ORIGINAL RESEARCH PAPER

Paediatrics

AREA POSTREMA SYNDROME VARIANT OF NMOSD

KEY WORDS:

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BACKGROUND:
Neuromyelitis optica spectrum disorder is an autoimmune disease of CNS that mainly affects optic nerve and spinal cord characterized by optic neuritis, brainstem lesions, diencephalic lesions, cerebral lesions and area postrema syndrome.

Most patients demonstrate a relapsing course with incomplete recovery between attacks, which leads to progressive disability. Pathogenesis is related to the production of AQP4-IgG antibodies by plasmablasts which leads to disruption of blood brain barrier, complement mediated astrocyte injury and secondary demyelination. Diagnosis is made by characteristic clinical manifestations, presence of serum AQP4-IgG antibodies or specific neuroimaging findings and exclusion of alternative diagnosis. Treatment is pulse dose steroids and plasmapheresis during acute attacks and long term immunosuppression for attack prevention.

Prognosis of patients with NMO depends on presence of antiaquaporin antibodies. Seronegative patients with NMO usually have a monophasic course. Seropositive patients tend to have relapsing remitting course and the long term prognosis is poor.

Case Report:
A 7year old female child came with complain of on and off vomiting for last 1 month, weight loss approximately 2-3kg in last one month and 2-3 episodes of choking in last 24hrs.

Patient presented in compensated shock and compromised respiration in form of pooling of oral secretions with decreased bilateral air entry and jerky respiration. CNS examination was suggestive of hypotonia in bilateral lower limb with diminished reflexes with bulbar dysfunction with nystagmus. Patient required mechanical ventilation.

MRI brain with spine contrast was suggestive of well marginated non enhancing lesion involving posterior part of medulla oblongata and cervico medullary junction on both sides of midline possibility of demyelinating lesion and normal MRI spine. Differentials kept were CNS infections, ADEM, NMO, Clinically isolated syndrome and myelin oligodendrocyte glycoprotein antibody associated disorder. Clinical and MRI correlation clinched the diagnosis of area postrema syndrome, confirmed by strongly positive AQP4-IgG antibodies. Case was managed with pulse dose corticosteroid (methylprednisolone) and monoclonal antibody (rituximab) accordingly.