

Photo Essay

Pigmentary Glaucoma with Retinochoroidal Pigmentation

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PRESENTATION

Pigmentary glaucoma (PG) is considered as an entity within the spectrum of secondary open angle glaucomas. The condition usually starts in the third or fourth decades of life with disruption of the iris pigment epithelium. The released pigment granules deposit on anterior segment structures such as the corneal endothelium, trabecular meshwork and lens/zonules. This stage is known as primary pigment dispersion syndrome (PPDS). Genetic studies have shown it to be an autosomal dominant disorder with incomplete penetrance. A susceptibility locus for PPDS has also been identified on chromosome 7q35-q36.^[1,2] Over a period of time, intraocular pressure (IOP) may increase due to progressive trabecular meshwork dysfunction, which may manifest as ocular hypertension or lead to PG. The probability of PPDS converting to PG is reported to be 10% at 5 years and 15% after 15 years of diagnosis.^[3] Sivaraman et al have cited the probability of progression from PPDS to PG at 10-50%.^[4]

Significant diagnostic signs for PPDS/PG are limited to the anterior segment. The classic triad of PPDS is the presence of Krukenberg spindles (KS), transillumination

iris defects (TID), and trabecular pigmentation.^[2,5-8] Qing et al reported the clinical characteristics of PG in Chinese patients and stated that TID is rare in that ethnic group, a finding which is corroborated by our case report.^[9]

A 54-year-old gentleman of Chinese origin was first seen at the ophthalmology clinic of Queen Elizabeth Hospital, Malaysia for painless, progressive blurring of vision in both eyes since a few months ago. There was no history of red eye, trauma, itching or any medication use. Medical and surgical history was unremarkable and no significant family history could be elicited.

On examination, uncorrected visual acuity was 6/9 in both eyes; corrected to 6/6 with -1.00 D sphere in the right eye and -1.50 D sphere in the left eye. In both eyes, the cornea had prominent KS [Figure 1a and b] and extensive pigment deposition on the anterior and posterior lens capsules [Figure 2a and b]. Fundus examination revealed cup/disc ratio of 0.8 in right eye and 0.9 in left eye [Figures 3 and 4]. Bilaterally, the retinal periphery showed extensive areas of pigmentary changes [Figures 5 and 6]. IOP was 30 mmHg in the right eye and 34 mmHg in the left eye. Gonioscopic examination showed open angles with extensive pigment deposition in both eyes [Figures 7 and 8]. Optical coherence tomography of the retinal nerve fiber layer is shown in Figure 9. A diagnosis of bilateral PG was entertained, and the patient was started on Timolol 0.5% twice daily in both eyes. One week later, IOP was 16mmHg in both eyes and he was advised to continue Timolol 0.5% twice daily in both eyes.

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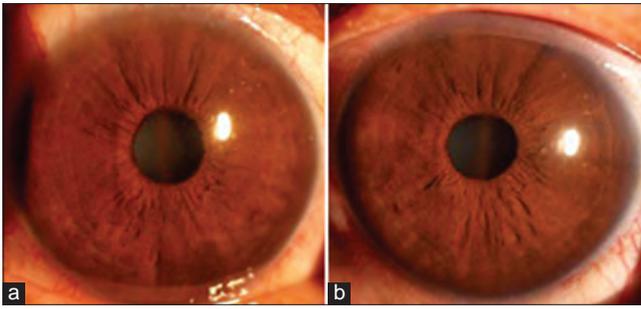


Figure 1. (a and b): Slit lamp images of the patient demonstrate Krukenberg spindles in right (a) and left (b) eyes.

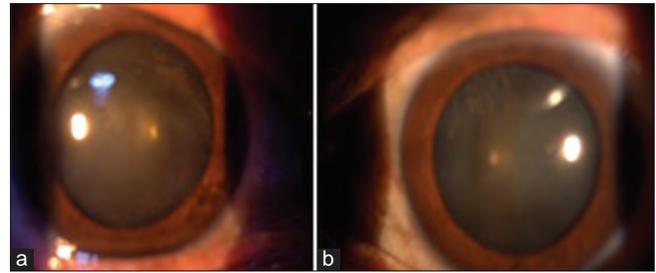


Figure 2. (a and b): Slit lamp image shows pigment deposition on the lens capsules in both eyes.



Figure 3. Fundus photograph of the right eye, note the glaucomatous cupping.



Figure 4. Fundus photograph of the left eye.

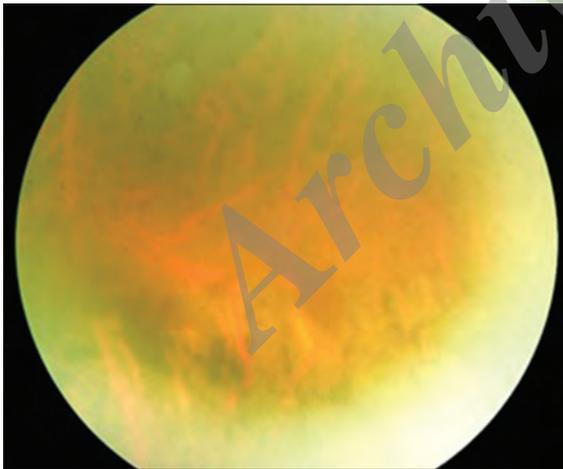


Figure 5. Retinal pigmentation in the periphery of the right eye.

