

Case Report

Pigment Dispersion Syndrome and Pigmentary Glaucoma in an Emmetropic Young Male

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Abstract

Introduction: Pigment dispersion Syndrome (PDS) is a disorder with an onset in mid-twenties. There occurs a disruption of the iris pigment epithelium and deposition of pigment granules throughout the anterior segment. The incidence of PDS is 4-8/100,000. This condition is more commonly seen In Caucasians and is considered to be rare in Indians. **Case:** A 33-year-old male presented with the complaint of headache for three months. He had normal vision in both eyes with visual acuity of 6/6.

Observation: Krukenberg's spindle, a classic sign of pigment dispersion syndrome was evident on slit-lamp examination over the posterior corneal surface. Gonioscopy revealed a heavy and uniformly pigmented trabecular meshwork. OCT (Optical Coherence Tomography) demonstrated a characteristic iris configuration in the form of a mid-peripheral posterior bowing of the iris. Retinal nerve fibre layer analysis done on OCT revealed glaucomatous thinning in the right eye and a more advanced defect in the left eye. A visual field examination revealed the field to be outside normal limits in both the eyes pointing towards a diagnosis of pigment dispersion glaucoma.

Conclusion: The purpose of presenting this case is to caution the clinicians to carefully examine young emmetropes who present with Krukenberg's spindle as it could be associated with PDS. 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 counseled regarding the hereditary nature of the syndrome.

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

Introduction

In pigment dispersion syndrome (PDS) there occurs a disruption of the iris pigment epithelium and deposition of the dispersed

pigment granules throughout the anterior segment (Campbell DG et al, 1996). The classic diagnostic triad consists of corneal pigmentation (Krukenberg's Spindle), slit like radial transillumination defects involving the mid-peripheral iris and dense trabecular pigmentation on gonioscopy (Speakman JS et al, 1981; Potash Sd et al and Chew SJ et al, 1994). The insertion of the iris is typically posterior and the peripheral iris has a concave configuration. We present a case report of pigment dispersion syndrome in both eyes of

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an emmetropic young male patient and the depiction of typical iris configuration with the help of OCT which is characteristically seen in pigment dispersion syndrome even in the absence of trans-illumination defects in the iris.

Case Report

A 33-year-old male presented to the Ophthalmology out-patient department with the complaint of headache for three months. He did not have any vision related problem. History of any form of ocular trauma, family history of headache, hypertension, diabetes mellitus or any other co-morbid condition was negative. Ocular examination findings are shown in table 1.

Krukenberg’s spindle (Figure 1), a classic sign of pigment dispersion syndrome was evident on slit-lamp examination over the posterior corneal surface. Gonioscopy revealed a heavy

and uniformly pigmented trabecular meshwork (Figure 2). The insertion of the iris was typically posterior and the peripheral iris demonstrated a concave configuration on AS-OCT (Figure 3). Retinal nerve fibre layer analysis done on OCT revealed findings as shown in figure 4. A visual field examination performed using 24-2 Threshold test (2010 Carl Zeiss medite, Humphrey field analyser 11 720-50030-5.1.2/5.1.2) revealed the field to be outside normal limits in both the eyes pointing towards a diagnosis of pigment dispersion glaucoma as evident in the figure 5 (a and b).

Our aim of treatment was to reverse the iris concavity, thereby preventing further pigment release and lowering the IOP. We therefore put the patient on a prostaglandin analogue and 1% pilocarpine eye drops .The patient did not come for follow up.

Table 1: Ocular examination findings in an emmetropic young male

Feature	Right eye	Left eye
Visual acuity	6/6	6/6
CCT (μ)	484	488
Keratometry (diopters)	K1:43.60	K1: 43.16
	K2:44.12	K2:43.38
Anterior chamber depth(mm)	3.96	3.95
Axial length (mm)	23.0	22.09
IOP (GAT) mm Hg	54.0	52.0
CCT: Central corneal thickness in microns; K1 and K2:keratometry readings in vertical and horizontal axes respectively; IOP: intraocular pressure; GAT: Goldmann Applanation Tonometry		

