Accessory iris membranes: Removing the veil

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ABSTRACT
Purpose: To report a case of a young adult with dense bilateral accessory iris membranes (AIMs).

Observations: AIMs can influence vision by multiple mechanisms. We discuss clinical examination and imaging considerations that can help parse optical and refractive complications to better guide intervention discussions. We also describe our surgical approach and perioperative management to help minimize trauma to the eye and maximize favorable surgical outcomes in these cases.

Conclusions and importance: This case highlights the excellent symptomatic, visual acuity and stereopsis gains that can be achieved following surgical intervention for this clinical entity, even in older patients.

1. Introduction

Up to 95% of newborns display incomplete involution of the anterior tunica vasculosa lentis. These are usually diaphanous, do not affect acuity, and regress by 1-year of age. Rarely, bilateral dense membranes covering much of the pupil remain after birth and fail to involute. These may represent a clinically-distinct entity caused by hyperplasia of the superficial mesenchymal layer of the anterior iris stroma, termed accessory iris membranes (AIMs). They are usually sporadic, but autosomal dominant inheritance can occur.

AIMs may influence vision through multiple mechanisms that can often be parsed. Deprivation amblyopia is nonprogressive in adulthood and typically unaffected by dilation due to drops or changes in ambient lighting. Severity is often modest, as small openings transmit light to the developing retina. Small apertures may even improve acuity by ameliorating uncorrected refractive error, either equally in all meridians via a pinhole effect when they are round, or preferentially in all-but-one meridian via a stenopac effect when they are ovoid; these effects too should not change over time, but may diminish with dilation. Excessively small apertures may impair acuity by diffraction, chromatic aberration and/or reduced retinal luminance; these too should not fluctuate over time, but in contrast may improve with dilation. Inadequate retinoscopic reflex and/or subjective refinement may manifest as reduced acuity that uniquely fluctuates over time. Lenticular-AIM attachments can induce cataractous changes that may be difficult to observe clinically with dense AIMs but may uniquely diminish acuity over time. Finally, closely-approximated AIM apertures can cause monocular diplopia that varies with working distance due to the Scheiner principle. Several factors can exist in a single patient, making careful examination and imaging essential to optimize visual acuity and guide intervention. Observation may be appropriate if AIMs are bilateral and symmetrical, apertures are adequate in size and location, and a fixation preference is absent in pre-verbal patients. Refractive correction and (precautionary) patching may need to be employed. Treatment options include pharmacological mydriasis, surgical membranectomy or laser membranotomy, each of which offer advantages and disadvantages.

2. Case report

A 20-year-old Asian male college student majoring in computer science, presented with a two-year progressive subjective decline in vision which was unimproved by new glasses, and visual confusion (seeing different images in each eye) at near but not distance. He also endorsed nyctalopia and difficulty adjusting to changes in ambient lighting, but no hemeralopia or photoaversion. His parents had also noted his use of an abnormal head posture during computer use which he felt helped alleviate the visual confusion. Medical and family histories were noncontributory. He was originally evaluated elsewhere at three- and six-months of age, but given good monocular tracking and no fixation...
preference, observation was advised. His only consequent intervention was full myopic refractive correction from 8-years of age. On examination, best-corrected visual acuity (BCVA) was 20/40\(^2\) in the right eye and 20/50\(^2\) in the left eye under ambient lighting conditions, with no measurable objective change under scotopic or photopic conditions, or following pharmacological dilation. Alignment was orthophoric, with full eye movements, fusion at both distance and near by Worth-4-dot, stereopsis of 140 seconds of arc, and no angle kappa. Intraocular pressure was normal in both eyes. Slit lamp biomicroscopy showed clear corneas, quiet anterior chambers, and bilateral symmetrical dense AIMs, with a very poor pharmacological dilation response in both eyes (Fig. 1a/b). Evaluation of lens clarity, macular appearance, and optic nerve structure were hence very limited. Cycloplegic refraction was \(0.75 + 0.75 \text{ axis } 165^\circ\) in the right eye and \(6.50 + 2.25 \text{ axis } 165^\circ\) in the left eye. Retinoscopy was not possible while subjective refinement was inconsistent. Anterior segment ultrasound biomicroscopy (AS-UBM) showed no evidence of iris-lenticular adhesions or lens opacification in either eye (Fig. 2a/b), and B-scan displayed no gross retinal or optic nerve anomalies.

