Bilateral spontaneous filtering blebs

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ABSTRACT

Purpose: To report a case of bilateral spontaneous filtering bleb, scleral thinning, microspherophakia, and mild craniofacial dysmorphosis.

Observations: An 18-year-old girl was referred to our clinic for evaluation of bilateral spontaneous filtering blebs. Her corrected distance visual acuity (CDVA) was 20/400 in the right eye (RE) and 20/100 in the left eye (LE). She had superior scleral thinning, multicystic filtering bleb, and microspherophakia bilaterally. The IOP was 9 mmHg in the RE and 8 mmHg in the LE. A mild craniofacial dysmorphosis including downward slanting of palpebral fissures and malar hypoplasia was present. There was no sign of corneal ectasia in tomography. Anterior segment optical coherence tomography revealed the filtering blebs as subconjunctival low reflective fluid-filled spaces. Due to severe scleral thinning un the RE we performed a tectonic scleral patch graft. 6 months after surgery the depth of the anterior chamber increased and CDVA improved.

Conclusion and importance: This case indicated that the formation of spontaneous filtering blebs may occur in a syndromic condition. In any case with this symptom, special attention should be paid to craniofacial features, sclera, crystalline lens, and IOP. Tectonic scleral patch graft could be a valuable option in selected patients.

1. Introduction

Filtering bleb is an aqueous filled subconjunctival space and routinely forms after trabeculectomy. It can also form after trauma or surgery, inadvertently.1 Spontaneous filtering blebs are rare. They are reported in association with ocular abnormalities like Axenfeld syndrome, Terrien’s marginal degeneration, microspherophakia, ectopia lentis, scleritis, and pellucid marginal corneal degeneration.2–8 There are reports of spontaneous filtering blebs in systemic conditions like scleroderma and Traboulssi syndrome.9–12 In this study, we present a case with mild craniofacial dysmorphosis, microspherophakia, and bilateral spontaneous filtering blebs.

2. Case report

An 18-year-old lady was referred to our clinic because of decreased vision and spontaneous filtering blebs. Her uncorrected visual acuity was finger counting at 10 cm bilaterally. Corrected distance visual acuity (CDVA) with a manifest refraction of −16.00–5.00 × 180 was 20/400 in the right eye. The manifest refraction of the left eye was −16.50–6.50 × 165 and the CDVA was 20/100. She had a downward slanting of palpebral fissures and malar hypoplasia on inspection (Fig. 1). In slit-lamp examination, there was a superior multicystic bleb associated with scleral thinning bilaterally without any history of trauma or surgery (Figs. 2 and 3). The IOP was 9 mmHg in the right eye and 8 mmHg in the left eye on Goldman applanation tonometry. In funduscopy cup to disc ratio was 5/10 in both eyes without any other retinal or vascular abnormalities. On dilated examination the entire circumference of a globular crystalline lens was visible (Figs. 4 and 5).

Besides, she mentioned a vague history of a spinal cyst that was under observation by an orthopedic surgeon. On physical examination, arachnodactyly, brachydactyly, joint or skin hyperextensibility, and heart murmurs were not noted. Also, Consultation with rheumatologists was done and no other rheumatologic abnormalities were found. The patient underwent electrocardiography and echocardiography. There was no sign of aortic root dilation and mitral valve prolapse which are common findings in Marfan Syndrome and the electrocardiography was normal. Serum and urine amino acids assay revealed no abnormality. Concerning family history, our patient was the first in her lineage and the marriage of her parents was not consanguineous.

Tomography (OCULUS GmbH, Wetzlar, Germany) showed Against the rule (ATR) astigmatism in the right eye and with the rule (WTR) astigmatism in the left eye (Figs. 6 and 7). There was no sign of corneal ectasia.
Anterior segment optical coherence tomography (AS-OCT, CASIA SS-1000; TOMEY, Nagoya city, Aichi, Japan) was done. Microspherophakia and low reflective subconjunctival spaces were thoroughly visible. No fistula connecting the anterior chamber to the subconjunctival space was found (Figs. 8 and 9). Anterior chamber depth based on AS-OCT was 1.23 mm and 1.39 mm in the right and left eye, respectively.

