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Case Report Open Access

Visual Outcome after Bilateral Implantation of Extended Depth of Focus Intraocular Lens in a Patient with Iris Microhemangiomas

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Abstract

Iris microhemangiomas (IMH) are acquired benign tumors of the iris stromal vasculature. We present the case of a 61-year-old diabetic woman with bilateral IMH and history of impaired pupillary response, in which excellent visual outcome was achieved after bilateral implantation of an extended depth of focus (EDOF) intraocular lens (IOL). On examination, her pupils demonstrated limited response to light and pharmacological mydriasis. There was history of spontaneous hyphema which resolved with medical therapy. She underwent uneventful bilateral phacoemulsification of visually significant cataracts, with implantation of an EDOF IOL in each eye. Post-operatively, the patient was satisfied with excellent uncorrected visual acuity both at distance and near. This case illustrates that EDOF IOLs can be a reasonable and effective option for cataract patients who suffer from iris pathology such as iris vascular anomalies and/or limited pupillary response.

Keywords: Iris microhemangiomas; Spontaneous hyphema; Pupillary response; Cataract surgery; Extended depth of focus intraocular lens

Introduction

Iris microhemangiomas (IMH), also known as iris vascular tufts or Cobb's tufts, consist of rare, acquired benign tumors of the iris stromal vasculature [1]. They appear as nodular clusters of small, tightly coiled, thin-walled blood vessels localized at the pupillary border [2]. Usually bilateral, they mostly affect individuals older than 50 years [1,3-5] and have been associated with certain systemic diseases including diabetes mellitus type II and myotonic dystrophy [5,6]. Histopathologically, IMH are defined as vascular hamartomas arising from the iris stroma, ranging from 15-150 μmin size [3,4,7-9]. They are clinically distinct from rubeosis iridis or neovascularization of the iris (NVI), which can occur as a complication of proliferative diabetic eye disease.

The clinical significance of IMH lies in their inherent predisposition to bleeding. Although they are generally asymptomatic, the most common presenting complaint consists of sudden decrease in visual acuity due to hyphema [10-12]. Several cases of spontaneous hyphema secondary to IMH have been reported in the literature [1,3,4,8,10,11,13-21]. In most occurrences, the hyphema appears as an isolated episode and can improve spontaneously with conservative management [1]. For recurrent cases however, laser photocoagulation or surgical iridectomy may provide definitive treatment [16,18].

In considering cataract surgery, patients with iris vascular anomalies such as IMH pose an additional challenge. First, these patients are at high risk for intra-and post-operative hemorrhage, as IMH are fragile vascular structures that can be prone to rupture. Second, since there is a well-known dependency between the visual performance of IOLs and pupil size, the potential benefits of certain presbyopia-correcting intraocular lenses (IOLs) remain less predictable in patients with impaired pupil responsiveness secondary to iris pathology [22]. With multifocal IOLs that utilize changes in pupil size to maintain focus across various distances and lighting conditions, iris abnormalities such as IMH may lead to uncertain and variable outcomes.

Newly developed IOLs tend to aim for a design that maintains lens performance regardless of changes in pupil size. The development of extended depth of focus (EDOF) technology has led to the appearance of novel presbyopia-correcting lens implants which have been shown to provide good visual outcomes across all distances, with high levels of spectacle independence and patient satisfaction after cataract surgery [23]. Although its novel technology is stated to provide stable performance in any lighting condition [24], no specific data has been published regarding the extent of its pupil independence and its impact on visual outcomes in patients with pupil abnormalities such as IMH.

We hereby present a case of bilateral IMH with history of spontaneous hyphema with limited pupil response, in which excellent visual outcome was achieved after bilateral implantation of an extended depth of focus IOL.

Case Report

A 61-year-old Caucasian woman was referred by her optometrist in February 2014 for sudden onset blurry vision in her right eye. On past medical history, the patient was known to have diabetes mellitus type II, hypertension, dyslipidemia, depression, as well as Raynaud's phenomenon. Her list of oral medications included Metformin, Quinapril/Hydrochlorothiazide, Atorvastatin, Sertraline, and Celecoxib. Upon initial examination, the patient's best-corrected visual acuity was measured at 20/25 on the right and 20/20 on the left. Her intraocular pressure (IOP) was measured at 17 mmHg and 15 mmHg in her right and left eyes, respectively. Slit lamp biomicroscopy revealed a 1 mm hyphema in the anterior chamber of her right eye and 4 small vascular tufts on the pupillary border of the iris in the right eye, and 8

