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CASE REPORT

Altitudinal Visual Defect as the Initial Sign of Optic Neuritis: A Case Report

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Abstract

Optic neuritis (ON) is the most common optic neuropathy in adults and is frequently seen together with multiple sclerosis, manifesting itself mainly as painful diffuse field loss or central scotomas. However, it could be encountered in various other inflammatory, demyelinating, infectious, and autoimmune conditions that could be broadly classified into two groups: typical and atypical ON. On the contrary, painless vision loss, especially altitudinal visual field defects (AVD), is commonly observed in ischemic optic neuropathies (ION), mostly in patients with vascular risk factors or history of giant cell arteritis. Although rare, AVD can be the initial sign of ON and inflammatory demyelinating process. Herein, we report a case of a 17-year-old patient with ON presenting with painless AVD and provide a brief review of the mechanisms involved in typical and atypical ON and ION.

Keywords: Optic neuritis, ischemic optic neuropathy, altitudinal visual defect

Introduction

Optic neuritis (ON) is the most common optic neuropathy in young adults and is characterized by the inflammation of the optic nerve. ON is seen in various demyelinating, infectious, and inflammatory conditions. Typical ON is generally associated with multiple sclerosis (MS), which manifests as painful subacute vision loss. On the contrary, ischemic optic neuropathies (ION) result in painless monocular vision loss with altitudinal visual field defects (AVD), commonly seen in older people with vascular risk factors. Herein, we present the case of a young female patient with painless AVD as the initial sign of ON and provide a brief review of ON and ION.

Case Report

A 17-year-old previously healthy female patient comes to the clinic due to blurry vision on her right eye that suddenly occurred two days ago. Eye movements were not painful, and she had no headache or any other neurological symptoms. She had no history of trauma and denied any recent infections, fever, or constitutional symptoms. Both her medical and family histories were unremarkable.

A detailed neurological examination was performed. Both direct and indirect pupillary light reflexes were normal; however, she had a mild relative afferent pupil defect on her right eye. Other cranial nerves and results of motor, sensory, and cerebellar examinations were normal. During the examination, she stated that for the last few days she would feel an electrical-like sensation extending down her spine when she flexed her neck, which was suggestive of Lhermitte's sign. On her ophthalmological examination, her intraocular pressure was normal, no signs of anterior compartment pathologies, retinitis, or vitritis were noted, and the optic disc was normal. Her visual acuity was 20/20 for both eyes on the Snellen chart. However, her visual field test revealed superior AVD in the right eye (Figure 1).

Complete blood count, B_{12} levels, comprehensive metabolic panel, erythrocyte sedimentation rate, angiotensin-converting enzyme level, and urinalysis parameters were in the normal range. *Borrelia*, *Treponema*, human T-cell leukemia virus, and human immunodeficiency virus serology were negative. Results of a wide range of autoantibody tests were normal. Chest X-ray imaging excluded infections and connective tissue

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disorders, and the findings were normal. A lumbar puncture was performed. The cerebrospinal fluid (CSF) was clear, with a normal opening pressure. CSF biochemistry and cytology were normal. Anti-aquaporin-4 antibody was found to be negative both in the CSF and serum. Anti-myelin oligodendrocyte glycoprotein immunoglobulin G (IgG) was negative. Nonetheless, CSF electrophoresis revealed oligoclonal bands, and the IgG index was 0.78. Visual evoked potential (VEP) revealed delayed P100 latency of the right eye. Cranial magnetic resonance imaging (MRI) demonstrated single ovoid hyperintensity perpendicular to the corpus callosum and contrast enhancement of the retroorbital segment of the right optic nerve, compatible with ON (Figure 2). The patient was admitted to the inpatient clinic and received 5 days of intravenous methylprednisolone 1,000 mg/day. After one month, the patient's vision was fully recovered, and her ophthalmological examination was completely normal. She had no other neurological deficits. Six months later, the follow-up MRI revealed new non-contrast enhancing hyperintense lesions in the right parietal white matter and C3-C4 vertebral level. Written informed consent of the patient was obtained.

