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Simultaneous Bilateral Nonarteritic Anterior Ischemic Optic Neuropathy in a Patient with a History of Diffuse Large B-Cell Lymphoma

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ABSTRACT

Background: Nonarteritic anterior ischemic optic neuropathy (NAAION) has a poorly understood etiology, and the onset of simultaneous bilateral NAAION in a patient <50 years without identifiable systemic risk factors is rare.

Case Report: We present the case of a patient with acute painless monocular vision loss and bilateral optic disc edema who subsequently developed painless vision loss in the fellow eye. The patient's history was significant for diffuse large B-cell lymphoma, and our pressing diagnostic concern was to determine if his vision loss and bilateral optic disc changes represented lymphomatous infiltrates. A complete ocular exam demonstrated findings consistent with simultaneous bilateral NAAION. After an extensive systemic workup for malignancy with central nervous system involvement, vasculitis, and other entities associated with NAAION, we determined that the patient's primary risk factor for developing bilateral ischemic optic neuropathies was his crowded optic discs.

Conclusion: This case supports the hypothesis that a crowded optic disc is a sufficient primary risk factor for developing NAAION.

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INTRODUCTION

Nonarteritic anterior ischemic optic neuropathy (NAAION) has a poorly understood etiology, but several risk factors have been identified, including atherosclerosis, diabetes mellitus, hyperlipidemia, hypertension, hypotension, hemoconcentration, hemodilution, hypercoagulable states, and a crowded optic disc.[1,2] Our patient presented with symptomatic vision loss and NAAION in one eye, and the fellow eye demonstrated pre-NAAION disc edema without vision loss. The onset of simultaneous bilateral NAAION in a patient <50 years of age and without identifiable systemic risk factors is rare and suggests that a crowded optic disc with edema is sufficient to precipitate ischemic optic neuropathy.[3]

CASE REPORT

A 49-year-old white male presented with acute painless vision loss in the left eye. His vision had been deteriorating during the previous 10 days. He stated he was feeling well otherwise and denied trauma, medication, or illicit drug use. A complete ocular exam demonstrated the following pertinent findings: visual acuity of 20/30 in each eye, full motility, and a left afferent pupillary defect. The anterior segment and vitreous were quiet without cell or flare in both eyes. A dilated fundus examination demonstrated crowded discs with optic disc edema in the right eye and optic disc edema with hemorrhage in the left eye (Figure 1). An intravenous fluorescein angiogram demonstrated absence of autofluorescence but late bilateral optic disc staining consistent with edema (Figure 2). A Humphrey Visual Field (HVF) (Carl Zeiss Meditec) 24-2 Swedish interactive threshold algorithm (SITA) standard test demonstrated general depression with an inferior arcuate scotoma in the left eye and a full field in the right eye (Figure 3).

The patient's medical history was significant for diffuse large B-cell lymphoma for which he had completed 6 cycles of etoposide, prednisone, Onco-

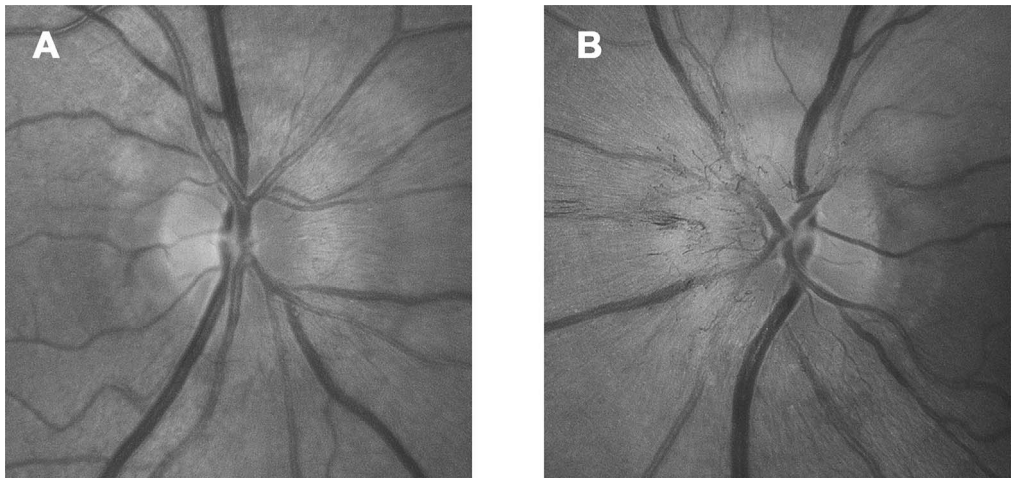

Figure 1. Fundus photographs demonstrate bilateral crowded discs with optic disc edema in the right eye (A) and optic disc edema with hemorrhage in the left eye (B).

vin (vincristine), cyclophosphamide, hydroxydaunorubicin (EPOCH)-rituximab chemotherapy 2 months prior to presentation. A recent follow-up with his oncologist deemed him clear of disease, and observation was recommended at this stage of recovery. Given the patient's history of lymphoma and bilateral optic disc edema, he was admitted and evaluated for central nervous system (CNS) metastatic disease. The patient underwent systemic evaluation with a complete blood count, serial lumbar punctures including opening pressure, and an infectious disease workup, all of which were normal. Magnetic resonance imaging of the brain and orbits demonstrated evidence of an abnormal edema signal and enhancement along the optic nerve pathway bilaterally but no signs of obstruction to cerebrospinal fluid flow. During the initial hospital admission and subsequently during the next 3 months, serial diagnostic positron emission tomography/computed tomography scans failed to identify signals suggestive of residual or recurrent neoplastic disease.

