ECTOPIA OF THE MACULA*

BY

IAN W. PAYNE AND R. PITTS CRICK

Royal Eye Hospital, London

ECTOPIA of the macula is a striking clinical condition but has been infrequently described, only fourteen cases having been reported: Krüger (1913), Triebenstein (1919), Beselin (1924, 1925), Bielschowsky (1930), Stein (1931), Friedman (1942), Cohen and Weisberg (1950), and Trevor-Roper (1952). Fuchs (1921) described a further eight specimens in which the diagnosis was made histologically after perforating injuries. Three patients have attended the Royal Eye hospital recently with the condition.

Case 1, a boy aged 12 years, was 7 weeks premature and his birth weight was $4\frac{3}{4}$ lb.; soon after birth there was a suspicion of "something wrong with his right eye" and at the age of 2 he was thought to have a definite right divergent squint. Glasses were then prescribed (Right -2 D sph.; Left -2.5 D sph.). At the age of 6 years the left eye was occluded, and he thereupon developed a convergent squint. He came under the care of Mr. T. M. Tyrrell at the Royal Eye Hospital in 1951 when he was 8 years old and was found to have an alternating convergent squint with a preference for fixation with the left eye. The following points were noted on examination:

(1) When the left eye was covered, the right fixed in an apparently divergent position. The angle kappa (measured on the perimeter) was $+20^\circ$ in the right eye and between $+1^\circ$ and $+2^\circ$ in the left, with no vertical deviation.

(2) Visual acuity: right eye with -3 D. sph. = 6/12; left eye with -3.25 D sph./, -1 D cyl. at 165° = 6/6.

(3) When the right fundus was examined an exaggerated temporal obliquity of the optic disc was found, and the retinal vessels were displaced so that they emerged from the temporal border; the nasal branches subsequently looped medially above and below. The macula showed no abnormality, though it was less clearly marked than that of the left eye. In the temporal periphery a dark grey area was visible behind which was a zone showing considerable pigmentary disturbance. The left fundus was normal. No relevant family history was obtained.

Examination of the binocular vision revealed simultaneous macular perception with very doubtful fusion, and in view of the unsatisfactory appearance which would have resulted from placing the visual axes parallel and the poor prognosis for binocular vision, it was decided to correct only the apparent squint at operation. The appearance of the right fundus, together with the history of prematurity, is strongly suggestive of an abortive retrolental fibroplasia (Fig. 1, opposite).

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FIG. 1.—Appearance of right fundus, Case 1.

Case 2, a male infant aged 20 months, was one of premature twins with a birth weight of $4\frac{1}{4}$ lb. He was 8 weeks premature and was kept in an oxygen tent for a week. A squint in the right eye had been noticed since the age of 3 months. He was first seen in March, 1954, when the right eye appeared to be densely amblyopic with a variable position, and the left eye which fixed a light well had an apparent divergence of 22° when fixing. A refraction under atropine was carried out and glasses were prescribed: Right—1 D cyl. at 180° ; Left—3 D sph.,—2 D cyl. at 90° .

Examination of the fundi under a general anaesthetic revealed the presence of an incomplete form of retrolental fibroplasia. In the right eye a retinal fold ran from the disc to the temporal periphery with traction of the margin of the disc and the central retina towards the temporal side. The left eye showed a less marked form of the same disturbance, displacement of the functioning macula of this eye being responsible for its apparent divergence. The fundus appearance was similar to that in Case 1.

Case 3, a man aged 34 years, was invalided from the Army in 1944 on account of retinal periphlebitis in both eyes. He attended the Royal Eye hospital in 1948 and at that time his fundi showed evidence of quiescent vasculitis, with degenerative changes in the periphery of the left retina, and an area in the right retina related to the termination of the inferior temporal vessels which showed recent haemorrhages and exudates and varicosities of the veins. A diathermy operation was performed on the right eye in the area of the vasculitis one year later, and the lesion appeared to be well scarred; 3 months later he complained of vertical diplopia and arrangements were made to investigate this, but the patient failed to attend hospital again until July, 1954, when he complained of some difficulty in reading. The following points were noted:

(1) An appearance of marked right hypotropia. The cover test revealed no deviation, however, and the Maddox rod test at 6 m. showed in fact right *hyperphoria* of 4 prism dioptres. This

was confirmed by a Hess screen test, in which there was no significant difference in the size or shape of the right and left fields. Though there was a tendency to right suppression, stereoscopic binocular vision was present.

