

CASE REPORT

Open Access

A case of dense pigment deposition of the posterior lens capsule



Igor Šivec Trampuž

Abstract

Background: Pigment dispersion syndrome (PDS) is a well-known entity which can lead to pigmentary glaucoma (PG). This case report presents a rare presentation of PG with bilateral dense pigment deposits of the posterior lens capsule.

Case presentation: A 72-year-old male came for his first appointment due to an asymmetric worsening of visual acuity. The examination showed unilaterally severely increased intraocular pressure, bilateral dense pigment deposition of the posterior lens capsule, and a shallow unilateral optic disk excavation. Gonioscopy revealed moderate pigmentation of the angle and a concave configuration of the peripheral iris in both eyes. The standard slit lamp examination showed no transillumination defects of either iris. Optical coherence tomography showed retinal nerve fiber layer (RNFL) thinning in the peripapillary and macular regions. An antiglaucoma medication was prescribed with a good lowering effect.

Conclusion: Pigment deposition of the posterior lens capsule, which has been rarely reported, is a possible important sign of PDS or PG.

Keywords: Pigmentary dispersion syndrome, Pigmentary glaucoma, Retinal nerve fiber layer, Intraocular pressure, Deposits, Posterior lens capsule, Optic neuropathy

Background

Pigment dispersion syndrome (PDS) is a relatively rare and peculiar entity that can lead to a secondary elevation of intraocular pressure (IOP) and cause pigmentary glaucoma (PG). PDS is characterized by dispersion of pigment of the anterior segment, including Krukenberg's spindles, iris transillumination defects, diffuse trabecular meshwork pigmentation, and a backward bowing of the iris [1–3]. It is hypothesized that the posterior bowing of the iris causes rubbing of the pigmented iris epithelium against lens structures, which in turn causes the liberation of pigment that reduces the outflow of the aqueous in the trabecular meshwork [4–7]. The majority of patients with PDS are asymptomatic; however, episodes of blurred vision, halos around light sources, and headaches

have been described [2, 8]. PDS typically affects myopic patients between 20 and 40 years of age and can be seen equally distributed between men and women [2, 9, 10]. It is reported that it affects Caucasians more often than other races [1, 9, 11]. Many cases are sporadic; however, there are families with apparent autosomal dominant inheritance [12, 13].

Unlike PDS, PG is more prevalent in men [1]. The features are similar to PDS with an additional elevated IOP and glaucomatous optic neuropathy. PG tends to be a high-tension type of glaucoma with 12.5% of patients having an IOP over 39 mmHg at the time of the diagnosis [10, 14]. The probability of conversion from PDS to PG is 10% at 5 years and 15% at 15 years. A lifetime risk of PDS progressing to PG has been estimated between 35 and 50% [10, 15]. Visual field progression is common [10, 14]. Unlike many other forms of glaucoma PG has a tendency to progress to a final quiescent phase (also

Correspondence: Sivec.igor@gmail.com

Morela Okulisti in Optika, Center for Eye Refractive Surgery, Tehnološki Park 21, 1000 Ljubljana, Slovenia



© The Author(s). 2020 **Open Access** This article is licensed under a Creative Commons Attribution 4.0 International License, which permits use, sharing, adaptation, distribution and reproduction in any medium or format, as long as you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons licence, and indicate if changes were made. The images or other third party material in this article are included in the article's Creative Commons licence, unless indicated otherwise in a credit line to the material. If material is not included in the article's Creative Commons licence and your intended use is not permitted by statutory regulation or exceeds the permitted use, you will need to obtain permission directly from the copyright holder. To view a copy of this licence, visit <http://creativecommons.org/licenses/by/4.0/>. The Creative Commons Public Domain Dedication waiver (<http://creativecommons.org/publicdomain/zero/1.0/>) applies to the data made available in this article, unless otherwise stated in a credit line to the data.

known as burn-out phase) with advancing age. Reduced pigment dispersion and IOP normalization have been reported with a reversal of iris transillumination defects [2, 16].

PDS has been reported to occur in 1.9% of a population undergoing glaucoma screening and it has been reported to represent 4.4% of all cases in a glaucoma department. Larger population-based studies would be necessary to enable a more exact estimation [1, 17].

