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Case Report

Unilateral Congenital Lenticular Pigmentation

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Keywords
Lenticular pigmentation · Pigmentary glaucoma · Pigment dispersion syndrome

Abstract
Introduction: Release of pigments in the anterior chamber is frequently observed in pigment dispersion syndrome, an autosomal dominant disorder marked by bilateral pigment deposition on the anterior and possibly posterior lens capsule, zonules of the lens, trabecular meshwork, and corneal endothelium, in addition to radial, spoke-like transillumination defects in the mid peripheral iris [J Ayub Med Coll Abbottabad. 2017;29(3):412–414 and Optom Vis Sci. 1995;72(10): 756–762]. Pigmentation of the anterior lens surface has also been associated with intraocular inflammation, pseudoexfoliation syndrome, siderosis, antipsychotic medication usage, and remnants of the tunica vasculosa lentis [Br J Ophthalmol. 1998;82(11):1344]. ***Case Presentation:*** A 23-year-old female presented to our eye clinic with chief complaint of mild blurring of vision in the right eye and inquired about refractive surgery. The patient denied any previous history of ocular inflammation, trauma, surgery, or use of topical or systemic medications. Slit-lamp examination of the right eye anterior segment was within normal limits except for the crystalline lens anterior capsular which showed confluent pigment deposits stellate in shape over the pupillary axis, whereas left eye examination was completely within normal limits. Ophthalmic examination of the posterior segment was normal in both eyes. Based on her previous ophthalmic history and slit-lamp examination of the right eye, a diagnosis of unilateral congenital lenticular pigmentation was made. ***Conclusion:*** Congenital lenticular pigmentation is a rare benign entity carrying no surgical indications with a relatively good visual response to optical correction. Recognition of this rare benign condition would add to the ophthalmologist's differential of ocular pigmentation and avoid unnecessary concern and follow-up in more potentially progressive disorders such as pigmentary glaucoma.

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Introduction

Release of pigments in the anterior chamber is frequently observed in pigment dispersion syndrome, an autosomal dominant disorder marked by bilateral pigment deposition on the anterior and possibly posterior lens capsule, zonules of the lens, trabecular meshwork, and corneal endothelium, in addition to radial, spoke-like transillumination defects in the mid-peripheral iris [1, 2]. Pigmentation of the anterior lens surface has also been associated with intraocular inflammation, pseudoexfoliation syndrome, siderosis, antipsychotic medication usage, and remnants of the tunica vasculosa lentis [3]. In this case report, we describe a rare ophthalmic entity presented as unique unilateral pigmentation of the anterior lens surface in a young myopic woman who otherwise, has a normal ophthalmic examination. We feel that recognition of this rare benign condition would add to the ophthalmologist's differential diagnosis of unilateral pigmentation and avoid unnecessary concern and follow-up in more potentially progressive disorders such as pigmentary glaucoma. To the best of our knowledge, this is the fourth report of congenital lenticular pigmentation, either unilateral or bilateral congenital lenticular pigmentation.

Case Presentation

A 23-year-old female presented to our eye clinic with chief complaint of mild blurring of vision in the right eye and inquired about refractive surgery. The patient denied any previous history of ocular inflammation, trauma, surgery, or use of topical or systemic medications. Snellen's visual acuity in the right eye was 0.05, which was corrected to 0.5 with −3.00 to 2.00 × 15 and from 0.5 in the left eye to 0.8 with plano −0.75 × 170. Intraocular pressure was 14 mm Hg in the right and 14 mm Hg the in left eye. Slit-lamp examination of the right eye anterior segment showed clear cornea, deep and quiet anterior chamber without iris transillumination defects, whereas crystalline lens anterior capsular showed confluent pigment deposits stellate in shape over the pupillary axis. Dilation the of right eye pupil showed no pigment deposits in the peripheral capsule, zonules, or on the posterior lens capsule or anterior hyaloid face (Fig. 1a, c). The examination of the left eye anterior segment was within normal limit before and after dilation. Gonioscopy revealed open angle with mild pigmentation and prominent iris processes of the right eye, whereas left eye showed open angle without remarkable findings (Fig. 2). Ophthalmic examination of the posterior segment was normal in both eyes. The patient was reassured and prescribed glasses, and booked for regular follow-up. Following discussion with the patient, she opted to have transepithelial photorefractive keratectomy for the right eye myopic astigmatism which improved her unaided vision to 0.8. Based on her previous ophthalmic history and slit-lamp examination of the right eye, a diagnosis of unilateral congenital lenticular pigmentation was made.

