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Neuromyelitis optica presenting with bitemporal hemianopia

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Abstract

Neuromyelitis optica (NMO) is an idiopathic immune-mediated inflammatory disease of the central nervous system (CNS) characterized by severe demyelination and axonal damage with a predilection for the optic nerves and spinal cord. In this case report, a 60-year-old woman presented with decreased vision. Visual field testing initially demonstrated a bitemporal inferior quadrantinopia which progressed to a bitemporal hemianopia. The patient did not have any focal neurologic deficits. Indirect immunofluorescence testing was performed and positive for serum anti-aquaporin 4 (AQP4) antibodies indicating a diagnosis of NMO. The bitemporal hemianopia largely resolved following intravenous steroids. However, she developed a relapse of her NMO requiring restarting of the intravenous steroids. The patient was diagnosed with chiasmal neuritis. A variety of visual field defects may be seen in optic neuropathies, but typically in NMO, a central scotoma or altitudinal hemianopia is the most frequent visual field defect in patients with optic neuritis; bitemporal hemianopia is exceedingly rare.

Introduction

Neuromyelitis optica (NMO) is an idiopathic immune-mediated inflammatory disease of the central nervous system (CNS) characterized by severe demyelination and axonal damage with a predilection for the optic nerves and spinal cord, the brain being relatively spared.[1] Though may be unilateral, it typically presents as a bilateral optic neuritis with subsequent development of transverse myelitis within days or weeks. A variety of visual fields may be seen in optic neuropathies but typically in NMO patients, with every optic neuritis attack, central scotoma and altitudinal hemianopia were seen most frequently.[2] In this case report, a 60-year-old woman presented with an atypical presentation of NMO with bitemporal hemianopia secondary to chiasmal neuritis with subsequent cerebral plaques.

Case Report

A 60-year-old woman with underlying hypertension, bronchial asthma, and a history of previously treated colon cancer, presented with a one-week history of reduced vision in the left eye, particularly in the lower half of her visual field. Both eyes were pseudophakic, without any known surgical or post-surgical complications. She denied any recent headache, nausea, or vomiting, and denied any limb weakness or altered sensation. There was also no history of prodromal flu-like illness, tinnitus, chronic cough, night sweats, or fevers.

Clinical examination revealed no signs of systemic illness. At presentation, best corrected visual acuity (BCVA) was 20/40 in the right eye and 20/80 in the left eye. Examination of the eye revealed normal anterior

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and posterior segments, with both optic discs appearing normal, without disc edema, hemorrhage, vessel obscuration, or edema (Figure 1). A relative afferent pupillary defect (RAPD) was not detected in either eye, but there was reduced red saturation and light brightness in the left eye as compared to the right.

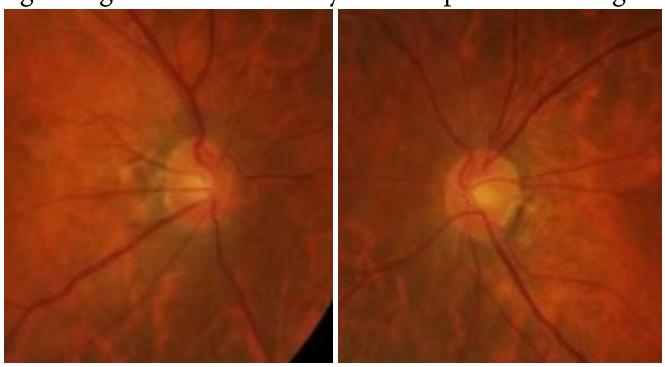

Figure 1

Upon presentation, ophthalmoscopy demonstrated normal optic discs without disc edema, hemorrhage, vessel obscuration, or pallor (right eye on left, left eye on right).

Visual field testing by confrontation revealed that the temporal halves of both visual fields were reduced. Optical coherence tomography (OCT) of the retinal nerve fiber layer did not demonstrate any optic disc abnormality.

