000).

Details of plasma volume extracted are shown in [Table/Fig-1]. The mean plasma volume exchanged was 2626 mL (1.1 plasma volumes), with 5% albumin as a replacement fluid, maintaining a 100% fluid balance. Prophylactic administration of 10% w/v calcium gluconate infusion was given at the rate of 10 mL/litre of replacement fluid. The patient was discharged a day after her last TPE session with a plan of rituximab therapy after a gap of three weeks.

On further follow-up after a fortnight of her discharge, she reported clinically significant improvement in her vision in the right eye by recognition of colour and faces. On fundus examination, papillary atrophy was present in both eyes. Best-corrected visual acuity on Snellen's chart was 20/70, and on perimetry, her visual field index was 61% in the right eye and 74% in the left eye.

TPE	Plasma volume extracted (mL)	Outcome on the next day of TPE
Session 1	1902*	No significant improvement
Session 2	2600	No significant improvement
Session 3	2634	No significant improvement
Session 4	2967	The vision started improving and projection of light could be appreciated.
Session 5	3028	Blurred vision in the right eye Finger counting possible using right eye. Recognition of vertical and horizontal lines at two feet distance. No colour recognition attained.
At two weeks follow-up, the patient had significant improvement in her vision. Colour vision was regained.		
*Line blockage occurred at 1902 mL, line adjusted before next procedure		

[Table/Fig-1]: Session-wise outcome of Therapeutic Plasma Exchange (TPE).

DISCUSSION

The NMO is a demyelinating inflammatory disorder of the spinal cord and optic nerve. It is diagnosed with optic neuritis, acute myelitis, and minimum two of the three supportive criteria, including contiguous spinal cord MRI lesions extending three or more vertebral segments, brain MRI negative for multiple sclerosis, and presence of Anti-AQP4 [1]. Optic neuritis in NMO is quite severe, which can be unilateral or bilateral with an unpredictable response to treatment and a high propensity for relapse. Approximately, 90% of patients with NMO have a relapsing course, with nearly 50% of patients become legally blind [2]. The disease worsens by incomplete recovery with each acute attack.

High-dose steroids usually manage acute attacks, and, if symptoms fail to resolve, TPE is added as a second-line of treatment. The TPE acts by elimination of inflammatory mediators, autoantibodies, complements, and different cytokines from the blood. The TPE can be considered as a first-line of treatment in patients with severe attacks who have responded well during earlier attacks [3]. As per ASFA guidelines 2019, TPE is considered as the second-line treatment in patients with weak or no response to steroid therapy as it removes antibodies while steroids have only the immuno-modulatory effect [2].

Merle H et al., on 16 patients and Yoshida H et al., in a 22-year-old woman in their study found TPE (as an add on) with sequential corticosteroids was more effective than steroids alone as similar to findings in the present case [4,5]. Assogba U et al., found that if the steroids are given on the same day of TPE, less than 1% of the circulating steroids are removed by TPE due to a large volume of distribution and short half-life of steroids. However, steroids should preferentially be infused after each of the TPE sessions [6].

Further, TPE should be started as soon as possible in addition to steroids as shown in the study done by Kleiter I et al., where they found apheresis as the first-line therapy (p=0.047) and the time from onset of attack to start of therapy in days (p=0.014) were strong predictors for complete recovery [7]. Bonnan M et al., in the study on 60 NMO patients found that the baseline impairment and delay in plasma exchange were the two major reasons associated with the improvement [8]. In the present case, TPE was delayed by 10 days. Early initiation of TPE in this case, might have resulted in a better outcome.

CONCLUSION(S)

The TPE is a possible treatment approach for acute attacks of NMO with loss of vision. The combined therapy of steroids and early plasma exchange can be a practical and balanced approach.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jul 01, 2020
- Manual Googling: Sep 01, 2020
- iThenticate Software: Sep 23, 2020 (4%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. NA

Date of Submission: **Jun 30, 2020**
Date of Peer Review: **Jul 28, 2020**
Date of Acceptance: **Sep 01, 2020**
Date of Publishing: **Oct 01, 2020**