

Clinical Reasoning:

A 61-year-old woman with a swollen optic nerve and progressive visual loss

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SECTION 1

A 61-year-old Caucasian woman was referred for evaluation of a nonarteritic anterior ischemic optic neuropathy (NAION) of the right eye. Two months before the referral, she experienced the acute onset of painless blurred vision in the right eye for which she sought the care of a local ophthalmologist. Visual acuity was recorded as 20/30 in the right eye with optic disc edema and multiple peripapillary flame hemorrhages. Erythrocyte sedimentation rate and C-reactive protein were within normal limits. MRI of the brain with contrast was read as normal. Her medical history was notable for arterial hypertension, hyperlipidemia, and carpal tunnel syndrome. Her systemic medications included atorvastatin, diltiazem, and spironolactone. She denied alcohol, tobacco, and IV drug use. Her family history was unremarkable. A complete review of systems was negative except for the blurred vision in the right eye.

When seen in the neuro-ophthalmology clinic, visual acuity was 20/70 in the right eye and 20/20 in the left eye. There was a right relative afferent pupillary defect. When presented with the Ishihara pseudoisochromatic color plates, she was unable to identify the control plate with the right eye and properly identified 7.5 of 10 color plates with the left eye. Eye movements were full in both eyes. Slit-lamp examination revealed early nuclear sclerosis of the crystalline lens in both eyes. Dilated fundus examination revealed florid optic disc edema with cotton wool spots and peripapillary flame hemorrhages in the right eye (not shown). The left optic nerve was normal. The peripheral retina and macula were normal in both eyes.

Questions for consideration:

1. What is the differential diagnosis of unilateral optic disc edema?
2. What further clinical testing would you perform?

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SECTION 2

This 61-year-old woman presents with progressive, painless vision loss in the right eye associated with right optic disc edema. The differential diagnosis of unilateral optic disc edema includes central retinal vein occlusion, optic neuritis, arteritic anterior ischemic optic neuropathy or NAION, a compressive optic neuropathy, and neuroretinitis. The history of visual loss, clinical examination, and paraclinical testing can narrow the differential diagnosis. In our patient, there were no peripheral intraretinal hemorrhages to suggest a retinal vein occlusion. Her age (older than 45 years) and lack of eye pain with extraocular movement makes optic neuritis unlikely. It would be very atypical for this to be arteritic ischemic optic neuropathy (i.e., giant cell arteritis) in the setting of normal erythrocyte sedimentation rate, normal C-reactive protein, and the absence

of headache and jaw claudication. Although the mean age of patients with NAION is 60 years, which fits with our patient, the progressive visual loss and persistent optic disc edema for 2 months are not characteristic of NAION.¹ Finally, she has no exudates in the macula in a star configuration that would be indicative of neuroretinitis.²

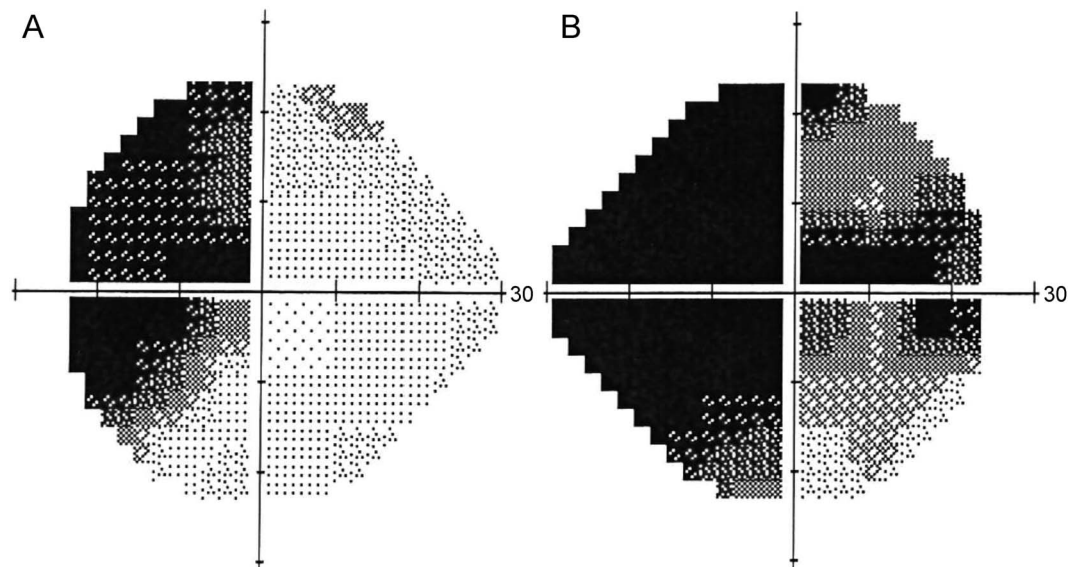
Automated visual field testing in the right eye demonstrated a central scotoma in addition to a defect in the nasal greater than the temporal field. In the left eye, there was a dense temporal defect, worse in the superior than in the inferior quadrants, respecting the vertical meridian (figure 1).

