OPTIC NERVE INFARCTION

BY *Paul R. Lichter,* MD

AND

John W. Henderson, MD, PHD

CUPPING AND ATROPHY OF THE OPTIC DISC WITH ASSOCIATED VISUAL FIELD LOSS implies glaucomatous damage. The ophthalmologist's reaction to such a finding is treatment aimed at reducing intraocular pressure. When such damage occurs with normal intraocular pressure, the term "low tension glaucoma" has been applied. In some instances of this type, reduction of the pressure is, nonetheless, advised in hope of preventing further disc damage.[1]

Recent emphasis has been place on optic disc hemorrhages with associated optic nerve cupping and visual field loss.[2-10] Some reports discuss the importance of these hemorrhages in eyes with normal intraocular pressures. Distinction is made between cases where the disc hemorrhage occurs in association with a serious systemic event such as acute blood loss, and where the hemorrhage occurs without other associated factors. In the first instance, resultant disc damage and field loss are not expected to progress, whereas in the latter instance they are.

There remains some ambiguity as to whether or not to treat individuals with a glaucomatous type of optic disc damage and field loss but normal intraocular pressure. The role of disc hemorrhages in such cases is also confusing. The fact that the word "glaucoma" is associated with many of these cases adds to the confusion. We have seen a number of patients with a specific type of optic disc damage and visual field loss which simulates glaucoma but which we believe may have nothing to do with glaucoma. Treatment is unnecessary since the problem is not progressive. This report will describe such findings and present 3 cases as examples.

REPORT OF CASES

CASE 1

A 76-year-old white woman was first examined in 1953 at age 52 for routine refraction. Her vision was 20/20, in both eyes. Refraction: RE +0.50 +1.00

*From the Department of Ophthalmology, University of Michigan Medical School, Ann Arbor, Michigan.

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×06; LE +0.25 +2.25 ×170. External, extraocular muscle, and slit lamp biomicroscopic examinations were all normal. Ophthalmoscopic examination was unremarkable. Intraocular pressure after cycloplegia was 20 mm Hg each eye (Schiøtz). General health was excellent.

The patient had no ocular problems until September, 1968 at age 67. At that time she complained of a "curtain" over the central vision in the left eye of sudden onset. Vision was RE 20/20; LE 20/25. Intraocular pressure was 18 mm Hg each eye (Schiøtz). Ocular examination was normal except for an area of optic disc atrophy at the 5 o'clock position in the left eye. A visual field on the tangent screen showed a dense superior arcuate visual field defect in the left eye. There was no associated hemodynamic crisis or other systemic event. Approximately two months later, however, the patient suffered a myocardial infarction.

In April, 1970, at age 69, a routine follow-up examination disclosed a hemorrhage on the optic disc margin of the right eye at the 7 o'clock position. Her visual field on the right was normal. The left disc and field had not changed. Intraocular pressure was 16 mm Hg each eye (Schiøtz).

In September, 1972, age 71, the right disc began to show an atrophic area on the rim at the 7 o'clock position identical to that in the left eye (Fig. 1). Visual fields now showed identical superior arcuate field defects (Fig. 2).

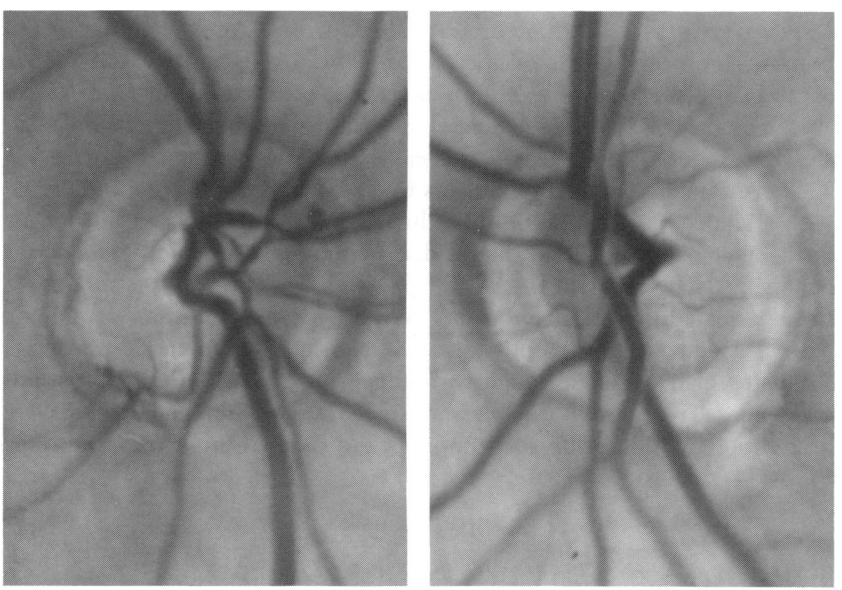

FIGURE 1

Right and left optic disc of Case #1. Both discs show a thin tongue of pallor extending to the inferotemporal disc border. The pattern of small vessels suggests a notch of the disc at this point. A peculiar collection of small vessels is seen associated with this area on the right disc. Such vessels have been seen in other cases of this type and can be a helpful clue as to the problem.

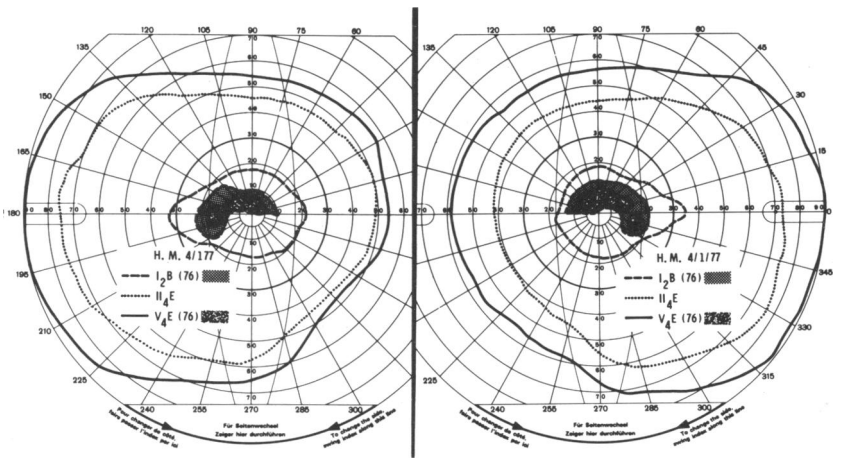

FIGURE 2
Right and left visual fields from Case #1 showing dense superior arcuate field defects close to fixation. Note the symmetry of these defects.

