Pigment Dispersion Syndrome: A Rare Case Report

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Abstract

Pigment dispersion syndrome (PDS) is a well-known clinical entity usually discovered during routine examination of an otherwise healthy adult eye and is associated with the development of pigmentary glaucoma. It's characteristic feature includes loss of iris contour and loss of pigment granules from the iris.A 55 year old female presented with complaints of painless, gradually progressive diminution of vision in both eyes. On ocular examination,both eyes showed pigment deposition on the corneal endothelium in a vertical spindle-shaped distribution suggestive of Krukenberg's spindle. On gonioscopy, there was presence of heavy pigmentation on the trabecular meshwork (Sampaolesi line).Regular IOP monitoring of this patient was advised for early detection of pigmentary glaucoma.

Key words: *Pigment dispersion syndrome, Krukenberg spindle, pigmentary glaucoma*

Introduction

Pigment dispersion syndrome is a rare disorder in young adults which is associated with development of pigmentary glaucoma.Its characteristic features include loss of iris contour and loss of pigment granules from the iris [1].

PDS results from iridozonular friction and some possible contribution from iridociliary process contact with resultant liberation of pigment from the posterior iris. The liberated pigment accumulates on the corneal endothelium, the front surface and circumferential furrows of the iris, the posterior lens capsule and lens zonules within the trabecular meshwork. This pigment dispersion is mainly due to aqueous convection currents [2,3]. When aqueous outflow is affected and intraocular pressure rises, optic nerve damage may ensue. This process is known as pigmentary glaucoma (PG). Pigment dispersion syndrome is found most commonly in patients between the ages of 30 and 50 years [4]. Myopia is frequently associated with pigment dispersion syndrome with a mean myopic correction of-3.9 diopters [4]. Hypermetropia with PDS has been rarely reported [5]. The risk of developing pigmentary glaucoma from pigment dispersion syndrome is about 10% in most series.About a third of patients with PDS eventually develop ocular hypertension or secondary open-angle glaucoma after 15years.Men are affected twice as frequently as women [6].

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Case Report

A 55 year old female presented with complaints of diminution of vision in both eyes since last one year. On examination, her vision in right eye was 6/36, while in left eye it was 6/24 improving with pinhole up to 6/9 in both eyes. Subjective refraction was +2.75 D spherical in Right eye and +2.00 D spherical in Left eye. Intraocular pressure was 17.3 in both eyes, recorded with Schiotz tonometer.

Corneas of both eyes showed pigment deposition on the endothelium in a vertical spindle-shaped distribution suggestive of Krukenberg's spindle. There was absence of transillumination defects in iris. On gonioscopy, presence of heavy pigmentation on the trabecular meshwork (Sampaolesi line) was observed.

Fundus examination revealed normal optic disk appearance. Visual field examination was within normal limits.

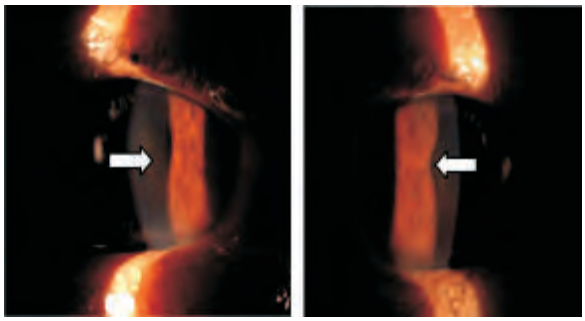

Fig 1: Right eye and left eye showing krukenberg's spindle

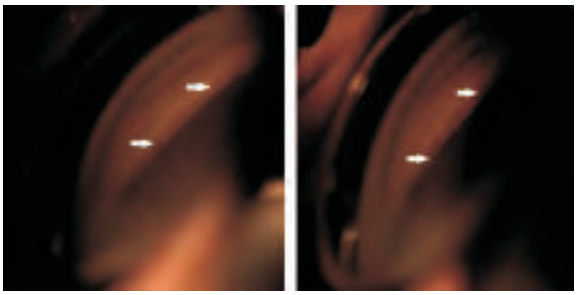

Fig 2: Right eye and left eye showing sampolesi line

Discussion

In humans, pigment dispersion has been shown to be inherited as an autosomal dominant trait, which suggests that specific gene defects may be responsible. One locus for this syndrome has been found on7q35-q36, 18 but the responsible gene has yet to be isolated. The high prevalence of this condition indicates that more than one gene may be responsible for this disorder. Two genes that contribute to a form of pigment dispersion syndrome and glaucoma in the DBA/2J mouse have been identified. These genes, TYRP1 and GPNMB, are involved in

melanosome metabolism; however, neither of these genes contributes to the disease in humans [7].

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. In a majority of the patients with PDS, there is a concave approach of the iris as it inserts into the anterior chamber angle [2,3]. The cause of this concavity remains unclear [2].

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. Laser iridotomy results in a planar configuration of iris but may not completely prevent pigment liberation. Argon laser trabeculoplasty produces better results in younger patients because of location of pigment in the trabecular meshwork [8].

Regular and frequent follow up of the patient was done for 1year so as to detect early pigmentary glaucoma. However, patient was lost to follow up after that. Association of PDS with hypermetropia in a female patient makes this a unique and rare case.

References

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