(2) The corrected visual acuity was 6/9 N.5 in the left eye and 6/12 N.8 in the right. Measurement of the angle kappa with the perimeter showed that the visual axis was 10° above and 8° external to the central pupillary line in the right eye. In the left eye, the angle kappa was $+2^\circ$.

(3) Examination of the fundi showed no change in the left eye since 1949. In the right fundus diathermy scars were visible at the periphery in the lower temporal quadrant, and a series of fine retinal folds suggesting tension lines ran from these scars to the disc. The upper temporal artery ran horizontally outwards from the disc and the macula was displaced downwards by about $1\frac{1}{2}$ disc diameters (Fig. 2).

FIG. 2.—Appearance of right fundus, Case 3, 1954.

(4) The visual fields (4/2000 white) were plotted on the Bjerrum screen. That for the left eye showed no anomaly, but the right blind spot was appreciably displaced downwards and outwards.

(5) The patient was shown the Amsler charts and stated that the horizontal lines above and to the right of the fixation point were inclined upwards and to the right.

(6) He had, incidentally, a marked facial asymmetry, the right eye being apparently displaced about 2 mm. below the left. X rays confirmed that the right lower orbital margin was 3.5 mm. below the left.

There has thus been a gradual downward displacement of the right macula, but the patient has compensated for this almost completely by the development of an apparent right hypotropia, the right hyperphoria representing the uncompensated moiety of the deviation of the visual axis. Thus the macula has suffered a marked displacement 10° down and about 6° outwards, which represents a distance of 2.9 mm. down and 1.7 mm. out, without appreciable interference with its function.

The macular displacement in this case might be ascribed to the contraction of diathermy

scars, but a fundus painting and a Bjerrum screen chart dating from the day before operation are available, and these show that the migration of the macula had already begun before the operation was performed. The blind spot was so placed that its upper border just touched the horizontal meridian. The painting shows that the macula was on the same level as the lower border of the disc, and that a few tension lines in the retina were already visible (Fig. 3).

FIG. 3.—Appearance of right fundus, Case 3, 1949, on the day before diathermy operation.

Discussion

The majority of recorded cases have been attributed to a developmental anomaly at the posterior pole of the eye. This would appear to be the most probable explanation in the two patients recorded by Triebenstein (1919), a mother and son with very similar fundus appearances, and also in one case which he quotes which was associated with familial microcephaly and microphthalmos. Triebenstein also mentions four cases in which ectopia of the macula was associated with pigmented choroidal lesions, colobomata, anomalies in the distribution of the retinal vessels, or bands in the retina or vitreous. These were all apparently congenital and presumably developmental anomalies, though it is often impossible to distinguish between intra-uterine inflammatory lesions and developmental defects. The description of the fundus lesions in some of these cases is reminiscent of the appearances in our Cases 1 and 2, and it is possible that some were due to retrolental fibroplasia. Friedman (1942) reported a case with a scar at the macula which was thought to be the result of a macular haemorrhage during delivery. In the patient reported by Bielschowsky (1930), the displacement of the maculae appeared to be due to the contraction of scars which he regarded

as evidence of old, probably congenital chorio-retinitis. A report by Blaxter (1951) concerned a girl born 3 weeks before term, with one "small eye" in which an upward deviation developed during the first 18 months of life. This was due to traction on the retina resulting from a focus of chorio-retinitis in the upper periphery. Retrolental fibroplasia is again a possibility.

In only three of the recorded cases was the condition known to be acquired. Stein (1931) reported a case in which a simple superior balloon detachment with a hole in the upper retina followed a blow to the eye in a myope. After ignipuncture, the macula was found to be displaced upwards, contraction of the scarred area being the apparent cause. Beselin (1924, 1925) described a 14-year-old myope on whom he operated for divergent strabismus. In the following 6 years he observed the development of a displacement of both maculae towards large choroidal scars in the periphery upwards and inwards. Trevor-Roper (1952) described a patient aged 21 years suffering from pulmonary tuberculosis, who had had a retinal detachment 9 months previously, in whom the macula was found to be drawn downwards by traction bands.

It is perhaps surprising that the macula is so rarely displaced after the reposition of extensive retinal detachments, and it may be that the circumstance which causes such displacement is a widespread inflammatory process involving the retina alone, rather than one restricted in area but extensive in depth, such as follows the surface application of diathermy to the sclera.

Summary

Three cases of ectopia of the macula are described. In two the cause is thought to be an abortive retrolental fibroplasia and the other is due to cicatricial contraction of the retina following retinal periphlebitis. Fourteen cases reported in the literature are summarized and the aetiology of this condition is discussed.

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