·Letter to the editor·

Corneal endothelial decompensation secondary to iridoschisis in degenerative myopic eyes: a case report

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Dear Sir,

I am doctor Hong-Bin Wang, from the Department of Ophthalmology of Beijing Friendship Hospital affiliated to Capital Medical University in Beijing, China. I write to present a case report of iridoschisis. Iridoschisis is a rare condition characterized by spontaneous splitting and separation of the layers of the iris stroma. It is mostly seen in both eyes in patients aged 60-80 years old, and mostly involves the inferior iris [1-3]. There is no genetic predisposition reported, and the pathogenesis remains poorly understood. In this report, we describe a case of corneal endothelial decompensation with bullous keratopathy secondary to iridoschisis, in degenerative myopic eyes with posterior staphyloma involving the macula.

A 74-year-old female visited our clinic complaining of acute and persistent pain in the left eye for 3 days duration. She had myopia and blurred vision for over 30 years with low corrected glasses. She denied any history of trauma, glaucoma or other eye diseases. She had history of hypertension, coronary artery disease and breast cancer surgery, her best-corrected visual acuity was 20/1000(count finger at eight feet) in the right eye and 20/100 in the left with -17.00D and -8.00D in the right and left eye, respectively. Her axial length was 27.6mm in the right eye, and 24.6mm in the left (Iomaster, Zeiss, German). Her intraocular pressure (IOP, Full auto Tonometer, TX-F, CANON, Japan) was 15mmHg in the right eye and 17mmHg in the left. Central corneal thickness was 501um in the right eye and 511um in the left (SP-3000, TOMEY, Japan). The central anterior chamber (AC) depth was 2.12mm in the right eye and 2.42mm in the left (UBM, SW-3200, Suowei, Tianjin, China)). Slit-lamp examination showed subtle iridoschisis could be detected in the inferior iris of right eye (Figure 1A). The free ends of the degenerated iris stromal fibers were seen floating in the anterior chamber with iridocorneal contact in the inferior quadrant of left eye(Figure 1B). Slit-lamp examination of the left cornea showed thickening of the stroma inferiorly with few bullae seen at the anterior stroma and a 2-mm round area of epithelial defect, staining with fluorescein, near the limbus indicating a ruptured bullous (Figure 1C). No stromal infiltrates seen. However, the central zone of the cornea was clear and the pupil was round and reactive in both eyes. The cortex of the lens was mildly cloudy. Fundus examination showed that there were optic disc crescents in both eyes. The Optic cup to disc ratio was 0.3. The posterior staphylomas involved the macular in right eye, while the left eye is better than right eye.There had lattice degeneration and pigmentary degenerations in the extreme retina periphery without retinal hole.Gonioscopy showed an open angle without the formation of synechias in the right eye. 2 weeks later, after the cornea epithelial defect recovered, the gonioscopy were done on left eye and showed an open angle without synechias.

Ultrasound biomicroscopy (UBM, SW-3200, Suowei, Tianjing, China) showed shallow peripheral anterior chamber in both eyes, and subtle separation of the iris stroma in the inferior of the right eye (Figure 2A); The iris stroma in the inferior quadrant of the left eye was separated with forward bowing (Figure 2B) and iridocorneal contact (Figure 2C). Corneal endothelial cell count (SP-2000P, TOPCON, Japan) was 739 cells /mm^2 in the left, (In the left eye, there were 746 cells /mm^2 in the superior quadrant and 709 cells /mm^2 in the inferiorand 1882 cells /mm^2 in the right eye. The endothelial cells of left eye were enlarged than the right eye with normal shape), Phasing test showed an IOP range of 11-18mmHg in the right eye and 11-19 mmHg in the left. Systemic physical examination was normal. Syphilis serologies were reported negative. The Clinical appearance was consistent with iridoschisis. The patients were given

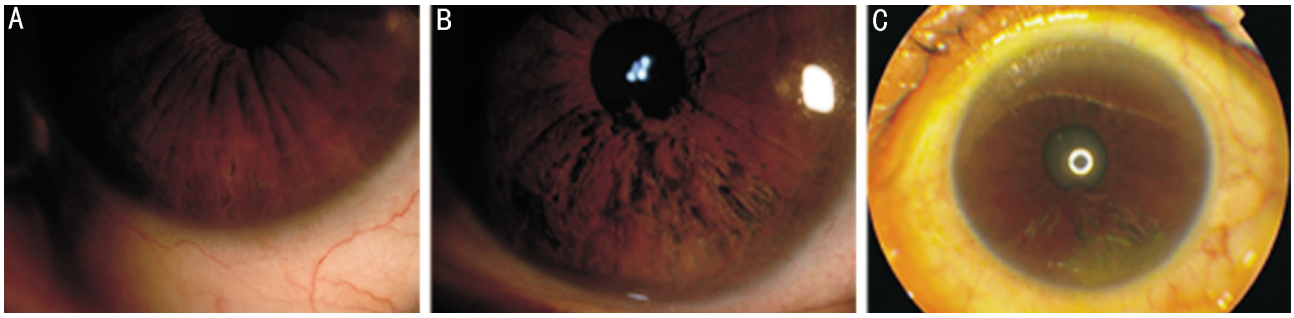

Figure 1 Iridoschisis slit-lamp examnation A:Subtle iridoschisis in inferior iris of right eye; B: Free ends of the degenerated iris stromal fibers in inferior quadrant of left eye; C:A bullous keratopathy at the inferior of the cornea, positive for fluorescein staining.

