Case Report:

Pigment dispersion syndrome

C.S. Sandhya, D. Murali Krishna, G. Vijay Bhaskar

Department of Ophthalmology, Sri Venkateswara Medical College, Tirupati

ABSTRACT

We report of the rare occurrence of pigment dispersion syndrome (PDS) with posterior subcapsular cataract in both eyes in a young male patient. The patient presented with complaints of progressive decrease in vision of one year duration. The patient also had high myopia with mild iridodonesis, phacodonesis and anterior insertion of zonules. Classical signs of PDS like Krukenberg's spindle on the posterior corneal surface were evident on slit lamp examination; transillumination defects in the iris could not be elicited by retroillumination as the iris was heavily pigmented. Gonioscopy revealed heavy and uniform pigmentation of trabecular meshwork. Evidence of a characteristic iris configuration on optical coherence tomography (OCT), namely, posterior bowing of iris in the mid periphery suggested the diagnosis of PDS. This case highlights the importance of OCT in identifying the iris configuration characteristically seen in PDS even in the absence of transillumination defects in the iris and reiterates the need to look for subtle signs like phacodonesis which are important when surgical intervention is planned.

Key words: *Pigment dispersion, Optical coherence tomography, Central corneal thickness, Krukenberg's spindle, Transillumination defects*

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INTRODUCTION

Pigment dispersion syndrome (PDS) is a unique and fascinating entity characterized by disruption of iris pigment epithelium and deposition of the dispersed pigment granules throughout the anterior segment.[1] The classic diagnostic triad consists of corneal pigmentation (Krukenberg's Spindle), slit like radial, mid-peripheral iris transillumination defects and dense trabecular pigmentation.[2-4] The iris insertion is typically posterior and the peripheral iris tends to have a concave configuration. We report a case of pigment dispersion syndrome with posterior subcapsular cataract in both eyes in a young male patient. This case highlights the importance of optical coherence tomography(OCT) in identifying the iris configuration characteristically seen in pigment dispersion syndrome even in the absence of transillumination defects in the iris.

CASE REPORT

A 35-year-old male presented to ophthalmology out-patient service with complaints of progressive decrease in vision of one year duration. The patient was otherwise asymptomatic; there was no history of ocular trauma or co-morbid conditions like diabetes mellitus. Ocular examination revealed similar findings in both the eyes (Table 1).

Table 1: Ocular parameters

Feature	Right eye	Left eye
Central corneal thickness (μ)	574	575
Keratometry	K1: 39D at 24º	K1:38D at 101º
	K2:40.25D at 114 º	K2: 39.50D at 11º
Anterior chamber depth (mm)	4.09	4.17
Axial length (mm)	27.02	26.74

CCT=Central corneal thickness in microns; K1=keratometry readings vertical axis in dioptres
K2=keratometry readings horizontal axis in dioptres

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Corresponding author: Dr C.S. Sandhya, Professor and Head, Department of Ophthalmology, Sri Venkateswara Medical College, Tirupati, India. **e-mail:** ophthalmologysvmc@gmail.com

Cornea showed Krukenberg's spindle in the central part (Figure 1). Keratometry indicated a flat cornea; the anterior chamber was deep. Iris was darker in colour with mild iridodonesis. Ectropion of uveal pigment was evident and transilumination defects were absent. Gonioscopy (Figure 2) revealed a wide open angle with increased trabecular pigmentation, posterior insertion of iris and posterior concavity of iris .

The pupil was mid-dilated and sluggishly reacting to light. Posterior subcapsular cataract (Figure 3) was present in both eyes, being more in left eye. There was deposition of uveal pigment on posterior lens surface in the periphery. On mydriatic administration, the zonules were found to be inserted more anteriorly on the lens surface (Figure 4).