Question for consideration:

1. Given the pattern of visual loss, where would you anticipate the location of the lesion?

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Figure 1 Automated static perimetry of the left (A) and right (B) eyes



In the left eye, there is a dense temporal defect, worse in the superior than in the inferior quadrants, respecting the vertical meridian. In the right eye, there is a central scotoma with a dense nasal and slightly less dense temporal defect. The pattern of visual field loss is consistent with an optic nerve–optic chiasm–optic tract syndrome.

SECTION 3

The pattern of vision loss is consistent with a left incongruous homonymous hemianopsia with a central scotoma in the right eye, localizing the lesion to the right optic nerve–optic chiasm–optic tract. The optic tract contains ipsilateral temporal retinal fibers and contralateral nasal retinal fibers that decussate in the optic chiasm. Each optic tract processes the contralateral visual field and, if damaged, can lead to a homonymous field defect, which often is incongruous. Therefore, an optic tract syndrome is characterized by a contralateral incongruous homonymous hemianopsia, as well as a contralateral relative afferent pupillary defect and contralateral bow-tie optic nerve pallor.³ The central scotoma in the right eye with a superotemporal hemianopsia in the contralateral left

eye suggests that part of the lesion involves the right optic nerve and anterior optic chiasm. The unique pattern of visual loss in our patient implicates, at a minimum, involvement of the optic chiasm.

The differential diagnosis of a lesion of the chiasm, from more common to less prevalent, includes intracranial mass (i.e., pituitary adenoma, suprasellar mass or cyst, meningioma, glioma), neoplasm (i.e., lymphoma, leukemia, optic nerve sheath meningioma, or optic nerve glioma), inflammation (i.e., chiasmal neuritis, sarcoidosis), head trauma, ophthalmic artery aneurysm, and infection (i.e., abscess, bacterial, fungal, or viral infection).

Question for consideration:

1. What studies and/or tests do you want to perform?

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SECTION 4

Two weeks later, the patient's vision declined to count fingers with persistent optic nerve head edema. A review of the outside MRI (that was previously read as normal) revealed abnormal enhancement of the intracranial portion of the right optic nerve. A repeat MRI of the brain and orbits revealed profound enhancement of the right optic nerve and the right aspect of the optic chiasm, extending to the right optic tract up to the lateral geniculate nucleus (figure 2). Because the lesion appeared well-delineated along the visual pathway, an intrinsic neoplasm of the optic pathways was highly suspected. A primary CNS neoplasm, such as lymphoma, would have more diffusely involved the brain parenchyma and/or the leptomeninges. Therefore, based on the neuroimaging findings, a lumbar puncture was not pursued. A right pterional craniotomy with exploration and excisional biopsy confirmed an optic pathway glioblastoma multiforme or malignant optic glioma of adulthood (MOGA).

