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Case Report

Filtering Pseudo-Bleb Secondary to Sutured Posterior Chamber Intraocular Lens with Implications for Ocular Surgery in Marfan Syndrome

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Abstract

A 51-year-old female Caucasian patient with a history of Marfan syndrome and multiple previous bilateral ocular surgeries presented with increasing discomfort, epiphora, and blurred vision in her right eye for a few months. On examination, we found an overhanging cystic Seidel-positive filtering pseudo-bleb with hypotony in her right eye and a smaller Seidel-negative filtering pseudo-bleb in the left eye secondary to sutured intraocular lens (IOL) in both eyes. Intraoperatively, two full-thickness scleral defects were found close to the limbus, suggesting a melting flap in the location of the previous sutured IOL implant in the right eye. The defects were plugged with two pieces of donor sclera and covered with a larger donor scleral patch, the ischemic conjunctiva was excised, and the remaining healthy conjunctiva was advanced and sutured along the limbus. At last follow-up, intraocular pressure and vision in the right eye increased to preoperative levels, and no pseudo-bleb or leak was detected.

Keywords: Marfan syndrome, pseudo-bleb, bleb leak, sutured intraocular lens

Introduction

Marfan syndrome is a congenital connective tissue abnormality caused by mutations in the fibrillin *(FBN1)* gene and affecting cardiovascular, musculoskeletal, and ocular structures.[1] Weakening of the sclera in Marfan syndrome poses a risk of primary (spontaneous scleral rupture) or secondary (following scleral incisions) pseudo-bleb.

Herein, we report a rare case of leaking pseudo-bleb secondary to sutured posterior chamber intraocular lens (IOL) in a patient with Marfan syndrome.

Case Report

A 51-year-old Caucasian woman presented with increasing discomfort, epiphora, and reduced vision in her right eye for a few months. She had a history of Marfan syndrome with multiple previous ocular surgeries. Twenty-six years ago, she underwent bilateral lens extractions and sutured posterior chamber IOL implantation. Seventeen years ago, her right IOL subluxated after eye rubbing and was treated with IOL removal and insertion of an iris-clip IOL. Eight years ago, the right eye developed corneal endothelial decompensation with secondary bullous keratopathy. As a result, the iris-clip IOL was removed and replaced with

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another sutured IOL. Six years ago, a right retinal detachment was detected and treated with pars plana vitrectomy and gas tamponade. Three years ago, bilateral superonasal filtering pseudo-blebs were noted with the right more prominent than the left. At that time, mild right bleb dysesthesia was present but both blebs were Seidel-negative.

At presentation to our clinic, her vision was 6/36 right and 6/6 left with intraocular pressures of 5 mmHg and 9 mmHg, respectively. The right eye displayed an overhanging cystic Seidel-positive filtering pseudo-bleb, corneal edema with Descemet's folds and endothelial decompensation. There were no clinical signs of blebitis or endophthalmitis. The sutured IOL was centered and the retina flat. On the left, a smaller Seidel-negative filtering pseudo-bleb was noted with clear cornea, quiet anterior chamber, centered IOL, and flat retina (Figure 1A-C). Ultrasound biomicroscopy (UBM) showed a suspected scleral fistula connecting the anterior chamber and subconjunctival space in both eyes (Figure 1D-F). Posterior segment optical coherence tomography revealed macular subretinal fluid in the right eye (Figure 3A).

The patient underwent surgery to the right eye to i) identify the source of aqueous flow into the subconjunctival space, ii) to close the defect allowing aqueous flow to the subconjunctival space, and iii) excise the ischemic leaking conjunctiva and reconstruct the ocular surface tissues. During surgery, two full-thickness scleral defects were identified close to the limbus, suggesting a melting flap in the location of the previous sutured IOL implant and matching the UBM findings (Figure 2A). The defects were plugged with two pieces of donor sclera and covered with a larger donor scleral patch, the ischemic conjunctiva was excised, and the remaining healthy conjunctiva was advanced and sutured along the limbus (Figure 2B). At last follow up, IOP and visual acuity in the right eye were 20 mmHg and 6/36, respectively, and no pseudo-bleb or aqueous leak was detected. Corneal edema persisted, but the macular subretinal fluid had significantly reduced (Figure 3B).