Case presentation

A 72-year old male was referred to our clinic for cataract surgery. The patient's medical history revealed bilateral asymmetric visual acuity worsening in the past 2 years with a generalized blur and no other associated symptoms. He had no previous eye surgery or trauma. His medical and family histories were unremarkable. He had no known allergies and took no regular medication.

The ophthalmic examination showed visual acuity (VA) of 0.4 and 1.0 (Snellen chart) in his right and left eye respectively. The subjective refraction was $-3.50 - 1.00 \times 80$ in the right eye and -2.25 in the left eye. Intraocular pressure measured with a Goldmann applanation tonometer was 48 mmHg in his right eye and 20 mmHg in his left eye. The patient had blue irises. The external examination was normal, but the slit lamp examination

revealed dense asymmetric pigmentation of the posterior lens capsule in both eyes, although the examination of the iris was unremarkable (Fig. 1). Gonioscopy showed bilateral wide-open angles with a moderate diffuse grade 2+ pigmentation using Scheie's grading, a pigmented line at Schwalbe's line similar to Sampaolesi's line, and a concavity of the peripheral iris. Dilated fundus examination showed a clear vitreous and a shallow extensive optic nerve head excavation in his right eye. He had pavingstone degenerative changes in the periphery; the rest of the fundus examination was unremarkable. The visual fields by standard static perimetry of the right eye showed generalized severe depression. Optical coherence tomography (OCT) (SS-OCT; DRI OCT Triton©Topcon, Japan) of his right eye showed generalized severe retinal nerve fiber layer (RNFL) thinning with a total thickness of 43 μm (Fig. 2). The left eye showed mild RNFL thinning with a total thickness of 89 μm . A diagnosis of pigmentary glaucoma of the right eye and pigment dispersion syndrome of the left eye was made.

Due to a severely elevated IOP we decided to prescribe the patient with a prostaglandin analogue (latanoprost once daily) for both eyes and a combination of an α_2 adrenergic agonist and a β adrenergic antagonist (brimonidine tartrate/timolol maleate twice daily) for his right eye. The patient had a good response to topical