Following discharge from the hospital, the patient returned 2 weeks later with a new complaint of painless vision loss in the right eye. A dilated eye exam demonstrated an interval change with worsening optic disc edema with hemorrhage in the right eye and improving optic disc edema in the left eye (Figure 4). An HVF 24-2 SITA standard test was performed and demonstrated new inferior and superior arcuate scotomas in the right eye and a stable inferior arcuate scotoma in the left eye.

During a 1-year period, serial ocular exams demonstrated resolution of bilateral optic disc edema with residual pallor and no further progression of visual field loss. The patient continued to do well without recurrence of his diffuse large B-cell lymphoma. The

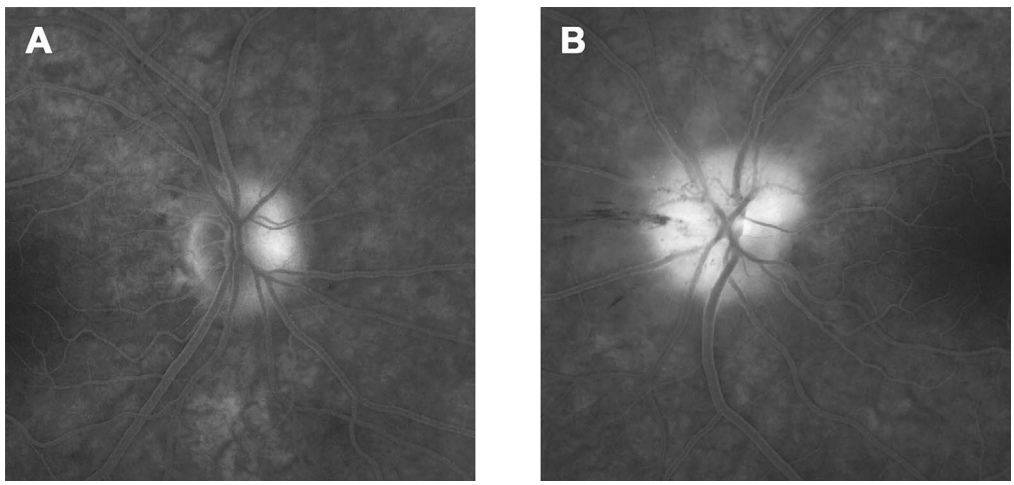

Figure 2. An intravenous fluorescein angiogram demonstrates late bilateral optic disc staining consistent with edema in the right eye (A) and left eye (B).

