BILATERAL ECCENTRIC FIXATION WITH NO OCULAR DEVIATION IN A CASE OF HEREDO-MACULAR DEGENERATION*

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Macular dystrophies are relatively uncommon and their classification is by no means agreed. We present this case therefore, not only as an addition to the literature on macular disturbances, but also as a report on an unusual feature, which does not appear to have been previously reported in cases of this type.

Case Report

A boy aged 10 years was referred to our Outpatient's Clinic in October, 1960, complaining that his visual acuity had begun to deteriorate about 12 months previously; he was now unable to distinguish writing on the school blackboard. His mother had observed over the last 6 months that he held reading matter very close to his face, and on subsequent visits she also reported that he preferred to study in dim illumination and always sought the darkest corner of the room in which to read.

It was not possible to establish with accuracy the exact time of onset of the visual disturbance, but when he had been examined at an ophthalmic school clinic in April, 1959, his visual acuity had been 6/6 in each eye. His general health was good.

Examination.—He was a thin, somewhat apathetic-looking boy of normal intelligence. The visual acuity in the right eye was 6/60, with −0.50 D sph., +0.50 D cyl., axis 90°, and he read N18 at 30 cm. and N5 at 15 cm; in the left eye it was 6/60, with −0.50 D sph., +0.50 D cyl., axis 90°, and he read N18 at 30 cm. and N5 at 15 cm.

The eyes were straight on cover test, with steady fixation. The cranial nerves were intact, and the pupillary reactions normal. The peripheral visual fields were full; it was not possible to demonstrate any scotomata. The ocular media were clear.

Fundus examination showed that the maculae had a slightly atrophic, granular appearance, and a tentative diagnosis of heredo-macular degeneration was made (Fig. 1, overleaf).

The boy was admitted to hospital for further investigation, and a skull x-ray, Wassermann reaction, and Price's precipitation reaction were all negative.

It was noticed on examination with the ophthalmoscope, by focusing the filament on the retina, that he apparently "fixed" above the macula with either eye, and this observation was confirmed by several observers using the visuscope. Individual and independent sketches were made which showed that the eccentric fixation spot was constant, almost identical in position in each eye, and situated about four degrees above the fovea.

When tested on the Co-ordinator (Cüppers, 1956), he reported that he could see Haidinger's brushes with a fair degree of clarity, and that they appeared to be centred on the central fixation spot of the slide, or on the point of a pencil, as required. As this observation was not in accordance with his behaviour on visuscopic examination, it was

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decided to make a record of his fixation by inserting a fixation target into the Zeiss-Nordenson fundus camera. This experimental target was made in the form of an X from black corneal suture silk supported in a metal ring, and it was positioned in the first image plane of the photographic system, so as to appear in simultaneous focus with the fundus image on the film.

The patient's pupils were dilated and he was asked to fix the junction of the cross. The fixation, as viewed through the optical system of the camera, was quite steady and constant in position, thus confirming the findings in the visusscopic examination.

This observation has been repeated on several occasions when fundus photographs have been taken.

Despite the somewhat unsatisfactory nature of the material forming the X, which gives rise to uneven diffraction patterns, it will be seen that the fixation point in each eye is approximately four degrees above the centre of the macula (Fig. 2, opposite).

The boy's colour sense seemed to be anomalous, in that he made a large number of mistakes in reading the Ishihara charts, but these mistakes were not typical of a red-green blindness.

Orthoptic Report

Maddox Rod.—Orthophoric.
Maddox Wing.—Orthophoric.
Cover Test.—Slight exophoria for near; no apparent deviation for distance.

Ocular Movements.—Full.
Convergence.—Normal.

Synoptophore.—Simultaneous macular perception (macular slides) 0° fixing either eye. Peripheral fusion at 0°. Central fusion slides joined at 0°, but central marks not visible. Abducts to −5°. Adducts to +15°. Stereoscopic vision appreciated.

Angle Kappa.—Within normal limits right and left.
Worth's Lights.—Binocular single vision response.

Progress.—A trial of telescopic visual aids produced no improvement in visual acuity at 6 metres.

No noticeable fundus changes have occurred during the past 12 months, neither has there been any definite change in the visual acuity.

Examinations of his parents, a 15-year-old sister, and a 5-year-old brother have revealed nothing abnormal.

Discussion

This patient showed bilateral macular degeneration with bilateral eccentric fixation, with no ocular deviation.

We suggest that this is a case of Stargardt's disease (recessive juvenile heredo-macular degeneration with atrophic reaction—Sorsby, 1951). This diagnosis is supported by the age at onset, the appearance of the maculae, and the normality of the parents.

The presence of fusion and a fusional range account for the absence of any ocular deviation. This fusion is probably peripheral in type, being related
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Fig. 1.—Appearance of fundus, showing an atrophic granular macula in each eye.

Fig. 2.—Fixation points situated approximately 4° above the macula in each eye.

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to a retinal correspondence centred on the true maculae. The retinal orientation may however be abnormal in both eyes, being centred on the pseudo-maculae. We have not been able to separate these two possibilities with any degree of certainty.

The low visual acuity is compatible with the degree of eccentricity of the pseudo-maculae, which also explains the relative hemeralopia.

The upward shift of fixation with no lateral shift accounts for the normal angle kappa in both eyes.

The correct projection of Haidinger’s brushes would appear to indicate that the eccentric fixation is still in the facultative phase in both eyes. It is anticipated that it will become obligatory. As a unilateral phenomenon this is well recognized (Lyle, 1950). Apart from this evidence, we have not been able to demonstrate fixation by the true maculae in this case.

It is hoped that our report on this patient will stimulate further investigation of fixation anomalies in cases of this type.

Summary

Bilateral eccentric fixation with no ocular deviation in a case of heredo-macular degeneration is described.

REFERENCES

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