Pit-like changes of the optic nerve head in open-angle glaucoma

RONALD L. RADIUS, A. EDWARD MAUMENEE, AND WILLIAM R. GREEN

From the Wilmer Ophthalmologic Institute, Johns Hopkins Hospital, Johns Hopkins University School of Medicine, Baltimore, Maryland, USA

SUMMARY Six patients with open-angle glaucoma and acquired pit-like changes in the optic nerve head are presented. In 1 patient evolution of the pit-like defect is documented. In all 6 patients progression of associated visual field deficits is described. It is suggested that such pit-like changes in selected patients with glaucoma may not represent congenital lesions but rather local, progressive nerve head disease, occurring particularly in response to raised intraocular pressure. The management of patients with optic nerve head pitting and the pathogenesis of glaucomatous optic neuropathy are discussed with respect to this observation.

Pitting of the optic nerve head is a well-recognised phenomenon representing localised ectasia of the nerve head. It is believed to be similar in pathophysiology to more extensive colobomas of the optic nerve and ocular tunics (Greear, 1942; Sugar, 1962; Duke-Elder, 1964; Walsh and Hoyt, 1969; Smith, 1977). Associated visual field defects have been described in cases of congenital pitting of the optic nerve head (Greear, 1942; Sugar, 1962; Duke-Elder, 1964; Walsh and Hoyt, 1969). Except in cases with transudation and sensory retinal detachment, however, such pathology is considered self-limited and stable.

The present report describes pitting of the optic nerve head in patients with open-angle glaucoma. Associated visual field defects and progressive ocular lesions in these cases are presented. The implications of these observations are discussed with respect to both management of optic nerve head pitting and pathophysiology of optic nerve head cupping.

Case reports

Case 1
This 57-year-old white man was first seen in 1967, at which time he was using both topical and systemic medications as therapy for open-angle glaucoma. Vision was 20/20 bilaterally. Both optic nerve heads showed cup-to-disc ratios of 0.40. An early nasal step defect was noted in the peripheral field of each eye. Intraocular pressures ranged from 25 to 36 mmHg. In 1969, filtration surgery was performed bilaterally. Intraocular pressure continued high postoperatively, and development of superior arcuate scotomata was noted bilaterally. A pit-like defect at the inferior margin of the left optic nerve head was noted coincident with this progressive field loss (Fig. 1). After the topical application of pilocarpine 4% was reinstated, visual function and disc pathology remained stable.

Case 2
This 36-year-old white man was struck in the right eye by a snowball. Immediately after this injury examination revealed a hyphaema, with vision reduced to hand motions. Anterior chamber haemorrhage cleared, and visual function returned to normal. The patient was seen in consultation for raised intraocular pressure. He was noted to have pressure elevations as high as 30 mmHg, probable angle recession, decreased facility of outflow, and slightly elongated but physiological cupping of the optic nerve head. Visual field examination with the Goldmann perimeter using the 1/2 and 1/4 test stimulus was completely normal. The left eye was unremarkable in all aspects.

During the subsequent 3 years pressure control remained difficult despite both topical and systemic medications. With persistent pressure elevation of greater than 30 mmHg filtration surgery was finally recommended. During this period of pressure
Fig. 1 Picture should be viewed when reader is wearing +10.00 spectacles in order to obtain stereoscopic view.

elevation, however, stereo disc photography documented progression of a pit-like slit at the inferior pole of the right optic nerve head (Fig. 2). Development of this defect corresponded to progression of an early arcuate scotoma in the superior nasal field of that right eye. Ophthalmoscopy revealed obvious nerve fibre layer loss (Fig. 2).

After filtration surgery, intraocular pressures remained below 22 mmHg, and no further progression in disc pathology or field loss was noted.

CASE 3
In 1960 this 69-year-old white woman first noted mild reduced vision of 20/30 in her right eye. Examination at that time showed immature cataract. Intraocular pressures were 14 mmHg bilaterally. The optic nerve heads were normal, and visual field examinations gave normal results. Rapid progression of the lenticular opacity reduced vision to 20/400. The patient underwent uneventful cataract surgery.

