Bilateral idiopathic spontaneous filtering bleb with ectopia lentis: A case report and review of literature

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A 26-year-old male presented with superior filtering bleb with scleral thinning, dislocated lens, and hypotony in both the eyes. His cornea was normal without any sign of ectasia, and there was no history of recurrent redness, trauma, or surgery in either eye. Anterior segment optical coherence tomography did not reveal communicating fistula between the anterior chamber and subconjunctival space. Physical examination and blood investigations did not reveal any systemic association. He was diagnosed to have spontaneous filtering bleb, which is a rare condition observed with ocular or systemic abnormalities.

Key words: Hypotony, lens dislocation, spontaneous filtering bleb

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Spontaneous subconjunctival filtering bleb is a rare occurrence and has been observed with ocular or systemic abnormalities such as Axenfeld syndrome,[1] Terrien marginal degeneration,[2,3] familial craniofacial dysmorphism,[4,5] scleritis,[6] and connective tissue diseases.[7] To the best of our knowledge, only few cases have been reported till date, and we report a 26-year-old patient with bilateral spontaneous filtering bleb.

Case Report

A 26-year-old male presented with defective vision in both eyes (BEs) since childhood. The patient had no history of previous ocular surgery, trauma, or systemic illness. His best-corrected visual acuity was 6/18 with + 11 + 1.5 × 90 in the right eye (RE) and 6/12 with + 11.5 + 2.5 × 90 in the left eye (LE). Slit-lamp examination showed a diffuse conjunctival bleb near the limbus extending from 10 to 4 o'clock in the RE [Fig. 1a and b] and 8 to 1 o'clock in the LE [Fig. 1d and e] with superior scleral thinning in BEs. Seidel's test was negative in BE. RE showed clear cornea with deep anterior chamber and inferonasal dislocated lens. LE showed iridocorneal adhesion in the paracentral zone with corneal scar, iris atrophy, and aphakic lens status. Dilated examination revealed microphakic-dislocated lens in the anterior chamber in the RE and inferonasal dislocated lens in the LE [Fig. 1c and f]. Anteriorly dislocated lens in the RE was reposited into the posterior chamber in the supine posture. His intraocular pressure was 6 and 3 mmHg in the RE and LE, respectively. On gonioscopy, angles were closed in BEs without evidence of ostium or cyclodialysis cleft. Fundus examination showed a cup

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to disc ratio of 0.5:1 with healthy neuroretinal rim, choroidal folds, and attached retina in BE.

Corneal topography and pachymetry (Orbscan, Bausch and Lomb, Orbtek Inc., UT, USA) were suggestive of oblique astigmatism in BE without evidence of corneal ectasia [Fig. 2a and b]. Anterior segment optical coherence tomography (DRI OCT Triton plus, Topcon, Tokyo, Japan) showed aqueous pockets in subconjunctival and intrascleral spaces in BEs [Fig. 3a and b]. There was no communicating fistula between the anterior chamber and subconjunctival space.

Physical examination did not reveal joint or skin hyperextensibility, brachydactyly, facial dysmorphic features, or marfanoid habitus. Blood investigations including rheumatoid factor, homocysteine, and lysine level were within normal limits. Antinuclear antibody test was negative. Electrocardiogram and echocardiogram were normal.

Discussion

First case of spontaneous filtering bleb was reported in a patient with Axenfeld syndrome by Nemet *et al*.[1] The patient had prominent Schwalbe line and iridocorneal adhesion in both the eyes. Authors have postulated that increased IOP leads to rupture of the corneoscleral junction resulting in the formation of filtering bleb. Our patient had iridocorneal adhesion only in the LE and did not have anterior embryotoxon in both the eyes. Spontaneous filtering bleb formation due to corneal pathologies such as Terrien and pellucid marginal degeneration have been reported by Soong *et al*.,[2] Munro *et al*.,[3] and Toriyama *et al*.[8] Mechanism proposed was the extreme distortion and stretching of the peripheral cornea resulting in rupture of the Descemet's

