Communications

Fundoscopic appearance of a nerve-fibre-bundle defect

WILLIAM F. HOYT, BERND SCHLICKE*, AND RONALD J. ECKELHOFF

Neuro-Ophthalmology Unit, Departments of Ophthalmology, Neurology, and Neurological Surgery, University of California, San Francisco

Focal destruction of nerve-fibre bundles is one of the pathological common denominators of diseases involving the optic nerve, the optic papilla, or the superficial layer of the retina. Since Bjerrum (1891) and Rönne (1909, 1913) first identified arcuate defects in the visual field as the perimetric correlates of defects in arcuate nerve fibres of the retina, quantitative perimetric studies have pre-empted ophthalmic interest for early diagnosis and assessment of these nerve-fibre-bundle defects (NFBDs). Direct visualization of NFBDs by ophthalmoscopy would provide the obvious advantages of an objective diagnostic technique, but the clinical feasibility of such a method has never been demonstrated.

Vogt (1913, 1917) pioneered direct ophthalmoscopic techniques for observation of retinal nerve fibres using a special type of illumination in which long (red) wave lengths were selectively filtered from white light emitted from a carbon arc. This *red-free* light greatly enhances visibility of superficial retinal reflexes, particularly the striated reflexes from the nerve-fibre bundles. We have adapted Vogt's method for evaluation of retinal-nerve-fibre abnormalities in patients with optic nerve disease and have repeatedly observed narrow slit-like gaps in the peripapillary nerve-fibre reflexes. In each instance there were corollary defects in the visual field. This report documents the fundoscopic appearance of small slit-like NFBDs in the arcuate nerve fibres of the retina, and describes the associated defects in the visual field as recorded by quantitative kinetic and static methods with the Tübingen Perimeter.

Clinical data

The patient was a 40-year-old Caucasian housewife who had experienced persistent dull pain behind her right eye and in her right brow for three weeks before blurring and dimness occurred in the inferior part of the visual field of the right eye. Her ophthalmologist found a dense inferior altitudinal defect that broke out from the blind spot in that eye; the ocular fundus and colour vision were normal and the visual acuity was 20/20. X-ray examinations of the sella turcica, optic foramina, and chest were normal, but studies of the spinal column showed mild rheumatoid osseous changes. The erythrocyte sedimentation rate was increased to 39 mm./hr. Prednisone (60 mg./day) administered for 10 days stopped the pain but did not change the scotoma.

When the patient was referred to the University Neuro-Ophthalmology Unit 4 weeks after her visual symptoms had begun, she reported clearing of the field defect. External ocular examination, ocular movements, and pupillary responses were normal. Colour vision with the pseudo-isochromatic plates (H-R-R) was normal in each eye.

Received for publication December 30, 1971

Address for reprints: Department of Neurological Surgery, University of California, Parnassus and Third Avenues, San Francisco, California 94122, U.S.A.

* Hearst Fellow in Ophthalmology, Universitäts Augenklinik, Tübingen, Germany

Fundoscopy findings

The colour and capillary network of the right optic disc appeared normal, but one superior nasal retinal arteriole appeared slightly narrow and irregular. However, when the superior temporal retina was examined with red-free light and a direct ophthalmoscope, two narrow arcuate ribbon-like parallel gaps in the striated peripapillary nerve-fibre layer became evident. These arcuate defects in the nerve-fibre layer were located below and parallel to the superior temporal vein and were approximately equal in width to the diameter of that vessel. They started at the disc margin between 10 and 11 o'clock, had a darker green colour than the adjacent nerve fibres, and coursed temporally for about $2\frac{1}{2}$ disc diameters before blending imperceptibly with the darkening retina. No other areas in the right and left fundus contained similar nerve-fibre defects or other abnormalities.

These arcuate gaps or grooves in the superior nerve-fibre bundles were recorded photographically with the Zeiss Fundus Camera on Kodak Plus-X film using $2\times$ magnification and a Wratten No. 65 (green) filter. The black and white negatives were printed on Kodak Ectomatic SC paper with a high contrast filter (Fig. 1).

Perimetric findings

No defects were found when the peripheral isopters of the visual fields were tested with the Goldmann perimeter. The central fields (0° – 30°) were examined by kinetic and static techniques with the Tübingen Perimeter.

Luminance of the bowl of the Tübingen instrument was set at 10 apostilbs (asb); the size of the test objects was 10 min. of arc; the size of the fixation point was 30 min. of arc. Before the examinations were begun the patient was thoroughly trained to give accurate and consistent test responses. Her performance during all the examinations was ideal.