Funduscopic examination was hampered by media being hazy due to posterior subcapsular cataract. Optic disc showed cup/disc ratio of 0.50:1. Neuroretinal rim was healthy. Peripapillary halo was seen. Foveal reflex was good. Peripheral retina was healthy on indirect ophthalmoscopy.

On applanation tonometry, intraocular pressure (IOP) was noted to be 10 and 20 mm of Hg in the right and left eyes respectively. Best corrected visual acuity was 6/18 in right eye and 1/60 in left eye. The intraocular power was 12D and 14.5 D suggestive of increased axial length. Optical coherence tomography (OCT) (RTVue, Model no: RT100, Software version 6.1.0.4, Optovue Inc, Fermont, CA, USA) revealed the characteristic findings of posterior bowing of iris in the mid periphery (Figures 5A and 5B).

DISCUSSION

The PDS is an autosomal dominant disorder with phenotypic onset beginning in most persons in mid-20s.[1] The disorder is characterized by disruption of iris pigment epithelium and deposition of pigment granules on the anterior segment structures. The incidence of PDS is 4-8 per 100,000 popu-

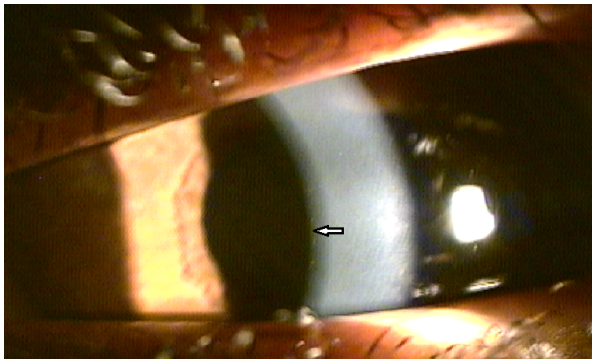

Figure 1: Slit lamp image showing Krukenberg's spindle (arrow) on back of central cornea

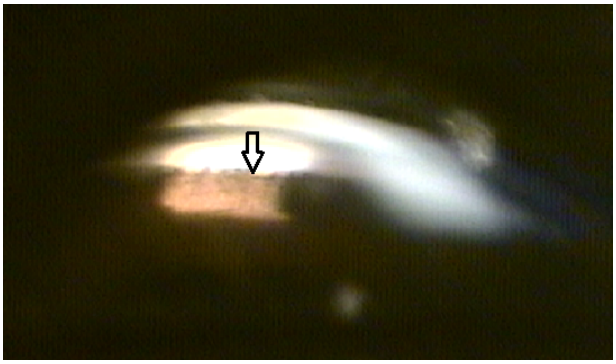

Figure 2: Gonioscopic picture showing heavy pigmentation of angle (arrow)