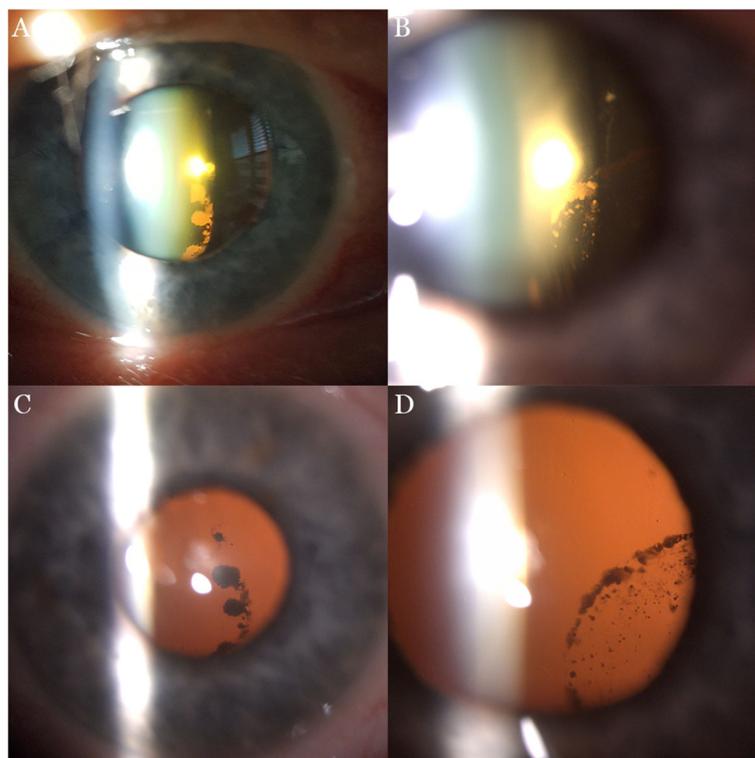
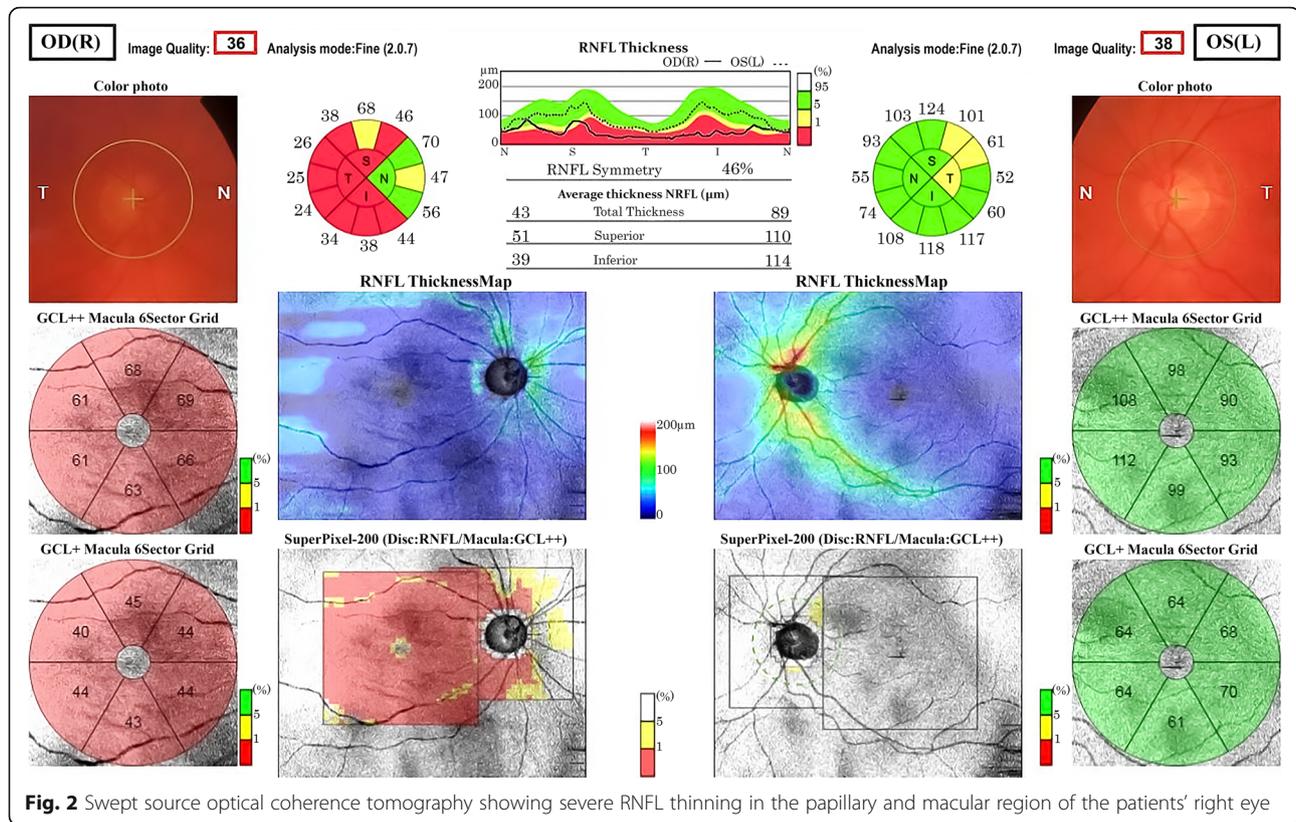


Fig. 1 Slit lamp images. **a** Pigment deposition in right eye. **b** Pigment deposition in left eye. **c** Retroillumination of right eye. **d** Retroillumination of left eye



medication. At his last follow up examination 6 months after the presentation, his VA was 0.5–0.6 and 1.0 and IOP 9 mmHg and 10 mmHg in his right and left eye respectively. OCT showed no additional RNFL thinning; also the visual fields defects were unchanged.

