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Case Report : Management of Recurrent Optic Neuritis in Young Patient

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Management of Recurrent Optic Neuritis in Young Patient
A Case Report

Abstract

Introduction :

Optic neuritis (ON) is an inflammatory condition of the optic nerve that accompanies a spectrum of systemic and neurological immune-mediated and infectious disorders. ON can be recurrent, with unilateral, bilateral presentation and diagnosis of recurrent cases can be challenging. The most common disorders associated with reccurent ON are multiple sclerosis (MS) and neuromyelitis optica spectrum disorder (NMOSD)

NMOSD previously known as Devic's disease, is an inflammatory CNS syndrome distinct from multiple sclerosis (MS). It is characterized by severe, immune-mediated demyelination and axonal damage predominantly targeting the optic nerves and spinal cord though rarely the brain is also involved. Most patients with NMOSD have autoantibodies against the water channel aquaporin-4(AQP4-Ab).

Purpose : To report a case of recurrent optic neuritis in young seronegative NMOSD patient

Case Report : A 23 years old woman, with gradually blurred vision on the left eye (LE) since two weeks before admission with pain in eye movement. She had two prior attack of optic neuritis and history of paraplegia which improved with corticosteroid treatment. Ophthalmological examination revealed visual acuity (VA) of the right eye (RE) was 1.0 and one meter finger counting on the LE, relative afferent pupillary defect (RAPD) grade 2 on LE, posterior segment revealed slightly pallor optic disc with defined margin on LE. Laboratory findings was within normal limit. MRI brain with contrast showed left optic neuritis and spinal cord showed and extensive transverse myelitis for C2-Th5 segment of the spine. Aquaporin-4 IgG showed negative result. She was diagnosed with recurrent typical optic neuritis on the LE + suspected NMO. She was hospitalized and got intravenous injection of methylprednisolone 250mg 4x perday. there were no improvement of LE VA, with complete recovery of pain in eye movement,

Conclusion : Recurrent optic neuritis is a challenging case, accurate diagnosis and etiology is crucial to determine treatment and to predict the prognosis of the patient. NMOSD can presents with acute unilateral optic neuritis and myelitis confirmed with clinical, radiologic and serologic examination.

I. Introduction

Optic neuritis (ON) is most commonly defined as demyelinating inflammation of the optic nerve. Loss of vision is accompanied by visual field defects and loss of color vision. Etiological causes of optic neuropathy can be infectious, ischemic, toxic, hereditary, autoimmune, metabolic, infiltrative, or compressive.In some cases ON can be recurrent, with unilateral or bilateral presentation. The diagnosis

of recurrent cases may be challenging and accurate prediction of the prognosis depends on correct diagnosis.[1-3]

Neuromyelitis optica is an immune-mediated chronic inflammatory disease of the central nervous system (CNS) that is associated with serum aquaporin-4 immunoglobulin G antibodies (AQP4-IgG). NMO was thought to be a clinical variant of MS in the past but the dicovery perivascular antibody complement deposition within active lesions and the subsequent discovery of specific autoantibodies serum of NMO patients indicated that humoral immunity is involved in the majority of cases. The hallmark manifestations are recurrent longitudinally extensive transverse myelitis and optic neuritis (ON). [1,3-6]

The International Panel for NMO Diagnosis (IPND) convened to develop revised diagnostic criteria using systematic literature reviews and electronic surveys to facilitate consensus. The new nomenclature defines the unifying term NMO spectrum disorders (NMOSD), which is stratified further by serologic testing (NMOSD with or without AQP4-IgG). More stringent clinical criteria, with additional neuroimaging findings, are required for diagnosis of NMOSD without AQP4-IgG or when serologic testing is unavailable. [2-4]

The incidence and prevalence of NMO were reported as little a 1:100.000 population among caucasian whereas in east asian popuplatin is a little bit higer at 3.5/100.000 population. Similar to other autoimmune disease, there is strong female predilection. This case report is intended to report a case of recurrent optic neuritis in a young seronegative NMOSD patient. [4-6]