DISCUSSION In 1973, 5 cases of MOGA and 10 additional cases from the literature were reviewed.⁴ The clinical presentation was one of rapid progressive vision loss resulting in a fatal outcome within several months. MOGAs are clinically very distinct from the more common benign childhood optic nerve gliomas

that are frequently associated with neurofibromatosis type I.⁵ A review of 44 published cases of MOGA revealed that 51% of cases were male, mean age at diagnosis was 54 years, and at onset 70% had unilateral decreased vision and 30% had bilateral vision loss. At presentation, 41% had optic disc edema, 43% had visual field defects, and 20% had ocular pain. The mean interval from initial symptoms to total blindness was 3.3 ± 2.8 months (range 2 weeks to 12 months).⁶

At presentation, MOGA may mimic optic neuritis, NAION, or arteritic anterior ischemic optic neuropathy, but as mentioned above, the history, clinical examination, and paraclinical testing can help differentiate these entities from one another. Patients with progressive visual loss associated with persistent optic disc edema should undergo neuroimaging, in particular cranial and orbital MRI with contrast and fat suppression technique.^{3,7} The neuroimaging appearance of MOGA is nonspecific; occasionally the optic nerve may be diffusely thickened and may show marked enhancement with heterogeneous and cystic areas.⁸

The prognosis for patients with MOGA is unfortunately very dismal. The median survival is approximately 12 months, with only 3% to 5% of patients considered long-term survivors, defined as survival greater than 3 years. Glioblastoma long-term survival is associated with younger age, good initial performance score, and *O*⁶-methylguanine methyltransferase promoter hypermethylation, a clinically significant molecular marker.⁹ Despite modern techniques of surgical resection, chemotherapy, and radiation, there has been no substantial improvement in the survival rate of patients with MOGA.

Follow-up. Our patient received multiple rounds of radiation and adjuvant chemotherapy; however, she eventually declined treatment, opting instead for hospice care. She died 10 months after initial presentation.

AUTHOR CONTRIBUTIONS

Paula Pecen: drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and will give final approval, acquisition of data. M. Tariq Bhatti: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and will give final approval, study supervision, obtaining funding.

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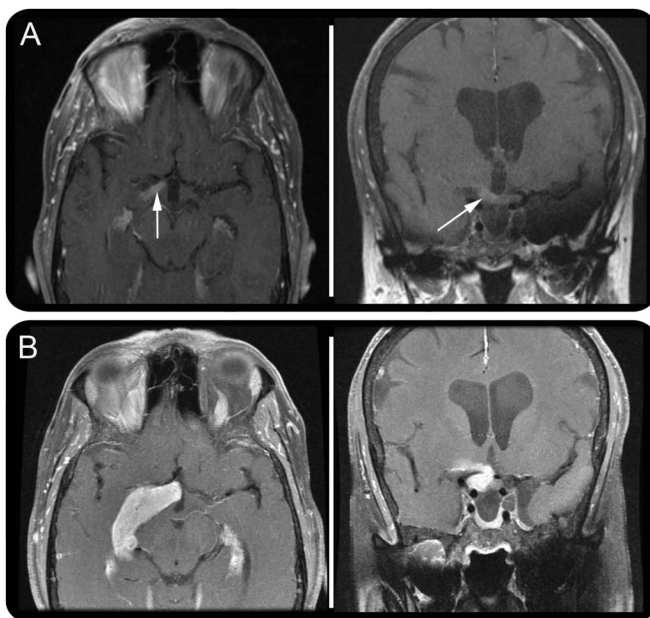
DISCLOSURE

P. Pecen reports no disclosures. M. Bhatti is a consultant for Alexion and Novartis. Go to Neurology.org for full disclosures.

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Figure 2 MRI of the brain and orbits with contrast at the level of the visual pathways, axial and coronal T1-weighted images



(A) Initial MRI was read as unremarkable, but repeat review revealed abnormal enhancement of the intracranial portion of the right optic tract and right side of the chiasm (arrows). (B) Follow-up MRI 6 weeks later demonstrates enhancement and enlargement of the right optic nerve, optic chiasm, and optic tract extending to the lateral geniculate nucleus.

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