Discussion

ON occurs through different mechanisms that could be categorized into two distinct groups: typical ON and atypical ON. The hallmark of typical ON is acute inflammatory demyelination, followed by axonal loss in the long term (1). It is mostly associated with MS; essentially, ON is the initial presentation in 20% of the MS, and 50% of patients with MS experienced ON during the disease course (2). Although its pathophysiological mechanism has not been fully elucidated, peripheral T-cell activation and migration toward the blood-brain barrier is thought to initiate this inflammatory process. This activation leads to a type 4 hypersensitivity-like reaction and results in myelin damage and axonal degeneration (3).

More than 90% of typical ON presents with painful monocular vision loss. Dyschromatopsia, reduced contrast acuity, and central visual defects are also among the common presentations of ON. According to the ON treatment trial, 97.5% of the patients experienced visual field defects, central scotomas being the most common entity, at some point of the disease course (4). Vision loss varies, ranging from mild to complete loss, can last up to 6 months, and mostly reaches its peak within two weeks after onset. It manifests mostly as retrobulbar neuritis in which the optic disc is normal or rarely as papillitis which involves the anterior optic nerve and results in disc swelling (5).

On the contrary, atypical ON is mostly caused by neuromyelitis optica spectrum disorders (NMOSD), other rare inflammatory conditions, and systemic diseases such as lupus, sarcoidosis, or Wegener's granulomatosis (3). NMOSD mostly presents with unilateral, sometimes bilateral rapidly sequential vision loss with painful eye movements. Although AVDs can be encountered in NMOSD, it is much more extensive, extending to the optic chiasm and optic tracts, and vision loss is much more persistent (6).

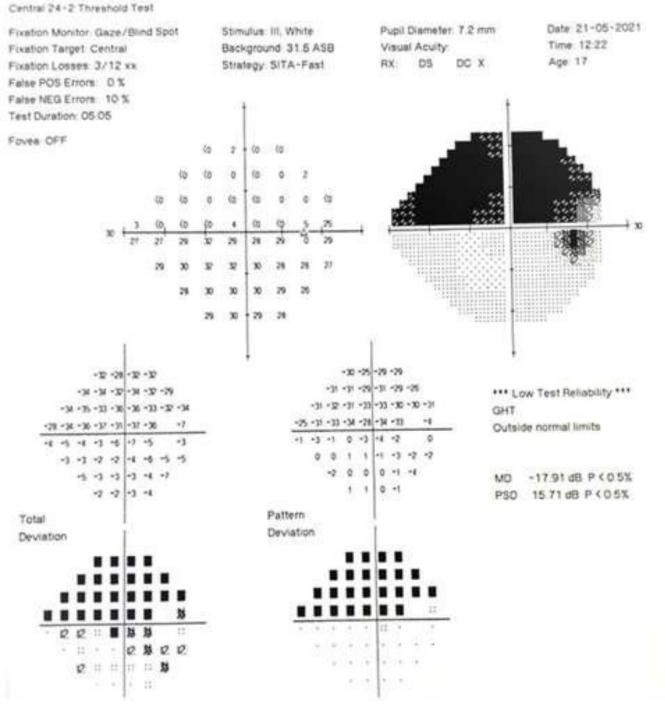

Figure 1. Visual field test revealing right superior altitudinal visual field defect