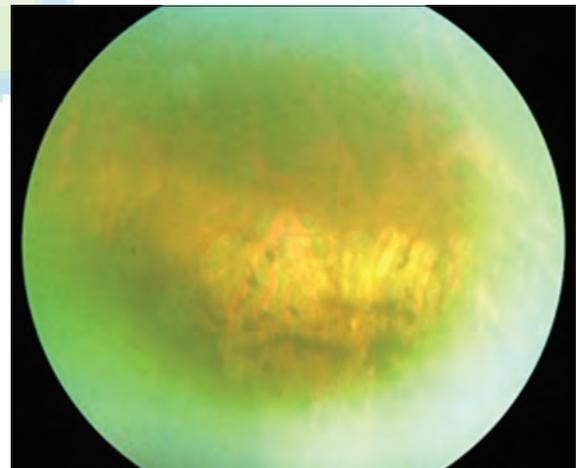


Figure 6. Retinal pigmentation in the periphery of the left eye.

DISCUSSION

Pigment dispersion in the eye could be part of the natural process of aging. The infant eye, especially the trabecular meshwork, is non-pigmented. However, with aging, various grades of pigmentation might become visible.^[10] Anterior segment pigment deposition in PPDS and PG has been widely reported in the literature, but pigment

deposition in the retinal periphery has not been reported so far.

In most cases, pigment dispersion is innocuous and does not cause any pathological changes in the eye. This condition, known as PPDS, is characterized by pigment deposition throughout the anterior segment. This can manifest as a KS, which is a vertically oriented line of pigment deposits on the corneal endothelium secondary to aqueous convection currents in the nasal and temporal halves of the anterior chamber.^[2,11,12] KS is more common

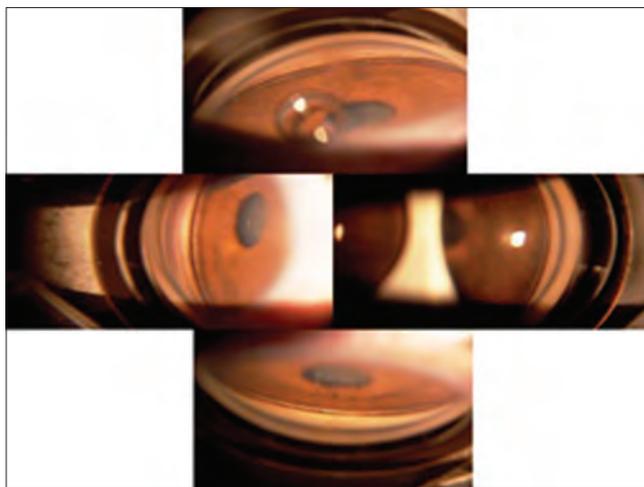


Figure 7. Gonioscopic appearance of the right eye, note the angle hyperpigmentation.

in females, suggesting a hormonal influence,^[13] and the presence of KS is associated with higher rate of conversion to glaucoma.^[6] Our patient had extensive pigment deposition on the corneal endothelium in the form of KS, on the anterior and posterior lens capsules, and in the angles bilaterally.

Pigment can also be deposited on the anterior iris surface, lens zonules and the anterior or posterior lens capsules. Accumulation of pigment may occur at the junction of zonules and posterior lens capsule, or medial to Wieger's ligament, which are called Zentmayer Ring and Scheie stripe, respectively.^[14-16]

Angle pigmentation is seen circumferentially, homogeneously or irregularly, and is more prominent in the inferior trabecular meshwork due to gravity. The pigment can be deposited at Schwalbe's line, producing a thin dark band resembling Sampaolesi's line seen in pseudoexfoliation syndrome. In asymmetric cases, glaucoma is found to be more severe in the eye with a greater degree of pigment dispersion and trabecular pigmentation.^[14,16]

Posterior segment changes reported in PPDS include lattice degeneration, retinal breaks, and retinal detachment. Scheie reported retinal detachment in 6.4% of patients diagnosed with PPDS; while, in Weseley's study, 20% of patients with PPDS had lattice degeneration, 11.7% had full thickness retinal breaks, and 3.3% had rhegmatogenous retinal detachments which required scleral buckling.^[7,17] Scuderi et al also reported retinoschisis, lattice degeneration and retinal detachment as the only retinal changes in PPDS.^[18] Similarly, Lehto and Vesti has reported lattice degeneration in 33.3% eyes with PPDS.^[19]

The peripheral pigmentary changes in our patient are unusual but quite plausible. It may be conjectured that the pigmentation seen in the posterior segment are in fact pigment granules which might have traveled by convection currents to those sites due to a more fluid

