

COMMUNICATIONS

PATHOLOGY OF POSTERIOR POLYMORPHOUS DEGENERATION OF THE CORNEA*†

BY

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POSTERIOR polymorphous degeneration of the cornea is an uncommon, non-progressive, hereditary condition which is usually bilateral; in the majority of cases there is only minimal visual impairment (Duke-Elder and Leigh, 1965). The condition was first described by Koepe (1916) as "keratitis bullosa interna", on account of the congenital pits on the posterior corneal surface. Koepe (1920) reported eighteen additional cases which he considered to be of congenital origin because of the absence of other ocular changes. Further cases have been reported by Triebenstein (1925), Freudenthal (1932), and Theodore (1939). Franceschetti, Klein, Forni, and Babel (1951) described the condition as showing "slight concavities, vesicles, and polymorphous opacities in the posterior limiting membrane". Its hereditary nature as a dominant trait is now well recognized (Freudenthal, 1932, in a father and son; Theodore, 1939, in a grand-mother, father, and daughter; McGee and Falls, 1953, in a father and son; Soukup, 1964, in two sisters). Franceschetti and Montresor (1960) described the condition in association with band-shaped degeneration of the cornea, and Bergman (1964) reported a case in which there was heterochromia of the brows and lashes.

The purpose of this paper is to describe for the first time the pathological changes of the condition.

Case Report

A man aged 46 years had had gradual deterioration of vision over a period of 9 years, which had been rapid for about the last year; 3 months before admission to hospital a dendritic ulcer of the right cornea had been treated successfully by I.D.U. drops. The younger of the patient's two daughters was myopic, but neither had any other visual complaints. Both his parents were dead and as far as he knew neither they nor his two brothers had suffered from any ocular disease.

Examination.—The visual acuity was 6/36 in the right eye and 6/60 in the left with glasses. Both corneae revealed multiple greyish opacities of various shapes and sizes at the level of Descemet's membrane. They were confined to the central portion of the cornea over an area of about 7 mm. in diameter; the periphery of the cornea was clear (Fig. 1, overleaf). Apart from an anterior stromal scar due to the dendritic ulcer in the right cornea, the remainder of both corneae and the other ocular structures were normal.

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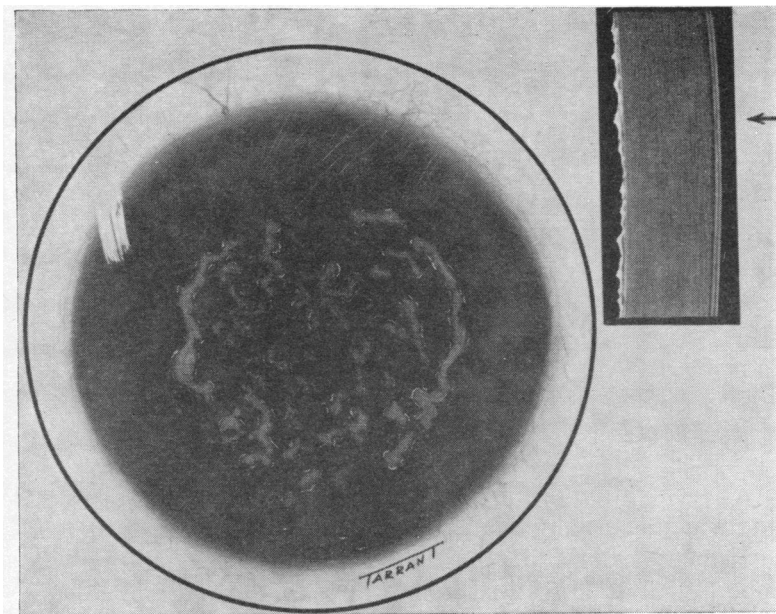
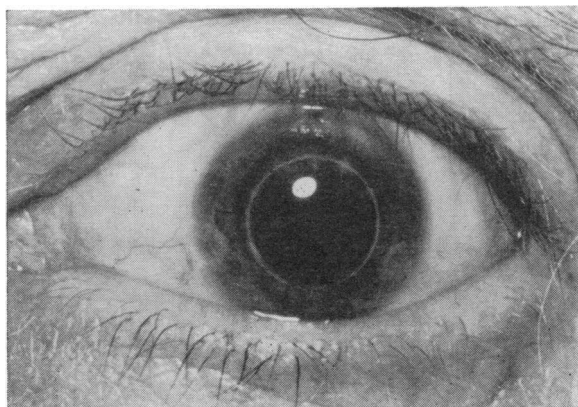


FIG. 1.—Left cornea, showing polymorphous opacities in Descemet's membrane.

