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CASE REPORT

Cyst Masquerading as Inadvertent Bleb After a Scleral-Fixated Intraocular Lens in Marfan Syndrome: A Case Report

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ABSTRACT

Introduction: To present a case of an epithelial inclusion cyst masquerading as an inadvertent bleb in a patient with Marfan syndrome.
Case Report: A woman with Marfan syndrome presented with a subluxed crystalline lens in her right eye, which progressively subluxed over the following 2 years. A lensectomy was performed with placement of an anterior chamber intraocular lens (IOL); however, the patient experienced blurred vision and photopsias and preferred IOL explantation. The IOL was removed and a scleral-fixated posterior chamber IOL was placed. Vision improved with an uncomplicated postoperative course. Five months later, the patient experienced sudden onset redness and sharp pain in this eye. A 3-mm cystic lesion with tan material was found over a prior scleral incision site. Intraocular pressure was normal and no aqueous leaked from the lesion. Owing to concerns of an infected inadvertent bleb, treatment with topical and oral antibiotics was started, but the lesion did not change in appearance and the patient experienced persistent pain. The lesion was surgically excised and histopathology revealed a conjunctival epithelial inclusion cyst with intralesional keratin. A month later, another conjunctival inclusion cyst developed and was excised per patient preference.
Conclusion: Marfan syndrome is characterized by defects in the *FBN1* gene and may theoretically lead to an abnormal sclera, increasing the risk of bleb formation after scleral incision. Distinguishing between a filtering bleb and an epithelial inclusion cyst is critical in patient care. Although retained keratin from a cyst may mimic a bleb with purulence, intraocular pressure, aqueous leakage, and response to topical antibiotics may help distinguish between the two.

Keywords: Blebitis; Epithelial inclusion cyst; Fibrillin; Marfan syndrome

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INTRODUCTION

Marfan syndrome is characterized by a gene mutation in the fibrillin *FBN1* gene [1]. Although certain ocular manifestations of Marfan syndrome such as lens subluxation are well documented, consequences of the *FBN1*

mutation on the sclera are relatively unassessed. The following case presents a Marfan patient who developed an epithelial inclusion cyst masquerading as an infected inadvertent bleb following placement of a scleral-fixated intraocular lens.

CASE REPORT

Informed consent was obtained from the patient prior to publication of this manuscript.

A 31-year-old Caucasian woman with a history of Marfan syndrome and an anterior chamber intraocular lens (IOL) in her left eye was referred to us for management of a subluxed crystalline lens in her right eye. She was complaining of floaters and photopsias in her right eye. On examination, a mild exotropia with left gaze preference was noted. Snellen visual acuity was 20/80 in the right eye, pinholing to 20/30 and 20/20 in the left eye. The right lens was superonasally subluxated with the lens still within the visual axis. The right eye was without intraocular inflammation, had an intraocular pressure (IOP) of 17 mmHg, and was without other abnormality aside from an area of peripheral congenital hypertrophy of the retinal pigment epithelium. The left eye had a centered anterior chamber IOL (ACIOL), IOP of 8 mmHg, a peripheral iridotomy, and a normal posterior segment.

Over the next 2 years, the patient's vision worsened in the right eye attributed to progressive crystalline lens subluxation. She subsequently underwent lensectomy with ACIOL implantation, but experienced persistent visual symptoms attributed to implant instability. She sought removal of the ACIOL and placement of a scleral-fixated IOL. This was performed using two 25-gauge trocars that were passed through bare sclera at an angle and into the vitreous cavity. End-gripping 25-gauge forceps were used to grab the tip of the IOL haptic and pull it through the trocar. The trocar was then removed leaving the haptic in the scleral tunnel. Postoperatively, she healed well and her central vision improved to 20/40.

Approximately 5 months later, the patient reported a sudden onset of redness and pain in the right eye while participating at a Marfan patient care conference abroad. She was evaluated by a local ophthalmologist who prescribed a topical antibiotic drop. Within a week, the patient returned to our center complaining of sharp, persistent periocular pain. Evaluation revealed the right eye to be mildly injected with a tender 3 mm × 3 mm lesion with tan layered material (Figs. 1, 2). There was no active leakage from the lesion and the anterior chamber of the right eye was deep and quiet. IOP and best corrected visual acuity (BCVA) were 15 mmHg and 20/200, respectively. The patient was referred to the glaucoma department for further management of a possibly infected inadvertent