Additional studies included gonioscopy which showed grade 2 open angles and tonography "C" values were RE 0.25; LE 0.24. Applanation tonometry was RE 13; LE 11 mm Hg. Interestingly, a small hemorrhage was noted at the 11 o'clock position on the right disc, and an unusual array of vessels was seen to be associated with the atrophic disc area at the 7 o'clock position on the right disc (Fig. 1).

In May, 1973, the disc hemorrhage at the 11 o'clock position on the right had disappeared. No disc atrophy or visual field defect has occurred associated with this disc hemorrhage.

The latest evaluation was in April, 1977. Vision was 20/20 each eye. Intraocular pressure by applanation was RE 13, LE 15 mm Hg. Repeat tonography showed a facility of outflow of RE 0.36; LE 0.26. Optic discs and visual fields were unchanged. The field defects continue to be quite symmetrical and show a solid central nucleus with relative defects spreading into a more complete arcuate pattern. Blood pressure was 180/110. The patient has angina and takes nitroglycerin. There have been no further myocardial infarctions and no episodes of acute blood loss.

CASE 2

A 44-year-old white man received a diagnosis of panhypopituitarism in 1964. Three years later, he was found to have a chromophobe adenoma and irradiation treatment was undertaken. Since then, the patient has done well on endocrine replacement therapy. Throughout these problems, the patient had an entirely normal ocular examination including 20/15 vision, both eyes. His visual fields and optic discs were normal although he was reported to have large, symmetrical physiologic cups (c/d ratio, 0.5 both eyes).

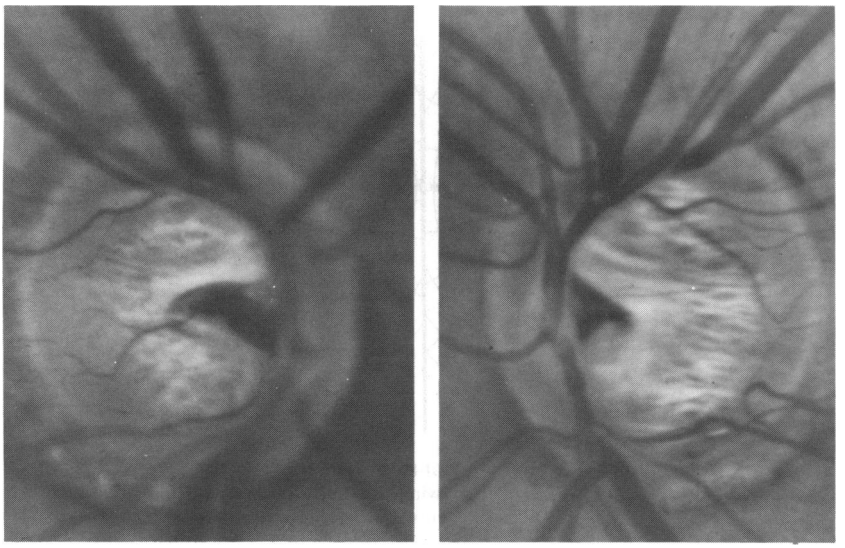

FIGURE 3
Right and left optic discs respectively of Case #2. The enlarged area of pallor on the left disc can be seen extending to the inferior rim. The vessel pattern is a bit unusual and different than the right disc.

In 1974 he reported a sudden onset of decreased vision on the left. He was not examined until three weeks later. He reported that the vision had not changed further. He had vision of 20/15 both eyes, uncorrected. External and slit lamp examination was unremarkable. Gonioscopy demonstrated wide open angles.

Intraocular pressure by applanation was 18 mm Hg in each eye and facility of outflow 0.29 each eye. Blood pressure measured 126/96 in a sitting position. Both optic discs showed the large cups described previously but there was a change in the inferior border of the left disc (Fig. 3). Visual fields on the Goldmann perimeter showed a dense superior arcuate defect close to fixation (Fig. 4).

Considering that the field change might be on the basis of pituitary tumor, plans were being made for carotid arteriography and pneumoencephalography. However, on evaluating the patient, we recognized this as an acute optic disc infarction and suggested that nothing further be done. There has been no change in status since the original event.

This patient does not have a history of acute hemodynamic crisis. However, the relationship of his pituitary insufficiency to his optic disc infarct is unknown.

CASE 3

A 47-year-old white woman noted a sudden onset of blurred vision in the left eye in March of 1976. No abnormalities were found other than an unusual appearance of the inferior temporal border of the left optic disc. This was associ-

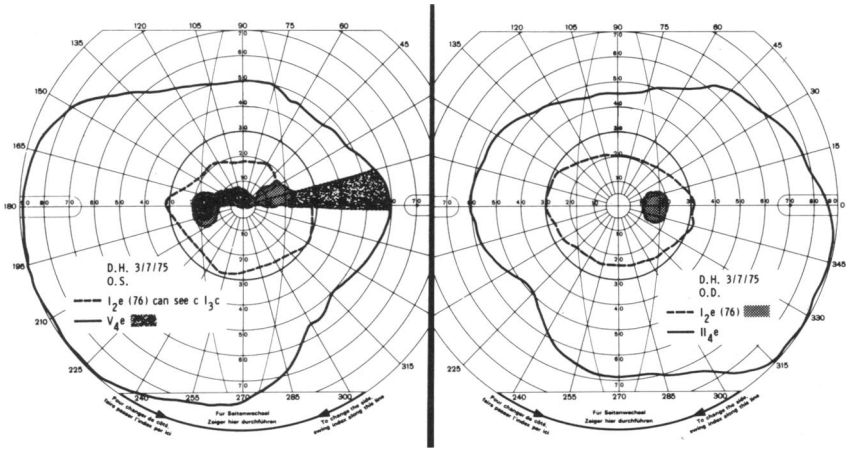

FIGURE 4
Visual fields of each eye of Case #2.

ated with a dense superior arcuate field defect. Intraocular pressure was 18 mm Hg on the right and 20 mm Hg on the left.

