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Original Research Paper

Neurology

A RARE CASE NEUROMYELITIS OPTICA SPECTRUM DISORDER

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ABSTRACT Neuromyelitis Optica spectrum disorder or Devic's disease is an autoimmune disease characterized by by acute inflammation of the optic nerve and spinal cord. A relapsing disease course is common, especially in untreated patients. In more than 80% cases it is caused by immunoglobulin G autoantibodies to aquaporin 4 the most abundant water channel protein in the central nervous system. Episodes of optic neuritis and myelitis can be simultaneous or successive.

KEYWORDS :

INTRODUCTION:

NMOSD selectively affects the optic nerve, spinal cord, and brain stem. This selectivity can be explained by the increased amount of AQP4 in these structures, and, furthermore, by the increased amount of AQP4 aggregates in the optic nerve and spinal cord.

NMOSD is caused by auto-antibodies targeting aquaporin 4 (AQP4), a channel protein in the cell membrane that allows water to pass through the membrane. AQP4 monomers form tetramers and the tetramers aggregate. AQP4 is found in astrocytes, which are the basis for the glymphatic system. Thus, NMOSD involving AQP4-IgG can be considered an astrocytopathy or autoimmune astrocytic channelopathy, since the astrocytes are semi-selectively destroyed.

Case Report:

A 21 year old female was brought to the Emergency department with complaints of giddiness , difficulty in walking and headache since 3-4 days. This was also associated with swaying to either sides in sitting position without support.

On examination patient was short statured moderately built conscious oriented to time place and person. She had a pulse rate of 88 beats/min, blood pressure of 100/70mmhg on right arm in supine position , RR 18 breaths/min, Spo2 98% at room air with RBS 119mg/dl. On CNS examination patient was conscious oriented with intact memory no motor deficit , no cranial nerve deformity with impaired co-ordination and ataxic gait having no sensory loss with normal deep tendon reflex. Rhomberg's sign was positive for the patient , she had nystagmus along with dysdiadochokinesia , past pointing, impaired knee heel test on left side. Cardiovascular system, respiratory system and per abdominal examination was unremarkable.

On clinical examination lesion was suspected to be in cerebellar region, hence an MRI brain plain was done , MRI brain plain done showed demyelinating lesion extending from the cerebellum to the cord having differentials of
1.Infective/Inflammatory aetiology
2.Transvesre myelitis
3.Spinal cord Infarction

Further contrast study was done along with c-spine which showed hyperintensity in the medulla oblongata and cervical cord up to C3 vertebral body level.

CSF examination done revealed a total cell count 15, Protein 109.48 sugar 83.1, Alb 0.03, CSF picture was s/o inflammatory aetiology,

Serum NMO antibody was sent for the patient which turned out to be positive.

Patient was started on MPS and then was given IvIg (100gms), later converted to oral steroids and Tab Azathioprine,.

Patient showed significant clinical improvement over a period of 5-6 days, she was able to sit up on her own and be able to maintain posture without support.

She was discharged after 2 weeks on Oral steroid and Tab Azathioprine.

DISCUSSION:

NMOSD is an autoimmune disease that causes severe demyelination, especially in the optic nerve and spinal cord with typical clinical manifestations of acute optic neuritis and transverse myelitis. It has been associated with serum AQP4-IgG [1], [2], [3], [4], [5], [6].

Eugene Devic (1858-1930) who first introduced the French term acute neuromyelitis optic "neuro-myélite optiquë aiugë" to show a new syndrome characterised by myelitis and acute optic neuritis. Lennon and Wingerchuck (2004) detected the presence of IgG-NMO or IgG-AQP4, the specific antibodies that distinguish NMOSD from MS [1], [7].

NMOSD is a rare syndrome with less than 1% demyelinating disease and the incidence varies in various countries. In general, the incidence of NMOSD ranges from 0.05-4.4 per 100,000 [1], [6]. It generally occurs in Asian, African and Hispanic descendants [6]. It is more dominant to attack female than the man with a ratio of 3-9: 1, and in adults age between 30.5 and 55.2 years, but can also occur children and elderly [1], [6], [8]. NMOSD cases have been reported in a 3-year-old and a 90-years-old [7].