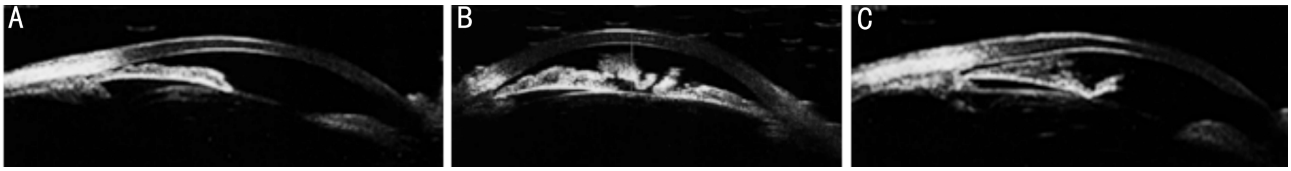

Figure 2 Ultrasound biomicroscopy showed shallow peripheral anterior chamber in both eyes A:Subtle separation of the iris stroma in the inferior of the right eye; B: Iris stroma in the inferior quadrant of the left eye; C:Iridocorneal contact.

Celluvisc (Allergen, USA) 3 times a day and BeiFuShu (bFGF) eye drops (ZhuHai Yisheng, China) 4 times a day with antibiotic eye drop to prevent infection. 1 week later, the epithelial defects were recovered.

Schmitt [1] first reported iris splitting into 2 layers in 1922. Then Loewenstein *et al* [2] coined the term "iridoschisis" and reported its pathologic changes in 1945. It is characterized by idiopathic fragmentation of the iris stroma, and the iris stroma separating into 2 layers and floating in the anterior chamber, mostly seen in the inferior iris of bilateral eyes, and mostly seen in senile people. It is usually accompanied with angle closure glaucoma, ocular trauma, syphilitic interstitial keratitis, lens subluxation and other congenital diseases[3-8]. Because the patient is asymptomatic and goes to clinic only after some complication occurs, it is very rarely encountered in clinical practice. The pathogenesis of iridoschisis still remains unclear. Using iris fluorescein angiography, Carnevalini *et al* [9] found normal perfusion in the area of iridoschisis, opposing the hypothesis of ischemic atrophy of the iris as the pathogenic mechanism [10]. Electron microscopic studies have shown thinning of the iris stroma and decreased collagen fibers with normal stromal vessels. Glaucoma occurs in about 65% of patients with iridoschisis[10,11], the 24-h IOP in this patient was 11-18mmHg in the right eye and 11-19mmHg in the left, For the glaucoma being more common in high myopic eyes thus, the IOP should be closely followed up.

Weseley and Freeman [12] were the first to report corneal changes associated with iridoschisis. They demonstrated focal endothelial cell loss overlying an area of iridoschisis in an 83-year-old woman. But there was no clinical evidence of iridocorneal contact, they speculated that intermittent iridocorneal touch might have caused these changes. Srinivasan *et al* [13] reported a case of bullous keratopathy accompanied with corneal decompensation secondary to iridoschisis, and they assumed it was caused by direct physical iridocorneal contact and believed that if has long standing iridocorneal contact, whole corneal endothelial decompensation will be the result. In our case, whole endothelial cell loss was observed, but iridocorneal contact and bullous keratopathy only observed in the inferior quadrant, while superior and center anterior chamber was deep, which demonstrated long-term iridocorneal contact in the inferior quadrant may lead to extensive whole corneal endothelial cell loss and resulted in corneal endothelial decompensation.

To the best of our knowledge, iridoschisis associated with degenerative myopia was not reported before. Degenerative myopia is thought to be the result of a combination of both genetic and environmental factors. It is also one of the manifestations of genetic disorders such as Marfan's, Ehler-Danlos syndromes, *etc* [14]. Although these two conditions (iridoschisis and degenerative myopia) can occur together by mere chance, it is also possible that hereditary factors might be involved in the pathogenesis of iridoschisis.

In summary, iridoschisis is a rare condition. To the best of our knowledge, iridoschisis associated with generalized reduction of corneal endothelial cell count in degenerative myopic eyes has not been reported before. We think iridoschisis and degenerative myopia although may occur together by mere chance. However, it is also possible that hereditary factors and collagen abnormalities might be involved in the pathogenesis of iridoschisis. For patients with iridoschisis, corneal endothelial cell count, ultrasound biomicroscopy and routine examination for glaucoma should be performed.

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