Optic Disc Cupping in Arteritic Anterior Ischemic Optic Neuropathy Resembles Glaucomatous Cupping

JERRY SEBAG, MD,* JOHN V. THOMAS, MD,† DAVID L. EPSTEIN, MD,† W. MORTON GRANT, MD†

Abstract: Five cases of anterior ischemic optic neuropathy secondary to biopsy-proven giant cell arteritis are presented. In each case, cupping of the optic disc, which closely resembled glaucomatous cupping, was observed in the affected eye. The presence of glaucoma was ruled out on the basis of normal intraocular pressures and normal tonographic measurements of outflow facility. These cases indicate that arteritic ischemic optic neuropathy may result in optic disc cupping, which closely resembles glaucomatous cupping. The similarities in the appearance of cupping of these discs with that seen in eyes with glaucoma suggest that the pathogenesis of cupping in glaucoma and in arteritic ischemic optic neuropathy may share some common mechanisms. [Key words: anterior ischemic optic neuropathy, cupping, giant cell arteritis, glaucoma, optic disc, temporal arteritis.] Ophthalmology 93:357-361, 1986

The pathogenesis of optic disc cupping in glaucoma is the subject of ongoing controversy.1,2 Proponents of a vascular mechanism cite the occurrence of cupping in patients with ischemic optic neuropathy as evidence in support of an ischemic etiology.2,3,4 In particular, it has been stated that patients with anterior ischemic optic neuropathy (AIN) due to giant cell arteritis, develop optic disc cupping.6 Hayreh has found that cupping developed in 11 out of 13 eyes with arteritic AION and did not occur in seven eyes with AION of nonarteritic etiology.5 It is not clear, however, that glaucoma was adequately ruled out in these patients.

Other studies have not found a relationship between AION and the development of cupping resembling that seen in eyes with glaucoma.7,8 Quigley and Anderson observed glaucomatous cupping in five of ten cases with AION due to giant cell arteritis.9 However, two of these five patients were found to have chronically elevated intraocular pressures and the remaining three had large physiologic cups in the contralateral eye. They concluded that the development of cupping resembling that seen in glaucoma was unusual after the occurrence of AION.

In this report, we present five patients in whom cupping of the optic disc clinically indistinguishable from glaucomatous cupping developed following well-documented episodes of acute AION secondary to biopsy-proven giant cell arteritis. In all cases, several intraocular pressure measurements, tonography studies of outflow facility, and follow-up evaluations failed to demonstrate the presence of glaucoma.

CASE REPORTS

Case 1. A 78-year-old white woman presented in July 1977 with a three-day history of sudden painless loss of vision in the right eye and tenderness in the right temporal area of the forehead of several months' duration. There was no history of diabetes, hypertension or massive hemorrhage, and no family history of glaucoma. Her visual acuity was hand motions temporally in the right eye and 20/25-2 in the left. On external examination, a right afferent pupillary defect was noted. Slit-lamp examination

From the Eye Research Institute of Retina Foundation,* Glaucoma Consultation Service † Department of Ophthalmology, Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston, Massachusetts.

Reprint requests to John V. Thomas, MD, 100 Charles River Plaza, Boston, MA 02114.
was unremarkable in the right eye and normal in the left.

The patient's visual acuity was 20/20 in the right eye and 1.0 in the left eye. The optic discs were normal in all four eyes. The patient's visual fields were normal in all four eyes. The patient was free of any systemic disease.

When examined by indirect ophthalmoscopy, the optic discs were normal in all four eyes. The disc margins were sharp and the discs were of normal size.

Case 2: Patient 2 was a 50-year-old male with a six-year history of diabetes mellitus. He also had hypertension.

His visual acuity was 20/30 in the right eye and 20/20 in the left eye. The patient's visual fields were normal in both eyes. The optic discs were normal in both eyes. The patient was free of any systemic disease.

When examined by indirect ophthalmoscopy, the optic discs were normal in both eyes. The disc margins were sharp and the discs were of normal size.