Discussion

The first filtering bleb cases secondary to sutured IOL in Marfan patients were reported by Rees et al.[2] Time to onset of the pseudo-blebs was 2 months in one of their cases and 2 years in the other. In our case, the time to onset appeared significantly longer (somewhere between 5 and 26 years prior to presentation). Our observation together with that of Rees et al.[2] suggests this complication may occur early or late after sutured IOL surgery when performed in individuals with Marfan syndrome. Additionally, our patient presented with a leaking bleb requiring surgical intervention, and her scleral wounds were thoroughly assessed by UBM, which was not reported in Rees' patients.[2] Other similar cases are rare in the literature. Turaga et al.[3] reported a spontaneous pseudo-bleb due to scleral rupture in Marfan syndrome. Conjunctival cyst was also documented to masquerade as a pseudo-bleb after sclera-fixated IOL implantation in Marfan syndrome.[1] Shanmugam et al.[4] described a case of pseudo-bleb secondary to vitrectomy, but that patient was diagnosed with Traboulsi syndrome. UBM has also been employed to describe the anterior segment findings of this syndrome.[5]

Pseudo-blebs may be problematic for several reasons, several of which were present in our case. An elevated pseudo-bleb can lead to dysesthesia and in turn reduce the patient's quality of life. The eye may develop structural changes related to hypotony, such as the maculopathy present in our patient, and this can adversely affect vision and contribute to ocular discomfort. Furthermore, an ischemic and/or leaking pseudo-bleb may predispose to blebitis or endophthalmitis, which is potentially blinding.

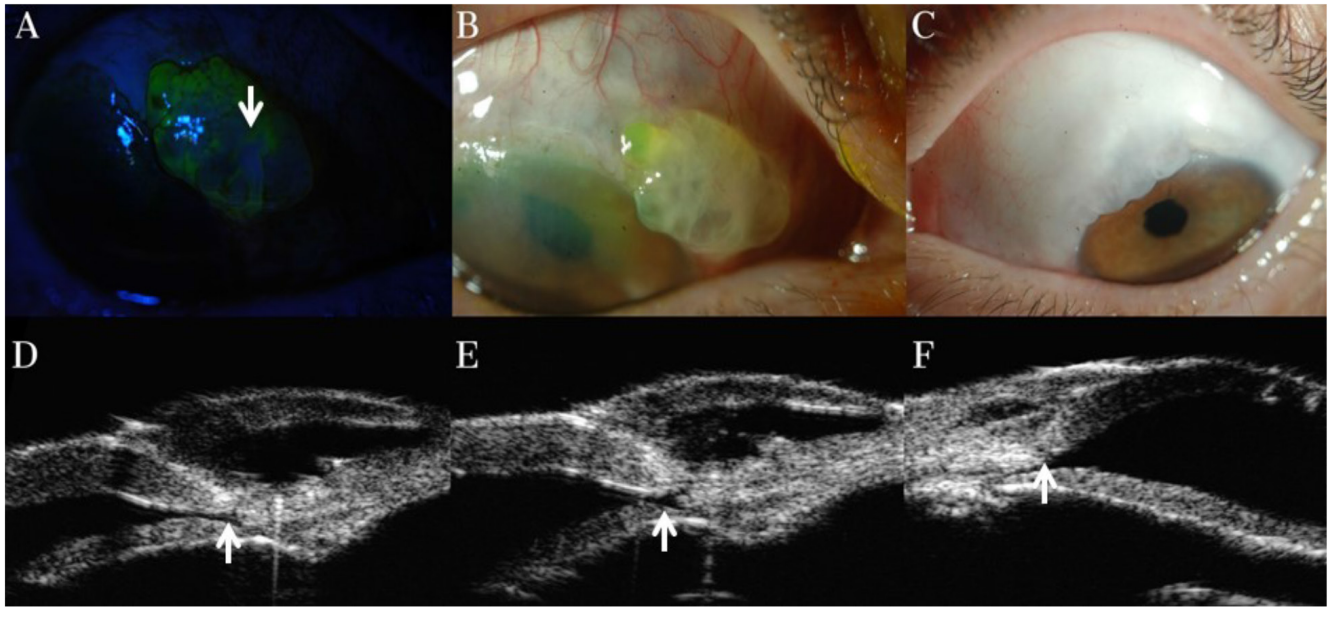

Figure 1. A-B) Right leaking filtering bleb. C) Left filtering bleb. D-F) Scleral defects in ultrasound biomicroscopy

