Different Pattern of presentation of pigment dispersion syndrome among Bangladeshi population

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Abstract

Purpose: *To report the clinical findings and characteristics of pigment dispersion syndrome (PDS) in Bangladeshi population.*

Method: *PDS suspects with any one of the following signs: corneal endothelial pigmentation, iris trans illumination defects (ITDs), pigment granule dusting on anterior iris surface, posterior iris bowing, trabecular meshwork (TM) pigmentation and lenticular or zonular pigmentation were evaluated for PDS at the Glaucoma specialty clinic at Chittagong Eye Infirmary and Training Complex. Diagnosis of PDS required at least two of the following signs: Krukenberg spindle, moderate-to-heavy TM pigmentation, and any degree of lenticular and/or zonular pigmentation.*

Result: *Thirty-six patients (24 males and 12 females) were identified as having PDS during a 1-year period, with a mean age of 35.5±7.0 years (range, 22–49). All but two eyes from two patients had myopia of 0.5 D or greater, with the mean spherical equivalent power of 5.20±5.80 D (range, 24.75±0.5). The average IOP at initial diagnosis was 33.7±10.5 mm Hg (range, 16–56). Thirty patients (83.3%) were found to have pigmentery glaucoma at their initial diagnosis. All patients showed homogenous increased TM pigmentation as well as lenticular and/or zonular pigmentation. 61.1% of patients had Krukenberg spindle. None of the patients exhibited spoke-like mid-peripheral transillumination defect except for trace-isolated transillumination in both eyes of the two patients.*

Conclusion: *The most common clinical findings in Bangladeshi PDS patients include corneal pigment granules, homogeneous TM pigmentation and pigment granule dusting on lens zonules and/or posterior peripheral lens surface. Transillumination defects are uncommon in Bangladeshi patients with PDS.*

Keywords: *Pigment, Glaucoma, Krukenberg, Pigmentation.*

Introduction

Pigment dispersion syndrome (PDS) is characterized by pigment granule dispersion from the iris pigment epithelium (IPE). The released pigment granules are transmitted by aqueous convection currents and deposit on the corneal endothelium, iris surface, trabecularmeshwork (TM), lens surfaces and zonules. Accumulation of pigment in the TM may lead to increased resistance to aqueous outflow and results in the development of pigmentary glaucoma (PG).[1, 2]

The prevalence of PDS among adult whites is as high as 2.45%.[3] Commonest clinical findings in white PDS patients include the triad of corneal krukenberg spindle, mid-peripheral iris transillumination defects, and homogenous TM pigmentation, usually found in young myopic male patients.[4, 5, 6, 7] The prevalence of PDS in blacks >7 years old was calculated to be 0.167%±0.013.[8] Clinical signs in dark PDS

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patients is narrower than that of typical and classic type in whites. Transillumination defects and anterior iris stromal pigment dusting are, however, not as common in blacks.[8, 9, 10, 11, 12, 13] To find out the clinical features of PDS in Bangladeshi, we carried out a prospective observational investigation on a series of 36Bangladeshipatients identified as having PDS according to the identical diagnostic criteria.

Patients and methods

Patient suspected as PDS at Glaucoma clinic at Chittagong Eye Infirmary and Training Complex from May 2015 to April 2018 were evaluated,If the patient had any one of the following signs: corneal endothelial pigmentation, anterior iris stromal pigment dusting, ITDs, posterior iris bowing, increased TM pigmentation, and pigment granule dusting on lens zonules or peripheral posterior surfacewere included. Detailed ophthalmic examinations included visual acuity measurement, IOP measurement, refraction, slit-lamp bio-microscopy pre- and post-mydriasis, gonioscopy, fundoscopic examination and automated Humphrey SITA standard 24-2 visual field test were done. Systemic and ocular medical history of each subject was also recorded.

TM pigmentation was graded according to Scheie's grading system. Corneal endothelial pigment dusting was described as Krukenberg spindle, diffusive pattern, none. If the patient had received anti-glaucoma medications before the examination, the IOP measured before medication was taken as the baseline IOP. Slit-lamp examination, gonioscopy and fundoscopic evaluation of the optic disc of all patients were performed and graded by the same physician to avoid inter-physician differences.

Diagnostic criteria for PDS included at least two of the following three signs: corneal Krukenberg spindle, homogenous moderate-to-heavy TM pigmentation and any degree of zonular and/or lenticular pigment granule dusting. Patients with a history of uveitis, trauma, ocular surgery or anterior segment laser treatment, or any evidence of exfoliation material were excluded. Patients with PDS were diagnosed with secondary glaucoma, if they had two or more of the following findings: initial IOP >21 mm Hg, glaucomatous optic nerve damage (increased cupping or abnormal disc appearance) or glaucomatous visual field loss.

Results

Out of 188 PDS suspectsthirty six patients (24 males and 12 females) were identified as having PDSaccording to criteria.Mean age of the PDS patients was 35±5.0 years (range, 22–49). Male to female ratio was 2:1.

All patients hadmyopia of 0.5 D or greater except 2.35 patients had increased baseline IOP of >21 mm Hg in at least one eye at the time of diagnosis with an average of 32±10 mm Hg.

Four patients had family history of glaucoma. Another 8 had family history of PDS. All of them were taking anti-glaucoma medications at the time of evaluation for PDS. Only 12 of the PDS patients had symptoms which were mostly occasional blurring of vision.

Bilateral Krukenberg spindle was found in16 patient and unilateral in 6. (Figure 1a). Of the remaining 14, three had trace diffuse corneal endothelial pigmentation and 8 had no corneal pigment dusting.

