

Visual Disorders in Optic Neuromyelitis: Report of Two Cases

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Abstract

Optic neuromyelitis is a disease caused by autoimmune disruption of the aquaporin 4 channels of central nervous system, whose involvement is more frequent over the spinal cord and optic nerves as well as in specific areas of the brain. Optic neuritis in optic neuromyelitis can occur simultaneously with transverse myelitis or separated by a variable time interval of days or even years. It may be unilateral or bilateral and could occur in recurrent forms. We describe a 35-year-old woman with visual loss due to acute bilateral optic neuritis and a 46-year-old woman with unilateral optic neuritis that were admitted at the neuro- ophthalmology service of the Cuban Ophthalmology Institute last year. Both were treated with intravenous methylprednisolone for five days. Three month later neurologic symptoms appeared, a magnetic resonance imaging was performed and the optic neuromyelitis diagnosis was confirmed. We show the results of the neuro-ophthalmology studies before and three month after the treatment.

Keywords: Optic neuromyelitis; Optic neuritis

Background

Optic Neuromyelitis (ONM) is a disease caused by autoimmune disruption of the aquaporin 4 channels (AQ4P) of central nervous system (CNS), whose involvement is more frequent over the spinal cord and optic nerves as well as in specific areas of the brain. Seropositivity for ONM-IgG and longitudinally extensive spinal cord lesions (3 or more spinal segments) are characteristic of ONM. Cuban population has an ONM prevalence of 0.52 per 100,000 habitants and an annual incidence average rate estimated of 0.053 per 100,000 habitants with no differences by ethnicity [1].

Optic neuritis in ONM can occur simultaneously with transverse myelitis or separated by a variable time interval of days or even years. It may be unilateral or bilateral and it could occur in recurrent forms, but few articles give a specific approach of the morphological characteristics of the optic nerve head in ONM. In this paper we describe two cases of patients with different type of optic neuritis as a presentation form of ONM. We showed the results of the neuro-ophthalmology studies before and three month after the treatment.

Case Presentations

Case 1

A healthy woman of 46 years old was presented to our clinic with vision loss in the right eye associated with pain at eye movements. Her ocular history included high myopia that was partially corrected with radial keratectomy 20 years ago. Her best corrected visual acuity (BCVA) with Bavie-Lovie Chart (LogMAR) was 1.0 in the right eye (RE) and 0.0 in the left eye (LE); contrast sensitivity (CS) with Pelli-Robson Chart was 0.00 in RE and 1.50 for LE. Color vision (CV) taken with Ishihara test was 0 in RE (affected eye) and 21 in LE.

Pupils were equally round and reactive to light with afferent pupillary defect in right eye. Dilated fundus examination was normal in both eyes.

The visual evoked potentials (VEP) showed significant delays in the P100 response and decreased amplitudes in RE.

A diagnosis of retrobulbar demyelinating optic neuritis (ON) was made.

Case 2

A healthy woman of 35 years old presented to our clinic with vision loss in both eyes that started in the left eye and four days later occurs more severe in the contralateral eye. The BCVA was hand motion in RE and counting finger in LE, CS and CV were not explorable due to severe visual impairment.

Pupils were equally round and slowly reactive to light without afferent pupillary defect on both eyes. Dilated fundus examination showed edema of the optic discs without vitreous cells.

The VEP showed significant delays in the P100 response and decreased amplitudes in both eyes.

A diagnosis of demyelinating bilateral optic neuritis was made.

Diagnosis

Both patients were treated with 1g/d of methylprednisolone for 5 days.

A magnetic resonance imaging (MRI) was made. The case 1 had MRI with lesions in the thalamus (Figure 1) and brainstem. In the second case the MRI showed a longitudinally-extensive transverse myelitis (LETM) lesion (Figure 2).

The diagnosis of ONM was performed using the diagnostic criteria of Miller et al. National Multiple Sclerosis Society (NMSS) task force criteria, 2008 [2].

