Sudden Visual Field Constriction Associated with Optic Disc Drusen

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We report two patients with optic disc drusen who suffered sudden, concentric constriction of the visual field. Visual acuity remained normal. The involved discs showed no swelling, hemorrhage, or other evidence of anterior ischemic optic neuropathy. We are unable to explain the mechanism or the pattern of visual field loss in these unusual cases.

Key Words: Optic disc drusen—Visual field constriction—Anterior ischemic optic neuropathy.

Although most patients with optic disc drusen are asymptomatic, defects in the field of vision can usually be evinced upon careful testing. In his classic monograph, Lorentzen (1) reported visual field changes in 87% of patients tested with the Goldmann perimeter. In a later study, Savino, Glaser, and Rosenberg (2) detected visual field loss in 71% of patients with visible drusen. The most common findings were nerve fiber bundle defects, generalized field constriction, and enlargement of the blind spot.

Visual field defects develop insidiously in the majority of patients with optic disc drusen. However, visual field loss may occur abruptly. Most authors have invoked anterior ischemic optic neuropathy to explain the phenomenon of sudden visual loss in patients with optic disc drusen. We describe two patients with optic disc drusen who experienced severe and sudden constriction of the visual field with preservation of normal visual acuity. The diagnosis of anterior ischemic optic neuropathy was tenable in neither patient.

CASE REPORTS

Case 1

An 18-year-old woman was noted on routine examination to have drusen of the left optic disc. Over the following decade, the drusen gradually became more prominent. At age 29, drusen also became evident in the right optic disc. Although the patient was asymptomatic, at age 31 she was tested with a Cooper Vision Auto-Perimeter 120 point screening program. The visual fields were normal, except for 12 points in the left eye missed along an inferior arcuate course. At age 35 the visual acuity was 20/20 OU, the visual fields were unchanged, and the patient remained without symptoms. Fundus photos were taken for the pa-
tient to share with future ophthalmologists who might be worried by the appearance of the optic discs (Fig. 1).

Less than two years later, at age 37, the patient reported that the vision in her left eye suddenly became blurry and dim upon arising from an office chair. On examination 2 days later, the visual acuity was 20/20 in both eyes. The Hardy-Rand-Rittler color plates were all identified correctly with each eye. A left afferent pupil defect was present. The visual fields were tested with a 30-2 threshold program using a Humphrey field analyzer. The visual field of the right eye was normal. The visual field of the left eye showed severe, concentric constriction with preservation of normal central retinal sensitivity (Fig. 2). The foveal threshold was 36 dB in each eye. The retina of the left eye was normal. The optic disc contained innumerable exposed and buried drusen, but no swelling or hemorrhage was evident (Fig. 3). Allowing for variation in photographic technique, we could see no difference in the appearance of the optic disc compared with the photograph taken 2 years earlier, which the patient carried in her purse (Fig. 1). A fluorescein angiogram showed normal perfusion of the left optic disc and retina (Fig. 4). The only abnormal feature was slow leakage from the left optic disc, which resulted in marked hyperfluorescence in late stages of the angiogram (Fig. 5). The patient received a brief course of prednisone, without benefit.

On examination a year later, the visual acuity was 20/20 in both eyes and the Hardy-Rand-Rittler plates were again identified correctly with each eye. A left afferent pupil defect was still present. Visual field testing of the left eye showed a foveal sensitivity of 38 dB and slight improvement of the visual field. The left optic disc appeared identical (Fig. 6) to the photograph taken the year before. The nerve fiber layer, except for the papillomacular bundle, appeared severely atrophic. A fluorescein angiogram showed late staining of drusen within the optic disc (Fig. 7), but the striking hyperfluorescence noted previously (Fig. 5) was absent.

![Fig. 1. Fundus photograph of the left eye in Case 1 taken on Feb. 19, 1989, 19 months prior to abrupt onset of visual field constriction. The disc appears choked with drusen.](image)

![Fig. 2. Visual field of the left eye in Case 1 tested using the 30-2 threshold program of the Humphrey field analyzer. The field shows severe, concentric constriction, but the macular thresholds are normal.](image)
Case 2

A 37-year-old woman was reported to have drusen in the right optic disc. At age 40, an ophthalmologist noted the presence of drusen in both optic discs. The visual acuity was 20/20 OU and the visual fields were normal when tested with a 3-mm white pin at a distance of 1 m from a tangent screen. The patient remained entirely free of symptoms until a year later, when she noted the sudden appearance of a "film" over the right eye. On examination the visual acuity was 20/20 in both eyes. An afferent pupil defect was present in the right eye. Upon testing at the tangent screen the visual field of the left eye was normal, but the visual field of the right eye showed concentric constriction that spared only the central 5-10°. Both optic discs contained drusen. The right optic disc appeared pale, but no hemorrhage or edema was present. A sedimentation rate, VDRL, carotid angiogram, and pneumoencephalogram were normal. The patient received prednisone, but the visual fields did not improve.