Figs 1-9. Legend, opposite page.
was unremarkable. Applanation tensions were 14 mmHg in the right eye and 16 mmHg in the left.

The patient was examined by Shirley Wray, MD, neuro-ophthalmology consultant at the Massachusetts Eye and Ear Infirmary, who described the right disc as being “pale and swollen” with uniform swelling and elevation of two dipters and attenuated arterioles. The left disc and both macular areas were normal. No carotid bruises were noted on auscultation and skull films were normal. The sedimentation rate was 78 mm and a right temporal artery biopsy was positive for giant cell arteritis. The patient was treated with oral prednisone with no improvement in visual acuity.

When examined in March 1979, the visual acuity was no light perception in the right eye and 20/40 in the left. The applanation tensions were 18 mmHg in both eyes. Tonography values were: $C_{40} = 0.26$, $P_{40}/C_{40} = 65$ in the right eye, and $C_{40} = 0.25$, $P_{40}/C_{40} = 60$ in the left. External and slit lamp examination of both eyes was unchanged from July 1977. The right disc showed a markedly enlarged cup with extension to the rim inferiorly and inferotemporally (Fig 1). Although not shown clearly in Figure 1, clinical examination revealed that a thin, pale neural rim was present elsewhere. The right macula was normal. The left disc was normal with a physiologic cup (Fig 2) and the left macula was normal. The authors examined the patient and agreed that without the benefit of history the right optic disc would be considered glaucomatously cupped.

Case 2. An 80-year-old white man presented in April 1973 with a six-day history of sudden onset of hazy vision in both eyes, the left eye being more severely affected than the right eye. He also complained of pain in the right eye.

His visual acuity was 20/200 in the right eye and counting fingers at one foot in the left. On external examination, a prominent vessel was noted in the right temporal area of the forehead and a left afferent papillary defect was noted. Slit-lamp examination showed minor anterior corneal and nuclear opacities (left greater than right). Applanation tensions were: 13 mmHg in the right eye and 14 mmHg in the left. The appearance of the discs were described as “papilledema 1+” in the right eye and “papilledema 2+ and 3+ with tiny hemorrhages” in the left. Both macular areas were normal. Tangent screen visual field examination in the right eye revealed a large inferior Bjerrum scotoma coming very close to fixation and a superonasal defect to a 9 mm white test object at 1 m. The visual field in the left eye revealed a small superotemporal island of vision where hand motions could be observed. Skull films were normal. The sedimentation rate was 55 mm per hour and a left temporal artery biopsy was positive for giant cell arteritis. The patient was treated with large doses of oral prednisone, with resulting improvement of vision in the right eye.

Six months later visual acuity was 20/40 in the right eye and counting fingers at two feet temporally in the left. There was an afferent papillary defect in the left eye. Applanation tensions were 13 mmHg in the right eye and 14 mmHg in the left. Both discs had temporal pallor, much more marked in the left eye. No cupping changes were noted.

When examined in March 1979 his visual acuity was 20/50 in the right eye and counting fingers at 1½ feet in the left. Applanation tensions were 11 mmHg in the right eye and 15 mmHg in the left. Tonography values were $C_{40} = 0.24$, $P_{40}/C_{40} = 41$ in the right eye, and $C_{40} = 0.15$, $P_{40}/C_{40} = 73$ in the left. Findings on external and slit-lamp examination were unchanged from April 1973. Examination of the right disc showed that the temporal half of the disc was pale (Fig 3). The nasal half of the disc had a pink color. There was shallow cupping of the disc to the temporal rim. The right macula was normal except for a few RPE defects. Examination of the left disc showed backward bowing of the lamina cribrosa and a cup which extended to the temporal rim (Fig 4). A few drusen were present in the left macula. Slight peripapillary chorioretinal atrophy was noted along the temporal margins of both discs. The authors examined the patient and agreed that without the benefit of history, the left optic disc would be considered glaucomatously cupped.