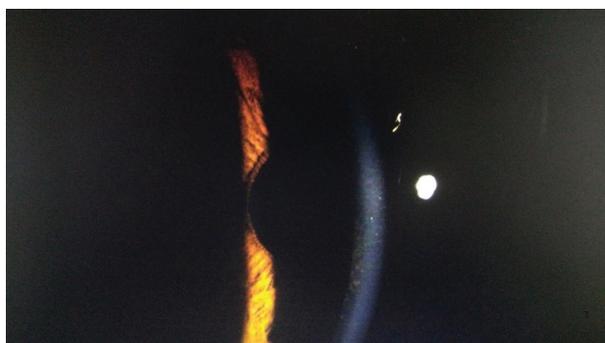


Figure 1: Cornea of the patient showing the presence of characteristic Krukenberg’s spindle in the central part

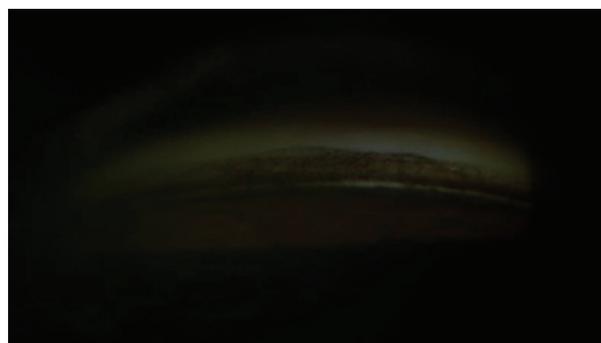


Figure 2: Gonioscopy showing a heavy and uniformly pigmented trabecular meshwork

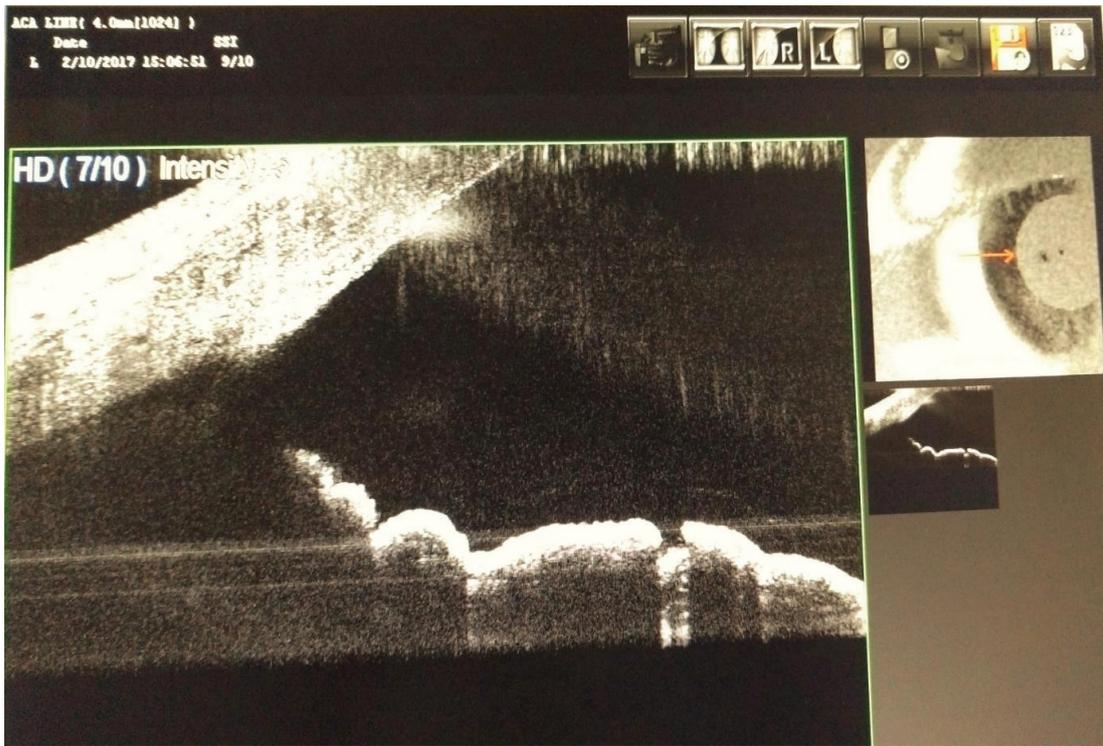


Figure 3: Anterior segment OCT (angle) showing posterior concavity of mid-peripheral iris