Full threshold central 24 degree Humphrey visual field (HVF) testing (using algorithm 2 on either side of the horizontal and vertical meridian, i.e., "24-2") disclosed bitemporal inferior quadrantinopia (Figure 2). Magnetic resonance imaging (MRI) of the orbit and brain was performed the following week to rule out a craniopharyngioma or a suprasellar lesion. The MRI was suggestive of bilateral optic neuritis with both intraorbital segments of the optic nerves thickened and enhanced (right 7 mm, left 6 mm); the right side of the optic chiasm appeared thicker compared to the left. No sellar or suprasellar masses were reported and the pituitary gland and infundibulum were normal (Figure 3). A computed tomography scan of her thorax, abdomen, and pelvis showed no signs of recurrence of her previous colon carcinoma.

Following the MRI, a repeat HVF was performed, demonstrating worsening of the visual field defect to a bitemporal hemianopia (Figure 4). The patient's BCVA had worsened to 20/80 in the right eye and 20/120 in the left eye. Following negative infectious disease screening, intravenous methylprednisolone at a dose of 250 mg four times per day was commenced.

A lumbar puncture was performed which drained clear cerebrospinal fluid (CSF) with an opening pressure of 7 cmH_2O. CSF cytology did not show any atypical cells and was negative for Cryptococcus neoformans. No capsulated yeast was seen upon staining with Indian ink. No oligoclonal bands were detected. Indirect immunofluorescence testing was performed and positive for serum anti-aquaporin 4 (AQP4) antibodies indicating a diagnosis of NMO. She was subsequently commenced on the immunosuppressant azathioprine at a dose of 25mg daily for 2 weeks; the dose was the increased to 50mg daily with close monitoring of her liver and renal functions.

A repeat HVF was done after completion of intravenous steroids, which showed improvement in her field loss (Figure 5). A spinal MRI was done which showed cervical spondylosis and degenerative disc disease with mid spinal canal stenosis at C4/C5, C5/C6 and L4/L5, but no enhancing spinal cord lesions were reported.

At her subsequent follow up appointment about a month later, she complained of a one-week history of vomiting, and not tolerating oral intake. She also reported new onset weakness and altered sensation in her left upper arm. She denies any ocular or visual symptoms. She then admitted to not taking any azathioprine nor her oral steroids as she was worried of the side effects. A repeat MRI showed new lesions at the left occipital and parietal lobes with resolved bilateral optic myelitis. She was restarted on intravenous methylprednisolone and treated as a relapse of her NMO.

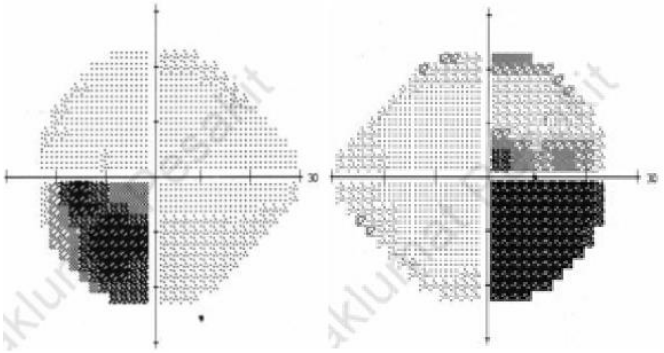

Figure 2
On presentation, Humphrey visual field testing demonstrated bitemporal inferior quadrantinopia (left visual field on left, right visual field on right).

