Plasma exchange for steroid unresponsive Devic's disease

Sir,

A 15-year-old girl presented with weakness of both lower limbs, sensory loss below costal margin, and bladder disturbances for 3 days; loss of vision in the left eye for 3 months with past history of similar illness (3 months back) that showed partial improvement in motor power, sensory, and autonomic disturbances with steroid therapy without any improvement in vision. Physical examination showed optic atrophy in the left eye. Muscle power in lower limbs was 1/5 in proximal and distal groups. Pain sensation below D_7 level was diminished. There was an exaggerated knee-jerk reaction and extensor plantar responses. Magnetic resonance imaging revealed demyelinating changes in right centrum semi-ovale of the brain and demyelination in spinal cord from C_4 level to D_9 level; visual evoked potential from left eye showed prolonged P_{100} latency with reduced amplitude. She was again treated with I.V. steroids for 5 days followed by oral steroids. As there was no improvement after 11 days of oral steroids, patient was treated with plasma exchange. She received a total of five sessions of plasma exchange on alternate days. The amount of plasma removed was calculated based on the formula: 0.065 × body weight (1% hematocrit) and it was about 1.7 l, which was replaced with four packs of fresh frozen plasma (each containing 180-220 ml of plasma), 100 ml of 20% albumin, and 1 l of normal saline. After third session, muscle power in the lower limbs improved to 3/5 in the proximal group of muscles and 2/5 in the distal group of muscles. Muscle power remained unchanged till discharge, i.e., after 10 days of completion of fifth session of plasma exchange, and the patient was able to walk with support. She maintaines her muscle power at 10 months follow up.

Devic's neuromyelitis optica (NMO) is an inflammatory disease of the central nervous system characterized by severe attacks of optic neuritis and myelitis.[1,2] The specific antibody called NMO-IgG was found in more than 70% of these patients. This antibody is targeted against aquaporin-4 (AQP-4) water channel,[2] widely expressed in the optic nerves, the spinal cord, and the peri-ventricular regions. Traditionally, the main stay of the treatment of the acute attack or the index event has been high-dose intravenous steroid. Plasma exchange found a significant role in patients who did not respond to corticosteroids in Devic's disease in the recent literature.[3-5] The proposed mechanism of benefit being removal of the antibodies and immune complexes and the reported recovery is to the extent of 40%.[4] In plasma exchange, the filter pore diameter measures upto 0.2 μm resulting in filtration of substances upto a molecular weight of approximately 3×10^6 Da, such as circulating immunoglobulins and immune complexes directed at components of the central and peripheral nervous system. Plasma exchange has been shown to reduce IgG, IgM, and total complement levels by 63.4%, 68.9%, and 57.1%, respectively, after one exchange and 80.1%, 79.5%, and 59.7%, respectively, after five exchanges.[2] Here in, we report the importance of plasma exchange in the management of a patient of steroid unresponsive Devic's disease.

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Quick Response Code:	Website: www.indianjnephrol.org
	DOI: 10.4103/0971-4065.106050