In July, 1976, the patient felt there was further visual loss although the objective examination was unchanged. Intraocular pressure was RE 17 mm Hg; LE 19 mm Hg. Epinephrine 2% drops were prescribed for the left eye twice daily.

In August, 1976 a small hemorrhage was seen at the 5:30 position on the left disc margin associated with a notching of the disc (Fig. 5). The field was unchanged. By January, 1977, the hemorrhage had disappeared.

We first examined this patient in March, 1977. Findings were unchanged from those previously reported. Intraocular pressure was 15 mm Hg both eyes. Facility of outflow was RE 0.38; LE 0.40. Gonioscopy demonstrated wide open angles in each eye. The right optic disc was unremarkable, but the left continued to show a tongue of pallor extending into a notch on the disc at the inferotemporal edge (Fig. 6). This corresponded to a dense superior arcuate field defect (Fig. 7). Fluorescein angiography showed avascularity in the area of the notch but was otherwise unremarkable. Since the patient was using epinephrine in each eye she was asked to stop this medication.

One month later, her intraocular pressure was 14 mm Hg both eyes. Repeat facility of outflow was RE 0.21; OE 0.23. The diagnosis was acute optic disc infarction and no treatment was advised.

DISCUSSION

This report is being made to call attention to what we believe is a specific entity which, if not recognized, could lead to unnecessary diagnostic studies and useless treatment. The patient often reports a definite and sudden alteration in vision just above fixation. Optic disc and visual field

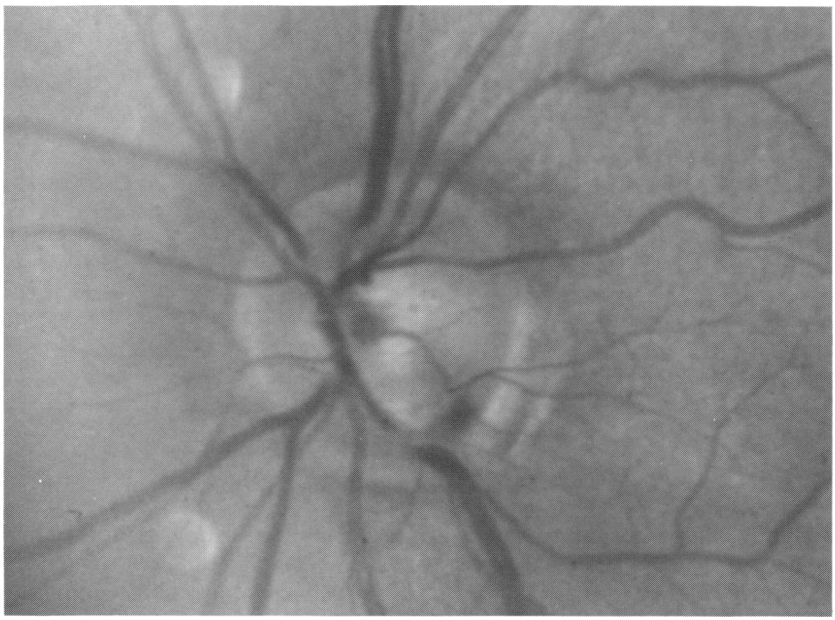

FIGURE 5
Left disc of Case #3 shows hemorrhage and notching of inferior temporal border.

changes confirm the patient's complaints. There is no evidence of abnormal aqueous dynamics. Systemic history may include previous vascular abnormalities.

There is a characteristic appearance to the optic disc which is not really typical of that usually seen in glaucoma. The disc has a subtle, shallow notch at the inferior temporal border with a tongue of pallor extending into the notch. There is often a peculiar appearance to capillaries in the damaged area possibly due to the shallow cupping and only minimal deviation of the course of the vessels. Another possibility is that these vessels are a form of neovascular alteration. When viewed monocularly, there is little evidence of a definite notch although the appearance is suspicious. However, binocular viewing confirms the presence of a definite loss of tissue at that point on the disc.

There may or may not be an associated hemorrhage of the rim tissue on the disc. If present, the hemorrhage occurs first and seems to disappear in a few weeks. The disc pallor, shallow depression, and visual field defect follow rapidly. Cases 1 and 3 showed this course of events. Case 2 did not demonstrate a hemorrhage.

