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Case Report

A case report on bilateral neuromyelitis optica

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ABSTRACT

Neuromyelitis optica (NMO, Devic disease) is an autoimmune inflammatory disorder of the central nervous system (CNS) in which the autoimmune system attacks myelin of the neurons located at the optic nerve and spinal cord, thus producing a simultaneously or sequential longitudinally extensive inflammation of the optic nerve (optic Neuritis) and spinal cord (myelitis). Early discrimination between NMO and multiple sclerosis is important because the two diseases have different natural histories and treatment regimens. Seropositivity for NMO-IgG and longitudinally extensive spinal cord lesions are characteristic of NMO. Despite the absence of a definitive therapeutic strategy for NMO syndrome, methylprednisolone pulse therapy is recommended in the acute phase. Treatment strategies in relapse phases are aimed at preventing relapses, and increasing evidence shows a better clinical response of immunosuppressive therapy than immuno-modulating therapy (a standard multiple sclerosis-modulating therapy). We have described a 32 years old girl who had visual loss due to acute optic neuritis before 15 days in right eye and followed by complete visual loss in left eye. NMO was diagnosed because of its characteristic longitudinal myelitis and positive NMO-IgG. After combine therapy with prednisolone and an immunosuppressant, the patient's medical condition was stable and no relapse symptoms were observed.

Keywords: Acute myelitis, Aquaporin-4, Demyelination, Devic disease, Neuromyelitis optica, Optic neuritis

INTRODUCTION

Neuromyelitis optica is an autoimmune inflammatory disorder of the central nervous system (CNS) in which the autoimmune system attacks myelin of the neurons located at the optic nerve and spinal cord, thus producing a simultaneously or sequential longitudinally extensive inflammation of the optic nerve (optic Neuritis) and spinal cord (myelitis).[1,2]

The association of acute or sub-acute loss of vision in one or both eyes caused by acute optic neuropathy preceded or followed within days to weeks by a transverse or ascending myelitis was initially describe by Allbutt and later by Devic. Devic called this condition Neuromyelitis optica but its subsequently became known as Devic's disease.[2] Devic's disease is now considered as an autoimmune channelopathy. Most NMO patient produce auto antibodies against aquaporin 4 (AQP-4) also known as NMO-IgG.[3]

CASE REPORT

A 32 year's old female presented with sudden and painless loss of vision in right eye followed by left eye within 15 days period. On examination Visual acuity in both eyes was limited to no perception of light. Torch light examination shows both eye pupils dilated (5mm) size and non-reacting to light. Slit lamp examination shows anterior segment was within normal limits Intraocular tension OD 21mmhg, OS 22.5mmhg on NON contact tonometer.

Fundus examination shows disc was generalized pallor in OD disc and OS disc show edematous, hyperemic, and elevated by one disc diameter with blurred nasal margin.

Rest detail was normal. Investigation CBC was normal, ESR- 18mm (by westergreen Method), CSF Examination show normal pattern oligoclonal bands. Orbital MRI with brain show bulky left optic nerve in entire length with hyper intense signal on stir image and show intense enhancement on post-contrast study.

Hyper intense signal is seen in the posterior part of intra orbital, intra canalicular and prechiasmatic segment of right optic nerve without significant post contrast enhancement.

Altered signal in both optic chiasmas with contrast enhancement in left half is seen. Indirect Immunofluorescence test shows positive NMO IgG antibodies in serum. Patient was treated with bolus doses of Inj methyl prednisolone, 1gm in 100ml normal saline intravenously given for 5days, followed by oral prednisolone at dosage 1mg/kg body weight daily with 7 days tapering along with systemic antacid and methyl cobalamin 5000 mcg per day.

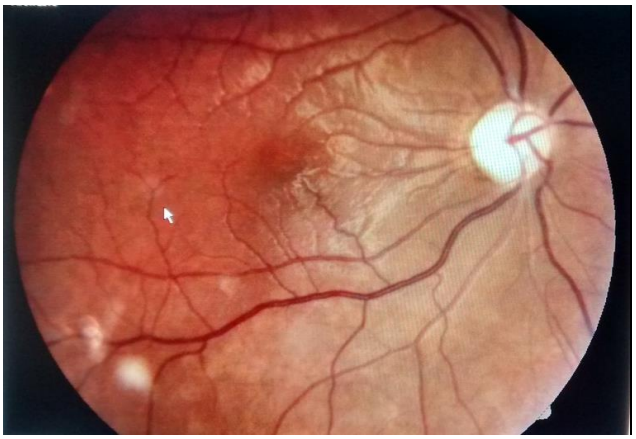

Figure 1: Right eye fundus on the day of admission.