Case 3. An 85-year-old white woman presented in August 1976 with a two-day history of blurred vision and a one-day history of no light perception in the left eye.

Her visual acuity was 20/40 in the right eye and no light perception in the left. No carotid bruises were heard on auscultation and a left afferent papillary defect was noted. Slit-lamp examination revealed bilateral nuclear sclerosis of her lenses. Tensions with a Schiotz tonometer were 17 mmHg in the right eye, and 17 mmHg in the left. The right disc was normal. The left disc was described as “pale and elevated with scattered splinter hemorrhages at the disc margin.” Both maculae were normal.

Visual field examination revealed a normal right visual field. The left visual field was unobtainable. The sedimentation rate was 100 mm per hour and a left temporal artery biopsy was positive for giant cell arteritis. The patient was treated with oral prednisone, but no improvement in vision resulted.

When examined in March 1979, her visual acuity was 20/60 in the right eye and no light perception in the left. Applanation tensions were 14 mmHg in both eyes. Tonography values were $C_{40} = 0.21$, $P_{40}/C_{40} = 67$ in the right eye, and $C_{40} = 0.18$, $P_{40}/C_{40} = 83$ in the left. External and slit-lamp examination findings were unchanged from August 1976. Examination of the right disc revealed normal color and a tiny physiologic cup (Fig 5). The right macula was normal. Examination of the left disc revealed definite sauceration of the cup to the rim inferotemporally and temporally (Fig 6). Although not clearly shown in Figure 6, clinical examination revealed slight pallor in the inferotemporal area of the disc. The left macula was normal. The authors examined the patient and agreed that without the benefit of history, the left optic disc would be considered glaucomatously cupped.

Case 4. An 82-year-old white man presented in September 1977 with a history of sudden visual loss in both eyes (right greater than left) that had occurred two weeks previously. His visual acuity was counting fingers at three feet in the right eye and at ten feet in the left. On external examination, a right afferent papillary defect was noted. Slit-lamp examination revealed bilateral nuclear selerotic lenses. Tensions with a Schiotz tonometer were 9 mmHg in both eyes. The right disc was described as being “raised with blurring of the margins 360.” A small
disc margin hemorrhage was noted at 5 o'clock. The temporal side of the left disc was described as "raised with indistinct borders". The maculae were normal in both eyes.

Visual field examination of the right eye revealed that a meaningful field could not be plotted with the largest test object on the Goldmann perimeter. Visual field examination of the left eye showed generalized constriction with an inferonasal defect. No carotid bruits could be auscultated. The sedimentation rate was 38 mm per hour and a right temporal artery biopsy was positive for giant cell arteritis. The patient was treated with large doses of oral prednisone, resulting in improvement of vision in both eyes.

When examined in March 1979, his visual acuity was 20/400 in the right eye and 20/60 in the left. Application tensions were 17 mmHg in both eyes. Tonography values were C55 = 0.16, Po/C55 = 69 in the right eye, and C55 = 0.20, Po/C55 = 69 in the left. Findings on external and slit-lamp examination were unchanged from September 1977. Examination of the right disc revealed a shallow depression of the cup extending to the supertemporal, temporal and inferotemporal rim. The lamina cribrosa was clearly visible and nasalization of vessels was present. Examination of the left disc revealed that it was normal with a physiologic cup. Both maculae were normal. The authors examined the patient and agreed that without the benefit of history, the right optic disc would be considered glaucomatously cupped.

Case 5. A 79-year-old white man presented in May 1974 with a seven-day history of intermittent superior altitudinal amaurosis in the left eye. His visual acuity was 20/40 in the right eye and hand motions at two feet in the left. On external examination a left afferent pupillary defect was noted. Slit-lamp examination showed early bilateral immature cataracts. Tensions with a Schiotz tonometer were 17 mmHg in both eyes.