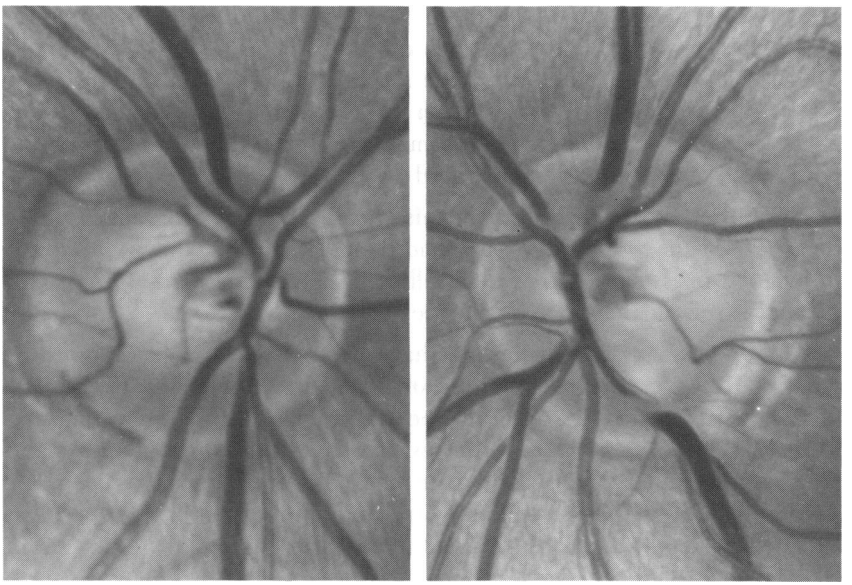

FIGURE 6

Right and left discs of Case #3 showing disappearance of disc hemorrhage on the left, but presence of notch and extension of pallor. A few fine vessels associated with this area can be seen to differ from the right disc. These vessels criss-cross and appear unusual. Careful scrutiny of this area of the retina reveals absence of nerve fibers corresponding to the disc defect (also seen in Figure 5).

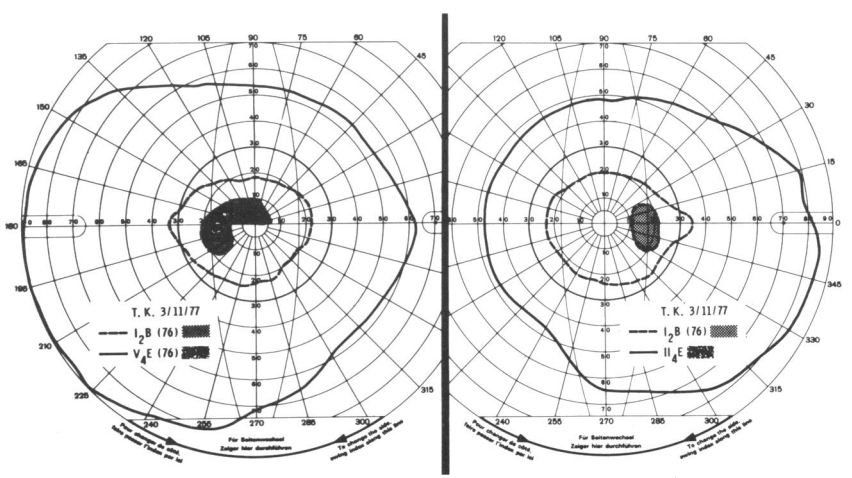

FIGURE 7

Right and left visual fields of Case #3.

The visual field defect is the result of a nerve fiber bundle loss, with the initial changes producing a small superior defect which gradually enlarges. There is an absolute nucleus to the scotoma with the remainder of the arcuate defect being relative. When the problem occurs bilaterally, one is impressed with the degree of symmetry present in disc appearance and field loss. This is well demonstrated by case 1.

The importance of recognizing this specific type of problem is that it does not appear to be progressive and does not warrant therapy to lower intraocular pressure. Rather than telling patients of a guarded prognosis, we feel it reasonable to be optimistic.

We feel certain that our findings have been observed and reported by others. However, such reports are obscured by their inclusion among a diverse set of cases. In addition the term "low tension" glaucoma has been used to cover all cases with low pressures having disc damage and field loss resembling that seen in glaucoma. Since this term is a receptacle for problems about which we know little and often can do little to resolve, a reader is likely to overlook the fact that all cases of so-called "low tension glaucoma" are not the same.[11]

It seems to us that the entity which we are describing has nothing to do with glaucoma. Rather there is a vascular accident in the prelaminar portion of the optic disc. More specifically this event occurs in the 7 o'clock position in the right disc and the 5 o'clock position in the left disc. This ischemic event may be associated with or heralded by a disc hemorrhage. In any event, atrophy of the optic disc ensues associated with a shallow notching. This damage to a nerve fiber bundle results in an arcuate visual field defect close to fixation.

Rather than calling this particular entity a type of "low tension" glaucoma, we prefer to use the term "acute inferotemporal prelaminar disc infarction syndrome." The acute nature of the optic disc damage and visual field loss is consistent with vascular accidents in other organs of the body. The optic nerve would not seem to have a special immunity to such an event.

Ischemic infarcts of small areas deep in the brain called lacunes are found in 11% of autopsy brains.[12] Although many of these are asymptomatic they are clearly related to systemic hypertension and cerebral atherosclerosis. Fisher describes evidence for embolism causing the lacunes. He uses as support the fact that emboli can be found in branches of the central retinal artery and that similar emboli could make their way deep into the brain. Whether lacunar infarcts of the brain are related to the infarcts we are describing in the optic disc is only speculative.

However, there are a number of questions left unanswered in our minds for which further studies and followup will be necessary. First, is the ischemic process in the optic nerve an isolated event or does it portend future vascular problems either in the optic nerve or elsewhere? Second, is there a difference between the disc cupping seen in primary open-angle glaucoma and the events we describe occurring at normal intraocular pressures? Third, why is the inferior temporal location on the disc apparently so susceptible to ischemia? Although these questions remain to be answered definitively, there are some clues already available from the literature and from the cases we have selected as examples.

We feel that the specific ischemic process we are describing is, indeed, an isolated event. Other studies on low tension glaucoma have pointed out types of patients where the field loss does not appear to be progressive.[5,7,9] These are cases where the disc damage and field loss occurred acutely, associated with a definite systemic event such as acute blood loss. Our patients may represent the same type of problem but not have an obvious systemic event associated with the acute nerve damage. The particular susceptibility of the optic nerve in the inferior temporal area may make a relatively minor ischemic episode a major event in terms of that particular portion of the nerve.

