Case Report

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Devic's Disease: The Confusing CNS Disease

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Abstract

Neuromyelitis optica spectrum is order (NMOSD) is a rare auto-antibody mediated disease of the central nervous system involving the central nervous system. The symptoms should alert the clinician to the diagnosis. Prompt diagnosis in the emergency department plays a crucial role as delay in diagnosis and initiation of management will lead to increased chances of morbidity and mortality. Approximately 80% of NMO patients have auto-antibodies against aquaporin-4, a water channel expressed on astrocytes. In this case report, we discuss a patient with NMO disease, the diagnosis and its management.

Keywords: Devic's Disease; Neuromyelitis Optica; Hemiparesis; Cerebrovascular Accident; Auto Antibody Aquaporin.

INTRODUCTION

The term neuromyelitis optica was first described by Eugène Devic and his doctoral student Fernand Gault in 1894 (hence the name).[1] It was earlier thought to be a variant of "optico-spinal" multiple sclerosis. In the year 2004 the causative antigenic target, the aquaporin-4 water channel was identified.[2] Recently, another antigenic target, myelin oligodendrocyte glycoprotein (MOG) was identified in these patients.[3] The involvement of the central nervous system is identified by bouts of intractable vomiting and hiccoughs (area postrema syndrome), severe optic neuritis and long segments of spinal cord inflammation (my elitis). In this case report, we discuss the difficulty in diagnosis, review of literature and the management of Neuromyelitis optica/ Devic's disease in detail.

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CASE

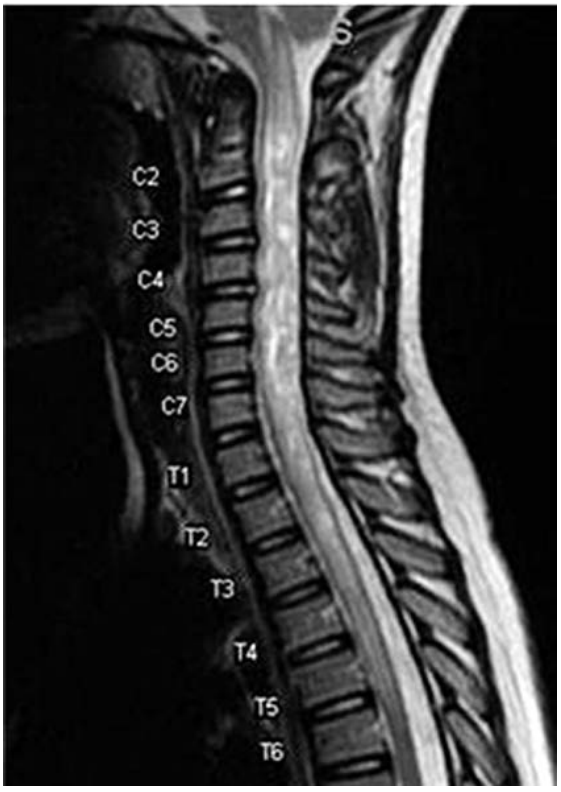
A 50 year old male, was brought to the emergency room with complaints of gradual on set of left side limb weakness (upper and lower limb) since the last 2 days. The patient today complained of reduced visual senses in left eye. His past medical history was significant for diabetes and hypertension. He was diagnosed with seizure is order 2 years back and was on antiepileptics measures since then, not missing any dose. While in the emergency room, the patient started having vomiting episodes, with

no air way compromise.

Patient was provisionally diagnosed with cerebrovascular accident and brain Magnetic resonance imaging (MRI) was done, which came out to be normal. Based on the MRI findings, the patient was tagged as a case of Transient Ischemic attack (TIA). he was admitted to the critical care unit for further evaluation. The limb weakness did not settle and improved, now involving the urinary bladder. Contrast MRI of the spine revealed "hyperintensity in the cervical and thoracic spinal cord suggestive of demyelination". The patient was started on a high dose of steroid (1 gram methylprednisolone). The condition deteriorated and the patient complained of dyspnea on day 4 of the disease on set. Patient serum was sent for aquaporin-4 antibody detection, which came out to be positive. He was now given plasma exchange therapy along with steroid dose and physiotherapy. Interestingly, the limb weakness improved and vomiting episodes settled after 2 cycles of plasma exchange therapy. A total of 6 plasma exchange cycles were given.

On day 9, the patient was discharged home and was called for routine physiotherapy sensations.

After 2 month follow-up, the patient's condition improved, the limb weakness not improved and the patient was able to walk with support and void urine byself with out any dependents.



DISCUSSION

The patient was diagnosed to have Neuromyelitis optica (also called as Dervic's disease). It is caused by the antibody AQP4-Abs (aquaporin-4) which are abundantly expressed in the central nervous system (astrocytes), opticnerve, spinal cord segments, brainstem.[4] The symptoms of the disease will depend on the area involved like optic neuritis (optic nerve) (spinal cord) longitudinally extensive transverse myelitis, acute brainstem syndrome (brain stem), area postrema syndrome (dorsal medulla) and acute diencephalic syndromes (thalamus/hypothalamus). Longitudinally extensive transverse myelitis (LETM) is the most specific presentation of NMOSD.[5] Although the term neuromyelitis optica is suggestive of exclusive spinal cord and optic nerve inflammation, the brain involvement might be seen in almost 60% of patients, though the majority of changes are non specific like hemipares is, encephalopathy and seizures.[6]

In the case of positive AQP4-Ab status, with one of the following, confirms the diagnosis: acute myelitis, opticneuritis, Acute brainstem syndrome (ABS), symptomatic narcolepsy or acute diencephalic clinical syndrome, symptomatic cerebral syndrome with NMOSD - typical brain lesions, Area postrema syndrome (APS): episode of other wise unexplained hiccups or nausea and vomiting. The acute treatment of an NMO consists of high dose steroids (HDS), plasma exchange (PLEX; 5 cycles) should be commenced[7] Long term therapy consists of prolonged immune suppression, the most common are: mycophenolate mofetil (MMF; 2–3grams/day) and azathioprine (AZA; 2.5–3mg/kg)[8], others like methotrexate, intravenous immunoglobulins tocilizumab, cyclo-phosphamide, tacrolimus, Medications to be avoided like natalizumab, beta-interferon and fingolimod as and ciclosporin.[8,9] When the yare involved in treatment of multiple scleros is and increase the Chances of neuro myelitisoptica.[9]

CONCLUSION

NMO is a rare butlife threatening disorder which needs prompt diagnosis and early initiation of treatment with high doses of steroids and plasma therapy. The early diagnosis and Treatment the loss Prevntion mortality and morbidity. The cause of NMO is Aquaporin-4, expressed in the central nervous system. A newer antibody myelin oligodendrocyte glycoprotein has been recently found to beacaus ativeagent. Life long immune

suppression is required in Patients with NMO disease.

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