The right disc was described as "pinpoint, flat, and healthy appearing". The left disc was described as "pale, swollen, with superficial hemorrhages, narrowed arterioles and slight venous engorgement" (Fig 7). Both macular areas were normal. Tangent screen visual field examination revealed a full field in the right eye and small temporal island of light perception in the left eye. There were no carotid bruits and skull x-rays were normal. The sedimentation rate was 60 mm per hour and a left temporal artery biopsy was positive for giant cell arteritis. The patient was started on large doses of oral prednisone. No improvement of vision occurred in the left eye.

When examined in November 1978 his visual acuity was 20/40 in the right eye and light perception temporally in the left. Application tensions were 14 mmHg in both eyes. Tonography values were C55 = 0.36, Po/C55 = 36 in the right eye, and C55 = 0.34, Po/C55 = 35 in the left. Findings on external and slit-lamp examination were unchanged from May 1974. Examination of the right disc revealed that it was normal with a physiologic cup (Fig 8). Examination of the left disc revealed a markedly enlarged, shallow cup extending to the rim (Fig 9). The authors examined the patient and agreed that without the benefit of history, the left optic disc would be considered glaucomatously cupped.

**DISCUSSION**

Glaucomatous damage to the optic disc is said to be characterized by one or more of the following: total cupping of the disc, a cup that extends upward or downward to touch the margin of the disc, or a cup that is vertically oval. Asymmetry of disc cupping, thinning of the neuroretinal rim without complete obliteration of the cup, and backward bowing of the lamina cribrosa are additional features of glaucomatous cupping. Perhaps the most important feature of the disc that needs to be carefully assessed when trying to distinguish ophthalmoscopically between glaucomatous cupping and nonglaucomatous cupping is the location of disc pallor.11 Pallor restricted to the area of the cup is typical of glaucomatous optic atrophy. Pallor that extends beyond the area of the cup and involving nonexcavated, neuroretinal rim is typical of nonglaucomatous optic atrophy. The diagnostic value of this distinction is underscored by the extension of pallor beyond the area of the cup in cases 1 and 3. However, when cupping extends to the rim (cases 2 and 5) this feature can no longer be evaluated.

Some authors have claimed that cupping that simulates glaucoma only occurs when there is a preexisting large physiologic cup.2 Quigley and Anderson found that in three of five cases of arteritic AION with cupping, there was a large physiologic cup in the fellow eye.9 In our series, there were no patients in whom the fellow eye had a large physiologic cup.

In spite of familiarity with the above-mentioned features of glaucomatous cupping, making a diagnosis of glaucoma from the appearance of the optic disc (without the benefit of history or other clinical findings) can be difficult. In a study performed without observer knowledge of clinical data, 44% of eyes with nonglaucomatous optic disc cupping were interpreted to have glaucomatous disc cupping by experienced disc examiners.11 This observed rate of misdiagnosis overestimates the frequency with which these two forms of pathologic cupping are confused ophthalmoscopically, since the "unknowns" in this study were preselected so that they closely simulated glaucomatous disc cupping.

Nevertheless, the fact remains that there are some cases of optic disc cupping which are clinically indistinguishable from optic disc cupping secondary to elevated intraocular pressure. The five cases presented in this report are examples of optic disc cupping secondary to ischemia which closely resemble glaucomatous cupping but in whom glaucoma has been ruled out. Cases such as these make it difficult to support the concept that glaucomatous cupping is solely due to elevated intraocular pressure. Indeed, backward bowing of the lamina cribrosa as a characteristic feature of advanced glaucomatous cupping has been related to this "mechanistic" theory of glaucomatous damage.12,13 Radulescu and MacManus have claimed that this finding is not present in nonglaucomatous cupping.9 However, in our series of patients, cases 2 and 5 exhibited prominent backward bowing of the lamina. This finding was less prominent but nevertheless present in cases 1 and 3.

