• Letter to the Editor •

Pulmonary hypertension causing uveal effusion syndrome: a case report

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Dear Editor,

I am writing this letter to present a case of uveal effusion syndrome (UES) caused by pulmonary hypertension found due to intermittent high intraocular pressure (IOP).

Pulmonary hypertension increases cardiac afterload due to elevated pulmonary vascular resistance, leading to right heart failure and death[1]. Notably, ocular complications of pulmonary hypertension have been reported less frequently[2-5]. Ophthalmologists need to increase the understanding of the disease so as to perform the proper treatment for these patients.

Ethical Approval The study was conducted in accordance with the principles of the Declaration of Helsinki. Written informed consent was obtained from the patient for publication of the images and data included in this article.

Case Presentation A 44-year-old male patient presented to ophthalmic clinic in March 2021 for intermittent high IOP and blurred vision for 1y. The patient had a family history of pulmonary hypertension for over 20y, with a maximum pulmonary artery pressure of 106 mm Hg and a significantly enlarged right heart. Six months ago, the patient visited our hospital for blurred vision in both eyes. The visual acuity was 20/100 in both eyes, and the best corrected visual acuity (BCVA) was 20/20 with the correction of -1.50 diopter sphere (DS) in both eyes, IOP was 26.6 and 26.3 mm Hg in the right eye and the left eye respectively. Slit lamp examination disclosed a markedly shallow anterior chamber both centrally and peripherally and no significant abnormalities in the iris and lens. There were dome-shaped choroidal detachments in all quadrants peripherally. Ultrasound biomicroscopy (UBM) revealed bilateral ciliary detachment with angle closure, and shallow central anterior chamber depth of 2.51 mm and 2.39 mm respectively (Figure 1A). Additionally, B-scan ultrasonography suggested bilateral ciliochoroidal detachment (Figure 1B). Due to dyspnea and hypoxia of the patient, fundus fluorescein angiography (FFA) and indocyanine green angiography (ICGA) were not performed. Furthermore, local cycloplegic agents were given to the eyes to lower IOP, and systemic glucocorticoids were applied for treatment. Visual acuity in both eyes recovered to 20/20, and IOP dropped to normal during the dosing period.

The ophthalmologic examination at this visit found the visual acuity of 20/20 in the right eye and 20/50 in the left eye, and with -1.25 DS correcting to 20/20. Following the administration of carteolol and brinzolamide drops, the IOP was 40.7 mm Hg on the right and 28.3 mm Hg on the left. Slit lamp examination showed mild conjunctival hyperemia, dilated episcleral venous, clear cornea without keratic precipitate in both eyes. The anterior chamber depth was shallower in the left eye than in the right eye. The pupils were round with 3 mm in diameter, and no obvious abnormalities of iris and lens were found in both eyes. Fundus examination showed a bilateral leopard-like fundus with normal optic discs (Figure 2A). Additionally, UBM demonstrated bilateral ciliary detachment with the central anterior chamber depth of 3.03 mm in the right eye and 2.62 mm in the left eye. The axial length was 23.98 mm in the right eye, and 23.59 mm in the left eye, while the thickness of the nasal sclera of the optic disc was 1.38 mm on the right and 1.45 mm on the left measured by B-scan ultrasonography. The optical coherence tomography (OCT), and visual field examination were unremarkable (Figure 2B, 2C). No significant abnormalities were seen on the CT of the orbit. After dilating the pupil with compound tropicamide drops, the anterior chamber depth became significantly deeper (Figure 3), at which point the IOP dropped to 28.3 and 18.5 mm Hg respectively. Furthermore, FFA did not show fluorescein leakage, and ICGA showed deep choroidal vascular tortuous dilatation and late fluorescence

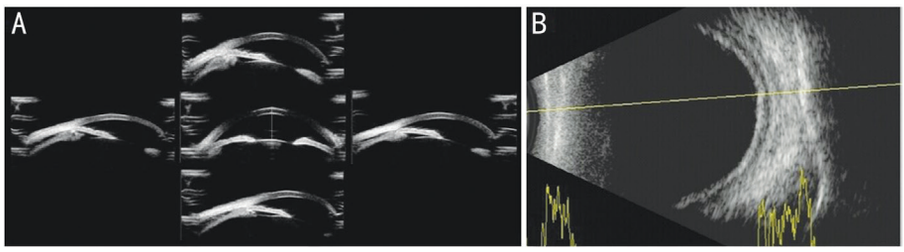

Figure 1 UBM and B-scan ultrasonography at the first visit A: UBM showeds hallow central anterior chamber, angle closure and ciliary detachment in four quadrants; B: B-scan ultrasound showed choroidal detachment. UBM: Ultrasound biomicroscopy.