It seems that there may be a difference between the disc damage associated with our cases and that found in patients with elevated intraocular pressure. One difference is that our description only includes hemorrhages at the inferotemporal position. We have not seen this particular entity with a hemorrhage in any other location. This is not to say such cannot occur, but our experience would suggest it is unusual at best. It is of interest that a report of cases with disc hemorrhage states that the hemorrhages nearly always occur at the inferotemporoal disc border.[4] Another difference is that disc hemorrhages may persist in patients with or without elevated intraocular pressure, but may not be associated with rapid axonal death and visual field loss. Figure 8 illustrates such a case.

Further, disc damage which occurs in chronic glaucoma is gradual with a slowly enlarging cup. The damage and notching we describe is acute. This acute type of damage can be seen in glaucomatous eyes, however (Fig. 9). An identical hemorrhage followed by notch and arcuate field defect seem in a glaucomatous eye may have the same meaning as the cases we describe — namely, nothing to do with glaucoma. However, we are not ready to state that such acute changes in eyes with elevated pressure should be ignored. Although disc hemorrhages were found associated with greater progression of field loss,[10] it is possible that such

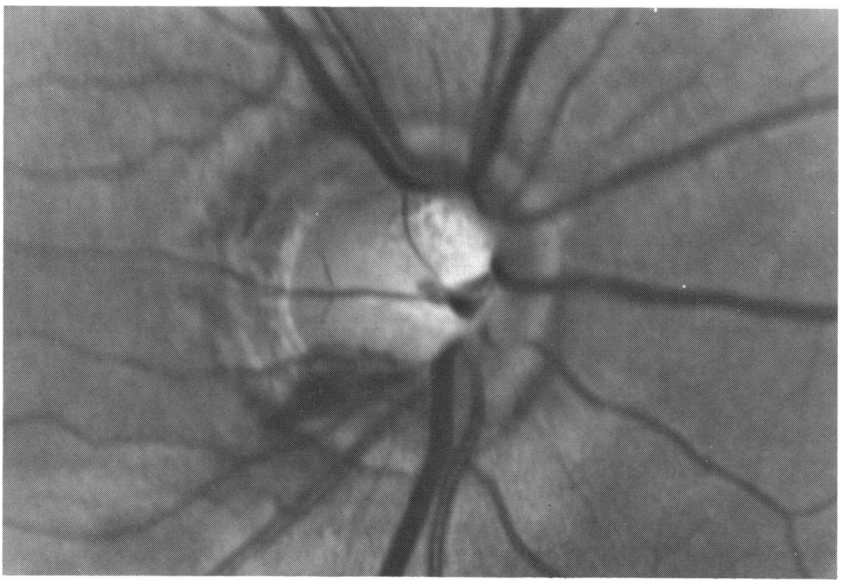

FIGURE 8
Inferotemporal disc margin hemorrhage in an ocular hypertensive. The hemorrhage has persisted for over one year and there is no associated visual field defect.

field loss is either related to the hemorrhage acutely as we describe, or is a sign of diffuse disc ischemia such as occurs in chronic glaucoma.

As to why the inferotemporal portion of the optic nerve is so susceptible to damage, one could speculate on the relationship of the arcuate nerve fiber bundles to the pupillomacular bundle. In dealing with a similar question, Ernest[13] points out that since the papillomacular bundle occupies the temporal portion of the nerve, the superior and inferior arcuate fibers are squeezed nasally. This packing of nerve fibers would create an increased metabolic demand of the superior and inferior disc areas. If there were a uniform sudden or gradual compromise in the disc vasculature, this increased metabolic need might account for specific damage confined to such areas.

Furthermore, the proximity to fixation of the superior arcuate defect in our cases, suggests that the ischemic event occurs in a vessel supplying nerve fibers up to but not including the papillomacular bundle. It would seem reasonable to assume that the macular fibers have their own specific vascular supply which does not involve adjacent vessels supplying the arcuate fibers. Although the precise end-vascular supply is not known for these areas, the watershed effect mentioned by Hayreh[14] as applying

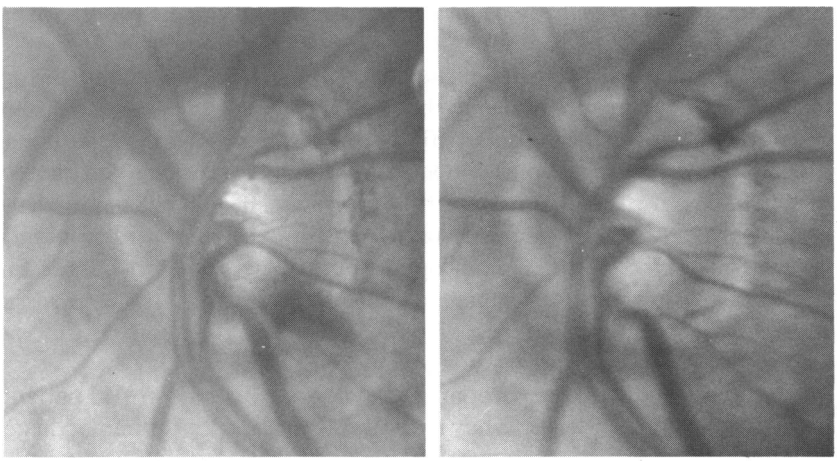

FIGURE 9
Composite of left optic disc of patient with ocular hypertension. Without treatment, intraocular pressure in mid 30s. On therapy, pressures in low 20s. Developed acute hemorrhage at inferotemporal margin of left disc. The hemorrhage cleared but a notch with pallor remained. This was associated with a superior arcuate field defect close to fixation which began as a relative defect and in a few weeks was dense. (The pupil dilated poorly due to long-term miotic therapy, which accounts for the imperfect quality of the photographs).

to the susceptibility of the choroidal circulation to elevated intraocular pressure may also apply at the disc. Hayreh also points out that branches directly from the posterior ciliary arteries seem to have a "sectoral distribution in the region of the lamina cribrosa with little anastomoses between adjacent arteries." He also feels that each branch supplies a specific sector of the nerve and involvement of one branch could produce a nerve fiber bundle defect.