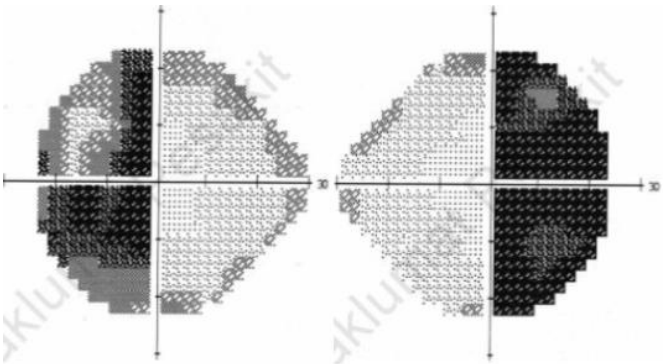

Figure 4
At one-week after presentation, Humphrey visual field testing demonstrated worsening to a bitemporal hemianopia (left visual field on left, right visual field on right).

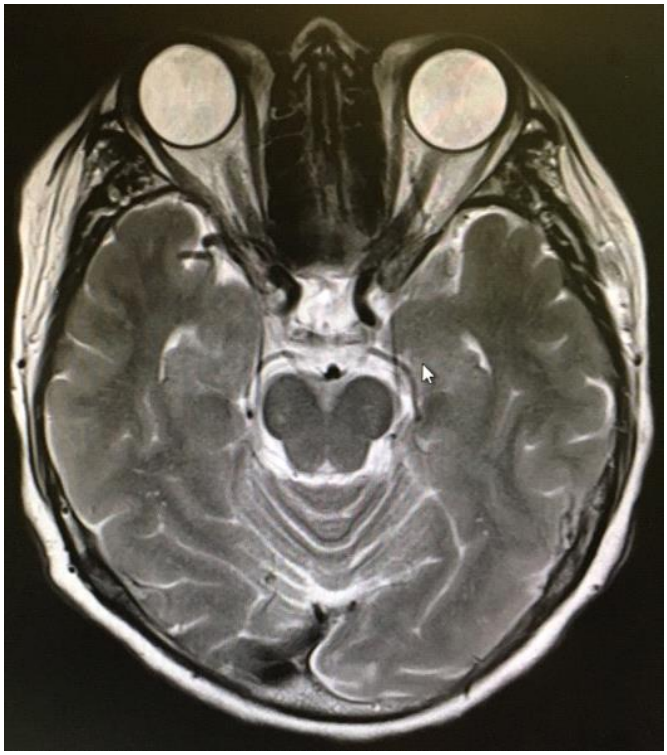

Figure 3
Axial section magnetic resonance imaging of the brain and orbits demonstrated optic nerve thickening of the intraorbital segments of the right and left optic nerves as well as enhancement of a thickening optic chiasm.

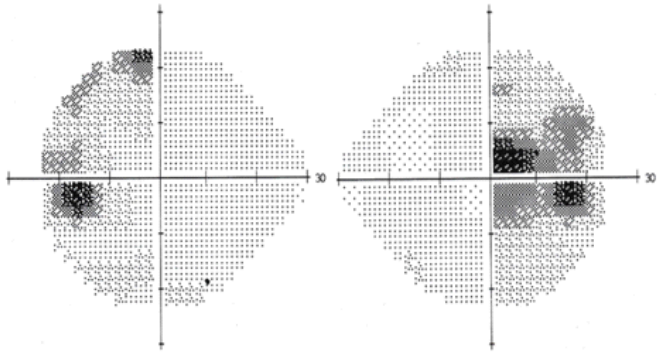

Figure 5
After three days of intravenous steroids, Humphrey visual field testing demonstrated improvement (left visual field on left, right visual field on right).

Discussion

The diagnosis of NMO may be difficult at onset of disease. The diagnostic criteria for NMO is optic neuritis (either unilateral or bilateral) with myelitis and at least two of the following criteria: a contiguous spinal cord lesion on MRI involving 3 or more vertebral segments, a brain MRI non-diagnostic for MS, positive testing for serum anti-aquaporin 4 (AQP4) antibodies. The presence of anti-aquaporin 4 (AQP4) antibodies confers 76% sensitivity and 94% specificity to NMO.[3]

