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Evaluation of Coexisting Optic Nerve Head Drusen and Glaucoma with Optical Coherence Tomography

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Abstract

Objective—Optic nerve head drusen often make evaluation of the nerve head difficult to interpret. In addition, visual field defects are known to occur in patients with optic disk drusen, resembling glaucomatous damage. The authors report two cases of coincident optic nerve head drusen and glaucoma, in which the use of optical coherence tomography (OCT) in evaluating the nerve fiber layer was beneficial.

Participants—Two patients with both optic nerve head drusen and glaucoma, one with primary open angle glaucoma, the other with pseudoexfoliation glaucoma were evaluated. Both patients had asymmetric optic disk drusen, with clinically visible drusen only in one eye.

Intervention—Ophthalmologic examination, color and red-free photography, automated Humphrey visual field testing and OCT were performed.

Results—Nerve fiber layer loss as measured by OCT was found to be greater than expected by the appearance of the optic nerve head and red-free photography, with visual fields consistent with findings in case 1. In case 2, visual fields were full, despite nerve fiber layer thinning seen by OCT and red-free photography.

Conclusions—There can be significant nerve fiber layer thinning in patients with both glaucoma and optic disk drusen, despite the appearance of the optic nerve head in these patients. The cup margin may be obscured by the drusen, giving rise to a falsely full-appearing disk. In such cases, OCT may provide a useful means to quantitatively measure the nerve fiber layer thickness and to aid in the management of these patients by detecting nerve fiber layer thinning earlier than would otherwise be possible.

Optic nerve head drusen (ONHD) are laminated, often calcified, hyaline bodies located in the prelaminar portion of the nerve head. Clinically, they appear as globular bodies that protrude from the disk and blur the disk margin. Small drusen embedded in the nerve may present only as a subtle elevation of the disk, giving rise to a pseudopapilledema appearance. They may obscure the physiologic cup, making it difficult to interpret optic disk cupping. Even in optic nerve heads not complicated by other factors, the variability in interpretation is significant.¹ We present two cases in which both glaucoma and optic nerve head drusen were observed. Through the use of optical coherence tomography (OCT), a new imaging technique that allows high-resolution (approximately 10 microns) cross-sectional imaging of the eye using infrared light, we were able to evaluate the nerve fiber layer (NFL) in these patients with uninterpretable optic nerve heads.

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Each author states that he or she has no proprietary interest in the development of this or competing instruments.

Optical Coherence Tomography Evaluation

After informed consent was obtained, OCT was performed. Optical coherence tomography technology, as well as image and data processing, have been described previously.²⁻⁷ Before OCT evaluation, both eyes of each of the two patients were dilated with 1 % tropicamide and 2.5% phenylephrine hydrochloride. Measurements were performed using a fiber-optically integrated Michelson interferometer with a short-coherence length superluminescent diode light source. A fiber-optic probe module from the OCT unit was coupled to a slit-lamp biomicroscope for in vivo tomography of the posterior segment of the eye. The beam was directed into the eye using computer-controlled galvanometric scanners that could scan arbitrary patterns. A 78-D Volk lens (Volk Optical, Mentor, OH) provided indirect imaging, with beam focus coincident with the slit-lamp image plane to permit simultaneous scanning and visualization of the eye via a charge-coupled device camera. A computer monitor provided a real-time display of the tomograph. Retinal thickness was quantified by computer for each scan in the image as the distance between the first reflection at the vitreoretinal interface and the anterior boundary of the red, reflective layer corresponding to the retinal pigment epithelium and choriocapillaris. Nerve fiber layer thickness was determined by computer. It was assumed to be correlated with the extent of the red, highly reflective layer at the vitreoretinal interface. Boundaries were located by an automated computer algorithm searching for the first points on each A-scan where the reflectivities exceeded a certain threshold. The inner limiting membrane was located by starting anteriorly and searching downward in the image. The posterior margin of the NFL was located by starting within the photoreceptor layer and searching upward in the image. Thresholds were determined separately by the computer for each scan in the image as two-thirds the maximum reflectivity in each smoothed A-scan evaluated on a logarithmic scale. The thickness of this layer corresponded to the thickness of the NFL measured histologically in previous studies.³

Linear interpolation was performed to remove gaps in the boundaries resulting from shadowing due to blood vessels. The boundaries chosen by the computer were overlaid on a false-color display of each image. Thus, the NFL boundary may not always appear to follow the reflectance in the scans.

Circular scans around the optic nerve were performed. A circle size of 3.38 mm was used, so as not to overlap the disk, giving approximately 750 microns of offset from the edge of the disk.

The circular scan around the optic nerve was performed with the subject fixating on an internal fixation light, shone in the eye being scanned, that could be offset from the scan. The offset information in the transverse direction between the fixation light and the scan pattern was recorded for automatic registration of subsequent scans. Near-infrared illumination (840 nm) was used in these scans.