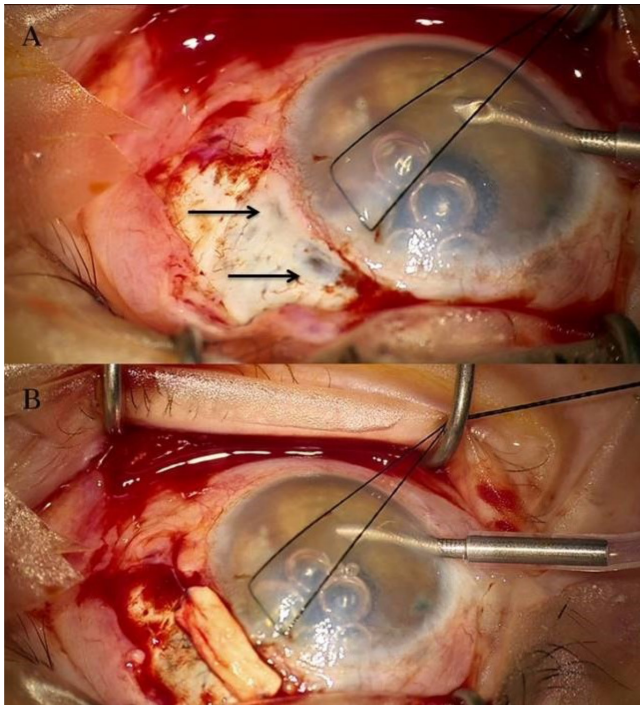

Figure 2. A) Intraoperative investigation of scleral wounds. B) Scleral patch graft

Both eyes of our patient presented with pseudo-blebs despite the fact that most surgeries had occurred on the right. This suggests the sutured IOL surgery was the likely contributing factor to scleral fistula development and subsequent pseudo-bleb formation. Therefore, scleral-fixated IOL surgery should be performed with caution. Alternative IOL choices may be preferable such as an iris-claw lens or three-piece IOL sutured to the iris.

It is interesting to note that both eyes in our patient, and those of other reported cases, developed cystic ischemic pseudo-blebs despite no exposure to anti-metabolites such as mitomycin C or 5-fluorouracil.[1-5] This suggests aqueous dynamics in the subconjunctival space plays a role in bleb morphology. This has been reported as a risk factor for bleb leak after trabeculectomy.[6] Hence, our case highlights the importance of directing aqueous flow posteriorly and over a broad area in bleb survival after filtration surgery.

Ethics

Informed Consent: Obtained.

Peer-review: Externally peer reviewed.

Authorship Contributions

Surgical and Medical Practices: Sidney Eye Hospital, Concept: T.H., Design: T.H., Data Collection or Processing: T.H., C.C., Analysis or Interpretation: T.H., C.C., Literature Search: T.H., C.C., Writing: T.H., C.C.

Conflict of Interest: No conflict of interest was declared by the authors.

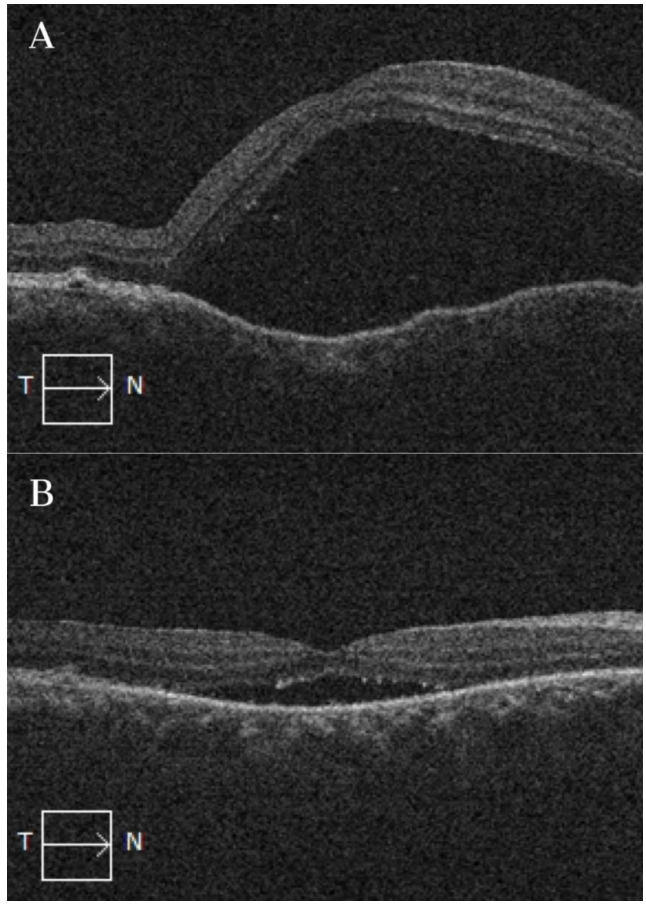

Figure 3. Subretinal fluid before (A) and after (B) the surgery

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