Pigment dispersion syndrome and pigmented pattern dystrophy of retinal pigment epithelium

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SUMMARY The 2 rare entities, pigmentary dispersion syndrome and pigmented pattern dystrophy of the retinal pigment epithelium, were found in a young male patient. Visual function was undisturbed.

Pigmentary dispersion syndrome is a rare entity found mainly in young myopic men. It is characterised by Krukenberg's spindles on the posterior corneal surface, loss of the pigment epithelial layer of the peripheral iris in a radial slit-like pattern, and deposition of pigment in the trabecular meshwork and peripheral lens surface and the anterior surface of the iris. In some cases of pigmentary dispersion glaucoma may occur. Since the original publication¹ the characteristics of the disease have been well described. This paper is a case report of a patient with pigmentary dispersion syndrome presenting in

addition with a pigmented pattern dystrophy of the retinal pigment epithelium. We believe this has not been previously reported.

Case report

A 34-year-old male presented with complaints of slight dimming of vision in both eyes for 4 weeks. No complaints of 'haloes' were made. His general health was excellent. On examination his visual acuity was 6/6 right eye and 6/9+ left eye (with plano right eye, -0.50 sphere left eye). Slit-lamp examination revealed bilateral Krukenberg's spindles (Fig. 1) with iris atrophy peripherally in a radial slit-like pattern (Fig. 2). Goldmann applanation pressures were 26

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Fig. 1 Krukenberg's spindle of the cornea.

Fig. 2 Radial iris atrophy.

Fig. 3 Gonioscopy reveals heavy pigmentation of the trabecular meshwork.

mmHg bilaterally. Gonioscopy disclosed bilateral grade 4 open angles with heavy pigmentation of the trabecular meshwork (Fig. 3). Funduscopy showed minimal cupping with healthy optic nerve heads bilaterally. Coarse pigmentary changes of no particular pattern were seen in the maculae of both eyes, with a decrease in the foveal reflexes (Fig. 4). Examination with the Goldmann 3-mirror lens showed the deep location of the pigments in the

Fig. 4B Pigmentary changes of the retinal pigment epithelium of the left macula.

retinal pigment epithelial layer. The retinal periphery and the retinal vessels were normal.

On fluorescein angiography a readily distinguishable reticular pattern in the form of a fish-net was seen in both maculae symmetrically (Fig. 5). The pattern on the angiogram was more distinct than the colour fundus photographs. The polygons formed by the reticular pattern measured less than 1 disc diameter. The pattern blocked fluorescence, with no

Fig. 4A Pigmentary changes of the retinal pigment epithelium of the right macula.

Fig. 5A Fluorescein angiogram of the reticular pattern of the pigmentary dystrophy in the right macula.

Fig. 5B Fluorescein angiogram of the reticular pattern of the pigmentary dystrophy in the left eye.

evidence of leakage or hyperfluorescence. The retinal periphery was again normal.

Visual fields on the Goldmann perimeter were normal. Colour vision, extensively tested with TMC (Tokyo Medical College), H-R-R, Ishihara (1970 ed.), Lanthony New Colour Test Box 6/4, Lanthony's desaturated D-15, Farnsworth-Munsell 100 hue test, and anomaloscope was normal. Electro-oculography was normal (1.87 right eye and 2.24 left eye.) The electroretinogram was normal.

Two subsequent intraocular pressure measurements were less than 20 mmHg bilaterally. The patient was not given antiglaucoma therapy. His 9 sibs and 2 young children also underwent ocular examination and were found to be entirely normal. His parents were not available for examination.

Discussion

This young man presented with the typical findings of pigmentary dispersion syndrome with ocular hypertension. The glaucoma which often accompanies this syndrome has been thought to be caused by the pigmentary obstruction of and damage to the trabecular meshwork. The cause of the loss of pigment from the iris is obscure. Theories have ranged from congenital atrophy or degeneration of the iris²⁻⁴ to actual mechanical rubbing between the peripheral concave iris and the zonules.⁵

Pigmentary dispersion syndrome is rare. Pigmented pattern dystrophy of the retinal pigment epithelium is also rare; it usually occurs in healthy

persons without ocular or systemic diseases. The only exception is the occurrence of pigmented pattern dystrophy of the retinal pigment epithelium in patients with myotonia dystrophia.^{6,7} Pigmented pattern dystrophy of the retinal pigment epithelium may be sporadic.^{8,9} Of the cases which are familial we consider there are 2 major categories, based on the hereditary patterns: autosomal dominant¹⁰⁻¹⁵ and autosomal recessive.^{16,17}

It may be a coincidence that this patient presented with 2 different pigmentary diseases. Embryologically, however, the pigmented cells of the iris are very similar to those of the retinal pigment epithelium. Both tissues are derived from the neural crests and both are formed of round clump cells with rod-shaped pigments concentrated near the nucleus.¹⁸ Could this patient represent a congenital or degenerative disorder of the generalised pigmented cells of the eye? The rarity of these disorders suggests their conjunction may be more than chance coincidence.

Mr A. L. Aan de Kerk made the photographs.

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