Plasma Exchange Therapy for Refractory Autoimmune Neurological Diseases

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Introduction

Immunosuppressive therapy is commonly used to treat autoimmune neurological disorders. Standard immunosuppression is frequently insufficient for complete recovery or preventing relapses in patients with refractory conditions. Other treatments are used to manage the disease in these patients. While the treatment of refractory cases varies depending on the disease, intravenous immunoglobulin, Plasma Exchange (PLEX), and immune-modulating therapies are commonly used. We focus on five autoimmune neurological disorders that were discussed at the 2018. Midlands Neurological Society meeting on PLEX in refractory neurology: Autoimmune Encephalitis (AE), Multiple Sclerosis (MS), Neuromyelitis Optica Spectrum Disorders (NMOSD), Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) and Myasthenia Gravis (MG). Inflammatory neuropathies are frequently difficult to diagnose, and while PLEX can be very effective in refractory autoimmune diseases, its ineffectiveness can be confounded. Numerous autoimmune disorders affect both the central and Peripheral Nervous Systems (PNS). They are distinguished by abnormal immune responses to antigens expressed in the nervous system. The presentation and severity of these diseases, however, can vary greatly.

Corticosteroids and immunosuppressive therapy (e.g., azathioprine, mycophenolate) are the standards of care for autoimmune neurological disorders in relapse. However, some patients are resistant to treatment (i.e. they do not respond to these treatments). Intravenous Immunoglobulins (IVIg) and plasma exchange are also commonly used to treat neurological disorders, with varying degrees of efficacy observed for various disorders. Immuno adsorption has recently emerged as a potential alternative to PLEX in the treatment of neurological disorders. While PLEX involves replacing the fluid with a blood solution such as fresh frozen plasma or albumin, Immuno adsorption is a blood purification process that uses a high-affinity adsorbent to remove humoral factors (i.e. disease-specific autoantibodies) from separated plasma. The Midlands Neurological Society meeting on PLEX in refractory neurology focused on five different autoimmune neurological disorders and the available treatment options, with a focus on the role of PLEX in patients with refractory disease. AE is generally characterized by impaired memory and cognition, frequently with seizures and/or a movement disorder, and occasionally with reduced consciousness or coma, but presentations vary greatly across sub-types. AE can be difficult to diagnose because its laboratory and imaging profiles are frequently normal. The incidence of AE is approximately 0.8/100,000 person-years. A small but significant proportion of AE patients are resistant to first- and second-line therapy and nearly all have residual cognitive deficits.

Steroids (e.g., methylprednisolone), IVIg, and PLEX, or immune adsorption are used as first-line treatments for AE, with corticosteroids being the most commonly used. Rituximab or cyclophosphamide are frequently used as second-line immunotherapy.

While corticosteroids are usually the first line of treatment, they are frequently insufficient to achieve adequate clinical improvements. In these cases, combining steroid treatment with PLEX or IVIg administration may be beneficial to achieve a synergistic effect. It has been demonstrated that concurrent PLEX and Intravenous Methylprednisolone (IVMP) treatment after transitioning to an IVIg regimen results in better short-term (1-2 months) outcomes than simultaneous IVIg and IVMP treatment without PLEX. PLEX is also thought to benefit AE by increasing the proliferation of autoantigen-specific B cells, making them more susceptible to immunosuppressants and chemotherapeutic agents. NMOSD is a CNS disorder that primarily affects the optic nerves (optic neuritis) and the spinal cord (myelitis), and is mediated in the majority of cases by aquaporin-4 Immunoglobulin G (IgG) Antibodies (AQP4-IgG). Myelin Oligodendrocyte Glycoprotein IgG Antibodies (MOG-IgG) are present in a proportion of patients with similar symptoms and are likely pathogenic. The incidence of NMOSD ranges from 0.053 person years to 0.4/100,000 person years. Acute relapses in MOG-IgG disease and AQP4-IgG NMOSD are currently treated similarly. Refractory NMOSD is defined as an incomplete or slow recovery from an acute attack despite corticosteroid treatment in this review. Refractory disease is defined as relapses despite treatment with corticosteroids, azathioprine (or mycophenolate), and rituximab for recurrent attacks. In some autoimmune neurological conditions, PLEX is standard first-line therapy. PLEX has also demonstrated high response rates in multiple settings following failure or relapse after corticosteroid treatment. However, diagnosing inflammatory neuropathies can be difficult, and the ineffectiveness of PLEX can be complicated by misdiagnosis. For example, POEMS is frequently misdiagnosed as CIDP, and while PLEX is widely used and effective in the treatment of CIDP, POEMS patients do not respond well to PLEX.

Not all refractory patients respond to PLEX even within the same disease. This difference could be attributed to different immunological aspects in different patients. Furthermore, correlations have been discovered between the efficacy of PLEX and various types of patterns within a single disease. One of the perceived advantages of IVIg over PLEX is its ease of use, particularly when PLEX is administered via a central line. However, using centrifugal PLEX machines rather than filtration-based ones allows for peripheral access in more than 70% of all cases, and can be as high as 90% for neurological patients. Peripheral access reduces the risk of PLEX-related vascular complications and significantly lowers the rate of catheter-induced infection. Peripheral access improves the safety and tolerability of PLEX and allows for an outpatient scenario. As previously stated, delaying PLEX initiation is associated with worse clinical outcomes. As a result, having proper planning and priority access for emergency patients makes this treatment option more accessible. Finally, both of these treatments have contraindications, such as hemodynamic instability, sepsis, hypersensitivity to albumin for PLEX and renal failure, hypercoagulable states, and immunoglobulin hypersensitivity for IVIg. More research is required to fully understand the biological mechanisms of PLEX in refractory neurological diseases, to better understand differences in response between diseases and subtypes, and to determine its role in therapy. Randomized clinical trials offer the most evidence to answer this question. Such studies, however, are difficult to conduct because the refractory disease is only a small subset of already rare conditions. The treatment of refractory neurological diseases can be difficult. We present a fair and balanced review, with an emphasis on the role of PLEX therapy. Further research into the mechanisms of action will aid in categorizing patients into groups that are more likely to benefit from the therapies discussed. Finally, peripheral access and seamless provision are critical components of using PLEX to treat refractory neurology patients.

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