Progressive Visual Field Loss in the Setting of Normalized IOP

BY SHAKEEL SHAREEF, MD, AND MORGAN RENNER, MD, MPH

CASE PRESENTATION

A 17-year-old girl diagnosed with common variable immunodeficiency hypogammaglobulinemia at birth presented at the Flaum Eye Institute in Rochester, New York, in August 2012 after starting hydroxychloroquine (Plaquenil; Sanofi Aventis US) 3 months prior. The patient had received a prescription for oral prednisone 5 mg for 5 years to treat a restrictive lung disease and allow her to breathe. An attempt was being made to wean her off steroids. Upon examination, a Humphrey 10-2 visual field test (Carl Zeiss Meditec) and macular optical coherence tomography (OCT) scan were within normal limits for both eyes.

One week later, after complaining of a headache, red eyes, and blurred vision, the patient discontinued hydroxychloroquine, and the symptoms resolved. The patient's internist restarted the medication. A recurrence of the same symptoms was noted 2 weeks later, and she was referred back for an evaluation. Upon examination, the patient's visual acuity was 20/40 OD and 20/30 OS, pachymetry readings were 625 to 640 µm OU, and IOP by applanation tonometry was 50 mm Hg OD and 48 mm Hg OS. Intense topical and oral hypotensive therapy was initiated followed by an immediate referral to the Glaucoma Service.

The patient displayed baseline dyspnea in the examination chair. A Humphrey 30-2 Swedish interactive thresholding algorithm standard visual field test revealed a dense inferior arcuate scotoma in the right eye and a ring scotoma sparing the central 10° in the left eye (Figure 1). OCT of the retinal nerve fiber layer revealed generalized loss to 1% of the normal nerve fiber layer in both eyes. The IOP measured 30 mm Hg OU. The pupil was reactive in the right eye but sluggish and larger in size in the left eye. Gonioscopy revealed open angles to the ciliary body band in both eyes. The patient started using brinzolamide ophthalmic suspension 1% (Azopt; Alcon) three times daily and travoprost (Travatan; Alcon) at bedtime in both eyes. We did not prescribe β-blockers.

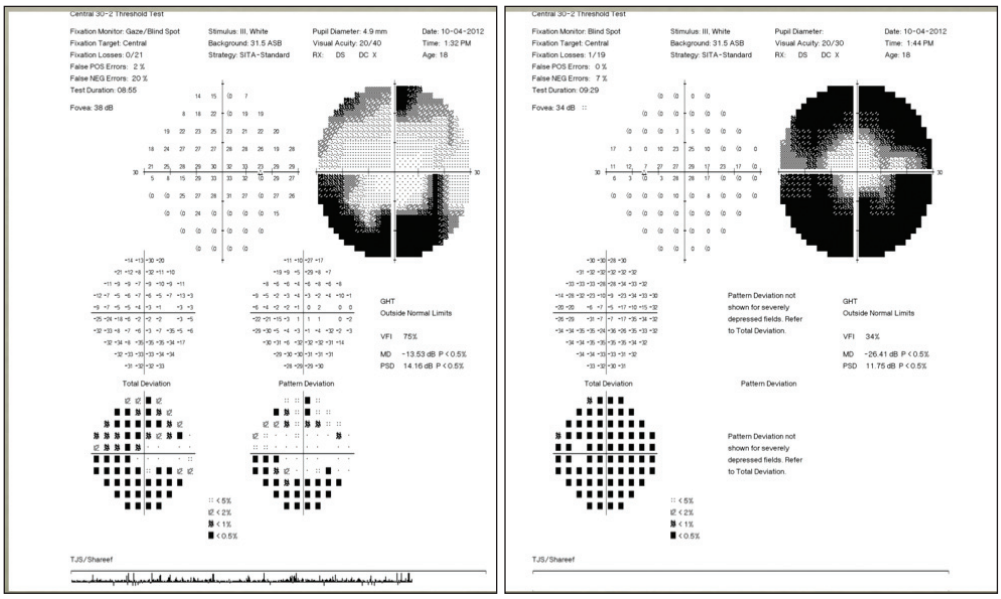

Figure 1. A Humphrey 30-2 Swedish interactive thresholding algorithm standard visual field test of the patient's right and left eyes following her referral.