In 1965 she returned for routine examination, at which time intraocular pressures remained below 20 mmHg. Tonography was moderately reduced, 0.17 µl/min-mmHg, and repeat field evaluations demonstrated general depression of field test stimuli but no specific defects.

In 1975 a definite pit-like defect of the optic nerve head was noted inferiorly. Visual field evaluation revealed a dense, superior arcuate scotoma when testing with the V/4 test stimulus. While intraocular pressure remained below 20 mmHg on several occasions, in the supine position this patient’s ocular tension rose to between 20 and 26 mmHg. Tonography revealed that facility of outflow had fallen to 0.10 µl/min-mmHg. After instituting topical therapy with pilocarpine 2%, no further deterioration in visual field function or progression in nerve head pathology was noted.

CASE 4
This 63-year-old white woman with a long history of therapy for open-angle glaucoma was first seen in consultation in 1962. At that time her visual acuity was right eye 20/60 and left eye 20/30. Intraocular pressures were 50 and 34 mmHg in right and left eyes. Facility of outflow was 0.06 µl/min-mmHg bilaterally. Visual field examination with the Goldmann perimeter showed general depression of all test isopters but no specific glaucomatous field defect. Optic nerve heads appeared normal, with a cup-to-disc ratio of 0.50. Patient intolerance of all topical and systemic treatment given so far prompted several subsequent attempts at filtration surgery, but intraocular pressures remained raised at 20 to 30 mmHg. Despite persistent pressure elevation no definite field deficit could be found. In 1972, how-

Fig. 2 Vertical elongation of the physical cup with a pit-like change seen inferiorly (white arrow) in this patient with secondary glaucoma and angle recession. Drop-out of nerve fibres was evident in an inferior arcuate pattern (double arrows). A superior arcuate field deficit was documented by perimetry.
ever, superior nasal field loss was noted in the left eye. A pit-like defect of the optic nerve head was seen inferiorly (Fig. 3).

CASE 5
This 72-year-old black woman had been treated since 1972 both topically and systemically in an effort to control increased pressure in both eyes. Vision in the right eye was reduced to 20/400 with unilateral high myopia and amblyopia. Vision in the left eye was 20/40 with mild cataractous lens changes. With poor patient compliance, pressure elevation persisted, ranging from between 15 and 25 mmHg.

A superior nasal field defect in the peripheral field of this patient's left eye gradually progressed. Ophthalmoscopy showed both optic nerve heads to be pale with increased cupping. Cup-to-disc ratios were right eye 0.70 and left eye 0.80. A pit-like defect similar to those described in previous patients was noted inferiorly.

CASE 6
This 74-year-old white woman was first seen in consultation in 1969 for examination of a post-operative filtration bleb following cataract surgery in the left eye. Vision in the right eye was 20/300 secondary to cataract. Best corrected acuity in the aphakic left eye was 20/30. Intraocular pressures were 5.5 and 10.0 with a 5.5 g weight. Visual fields tested with the Goldmann perimeter were full in both eyes. Ophthalmoscopy showed vertical elongation of the optic cup in the right eye.

After an uneventful operation for cataract in the right eye vision in that eye improved to 20/20. During the next several years at many follow-up examinations pressures remained below 20 mmHg, although tonography was moderately reduced in both eyes, 0.18 and 0.20 μl/min-mmHg.

When the patient was seen again in 1976 definite pitting of the optic nerve head of the right eye was noted inferiorly. Repeat visual field examination showed an early superior arcuate scotoma (1/4 test stimulus) as well as a peripheral nasal step defect.

Discussion
Pitting of the optic nerve head is well described (Greear, 1942; Sugar, 1962; Duke-Elder, 1964; Walsh and Hoyt, 1969; Smith, 1977). These ectasias characteristically involve superior and inferior temporal margins of the disc tissue. Associated field defects, occasionally arcuate in nature, have been described (Duke-Elder, 1964; Walsh and Hoyt, 1969; Smith, 1977). These pits are generally felt to represent localised colobomas of the optic nerve head and are congenital. Consequently, associated visual field deficits are considered to be characteristically benign or at least non-progressive (Walsh and Hoyt, 1969; Smith, 1977). Instances of acquired pitting of the optic nerve head have been noted as well, but implications of this observation have not been adequately stressed (Gayer, 1951; Lichter, 1978).

