A Rare Case of Devic's Disease Presented with Reversible Optic Neuritis

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Abstract- Neuromyelitis optica or Devic’s disease is a rare inflammatory demyelinating autoimmune disease of the central nervous system which affects the spinal cord and optic nerves and usually associated with increased disability and morbidity.

Keywords– Devic’s, Demyelinating

INTRODUCTION
Neuromyelitis optica (NMO), also known as Devic’s disease, is a serious, idiopathic and inflammatory demyelinating syndrome of the central nervous system (CNS). This devastating disease is classically characterized by selective and severe attacks of the optic nerve and spinal cord with or without recovery, and is potentially fatal. It causes sudden loss of vision in one or both eyes, varying degrees of weakness or paralysis in the legs or arms, loss of sensation and/or bladder and bowel dysfunction. Although originally considered as a variation of multiple sclerosis (MS), clinical, radiological, pathological and especially immunological data have led to a novel definition of this clinical entity. Early differentiation between NMO and MS is particularly important because the course of NMO is more severe and the treatment strategies for attack prevention are different. Immunotherapy approved for MS treatment is ineffective and appears to aggravate NMO. Immunosuppressive therapy is the treatment of choice for reducing NMO relapses. The association of NMO with the specific biomarker AQP4-IgG is considered as an additional criterion supporting the diagnosis (sensitivity and specificity for NMO respectively 91% and 100%)

CASE REPORT
We present here a case of a 32-year-old female patient, coming from a poor socioeconomic background, complaining of weakness in the lower limbs and urine retention. The medical history of our patient includes a gradual loss of vision over period of 3 months, but not losing vision completely. No neurological diseases were reported in the family. The patient was hospitalized before in 2022 due to weakness in the lower limbs and blurring of vision. MRI scan of the brain showed no lesions. The patient’s financial constraints have been a great hurdle in the diagnosis and management of her condition, but ever since then she has experienced varying degrees of paralysis in the upper and lower limb, urinary retention and constipation every year and has been hospitalized to treat these symptoms. Before being admitted here, she has had admission 4 times previously in Government Hospitals with no definitive diagnosis. Previous drug history was unknown. No any documents were available. 15 days ago, she presented to the hospital complaining of progressive weakness in her legs and urine retention. Neurological exam showed grade 5 strength in upper limbs and grade 3 strength in the lower limbs with hyper-reflexia. The patient was vitally and hemodynamically stable but her MRI Contrast Brain Revealed Multiple altered signal intensity areas in B/L frontal corona radiata, centrum semiovale and periventricular white matter appearing hyperintense on FLAIR images without true diffusion restriction. Three Differential Diagnosis were made on the basis of MRI Brain Contrast Results (1) ADEM (2) NMO (3) MOGD. CSF examination was done to rule out MS. Neuromyelitis Optica was suspected. Advanced immunological tests were performed at Pacific institute of medical sciences confirmed the presence of NMO (Aquaporin 4 Antibodies and Diagnosis of NMO was made. Improvement in her symptoms was seen after she was started on 5mg of prednisolone and during follow up azathioprine was added for prophylaxis for Relapse.
T2-weighted MRI of the thoracic spine revealed high-intensity signal changes in the spinal cord involving both halves extending from D2 to D9. Similar changes were noted in the cord at L1 L2. Diffuse edematous signals in entire cervico-dorsal cord with mild post-contrast enhancement was observed on T2 weighted MRI of the thoracic spine.
DISCUSSION

Through this study, we highlight the complexity of diagnosing neuromyelitis optica and the treatment challenges when socioeconomic factors are a hurdle. Because of lack of funding in public hospitals, plasmapheresis and the advanced immunological test of NMO was delayed. Patients’ family was also reluctant to go through these tests and procedures due to financial constraints. NMO is quite similar to the opticospinal form of MS hence it can be misdiagnosed, however, the course of NMO is more acute. Optic neuritis is severe with poor prognosis, our patient had a very severe case of optic neuritis and lost her vision reversibly.

NMO Ig antibody is a highly specific diagnostic marker for NMO. NMO is a rare pathological entity with neurological manifestations. Previous literature suggests that in patients’ with SLE or Sjogren’s syndrome positive NMO IgG is not a secondary effect but...
possibly these patients have two autoimmune disorders (10,11). In other studies, the presence of NMO IgG has been considered as a predictor for the manifestation of NMO spectrum disorder and predicts relapse of the disease. Co-existence of NMO is other autoimmune disorders either organ-specific or non-specific results in a poorer prognosis. The current diagnostic criteria for NMO requires the presence of optic neuritis and acute transverse myelitis along with supportive criteria which include aquaporin 4 seropositivity normal brain magnetic resonance imaging or not meeting the criteria for multiple sclerosis and longitudinally extensive cord lesion extending over 3 or more vertebral segments.

CONCLUSION
Neuromyelitis optica (NMO; also known as Devic syndrome) is a clinical syndrome characterized by attacks of acute optic neuritis and transverse myelitis. In most patients, NMO is caused by pathogenetic serum IgG autoantibodies to aquaporin 4 (AQP4), the most abundant water-channel protein in the central nervous system. In a subset of patients negative for AQP4-IgG, pathogenetic serum IgG antibodies to myelin oligodendrocyte glycoprotein, an antigen in the outer myelin sheath of central nervous system neurons, are present

REFERENCES: