

Disease Progression in Glaucoma

Some diagnostic pearls and controversies in management.

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CASE PRESENTATION

A 75-year-old white female was originally diagnosed with asymmetric open-angle glaucoma 17 years ago. At the time of diagnosis, her visual acuity was 20/25 OU with an IOP of 22 mm Hg OD and 27 mm Hg OS. Although the right optic disc appeared healthy, there was extensive excavation of the optic disc inferiorly in her left eye with a defect in the retinal nerve fiber layer (RNFL). Standard automated perimetry revealed a full visual field in her right eye and a dense, superior arcuate visual field defect that split fixation in her left eye (Figure 1).

Progressive loss of the left neural rim and widening of the visual field defect occurred despite maximal tolerated medical therapy and argon laser trabeculoplasty. A trabeculectomy with mitomycin C in the patient's left eye yielded an IOP of between 8 and 10 mm Hg. She subsequently developed a cataract requiring extraction. Her visual acuity improved initially to 20/30 OS but subsequently declined to 20/60 with the concurrent development of an epiretinal membrane. During a consultation, a vitreoretinal surgeon recommended a pars plana vitrectomy with epiretinal membrane peeling, but the patient elected not to proceed.

Comments on RNFL Defects and Epiretinal Membranes After Trabeculectomy

DSM: I would be interested to see whether the appearance of the RNFL defect changed after the IOP dropped. I have observed dramatic alterations, especially in young patients, but they are usually less obvious in adults. Changes in the defect's appearance occur when the live retina around the dead area swells, presumably due to axonal transport dysfunction,¹ and makes the nerve fiber defect more obvious. This phenomenon is similar to how the disc swells in

reaction to very low pressures.

RNW: Although I have not recognized this clinically, it has been reported that retinal height (as measured with the Heidelberg Retina Tomograph; Heidelberg Engineering GmbH, Dossenheim, Germany) and RNFL thickness (as measured with the scanning laser polarimeter² [Carl Zeiss Meditec Inc., Dublin, CA] or ocular

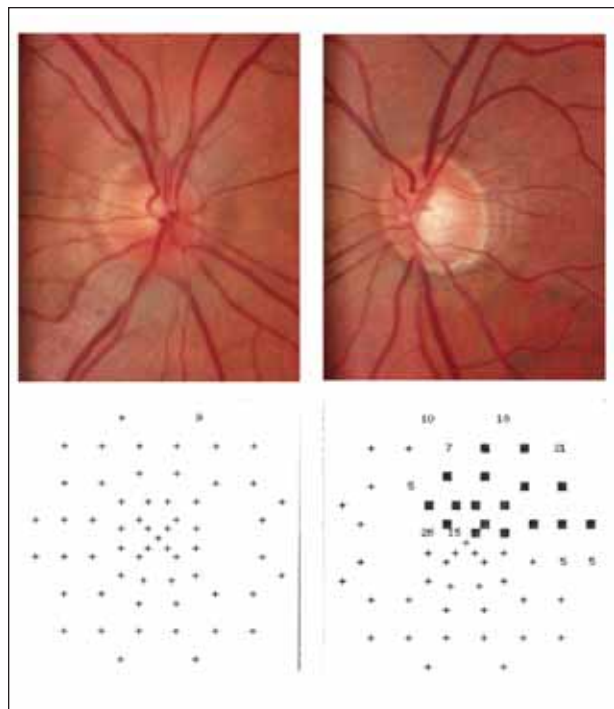


Figure 1. These optic disc photographs were taken in 1989 with corresponding difference plots from an Octopus 2000 (program G1) automated visual field (Interzeag AG, Schlieren, Switzerland). The patient's left eye displays marked thinning of the rim with a superior arcuate visual field defect and split fixation.

coherence tomography³) can increase after trabeculectomy. If these phenomena do occur, could a change in RNFL thickness guide our estimation of target IOP? An answer to this question is not known.

Comments on the Visual Symptoms and the Epiretinal Membrane

JGC: Epiretinal membranes typically cause visual distortion but not a loss of vision. Because the patient in this case did not complain of distortion, I think that her vision loss was due to glaucoma and not the epiretinal membrane.