Two weeks later, the patient's IOP measured 29 mm Hg OD and 37 mm Hg OS. In November 2012, we performed a

TABLE. SUMMARY OF IOP (OCTOBER 2012-SEPTEMBER 2013)

Date	OD (mm Hg)	OS (mm Hg)
October 4, 2012	50	48
November 8, 2012	38	7 (status post trabeculectomy November 7)
November 15, 2012	42	5
December 6, 2012	10 (status post trabeculectomy December 5)	7
December 13, 2012	8	5
February 5, 2013	6	3
April 2, 2013	6	5
August 30, 2013	11	4 (drop in visual acuity to 20/400)
September 27, 2013	10	2 (visual loss 11 months status post trabeculectomy)

trabeculectomy and applied a cotton pledget soaked in a 0.2-mg/mL solution of mitomycin C for 2.5 minutes to the left eye. The IOP normalized. We performed a similar surgery 1 month later on the right eye (IOP was 42 mm Hg), which resulted in good postoperative IOP control. The IOP remained in the single digits for both eyes through April 2013 (Table).

In August 2013, the visual acuity in the patient's left eye dropped to 20/200. Peripheral iridocorneal touch was noted with two to three times corneal thickness anterior chamber (AC) depth observed centrally. A dilated examination of the left eye was negative for choroidal effusions, and macular OCT was normal for both eyes. The AC remained within normal limits in the right eye. Goldmann visual field testing revealed a central island of vision of less than 5° in the left eye and marked constriction in the right eye (Figure 2A). In September 2013, a repeat Goldmann visual field test showed further disease progression in the right eye, and despite a visual acuity of 20/150, the left eye did not respond to any stimulus (Figure 2B).

HOW WOULD YOU PROCEED?

- Would you evaluate this patient's blindness for a nonglaucomatous etiology(ies), and if so, what workup would you propose?
- Would you take any steps to prevent the right eye from succumbing to a similar outcome as the left?

CLINICAL COURSE

With input from other medical subspecialists, the patient underwent a thorough workup, including blood pressure and pulse monitoring by her internist to rule out ocular hypoxia/hypoperfusion, imaging studies by a neuro-ophthalmologist to rule out an extraocular cause, a retinal consultation to rule out autoimmune retinopathy, and sleep laboratory studies to assess her for sleep apnea (Figure 3).

OUTCOME

The patient's blood pressure and pulse remained within the normal range. Magnetic resonance imaging did not identify intracerebral or intraorbital compressive or aneurysmal lesions to account for a progressive neuropathy. An electroretinogram (ERG) revealed no evidence of autoimmune retinopathy or retinal degeneration under photopic and scotopic conditions. An overnight sleep study revealed 14 respiratory events with an obstructive apnea-hypopnea index of 2.1 events per hour. Pulmonary function tests showed a forced expiratory volume in 1 second 35% to 39% and a forced vital capacity 40% of predicted. Oxyhemoglobin saturations were within the low 90s with sustained drifts into the upper 80s. The patient's oxygen saturation dropped below 90% for 7.9% of the night. These findings were consistent with sleep hypoxemia and mild obstructive sleep apnea. Home oxygen therapy at night was recommended for the patient.

Despite the therapy, the most recent visual field test of the patient's right eye (August 2014) revealed visual field progression similar to in the left eye (Figure 2B). Upon consultation with the American Glaucoma Society, there was a unanimous recommendation that the patient be referred for a low vision evaluation.

DISCUSSION

Progressive visual loss in both of this patient's eyes occurred despite a normalized IOP in the single digits for

9 to 10 months after a trabeculectomy. She succumbed to blindness in the left eye within 1 year of surgery. Several publications have described central visual loss in patients presenting with preoperative split fixation (Figure 1).[1-3] Central visual loss has also been reported to correlate with a high mean preoperative IOP.[2] In addition, hypotony maculopathy has been attributed to central visual loss.[2,3] The patient in this case had a shallow AC in the left eye, a normal macular OCT scan, and an absence of choroidals. The AC in the right eye remained within normal limits. Given her restrictive lung disease, she experienced daily bouts of excessive coughing, which might have contributed to hypotony. Surgical options were limited in the setting of restrictive lung disease and

