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Pigment Dispersion Syndrome with Pigmentary Glaucoma - A Rare Case Report

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Abstract: ***Pigment dispersion syndrome (PDS) and pigmentary glaucoma (PG) is a spectrum of same disease characterized by excessive pigment liberation throughout the anterior segment of the eye. The disease is more prevelent among young myopic males. Here we present a case of Pigment dispersion with pigmentary glaucoma in a 48 year old hypermetropic female patient. All the classical sign of PDS were present like Krukenbergs spindle, pigments in anterior chamber, increased pigmentation of trabecular meshwork in both the eyes and Pigmentary glaucoma in right eye.***

Keywords: PDS, Pigment dispersion syndrome, Krukenberg's Spindle, Transillumination defects, Pigmentary Glaucoma, Glaucoma, Trabecular pigmentation

1. Introduction

Pigment dispersion syndrome (PDS) is characterized by disturbance pigment epitheliumof iris and toppling of the dispersed pigment granules all over the anterior segment. The Pigment dispersion syndrome consists of a classic diagnostic triad Krukenbergs spindle, Iris transillumination defects and Trabecular pigmentation (1). The insertion of iris is posterior and concave at periphery. We report a case of pigment dispersion syndrome in a 48 years female patient.

2. Case

A 50 years old female presented with complaints of headache for past 2 months. On examination her best corrected distance vision was 6/6 in BE with +1.00 DSP and N6 for near vision with addition of + 2.00DSP. She had no systemic illness. On Slit lamp examination, Krukenberg's spindle was present on back of cornea in BE(figure 1).

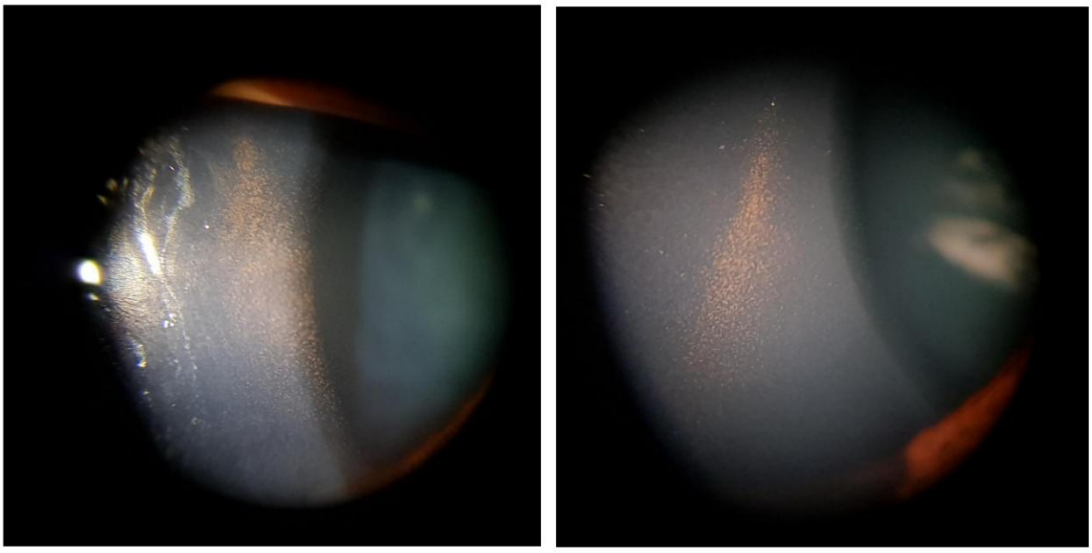

Figure 1: Both eye Krukenbergs spindle Back of cornea

Pigment dispersion was more diffuse covering more than half of cornea in RE and linear spindle shaped in LE. Pigment granules were seen in Anterior chamber. Iris transillumination was not seen. On Gonioscopy angles were open in BE with pigmented trabecular meshwork which was more in RE compared to left eye. CCT corrected IOP by AT was 24 mm of Hg in RE and 19 mm of Hg in LE. On Fundus examination, CD Ratio of 0.5, laminar dot sign and peripapillary atrophy was noted in RE while in LE CD Ratio was 0.3. Humphery Automated Perimetry using 30-2 programme revealed a visual field defect in superior and inferior quadrant in RE (figure 2) where as LE was normal

(figure 3). So a diagnosis of Pigment Dispersion syndrome with RE Pigmentary Glaucoma was made. Patient was started on Topical Latanoprost eye drops in RE. IOP reduced to 22mm of Hg after 2 weeks of follow up. Family history was not significant. All her siblings and children were advised for screening of glaucoma. Patient was explained regarding the seriousness of the disease and chance of other eye getting involved in the future was explained. Importance of lifelong regular follow up and continuation of drops to prevent complete blindness do to glaucoma was also stressed upon.

