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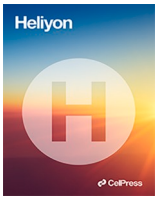

Case report

Macrophakia: The characterization of a novel lens condition

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ABSTRACT

Purpose: To present a novel characterization of a lens condition termed ***macrophakia*** which includes large or proportionally large lenses with shallow anterior chamber dimensions that are statistically deviant of a normal population.

Observations: We identified five eyes from three cases to have significantly large lens parameters and small anterior chamber depths. In all five eyes, the anterior chamber depth was less than 2 mm and the anterior chamber depth to lens thickness ratio was two standard deviations outside the normative range of lens and chamber measurements. These large lenses were all observed in the absence of typical pathology and other biometric abnormalities.

Conclusions and importance: A novel lens characterization termed ***macrophakia*** is proposed to describe large or proportionately large lenses. Naming this condition is clinically relevant and will enhance cataract evaluations and patient education.

1. Introduction

The human anterior chamber and crystalline lens differentially grow with age. In certain conditions, a statistically smaller anterior chamber depth (ACD) coupled with a growing lens can result in angle closure. In other situations, abnormalities in the lens-zonular apparatus can be predicted by careful slit lamp examination and irregularities on optical biometry. Herein, we present three cases with abnormally large lenses in the context of anterior chamber dimensions in the absence of any known pathology.

2. Findings

2.1. Case 1

A 53-year-old white female was originally referred to our glaucoma clinic for evaluation and management of bilateral narrow angles.

She had a history of myopia, chronic uveitis, and dry eyes. There was no history of previous ocular surgeries, trauma, or glaucoma. She had no family history of glaucoma. She had a remote, but not current, history of topiramate use, and no history of sulfa drug use. Patient was myopic and her refraction was OD: $-5.50 + 2.75 \times 180$ and OS: $-4.25 + 2.00 \times 165$.

On examination, best corrected visual acuity was 20/25 in both eyes. Intraocular pressures (IOPs) were 12 mm Hg in both eyes. Anterior segment exam revealed shallow anterior chambers bilaterally. Gonioscopy demonstrated 270° of appositional closure bilaterally. Ultrasound biomicroscopy (UBM) demonstrated pupillary block but was otherwise negative for effusions, masses, or

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anterior rotation of the ciliary body. Optical biometry revealed axial lengths (AL) of 24.06 mm and 24.18 mm and anterior chamber depths of 2.16 mm and 2.17 mm. RNFL was intact. Anterior segment OCT further showed shallow chambers with a large anterior portion of the lens (Fig. 1A and B).

Patient was diagnosed with atypical secondary angle closure due to her normal axial length, myopic refraction but extremely anterior lens presence, and short anterior chamber depth (ACD).

Laser peripheral iridotomies (LPI) were completed. Post-iridotomy gonioscopy showed the angle open to pigmented trabecular meshwork 360° circumferentially.

The patient presented to the glaucoma clinic approximately 4 years later for cataract evaluation. Cataracts were visually significant bilaterally, and both graded as 1–2+ nuclear sclerotic (NS) with central posterior subcapsular cataract (PSC). Biometry was obtained and revealed shallow ACDs (Table 1).

Cataract extraction and insertion of the intraocular lens of the right eye was completed without complications. Prior to surgery, there was concern that this was in fact a case of microspherophakia, but during surgery, with excellent dilation, there was no zonulopathy and the lens had a typical equatorial diameter, but also a very large anterior-posterior diameter.

Prior to cataract surgery of the left eye, slit lamp photos and repeat anterior segment OCT were obtained, as seen in Fig. 2A and B.

2.2. Case 2

A 64-year-old white male was referred to our clinic by his primary care provider for glaucoma and cataract evaluation.

Ocular history was significant for Fuchs Dystrophy and a prior LPI in the left eye. The patient had no other history of ocular surgeries. He had no history of topiramate or sulfa-drug use. The patient had a positive family history of glaucoma in his mother and sister. The patient's refraction was OD: plano $+1.00 \times 073$ and OS: $-2.00 + 2.00 \times 093$, and he did not experience any recent shifts in vision.

Best corrected visual acuity was 20/25 and 20/40 in the right and left eye, respectively. IOPs were 12 and 11 mm Hg. Mild guttata were seen bilaterally. Anterior segment exam revealed shallow anterior chambers. Cataracts were visually significant in both eyes and both graded as 2.5+ NS.

Gonioscopy demonstrated 360° of appositional closure bilaterally. RNFL was intact. Biometry revealed shallow ACD (Table 1). Axial lengths were 22.46 mm and 22.74 mm, OD and OS respectively.