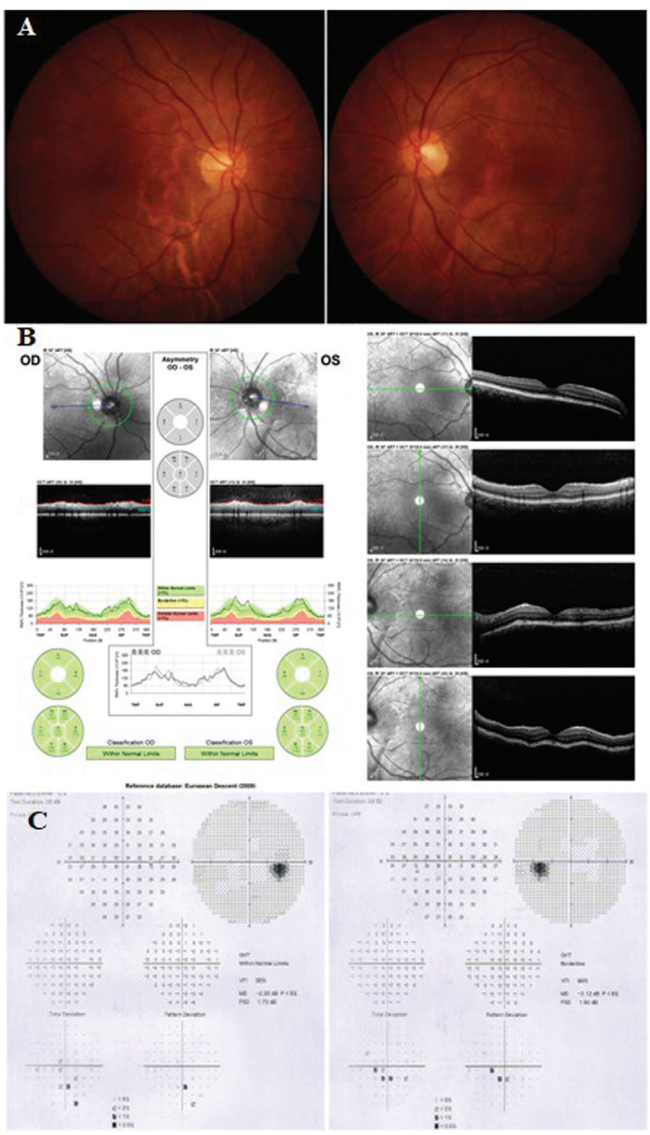

Figure 2 Color fundus photography, OCT and visual field images A: Color fundus photography showed leopard-like fundus with normal optic discs; B: OCT revealed the normal shape of optic disc and macular fovea; C: Humphrey visual field was normal. OCT: Optical coherence tomography.

inhomogeneity (Figure 4), which led to the diagnosis of UES in this patient.

The eye was treated with topical atropine ophthalmic gel to paralyze the ciliary muscle, and carteolol drops were administered to reduce aqueous humor production.

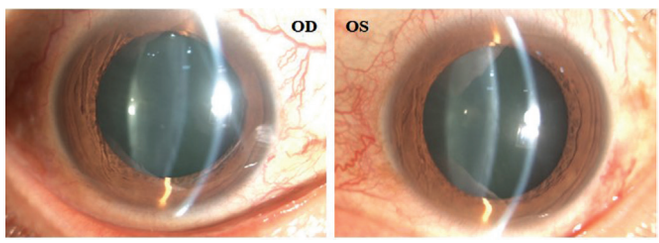

Figure 3 Biomicroscopic images Mild conjunctival hyperemia and dilated episcleral venous. The anterior chamber depth of both eyes became significantly deeper after pupil dilation.