Case Reports

Case 1

An 84-year-old white woman was initially referred to the glaucoma service at the New England Eye Center for evaluation of poorly controlled primary open-angle glaucoma in both eyes treated with betaxolol 0.5% in the right eye twice daily. Medical history was significant for diet-controlled diabetes, hypertension, cardiac arrhythmia, and depression. Her medications consisted of furosemide, digoxin, nicardipine, and sertraline hydrochloride. Her ocular history was notable for a retinal detachment repair in the right eye and a cataract extraction in the left eye 5 years previously.

In April 1995, visual acuity was light perception in the right eye and 20/70 in the left eye without correction. Intraocular pressures were 31 mmHg in the right eye and 38 mmHg in the left eye. Corneal endothelial pigment was seen in both eyes. By Koeppel gonioscopy, angles were open to ciliary body band in both eyes. Posterior chamber intraocular lens implants were in place in both eyes.

Fundus examination revealed a pale, atrophic optic disk in the right eye with attenuated vessels (Fig 1 A). The optic disk in the left eye was notable for multiple drusen, particularly in the superior portion of the optic nerve, with no physiologic cup discernible. An associated NFL hemorrhage was seen in the inferior arcade (Fig 1B). Visual field of the left eye showed a dense inferior arcuate scotoma (Fig 2). Visual field of the right eye could not be obtained given light perception vision. Nerve fiber layer photography performed 1 week later showed diffuse, generalized NFL loss in both eyes (Fig 3).

The patient began receiving apraclonidine 0.5% three times a day in both eyes and changed from betaxolol to levobunolol 0.5% twice daily in both eyes. She returned 3 weeks later with a decreased intraocular pressure of 16 mmHg in the right eye and 23 mmHg in the left eye. Thickness measured by OCT at this time showed markedly thinned NFL in both eyes compared to normal NFL measurements (Figs 4 and 5). Argon laser trabeculoplasty was performed 180° in both eyes with resultant intraocular pressure 6 months later of 14 mmHg in the right eye and 13 mmHg in the left eye.

Case 2

A 70-year-old woman was referred to the glaucoma service with a history of glaucoma for the previous 8 years. She recently developed an allergy to her timolol eye drops. In addition, she had a history of intolerance to pilocarpine due to visual side effects. Argon laser trabeculoplasty 360° had been performed on her left eye. She was taking no systemic medications other than multivitamins. She had an allergy to sulfa medications.

On examination, visual acuity was 20/20 in each eye. Intraocular pressures were 26 mmHg in the right eye and 34 mmHg in the left eye. Humphrey visual fields performed at another office 3 months before the initial examination revealed full visual fields bilaterally. Slit-lamp examination showed bilateral pseudoexfoliation material on the anterior lens capsules. The angles were open 360 degrees to ciliary body band bilaterally by Koeppel gonioscopy. Fundus examination showed a cup/disk ratio of 0.50 in the right eye with an intact neuroretinal rim, and ONHD in the left eye with peripapillary atrophy and no visible cupping.

The patient was diagnosed with pseudoexfoliation glaucoma. She was started on carteolol 1% in both eyes twice daily. At her 2-week follow-up visit, she was tolerating the carteolol well, with intraocular pressures of 20 mmHg in the right eye and 26 mmHg in the left eye. She was advised to continue the medical regimen and scheduled for follow-up in 3 months. At that time, intraocular pressures were 25 mmHg in the right eye and 30 mmHg in the left eye. Optic nerve heads appeared unchanged, with drusen abundant in the inferior quadrant in the left eye (Fig 6). Although Humphrey visual fields were full in both eyes, red-free NFL photography demonstrated diffuse NFL thinning in the left eye (Fig 7). No focal thinning was identified with red-free photography in the left eye. Optical coherence tomography measurements, however, revealed a much thinner inferior NFL in the left eye (86 microns) compared to the inferior NFL in the right eye (152 microns), in addition to the generalized thinning in the left eye (Fig 8). Dorzolamide 2% three times daily in both eyes was added with subsequent decrease in intraocular measurements of 17 mmHg in both eyes.

Discussion

Patients with ONHD pose a challenging diagnostic dilemma and are often difficult to manage because of uninterpretable optic nerve head appearance. Patients may have visual field defects secondary to the presence of ONHD,^{8–10} and may be diagnosed with glaucoma. Conversely, other patients are not diagnosed with glaucoma because of the alteration in the optic nerve head appearance by ONHD, with diminution of the optic nerve cup. The extent of NFL thinning may not be realized in these cases. With OCT, it is possible to measure the thickness of the NFL directly. Optical coherence tomography allows in vivo visualization of cross-sectional images of the retina and NFL at histologic resolution. Schuman et al have shown that NFL thickness by OCT measurements correlate highly with visual function as measured by visual fields.²

Nerve fiber layer thinning has been shown to be the most sensitive indicator of glaucomatous damage, preceding both measurable visual function loss and detectable changes in optic nerve appearance.^{11–13} In our two patients with coincident ONHD and glaucoma, in which the optic nerve heads appeared full, we found OCT to be a useful tool in evaluating generalized NFL thinning, as well as specific, localized areas of NFL thinning. We were unable to obtain the same information with the traditional methods of glaucoma evaluation.