The anatomical relationship of the choroidal and retinal circulation to the optic disc were reviewed and studied by Anderson and Braverman.[16] They stressed the relationship of the disc circulation to the retina-optic nerve vascular system with less emphasis given to a relationship to the choroidal circulation. In any case they point out that the effect of ischemia on glaucomatous optic disc change is still uncertain.

In a sophisticated study of fluorescein angiography in normal individuals, patients with glaucoma, and patients with low tension glaucoma, Spaeth[17] found a frequent occurrence of persistent hypoperfusion of the inferotemporal portion of the disc in patients with low tension glaucoma. His Case 4 was similar to our first case, although a diurnal pressure curve did show a single rise to 23 mm Hg. Findings in this case suggested to Spaeth that hypoperfusion may be the cause of the initial vision loss

and that disc hemorrhage may not be the primary pathologic event causing the loss of vision.

In a report on low-tension glaucoma, Drance[5] reviewed the history of the disorder and pointed out that the term emcompasses a number of entities having differing sets of findings. He mentions that there may be many ways a disc becomes cupped and atrophic. He observed "notching of the neuroretinal rim after the appearnce of a small hemorrhage on the disc with the appearance of a corresponding nerve fiber bundle defect considered to be a small optic nerve infarction." In 59 percent of his cases, low tension glaucoma did not progress. Drance stresses that lack of progression is expected when a hemodynamic crisis was associated with sudden disc damage and field loss.

Chumbley and Brubaker[9] reported a retrospective study of 45 patients with low tension glaucoma. They note that 10% had splinter hemorrhages on the disc which they suggest may represent infarction or vascular insufficiency to the disc. Also noted was that a better prognosis existed for individuals with sudden, permanent damage and an associated hemodynamic event. Very likely, some of these cases may have been identical to the syndrome which we are describing.

It may be reasonable to question the relationship of ischemic optic neuropathy to the entity we are emphasizing. Typically, ischemic optic neuropathy shows a pale, edematous optic disc.[18] Hemorrhage is common. Although our cases sometimes demonstrate associated disc hemorrhage, there is no edema noted. Further it is extremely unusual to see notching of the disc with ischemic optic neuropathy. Only atrophy occurs. On the other hand, our cases show both cupping and atrophy.

Whether or not our cases could represent a form of cranial arteritis is also questionable. Such patients with optic disc involvement typically show edema and hemorrhage, the findings of ischemic optic neruopathy. We did not perform sedimentation rates on our patients, but there were certainly no other signs or symptoms of cranial arteritis. Nevertheless, cases of "occult" temporal arteritis have been reported where only ocular involvement is seen.[19]

Although the very broad term of ischemic optic neuropathy could describe our cases, that term has come to be associated with a specific set of findings which our cases fail to demonstrate. Thus we feel the use of that term to be inappropriate.

In any event, although the cause of the syndrome is in doubt, our cases seem to be supported by other reports of similar cases where acute optic disc damage occurs and does not progress. Such cases are unusual but far from rare. Being aware of the specific syndrome of acute

inferotemporal prelaminar optic disc infarction should be helpful to all ophthalmologists.

SUMMARY

A specific type of optic disc damage and visual field loss has been emphasized. The changes occur acutely at the inferotemporal disc margin and do not seem to progress. Such patients have normal intraocular pressures.

We have termed this entity "acute inferotemporal prelaminar optic disc infarction syndrome" in an effort to avoid using the word glaucoma. Although similar events can occur in glaucomatous eyes, we feel that such changes are not progressive and that sophisticated diagnostic studies should be avoided. Three cases are presented as examples.

ACKNOWLEDGEMENT

Case 3 was seen through the courtesy of Richard F. Brubaker, MD, and Figure 5 was kindly provided by him.

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DISCUSSION

Dr Steven M. Podos. Faced with a patient who demonstrates optic nervehead cupping, optic atrophy, and arcuate field loss in the apparent absence of elevated intraocular pressure, the ophthalmologist often gropes for an explanation consistent with preconceived notions about the glaucomatous process and shudders at the prospect of treating potential progression. Often the wastebasket appellation of low tension glaucoma is employed to characterize this situation.

Before resorting to this diagnosis it is appropriate to verify the presence of low intraocular pressures, past and present. Some of these individuals may represent old glaucoma that is currently nonprogressive, damage caused by prior attacks of angle-closure, secondary glaucoma or corticosteroid-related phenomena. Others also can be suspected of having "high-tension glaucoma" when increased intraocular pressures are found at odd hours on diurnal testing, low outflow facility is present on tonography, or topical corticosteroids elicit a significant elevation of intraocular pressure. Still others when evaluated neurologically demonstrate conditions in the orbit or intracranially that can mimic glaucomatous damage. Finally one is left with a category of vascular etiologies.

The vascular hypothesis may explain the pathogenesis of optic nerve damage in glaucoma (Phelps CD: *Current Concepts in Ophthalmology,* Vol. III, F.C. Blodi ed., C.V. Mosby Co., St. Louis, 1972, p 142). An imbalance between intraocular pressure and the arterial pressure of the blood vessels entering the eye is postulated. Although this explanation does not account for individual differences in pressure sensitivity or selective ischemia of the arcuate nerve fibers, it is appealing as it relates well to the known observations of glaucomatous progression in recently treated systemic hypertensive patients and the occurrence of glaucomatous damage in patients with intraocular pressures varying from low to high.

In studying the patient with low-tension glaucoma, if high tensions or neurologic disease are not found, one must turn to what little information is available about such vascular factors. The thorough studies of Drance (Drance SM: Br J Ophthalomol 56:229, 1972) indicate an association with hemodynamic crises, low systemic blood pressure, possible hematologic abnormalities, cardiovascular disease, carotid insufficiency, and diabetes mellitus. The common denominator here is ischemia. Notable in this condition is the frequent finding of small hemorrhages of the optic nervehead rim, which evolve into notching weeks to months after the hemorrhage disappears. The hemorrhages are not always inferior in

location [slides 1-5]. Is this ischemic optic neuropathy, disc infarction, or glaucoma, or are they one and the same?