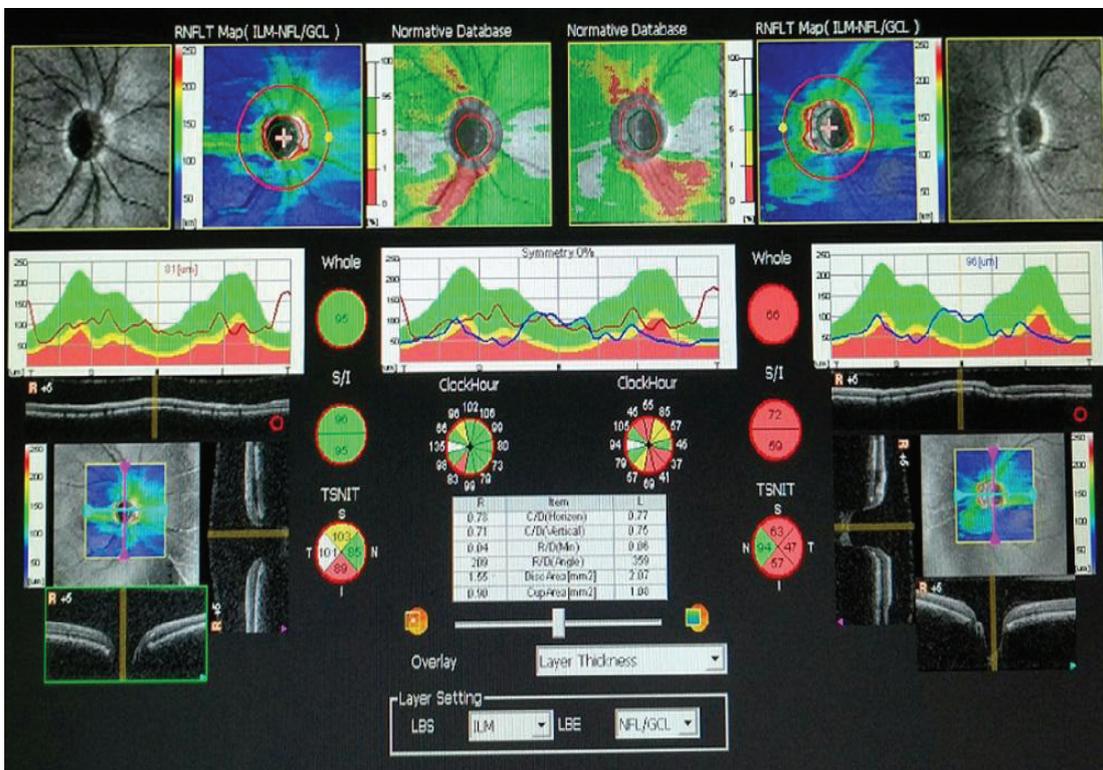


Figure 4: Retinal nerve fibre layer analysis done on OCT revealed glaucomatous thinning in the right eye and a more advanced defect in the left eye