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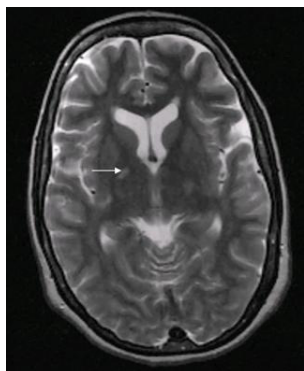


Figure 1: Brain MRI with demyelinating lesions (case 1).



Figure 2: Spinal cord MRI with longitudinally-extensive transverse myelitis lesion (case 2).

Clinical Course

Case 1

Three months later, the patient improved visual function slightly on the right eye. BCVA was 0.6 (LogMAR units), CS improved from 0.00 to 0.90 and CV was 1. Left eye remained normal in psychophysical studies (BCVA, CS, CV)

The visual field (VF) in figure 3 had a scotoma in RE and the VEP showed significant delays in the P100 response with normal amplitudes in LE. Optical coherence tomography (OCT) in figure 4, showed thinning of retinal nerve fiber layer (RNFL) in the upper and lower quadrants in RE. Dilated fundus examination (Figure 5) of RE had attenuation of arterioles in the peripapillary retina (black arrow) and optic disc atrophy (white arrow); dilated fundus exam of LE was unremarkable at that time.

Case 2

Three months later, the patient returned to our clinic complaining of acute urinary retention and complete transverse myelitis, but improved visual function in the right eye with BCVA 0.0 (LogMAR units), CS 1.50 and CV 21. The left eye had lower recovery with BCVA in 0.3, CS in 1.05 and CV in 1. The visual field in figure 6 showed an inferior altitudinal defect in both eyes. OCT in figure 7, presented significant RNFL thinning in superior quadrants in RE. The RNFL thickness also declined in most quadrants in the left eye. Dilated fundus examination (Figure 8) in both eyes showed significant attenuation of arterioles with changes in vascular retinal walls (white arrow) and bilateral optic disc atrophy.

Discussion

The improvement of visual function after an attack of optic neuritis

associated with ONM is less than occurs after other causes of optic neuritis. Visual field defects have been described of different types, but

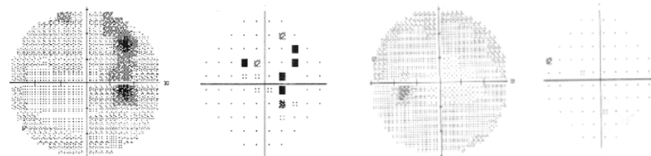


Figure 3: HVF 30.2 of both eyes. Scotoma in right eye (case 1).

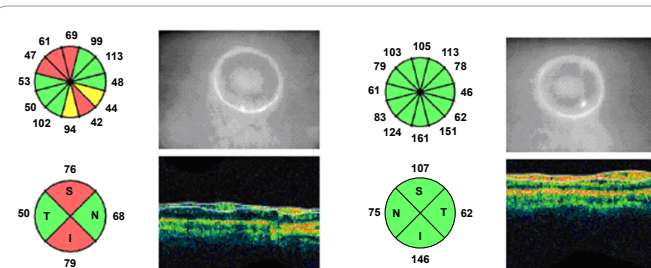


Figure 4: OCT (Stratus 3000) with significant RNFL thickness reduction in superior and inferior quadrants in right eye. Normal left eye (case 1).

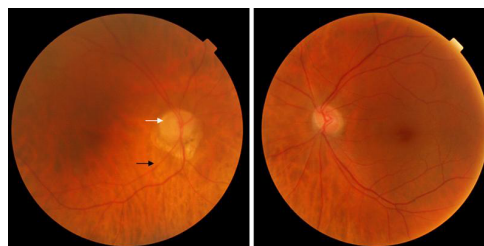


Figure 5: Retinography of right eye with peripapillary arteriolar attenuation (black arrow) and optic disc atrophy (white arrow). Normal left eye (case 1).