II. Case Report

A 24-year-old woman came to the Cicendo Eye Hospital on 3rd November, 2020, with the chief complaint of gradually blurry vision on the left eye (LE) since 2 weeks before admission, with pain in eye movement. Patient denied histories of fever, flu-like syndrome, head trauma, severe headache, nausea and projectile vomiting, unexplainable hiccups, consuming alcohol or long-term medication, recurrent red eye, nor history of surgery. She had no systemic disease such as hypertension, diabetes mellitus nor hypercholesterolemia. Patient had completed

Tuberculosis (TB) treatment 15 years prior, two attacks of optic neuritis on (LE), and paraplegia. The patient mentioned that there were clinical improvement with corticosteroid treatment on those attacks.

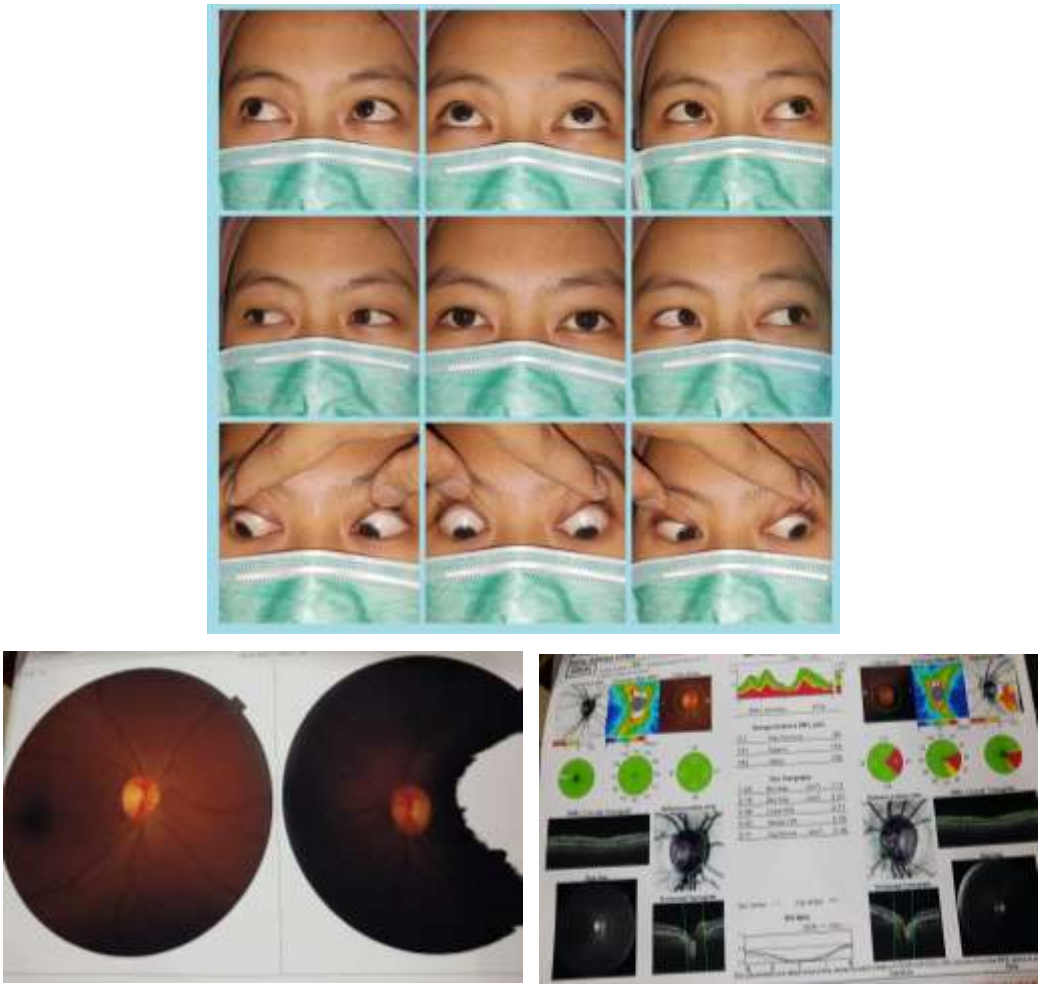


Figure 2.1 Eye movement of the patient showed no eye movement restrictionwith fundus photograph and OCT of the patient showed slightly pallor left optic disc with decreased of Retinal nerve fiber layer thickness on LE