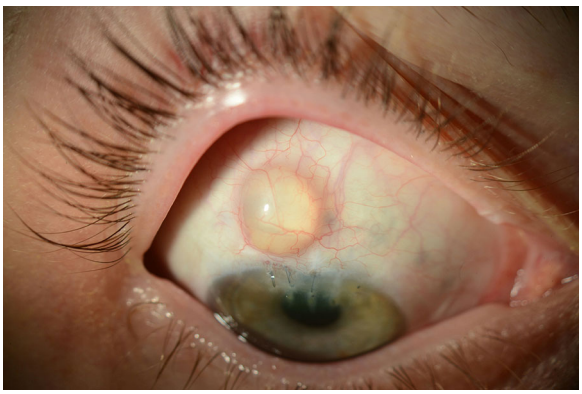

Fig. 1 Slit-lamp photograph depicting a 3 mm × 3 mm cystic lesion abutting the limbus that contains a layer of tan-colored material. The lesion failed to demonstrate active leakage with fluorescein testing

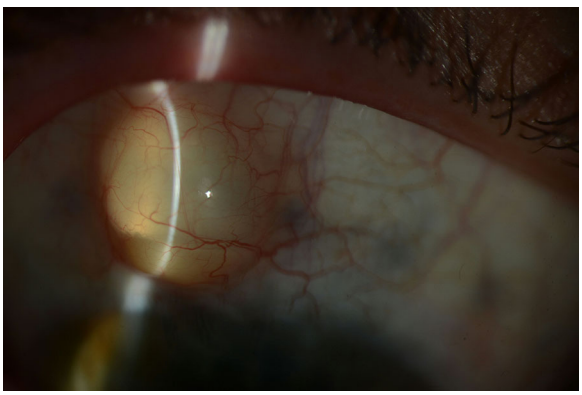

Fig. 2 Appearance of the tan-colored lesion under slit lamp examination

filtration bleb. Antibiotic therapy was switched to broad spectrum fortified antibiotics as well as oral antibiotics. Despite broadened antibiotic treatment the conjunctival injection and lesion appeared unchanged, with persistent tan material within. The patient's periocular pain worsened over the course of 2 weeks. A stable appearance on antibiotics as well as normal intraocular pressure and examination led to a high suspicion for a conjunctival epithelial cyst. The lesion was surgically excised at the patient's request to relieve the pain. Histopathologic analysis confirmed the diagnosis of conjunctival epithelial inclusion cyst (Fig. 3). A month after cyst excision, the patient developed a similar lesion a few millimeters beyond the original site (Fig. 4). BCVA in the right eye was 20/500 and the anterior chamber was deep and quiet. Again, to alleviate pain, the lesion was excised uneventfully with copious mechanical debridement of local epithelium. Histopathologic examination revealed a conjunctival epithelial inclusion cyst.

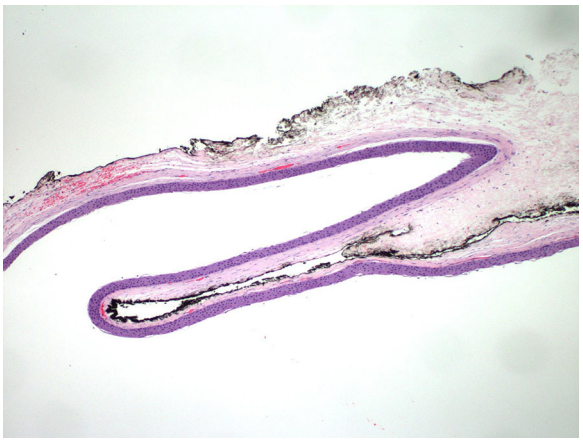

Fig. 3 Histopathological examination reveals a cyst lined by non-keratinizing stratified squamous epithelium, confirming diagnosis of epithelial inclusion cyst

Fig. 4 Reappearance of epithelial inclusion cyst

DISCUSSION

Marfan syndrome is a connective tissue disorder that arises because of congenital mutations in the fibrillin *FBN1* gene. Fibrillin provides a scaffold-like structure on which elastin fibers may assemble. Fibrillinopathies caused by the *FBN1* mutation include various cardiac valvular and vascular disorders accompanied by musculoskeletal defects. *FBN1* gene mutations may also affect certain ocular structures such as zonular fibers, retina, and sclera. Cardinal ophthalmic findings include lens subluxation, refractive and axial myopia, early onset cataracts, and retinal detachment [1].