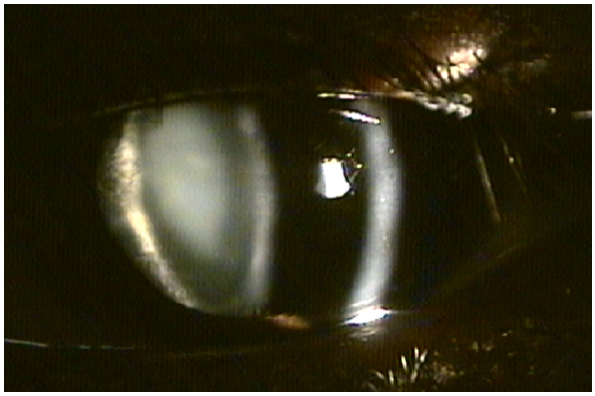

Figure 3: Slit lamp image showing posterior sub capsular cataract

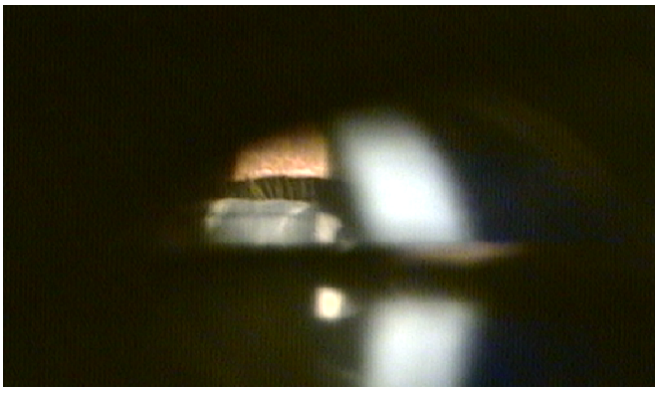

Figure 4: Gonioscopic picture showing anterior insertion of Zonules

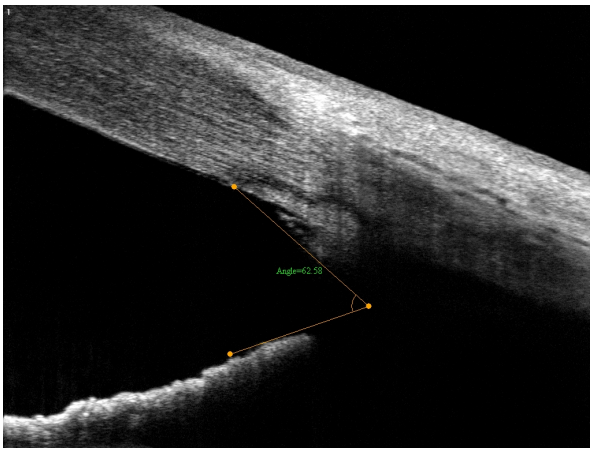

Figure 5A: Anterior segment OCT(angle) right eye showing posterior concavity of mid peripheral iris

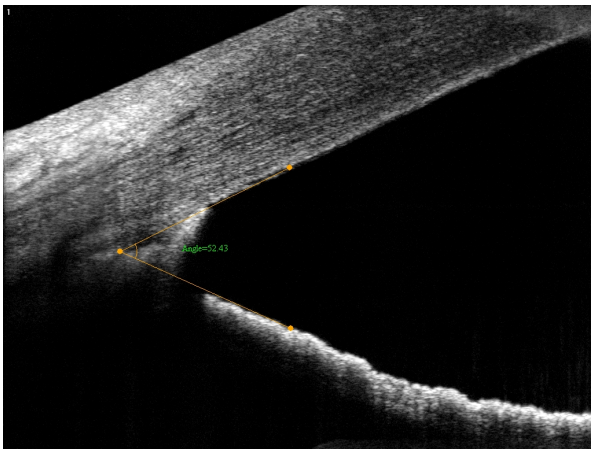

Figure 5B: Anterior segment OCT (angle) left eye showing posterior concavity of mid peripheral iris

lation. This condition is more commonly seen in Caucasians and is considered to be rare in Indians.[5] PDS is typically a bilateral disease, although asymmetry may occur. Men and women are equally affected by PDS. Men are significantly younger than women at the time of diagnosis of the disease. About 60% - 80% of patients are myopes and 20% are emmetropes.[1]

The syndrome is characterized by the triad of deposition of pigment on the posterior corneal surface in a vertical line (Krukenberg's spindle), wide open angles on gonioscopy with uniform and heavy pigmentation of trabecular meshwork, slit like radial transillumination defects in the iris. Other ocular findings commonly observed are: relatively flat cornea, deep anterior chamber,and wide open angles.[2] The iris is inserted posteriorly and shows a concave configuration in the mid periphery. This feature is best demonstrated by ultrasound biomicroscopy (UBM) or anterior segment OCT.[3,4] Pigment accumulation on the anterior surface of the iris often appears as concentric rings within the iris furrows. Pigment is also deposited on the posterior surface of the lens in the region of contact between the anterior hyaloids face and posterior lens capsule. Visualization of this circular ring or arc of pigmentation requires pupillary dilatation and is considered pathognomonic of PDS.[6] Patients with PDS and pigmentary glaucoma are at increased risk for retinal detachment and this may occur in 6%-7% of individuals. Retinal breaks and retinal detachment may occur twice frequently in these eyes.[3,4]