The great difficulties in further lowering the intraocular pressure of such a patient and the questions of the efficacy of such therapy warrant further studies of this issue. In 1973, Drance and co-workers (Drance SM, Morgan RW, Sweeney VP: NEJM 288:392-395, 1973) reported on 38 patients with low-tension glaucoma. Of these, ten had a history of shock prior to recognition of field loss and optic disc appearance characteristic of glaucoma. Etiologies of the shock included gastrointestinal bleeding, cardiac arrest, myocardial infarction, and uterine bleeding. Only one of these ten patients had progression of field loss as compared to 18 of the 28 patients with no history of hemodynamic crisis. This definition of a portion of the spectrum of low-tension glaucoma has important therapeutic implications. The patient with a history of hemodynamic episodes associated with a true low-tension glaucoma syndrome and no evidence of neurologic causes may be followed closely off therapy. Progressive damage is less likely to occur in this individual and ocular hypotensive agents or further diagnostic workup may be withheld until evidence of progression. Similarly, the abnormality at the inferior margin of the optic disc described in the present paper also may indicate a more benign course [slide 6].

The ability to predict which patients with the low-tension glaucoma picture have had an acute, nonprogressive insult and which ones will deteriorate is crucial. We are indebted to Doctors Lichter and Henderson for bringing yet another such predictive finding to our attention.

DR GEORGE SPAETH. Dr Harrington, Ladies, and Gentlemen. I would like to congratulate Dr Lichter for presenting what I think is an important paper. We believe very strongly that this is indeed a specific finding. We would like to give this a different name than Dr Lichter has used. In our exhibit at the AMA meeting in 1974 we presented 22 cases which we felt fulfilled criteria similar to those he mentioned. We have also published material in this regard. (*Tran Am Acad Ophthalmol Otolaryngol* 81:217, 1976; *Br J Ophthalmol* 61:126, 1977) We called this entity "focal ischemic glaucoma." The points that Dr Lichter made bear repeating. Specifically, the visual field loss is highly characteristic. It does not usually break through to the periphery; it is a dense paracentral scotoma just as he showed. About another point, however, we have some disagreement. We have found progression of the visual field loss in almost one-half of these patients. One individual comes to mind immediately. She had a paracentral scotoma in one eye and a perfectly normal field in the other eye. During the next years the scotoma in the one eye became denser and enlarged slightly though did not greatly involve the periphery. New field loss developed in the other eye. Of 18 cases of this specific entity being reported in a new publication, 8 have shown progression.

Regarding the work-up of these patients, we believe they should have a glucose tolerance test since approximately one-half will show a positive glucose tolerance

test; many are unaware of their diabetes at the time of recognition of their focal ischemic glaucoma.

Regarding the pathogenesis of the entity, I am not convinced that the development of the hemorrhage is a sign of an infarction. One individual who was being studied with fluorescein angiography showed no hemorrhage, but between the time time of the initial disc photography and the later fluorescein angiography (on the same day), a hemorrhage developed; this showed in fluorescein angiography. (*Tran Am Opthalmol Soc* 73:491, 1975) Yet, visual field examination done at that time and repeated about one week later showed no visual field defects. We have also other cases in which hemorrhage developed without appearance of a new or extended visual field defect.

We can consider "focal ischemic glaucoma" a specific entity. Is it really "glaucoma?" Is it an infarction? The answer is uncertain. The patients, however, do deserve careful follow-up and we believe should be treated as if they have glaucoma.

Dr J Lawton Smith. I gave a paper at the Wilmer Meeting on April 22, 1976 entitled "*The 6:45 Syndrome.*" When I read the abstract of Dr Lichter's paper in the program of this meeting, it was immediately apparent to me that we were both looking at exactly the same clinical entity, but with different interpretations. Many patients with "low tension glaucoma" are referred for neuro-ophthalmologic examination to rule out other causes for atrophic cupping and visual field loss. I have found that with a meticulous Hruby lens examination, many of these patients have small gray oval subtle lower temporal optic disc lesions, which I have interpreted as congenital pits of the nerve. The following case is an example.

This patient was a 45-year-old lady who had worn contact lenses for myopia for 16 years comfortably. About six years before, she had noted a little blurring in the left eye. She was seen in New York with 20/25 acuity and a cecocentral field defect in the involved eye. Neurologic studies were negative. Dr Max Chamlin did her fields every six months for the next six years and noted no change whatever. She saw another ophthalmologist and the question of "low tension glaucoma" was raised. The patient was admitted to Manhattan Eye and Ear Hospital, found to have a normal diurnal curve, negative glucose tolerance test, and later had a negative EMI scan. She was later seen at the Wilmer Institute, had a repeat diurnal curve including measurements at 0200 in the morning, and all the tensions were normal.

When we saw her on September 18, 1975, the corrected vision was 20/15 in the right eye and 20/25 in the left eye, the applanation tensions were 15, the peripheral fields were normal, but she had characteristic central field defects in both eyes, which I like to call "the pistol." This is a ceco-upper paracentral scotoma, which looks like an old time pirate's pistol. Fundus photographs revealed that with only a quick look, no notable abnormality might be seen. However, with enlarged stereoscopic pictures of the discs, a small hemorrhage was seen at the 6:45 position in the right eye coming out of a little gray oval slit. The left eye had a small pit at the 3 o'clock position with a small vessel going across

its base. I interpreted the field defects as due to these subtle pits and reassured her. Six months later there was no change in field vision, and the hemorrhage in right eye had cleared. I have now seen approximately eight of these cases since last year and all show the same thing, varying only in degree.