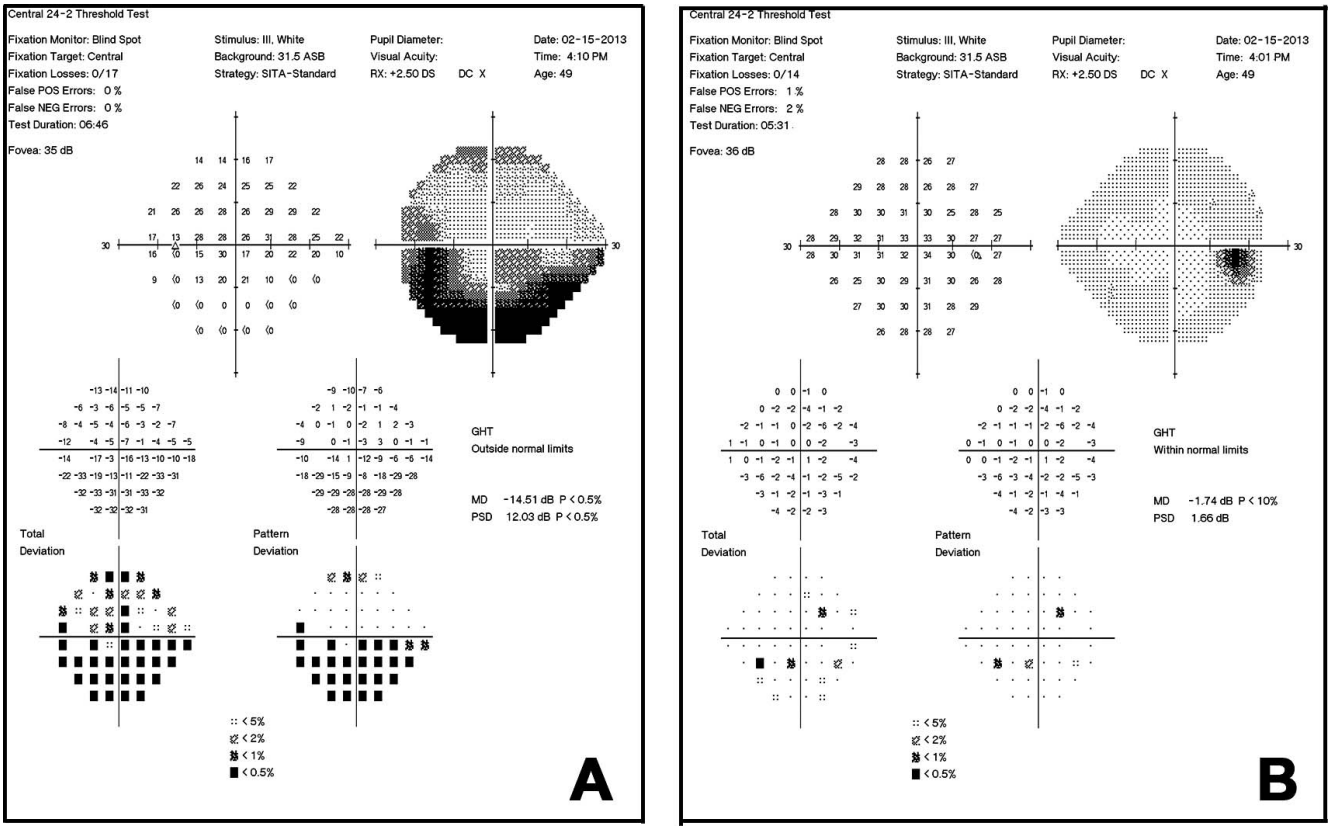

Figure 3. Humphrey Visual Field 24-2 Swedish interactive threshold algorithm standard test demonstrates general depression with an inferior arcuate scotoma in the left eye (A) and a full field in the right eye (B).

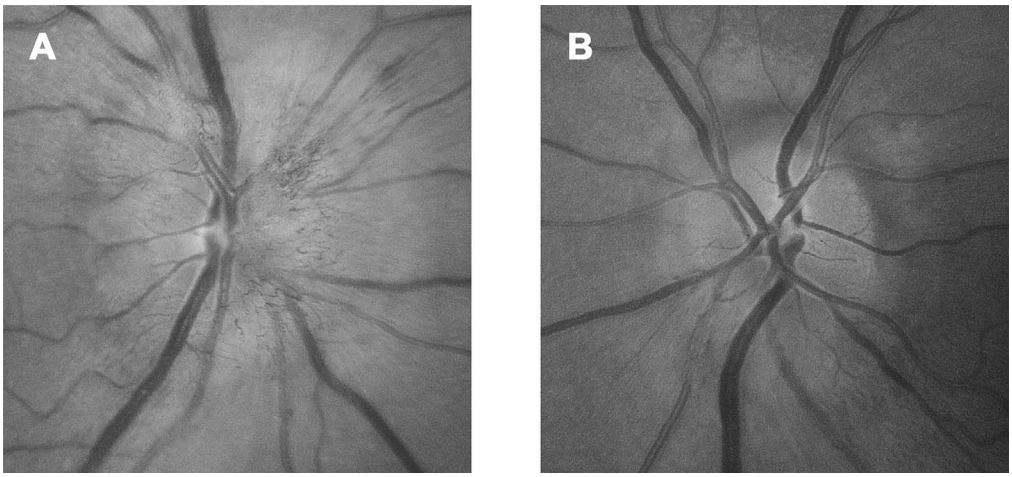

Figure 4. Fundus photographs demonstrate an interval change with worsening optic disc edema with hemorrhage in the right eye (A) and improving optic disc edema in the left eye (B).

diagnosis of bilateral NAAION was established, and the concern for CNS lymphoma was extinguished.

DISCUSSION

The presentation of simultaneous, asymmetric, bilateral NAAION is uncommon and is described in the literature as associated with systemic conditions that cause altered blood flow to the optic disc, such as essential thrombocytosis,[4] hemorrhage,[5] and use of sildenafil.[6] Given our patient's history, significant for diffuse large B-cell lymphoma, we performed an extensive medical and imaging workup that revealed no known risk factors for NAAION, including medication use, sleep apnea, optic disc drusen, blood disorders, CNS fluid abnormalities, autoimmune markers, cranial masses, or infection. However, the diagnosis of intraocular lymphoma cannot be completely excluded. For this reason, the recommendation for patients at risk of intraocular malignancy is to undergo long-term follow-up and surveillance.

Of interest in our patient's presentation is that the asymptomatic fellow eye demonstrated optic disc edema and 2 weeks later developed vision loss. This observation provides further evidence that disc edema in a crowded nerve head is intimately related to the pathogenesis of NAAION.[7] The risk factor for developing bilateral NAAION in our patient $<$50 years of age was crowded optic discs.

CONCLUSION

This case supports the hypothesis that a crowded optic disc is a sufficient primary risk factor for developing NAAION.

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