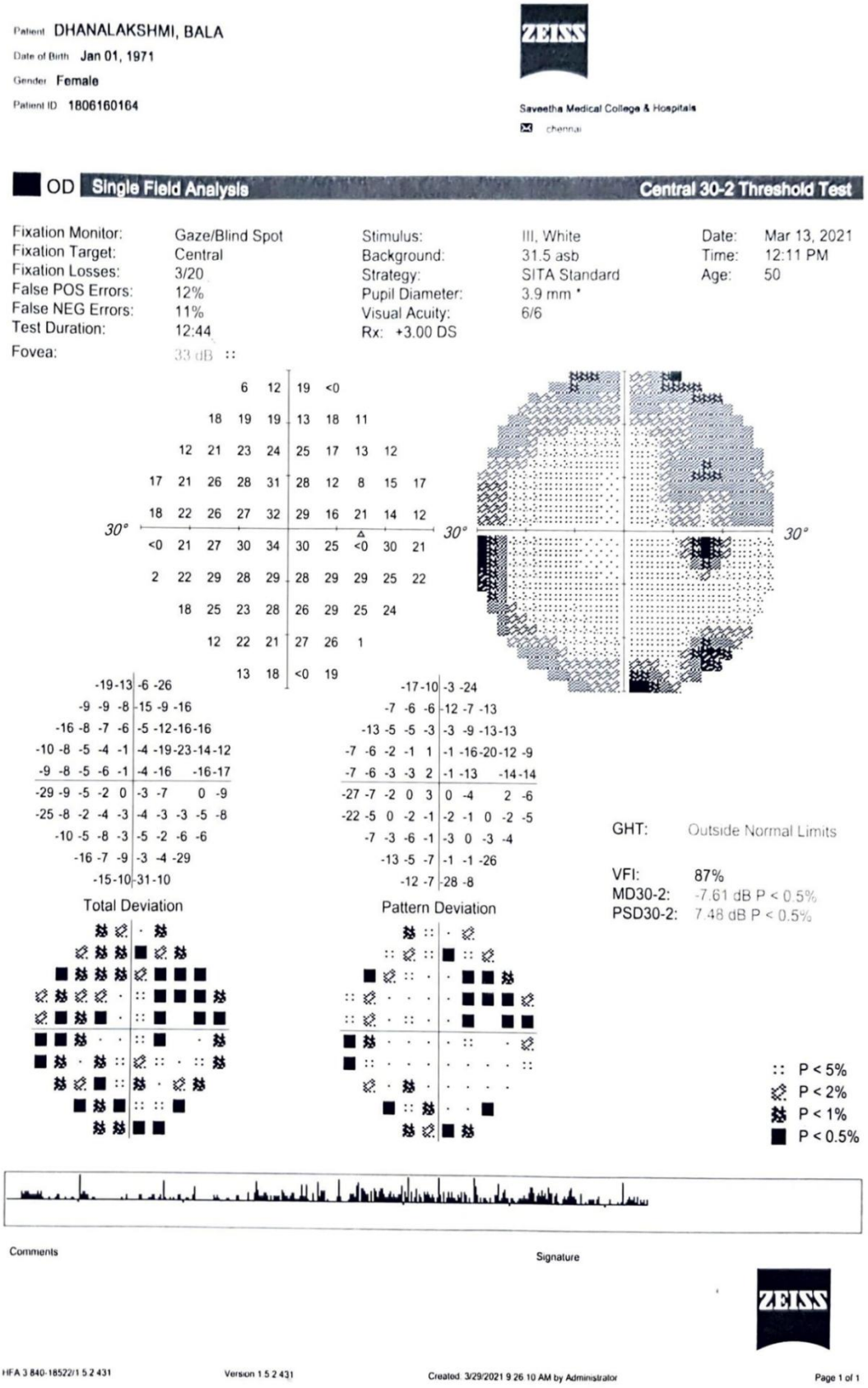

Figure 2: Right eye HFA revealed a visual field defect in superior quadrant and inferior quadrant