Physical examination on 3rd of November 2020 showed normal vital signs, with 46 kg of body weight. Ophthalmological examination revealed visual acuity of the right eye (RE) was 1.0 and LE was one-meter finger counting. Intraocular pressure (IOP) within normal limit, no restriction of eye movement but there were pain on LE when looking to both sides, Hirschberg test showed ortothropia. Anterior segment on the RLE were normal, with decreased pupillary light reflex and relative

afferent pupillary defect (RAPD) grade 2 on LE. Posterior segment with indirect funduscopy revealed normal optic disc apprearance on RE, slightly pallor optic disc with defined margin on LE. Ancillary examinations such as colour vision with Ishihara, amsler grid and contrast sensitivity test showed normal result on the RE, meanwhile they were hard to evaluate on the LE.

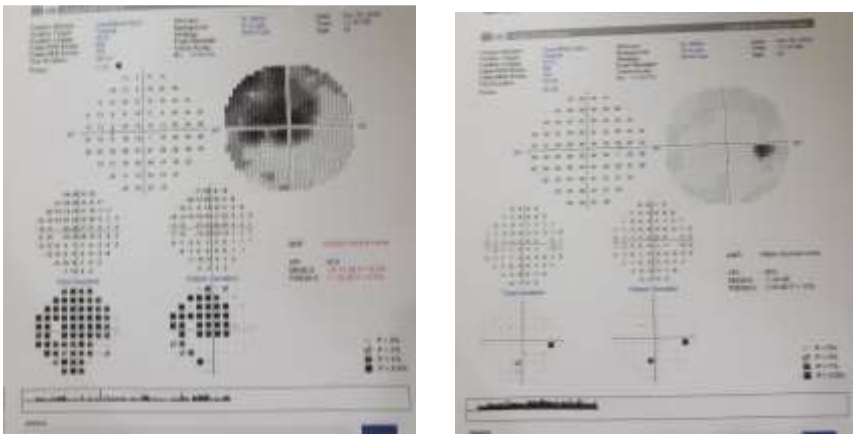


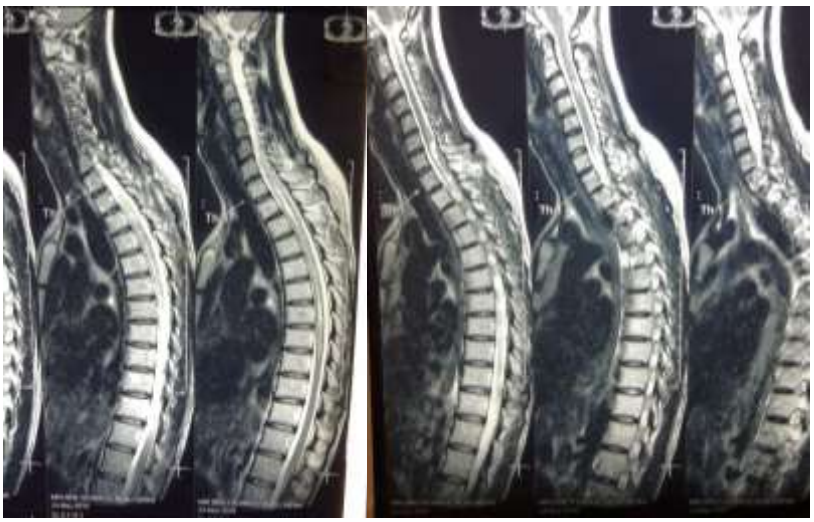
Figure 2.2 Visual field testing using showed normal result on RE, and central scotoma on LE

Neurological examinations were within normal limit with normal motoric strength and no other cranial nerves symptoms present. Optical coherence tomography showed decreased of temporal retinal nerve fiber thickness and visual field test showed normal result in RE, and central scotoma on LE.