Discussion and conclusion

Pigment deposition of the more central aspect of the posterior lens capsule can be the first sign of pigmentary glaucoma or pigment deposition syndrome. This clinical presentation has been rarely reported. A search of published literature using PubMed and the Cochrane Registry revealed six cases with similar clinical presentation; however, they all described cases that had unilateral pigment deposits. Roberts et al. [18], Lin et al. [19], Turgut et al. [20], and Nagarajaiah et al. [21] described cases of unilateral pigment deposition on the posterior lens capsule without ocular trauma. Al-Mezaine [22] described a case of pigment deposition after blunt ocular trauma. There is only one documented case of bilateral pigment deposition of the posterior lens capsule published by Canestraro et al. [23]. The patient presented in this latter case study had optic nerve hypoplasia which made the assessment of the damage done by the glaucoma difficult. The hypothesized explanations of the etiology of pigment deposits of the posterior lens capsule are very similar across different authors. However, due to only a small number of reported cases, it is still too early to say that we understand the

pathophysiological mechanisms behind this pathological finding. An intact anterior hyaloid face exhibits a firm annular adhesion to the posterior lens capsule at Wieger's ligament [24]. This forms a natural barrier which prevents pigment from traveling from the posterior chamber to the polar retrolental space. The pigmentation of the lens capsule is classically in the periphery in the shape of Scheie's or Zentmayer's line in pigment dispersion syndrome [25–27]. This means that an anatomical anomaly or a defect in Wiegers ligament would have to be present and for the aqueous from the posterior chamber to be able to flow into the retrolental space [18–20, 22]. Canestraro et al. [23] proposed that anatomical changes due to the proximity of the posterior zonulae insertions and Wieger's ligament are to be blamed.

The identification of pigment deposition of the posterior lens capsule is important especially if other signs of PDS are subtle. This is especially true if the patient has no transillumination defects or Krukenberg's spindle. One could easily give the patient a diagnosis of primary open angle glaucoma; however, it has a different natural course and surgical treatment modalities from PG which makes it an important entity to diagnose correctly.

Abbreviations

PDS: Pigment dispersion syndrome; PG: Pigmentary glaucoma; IOP: Intraocular pressure; RNFL: Retinal nerve fiber layer; OCT: Optical coherence tomography

Acknowledgements

Not applicable.

Author's contributions

The author(s) read and approved the final manuscript.

Funding

The author does not receive any financial support for the publication of this manuscript.

Availability of data and materials

All data generated or analysed during this study are included in this article.

Ethics approval and consent to participate

Not applicable.

Consent for publication

The written consent to publish this case report with images and clinical information was obtained from the patient.

Competing interests

The author has no competing interests.