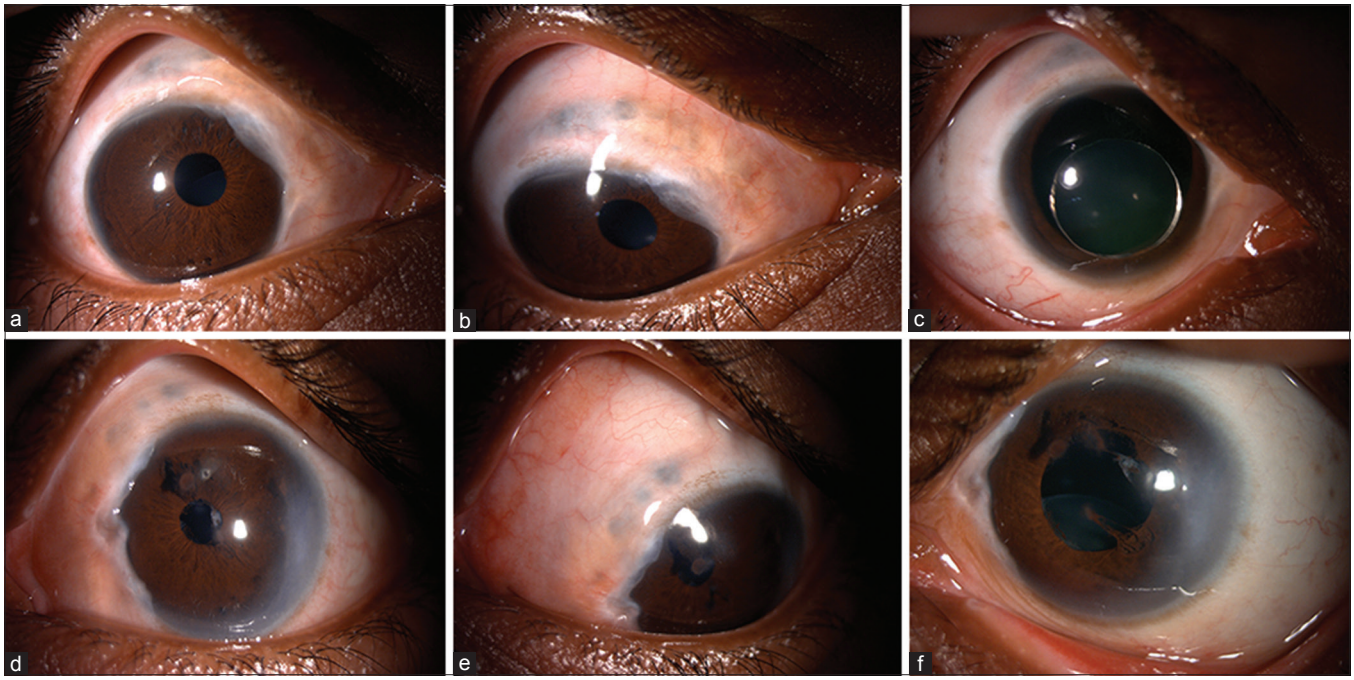

Figure 1: Slit-lamp photograph of the right eye showing inferonasal dislocated lens, superonasal diffuse bleb with scleral thinning (a and b) and anterior dislocated microphakic lens on dilatation (c). Left eye showing iridocorneal adhesion and aphakic lens status, superonasal diffuse bleb with scleral thinning (d and e) and inferonasal dislocated lens on dilatation (f)