Till date, the pathogenesis of NMOSD is still not fully understood [7], [9]. Antibodies to AQP4 play a key role in the pathogenesis of NMOSD. AQP4 is a water channel that is mostly expressed on podocytes of astrocytic cell membrane forming part of the blood-brain barrier [1], [7], [9].

Cerebrospinal fluid examination in positive NMOSD patients with AQP4-Ab can be found moderate with normal pleocytosis in about 40% of cases during acute recurrence. Oligoclonal bands (OCB) are usually not found, the intrathecal polyspecific antiviral immune response against-Measles, Rubella and Varicella-Zoster viruses (MRZ reaction) are negative, and increased glial fibrillary acidic protein and neurofilament heavy chain (nfH) are commonly found [8]. In

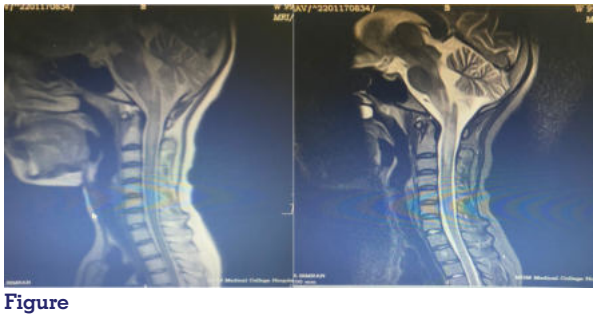
this patient, the cerebrospinal fluid examination gave an impression of inflammatory aetilogy.

The MRI examination on the spinal cord in NMOSD have typical features of longitudinal extensive transversal myelitis (LETM), a lesion that extends over 3 or more segments of the adjacent spinal cord [1], [5], [13], [14]. MRI of the optic nerve can be seen as hyperintensity in optic neuritis and tends to have more posterior involvement of the optic nerve. Brain MRI features can vary, such as normal or periependimal lesions surrounding the ventricular system, dorsal brain stem lesions bordering the fourth ventricle, periependymal lesions that surround the lateral ventricles, white matter hemispheres, lesions involving the corticospinal tract, non-specific lesions and enhancing lesions [14], [15]. In this patient, an MRI examination of the spinal cord showed with the features of myelitis involving cerebellum extending till C3while a brain MRI examination was normal.

Therapy in NMOSD consists of acute exacerbation phase therapy to reduce the risk of relapse and long-term care [10], [20]. Treatment options for prevention of relapse include oral corticosteroids, immunosuppressant therapy, TPE, immunomodulatory therapy, and other new therapies. Azathioprine is the main treatment option for preventing relapse at a dose of 75-100 mg/day and is more effective when combined with oral prednisolone (1 mg/kg/day). Evaluation on haematology and long-term side effects including gastrointestinal complaints, leukopenia, infections, allergies, haematological general disorders, and congenital disorders are required [10].

Corticosteroids are the main choice in the acute phase. Intravenous methylprednisolone is administered with a dose of 1-1.5 grams in 3-5 days [1], [2], [6], [7], [8]. Intravenous dexamethasone at a dose of 5 mg can be also a choice of corticosteroids [21]. Therapeutic plasma exchange (TPE) can be considered if the patient's condition does not improve or neurological symptoms worsen. Therapeutic plasma exchange dosage is carried out by giving 5-7 cycles in a period of 2 weeks with a dose of 1-1.5 plasma per time TPE [1], [6], [8], [9], [10], [20]. In this case plasma exchange was not possible due to cost issues hence IvIg was given.

The probability of recurrence of disease activity is greater than 90% [21]. Attacks on NMOSD can be very severe, NMOSD can be life-threatening if the lesion extends to the cervical spinal cord and brain stem because it has the potential to cause respiratory failure [1], [7].



Figure

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