Received: 25 June 2020 Accepted: 10 November 2020

Published online: 19 November 2020

References

- Scheie HG, Cameron JD. Pigment dispersion syndrome: a clinical study. *Br J Ophthalmol*. 1981;65:264–9.
- Niyadurupola N, Broadway DC. Pigment dispersion syndrome and pigmentary glaucoma – a major review. *Clin Exp Ophthalmol*. 2008;36(9):868–82.
- Scuderi G, Contestabile MT, Scuderi L, Librando A, Fenicia V, Rahimi S. Pigment dispersion syndrome and pigmentary glaucoma: a review and update. *Int Ophthalmol*. 2018;39(7):1651–62.
- Campbell DG. Pigmentary dispersion and glaucoma: a new theory. *Arch Ophthalmol*. 1979;97:1667–72.
- Potash SD, Tello C, Liebmann J, Ritch R. Ultrasound biomicroscopy in pigment dispersion syndrome. *Ophthalmol*. 1994;101(2):332–9.
- Ritch R. A unification hypothesis of pigment dispersion syndrome. *Trans Am Ophthalmol Soc*. 1996;94:381–409.
- Okafor K, Vinod K, Gedde SJ. Update on pigment dispersion syndrome and pigmentary glaucoma. *Curr Op Ophthalmol*. 2017;28(2):154–60.
- Sivaraman KR, Patel CG, Vajaranant TS, Aref AA. Secondary pigmentary glaucoma in patients with underlying primary pigment dispersion syndrome. *Clin Ophthalmol*. 2013;7:561–6.
- Sugar HS. Pigmentary glaucoma: a 25-year review. *Am J Ophthalmol*. 1966;62:499–507.
- Siddiqui Y, Ten Hulzen RD, Cameron JD, Hodge DO. What is the risk of developing pigmentary glaucoma from pigment dispersion syndrome? *Am J Ophthalmol*. 2003;135:94–9.
- Farrar SM, Shields MB. Current concepts in pigmentary glaucoma. *Surv Ophthalmol*. 1993;37(4):233–52.
- Tandon A, Zhang Z, Fingert JH, Kwon YH, Wang K, Alward WLM. The heritability of pigment dispersion syndrome and pigmentary glaucoma. *Am J Ophthalmol*. 2019;202:55–61.
- Lahola-Chomiak AA, Walter MA. Molecular genetics of pigment dispersion syndrome and pigmentary glaucoma: new insights into mechanisms. *J Ophthalmol*. 2018;2018:5926906.
- Migliazzo CV, Shaffer RN, Nykin R, Magee S. Long-term analysis of pigmentary dispersion syndrome and pigmentary glaucoma. *Ophthalmol*. 1986;93:1528–36.
- Goyeneche HFG, Hernandez-Mendieta DP, Rodriguez DA, Sepulveda AI, Toledo JD. Pigment dispersion syndrome progression to pigmentary glaucoma in a Latin American population. *J Curr Glaucoma Pract*. 2015;9(3):69–72.
- Speakman JS. Pigmentary dispersion. *Br J Ophthalmol*. 1981;65:249–51.
- Ritch R, Steinberger D, Liebmann JM. Prevalence of pigment dispersion syndrome in a population undergoing glaucoma screening. *Am J Ophthalmol*. 1993;115(6):707–10.
- Roberts DK, Miller E, Kim LS. Pigmentation of the posterior lens capsule central to Wieger's ligament and the Scheie line: a possible indication of the pigment dispersion syndrome. *Optom Vis Sci*. 1995;72(10):756–62.
- Lin DY, Volpicelli M, Singh K. Dense pigmentation of the posterior lens capsule associated with the pigment dispersion syndrome. *J Glaucoma*. 2003;12(6):491–3.
- Turgut B, Türkçüoğlu P, Deniz N, Catak O. Annular and central heavy pigment deposition on the posterior lens capsule in the pigment dispersion syndrome: pigment deposition on the posterior lens capsule in the pigment dispersion syndrome. *Int Ophthalmol*. 2007;6:441–5.
- Nagarajaiah S, Shun-Shin GA. Pigment deposition on the central aspect of the posterior lens capsule in pigmentary dispersion. *Digit J Ophthalmol*. 2011;17(4):69–71.
- Al-Mezaine HS. Central posterior capsule pigmentation in a patient with pigment dispersion and previous ocular trauma: a case report. *Indian J Ophthalmol*. 2010;58(4):336–7.
- Canestraro J, Sherman J. Curvilinear, symmetrical, and profound pigment deposition on the posterior lens capsule in a patient with bilateral pigmentary dispersion syndrome. *Eye Brain*. 2018;10:79–84.
- Hilding AC. Normal vitreous, its attachments and dynamics during ocular movement. *Arch Ophthalmol*. 1954;52:47–514.
- Zentmayer W. Association of an annular band of pigment on posterior capsule of lens with a Krukenberg spindle. *Arch Ophthalmol*. 1938;20:52–7.
- Vail D. The Zonule of Zinn and ligament of Wieger. Their importance in the mechanics of intracapsular extraction of cataract. *Trans Ophthalmol Soc UK*. 1957;77:441–99.
- Lichter PR, Shaffer RN. Diagnostic and prognostic signs in pigmentary glaucoma. *Trans Am Acad Ophthalmol Otolaryngol*. 1970;74:984–98.

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Ready to submit your research? Choose BMC and benefit from:

- fast, convenient online submission
- thorough peer review by experienced researchers in your field
- rapid publication on acceptance
- support for research data, including large and complex data types
- gold Open Access which fosters wider collaboration and increased citations
- maximum visibility for your research: over 100M website views per year

At BMC, research is always in progress.

Learn more biomedcentral.com/submissions