similar small vascular lesions on the left. Source of the hyphema in the right eye could be seen as active pulsatile hemorrhage from one of the superior vascular tufts, suggestive of arterial nature of the hemorrhage. Limited pupil dilation was noticed on both sides despite application of Phenylephrine 2.5% and Tropicamide 1% drops (from 3 mm diameter under scotopic condition to 3.5 mm after application of drops). Remainder of the examination was unremarkable, and no iris neovascularization was identified. Dilated fundus examination also did not reveal any sign of diabetic retinopathy. The patient was diagnosed with bilateral iris microhemangiomas (IMH) with associated spontaneous hyphema in her right eye. She was treated with a course of topical Prednisolone 1% and Homatropine 2%, and the hyphema resolved after one week with no later recurrence. The patient's vision returned to 20/20 and she was discharged back to her optometrist for routine follow-up and monitoring.

Over two years later, the patient was referred for progressive vision loss secondary to cataracts. Best-corrected visual acuity had decreased to 20/200 on the right and 20/30 on the left. Visually significant nuclear sclerotic cataracts were noticed bilaterally with dense posterior subcapsular cataract changes on the right. Limited pupil response to light and pharmacological agents was noticed as seen in initial visit. Risks and benefits of cataract surgery were discussed, including risk of intra-operative or post-operative hemorrhage from IMH. IOL options were discussed with the patient and she opted to have the Tecnis Symfony IOL (Johnson & Johnson Vision Care, Inc., Santa Ana, California, USA) in each eye.

Phacoemulsification with posterior chamber intra-capsular IOL implantation was performed for the right eye followed by the left one month later. For each eye, pupil dilation was again limited despite application of phenylephrine drops pre-operatively (10% minims) and intracamerally 0.1 cc (0.03%, diluted with balanced saline solution). Phenylephrine was injected into the anterior chamber in the beginning of the case not just as attempt to dilate pupil but also for vasoconstriction. A 6.25 mm Malyugin ring (MicroSurgical Technology, Redmond, Washington, USA) was used to maintain sufficiently dilated pupil throughout the case and the ring was removed at the end. Care was taken during placement and removal of the ring to avoid significant trauma to the vascular tufts. A Tecnis Symfony IOL (ZXR00) 18.0 D and 18.5 D was implanted in the right and left eye, respectively. Both procedures were tolerated well with no occurrence of hemorrhage or other intra-operative complications.

Post-operatively, no hyphema was present for each eye. Post-operative iritis resolved for each eye after a course of topical corticosteroid (with gradual taper) and non-steroidal anti-inflammatory drug (NSAID). At post-operative day 3 follow-up for the left eye, a new bullous area of retinoschisis was detected temporally in the periphery. Though patient was asymptomatic, given the suspected acute and unilateral nature of the retinoschisis, it was treated with Argon laser retinopexy to prevent expansion of the area of concern.

At final post-operative follow-up (5 months for right eye, 4 months for the left eye), the patient expressed high satisfaction with the results and reported no issues with either distance or near-vision activities. Despite limited pupil response as seen in baseline, the uncorrected distance visual acuity was 20/15-1 in the right eye and 20/20 in the left eye, and the uncorrected near visual acuity was J1 in each eye at 2 feet. The Symfony IOL was stable in the capsular bag with centrated position at the visual axis in each eye (Figure 1). Interestingly, some of the IMHs in each eye had possibly involuted at the pupil margin as only 2 vascular tufts remained visible at the pupil margin of each eye (Figure 2). Her intraocular pressure (IOP) remained normal at 19 mmHg on the right and 17 mmHg on the left. She did not show any sign of rebound iritis nor hemorrhage and had been tapered off all post-operative medications. Optical coherence tomography (OCT) of the macula showed no evidence of macular edema in either eye. She was discharged back to her optometrist for annual monitoring.