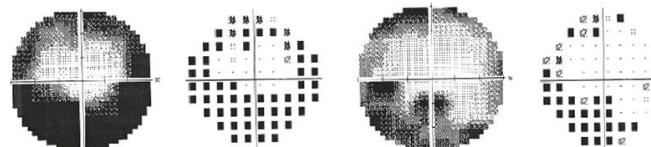


Figure 6: HVF 30.2 of both eyes with inferior altitudinal defect (case 2).

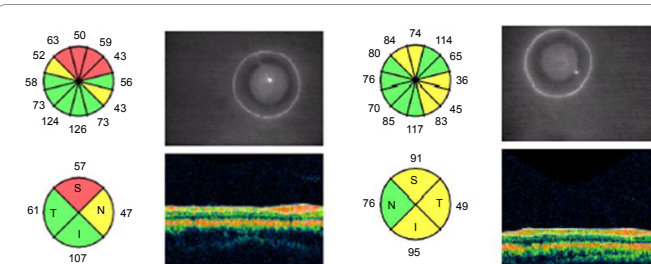


Figure 7: OCT (Stratus 3000) with significant RNFL thickness reduction in superior quadrant of right eye and less reduction in almost all quadrants of left eye (case 2).

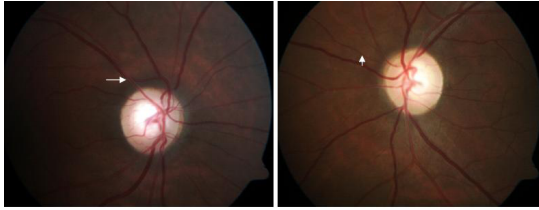


Figure 8: Retinography of both eyes with arteriolar attenuation, changes in retinal vascular walls (white arrow) and optic disc atrophy.

in patients with ONM altitudinal hemianopsia may be characteristic [3]. The most important features of fundus examination are the pallor of the optic disc that may vary from mild to severe segmental pallor to overall pallor and atrophy associated with attenuation of retinal blood vessels. The narrowing of the arterioles was severe enough in the second case as to give the appearance thickened vascular wall. RNFL thinning is well documented in this condition [4,5]. RNFL thinning in ONM involves all quadrants and sometimes preserves axons that mediate central vision, but the upper and lower quadrants are affected more severely.

AQP4 is expressed on the abluminal surface of endothelial cells in unfenestrated capillaries from the CNS, as well as in the astrocytic end feet that supply the tight junction of the blood–brain barrier. Astrocytes and their processes not only constitute a large part of optic nerve parenchyma in positions adjacent to vessels and neurons but rather are present in the adventitia of the central retinal vein and artery [6]. Therefore, some of the arteriolar changes described here, may result from direct vascular inflammation due to anti-AQP4 autoantibodies. The prelaminar region is principally supplied by direct branches of the short posterior ciliary arteries and branches of Zinn-Haller circle that are related with the end feet astrocytic. It has been suggested that an ischemic mechanism mediated by anti-aquaporin-4 antibody may play a role and explain the altitudinal hemianopia that could be seen in these patients, similar to ischemic optic neuropathy. Perhaps one reason that optic neuritis anti-AQP4-specific antibodies might initially be more prevalent than a longitudinally-extensive transverse myelitis in

ONM is that the tissues of the optic nerve are more sensitive to volume changes induced by AQP4 dysfunction than other areas of the CNS [7].

The global pattern of RNFL thinning may be causally related to vascular changes described. Vascular-mediated optic neuropathies such as glaucoma and non-arteritic anterior ischemic optic neuropathy are known to cause injury to the arcuate fibres of the RNFL. Therefore the global pattern of RNFL thinning is more consistent with a vascular process [4]. We speculate that vasculopathy may play a direct role in tissue injury in ONM.

Conclusion

In these case reports we wanted to emphasize different forms of presentation of optic neuritis in patients with ONM and the distinctive pattern of RNFL and vascular damage that could be an important feature in the differential diagnosis of other demyelinating diseases. In conclusion, it is important to say that accurate and early diagnosis and distinction from multiple sclerosis is critical to facilitate the initiation of immunosuppressive therapy to prevent attacks.

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