Due to severe scleral thinning and the shallowness of the anterior chamber in the right eye, we considered a tectonic scleral patch graft. During surgery, severe scleral thinning was noted. A large patch graft of sclera around 120° was sutured superiorly and the conjunctiva was fixed again on the limbus.

Due to the COVID-19 pandemic, she didn’t have regular follow up visits, unfortunately. But 6 months after surgery, we visited the patient again. CDVA of the right eye with a manifest refraction of −15.00 −6.5 × 88 was 20/80. A well-positioned scleral patch graft was visible in the slit lamp examination and AS-OCT (Figs. 10 and 11). Also, the depth of the anterior chamber was increased to 2.46 mm (Fig. 12).

3. Discussion

In this report, we presented a case with bilateral spontaneous filtering blebs, microspherophakia, and mild craniofacial dysmorphism. The first case of spontaneous filtering bleb was reported in Axenfeld syndrome. As glaucoma is a serious complication in this condition the postulation of Nemet et al. for bleb formation in their case was the rupture of abnormal tissue in the corneoscleral junction due to high IOP. Soon et al. reported four cases of Terrien’s marginal degeneration with spontaneous rupture of Descemet’s membrane who consequently developed corneal hydrops. In two cases intracorneal fluid extended peripherally and formed subconjunctival filtering bleb. The common pathology in cases of Terrien’s and pellucid marginal degeneration is the abnormal tissue of the corneal periphery. Our patient didn’t have any sign of axenfeld syndrome, pellucid, or Terrien’s marginal degeneration.

A case of spontaneous filtering bleb in a microspherophakic patient with mild craniofacial dysmorphism reported in 1991 by Pasquale et al. Similar to our patient, their case didn’t have physical signs of Marfan, Weill-Marchesani, or Ehlers-Danlos syndrome. The authors stated that
Fig. 6. Against the rule astigmatism in right eye.
Fig. 7. With the rule astigmatism in left eye.
the reason for conjunctival filtration is unclear.

In a case that was reported by Mantravadi et al., anterior scleritis and scleral thinning led to spontaneous filtering bleb in a 40-year-old woman suffering from systemic lupus erythematosus.

Chandran et al. reported a case of bilateral spontaneous filtering bleb, hypotony, ectopia lentis, scleral thinning and decreased vision (6). He didn’t have corneal ectatic disorders or systemic anomaly. Their proposed mechanism was seeping of aqueous humor through the scleral thinning.

Spontaneous filtering blebs are reported in association with systemic conditions like scleroderma and Traboulsi syndrome. First Shawaf et al. reported 6 members of a Druze Lebanese family with mild facial dysmorphism, subluxation of the crystalline lenses, iridocorneal adhesions, and patchy areas of iris atrophy (10). Two members had spontaneous filtering blebs, one bilaterally and the other unilaterally. There wasn’t any systemic condition related to the subluxation of the crystalline lens. They proposed an autosomal recessive pattern for this so-called syndrome. Then Haddad et al. reported four members of another Druze Lebanese family with the syndrome of lens dislocation, spontaneous filtering blebs, anterior segment abnormalities, and a distinctive facial appearance. They called it Traboulsi syndrome.

Similarly, our patient had mild facial dysmorphism, microspherophakia, scleral thinning, and bilateral superior spontaneous filtering blebs. Physical examination and laboratory experiments in our patient only revealed a mild craniofacial dysmorphism. Family history was unremarkable, proposing an autosomal recessive pattern. Albeit, we didn’t perform genetic testing. Crystalline lens abnormality may be a spectrum from microspherophakia to subluxation in this syndrome. Aqueous humor crossing through the thin sclera is the presumed mechanism for bleb formation. In cases with scleral thinning reinforcing sclera with a patch graft may be an appropriate treatment option.

Fig. 8. A. Multicystic bleb in the right eye. B. Anterior chamber depth before surgery in the right eye.

Fig. 9. A. Multicystic bleb in the left eye. B. Anterior chamber depth in the left eye.

Fig. 10. Note the scleral patch graft and anterior chamber depth 6 months after surgery.
Patient consent
A written consent form was taken from the patient.

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

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Declaration of competing interest
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