Treatment.—A 6 mm. perforating keratoplasty was performed by Mr. A. Lister on the left eye, and the graft was retained with direct sutures (Fig. 2). The post-operative course was uneventful, and at the time of writing, one year later, the visual acuity in the left eye is 6/6, N.5. with glasses.

FIG. 2.—Post-operative photograph of left eye taken one year after keratoplasty.



Pathological Findings

The corneal disc was fixed in 10 per cent. formol saline and sections were stained with haematoxylin and eosin, periodic acid-Schiff (PAS), and Alcian blue. The pathological changes were confined to Descemet's membrane and the endothelium, while the epithelium, Bowman's membrane, and the stroma were normal. Approximately four excrescences were seen on Descemet's membrane in each section (Fig. 3, opposite). These were fusiform in shape, bulged into the anterior chamber, and were PAS-positive; each swelling showed, at a maximum, two dozen irregular empty vacuoles (Fig. 4, opposite), and Descemet's membrane between the excrescences was normal in some places and thickened in others. The endothelium was thinned over the excrescences with flattening of its nuclei (Fig. 5, opposite).

FIG. 3.—Fusiform swellings of Descemet's membrane. Haematoxylin and eosin. $\times 75$.

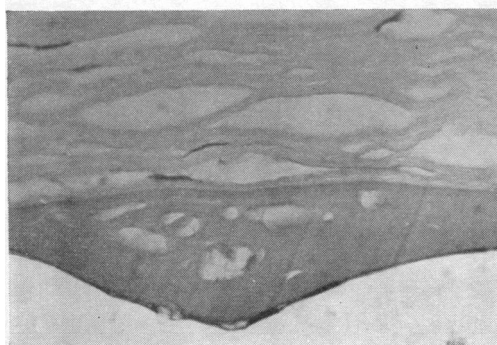
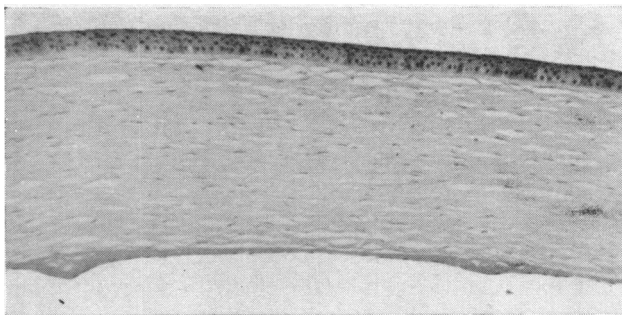


FIG. 4.—Irregular vacuoles in Descemet's membrane. Haematoxylin and eosin. $\times 400$.

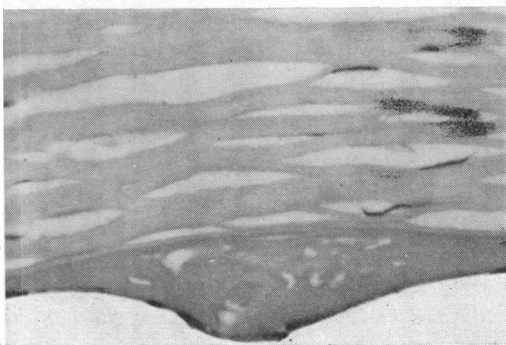


FIG. 5.—Thinning of the endothelium over an excrescence in Descemet's membrane. Haematoxylin and eosin. $\times 400$.

Differential Diagnosis.—Guttate swellings of Descemet's membrane are seen in the eyes of older persons where they are situated peripherally. Centrally-placed swellings may be seen in Fuchs's corneal dystrophy and in some cases of macular dystrophy. There was no clinical evidence of any of these three conditions in the case described. Histologically, the swellings in this case were fusiform in shape and showed numerous vacuoles. On the other hand, the guttate swellings of Descemet's membrane in Fuchs's dystrophy are hemispherical or flat-topped, and besides being swollen, show no vacuolation. Moreover, the stromal and epithelial oedema of Fuchs's dystrophy was absent. It was clearly not macular dystrophy of the cornea because of the absence of the typical accumulations of acid mucopolysaccharide in the corneal stroma, the corneal fibroblasts and the endothelium.

Discussion

As we have already indicated, the pathological changes in posterior polymorphous degeneration of the cornea are confined to Descemet's membrane. The endothelium in the case described appeared to be normal except for some thinning over the excrescences, but too much reliance cannot be placed on the microscopical appearances of formalin-fixed endothelium.

It is now generally agreed that in the normal cornea Descemet's membrane is derived in some way from the endothelium, to which its relationship is that of a basement membrane (Chi, Teng, and Katzin, 1958). The formation of the swellings on Descemet's membrane in posterior polymorphous degeneration appears to be different from that in Fuchs's endothelial dystrophy. Nevertheless, although guttate swellings of Descemet's membrane are not a constant finding in Fuchs's dystrophy, the

membrane is always markedly thickened, so that in both conditions it may be supposed that the endothelium is primarily involved.