Case 1 demonstrated marked asymmetry of optic nerve appearance due to ONHD, in which the right optic nerve appeared to have advanced cupping from glaucomatous damage, although the left optic nerve appeared undamaged and full. By OCT measurements, significant NFL thinning was evident in the left eye, with measurable NFL remaining only in the inferior quadrant at 45 microns and the temporal quadrant at 48 microns. This was consistent with the superonasal sparing seen on visual field testing. However, this partial sparing of NFL was not evident by red-free photography, which revealed diffuse, generalized NFL loss, with no focal areas of NFL thinning seen.

In case 2, OCT measurements showed generalized NFL thinning in the left eye. In addition, the inferior quadrant in the left eye at 86 microns was much thinner than the corresponding inferior quadrant in the right eye at 152 microns. This was the quadrant in which the drusen in the left eye were most abundant clinically. Thus, although the optic nerve of the left eye appeared to have less cupping, the NFL by OCT was thinner in comparison to the right eye, particularly in the region in which the drusen were located. The presence of a small optic nerve head cup did not correspond necessarily with less NFL loss. At this level of NFL thinning, visual fields remained full, although a clear difference was seen in the NFL thickness by OCT and red-free photography between the two eyes.

Our conventional means of monitoring glaucoma using visual field testing and red-free photography were not as sensitive in the evaluation of our two patients with ONHD. Nerve fiber layer interpretation by red-free photography has been known to be difficult not only because of factors such as hypopigmented fundi, but also because of the lack of clarity in the photographs.¹⁴ These factors are not an issue with OCT evaluation of the NFL, and interpretation is not variable among different observers because OCT quantifies NFL thickness.

Optical coherence tomography provided a more accurate assessment of NFL than either visual field testing or red-free photography. Furthermore, we were able to quantify NFL thickness objectively, allowing early detection of NFL thinning and identification of specific areas of thinning. This allowed us to recognize eyes requiring closer attention and lower intraocular pressures than we may have assumed based on our traditional assessment with optic nerve appearance and visual field testing.

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Figure 1.

Case 1. **A**, Optic nerve photograph shows severely cupped optic nerve head in the right eye with no drusen evident. **B**, ONHD are most concentrated in the superior and nasal regions in the left eye (arrows). The optic nerve appears full with no clinical evidence of glaucomatous cupping. Associated retinal heme is seen inferotemporally in the left eye.

Figure 2.

Case 1. Dense inferior arcuate scotoma demonstrated on automated visual field testing of left eye. The scotoma corresponds to the ONHD location in the superonasal quadrant of the optic nerve.

Figure 3.

Case 1. **A**, Red-free photography reveals atrophic optic nerve and diffuse NFL loss in the right eye. **B**, Red-free photography shows diffuse NFL loss. Note retinal hemorrhage has diminished. Arrows indicate areas of drusen.

Figure 4.

Example of normal NFL as measured by OCT. The NFL tomograph is represented by the most superficial red reflectance layer. The inferior NFL is seen between the arrows. The average measurement of each of the quadrants is seen in the center circular scan, below the tomograph. Arrowhead points to the numerical value (in microns) of the inferior quadrant. The numerical NFL measurements of each clock hour are seen on the right circular scan.

Figure 5.

Case 1. **A**, The right eye has virtually no measurable NFL present by OCT measurements, as expected by the appearance of the optic nerve in the right eye. The temporal and inferior quadrants show some measurable NFL (arrowheads). Note the thinned superficial reflectance layer. The arrows point to the NFL at its thickest portion on the tomograph. **B**, In the left eye, the NFL is thinnest in the superonasal area in which the drusen are most abundant. Arrows point to region in which the NFL is spared — the inferotemporal region. The corresponding numerical value is seen by the arrow-heads inferiorly (45 microns) and temporally (47 microns). Note the extremely thinned NFL in the left eye despite the full appearance of the optic nerve seen in Figure 1B.

Figure 6.

Case 2. **A**, Optic nerve photograph in the right eye shows cup/disk ratio of 0.5 with an intact neuroretinal rim. **B**, Optic nerve photograph in the left eye reveals drusen inferiorly (arrows). Note the full appearance of the optic disk, particularly inferiorly in the region of the drusen. No cupping is evident in the left eye.

Figure 7.

Case 2. **A**, The NFL in the right eye on red-free photography appears normal, with intact NFL striations. **B**, Red-free photography in the left eye shows diffuse NFL loss, with no focal defects seen.

Figure 8.

Case 2. **A**, The NFL in the right eye by OCT measurements in the right eye is thick inferiorly (arrows). Arrowhead points to numerical value of inferior NFL thickness (152 microns). **B**, Despite the full appearance of the optic nerve in the left eye, the NFL is thinned in the left eye compared to the right eye, particularly in the inferior quadrant (arrows). Arrowhead points to numerical value of average NFL thickness in the inferior quadrant (86 microns).