A presentation of reduced vision in the patient's left field of vision triggered the formal HVF testing. A

lack of any signs of optic disc edema or inflammation upon fundoscopy, lack of any relative afferent pupillary defect, a normal retinal nerve fiber l ayer on OCT imaging, and HVF testing demonstrating bitemporal inferior quadrantinopia suggested the possibility of a space occupying lesion potentially at the chiasm, and argued against a diagnosis of optic neuritis. Therefore, an MRI was requested for the patient; in the absence of SOL symptoms such as headache, vomiting, or limb weakness, the earliest available MRI for her was one week later. When the MRI scans then showed signs suggestive of a bilateral chiasmal optic neuritis, she was immediately commenced on intravenous steroids. However, at that point, her repeat HVF as well as her visual acuity had demonstrated significant deterioration, suggesting the rapid progress of her disease. Fortunately, following completion of the steroid regime, her HVF and vision had improved. A contrast sensitivity test however was not done on the patient at baseline and post treatment.

Central scotoma is recognized to be a typical visual field defect pattern of optic neuritis in patients with multiple sclerosis (MS). In a Japanese study by Nakajima, et al. NMO patients showed a higher incidence of non-central scotoma formation than MS patients, and, in this group of patients, altitudinal hemianopia was the more common defect.[1] An altitudinal visual field defect is suggestive of ischemic optic neuropathy, which occasionally results from posterior ciliary artery occlusion.[4] It is thus suggested that an ischemic mechanism mediated by anti-aquaporin 4 (AQP4) antibodies may have a role in optic neuritis for NMO patients.[1] NMO patients showed more vascular changes of optic neuritis compared to MS patients and it is postulated that these changes may result from direct vascular inflammation mediated by anti-AQP4 antibody.[5,6]

This however does not explain the bitemporal hemianopia in our patient. The initial presentation of bitemporal inferior quadrantinopia suggests a lesion affecting the superior decussating fibers of the chiasm with functional interference in the superior portion.[7] Compressive lesions are typically more relentless, with slow depression of monocular or binocular function. Intrinsic lesions, including demyelinating disease, are usually acute in onset, often bilateral blurred vision as demonstrated in this patient. The Optic Neuritis Treatment Trial (ONTT) demonstrated that 5.1% of the 448 participants with acute optic neuritis had intrinsic lesions of the chiasm in demyelinating disease.[8] The exact mechanism by which the chiasm is targeted in optic neuritis is unknown but majority are associated with inflammatory demyelinating disorders, such as NMO and to a lesser extent MS.[9,10] Although the visual prognosis of patients with idiopathic chiasmal optic neuritis are generally good, patients with NMO and its partial or intermediate forms, termed NMO spectrum disorders (NMOSD) tend to have severe and irreversible visual impairment.[11] Fortunately, in the case of this patient, her visual acuity and visual field on HVF improved significantly post treatment with intravenous methylprednisolone.

It is important to distinguish NMO from MS as NMO does not respond to the immunomodulatory treatments that are used in MS. Case series and observational studies have demonstrated that azathioprine in combination with oral steroids reduces the frequency of attacks.[12,13] In this case, however, the patient did not take the prescribed azathioprine, which subsequently led to a relapse of the disease within less than 2 months from the initial onset. Traditionally, NMO is believed to differ from MS in that NMO has no brain lesions, although studies have reported a higher frequency of non-specific brain MRI abnormalities in NMO patients. More recent studies, however, have showed the involvement of area postrema and the optic chiasm in NMOSD.[10,14,15]

In our reported patient, new cerebral lesions were noted at the time of her relapse. This case illustrates an atypical case of AQP4-seropositive NMO with bitemporal inferior quadrantinopia on presentation,

progressing to bitemporal hemianopsia, with chiasmal optic neuritis, who demonstrated good visual recovery after treatment with intravenous steroids and immunosuppression. Awareness of atypical presentations of demyelinating diseases is important in order to initiate timely and appropriate disease-modifying management.

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