Chi, Teng, and Katzin (1962) reported the presence of cytoplasmic bodies in the endothelial cells in Fuchs's dystrophy; according to them these bodies increase in size and coalesce with Descemet's membrane to form nodular excrescences. Posteriorly, the cytoplasm of the endothelial cells thins over the top of the swellings and the nucleus is pushed aside. Eventually the endothelial cell bursts. Some doubt about this hypothetical coalescence between the cytoplasmic bodies and Descemet's membrane has been thrown by the electron microscopical studies of Kayes and Holmberg (1964), who suggested that the cytoplasmic bodies could be accounted for by the projections being cut off during sectioning, giving the impression of an intracellular inclusion on light microscopy. They found vacuoles of different sizes in the endothelium in Fuchs's dystrophy, the larger being lined by cell membrane were probably intercellular, whereas the smaller vacuoles had no such lining and were probably intracellular. Occasional vacuoles were in contact with Descemet's membrane, and others, which occasionally broke down, with the anterior chamber. These changes in the endothelium lead to the laying down of thickenings of Descemet's membrane and to a disturbance of normal corneal physiology, resulting in oedema of the corneal stroma and epithelium. Oedema, however, is not a feature of posterior polymorphous degeneration and it is therefore reasonable to assume that in this condition the severe and widespread endothelial damage of Fuchs's dystrophy does not occur. It is much more likely that the Descemet swellings are formed in the same way as the peripheral guttate swellings of the ageing cornea (Hassall-Henle warts), which are also unassociated with oedema of the cornea, and appear to be due to the excessive deposition of material by the endothelial cells (Duke-Elder and Leigh, 1965), similar to the formation of nodules on the lens capsule and Bruch's membrane (Leber, 1879; Salzmann, 1912). The vacuoles seen in the fusiform swellings in posterior polymorphous degeneration are probably due to the uneven laying down of material for Descemet's membrane by the endothelial cells, and it is interesting to note that similar but much smaller fissures and channels have been demonstrated in Hassall-Henle warts by the electron microscopical studies of Feeney and Garron (1961).

Summary

The pathological changes in a corneal disc from a case of bilateral posterior polymorphous degeneration of the cornea are described for the first time. The visual impairment on the left side was treated successfully by corneal grafting, the visual acuity being 6/6 one year after operation. The historical background of the condition has been recorded. The lesions on Descemet's membrane are similar to the Hassall-Henle warts seen in ageing and to the guttate swellings of Fuchs's dystrophy. Evidence has been put forward to support the likelihood that their formation is similar to those of Hassall-Henle warts.

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REFERENCES

- BERGMAN, G. D. (1964). *Amer. J. Ophthalm.*, **58**, 125.
- CHI, H. H., TENG, C. C., and KATZIN, H. M. (1958). *Ibid.*, **45**, 518.
- , ———, ——— (1962). *Ibid.*, **53**, 215.
- DUKE-ELDER, S., and LEIGH, A. G. (1965). "System of Ophthalmology", vol. 8, pt 2, pp. 873, 955. Kimpton, London.
- FEENEY, M. L., and GARRON, L. K. (1961). In "The Structure of the Eye", ed. G. K. Smelser, p. 372. Academic Press, New York and London.
- FRANCESCHETTI, A., KLEIN, D., FORNI, S., and BABEL, J. (1951). "XVI Concilium Ophthalmologicum, 1950, Britannia Acta", vol. 1, p. 157. B.M.A., London.
- and MONTRESOR, D. (1960). *Boll. Oculist.*, **39**, 785.
- FREUDENTHAL, E. (1932). *Z. Augenheilk.*, **78**, 224.
- KAYES, J., and HOLMBERG, A. (1964). *Invest. Ophthalm.*, **3**, 47.
- KOEPPE, L. (1916). *v. Graefes Arch. Ophthalm.*, **91**, 375.
- (1920). "Die Mikroskopie des lebenden Auges", vol. 1. Springer, Berlin.
- LEBER, T. (1879). *v. Graefes Arch. Ophthalm.*, **25**, (1), 257.
- MCGEE, H. B., and FALLS, H. F. (1953). *A.M.A. Arch. Ophthalm.*, **50**, 462.
- SALZMANN, M. (1912). "Anatomie und Histologie des menschlichen Augapfels". Deuticke, Leipzig.
- SOUKUP, F. (1964). *Čsl. Ofthal.*, **20**, 181.
- THEODORE, F. H. (1939). *Arch. Ophthalm. (Chicago)*, **21**, 626.
- TRIEBENSTEIN, O. (1925). *Klin. Mbl. Augenheilk.*, **74**, 777.