HETEROTOPIA OF THE MACULA*

BY

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DISPLACEMENT of the macula may result from defective growth of the posterior segment of the eye (Mann, 1957; Duke-Elder, 1964) and may be associated with other congenital abnormalities.

Derzhavin (1896) described the first recorded case, with a vascularized band extending from the whole width of the optic disc to the periphery of the retina, which at first formed a retinal fold and then broke free from the retina as a band extending forwards into the vitreous. It has been suggested that this represented remnants of the hyaloid artery. Gröndahl (1963) has reported a child with unilateral heterotopia of the macula due to atypical ablatio falciformis congenita. The child described by Woillez, François, and Lebrun (1960) had a vascularized retinal traction band and was probably a further case of atypical falciform congenital fold producing heterotopia of the macula. Triebenstein (1919) ascribed his two cases, in a mother and her son, to congenital causes of a developmental anomaly, though the description of the fundal lesions would suggest that they were post-inflammatory. Cohen and Weisberg (1950) thought that their case of bilateral heterotopia of the macula was due to disturbances in the rate of growth and maturation of the retinal structures.

Post-operative displacement of the macula has been known to follow retinal detachment surgery, as in the case described by Stein (1931).

The majority of cases of macular heterotopia are due to traction bands (Duke-Elder, 1964), which may result from trauma, as in the case described by Friedman (1942), where a macular scar was thought to have followed a retinal haemorrhage during birth.

Incomplete retrolental fibroplasia has been incriminated as the cause in many of the cases recorded in the literature, particularly in two of the cases mentioned by Payne and Pitts-Crick (1956) and most or all of the eight cases reported by Rados and Scholz (1958). Blaxter (1951) described a unilateral case which he ascribed to old chorioretinitis, but the child had been born prematurely and a vertical squint involving the affected eye, which was slightly microphthalmic, had been noted since the age of 18 months. Hugonnier and Royer (1958) reported a further bilateral case due to incomplete retrolental fibroplasia in a child, who at the time of presentation had a uveitis associated with a primary tuberculous complex. The uveitis settled rapidly on steroid and antibiotic therapy and the ectopic maculae, associated with retinal folds due to incomplete retrolental fibroplasia, could then be seen. This case and another of uncertain aetiology were mentioned in greater detail by Michel (1959) in his thesis on ectopia of the macula in which he gives an extensive review of the literature.

The third case reported by Payne and Pitts-Crick (1956) appears to have begun as a result of retinal periphlebitis and subsequently to have been exaggerated by a diathermy operation performed for the periphlebitis.

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Post-inflammatory displacement of the macula has accounted for several cases (Biel-schowsky, 1930; Foulds, 1956). Beselin (1924) described a myope with a divergent squint for which he had had surgery, whose maculae were both displaced towards superior nasal choroidal scars, which were probably of congenital inflammatory origin. Nauheim (1960) reported six cases of varied aetiology, including incomplete retrolental fibroplasia, chorio-retinitis, and angioma.

Trevor-Roper (1952) described a case in which displacement of the macula followed the spontaneous resolution of a retinal detachment in a patient suffering from pulmonary tuberculosis. Vitreous bands associated with a probable small localized retinal detachment were present in the case reported by Horwich and Linfesty (1956).

The case to be described presented a fundal lesion somewhat resembling the end-result of infestation of the eye by *Toxocara* larva.

Case Report

A 28-year-old male was first seen at the R.A.F. Hospital, Ely, in May, 1964, complaining of tiredness of the eyes and vertical double vision for the past 9 to 10 years. The symptoms were most noticeable on performing close work and he had been prescribed reading glasses in the past but had had no other therapy. His general health was good and he had never suffered any ocular injury or disease. He had not been born prematurely. There was no significant family history.

Examination.—The visual acuity was 6/18 improving to 6/5 with +1 D sph., +1 D cyl., axis 15° in the right eye, and 6/5 improving to 6/4 with +0.5 D cyl., axis 180° in the left.

There was an apparent right hypertropia with slight right divergent squint (Fig. 1), but cover testing revealed an actual right hypotropia. The ocular movements and convergence appeared to be full. An apparent vertical eccentric fixation of the right eye, noted during the cover test, was shown to be due to a large vertical angle kappa when the visuscope revealed foveal fixation in both eyes.