The patient and his family were keen to pursue intervention, although caution was advised given the subjective decline in vision of unclear etiology, significant anisometropia, and risk of complications (e.g. hyphema, cataract creation or exacerbation). Despite this, the patient wished to proceed, so bilateral sequential surgical membranectomy was planned, starting with the left eye on one day and proceeding to the right eye at a later date only if symptomatic improvement without complication was achieved in the first eye. Pre-operative and intra-operative sympathomimetic agents (i.e. phenylephrine, epinephrine) were avoided to allow identification and intraoperative treatment of bleeding from severed iris vessels. Viscoelastic was placed under the AIM to stretch the fronds attached to the iris collarette, break any diaphanous connections to the lens capsule that were not detectable on AS-UBM, and create space to protect the lens. AIM fronds were severed at the iris collarette junction using 23G microscissors (MST, Redmond, WA) and the AIM was extracted en-masse through a 1.0mm paracentesis using 23G microforceps (MST). Minimal bleeding occurred in either eye without need for intraocular cautery. BCVA in the patient’s left eye at his post-operative week 2 visit improved to 20/30\(^3\) with pinhole, with no evidence of post-operative complications (Fig. 1d). Hence the decision was made to proceed with surgical membranectomy of the right eye 4 weeks later (Fig. 1a/c) using the same approach. Post-operative eye drops in each eye included prednisolone acetate 1% four times daily for 1 week followed by a taper by 1 drop per week, and moxifloxacin 0.5% four times daily for 1 week, but no cycloplegic or miotic agents.

On his last post-operative visit, BCVA improved to 20/20\(^3\) in the right eye and 20/30\(^2\) in the left eye. Final refraction was \(-2.50 + 0.75 \text{ axis } 025^\circ\) and \(-6.25 + 2.50 \text{ axis } 175^\circ\), respectively. Stereopsis improved to 20 seconds of arc. Both lenses were clear, with normal retina and optic nerve in the right eye, and a mild myopic retinal appearance with tilted disk and scleral crescent in the left eye. His nyctalopia, issues with ambient lighting, visual confusion at near and intermittent torticollis all fully resolved following surgery.

3. Discussion

Accessory iris membranes are a rare entity that may cause reduced visual function via multiple mechanisms. Our patient presented with somewhat atypical symptoms, including a subjective gradual decline in vision over 2 years that did not improve with refraction, more difficulty seeing at night, and perceiving different images out of his 2 eyes at near, which he could resolve with a mild head turn. All resolved following AIM removal, suggesting a causal association.

Given reduced vision in both eyes, the density and central involvement of his AIMS, as well as his anisometropia, we suspected a component of deprivation and/or refractive amblyopia, which was supported by a review of old notes indicating reduced but relatively stable visual acuity over the past several years. The absence of a subjective or objective decline in acuity following pharmacological dilation or...
reduced ambient lighting argued against a role for pinhole or stenopaeic effects from his AIMs augmenting his vision, while the lack of improvement in acuity following dilation ruled out reduced retinal illumination from his AIMs as an impediment to his vision. While his poor dilation response somewhat clouded this interpretation, we would expect such effects to have remained stable over time, and hence sought at an alternate cause of his subjective decline in vision. Despite our best attempts, we have to accept that suboptimal refraction may have at least partially contributed to his subjective decline in vision, given that both the sphere and cylinder axis changed significantly after surgery in his better-seeing right eye. Given his subjective decline in vision and increasing issues seeing in the dark, cataract development was a concern, although there was no evidence of this in either eye after surgery.

Patient consent

Written consent was not obtained as the report contains no personal identifying information.

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Authorship

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

4. Conclusions

This case highlights that surgical removal of AIMs can provide significant symptomatic, acuity and stereopsis gains, even in the setting of anisometropia, that optimal subjective refraction is challenging even in high-functioning adults with AIMs, and that a subjective decline in vision may not portend neutral or negative outcomes after membranectomy. However, it should be noted that such improvements may not be universal, and each case needs to be examined separately from prior experience and outcomes to avoid unnecessary interventions and/or failed expectations.
Declaration of competing interest

The following authors have no financial disclosures: OL, KM, RM, JJD.

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