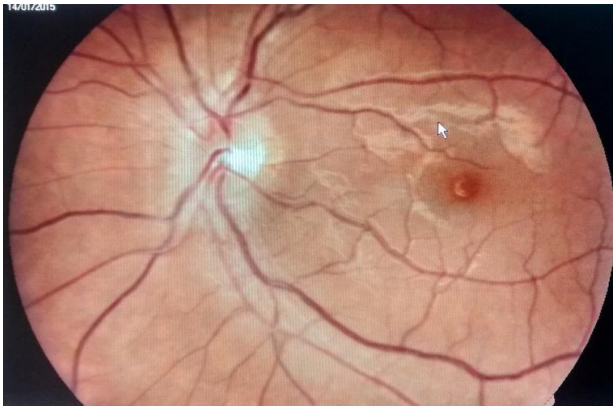

Figure 2: Left eye fundus on the day of admission

For the first 3 dose patient do not show any response but from fifth day onward patient shows some response in the left eye, she could achieve Visual acuity finger counting near face with sluggish pupillary response in left eye but no response in right eye.

Patient is also referred to oncologist for immunosuppressant therapy to prevent the recurrence of attack.

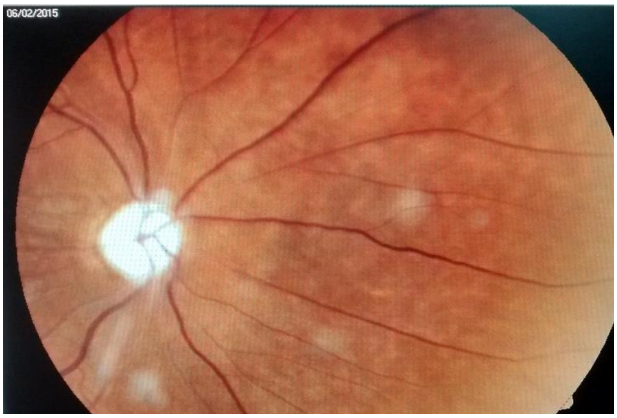

Figure 3: Right eye fundus after treatment.

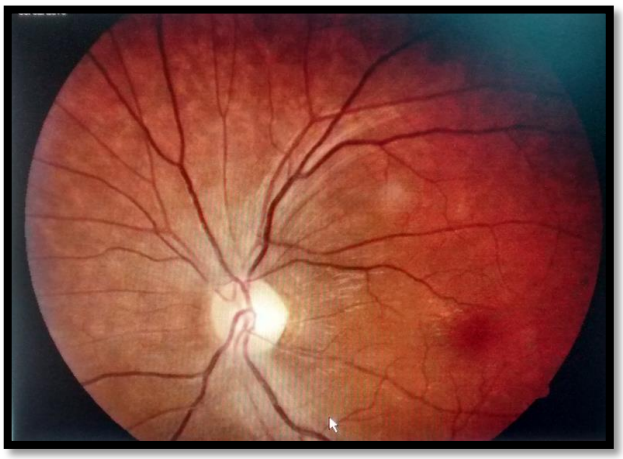

Figure 4: Left eye fundus after treatment.

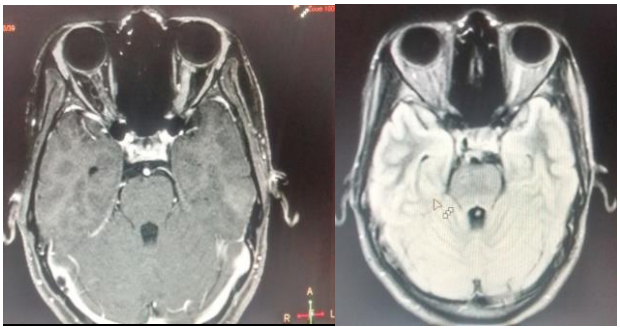

Figure 5: Orbital MRI with brain show bulky left optic nerve in entire length with hyper intense signal or stir image and show intense enhancement on post contract study.