Other helpful signs are a partial inferior Fuchs' coloboma, slight tilting of the disc, or very subtle signs of a congenital variant of the disc. It is important to remember that you do not have to have a serous detachment of the macula to have a visual field defect with a congenital pit of the nerve. Kranenburg (*Arch Ophthalmol* 64:912, 1960) gave a classic paper on congenital pits of the nerve and shows the cecoparacentral scotoma in that paper, which is an exact replica of "the pistol," which we are now seeing in these cases. It is obvious that this syndrome may occur with normal tension, with ocular hypertension, and with glaucoma. However, I believe that the *"6:45 syndrome"* is the best name we have at the moment (of course, in the left eye the defect is at 5:15) as it tells you where to look at a disc with the Hruby lens when you see an unexplained visual loss, and particularly in a patient suspected of having "low tension glaucoma." Further study of the families of these patients will be helpful in telling whether there is a congenital pit present or whether the defect is due to optic nerve infarction, as Dr Lichter and Henderson have suggested. I believe this is an extremely important clinical paper and congratulate the authors on their presentation.

Dr Robert N. Shaffer. I have enjoyed this excellent paper by Dr Lichter and Dr Henderson. Their interesting observations may serve to help us understand more about the mechanism of nerve damage in open-angle glaucoma.

I would like to present a case history of a 52-year-old woman who was first seen in our office in February of 1970. She had a definite open-angle glaucoma with a left disc which was excavated to the lower temporal margin and a corresponding upper arcuate scotoma which has persisted to the present time. Her intraocular pressure in both eyes was in the low 20's. She had mild systemic hypertension and mild diabetes, controlled by diet. Her right disc appeared normal with a cup-disc ratio of 0.3. Seven months later she was found to have a hemorrhage on the lower temporal disc margin with a relative upper arcuate scotoma in the right eye. Six months later there was a mild hollowing of the lower temporal disc and the scotoma had become absolute. Had we not seen the hemorrhage this case would be similar to those described in this paper, particularly if there had been no further damage. Unfortunately she again had a disc hemorrhage in 1972 and a third one in 1974. The optic cup in the right eye slowly increased until there is no lower temporal neural rim, matching the left disc. A large dense upper arcuate scotoma is present in both eyes.

I suspect that Dr Maumenee will say this nerve damage is due to localized slippage of the lamina cribrosa. Personally I think we are seeing a surface manifestation of tiny infarcts in the nerve head. These may be limited to one episode, or repeated incidents can result in progressive cupping and advancing arcuate scotomas which are typical of uncontrolled open-angled glaucoma.

DR JOHN WOODWORTH HENDERSON. An additional patient was recently seen in our department. A 54-year-old man had noted a partial field defect affecting the left edge of words in the left eye in October, 1976. In February 1977 a new field defect was noted in the right eye. He was referred to Dr Richard A. Lewis for possible neurologic disease. The history elicited no related systemic illness or family history of glaucoma. His blood pressure was 145/85. Vision was 20/15-2 OD, 20/20-2 OS corrected. The visual field showed bilateral superior arcuate scotomas extending from the blind spots. Applanation tensions were 16 mm Hg OU, dilated and undilated. The outflow facility was 0.24 in each eye. Ophthalmoscopic examination showed symmetrical inferior notching of the nerve heads as noted in the slides.

The symmetry of the condition in certain patients (such as Case 1 in our paper) is of interest. In case 1, I watched the process develop in the second eye after hemorrhage along the disc margin to become an exact mirror image in the visual field and in the appearance of the disc. The field defects have been stable for a number of years since onset, and there has never been any evidence of glaucoma.

The patient just shown has not had sufficient follow-up as yet to prove stability. Interestingly, he has a twin brother who has not yet been examined.

DR RALPH LEVENE. I enjoyed the paper and I wish to bring up one point. In my experience, the pressures in these eyes usually have peaks over 15, that is from 16 to 20. Since the normal range of intraocular pressure runs from 10 to 20 with a mean of 15 this may be a significant observation and we must consider the normative concept of Friedenwald and others. If someone starts with a pressure of 10 or 11 and subsequently develops a pressure of 16 the latter may be normal in one sense but not for that patient. What have you found in this regard?

DR PAUL LICHTER. I would like to thank Dr Podos for his discussion as well as the others who have added a lot to this paper. I want to reemphasize one thing about this entity and that is that it is acute. In most cases of glaucoma when we see changes occur, they are not acute but are gradual. The changes are usually progressive. Not so in our infarct cases.

Regarding the comments about treating these patients, I think it is not necessary to treat the patient and to make the patient worry. On the other hand, if an entity similar to our cases is shown to be progressive, as Dr Shaffer showed in his case, then, of course, we are really groping as to what to do. We want to do something to improve the vascularity of the optic disc. The only thing we know to do is perhaps to improve the circulation by lowering the intraocular pressure with medication. Can I have the slides please.

As to the concept of an optic nerve pit, being responsible for our findings, Dr Henderson mentioned that we thought about this possibility and even mentioned it on the record of the first case that we presented. We finally decided that these were not pits, although resembling them in some ways.

[Slide] This is a disc from one of the cases we showed you. It does not look like a pit. In the cases that Dr Smith discussed, he is talking about very subtle pits.

I brought along a few photographs of optic disc pits thinking perhaps someone might raise this point. [slide] This is, of course, one with macular edema, and I don't think we would confuse it with our cases. [slide] Here's still another in stereo; you can see a very huge depression of the disc extending into this pit and this patient did have a dense superior arcuate defect much like the cases we showed. However this pit is at the 9 o'clock position on the disc. Here's one in the 5-o'clock position but not really subtle. The color is not at all like that in the cases we presented, nor is the pallor the same.

We are not certain as to what the disc defect is, but we have seen cases where we have documented that the disc has been entirely normal before the infarct occurs. Thus, if some of the cases are indeed pits, they are acquired and most unusual.

We thank the program committee for the opportunity to present this paper.