

Seropositive Neuromyelitis Optica (Devic's Disease): A Rare Case Report

MD. GOLAM-UR-RAHAMAN,¹ AZIZUL HAQUE,¹ SHUVRA ADHIKARY,² CHOWDHURY TAMANNA TABASSUM,² AHMEDUL KABIR,³ SUDIP RANJAN DEB⁴

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. The purpose of this case report is to present this rare disease and focus on diagnostic criteria. NMO is often misdiagnosed as Multiple Sclerosis. The discovery of neuromyelitis optica (NMO) immunoglobulin G (IgG), directed against aquaporin-4 (AQP4), has dramatically changed the clinical definition of NMO and is important in the diagnostic criteria of this disease. Also longitudinally extensive spinal cord lesions (3 or more spinal segments) are characteristic of NMO. Here we report a case of a 14 year old girl presented with weakness of all four limbs with left sided complete blindness. NMO was diagnosed because of characteristic MRI finding and positive Aquaporin 4 IgG positive.

Keywords: Neuromyelitis optica, Devic's disease, Immunoglobulin G (IgG), aquaporin-4 (AQP4).

Introduction:

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.¹ It is controversial whether NMO is a variant of multiple sclerosis (MS) or is a distinct disease. The advent of NMO antibody has permitted clearer differentiation between NMO and MS. It has increased the accuracy of diagnosis, allowing differentiation of the two disorders in many cases where it was not previously possible. The clinical features of NMO are female-predominance, negative oligoclonal IgG band, and longitudinally extensive and centrally located lesions in the spinal cord.² This disease is more prevalent in Black, Asian, and Indian populations. It is important to differentiate NMO from MS early because NMO (especially relapsing NMO) has a more severe morbidity than MS and standard MS-modifying therapies may not be effective on NMO. Here

we report a 14 year old girl with seropositive Neuromyelitis Optica (AQP4 IgG positive).

Case Report:

A 14 year old girl presented to us with bilateral dimness of vision for 3 and half months and progressive weakness in all four limbs for 20 days. Dimness of vision was more severe in left eye and progressed to complete blindness. Weakness started from right upper limb and gradually progressed to all four limbs over next day, which made her walking difficult and she became bedridden. She had been previously well with no known chronic medical conditions, like hypertension or diabetes mellitus, and had no preceding vaccinations or viral infections noted. There was no history of trauma, bony tenderness, vertigo, convulsion, facial pain, headache, fever, diarrhea, jaundice, joint pain, oral ulceration, drug abuse, allergy, sexual exposure or contact with TB patient. Her family history does not suggestive of such type of illness.

On Examination she was ill looking and mildly anemic. Examination of the nervous system there was 2nd and 3rd nerve palsy on left eye evident by complete loss of vision, loss of light reflex and dilated pupil. On fundoscopy revealed optic atrophy on left eye and early changes of optic atrophy on right eye. Muscle tone was increased on all four limbs. Muscle power of both limbs of right side were 2/5 and both limbs of left side were 3/5. All jerks of both upper and lower limbs were exaggerated with bilateral extensor plantars. Patellar and Ankle clonus was present. There was impaired sensation of pain, touch, temperature upto T₁ level.

1. Indoor Medical Officer, Department of Medicine, Dhaka Medical College & Hospital, Dhaka.
2. Medical Officer, Department of Medicine, Dhaka Medical College & Hospital, Dhaka
3. Associate Professor, Department of Medicine, Dhaka Medical College & Hospital, Dhaka.
4. Assistant professor, Department of Medicine, Dhaka Medical College & Hospital, Dhaka.

Corresponding author: Dr. Ahmedul Kabir, Associate Professor, Department of Medicine, Dhaka Medical College & Hospital, Dhaka. e-mail: ahmedul_986@yahoo.com.