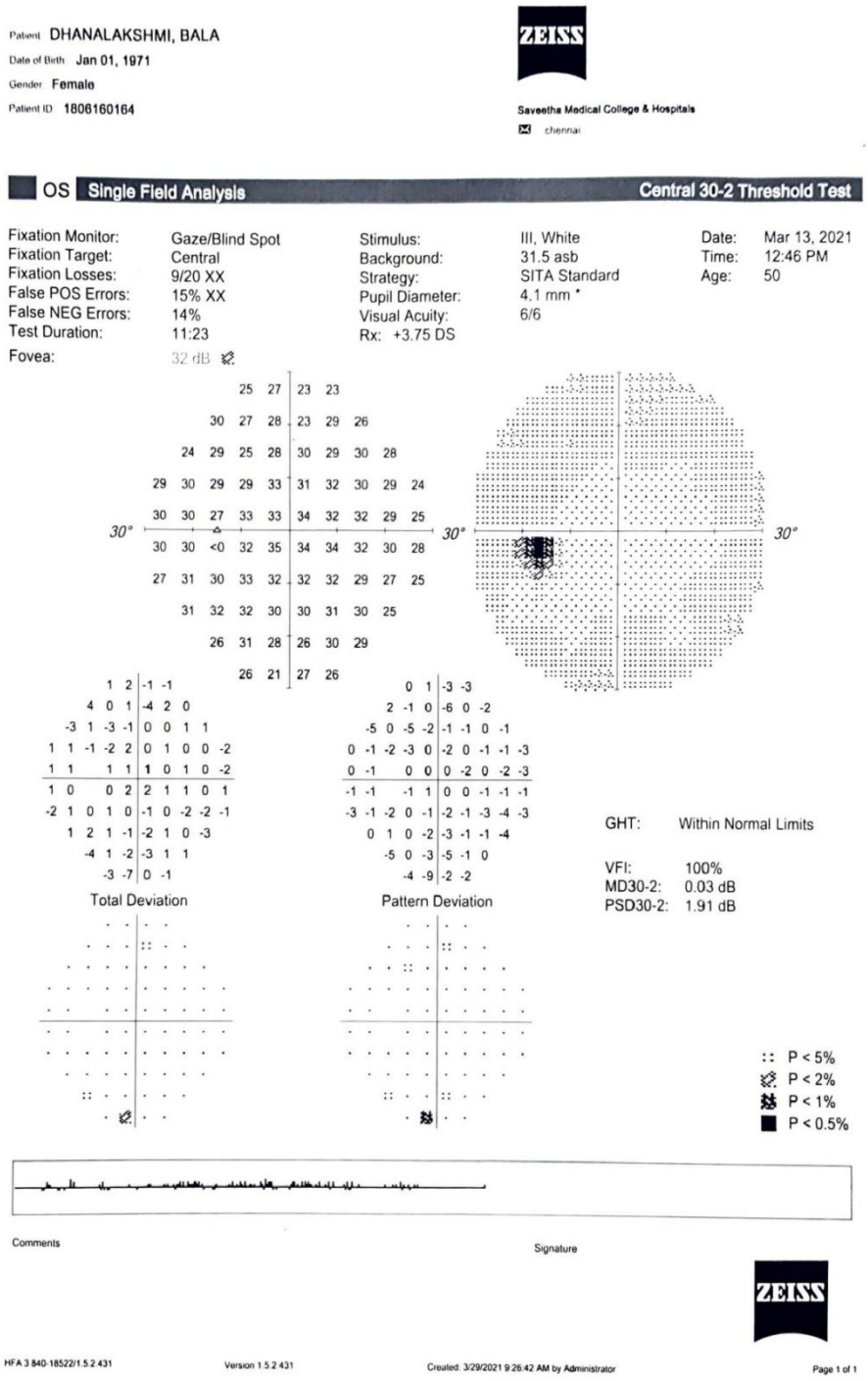

Figure 3: Left eye HFA shows no field defects

3. Discussion

Pigment Dispersion syndrome is an autosomal dominant disorder. Thisis characterized by disturbance of iris pigment epithelium and deposition of pigment granules on the anterior segment structures. Incidence is 4 per 1 lakh population and rare India. Usually bilateral but asymmetry is also noted. Both sexes are equally effected. About 60-80% are myopes and 20% emmetropes. This consists of triad Krukenberg's spindle, Iris transillumination defects and Trabecular pigmentation. (1),(2).

Other finding are Flat Cornea, Deep anterior chamber and wide open angles. Iris is posterior inserted and exhibits concave configuration in the mid periphery this is demonstrated by ultrasound biomicroscopy and anterior segment OCT. Pigments are also will be deposited in posterior surface of lens. Patients with PDS are in increased risk of retinal detachment and retinal breaks.

The concave iris configuration will increases the contact in between iris and anterior zonular apparatus. This contact is increased while blinking, accommodation, pupillary dilatation and exertion causes liberation of pigments in to anterior chamber.(8) This pigments blocks trabecular meshwork which gives rise to increase in IOP and Glaucoma.(3)

Treatment should be started early to prevent Glaucomatous changes. (4) Miotic treatment causes conves iris configuration and completely inhibits pigmments disperstion. Argon laser trabeculoplasty gives good results in young patients. (5)

Usually maximum patients with PDS may remain undetected. (6)

Patients with Krukenberg's spindle and normal IOP are treated as normal. (9), (7). But these patients should be aware of future consequences, hereditary nature of the disease, importance of regular follow up and avoidance of vigorous exercise which liberate pigments.

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