Cataract surgery was completed without complications. However, a full appreciation of the proximity of the lens to the cornea was noted when the paracentesis blade nearly penetrated the lens capsule immediately upon entry into the anterior chamber. Vistante OCT was obtained prior to surgery of the right eye, revealing a remarkably shallow anterior chamber (AC) in the absence of other known pathology or insult (Fig. 3).

2.3. Case 3

A 72-year-old white male presented to our clinic for cataract evaluation. He was being followed by an outside provider for suspicion of glaucoma. Medical history was extensive and notable for insulin-independent type 2 diabetes and meningioma resection. Ocular surgical history included strabismus surgery in the right eye and bilateral upper lid blepharoplasty. He had no known history of topiramate or sulfa drug use.

Family history was notable for glaucoma and macular degeneration. The patient was myopic and refraction was OD: $-1.00 + 2.00 \times 178$ and OS: $-4.25 + 1.75 \times 001$.

Best corrected visual acuity was 20/40 -2 and 20/50. IOPs were 14 and 13 mm Hg. Guttata was noted bilaterally. Anterior segment exam and OCT revealed shallow anterior chambers (Fig. 4A and B). Cataracts were visually significant in both eyes and graded as 2+ NS and 2+ cortical cataract with cortical spokes. Biometry revealed shallow ACD in the left eye (Table 1). Axial lengths were 23.52 mm and 23.57 mm, OD and OS respectively.

Cataract surgery was completed in both eyes. Once again, as in case 2, a full appreciation of the proximity of the lens to the cornea was noted when the paracentesis blade nearly penetrated the lens capsule immediately upon entry into the anterior chamber. Iris immediately prolapsed into the paracentesis. The left eye cataract extraction post-operative course was complicated by Irvine-Gass syndrome.

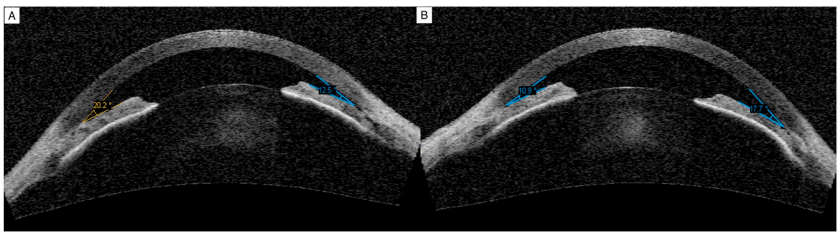

Fig. 1. a. Anterior segment OCT of the right eye revealing a shallow anterior chamber and prominent anterior aspect of the lens.
b. Anterior segment OCT of the left eye revealing a shallow anterior chamber and prominent anterior aspect of the lens.

Table 1
Biometry values for case 1, case 2, and case 3.

	Case 1 OD	Case 1 OS	Case 2 OD	Case 2 OS	Case 3 OD	Case 3 OS
Age/Gender	58/Female		67/Male		74/Male	
Previous Disease	Uveitis, nonspecific autoimmune disease, HLD, OSA, HTN, mild MR and TR, fibromyalgia		Fuchs Dystrophy, HTN, HLD, melanoma, coronary artery disease, neuropathy		Fuchs dystrophy, dry eye syndrome meningioma resection, DM, HTN, HLD, sleep apnea, diverticulosis	
Axial Length (AL)	24.16 mm	24.26 mm	22.46 mm	22.74 mm	23.52 mm	23.57 mm
Anterior Chamber Depth (ACD)	1.71 mm	1.84 mm	1.89 mm	1.89 mm	2.22 mm	1.83 mm
Lens Thickness (LT)	6.49 mm	6.36 mm	4.99 mm	5.39 mm	5.22 mm	5.00 mm
ACD/LT Ratio	0.263	0.289	0.379	0.351	0.425	0.366

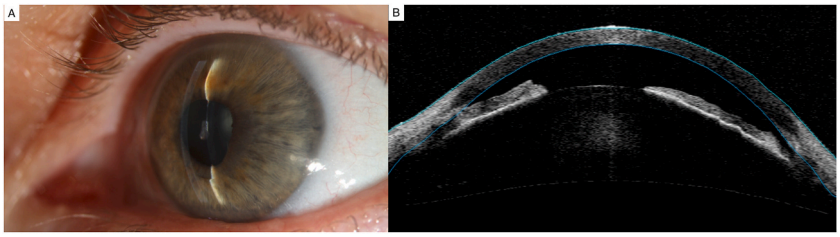

Fig. 2. a. Slit lamp photograph of the left eye revealing a markedly shallow anterior chamber.
b. Anterior segment OCT of the left eye highlighting a similarly shallow anterior chamber and prominent anterior aspect of the lens.