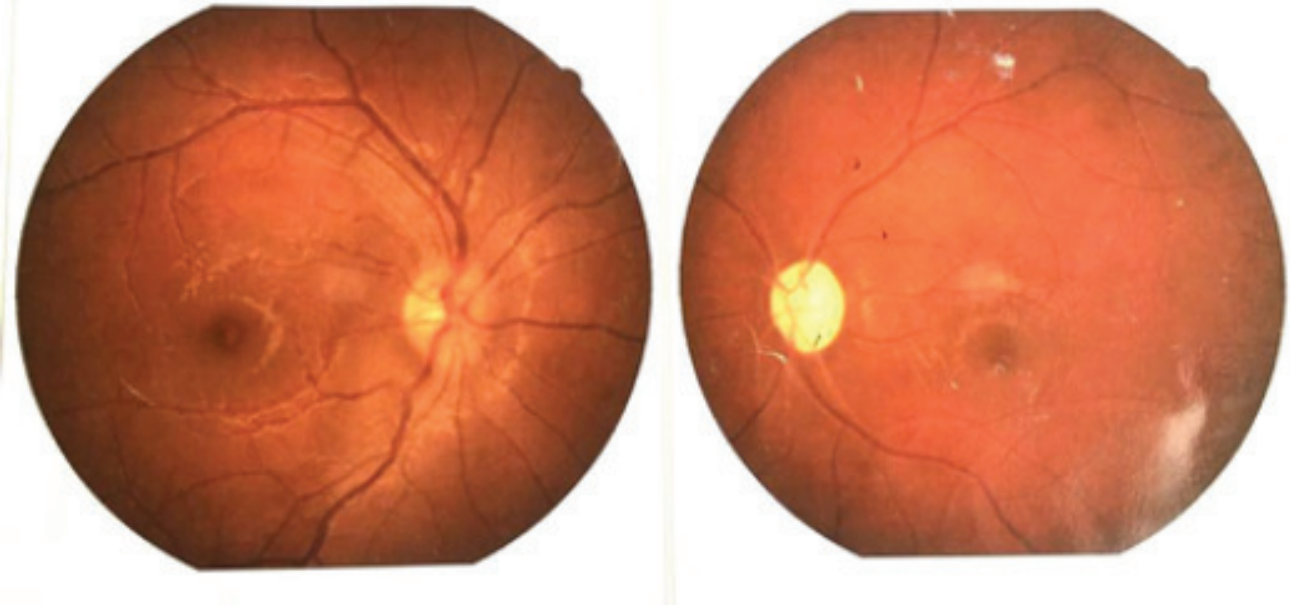


Fig.-1: Color fundus photography showing optical atrophy on left eye

Investigations Report showed ESR 42 mm in 1st hr with Hb 10.4 g/dL, total count of wbc 8,400/cmm. On Differential count neutrophil was 58%, L-38%,M-1%,E-3%,B-0%. Chest Xray was normal. S.lipid profile was within normal limit. A lumbar puncture performed showed the cerebrospinal fluid (CSF) was haemorrhagic, the CSF protein content was elevated at 1.2g/l, WBC count in the CSF was 1/cmm and RBC was 1100/cmm and oligoclonal band was negative.

MRI of cervical and thoracic spine revealed a longitudinal area of hyper intensity and spinal cord thickening from C3 to T1 in T2-weighted image. MRI of brain revealed no abnormality. Because of a high suspicion of NMO, Aquaporin 4 IgG was assessed and the result was positive.

So we started high-dose intravenous methylprednisolone (1 g/day) for 5 days, and then oral prednisolone 1 mg/kg body weight was maintained with immunosuppressant azathioprine. Her muscle power greatly improved over a period of 2 weeks and overall patient clinically improved.

Discussion:

Neuromyelitis optica or Devic's disease is a severe idiopathic immune-mediated inflammatory demyelinating disease that predominantly involves the optic nerves and spinal cord. The cardinal features of the disorder are longitudinally extensive transverse myelitis and Optic Neuritis(ON). These two clinical events can occur simultaneously or can be separated by many years. ON can be unilateral or bilateral and can occur before or after an attack of myelitis. More than 90% of patients with NMO have repeated relapses

(relapsing NMO) rather than monophasic disease. NMO is often confused with multiple sclerosis. Early discrimination between NMO and multiple sclerosis is important because the two diseases have different natural histories and treatment regimens.¹

Recently, NMO-IgG was found in the serum specifically from patients with NMO, and its target antigen was identified as AQP4 water channel protein.² NMO-IgG is 73% sensitive and 91% specific for distinguishing NMO from optic-spinal presentations of classical MS.³

NMO also can be distinguished from multiple sclerosis on the basis of several characteristics [progress more severe, no fever at onset, cerebral and cerebellar lesions absent on MRI, no oligoclonal bands shown on electrophoresis of the cerebrospinal fluid (CSF) and no elevation of albumin levels in the CSF]. In fact, CSF analysis is of little benefit in making the diagnosis. Devic's syndrome is also characterized by longitudinal rather than transverse myelitis.⁴

There are two major types of neuromyelitis optica. In the first type, optic neuritis and myelitis episodes tend to come very close together often within days or weeks and there is no recurrence after the initial flurry of symptoms. In the second form, repeated episodes of optic neuritis and myelitis that are separated by months or years occur.