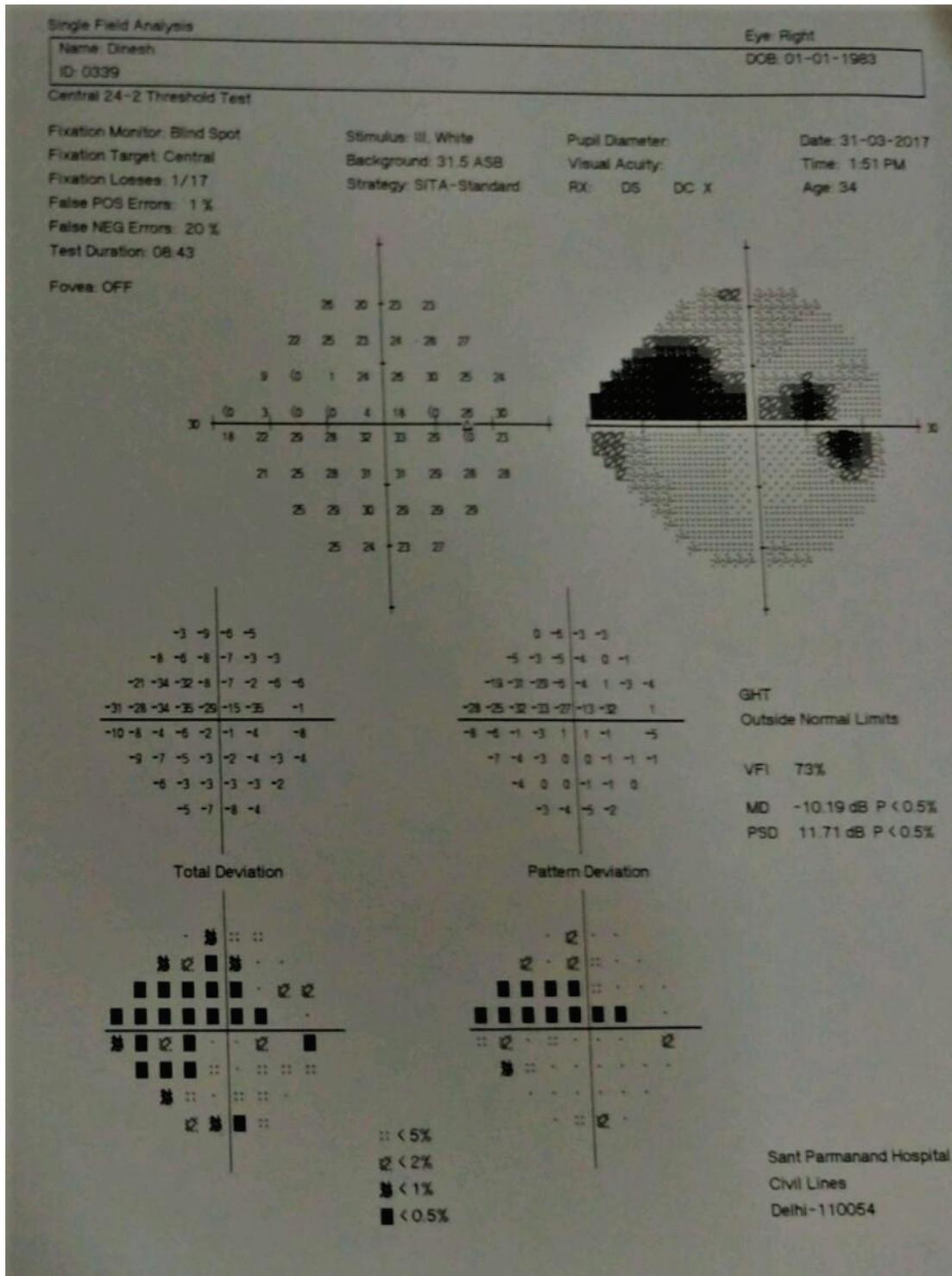


Figure 5a: Visual field examination findings of right eye

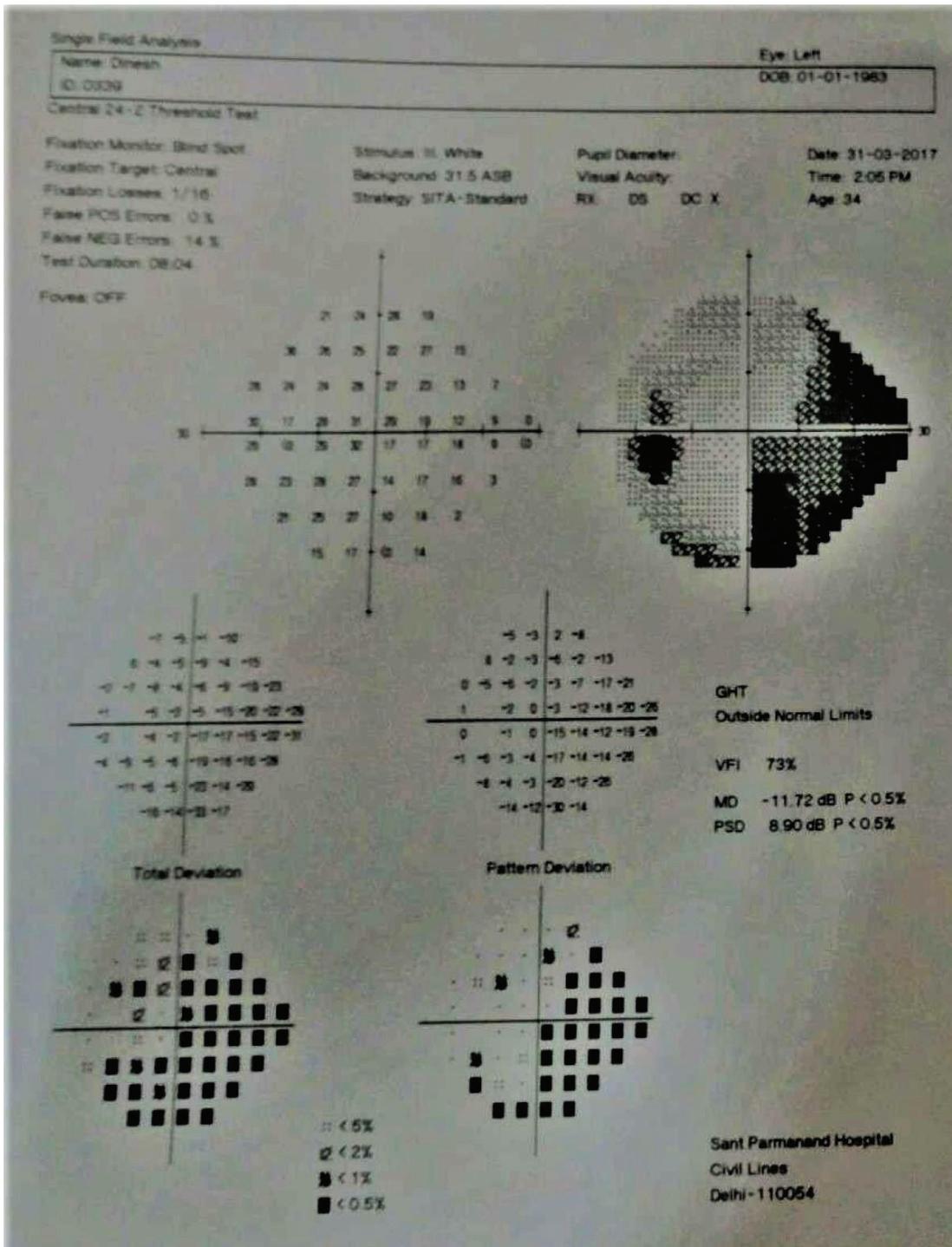


Figure 5b: Visual field examination findings of left eye

Discussion

The PDS is an autosomal dominant disorder with phenotypic onset beginning in most people in mid –twenties. (Campbell DG et al, 1996). The incidence of PDS is 4-8 per 100,000. This condition is more commonly seen In Caucasians and is considered to be rare in Indians (Krishnadas R et al, 2001). The presentation of PDS is generally bilateral, although asymmetry can also be found in some cases. There is an equal incidence of PDS in both men and women. However, men are effected at a younger age than women. Other ocular findings commonly that can be seen are: a relatively flat cornea and a deep anterior chamber. (Speakman JS et al, 1981).Pigment is also deposited on the posterior surface of the lens in the region of contact between the anterior hyaloid face and posterior lens capsule. Visualization of this circular ring or arc of pigmentation requires pupillary dilatation and is considered characteristic of PDS. (Zentmayer W, 1938) 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 as frequently in these eyes. (Potash SD et al, 1994; Chew SJ et al, 1994)The concave iris configuration increases the contact between the iris and the anterior zonular apparatus. Anterior segment OCT (angle) shows posterior concavity of mid peripheral iris. This abnormal iridozonular contact is exaggerated by blinking, pupillary dilatation, accommodation and exercise causing liberation of iris pigment into the anterior chamber. The iris assumes a convex configuration when blinking is inhibited, which is immediately reversed when blinking occurs. The aqueous is pushed from the posterior to the anterior chamber upon blinking, as the process acts as a mechanical pump. (Chew SJ et al, 1994; Liebman JM et al, 1995). Once in the anterior chamber, backflow of aqueous is inhibited by the abnormal iridolenticular contact, which

results in a reverse pupillary block, further increasing iris concavity .The liberated pigment blocks the trabecular meshwork causing rise in intraocular pressure and glaucoma. There occurs a decline in the severity of PDS in middle age as pigment liberation decreases. In order to prevent glaucomatous damage treatment should be started early in PDS and the aim of treatment should be to prevent the progression of disease rather than mere lowering of intraocular pressure (IOP). Miotics act by producing a convex iris configuration thereby inhibiting pigment release. (Campbell DG, 1979).Laser iridotomy results in a planar configuration of iris but may not completely prevent pigment liberation. (Gandolfi SA et al, 1996).

Conclusion

The aim of presenting this case report is to make the clinicians aware of a possible association of PDS in young emmetropes 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 pigment release and progression of glaucoma.

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