The topographic central visual field examination (kinetic method) gave normal findings in the left eye and an inferior, relative, arcuate-type scotoma with nasal step in the right eye (Fig. 1). Also in the right eye, responses to a moving target of a relative luminance of 16 asb were the typical "flickering" or "in-and-out" type that is characteristic of NFBDs; these responses were found all over the scotomatous zone, especially nasally. Two narrow, absolute, island-shaped scotomas were found near the nasal step. A relative, band-shaped scotoma was found in the Bjerrum area inferiorly between the blind spot and the vertical meridian. No arcuate defects were found superiorly in the right visual field (Fig. 1).

Static profiles of the visual fields were recorded along several superior and inferior radial meridians. Particular attention was paid to:

- (1) The 307° meridian, which corresponded to the fundoscopically observed NFBDs;
- (2) The 195° meridian, which included the area of the kinetically determined island-shaped scotomas in the nasal step (Fig. 1).

The *static profile at the 307° meridian* (Fig. 1, inset A on the *right* margin) revealed a relative area of depressed sensitivity at about 13° from fixation. The *static profile at the 195° meridian* (Fig. 1, inset A on the *left* margin) showed two absolute depressions in sensitivity at 15° and 18° which corresponded to the two scotomas in the nasal step.

These static profiles at the two positions in the inferior visual field were redetermined 30 days later when the findings were similar except that the pit-like depressions in sensitivity in the 195° profile were now relative instead of absolute, and that a third relative depression was present at 21° (Fig. 1, inset B on the *left* margin).

Diagnosis

The patient's ocular findings indicated focal involvement of the right optic nerve. The cause of the disease could not be established with certainty, but a vascular aetiology was suggested by the following:

- (1) Prompt relief of pain after steroid administration;

FIG. 1 Retinal nerve-fibre-bundle defects. The red-free fundus photograph shows a double, dark, slit-like gap (between white arrows) in the superior arcuate fibres and extending to the disc margin.

- (2) Increased erythrocyte sedimentation rate;
- (3) Radiological signs of rheumatoid spondylitis;
- (4) Altitudinal scotoma with preservation of central acuity and colour vision;
- (5) Suspicious narrowing of a retinal arteriole.

Discussion

In reporting on red-free retinal-nerve-fibre signs in toxic amblyopia and traumatic optic atrophy, Vogt (1921) introduced an entirely new and refined dimension to ophthalmic diagnosis. The technical difficulty of applying his method in the 1920s may explain why the practical neuro-ophthalmological significance of his discovery never received the attention and acclaim that were its due. Only recently have Ito, Ozawa, Suga, and Mizuno (1969) reported their preliminary observations of vascular and nerve-fibre changes in papillitis using red-free direct ophthalmoscopy and magnifying fundus photography. Hoyt, Rios-Montenegro, Behrens, and Eckelhoff (1972) applied similar methods to identify the characteristic patterns in the retinal-nerve-fibre layers in patients with congenital hemianopia (homonymous hemioptic hypoplasia).

Recently, we have been using red-free ophthalmoscopic illumination routinely in our evaluation of patients with optic nerve involvement. Whenever we find a slit-like gap in the arcuate or radial nerve fibres, we also find a corresponding arcuate defect in the visual field (Fig. 2, A,B,C).

Retinal NFBDs from focal retinal infarction extend peripherally in the fundus (retrograde axonal degeneration) and centrally (Wallerian or ascending degeneration) (Fig. 2, A). Other NFBDs in the nerve-fibre layer exemplify retrograde (descending) axonal degeneration only; these occur with optic nerve involvement at any level (Fig. 2, B,C). Factors aiding observation of NFBDs are a well-dilated pupil, a well-pigmented fundus, and a direct ophthalmoscope with a red-free filter and a bright light source. Photographic documentation of NFBDs requires precise focusing, magnification of the nerve-fibre reflexes, and a black and white photographic paper that enhances contrast between the image of the NFBD and adjacent nerve fibres.

CORRELATION OF PERIMETRIC AND RETINAL SIGNS

Prediction of all details of a patient's visual field defect from the appearance of an NFBD in his fundus is not possible with the ophthalmoscopic or photographic method used in this study. However, as exemplified by our patient, the retinal signs satisfactorily explain

FIG. 2 (A, B, C) *Fundoscopic appearance of nerve-fibre-bundle defects photographed in red-free light.*

(A) A focal infarct (inf.) in the superficial retina has caused ascending (Wallerian) degeneration of a strip of nerve fibres (arrows) between the lesion and disc margin. Peripheral to the lesion there is a dark expanding wedge (on far right) representing retrograde axonal degeneration.