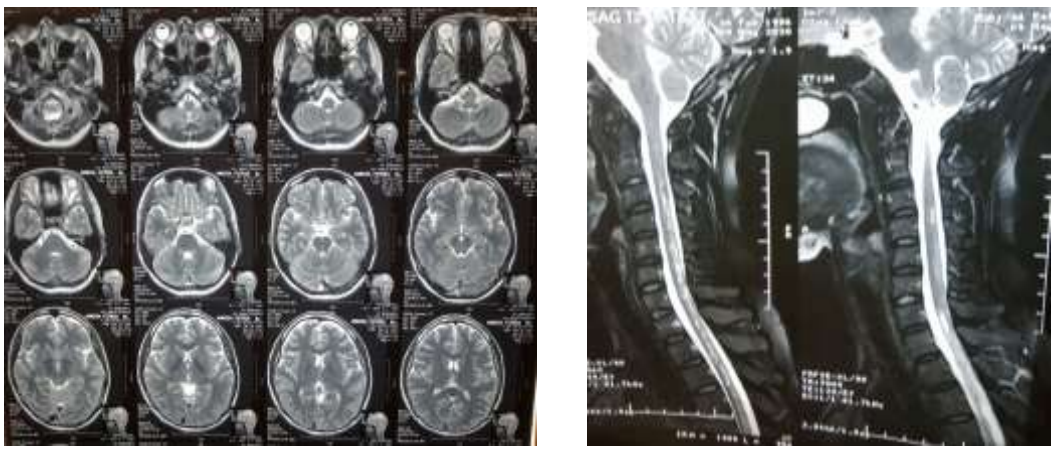
Prior radiological examination showed no significant finding on brain MRI, but showed myelopathy in the level of C3-4 and C7-Th 9 in spine MRI that was conducted one-year prior and was repeated on 6 months before admission that showed extensive transverse myelitis cervicothoracal C2-T5. Chest x ray taken on admission day showed no abnormalities. routine haematologic examination showed no C reactive protein elevation, normal eritrocyte sedimentation rate, and normal leukocyte level, and serologic tesing of toxoplasmosis and CMV infection showed non-reactive result.

Patient was diagnosed with recurrent Optic neuritis LE + suspected NMO and was admitted to receive intravenous injection of methylprednisolone 4x250 mg, omeprazole iv 2x40mg, mecobalamine iv 1x500mcg, calcium supplement tablet 3x perday. On the third day of hospitalization the patient reported an improvement of

eye movement pain, whereas her visual acuity did not improve. After 12th injection of methylprednisolone she was discharged and was treated with methylprednisolone tablet 1 mg/kgBW/day (48 mg once perday), omeprazole 1x40 mg, calcium tablet 3x 1, and mecobalamine tablet 1x500 mcg. She was planned to do repeat MRI brain scan with contrast and follow up visit 1 week later to neuro-ophthalmology unit.



(a)



(b)

Figure 2.3 Previous spinal cord MRI showed myelopathy in C2-4 C7-Th9 (a), Brain MRI and spinal cord showed signs of left optic neuritis and extensive transverse myelitis in the spinal cord from C2-Th5 (b)

On follow up visit, patient did not report any side effect of the given treatment, the pain in eye movement has healed completely while the visual acuity still feels the same, ophthalmological examination showed visual acuity of the RE was 1.0 and the LE was still one-meter finger counting. IOP was within normal limit. Eye movement showed no restriction and no pain, Hirschberg test showed ortothropia. Anterior segment on the RLE showed similar result compared to before admission. Posterior segment with indirect funduscopy revealed slightly pallor optic disc with defined margin on LE. Ancillary examinations such as colour vision with Ishihara, amsler grid and contrast sensitivity test showed normal result on the RE, hard to evaluate on LE.The Brain MRI showed left intracranial and intraorbital optic neuritis with no sign of demyelination process in brain or brainstem. Patient underwent tapering off of methyl prednisolone by 8 mg every week and consulted to the neurology department to underwent aquaporin 4 examination which showed negative result. The prognosis of this patient is quo ad vitam Dubia, and quo ad fungsionam is dubia ad malam.