Additionally, retrobulbar injections of dexamethasone 5 mg were administered for anti-inflammatory treatment. Moreover, a combined cardiopulmonary consultation was performed on the patient. During the treatment period, the patient demonstrated significant improvement in cardiopulmonary function, alongside marked alleviation of ocular symptoms. A repeat ophthalmologic examination revealed visual acuity of 20/32 in both eyes with the using of atropine, and IOP of 18 mm Hg in both eyes, UBM showing normal central anterior chamber depth of 3.18 mm in the right eye and 3.23 mm in the left eye (Figure 5A), and reset of the detached ciliary body in each quadrant, and B-Scan ultrasonography indicating disappearance of choroidal detachment (Figure 5B). Two month later, the visual acuity of this patient was 20/20, and the IOP was normal without any eye drops.

DISCUSSION

Pulmonary hypertension is defined as mean pulmonary artery pressure ≥25 mm Hg measured by right heart catheterization at sea level and resting state, mainly manifesting clinically as dyspnea, chest tightness, chest pain, *etc*[1]. Genetic, developmental, and infectious factors can lead to the development of the disease[5-7]. In this case, there is a family history of pulmonary hypertension and no other possible causative factors. Thus, the patient was considered to have genetic factors related to pulmonary hypertension.

Prolonged pulmonary hypertension can lead to systemic hypoxemia and right heart failure, which in turn increased venous pressure and reduced venous return in multiple extra-pulmonary organ systems, ultimately causing ocular complications when the pressure in the superior vena cava

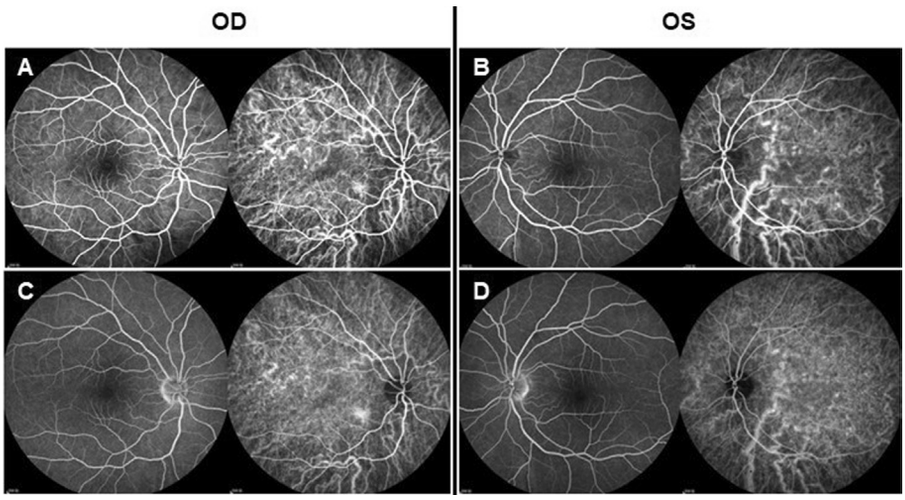

Figure 4 FFA and IGGA images A, B: FFA showed no fluorescein leakage, and ICGA showed the tortuous and dilatate vessels in deep choroid in early phase; C, D: FFA showed no fluorescein leakage, and ICGA showed inhomogeneous fluorescence in later phase. FFA: Fundus fluorescein angiography; ICGA: Indocyanine green angiography.

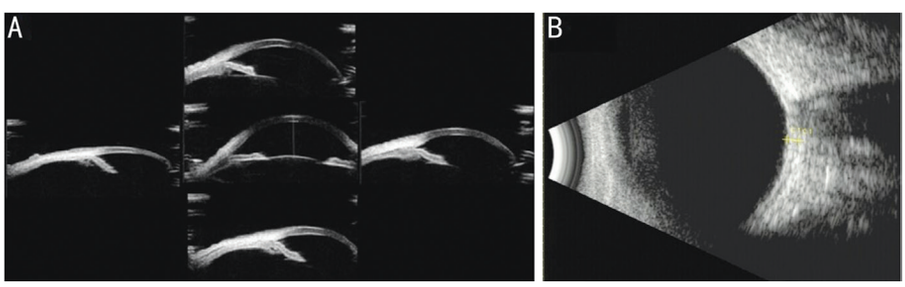

Figure 5 UBM and B-scan ultrasonography after the treatment A: UBM revealed the central anterior chamber was deeper than before, the detached ciliary body was restored and the anterior chamber angles were open; B: B-scan ultrasonography showed the choroidal detachment was disappeared. UBM: Ultrasound biomicroscopy.