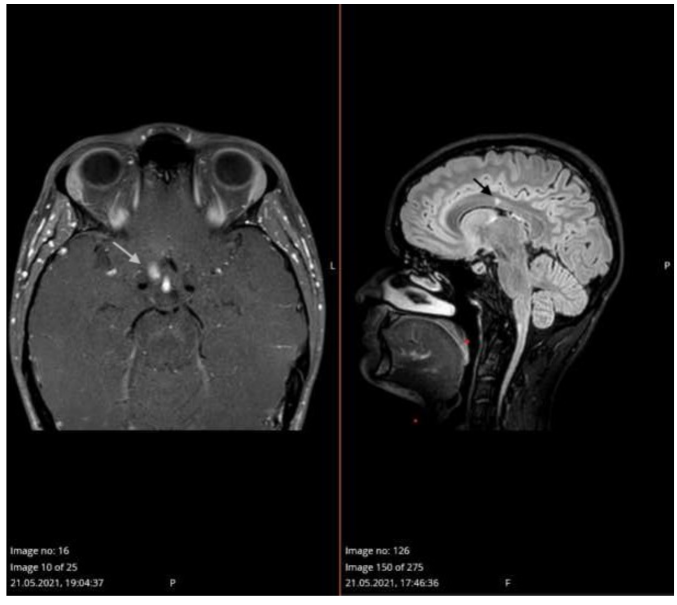

Figure 2. Orbital MRI showing contrast enhancing lesion in the right optic nerve (left) and cranial sagittal MRI showing single ovoid hyperintensity perpendicular to the corpus callosum (right) in T2-weighted images

MRI: Magnetic resonance imaging

Painless unilateral vision loss with AVD mostly suggests ION. Anterior ION (AION) is the most common cause of vision loss in older people and is divided into arteritic and non-arteritic (7). Arteritic AION is almost always seen with giant cell arteritis, whereas non-arteritic AION is seen in patients with vasculopathic risk factors such as diabetes, hypercholesterolemia, and hypertension. Fundoscopic examination typically reveals optic disc edema. Taken together, our patient's age, medical history, and fundoscopic examination are not compatible with AION. Although posterior ION presents with normal optic disc findings, as observed in our patient, the absence of recent ocular surgery or history of giant cell arteritis excludes this diagnosis (8).

At her initial admission, her MRI findings did not fulfill the McDonald criteria; however, the positive findings on her follow-up MRIs in the subsequent year, together with CSF, VEP, and laboratory findings, raised strong suspicion for MS.

Conclusion

Although the painless nature of the visual loss and its characteristic findings mostly suggest an ischemic or vasculitis cause, our patient represents a rare phenomenon. This case emphasized that AVDs should be considered in the decision-making process and could be observed as the foremost findings of an inflammatory process.

Ethics

Informed Consent: Written informed consent of the patient was obtained.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: R.C.A., H.S., Design: R.C.A., C.B., S.C., H.S., Data Collection or Processing: R.C.A., C.B., H.S., Analysis or Interpretation: R.C.A., H.S., Literature Search: R.C.A., C.B., H.S., Writing: R.C.A., S.C., H.S.

Conflict of Interest: No conflict of interest was declared by the authors.

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References

1. Toosy AT, Mason DF, Miller DH. Optic neuritis. Lancet Neurol 2014;13:83-99.
2. Kale N. Optic neuritis as an early sign of multiple sclerosis. Eye Brain 2016;8:195-202.
3. Pau D, Al Zubidi N, Yalamanchili S, Plant GT, Lee AG. Optic neuritis. Eye (Lond) 2011;25:833-842.
4. Frohman EM, Frohman TC, Zee DS, McColl R, Galetta S. The neuro-ophthalmology of multiple sclerosis. Lancet Neurol 2005;4:111-121.
5. Wilhelm H, Schabet M. The Diagnosis and Treatment of Optic Neuritis. Dtsch Arztebl Int 2015;112:616-625; quiz 626.
6. Levin MH, Bennett JL, Verkman AS. Optic neuritis in neuromyelitis optica. Prog Retin Eye Res 2013;36:159-171.
7. Morrow MJ. Ischemic Optic Neuropathy. Continuum (Minneap Minn) 2019;25:1215-1235.
8. Atkins EJ, Bruce BB, Newman NJ, Biousse V. Treatment of Nonarteritic Anterior ischemic optic neuropathy. Sur Ophthalmol 2010;55:47-63.