DISCUSSION

Recent advances in the clinical, neuroimaging, laboratory and pathological hallmark have established that NMO is distinct demyelination diseases of CNS from MS. NMO can be reliably differentiated from MS at early stage using diagnostic criteria proposed by wingerchuk in 2006 which define NMO.[1,4,5]

Table 1: Biological characteristic of multiple sclerosis (MS) and neuromyelitis optica (NMO).

Cerebral spinal fluid	Usually a normal cells count or modest	Relevant in acute attack (>50cells/mm^3)
	MS	**NMO**
Pleocytosis	Pleocytosis (<50cells/mm^3)with predominance of lymphocytes	Dominated by neutrophils, (high specificity>95%, low sensitivity<30%)
Oligoclonal bands (%)	85-90	20-30
Serum Anti-nuclear abs (%)	20-30	>50
Other auto-abs(systemic autoimmune dis's	Rare	Frequent
NMO-IgG	Absent	High sensitivity (73%) High specificity (91%)

Abs= antibodies; NMO-IgG=neuromyelitis optica-immunoglobin G.

Table 2: Characteristics of monophasic and relapsing neuromyelitis optica.

Monophasic Frequency (%)	Less common (20)	Relapsing More common (80)
Age of onset (Year; median)	29	39
Sex ratio of females (%)	50	80−90
History of autoimmune disease	Uncommon	Approximately 50%
ON or myelitis (%)	48	90
Bilateral ON (%)	17	8
Simultaneous ON + myelitis (%)	31	0
Respiratory failure	Rare	Approximately 1/3
5-yr mortality rate (%)	10	32
Recovery	Good	Fair

ON = optic neuritis.

One or both the following characteristic:

- Optic neuritis
- Acute myelitis

2 of the following 3 characteristic

- Disease onset brain magnetic resonance imaging that is non-diagnostic for MS,
- Contiguous spinal cord MRI lesion extending over 3 or more vertebral segment, and
- NMO-IgG seropositive status.

Revised diagnostic criteria for neuromyelitis optica (2009)[4-6]

Definite NMO

- Optic neuritis
- Acute myelitis

At least 2 of the 3 supportive criteria

- Contiguous spinal cord MRI lesion extends ≥ 3
- Vertebral segments
- Brain MRI does not meet diagnostic criteria for
- Multiple sclerosis
- NMO-seropositive status

NMO = neuromyelitis optica; MRI = magnetic resonance imaging.

He also added that although brain MRI findings are generally either negative or nonspecific in NMO, brain lesions do not preclude the diagnosis. CSF pleocytosis or neutrophilia and the occurrence of severe, fixed, attack related motor weakness were also validated as characteristic feature of NMO but less diagnostic power. As far as treatment is concern there is no specific treatment for NMO although as noted one should considered the implication of the immuno-pathophysiology in rendering early and perhaps selective

treatment based on consideration of the uniqueness of this diseases process. Supportive care is crucial to ensure survival in patient with severe myelitis. The use of intravenous corticosteroid may lessen the severity of the attack and increase the speed of recovery of both visual and motor function. Administration of IVIG allowed discontinuation of steroid without further ophthalmic or neurological detoriation or recurrent diseases. With recent knowledge that humoral effector mechanism have a central role in NMO, acute therapeutic intervention may focus on interrupting antibody/ complement- dependent effector mechanism which include plasmapheries.[2,7] In patient with NMO, it may be worthwhile to considered treatment with highly active immunosuppressant early in diseases, reducing the immunotherapeutic regime as soon as the patient has achieved stabilization or remission. The recovery is rarely complete but may be substantial particularly as regards vision.[7] Visual recovery usually occurs with several weeks to month after the onset of visual loss.

CONCLUSION

In this patient NMO was diagnosed early because of optic neuritis and positive MRI orbit with brain and positive NMO-IgG seropositive status. After combine therapy with prednisolone and an immunosuppressant patient was medically stable and no relapse of symptoms were observed till now. Patient achieved one meter finger count in left eye and no response in right eye, suggestive of optic nerve atrophy in right eye.

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Ethical approval: Not required

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