American Journal of Ophthalmology Case Reports 11 (2018) 119–120

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.elsevier.com/locate/ajoc

Bilateral, presumed congenital ectropion uveae in a patient with pathologic myopia

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ARTICLE INFO

Keywords:
Ectropion uvea
Pathologic myopia
Lattice degeneration
Congenital

1. Clinical scenario

A 24-year-old woman with bilateral pathologic myopia (spherical equivalent [SE], −21.0 diopters in both eyes) and lattice degeneration was referred for retinal evaluation. She presented with 20/200 vision in both eyes secondary to myopic maculopathy. Intraocular pressure, pupils, visual fields, and motility were normal. Slit lamp examination was significant for bilateral 360-degree ectropion uveae without iris crypts or abnormal vessels (Fig. 1; arrows). The patient reported having this iris finding for as long as she could remember. She had no other medical history and did not have a personal or family history of glaucoma or neurofibromatosis. Gonioscopy was normal (i.e., no anterior synechiae, abnormal iris insertion or dysgenesis, or neovascularization). Over the subsequent six years of follow-up, the patient underwent multiple surgeries and in-office procedures for retinal detachment repair in both eyes. She required a lensectomy and was left aphakic during repair of re-detachment in the right eye.

2. Discussion

Ectropion uveae results from displacement of the posterior pigmented iris epithelium onto the anterior iris surface.[1] It is most commonly acquired, but may be seen in isolation, as part of congenital iris ectropion syndrome, or in association with syndromes of anterior segment dysgenesis or with systemic syndromes.

Acquired ectropion uveae develops due to a contractile neovascular membrane and typically results in iris atrophy and changes to the iris sphincter and dilator muscles.[2] This neovascular membrane develops most commonly secondary to proliferative diabetic retinopathy, retinal vein occlusion, or ocular ischemic syndrome, although other etiologies such as inflammation, trauma, neoplasm, or intraocular surgery should also be considered. The iris may be irregular and poorly reactive due to posterior synechiae and iris muscle abnormalities.

Congenital ectropion uveae (CEU) is rare, non-progressive, and is typically unilateral, although bilateral cases have been reported.[1] It results from iris pigment hyperplasia extending onto the anterior iris surface, which may be caused by endothelium over-activity and/or delayed developmental arrest of neural crest cells (NCC). The iris typically remains round and reactive, and has a smooth, glassy, and cryptless appearance. This condition is associated with glaucoma and mild ipsilateral ptosis due to Müller's muscle dysfunction (NCC origin).

3. Conclusion

Ectropion uveae represents anteriorization of pigmented epithelium onto the iris surface. It is most commonly seen in eyes with iris neovascularization (i.e., acquired), which is managed according to the specific etiology. Congenital ectropion uveae is a distinct subtype with a specific patient population that typically develops glaucoma from drainage angle dysgenesis. It is important to closely follow these patients, as glaucoma may not develop until adulthood.

Based on the non-progressive nature and morphological characteristics of the iris, our patient was diagnosed with isolated, bilateral, presumed CEU. After a thorough literature review, we believe this is the

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https://doi.org/10.1016/j.ajoc.2018.06.017
Received 5 January 2018; Received in revised form 22 April 2018; Accepted 19 June 2018
Available online 20 June 2018

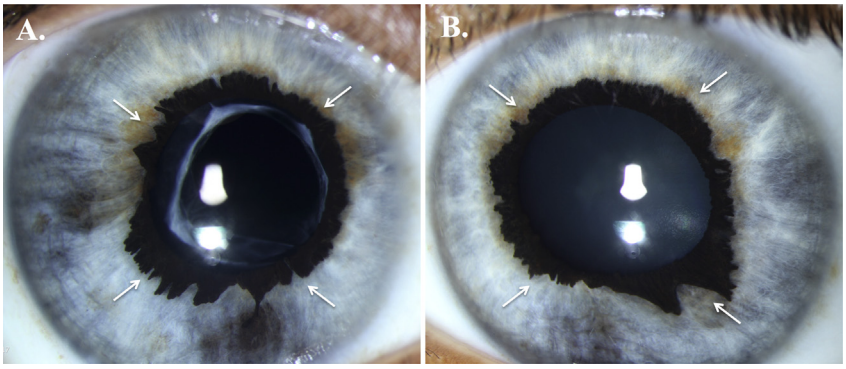

Fig. 1. Three-hundred-sixty degree ectropion uveae in the right (A) and left (B) eyes. The irides are characterized by a glassy and smooth appearance without obvious crypts or abnormal vessels. The right eye (A) is aphakic while the left eye (B) is phakic.

first reported association of presumed CEU with pathologic myopia. A similar case of unilateral CEU in a patient with vitreoretinal degeneration (SE, −6.0 diopter) has been described.[3] Although it is tempting to hypothesize how CEU can be associated with pathologic myopia, an underlying unifying abnormality that connects these two findings has not been identified.

Acknowledgements and disclosures

Funding: The Heed Ophthalmic Foundation (AMK).

Conflicts of interest: The following authors have no financial disclosures: AMK, BJK.

Authorship: All authors attest that they meet the current ICMJE criteria for authorship.

Acknowledgements: None.

Patient consent: Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.ajoc.2018.06.017.

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