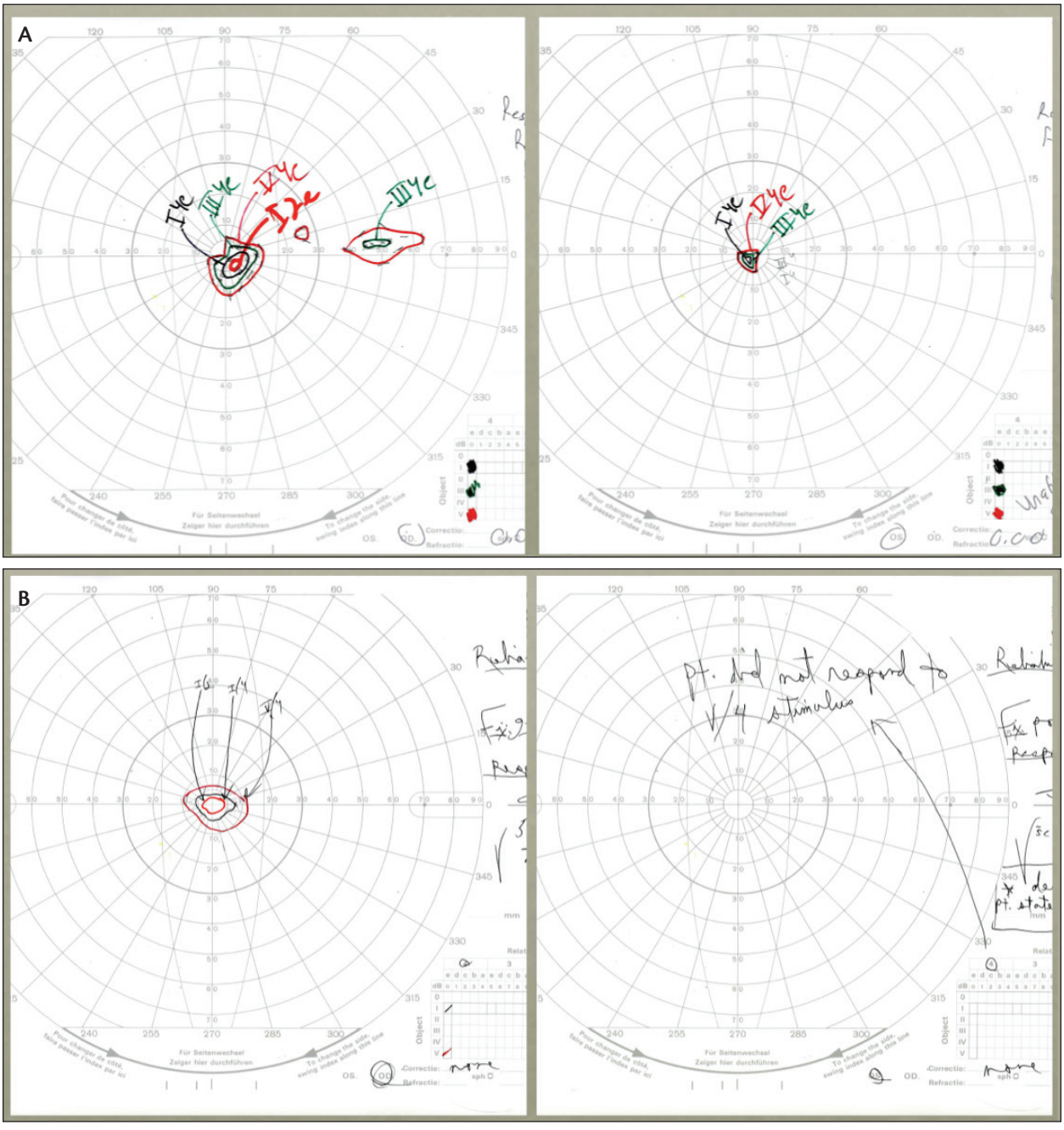

Figure 2. Right and left eyes, respectively. Note that marked constriction to within 10° and split fixation in the left eye is greater than in the right eye, with a temporal island in the right eye (A). Repeat Goldmann visual field testing revealed the loss of a temporal island in the right eye, and the left eye was unresponsive to stimulus (B).

