ORIGINAL ARTICLE

DETERMINATION OF ASSOCIATION OF PIGMENTARY GLAUCOMA WITH PIGMENT DISPERSION SYNDROME

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Background: Pigment Dispersion Syndrome (PDS) is an autosomal dominant disorder of white males between 20 to 40 years of age characterized by deposition of pigment on the lens, zonules of lens, trabecular meshwork and corneal endothelium (Krukenberg's spindle) in addition to radial, spoke like transillumination defects in the mid peripheral iris. This study was conducted to determine the frequency of occurrence of Pigmentary Glaucoma in patients with Pigment Dispersion Syndrome (PDS). **Methods:** This longitudinal follow up study included patients presenting with Krukenberg's spindle on the endothelial side of cornea and pigmentation of angle of anterior chamber seen on slit lamp examination and gonioscopy. **Results:** Seventy-two cases of PDS were included in the study, amongst them 63 (87.50%) were males. Mean age was 35.00±6.54 years (range 24–46 years). Forty-seven (65.28%) patients had an IOP in the range of 10–14 mmHg, 22 (30.56%) patients had an IOP in the range of 15–18 mmHg and 3 (4.17%) patients developed an IOP of greater than 19 mmHg. Fundoscopy showed myopic degeneration in 49 (68.06%) patients and optic disc cupping in 3 (4.17%) patients. Four (5.56%) patients had refractive error between +1D to +3D, 9 (12.50%) patients had refractive error between -1D to -4D, 21 (29.17%) patients had refractive error between -5 D to -8 D and 38 (52.78%) patients had refractive error between -9 D to -12 D. Our study showed that one patient having PDS developed glaucoma at 5 years of follow up and three patients developed glaucoma at 14 years of follow up. **Conclusion:** On the basis of this study we conclude that early onset primary open angle glaucoma associated with PDS or Juvenile glaucoma associated with PDS might have been mistaken as Pigmentary Glaucoma in Pakistani patients and a distinct entity in the form of Pigmentary Glaucoma may be non-existent.

Keywords: Pigment dispersion syndrome; Pigmentary glaucoma; Open angle glaucoma; Pigment epithelium; Trabecular meshwork; Trans-illumination

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INTRODUCTION

Pigment Dispersion Syndrome (PDS) is an autosomal dominant disorder of white males between 20–40 years of age characterized by deposition of pigment on the lens, zonules of lens, trabecular meshwork and corneal endothelium (Krukenberg's spindle) in addition to radial, spoke like trans-illumination defects in the mid peripheral iris.[1] Pigment may also deposit in the posterior lens capsule called as the Zentmayer's ring and in the trabecular meshwork above the Schwalbe's line which is called as the Sampaolesi's line.[2] Association of PDS with glaucoma was reported in 1949 in two young myopic males with findings of hyperpigmented trabecular meshwork and deposition of pigment over endothelial side of cornea in the form of more or less vertical spindle.[3]

Myopia is supposed to be the prime risk factor for development of PDS and posterior bowing of iris is blamed as the prime cause of pigment dispersion, mechanism being release of the pigment from pigment epithelium of the concave iris, consequent upon its rubbing effect on the anterior zonular fibres of the lens.[4,5] Two different pigments, melanin (brown coloured) and lipochrome (yellowish-brown coloured) are present in the human iris and are responsible for the iris color.[6] If no pigment is present in the iris, its colour looks like pink, if very small amount of pigment is present then the iris appears blue, if small amount of pigment is present then the iris appears green, if moderate amount of pigment is present then the iris appears hazel and if plenty of pigment is present then the iris appears brown.[7]

With the development and advancements in the field of genetics it is now known that the iris colour in human beings is controlled by at least 16 different genes and how they regulate and control the development of iris colour is a complex process.[8] Most of the limited studies performed on this topic up till now show that the risk of progression of PDS to pigmentary glaucoma is about 10%.[9] We took up PDS for study, to assess the frequency of occurrence of pigmentary glaucoma in Pakistani patients.

MATERIAL AND METHODS

Prospective follow up of 72 cases of PDS was done to see development of pigmentary glaucoma in these cases. The study was conducted at Ophthalmology Department, Chandka Medical College Larkana, Pakistan, from August 2001 to March 2015. Patients presenting with Krukenberg's spindle on the endothelial side of cornea and pigmentation of angle of anterior chamber seen on gonioscopy were considered as cases of PDS. Presence of pigmentation on other parts of the anterior segment and trans-illumination defects in the iris were taken as additional points in favour of the diagnosis of PDS. Patients with presence of secondary pigment dispersion associated with causes like, pseudoexfoliation, iris cyst, nevus, malignant melanoma, intraocular inflammation, intraocular surgery, ocular trauma and irradiation were excluded from the study. Slit lamp examination, Applanation tonometry, gonioscopy, fundoscopy and Refraction was performed on every case on two monthly follow up visits. Automated Perimetry was performed on every case at six monthly intervals. SPSS version 20 was used for data entry and analysis.