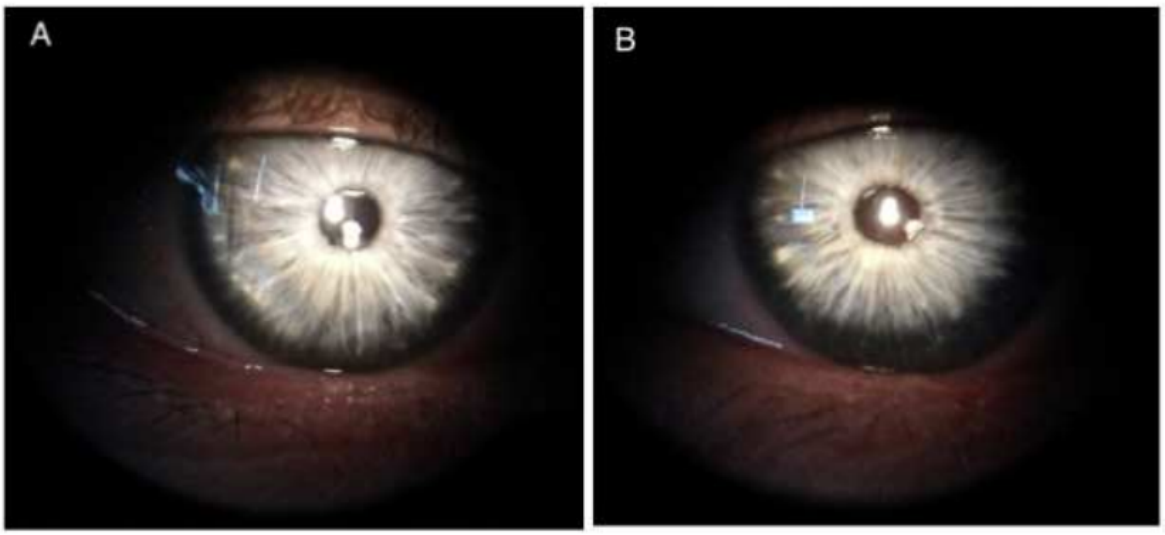

Figure 1: Post-operative anterior segment photographs. Photograph for the right (A) and left (B) eyes after bilateral phacoemulsification and implantation of an extended depth of focus intraocular lens. The lens implant can be seen centrated in the posterior chamber behind the pupil in each eye

Discussion

With the introduction of presbyopia-correcting IOLs, patients undergoing cataract surgery can now benefit from both distance and near-vision correction without having to wear glasses. These lenses provide a significant advantage over traditional monofocal IOLs, with which patients need to rely on spectacle use for near-vision activities. Presbyopia-correcting lenses can be divided into multifocal and accommodative types. Multifocal lenses are designed to produce multiple focal points through different zones of lens power [25], whereas accommodative lenses rely on ciliary muscle contraction to change power across different distances [26]. More specifically, multifocal IOLs can be further categorized as refractive or diffractive. Refractive lenses possess multiple concentric zones which create different focal points to provide uncorrected vision across all distances. Although these IOLs provide good visual acuity at both distance and near, their zonal design induces a certain limitation based on pupillary diameter [27]. On the other hand, diffractive IOLs are inherently designed to provide pseudoaccommodation by focusing the light across all distances regardless of pupil size [28].