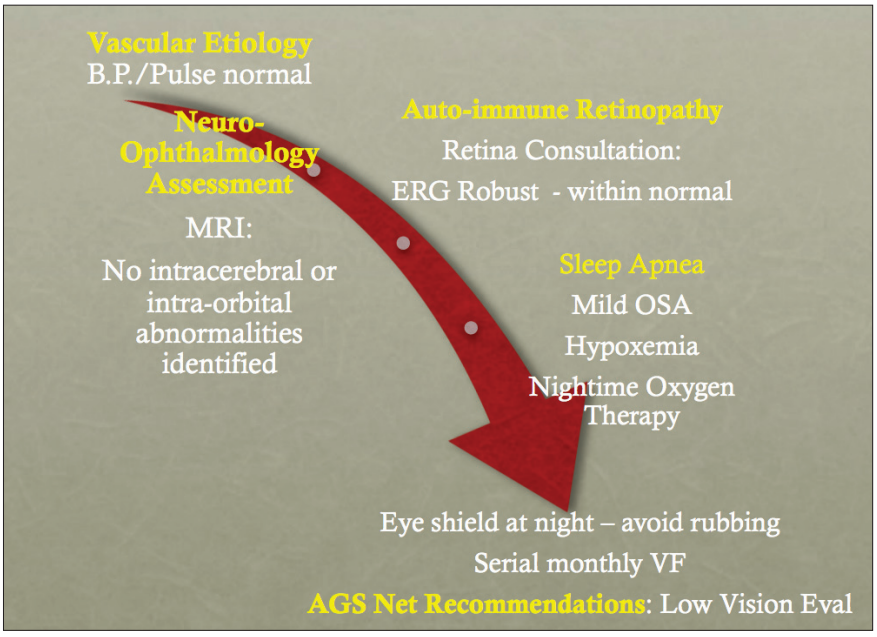

Figure 3. Differential of visual loss despite normalized IOP due to nonglaucomatous etiology. Abbreviations: BP, blood pressure; MRI, magnetic resonance imaging; ERG, electroretinogram; OSA, obstructive sleep apnea; VF, visual field; AGS, American Glaucoma Society.

multiple ER visits and hospitalizations.

Pecora et al described progressive visual field loss in both eyes with blindness in the left eye over 4 years in a patient with a normal IOP.[4] After an exhaustive workup, the patient was reported to engage in excessive eye rubbing 10 to 20 times a day. With cessation of the rubbing, the visual field progression stopped in the right eye, resulting in stable visual field tests over 8 years. Our patient was noted to sleep 13 to 14 hours each night with frequent napping. We recommended an eye shield for the right eye to prevent either rubbing or external pressure while sleeping on a pillow.

Nonglaucomatous causes (Figure 3) such as vascular predictors of visual field progression include low systolic perfusion and systolic blood pressures and cardiovascular disease, which may contribute to ocular ischemia.[5] Our patient's serial pulse and blood pressure measurements were within normal limits.

Rapid visual field loss in both eyes despite IOP control over a 4-year span has been reported in a patient with open-angle glaucoma.[6] Immunomediated retinopathy characterized by rapidly decreasing vision was diagnosed with an abnormal ERG and antiretinal antibodies found in blood samples. Our patient had a normal ERG.

The presence of glaucoma-like cupping has been associated with neuro-ophthalmic conditions in the setting of a normal IOP.[7] These bulging areas may arise from aneurysmal compression of the optic nerve or compressive lesions involving the anterior visual pathway.[8] Magnetic resonance imaging was negative for these findings.

Given the complex nature of this case, it is possible that a combination of glaucomatous and nonglaucomatous processes caused the patient's blindness. Progressive lung disease may contribute further to her ocular ischemia with a guarded visual prognosis. It is important to help patients such as this one to maintain a positive attitude despite vision loss and to emphasize that there is life beyond blindness. In coordination with social services, the patient has been trained to use a walking cane, and she has enrolled in classes to learn Braille. Despite ophthalmologists' most valiant of efforts, some patients will succumb to blindness. It is a relief that cases such as this one are relatively uncommon. ■

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