

Abstract

Neuromyelitis optica spectrum disorder (NMOSD) or Devic's syndrome[6] is a rare autoimmune mediated demyelinating condition[5,6] that affects central nervous system. Autoantibodies are directed against aquaporin-4[5,6] water channels which present in foot process of astrocytes, which situate in higher concentration in optic nerve, brain stem and spinal cord. Therefore, demyelination mostly occurs in these sites resulted in optic neuritis, area postrema syndrome and transverse myelitis respectively[1]. Usually, patients present with transverse myelitis alone or optic neuritis alone or both together. The natural history of NMOSD is relapsing and progressing, that is, more the number of relapses more the chance of disability[1], which leads to permanent disability if not treated aggressively. Therefore, early initiation of definitive treatment is mandatory.

Acute episodes are treated with corticosteroids, specially intravenous methylprednisolone pulse followed by tapering course of oral steroids, therapeutic plasma exchange (PLEX) and intravenous immunoglobulins (IVIG). Disease modifying treatment would be continued afterwards. Azathioprine, mycophenolate mofetil and rituximab are the most commonly used disease modifying agents[1].