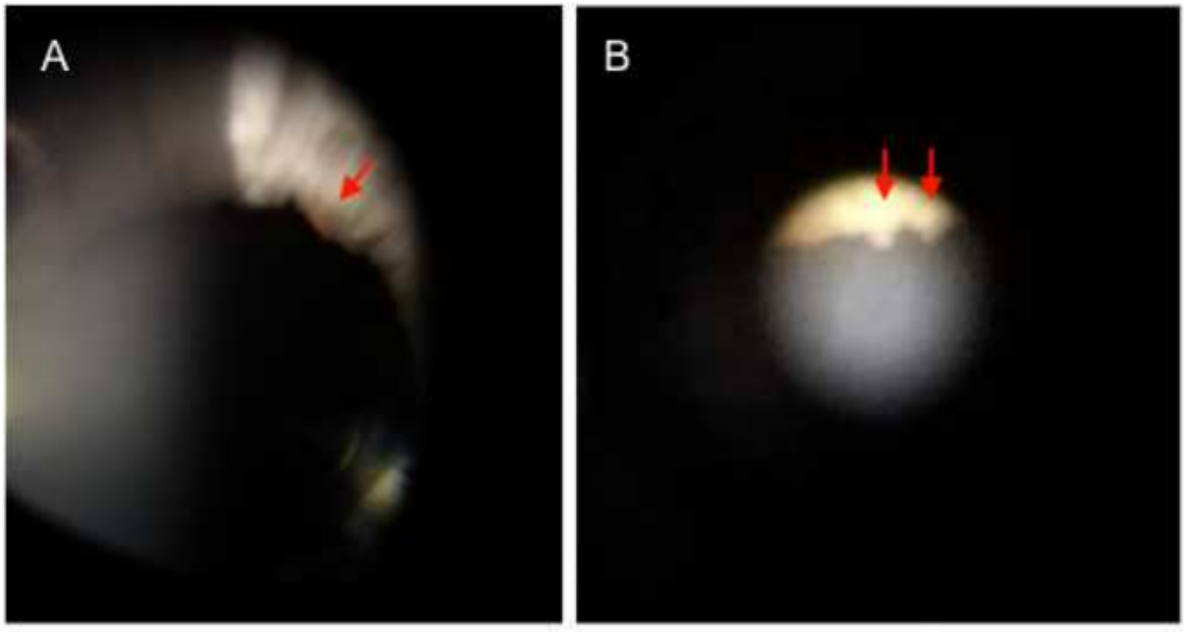

Figure 2: High magnification photographs of the iris microhemangiomas in each eye. Photograph for the right (A) and left (B) eyes, showing the superior aspect of the pupillary aperture with visible iris vascular lesions. The microhemangiomas can be seen as small vascular tufts at the pupil margin (indicated by red arrows).

The Tecnis Symfony IOL is a diffractive lens designed with an anterior aspheric surface and a posterior achromatic surface with diffractive rings that provide an elongated focal area [23]. Since this lens does not rely on a multi-zone design to maintain focus across various distances, it is stated to be pupil-independent and to provide optimal performance in both photopic and mesopic conditions [24]. Despite the theoretical basis behind this potential advantage, concrete data assessing the validity of pupil independence with diffractive lenses remains limited.

Given the history of diabetes mellitus in this patient, the differential diagnosis of the observed iris vascular lesions included NVI, which can be a sign of underlying proliferative diabetic retinopathy. However, dilated fundus examination performed at the initial encounter revealed no sign of diabetic retinopathy or neovascularization, which made this diagnosis less likely. Moreover, the characteristic appearance of these small clusters of tightly coiled blood vessels was strongly suggestive of iris microhemangiomas as opposed to NVI.

IMH are benign vascular tumors of the iris stroma which are prone to rupture and can lead to spontaneous bleeding in the anterior chamber. The patient reported here was at higher risk for the development of IMH given the known association between IMH and diabetes. At final post-operative follow-up, some of the IMHs initially observed in each eye had possibly involuted, as demonstrated by the smaller number of visible lesions at the pupil margin. Spontaneous regression has been previously reported as part of the natural history for other types of iris vascular anomalies, including iris capillary hemangioma [29], iris cavernous hemangioma [30], and iris varix [31]. Although there was no obvious rupture of any IMH during phacoemulsification for this patient, surgical manipulation may have played a role in the regression of these lesions. Some of the vascular tufts could have rolled back posteriorly behind the pupil margin during recovery, after the use of a pupil retraction device intra-operatively.

The patient presented here displayed impaired pupillary reactivity to light as well as limited response to pharmacological mydriasis. To our knowledge, no evidence has been published to date regarding the association between iris vascular anomalies such as IMH and limited pupillary response. However, long-standing diabetes has been associated with poorer response to mydriatic agents such as

Tropicamide and Phenylephrine [32]. This may be a result of damage to the autonomic nervous system which occurs as one of the microvascular complications of diabetes [33]. Also, accumulation of glycogen in the iris stroma as well as modifications to the iris muscles may contribute to limited pupillary dilation in diabetic patients [34].

When a cataract patient displays iris abnormalities causing potentially impaired pupil responsiveness, one may question the reliability of lenses that are limited by pupillary diameter. As such, choosing a lens with a pupil-independent design, such as certain diffractive IOLs, appears to be a strategic option in these individuals. The patient reported here had been diagnosed with bilateral IMH several years prior to her cataract extraction. She had suffered from a spontaneous hyphema in her right eye, which remains one of the potential complications of iris vascular anomalies. Despite her known iris pathology, she displayed excellent uncorrected distance and near visual acuity after bilateral implantation of the Tecnis Symfony IOL. There was no subjective concern regarding light adaptation or visual changes in different lighting conditions.

To our knowledge, this is the first report of successful visual outcome after bilateral EDOF IOL implantation in a patient with bilateral IMH. This example illustrates that EDOF IOLs can be a reasonable IOL option that should be considered for cataract patients who suffer from iris pathology such as iris vascular anomalies and/or limited pupillary response.

Disclosure

The authors report no conflicts of interest in this work.

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