NEUROMYELITIS OPTICA- A RARE CASE

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PRESENTATION OF CASE

A 48 yrs. old female patient presented with complaints of unexplained hiccups & vomiting since 5 months. She complained of tingling sensation and numbness all over the body & imbalance while walking. She gave history of multiple joint pains & back pain. She had consulted various specialists before she had visited us but with no relief of symptoms. She also had diminution of vision in the right eye, associated with pain which was insidious in onset & progressive since 2 months. History of intermittent fever was present. No complaints of urinary incontinence. Medical history of the patient included hypertension for which she was on Tab. Telmisartan 40 mg OD. No Family history of neurological diseases, autoimmune disease was noted. On examination, patient was conscious oriented to time, place & person. She had muscle power grade 5 on the Medical Research Council (MRC) scale in all the four limbs, with normal reflexes.

Image 1. Fundoscopy Revealing Right Eye Papilloedema

In MRI Brain, Hyper intense signals in pons. Optic Neuritis – Mild thickening of optic nerves with perineural hyper intense signal on either side. Mild FLAIR hyperintense signal in the bilateral frontal periventricular white matter – may be demyelinating.

PATHOLOGICAL DISCUSSION

Patients CBC, Electrolytes, TFT & Urine routine were within normal range. Patient had hypoproteinaemia with a low albumin. The patient had a normal antinuclear antibody (ANA) titre. Antibodies to double-stranded DNA were undetectable. p-ANCA & c-ANCA titres were normal. Cerebrospinal fluid analysis was unremarkable, with normal protein, glucose, no oligoclonal bands and no pleocytosis. Testing for Antibodies to HIV, Hepatitis B & Hepatitis C were negative. Fundoscopy revealed right eye papilloedema.

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Image 2.1. MRI Brain

Image 2.2. MRI Brain

Image 2.3. MRI Brain

In MRI Spine, we observed Straightening of lordosis appears to be due to paraspinal muscle spasm. Hyper intense signals in cervico-medullary junction extending posterosuperiorly into Area Postrema. Mild disc bulges at C4-C5 & C5-C6 levels with osteophyte indenting on anterior subarachnoid space without significant nerve root compression & with mild canal narrowing. Diffuse asymmetrical bulge at C6-C7 level indenting anterior subarachnoid space & abutting left nerve root, mild canal & left foraminal narrowing seen.

Image 3.1. MRI Spine

Image 3.2. MRI Spine

Image 3.3. MRI Spine

Image 3.4. MRI Spine

The term Neuromyelitis optica (NMO) was proposed in 1894 by Gault and Devic.¹ Neuromyelitis optica (NMO), also known as Devic's syndrome or Devic's disease, is an immune-mediated demyelinating central nervous system disease that preferentially affects the spinal cord and optic nerves.² Initially NMO was considered as a variant of Multiple Sclerosis. But then many Studies have shown it to be a distinct entity. NMO is a Humoral mediated autoimmune disease whereas Multiple sclerosis is a CD4 T + cell mediated autoimmune disease. MS lesions usually do not exceed the length of one vertebral corpus in the spinal cord, whereas in NMO, the lesions involve at least 3 segments, and may also include cavitations.³ It is important to differentiate NMO from MS early because NMO has a more severe morbidity than MS. Neuromyelitis optica (NMO) immunoglobulin G (IgG), is directed against aquaporin-4 (AQP4) which is present in the Astrocytic Foot process. Sensitivity and specificity of NMO IgG for NMO is 91% and 100% respectively.⁴ The Autoantibody initiates various events which lead to severe inflammatory reaction & tissue destruction.

NMO is an aggressive inflammatory disorder characterised by recurrent attacks of Myelitis & Optic Neuritis. It is more common in women than men. The disease typically begins in adulthood but can arise at any age. When autoantibody binds against the Aquaporin 4 channel there occurs hyper permeability at the blood brain barrier, perivascular inflammation, astrocyte damage and inflammation which form the main components of pathogenesis in NMO.⁵ Patients with positive Anti NMO antibodies usually have a recurrent & aggressive course of disease. Whereas patients with negative Anti NMO antibodies are more likely to have a Monophasic course.

DISCUSSION OF MANAGEMENT

Therapies with proven efficacy in Multiple Sclerosis have not appeared to be useful in NMO. Acute attacks are usually treated with high dose glucocorticoids. Plasmapheresis is used as an empirical therapy in patients not responding to glucocorticoid therapy.^{6,7} Few regimens have been proposed to prevent relapses which include usage of Mycophenolate Mofetil, Rituximab⁸ or a combination of Glucocorticoids plus Azathioprine. Early diagnosis and treatment are quite important in NMO as in untreated patients it is quite disabling as time progresses.

The patient was started on IV Methyl prednisolone, Azathioprine & Oxcarbazepine. Later started on Oral Prednisolone for maintenance. There was a bit of symptomatic relief to the patient with partial recovery of vision. Later she underwent Rituximab therapy.

FINAL DIAGNOSIS

Area Postrema Syndrome was diagnosed based on both symptoms and lesions in MRI. Anti NMO antibodies were detected in the serum. Anti-Myelin Oligodendrocyte Glycoprotein antibodies were not detected. Diagnosis of NMO was made according to the 2015 NMOSD Criteria by Wingerchuk.⁹

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