and ophthalmic veins increases[8]. In the patient reported in this study, increased pressure in the superior vena cava and ophthalmic veins leads to obstruction of ocular venous return and impaired choroidal circulation. When the filtration rate of the choroidal capillary bed exceeds the normal transscleral outflow, fluid will accumulate in the choroidal interlaminar and suprachoroidal space. Consequently, choroidal thickening and exudative ciliary choroidal detachment will occur. Additionally, ciliary detachment can lead to shallowing of the anterior chamber or even closure of the angle and, consequently, elevated IOP due to increased scleral venous pressure[8-9]. Comparing the visual acuity, IOP, and anterior chamber depth of both eyes before and after the onset of the disease, it was found that the shallow anterior chamber caused the decreased visual acuity of this patient, and the visual acuity could return to normal with the recovery of the anterior chamber depth. Clinically used IOP-lowering therapies include reducing aqueous humor production, promoting aqueous humor drainage, and hypertonic dehydration[10]. However, clinically used IOP-lowering therapies are ineffective in lowering IOP in this disease. Treatment requires joint cardiopulmonary consultation to lower scleral venous pressure based on the treatment of the primary disease, giving topical glucocorticoids and ciliary muscle paralyzing agents to the eye. With the recovery of ciliochoroidal detachment and absorption of fluid in the interlaminar and suprachoroidal space, IOP can return to normal.

Scheider *et al*[11] first reported the development of UES after continuous oxygen therapy in a patient with advanced primary pulmonary hypertension in 1991, suggesting that pulmonary hypertension can lead to the development of UES, and several similar cases have been reported subsequently[5,8]. UES is a syndrome dominated by fundus changes, such as uveal detachment and serous retinal detachment due to impaired vortex venous return and/or abnormal scleral histology. Notably, this is clinically rare, has atypical symptoms, and needs to be differentiated from central placental chorioretinopathy and Vogt-Koyanagi-Harada Disease[12-13]. The combination of exudative retinal detachment, superficial scleral vasodilatation, and FFA showing leopard spot-like fundus changes may further aid in diagnosing UES[14-15]. The patient reported in this study presented clinically with elevated IOP due to ciliary detachment, choroidal detachment, an angle closure, and dilatation of superficial scleral vessels, and

a fundus image showing a leopard-like fundus that met the diagnostic criteria for UES.

In addition to UES, pulmonary hypertension can lead to other ocular complications, including central retinal vein occlusion (CRVO), exudative retinal detachment, and retinal neovascularization[16-18]. CRVO and serous retinal detachment are associated with elevated systemic venous pressure and stagnation of blood flow due to obstructed venous return[18]. Retinal neovascularization is also associated with systemic hypoxemia due to pulmonary hypertension. Hypoxia is the primary stimulus for releasing angiogenic factors such as hypoxia-inducible factor-1a and vascular endothelial growth factor from ischemic tissues. These transcription factors are released during hypoxia to stimulate neovascularization in ischemic tissues, including the retina[18]. In addition, when the choroidal capillary bed filtration rate exceeds the normal choroidal outflow, fluid accumulates not only in the choroidal and suprachoroidal spaces but also in potential spaces such as the subretinal, leading to the development of exudative retinal detachment[17].

Notably, pulmonary hypertension can lead to UES, a rare clinical condition, inducing a forward displacement of the lens-iris diaphragm with transient myopia and intermittent high IOP. Therefore, ophthalmologists need to be more aware of this type of disease to better guide their patients in subsequent treatment and follow-up, and multidisciplinary collaboration in treatment is necessary and appropriate.

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