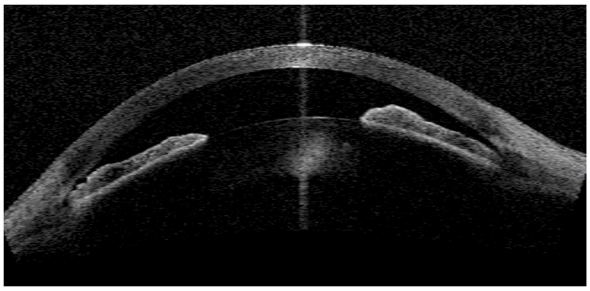

Fig. 3. Anterior segment OCT of the right eye revealing a shallow anterior chamber and prominent anterior aspect of the lens.

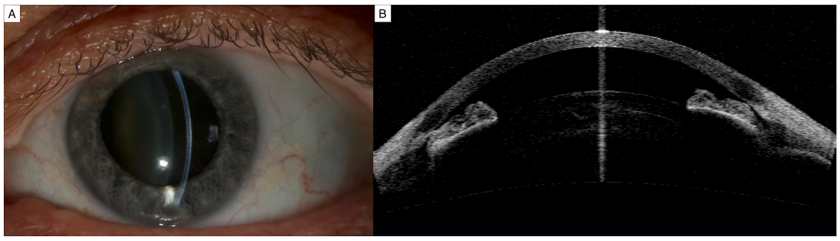

Fig. 4. a. Slit lamp photograph of the left eye revealing a markedly shallow anterior chamber.
b. Anterior segment OCT of the left eye revealing a shallow anterior chamber and prominent anterior aspect of the lens.

3. Discussion

Noting optical biometry is crucial in cataract surgery evaluation. Conditions such as microspherophakia or nanophthalmos can result in complex surgeries, and for this reason, characterizing unique anatomic conditions is advantageous to the cataract surgeon. Our three mentioned cases demonstrate unusually large lenses, either in their absolute size or in their relative size within the anterior chamber in the absence of other pathology. We used a reference study from Popov et al. which used the Lenstar LS900 to measure biometry in a Caucasian population and found average eye measurements as follows: AL: 23.33±1.01, lens thickness (LT): 4.57± 0.43, and ACD: 3.08 ± 0.43 [1]. All our cases had measurements that fell well beyond two standard deviations of a normal distribution of lens and chamber measurements. After literature review, we concluded that such dimensions in our subjects have been previously

undefined. We defined a novel condition termed ***macrophakia***.

We defined a small ACD:LT as a criterion for macrophakia. A *small* ratio was a *shallow ACD* (two SD below average ACD) and a *thick lens* (two SD above average LT). From the reference study, a normative value of this ratio is 0.674. The smallest ratio still within the normative range is 0.409. Case 1 had ratios of OD: 0.263 and OS: 0.289. Case 2 had ratios of OD: 0.379 and OS: 0.351. Case 3 OS had a value of 0.366.

In addition to ACD:LT ratio, we consider an ACD of less than 2 mm as a criterion for macrophakia. All five of our eyes fit this parameter.

We consider macrophakia as a physiologic state that can lead to a disease state or complicated anterior segment surgery. As such, we excluded from this condition of macrophakia any other pathological state that could have led to these lens parameters. For example, nanophthalmos and microspherophakia can present with shallow anterior chambers and a high lens/eye volume ratio, but our subjects' normal axial length and equatorial diameter eliminated such diagnoses [2]. Ciliary body disorders such as cysts, effusions, and masses as well as lens abnormalities such as lenticonus, trauma, intumescent and hypermature cataracts, and any zonulopathy or capsular violations were also excluded. These disorders were not noted clinically; however, one limitation includes the lack of UBM imaging available in cases 2 and 3, and will be a worthwhile addition in noting future cases of macrophakia.

Diabetes also plays a role in changing lens parameters. Eyes in type one diabetics have been found to have smaller ACDs and greater lens thickness, but smaller equatorial diameters, making the lens more round [3]. In addition, periods of hyperglycemia can also cause lenticular swelling. Excess glucose is converted to sorbitol in the lens and accumulates due to its poor permeability. This leads to osmotic changes that cause lens fibers to degenerate. The accumulated sorbitol creates a hyperosmotic gradient, resulting in the infusion of fluid and swelling. This osmotic stress is implicated in cataract formation and intumescence with longstanding poor glycemic control [4].

This did not confound measurements of Case 1 and 2. Case 3 did have a diagnosis of type 2 diabetes, but it was noted to be diet-controlled and not insulin dependent. Furthermore, the patient had no noted history of hyperglycemic episodes that may lead to intumescence in the lens. For this reason, we did not consider his diagnosis of diabetes to play a role in his abnormal lens parameters.