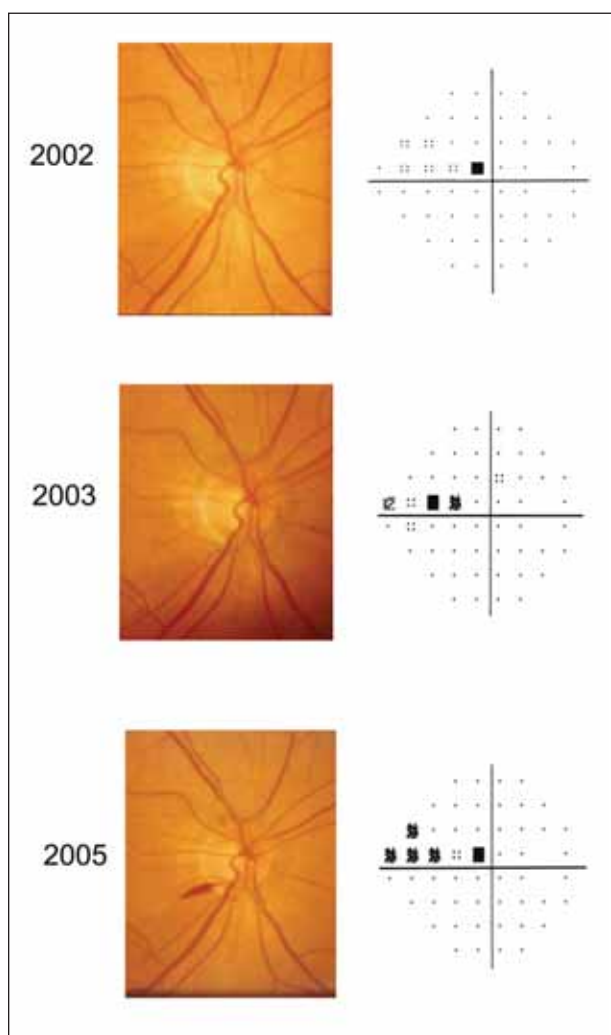


Figure 2. These optic disc photographs of the patient's right eye were taken from 2002 to 2005 with corresponding pattern deviation plots (from a 24-2 Humphrey Visual Field using the Swedish Interactive Threshold Algorithm-Standard [Carl Zeiss Meditec Inc.]). The development of the visual field defect corresponds with the progression of the optic rim's thinning, RNFL defects, and disc hemorrhages.

"With respect to the pathophysiology of disc hemorrhages, what has always made the most sense to me is a collapse of the lamina with a stretching and rupturing of the vessels."

—Donald S. Minckler, MD

Three years ago, the patient's right eye started to show evidence of glaucomatous progression, despite maximal tolerated medical therapy and argon laser trabeculoplasty. Initially, the physician noticed thinning of the inferior neural rim and RNFL defects, followed by recurrent optic disc hemorrhages and the development of a nasal defect on standard automated perimetry (Figure 2). The nasal defect then grew in size and depth so that it encroached on fixation at her last examination.

Comments on Disc Hemorrhages in Low-Tension Glaucoma

DSM: There is a nice correlation between the disc hemorrhages and the change in the visual field of this patient's right eye.⁴ A clear correlation is not always present.

With respect to the pathophysiology of disc hemorrhages, what has always made the most sense to me is a collapse of the lamina with a stretching and rupturing of the vessels. I do not believe that a disc hemorrhage⁴ causes or aggravates RNFL injury but rather that the hemorrhage reflects the collapse of the disc floor after axonal or glial injury. One may see disc hemorrhages in normal eyes as well as in areas corresponding to already dead retina.

Research has demonstrated relatively high percentages of disc hemorrhages in Japanese and white individuals with low-tension glaucoma.⁵⁻⁸ One might argue that patients who have low-tension glaucoma have vessels that are more prone to rupture at lower pressures.

JGC: One should be aware, however, of the potential for selection bias, because patients with low pressures are much more likely to be diagnosed with glaucoma on the basis of a disc hemorrhage.

RNW: Low-tension glaucoma is uncommon in white patients. When considering 24-hour, habitual (sitting during the day, supine at night) IOP measurements in a non-Asian patient, it is unusual for peak IOP to be less than 21 mm Hg.

The physician recommended trabeculectomy with mitomycin C for the patient's right eye on a number of occasions over a 3-year period, but the patient remained reluctant to proceed due to the preservation of good visual acuity in her right eye and poor vision in her left eye. She sought additional opinions concerning the timing of surgery from other glaucoma specialists.

Comments on Management

JGC: What would you currently recommend for this patient? She still has split fixation in her left eye but with moderately worse vision. Her right eye is now developing progressive rim loss, visual field defects that are encroaching on fixation, and repeated disc hemorrhages.

"It may only be possible to confirm the adequate control of IOP through long-term follow-up and assessment of disease progression."

DSM: It appears that the trabeculectomy in the patient's left eye has largely stabilized her disease. Although her visual acuity has decreased, glaucoma has not progressed. I would not recommend peeling the epiretinal membrane if she has no metamorphopsia and her vision loss seems to be due to glaucoma. The procedure would pose a significant risk of increasing her IOP from surgical manipulation and inflammation. Given the evidence of glaucomatous progression in her right eye, there is a strong case for moving ahead with filtration surgery.

RNW: Several glaucoma specialists had seen this patient previously for a second opinion, but they did not have the benefit of the information available here from long-term follow-up. Their opinions at that time were not to proceed with trabeculectomy. This situation shows well how conclusions about management are highly dependent on the available data.

CONCLUSION

Practitioners can make a definitive diagnosis of glaucoma when there is demonstrable progression of the disease. Such evidence can take the form of an excavated optic nerve head, the appearance or widening of RNFL defects, or the progression of visual field defects.

It may only be possible to confirm the adequate control of IOP through long-term follow-up and assessment of disease progression. Glaucoma specialists are often placed in the difficult position of providing opinions on diagnosis or management with little or no previous information about the course of the disease. This situation can lead to diagnoses and treatment plans that differ from those they would make if a complete history were available. □

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