RESULTS

Seventy-two cases of PDS according to the inclusion criteria were included in the study, amongst them 63 (87.50%) were males and 9 (12.50%) were females. Mean age of patients was 35.00±6.54 years with a range of 24–46 years. Anterior segment findings on slit lamp examination and gonioscopy showed that all the 72 patients had trabecular meshwork pigmentation, 46 (63.89%) patients had Krukenberg's spindle, 19 (26.38%) patients had trans-illumination defects and 16 (22.22%) patients had lens surface pigmentation (Figure-1). Forty-seven (65.28%) patients had an IOP in the range of 10-14 mmHg, 22 (30.56%) patients had an IOP in the range of 15–18 mmHg and 3 (4.17%) patients developed an IOP of greater than 19 mmHg. Fundoscopy showed myopic degeneration in 49 (68.06%) patients and optic disc cupping in 3 (4.17%) patients. Table-1 shows frequency of pigmentation. These three patients also showed visual field defects on Perimetry. Four (5.56%) patients had refractive error between +1D to +3D, 9 (12.50%) patients had refractive error between -1D to -4D, 21 (29.17%) patients had refractive error between -5 D to -8 D and 38 (52.78%) patients had refractive error between -9 D to -12 D. A total of 31 (43.06%) patients were lost to follow up. Seven (9.72%) patients were lost to follow up in the first four years, 4 (5.56%) more were lost to follow up in the fifth year, 8 (11.11%) patients were lost to follow up in the next five years and another 12 (16.67%) patients were lost to follow up in the last four years. A total of 41 (56.94%) patients completed the follow up. Our study showed that one patient having PDS developed glaucoma at 5 years of follow up and 3 patients developed pigmentary glaucoma at 14 years of follow up. Out of the 3 patients developing glaucoma, 2 were males.

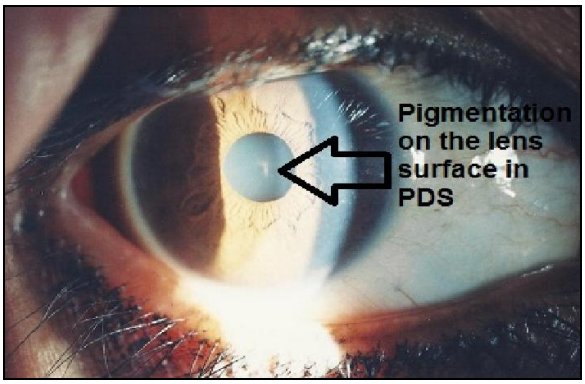

Figure-1: Pigmentation on anterior lens capsule in PDS

Table-1: Anterior and posterior segment findings

Anterior segment	No. of patients	Percentage
Krukenberg's spindle	46	63.89
Transillumination defects	19	26.38
Pigmentation of trabecular meshwork	72	100
Lens surface pigmentation	16	22.22
Posterior segment		
Myopic degeneration	49	68.06
Optic disc changes including cupping, atrophy, swelling	3	4.17

DISCUSSION

Our study indicates that Pigment dispersion syndrome is more common in young to middle aged myopic males as is reported in previous studies.[9,12,19,20] In our study, 1.64% patients developed pigmentary glaucoma at 5 years and 7.32% patients developed pigmentary glaucoma at 14 years. Siddiqui Y *et al*[12] has reported the risk of developing pigmentary glaucoma from PDS as 10% at 5 years and 15% at 15 years which is higher compared with our study. According to Sivaraman KR *et al*[13] it is difficult to predict which patients will progress to pigmentary glaucoma from PDS. Few questions arise at this stage as to why should the pigment block the trabecular meshwork when nature has made numerous arrangements to clear the pigment deposited on trabecular meshwork like macrophages, wandering histiocytes and trabecular endothelial cells all take part in clearing the trabecular meshwork and anterior chamber angle by way of phagocytosis.[13,14] Why even the drugs which increase the size of melanosomes like prostaglandin types of medications do not cause pigmentary glaucoma? On the contrary pigmentary glaucoma can be well treated by prostaglandin like medications.[18] Estimated prevalence of primary open angle glaucoma varies from 1.1–3.9% in different studies[15–17], which is close to the prevalence

of pigmentary glaucoma reported in this study. Alvarado JA *et al*[10] reports that a common pathophysiological mechanism is operative for the development of both primary open angle glaucoma and pigmentary glaucoma. On the basis of review of literature and the present study one can reconsider the occurrence of pigmentary glaucoma as a distinct entity. It seems that early onset primary open angle glaucoma associated with PDS or juvenile glaucoma associated with PDS has been mistaken as pigmentary glaucoma and in reality, a distinct entity like pigmentary glaucoma may be non-existent. However, the higher ratio of patients lost to follow up, 43%, may affect the results of this study and more studies are required to prove this opinion.

CONCLUSION

Forty one out of the 72 patients completed the fourteen years follow up study. Three cases out of the 41cases completing the follow up developed the so called pigmentary glaucoma, although all patients had enough pigment deposition on their trabecular meshwork's. The study shows that 1.64% patients developed pigmentary glaucoma at 5 years and 7.32% patients developed pigmentary glaucoma at 14 years. Juvenile glaucoma which occurs due to decompensation of a less mature trabecular meshwork in twenties of life when associated with PDS may have been considered as pigmentary glaucoma in mistake or the other possibility of early onset primary open angle glaucoma associated with PDS mistaken as pigmentary glaucoma seems to exist.

Recommendation: We recommend the alternate term for Pigmentary Glaucoma as "Primary Open Angle Glaucoma with PDS".

AUTHORS' CONTRIBUTION

IAS: Substantial contribution to conception and design, acquisition of data, analysis and interpretation of data for intellectual content and final approval of the study to be published. SAS: Drafted and revised the manuscript, reviewed the figures, contributed to conception and design, acquisition of data, analysis and interpretation of data. PRN: Contributed to acquisition of data, analysis and interpretation of data and revised the study critically. SAA: Reviewed the manuscript and contributed to acquisition of data. NAK: Contributed to acquisition of data.

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