The refractive error of the eyes was also a point of interest. Shallow anterior chambers tend to be seen more in hyperopic eyes, while myopic eyes tend to have deep chambers [5]. Our study eyes were myopic and emmetropic. Our subjects' shallow chambers were not in line with typical myopic eyes. When there is presence of a shallow anterior chamber and a myopic shift, there is a strong concern for a ciliary body effusion, lens dislocation, or malignant glaucoma. However, none of those situations applied in either of these cases. Further, variants of hyperopia and nanophthalmos are described with normal and abnormal intraocular dimensions. Clinically, these patients have lenses that are too large for their eye in an otherwise non-pathologic sense. However, these conditions are well-known and therefore we consider them to be a different entity to the patients described above.

One additional factor to consider is age. Lens thickness is known to increase with time [6,7]. Equatorial epithelial cells in the lens continue to proliferate into new fiber cells. These are then added in concentric layers around the older fiber cells. In this way, the lens continues to grow as we age [8].

As seen in Case 1, the ACD decreased when the patient returned for cataract surgery four years after initial presentation. Though we did not have lens thickness data from her initial presentation, we can hypothesize that the anterior chamber depth decrease was due to her lens thickness increasing, especially since she had a patent peripheral iridotomy. Even when stratifying by age and comparing it to the reference study, our cases' ACD:LT ratios still fall below the range of normative values. However, we may consider that this definition of macrophakia may progress as a patient ages and lens thickness increases. One limitation of this study includes the lack of biometric data from our cases when they were younger. In fact, we suspect that the ACD:LT ratio may have been normal at some point earlier in their lives. However, the fact that lenses grow into this size and ratio does not preclude this as a separate diagnostic entity.

4. Conclusion

We present three cases in which we define a novel entity known as ***macrophakia***. Our conditions for this condition include ACD: LT ratio two standard deviations below the normal means (as found by the Lenstar study in the Caucasian population) and an anterior chamber depth of less than 2 mm, while meeting inclusionary criteria as documented above. This characterization will be advantageous for the cataract surgeon when describing eyes with these unique anatomic parameters, as well as for patient education. In addition, by formally naming this condition, future cases of macrophakia can also be noted, and additional imaging and testing can be obtained for further characterization.

Patient consent

Signed patient consent was obtained for all three cases.

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Ethics declaration

The UNC IRB granted this research exemption status. All participants provided informed, written consent to participate in the

study.

Data availability

Data in this study has not been deposited into a publicly available repository. Data is included in the article.

CRediT authorship contribution statement

Gulrukh Shaheen: Writing – review & editing, Writing – original draft, Data curation. **Joshua Paul:** Writing – review & editing, Writing – original draft, Investigation, Conceptualization. **David Fleischman:** Writing – review & editing, Writing – original draft, Supervision, Investigation, Conceptualization.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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None

References

[1] I. Popov, I. Waczulikova, J. Stefanickova, J. Valaskova, D. Tomcikova, H. Shiwani, et al., Analysis of biometric parameters of 2340 eyes measured with optical biometer Lenstar LS900 in a Caucasian population, Eur. J. Ophthalmol. 32 (1) (2022) 213–220.
[2] N. Ally, S. Ismail, H.D. Alli, Prevalence of complications in eyes with nanophthalmos or microphthalmos: protocol for a systematic review and meta-analysis, Syst. Rev. 11 (1) (2022) 25.
[3] X. Adnan, M. Suheimat, N. Efron, K. Edwards, N. Pritchard, A. Mathur, et al., Biometry of eyes in type 1 diabetes, Biomed. Opt. Express 6 (3) (2015) 702–715.
[4] A. Pollreisz, U. Schmidt-Erfurth, Diabetic cataract-pathogenesis, epidemiology and treatment, J. Ophthalmol. 2010 (2010) 608751.
[5] X. Liu, H. Ye, Q. Zhang, X. Cai, W. Yu, S. Yu, et al., Association between myopia, biometry and occludable angle: the Jiangning Eye Study, PLoS One 11 (10) (2016) e0165281.
[6] D.A. Atchison, E.L. Markwell, S. Kasthurirangan, J.M. Pope, G. Smith, P.G. Swann, Age-related changes in optical and biometric characteristics of emmetropic eyes, J. Vis. 8 (4) (2008) 1–20.
[7] K. Richdale, M.A. Bullimore, K. Zadnik, Lens thickness with age and accommodation by optical coherence tomography, Ophthalmic Physiol. Opt. 28 (5) (2008) 441–447.
[8] C. Cheng, J. Parreno, R.B. Nowak, S. Biswas, K. Wang, M. Hoshino, et al., Age-related changes in eye lens biomechanics, morphology, refractive index and transparency, Aging (Albany NY) 11 (24) (2019) 12497–12531.