(B) Multiple gaps in nerve-fibre layer (arrows) and temporal disc pallor as the sequelae of acute optic neuritis. In addition to depression of central vision, there were multiple arcuate scotomas in the inferior "Bjerrum area" of the visual field.

(C) Multiple dark wedge-like defects (arrows) among the arcuate fibres paralleling the course of the superior temporal vessels.

There were corollary arcuate scotomas in the "Bjerrum area" of the visual field.

This was the asymptomatic right eye of a 65-year old patient with recent idiopathic optic disc infarction in the left eye.

most of the perimetric findings. The parallel slit-like gaps in the superior temporal nerve fibres (arcuate retinal fibres) correlated best with the two absolute scotomas in the inferior nasal step. The general depression in the inferior visual field exceeded the area

which could be predicted from the size and location of the retinal NFBD alone. This disproportion between perimetric and fundoscopic signs may indicate:

- (1) Conduction block in axons scattered through all of the superior arcuate fibre zone;
- (2) Scattered degeneration of axons through all of the superior arcuate fibre zone;
- (3) A combination of both.

One month after the initial perimetric examinations, the two slit-shaped scotomas in the inferior nasal step changed (improved) from absolute to relative. Whether the third pit in the static profile at the 195° meridian, recorded at the second examination (Fig. 1, inset B on the *left* margin), represented a newly acquired scotoma or one overlooked at the initial examination is not clear.

Interpretation of the relative arcuate scotoma in the inferior *temporal* field and its correlation with the visible nerve-fibre signs is speculative. Differentiation of an angioscotoma and an acquired scotoma from our static perimetric data is difficult. The more peripheral pit in the static profile at 307° (Fig. 1, inset A on the *right* margin) is located at 16° – 17° from fixation and is consistent with a scotoma produced by the superior temporal vein. The pit at 13° is probably abnormal and not a part of an angioscotoma as described by Weekers (1969) for the following reasons:

- (i) In several kinetic perimetric examinations the arcuate scotoma was broader than an angioscotoma and lacked connection with the blind spot.
- (ii) The sloping entry of the pit at 13° is unlike the profile of an angioscotoma but is consistent with the visible NFBD.
- (iii) The “flickering” sensation in the Bjerrum area determined by kinetic testing techniques covered a longer and wider zone than an angioscotoma.

Three generalizations seem warranted from our perimetric studies:

- (1) Every visible NFBD has an analogue in the visual field;
- (2) The size of the field defect will equal or exceed the area predicted from the size of the retinal NFBD;
- (3) The densest portion of the field defect will lie further from the blind spot than would be predicted ophthalmoscopically from the distance between the NFBD and the optic disc.

Small arcuate field defects are easily overlooked during a routine perimetric examination, but the finding of an NFBD by red-free ophthalmoscopy can direct the perimetrist's attention to the areas of the visual field in which such a scotoma will be located.

Conclusions and summary

This fundoscopic and perimetric correlative study indicates that a nerve-fibre-bundle defect, previously only a clinical perimetric concept, is an objective sign that can be seen by direct ophthalmoscopy using red-free illumination. It consists of a slit-like gap in the arcuate, or occasionally the radial, nerve-fibre pattern of the retina, and it is easily documented by magnified fundus photography on black and white film. Its presence always signifies permanent involvement of optic axons even in the patient without previous history or other fundoscopic signs of optic nerve disease.

References

- BJERRUM, J. (1891) *X int. med. Congr., Berlin*, 1890, Band IV, 10th Abtheilung: Augenheilkunde, p. 66. Hirschwald, Berlin

- HIOYT, W. F., RIOS-MONTENEGRO, E. N., BEHRENS, M. M., and ECKELHOFF, R. J. (1972) *Brit. J. Ophthalm.*, **56**, 537
- ITO, H., OZAWA, K., SUGA, S., and MIZUNO, K. (1969) *Folia ophthalm.*, **20**, 282
- RÖNNE, H. (1909) *v. Graefes Arch. Ophthalm.*, **71**, 481
- (1913) *Arch. Augenheilk.*, **74**, 180
- VOGT, A. (1913) *v. Graefes Arch. Ophthalm.*, **84**, 293
- (1917) *Klin. Mbl. Augenheilk.*, **58**, 399
- (1921) *Ibid.*, **66**, 718
- WEEKERS, J. F. (1969) *Arch. Ophthalm.*, **29**, 47