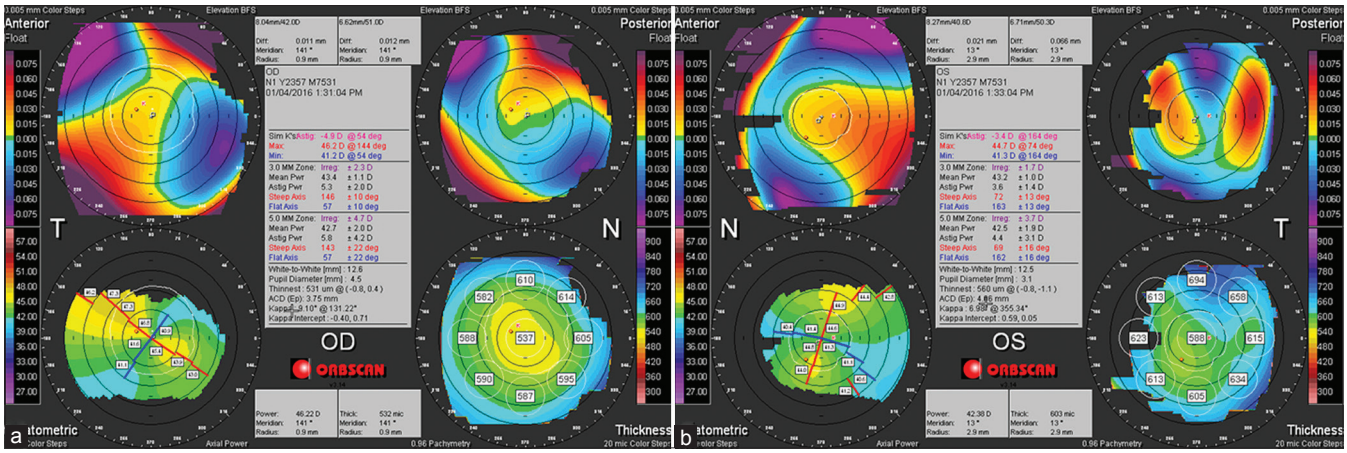

Figure 2: Corneal topography image of the right (a) and left (b) eye showing oblique astigmatism

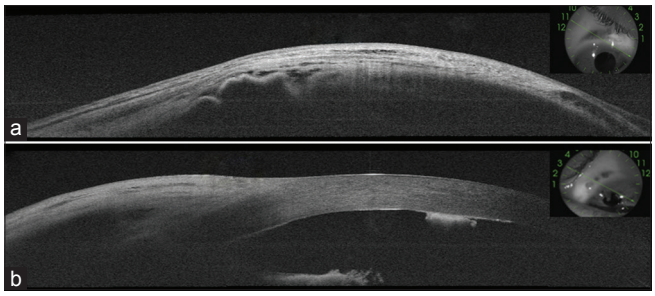

Figure 3: Anterior segment optical coherence tomography of the right (a) and left eye (b) showing aqueous pockets in subconjunctival and intrascleral spaces

membrane and bleb formation. Our patient had normal cornea without signs of thinning or degeneration.

Shawaf *et al*.[4] and Haddad *et al*.[5] have reported two unrelated families of Lebanese Druze community with syndrome of ectopia lentis, spontaneous filtering bleb, and craniofacial dysmorphism. Dysmorphic features were dental crowding, large beaked nose, and antimongoloid slanting of the palpebral fissures. Authors concluded that the syndrome is due to abnormality in the connective tissue development and it is inherited in an autosomal recessive manner. Pasquale *et al*.[9] have reported spontaneous filtering bleb in a patient with microspherophakia and dysmorphic features in the form of malar hypoplasia, frontal bossing, and septal deviation. Our patient had filtering bleb and microphakic-dislocated lens, but there was no facial dysmorphic features.

Mantravadi and Stock[6] have reported spontaneous filtering bleb with scleral thinning as a consequence of scleritis in a female with systemic lupus erythematosus. Gerke *et al*.[7] have reported spontaneous filtering bleb, ocular hypotension, inferior oblique and superior rectus paralysis, and madarosis in a patient with scleroderma. There were no signs of systemic connective tissue abnormality in our patient. Two patients with spontaneous filtering bleb without any other ocular or systemic abnormalities have been reported by Munro *et al*.[3] and Tatham *et al*.[10]

Conclusion

Our patient had bilateral superior filtering bleb with scleral thinning, dislocated lens, and hypotony for which the cause could not be elucidated and the probable mechanism of bleb formation would be seeping of aqueous through the area of scleral thinning.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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