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 publication 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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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 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
Discussion

The retinal pigment epithelium is an important component of the retina. Its function includes the phagocytosis of photoreceptor outer segments, the production of vitamin D3, and the maintenance of the photoreceptor layer. Dystrophy of the retinal pigment epithelium can lead to various visual impairments, including macular degeneration, tapetoretinal dystrophy, and pigmentary retinopathy.

In its congenital form, this condition is usually inherited as an autosomal recessive trait. However, in cases where the condition occurs sporadically, it may be due to a de novo mutation. The symptoms can range from mild to severe, and the onset may be either early or late in life. The treatment options for dystrophy of the retinal pigment epithelium are limited and depend on the severity of the condition.

It is essential to diagnose and manage these conditions early to prevent further damage to the retina and to improve the quality of life of affected individuals. Further research is needed to develop more effective therapies for these conditions.
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