On subsequent examinations the patient reported no further change in the vision in the right eye. When retested 16 years later, at age 56, the
vision was still 20/20 in both eyes. The visual field of the right eye showed severe concentric constriction, sparing only 5° toward the temporal side of fixation (Fig. 8). The foveal sensitivity was 33 dB. The right optic disc appeared pale and filled with drusen. The nerve fiber layer was absent, except for the papillomacular bundle.

DISCUSSION

Previous investigators have noted a poor correlation between the location of drusen in the optic disc and the pattern of scotomata in the visual field (1-4). This lack of correspondence argues against mechanical compression or erosion of nerve fibers as a simple explanation for the visual field defects associated with optic disc drusen. It is conceivable, though, that buried drusen unseen with the ophthalmoscope might correlate better with the pattern of visual field loss. Spencer (5) has proposed that a defect in axoplasmic transport at the optic disc is the cause of drusen formation. According to his view, impaired axoplasmic transport leads to axonal death, calcium deposition, and the appearance of drusen within the optic disc. Tso (6) favors a similar mechanism, although he believes a dis-

FIG. 5. Later stage (388.9 seconds) of fluorescein angiogram in Case 1 showing hyperfluorescence of the left optic disc.

FIG. 6. Follow-up fundus photograph taken Aug. 27, 1991 in Case 1 showing no change in the appearance of the left optic disc, except for loss of the nerve fiber layer outside the papillomacular bundle, which cannot be appreciated on these photographs.
order of axonal metabolism, rather than axonal transport, is responsible for the accumulation of disc drusen. This debate—whether drusen are the cause, or merely a conspicuous by-product, of axonal degeneration—underscores the fact that the mechanism of visual loss in patients with optic disc drusen is still a mystery.

Infrequently, vision loss occurs suddenly in patients with optic disc drusen. The mechanism is unknown. Anterior ischemic optic neuropathy has been offered as an explanation (7–11). Sudden vision loss from disc drusen is usually painless, non-progressive, and may be accompanied by disc edema and hemorrhage. Fluorescein angiography has documented hypoperfusion of the optic disc (8). These features are all characteristic of anterior ischemic optic neuropathy. Moreover, optic discs that contain drusen are usually small, crowded, and lack a physiological cup. Absence of a physiological cup is linked to an increased risk of anterior ischemic optic neuropathy (12–14).

Although anterior ischemic optic neuropathy may provide a satisfactory explanation for the occurrence of sudden vision loss in some patients with optic disc drusen, this diagnosis is difficult to reconcile with the findings in our patients. When examined soon after acute loss of visual field, neither patient had hemorrhage or edema of the optic disc. The diagnosis of anterior ischemic optic neuropathy requires ophthalmoscopic signs of optic disc ischemia. Fortuitously, disc photos were taken in the first patient 2 years before vision loss (Fig. 1). Using these photos for comparison, we could detect no change in the appearance of the optic disc after acute vision loss (Fig. 3). Moreover, 1 year later (Fig. 6), we observed no attenuation of the retinal arteriolar circulation. This sign typically appears as a late sequela of anterior ischemic optic neuropathy (15).

In our first patient, the fluorescein angiogram failed to show hypoperfusion of the optic disc. In a fluorescein study, Karel and coworkers (16) found no filling defects in the disc circulation in patients with sudden visual loss and disc drusen. These
findings neither support nor refute the diagnosis of anterior ischemic optic neuropathy, as much as Kommerell and coworkers (17) have obtained normal fluorescein angiograms of the optic disc circulation in patients with anterior ischemic optic neuropathy.

In our patients, the most striking feature of the drusen-associated visual loss was the sudden and severe loss of peripheral vision with preservation of normal visual acuity. This pattern of concentric visual field constriction is not consistent with anterior ischemic optic neuropathy. Indeed, it is difficult to explain in anatomical terms how hypoperfusion of the posterior ciliary arterial supply could produce such a pattern of visual field loss.

Occasionally, patients report "acute" visual field loss, but in fact have only acutely become aware of long-standing, insidious visual field loss. We doubt this occurred in our patients. Both patients were under regular ophthalmological surveillance, and their visual fields were documented by screening tests prior to their episodes of abrupt visual loss.

In many optic neuropathies, such as those caused by toxins, nutritional deficiencies, or mitochondrial DNA mutations, the small fibers of the papillomacular bundle are selectively destroyed. The converse appears to be true in drusen-associated optic neuropathy: central visual acuity remains normal even when visual field loss is severe (18). This observation is frequently referred to as "Rucker's rule." Like most dictums in medicine, it is fallible. Central acuity loss from drusen has been well documented in several reports (9,19,20). These noteworthy exceptions aside, central vision tends to be preserved in patients with optic disc drusen. This finding implies that the small-caliber axons from ganglion cells in the macula enjoy some degree of immunity from the optic neuropathy associated with optic disc drusen. Understanding why these fibers are relatively spared may provide a key to discovering the cause of visual field loss in patients with optic disc drusen. The findings described in this paper indicate that there may be a propensity to spare the papillomacular bundle even when visual field loss is sudden. We caution against the assumption that sudden visual loss in patients with disc drusen represents a manifestation of anterior ischemic optic neuropathy.

REFERENCES