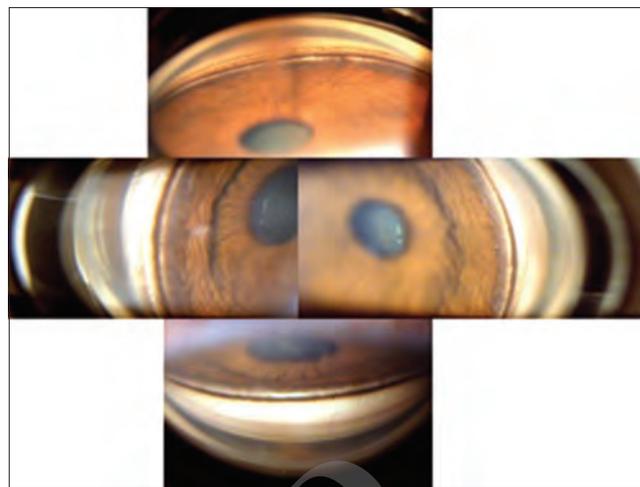


Figure 8. Gonioscopic appearance of the left eye.

vitreous in these myopic patients. Thus, this pigment may have deposited on the retina similar to the pigment deposition seen on the cornea, angles, and lens capsules. In another case report, Nagarajah and Shun-Shin stated that the presence of Scheie's line in PPDS demonstrates some flow of pigment from the anterior to the posterior chamber.^[20]

Chew and Deutman presented a case of PPDS associated with pigmented pattern dystrophy of the retinal pigment epithelium (RPE) at the macula. Such involvement of the iris and RPE has been explained on the basis of their common embryologic origin from the neural crest. The condition was assumed to represent a generalized congenital or degenerative disorder of pigment cells in the eye.^[21] Piccolino et al have reported two brothers diagnosed with PG and widespread salt-and-pepper RPE mottling and pigment clumping. According to the authors, an inherited defect in the pigment epithelium could be responsible for the changes seen in both the anterior and posterior segments.^[22]

Scuderi et al and also Greenstein et al have studied electrooculograms from patients with PPDS and PG and found mean Arden ratios for this group to be significantly lower than normal controls as well as patients with ocular hypertension or primary open angle glaucoma. These studies seem to support the hypothesis that integrity of the RPE/photoreceptor complex is affected in patients with PPDS/PG.^[23,24]

Management of PG is similar to other primary open angle glaucomas. Miotics may reduce irido-zonular contact and facilitate aqueous outflow; however, they may exacerbate myopia. Laser trabeculoplasty, laser iridotomy, and trabeculectomy might be performed as required. The prognosis for such patients is usually good with a reduction of IOP either with medical treatment or spontaneously over time.

In summary, although pigmentation of the anterior segment in PG has widely been reported, we are

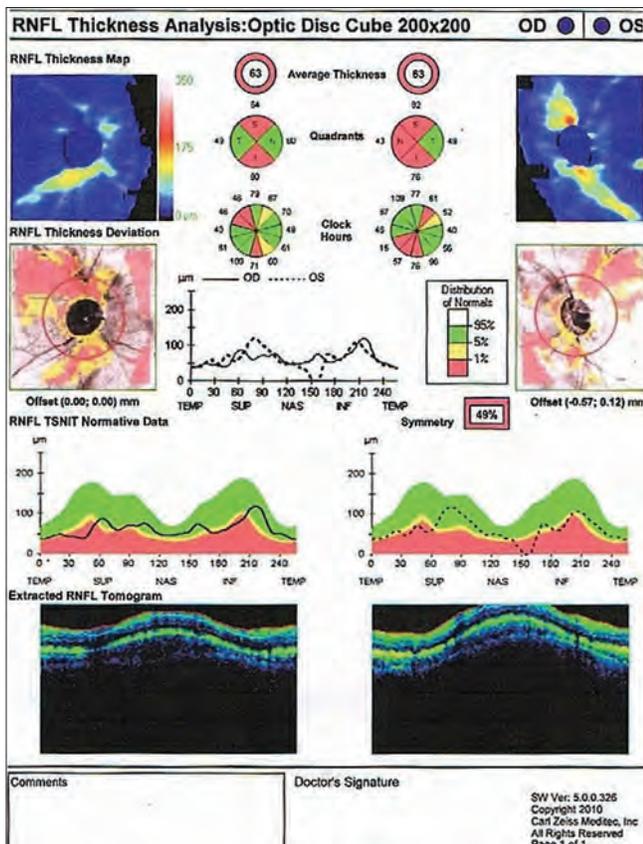


Figure 9. Retinal nerve fiber layer thickness, determined by optical coherence tomography.

unaware of any report documenting pigmentation of the peripheral retina. This could be due to movement of iris pigment granules to the posterior segment or a primary disorder of the retinal pigmentary epithelium.

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Conflicts of Interest

There are no conflicts of interest.

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