Discussion

In the absence of signs of intraocular inflammation and other causes of pigmentary dispersion, it has been postulated that the pigmented cells were implanted on the lens surface in utero from the developing iris pigment epithelium. Migration of some of these implanted cells into the visual axis is a possibility and they appear nonprogressive and visually insignificant. It is known that paraxial pigmentation is less common than radial distribution [3]; our case shows particularly unusual finding of axial pigment deposits. Eyes with pigmentary glaucoma are more prone to irido-lenticular touch, reverse pupillary block, and backward

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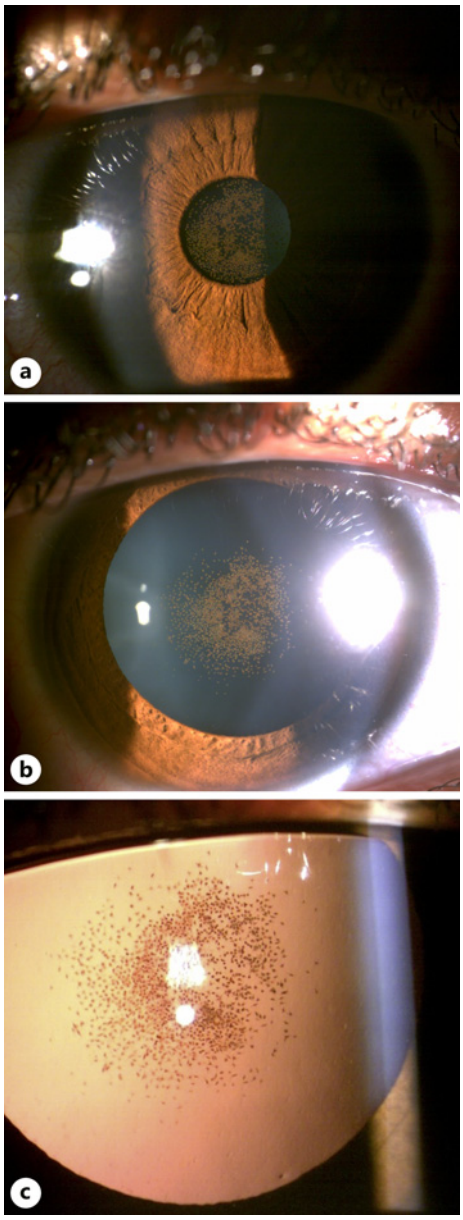

Fig. 1. a–c Slit-lamp photograph of the right eye showing central pigmented cells deposition over the anterior crystalline lens capsule (before dilation (**a**), after dilation (**b**), and retro-illumination (**c**)).

posterior bowing. These eyes are more likely to be bilateral, mainly male, and myopic. A reverse pupillary block might occur in eyes at risk as a result of physiological processes such as accommodation, blinking, exercise, head motions, or certain head positions. This results in high AC pressure, which further causes iris apposition to the surface of the anterior lens, raising IOP [2, 4]. Spindle-shaped brownish pigmentary deposits are observed on the anterior lens capsule during slit-lamp examination in patient with congenital lenticular pigmentation. This rare condition is a benign entity carrying no surgical indications with a relatively good visual response to optical correction [5]. However, it is important to note that Mangan et al. [6] reported a case of an eight-year-old girl with a unilateral epicapsular star and secondary visual impairment. In this case report, we describe a rare ophthalmic entity – unilateral pigmentation of the anterior lens surface – in a young myopic woman who otherwise, has a normal ophthalmic examination. Recognition of this rare benign condition would add to the ophthalmologist's differential of ocular pigmentation and avoid unnecessary concern and

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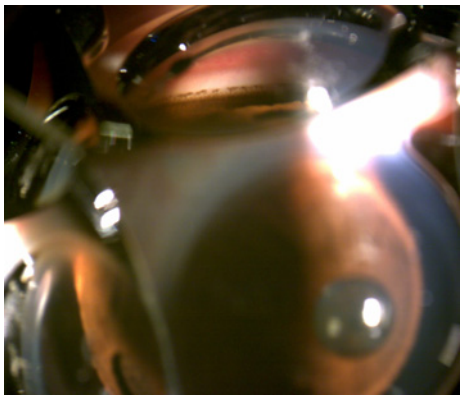

Fig. 2. Gonioscopy of the right eye demonstrating open angle with patchy pigmentation at, and anterior, to Schwalbe's line along with prominent iris processes.

follow-up in more potentially progressive disorders such as pigmentary glaucoma. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000534927).

Conclusions

Congenital lenticular pigmentation is a rare benign entity carrying no surgical indications with a relatively good visual response to optical correction. Recognition of this rare benign condition would add to the ophthalmologist's differential of ocular pigmentation and avoid unnecessary concern and follow-up in more potentially progressive disorders such as pigmentary glaucoma.

Acknowledgments

The patient's cooperation and the assistance of Dr. Doaa Alnihmi in obtaining clinical photos are appreciated by the authors.

Statement of Ethics

Research was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. This study protocol was reviewed and approved by the Saggaf Eye Center Research Ethics Committee; approval number 405. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

There are no conflicts of interest.

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Author Contributions

All authors contributed to the conception or design of the work and acquisition of the data; drafting the work and revising it critically for intellectual content; and final approval of the manuscript for publication. Yahya Al-Najmi wrote the concept and performed data analysis and interpretation. Abdulrahman Alsaggaf and Maram E. A. Abdalla Elsayed, co-writing of the manuscript and literature search. Mohammed Albeedh contributed to critical review of the article and reviewed the manuscript for final approval.

Data Availability Statement

All data generated or analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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