The concave iris configuration increases the contact between the iris and the anterior zonular apparatus. This abnormal iridozonular contact is exaggerated by blinking, pupillary dilatation, accommodation and exercise causing liberation of iris pigment into the anterior chamber. When blinking is inhibited, the iris assumes a convex configuration which is immediately reversed upon blinking. The act of blinking acts as a mechanical pump to push aqueous from the posterior to the anterior chamber.[4,7] Once in the anterior chamber, backflow of aqueous is prevented by the abnormal iridolenticular contact, which produces a reverse papillary block, further enhancing iris concavity. The liberated pigment blocks the trabecular meshwork causing rise in intraocular pressure and glaucoma. The severity of PDS decreases in middle age when pigment liberation ceases.

In PDS, treatment should begin early in order to prevent the development of glaucomatous damage and should aim at preventing the progression of disease than mere lowering of intraocular pressure (IOP). Miotic treatment produces a convex iris configuration, completely inhibiting pigment liberation.[8] Laser iridotomy results in a planar configuration of iris but may not completely prevent pigment liberation.[9] Argon laser trabeculoplasty

produces better results in younger patients because of location of pigment in the trabecular meshwork.[10]

The purpose of presenting this case is to caution clinicians regarding the possible association of PDS in young myopes with Krukenberg's spindle. Many patients with PDS remain undetected, while those with glaucoma are misdiagnosed as having juvenile onset glaucoma or primary open angle glaucoma. Those patients with Krukenberg's spindle and without elevated IOP are often treated as normal. These patients must be cautioned regarding possible future consequences of the disease and the hereditary nature of the syndrome. They must be educated about the importance of regular eye checkups and the possible avoidance of vigorous exercise which may cause liberation of pigment and progression of glaucoma. The case also highlights the importance of looking for subtle signs like phacodonesis and iridodonesis which may be due to increased axial length of the eyeball or may be an associated feature of PDS.

REFERENCES

1. Campbell DG, Schertzer RM. Pathophysiology of pigment dispersion syndrome and pigmentary glaucoma. Curr Opin Ophthalmol 1995;6:96-101.
2. Speakman JS. Pigmentary dispersion. Br J Ophthalmol 1981;65:249-51.
3. Potash SD, Tello C, Liebmann J, Ritch R. Ultrasound biomicroscopy in pigment dispersion syndrome. Ophthalmology 1994;101:332-9.
4. Chew SJ, Tello C, Wallman J, Ritch R. Blinking indents the cornea and reduces anterior chamber volume as shown by ultrasound biomicroscopy. Invest Ophthalmol Vis Sci 1994;35(suppl):1573.
5. Krishnadas R, Ramakrishnan R. Secondary glaucomas: the tasks ahead. Community Eye Health 2001;14:40-2.
6. Zentmayer W. Association of an annular band of pigment on posterior capsule of lens with a Krukenberg spindle. Arch Ophthalmol 1938;20:52-7.
7. Liebmann JM, Tello C, Chew SJ, Cohen H, Ritch R. Prevention of blinking alters iris configuration in pigment dispersion syndrome and in normal eyes. Ophthalmology 1995;102:446-55.
8. Campbell DG. Pigmentary dispersion and glaucoma: a new theory. Arch Ophthalmol 1979;97:1667-72.
9. Gandolfi SA, Vecchi M. Effect of a YAG laser iridotomy on intraocular pressure in pigment dispersion syndrome. Ophthalmology 1996;103:1693-5.
10. Ritch R, Liebmann J, Robin A, Pollack IP, Harrison R, Levene RZ, et al. Argon laser trabeculoplasty in pigmentary glaucoma. Ophthalmology. 1993;100:909-13.