The sclera is the predominant connective tissue of the eye. Anteriorly, the sclera is connected to the conjunctiva under the enclosure of the episclera. Elsewhere in the eye, the sclera is connected to Tenon's capsule. The sclera is composed of 0.64% elastin, 28.8% collagen, and 68% water by weight. Other proteins including cellular components and proteoglycans make up less than 4% of the remaining weight of the sclera. It is hypothesized that the elastin-fibrillary composition of the sclera plays a role in helping the eye resist deformation when acted upon by the extraocular muscles [2]. Despite the important role of fibrillin in the sclera, scleral abnormalities due to the *FBN1* gene mutation have not been extensively studied or documented. Meire et al. reported findings of blue sclera in a small subset of pediatric patients [3] and Turaga et al. published a case report on recurrent spontaneous scleral rupture in a Marfan patient [4]. Given thin or abnormal sclera in Marfan syndrome, a theoretical risk of bleb formation may exist after scleral incisions.

Distinguishing a filtering bleb from an epithelial inclusion cyst is vital in preventing bleb-associated hypotony, spontaneous rupture, and endophthalmitis. Krishnacharya described

distinguishing characteristics of blebs as larger and more luminous than conjunctival inclusion cysts. However, these general features may not be consistently accurate diagnostic criteria [5]. For example, Pawar et al. reported a patient with a small, less luminous bleb masquerading as an inclusion cyst following small incision cataract surgery (SICS) [6]. Epithelial inclusion cysts are most commonly acquired after surgery or trauma due to implantation of conjunctival epithelium [7]. During scleral fixation intraocular lens surgery as in this case, epithelial cells can be seeded if a 25- or 23-gauge trocar is placed through the conjunctiva and sclera into the vitreous. To prevent against this, the conjunctiva may be removed prior to trocar insertion.

Epithelial inclusion cysts are lined by non-keratinized stratified squamous epithelium with possible interspersed goblet cells. In some cases, keratin production by the epithelial cells in the cyst wall may lead to accumulation of tan-colored debris, which may mimic the appearance of purulent material. These cysts may therefore mimic an infected filtering bleb [8]. IOP may help distinguish the entities as an epithelial inclusion cyst lacks a connection to the anterior chamber and would likely present with minimal fluctuation of IOP. Furthermore, blebs may present with conjunctival epithelial edema. Excision and histopathological examination of a lesion in question is the gold standard for diagnosis.

Although there exists no literature regarding the prevalence of postoperative epithelial cysts or inadvertent bleb formation complications in Marfan patients, these complications have been reported in patients following manual SICS [5]. Interestingly, the incidence of inadvertent bleb formation prior to the development SICS had been reported to be between 1% and 7.7% [8]. Cases of epithelial inclusion cysts have also been reported following pars plana vitrectomy and strabismus surgery [9, 10]. To minimize postoperative cyst formation following SICS, Narayanappa et al. recommend carefully reflecting the conjunctiva before construction of the scleral wound. They believe it may also be beneficial to avoid contact between the IOL and conjunctiva during the procedure [11].

When considering cyst excision, it is important to maintain an equanimous mind-set and keep the tips of the corneal scissors away from the cyst during dissection in order to avoid cyst rupture to prevent recurrence. Thatte and Gupta suggest preserving conjunctiva above the cyst to allow for a firm grasp of the cyst while dissecting [12]. These considerations were kept during our patient's cyst excision and it was removed intact. Debridement of the scleral bed was also performed to remove any potentially seeded epithelial cells.

CONCLUSION

Fibrillin is a connective tissue protein that serves as the scaffold for elastin fibers. In the sclera, fibrillin is hypothesized to provide elasticity and prevent deformation of the eye during ocular movements [2]. Patients diagnosed with Marfan syndrome have defects in the fibrillin *FBN1* gene, and may have an abnormal sclera, theoretically leading to an increased risk of bleb creation after a scleral incision [1]. In this case, an epithelial inclusion cyst with layered keratin was clinically supported by the lack of wound leakage (a blebitis would likely require an external conduit for bacterial invasion), normal intraocular pressure, and lack of response to topical and oral antibiotics. Utilizing these criteria to differentiate between a filtering bleb and epithelial inclusion cyst may help in clinical diagnosis and prevent against bleb-associated ocular complications.

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Compliance with Ethics Guidelines. Informed consent was obtained from the patient prior to submission of this manuscript.

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