One case in our series indicates that there may be a relationship between the severity of AION and the development of disc cupping. An acute ischemic event occurred simultaneously in both eyes of case 2 resulting in considerably more disc swelling in the left eye. Following the acute episode, the left eye was more far advanced, as evidenced by marked decrease in visual acuity and extensive visual field loss. Both eyes developed disc pallor (Fig 3). Although initially, there were no cupping changes, the more severely affected eye went on to develop cupping indistinguishable from that observed in glaucoma (Fig 4).
The association of optic disc cupping with AION does not provide direct evidence of an ischemic etiology for glaucomatous optic disc cupping. However, this association and the close resemblance of optic disc cupping resulting from glaucoma and from arteritic AION needs to be accounted for when considering the pathogenesis of true glaucomatous cupping.

REFERENCES


Discussion
by
Jonathan D. Trobe, MD

The authors present five patients with optic disc cupping caused by infarction related to biopsy proven giant cell arteritis. They contend that the excavation visible fundoscopically is indistinguishable from that found in primary open-angle glaucoma.

This point has been made before. As the authors acknowledge, Quigley and Anderson suggested in 1977 that five of ten patients with arteritic ischemic optic neuropathy showed glaucomatous disc excavation. Their findings were muted by the fact that two of five patients had had elevated intraocular pressures, and the other three had large physiologic cups in the uninvolved eyes. Because the five patients presented have no such confounding attributes, the authors have provided valuable evidence that more than one disease produces an etched optic nervehead.

Arteritic ischemic optic neuropathy is not the only disease to produce disc excavation mimicking glaucoma. In 1937, Dalsgaard-Nielsen, and in 1950, Blazar and Scheie used the term pseudoglaucoma for disc cupping in chiasmal tumors, syphilitic optic neuritis, and congenital disc anomalies. They found disc cupping in 6 of 61 eyes with nonarteritic ischemic optic neuropathy. They believed that neuro-retinal rim pallor was the feature that separated these nonglaucomatous cases from the glaucomatous cases.

In 1978, Radius and Maumenee came to slightly different conclusions. In their study of cup-disc ratios from stereo fundus photographs and ophthalmoscopy of patients with nonglaucomatous optic atrophy, they found that "Few if any new heads demonstrated characteristic glaucomatous disc changes including disc asymmetry, vertical elongation, backward bowing of the lamina cribrosa, and notching of the neuro-retinal rim." They stated that those few cases with glaucoma-like cupping were found to have large physiologic excavation in the normal fellow eye.

Their conclusions may have been influenced by knowing the correct diagnosis in advance. In 1980, I asked three experienced fundus gazers to view stereo fundus photographs of 80 patients with optic disc cupping caused either by glaucoma or by other optic nerve diseases. Without knowing the diagnosis, they were instructed to distinguish glaucoma from nonglaucoma and to indicate whether they saw neuro-retinal rim pallor, thinning or obliteration, or laminar dots. At least one observer incorrectly diagnosed glaucoma in nearly half of the 29 eyes of patients with disc excavation caused by either compressive or hereditary optic neuropathy. Neuro-retinal rim pallor proved to be the most helpful differential sign, being 94% specific for nonglaucomatous excavation. Focal or diffuse obliteration of the rim was 87% specific for glaucoma. Thinning of the rim was only 47% specific for glaucoma, and laminar dots was worthless as a differential sign. Had the observers been aware of the differential value of neuro-retinal rim pallor, they would have improved their score greatly; but it would not have been perfect, because there were some cases of complete rim obliteration caused by nonglaucomatous diseases. Complete rim obliteration also occurred in two of the five cases presented by the authors.

What is the significance of this information? First, clinicians must beware not to overdiagnose glaucoma on the basis of cupping alone. Second, neuro-retinal rim pallor together with cupping is a combination rarely seen in glaucoma. We must remember that the optic nervehead has only limited ways of reacting to a variety of insults. Evidently the only truly specific feature of glaucomatous cupping is that the surviving rim tissue stays in the pink.

References