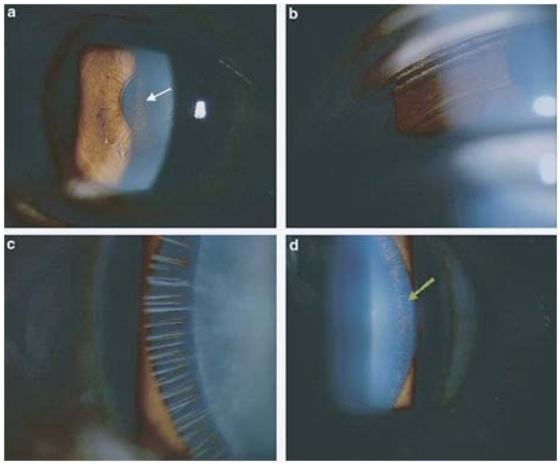

Figure 1 : (a)Triangular Krukenberg spindle (white arrow) (b) Trabecular meshwork pigmentation . (c) Pigment granules on lens zonules. (d) Pigment accumulated at the zonular attachments to the lens, where it formed a Zentmayer ring, also referred as Scheie's line (green arrow).

Posterior bowing of the mid-peripheral iris was found except 2 cases. The slit beam was projected vertically through the centre of the pupil at 30–45 degrees.

In 4 patients short slit-like transillumination defects were visualized in iris crypts.

Homogeneous TM pigmentation and pigment granule dusting on lens zonules and/or peripheral posterior lens surface were seen in all patients .On slit lamp exam all patients showed different extent of pigment on lens zonules (Figure 1c) and/or posterior peripheral surface (Zentmayer ring) (Figure 1d).

30 out of 36 patient(83%) developed glaucoma.Out of 30,20 patients were bilateral and 10 patients were unilateral. 4 patients presented with blindness at the time of initial presentation. They had severe constriction of visual field in bilateral eyes.

Discussion

Among two phenotypes of PDS,one is the classic type in whites characterized by the triad of Krukenberg spindle, trans illumination defects and homogeneous TM pigmentation. Posterior iris bowing, pigment granule dusting on peripheral posterior lens surface and/or zonules, as well as anterior iris stromal pigment dusting are also common findings in white PDS patients.[3, 6, 7, 14] The other phenotype is the one found in blacks. Black patients may not have mid-peripheral radial ITDs.[8, 9, 10, 11, 12, 13] The Results of our study have shown that the clinical features of PDS in Bangladeshis are more like that of blacks. Homogeneous TM pigmentation and posterior lenticular and/or zonular pigment granule dusting are the most common findings in both black and Bangladeshi PDS patients. In all, 61% of Bangladeshi PDS patients in our series have Krukenberg spindle. In black patients, this percentage was 57.1%.[8] Nevertheless, there are differences in clinical findings between black and Bangladeshi PDS patients. Posterior iris bowing was very common in Bangladeshi PDS patients seen in 94% of the patients, but is rare in black patients.[8] Trace clusters of pigment granules on inferior surface of the iris were discerned in 6 of the 36 (16%) patients.

The gender and age distribution of these PDS patients was similar to white and black PDS patients. All the patients were detected between 20 and 50 years of age, with a 2:1 male to female ratio.

Black PDS has no ITDs due to dark iris.[8, 11, 13] There are a greater content of pigment granules in melanocytes and iris stroma in dark irides than in light ones can block an iris transillumination defect. Bangladeshi iris colour is usually brown or dark brown for which there is usual absence of ITDs. Infra-red imaging technique, which has demonstrated to be helpful in detecting ITDs in black PDS patients,[11, 13] may pick up iris defects in our patients too.

To our knowledge, there has been no published data regarding clinical features of PDS in Bangladeshipatients.Theoretically it's a rare disease. But the results of this study is so alarming that, this may not be the actual situation. In this study, we found that PDS patients comprised 1.1% (36 out of 3264) of all outpatients in Glaucoma clinic of CEITC. This is much more common than we expected. So it might be underestimated.There is no typical ITDs in Bangladeshi populations.For this, PDS is missed from doctor's attention. 12 of the PDS patients in this study had been diagnosed with primary open angle glaucoma before referral to the authors and this is a common practice so far. Secondly, even though Krukenberg spindle is seen in 61% ofour PDS patients, it is not obvious because of the dark background of the highly pigmented iris, especially when the pigmentation was minimal. This pigment does not do any harm of endothelial function like other study. Stromal pigment dusting was absent .Krukenberg spindle, ITDs, and anterior iris stromal pigment granule dusting are all important signs of PDS and in pigmentary glaucoma. But none of them is easily detected in Bangladeshi PDS patients. This may lead to missing of some cases. Lack of early diagnosis, most of them presents with raised IOP and optic nerve damage.In our study 83% of patients had PG and 94% of them had increased

IOP at their initial diagnosis of PDS. These percentages are much higher than white PDS patients.[3, 6, 7, 14, 15]

Commonest clinical findings in Bangladeshi PDS patients are homogeneous moderate-to-heavy TM pigmentation and pigment granule dusting on peripheral posterior lens surface and/or zonules. Radial mid-peripheral ITDs are rare in this patient population. 61% of the patients had Krukenberg spindle. Among them, 83% had PG at the time of initial diagnosis of PDS. So the clinical findings of PDS in Bangladeshi populationis really alarming because they may have glaucoma or may develop glaucoma within a very short time.

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