FIG. 1.—Apparent right hypertropia.

FIG. 2.—Right fundus, showing glial tissue mass and finer band with displaced macula and inferior temporal vessels.

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against any true achievement of symptomatic improvement. He was therefore advised not to wear glasses and to accept his diplopia.

Summary

Case reports of heterotopia of the macula are reviewed and a further case of post-inflammatory vertical displacement of the macula and inferior temporal vessels is described. The similarity of the associated fundal lesion to certain cases of *Toxocara* larval infestation of the eye is pointed out, though confirmation of the diagnosis was not obtained.

I wish to thank the Director General of Medical Services, R.A.F., for permission to publish this case. I am indebted to Prof. W. S. Foulds for the interest he took in the case and his suggestions as to the aetiology. I thank Prof. A. W. Woodruff for kindly supplying the *Toxocara canis* antigen, Mrs. R. Graver for the orthoptic report, the Medical Illustration Dept. of the Institute of Ophthalmology for the fundus painting, and the Clinical Photography Dept., R.A.F. Hospital, Ely, for the photograph. I am grateful for Mr. K. Rubinstein's considerable help in the translation of the German language literature.

REFERENCES

- ASHTON, N. (1960). *Brit. J. Ophthalm.*, **44**, 129.
 BESELIN, O. (1924). *Klin. Mbl. Augenheilk.*, **73**, 691.
 BIELSCHOWSKY, A. (1930). *Ibid.*, **84**, 755.
 BLAXTER, P. L. (1951). *Trans. ophthalm. Soc. U.K.*, **71**, 231.
 COHEN, I. J., and WEISBERG, H. K. (1950). *Arch. Ophthalm. (Chicago)*, **44**, 419.
 DERZHAVIN, V. (1896). *Vestn. Oftal.*, **13**, 22 (*Jber. Leist. Ophthalm.*, **27**, 244).
 DUGUID, I. M. (1961a). *Brit. J. Ophthalm.*, **45**, 705.
 ——— (1961b). *Ibid.*, **45**, 789.
 DUKE-ELDER, S. (1964). "System of Ophthalmology", vol. 3, Part 2, p. 654. Kimpton, London.
 FOULDS, W. S. (1956). *Brit. J. Ophthalm.*, **40**, 571.
 FRIEDMAN, B. (1942). *Arch. Ophthalm. (Chicago)*, **28**, 444.
 GRÖNDAHL, J. (1963). *Acta ophthalm. (Kbh.)*, **41**, 259.
 HORWICH, H., and LINFESTY, J. W. (1956). *A.M.A. Arch. Ophthalm.*, **56**, 194.
 HUGONNIER, R., and ROYER, J. (1958). *Bull. Soc. Ophtal. Fr.*, p. 583.
 MANN, I. (1957). "Developmental Abnormalities of the Eye", 2nd ed., p. 143. B.M.A., London.
 MICHEL, J. (1959). "Contribution à l'étude des ectopies de la macule". Thèse Lyon.
 NAUHEIM, J. S. (1960). *A.M.A. Arch. Ophthalm.*, **63**, 144.
 PAYNE, I. W., and PITTS-CRICK, R. (1956). *Brit. J. Ophthalm.*, **40**, 492.
 RADOS, W. T., and SCHOLZ, R. O. (1958). *Amer. J. Ophthalm.*, **45**, 683.
 STEIN, R. (1931). *Klin. Mbl. Augenheilk.*, **86**, 188.
 TREVOR-ROPER, P. D. (1952). *Trans. ophthalm. Soc. U.K.*, **72**, 671.
 TRIEBENSTEIN, O. (1919). *Klin. Mbl. Augenheilk.*, **62**, 442.
 WOILLEZ, M., FRANÇOIS, P., and LEBRUN, H. (1960). *Bull. Soc. Ophtal. Fr.*, p. 383.
 WOODRUFF, A. W., THACKER, C. K., and SHAH, A. I. (1964). *Brit. med. J.*, **1**, 1001.