We have described 6 patients with open-angle glaucoma associated with pit-like defects of the optic nerve head. Although these ectasias may represent congenital pitting of the optic nerve head, in at least one instance (Case 2) stereo disc photography documented evolution of the abnormality. In each instance progression of associated field defects suggested acquired rather than congenital lesions.

In addition to this clinical material we have found pit-like areas of ectasia in the lamina cribrosa in eyes with advanced glaucoma obtained either at necropsy or as postenucleation specimens (Fig. 4). These defects were located at either the inferior or superior poles of the disc and were most easily identified when eyes were sectioned vertically. In no instance was there a good clinical description of the optic nerve head before the specimen was obtained. Eight such eyes have been studied histologically, and details of these experiments will be presented in a later report.

The inferences to be drawn from these observations are several. The tendency for these pit-like changes to occur at the vertical poles of the disc

Fig. 3 Inferior pit-like change was noted in the optic nerve head of this patient (black arrow) with open-angle glaucoma. The disc change corresponded to a dense arcuate scotoma of the superior visual field.
suggests an anatomical explanation for the arcuate field defect so characteristic of glaucoma. If the lamina cribrosa were structurally weaker and more prone to mechanical distortion at the superior and inferior disc poles, compression of axons traversing this area might result in ascending and descending optic atrophy.

The basic mechanism which might produce such a local outpouching is not certain. Whether such changes are primarily in response to raised intraocular pressure or represent secondary changes following ischaemic necrosis of neural tissue (Lichter, 1978) cannot be determined from available material. However, the histological evidence of localised areas of ectasia in the lamina cribrosa suggests that a localised infarct in the nerve fibre layer at the edge of the disc is not the cause of the pit-like change. The blood supplies to these 2 areas are independent, although they do anastomose (Lieberman et al., 1976; Minckler et al., 1977). Nor is it evident how such ectasia at the lamina might damage axons mechanically. It is known, however, that acute elevation of intraocular pressure in experimental animals can produce a blockage of axonal transport at the lamina cribrosa (Lieberman et al., 1976). Whether such blockage results from mechanical compression of individual axons or from focal ischaemia of the optic nerve, and whether such observations after acute elevation of intraocular pressure relate to ultimate atrophy of nerve fibres in chronic glaucoma, is not clear. It is hoped that better experimental glaucoma models and more exact clinicopathological correlation may help answer such questions.

A further importance of these observations is suggested by two of our patients in whom pressure elevation was not detected (Cases 3 and 6). In Case 3, however, decreased outflow facility and increased intraocular pressure in the supine position suggested that aqueous dynamics were abnormal. These 2 patients suggest that pit-like changes of the optic nerve might occur at relatively low pressures in some individuals. Variation in resistance of the lamina cribrosa might explain why certain individuals develop extensive nerve head cupping at presumably low elevation of intraocular pressures. It likewise suggests why other individuals with more resistant lamina, such as Case 4, can withstand high intraocular pressures for many years before suffering glaucomatous field loss.

Finally, apart from speculation on the causation of these pit-like changes in some patients with glaucoma it is important to recognise that, in some individuals, pit-like changes involving the optic nerve head represent progressive lesions, and those patients must be observed for the development of associated visual field defects, especially when the intraocular pressure is also raised. Even in patients with only intermittently raised intraocular pressures pit-like lesions of the optic nerve head may be associated with progressive field dysfunction and disease of the optic nerve head.

References


Pit-like changes of the optic nerve head in open-angle glaucoma

Pit-like changes of the optic nerve head in open-angle glaucoma.

R. L. Radius, A. E. Maumenee and W. R. Green

doi: 10.1136/bjo.62.6.389

Updated information and services can be found at:
http://bjo.bmj.com/content/62/6/389

Email alerting service

These include:
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/