The combination of neurological impairments which occur in patients with neuromyelitis optica can also be seen in multiple sclerosis (MS), acute disseminated encephalomyelitis (ADEM), systemic lupus erythematosus

(SLE) and Sjögren syndrome.⁵ In rare cases viral illnesses or immunizations precede the clinical onset.⁶ In our case patient had no history of preceding vaccinations or viral infections.

Patients with myelitis should have a cervical and thoracic spinal cord MRI scan to determine whether there is a longitudinally extensive cord lesion, which is the second major supportive diagnostic criterion for NMO. A radiological feature of NMO is a longitudinally extensive cord lesion, often extending over three or more spinal segments, but MS involves fewer than two spinal segments.^{7,8} In our Patient MRI of cervical and thoracic spine revealed a longitudinal area of hyperintensity and spinal cord thickening from C3-T1 in T2-weighted image which is consistent with the radiographic diagnostic criteria for NMO.

The first line treatment of acute attack of NMO is high dose intravenous methylprednisolone 1g daily for at least three to five days. Plasmapheresis or cyclophosphamide may be considered if there is no clinical improvement with steroid therapy alone. Oral prednisolone, Azathioprine, rituximab, mycophenolate mofetil, methotrexate, prednisone or mitoxantrone can be used as maintenance therapy. However, in this case, our patient was improved with I/V methyl prednisolone 1g daily for 5 days initially and Oral prednisolone and azathioprine as maintenance therapy.⁹

Conclusion:

In conclusion, NMO should be thought of in the differential diagnosis in any patient presenting with transverse myelitis and/or optic neuritis regardless of the gender and race. Rapid diagnosis and early initiation of aggressive immunosuppressive treatment are essential in most NMO cases.

Conflict of interest: None.

References:

1. Chia WC, Wang JN, Lai MC. Neuromyelitis Optica: A Case Report. *Pediatr Neonatol* 2010;51(6):347-352.
2. Loss of Aquaporin-4 in Active Perivascular Lesions in Neuromyelitis Optica: A Case Report. Misu T, Fujihara K, Nakamura M, Murakami K, Endo M, Konno H and Itoyama Y. *Tohoku J. Exp. Med.* 2006;209:269-275.
3. Petelin Z, Hajnsek S, Basic S, Sporis D, Pavlisa G and Nankovic S. Patient with neuromyelitis optica and inflammatory demyelinating lesions comprising whole spinal cord from C2 level till conus: case report. *BMC Neurology* 2009;56. DOI: 10.1186/1471-2377-9-56
4. Gibbs AN, Moroney J, Foley Nolan D and O'Connell PG. Neuromyelitis optica (Devic's syndrome) in systemic lupus erythematosus: a case report. *Rheumatology* 2002;41(4):470-471. doi:10.1093/rheumatology/41.4.470.
5. A case of neuromyelitis optica (Devic's disease). Kumuduni SGP, Dissanayaka A. *Galle Medical Journal* 2008;13(1).
6. Devic's neuromyelitis optica: a critical review, Lana-Peixoto MA, Arq Neuropsiquiatr. 2008 Mar;66(1):120-38.
7. Jacob A, Boggild M. Neuromyelitis optica. *Pract Neurol* 2006; 6:180-4.
8. Nakashima I, Fujihara K, Miyazawa I, et al. Clinical and MRI features of Japanese patients with multiple sclerosis positive for NMO-IgG. *J Neurol Neurosurg Psychiatry* 2006;77:1073-5.
9. Thongpooswan S, Chapagain B and Bandagi S. A Rare Case of Neuromyelitis Optica Spectrum Disorder in Patient with Sjogren's Syndrome. *Case Reports in Rheumatology*. vol. 2014, Article ID 158165, 3 pages, 2014. doi:10.1155/2014/158165.