III. Discussion.

Recurrent ON is a challenging case and prompt acurate diagnosis needs various examination including clinical, radiologic and serologic examination. it is usually associated with conditions such as multiple sclerosis, neuromyelitis optica, myelin oligodendrocyte glycoprotein (MOG), or it can present as an idiopathic case. Cases of a primary demyelinating process in the optic nerve or the central nervous system is not the cause of unilateral or bilateral anterior or retrobulbar optic neuritis. Those cases can arise from underlying systemic systemic infection, vaccination, or systemic inflammatory disease.[3,5-8]

Recurrent ON is one of the hallmarks of NMO together with longitudinally extensive transverse myelitis. Optic neuritis associated with NMO is considered chronic demyelinating optic neuritis with the difference between NMO and MS, NMO has destructive lesions on both white and grey matters of the CNS in contrast of only white matter in MS. Wingerchuck Et al proposed a diagnostic criterion in 2006 with high specifity if there in addition to a history of at least one episode of

ON and one episode of myelitis, two of the following three supporting criteria are met: 1. Continuous spinal cord MRI lesion extending over three or more vertebral segments 2. Brain MRI not meeting criteria for MS1 at disease onset, and NMO-IgG seropositive status. [4,8-10]

The patient in this case report has had three episodes of optic neuritis, one episode of myelitis, in addition of MRI examination showed continuous spinal cord lesion and the MS criterion was not met. Although the AQP4-IgG was negative, according to wingerchuck criteria this patient can be diagnosed with NMOSD. In a testing for MOG antibody was suggested but owing to the lack of available testing the examination was not carried out. Treatment for NMO includes management of acute attacks to promote recovery, prevention of NMO exacerbations (i.e. initiation of long-term maintenance immunosuppression), prevention and monitoring of adverse effects. During acute attack in NMO, intravenous methylprednisolone is the choice according to ONTT. The ONTT is still relevant for treatment guideline in optic neuritis because it remains the landmark treatment of optic neuritis. The ONTT demonstrated that corticosteroid therapy for optic neuritis had no long-term beneficial effect on vision, although the use of intravenous methylprednisolone, 250 mg every 6 hours for 3 days, followed by oral prednisone, 1 mg/kg/day for 11 days, sped recovery by 1-2 weeks.[10-12]

This patient was treated with intravenous methylprednisolone 250 mg every 6 hours for 3 days followed by oral methylprednisolone 1 mg/kg/day for 7 days and tappered-off by 8 mg per week in the following weeks, her visual acuity, ad other ancillary testing such as ishihara, amsler gerid and contrast examination did not improve but she reported the eye movement pain has healed.

A curative treatment for NMO does not exist to date. Instead, the main treatment goals are remission and improvement of relapse-associated symptoms, long-term stabilization of disease course by means of relapse prevention, and Symptomatic therapy of residual symptoms. Treatments for seronegative NMO patients is still debated, but infectious, parainfectious, metabolic, or paraneoplastic causes must definitely be ruled out before considering immunosuppressive treatments. Azatriopin and rituximab is currently most widely used therapies in NMO. [4,11-14]

This patient did not have any improvement in her visual acuity owing to the fact that there was optic atrophy present and this is her third ON attack. while her eye movement pain was healed. The visual prognosis of this patient is ad malam. and the prognosis for life is dubia due to the recurrency could still happen and threaten her life, but the current mortality rate is around 10%

V. Conclusion

Reccurent optic neuropathy is a challenging case and accurate diagnosis is crucial to determine treatment and to predict the prognosis of the patient. Etiology of which the most common form are multiple sclerosis and neuromyelitis optica. NMO presents with either acute, chronic relapsing unilateral or bilateral optic neuritis and myelitis confirmed with MRI findings based on the criteria by wingerchuck in 2006. NMO and MS are two disease with similarity in clinical presentation but different in patomechanism, MRI finding, treatment and prognosis. underscoring the importance of early diagnosis and appropriate treatment.

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