American Journal of Ophthalmology Case Reports 20 (2020) 100878

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.ajocasereports.com/

Secondary open-angle glaucoma and serous macular detachment associated with pulmonary hypertension

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ARTICLE INFO

Keywords:
Episcleral venous pressure
Pulmonary hypertension
Secondary open angle glaucoma

ABSTRACT

Purpose: We describe a patient with secondary open-angle glaucoma and serous macular detachment associated with pulmonary hypertension.

Observations: A 59 year-old male with pulmonary hypertension presented with vision loss and was noted to have bilateral engorged epibulbar vessels, blood in Schlemm's canal, elevated intraocular pressure, retinal venous tortuosity and serous retinal detachments. Enhanced depth optical coherence tomography (ED-OCT) showed bilateral serous macular detachments with marked choroidal thickening. Fluorescein angiography and indocyanine green angiography revealed choroidal vascular congestion and engorgement. Improvement of subretinal fluid was achieved with systemic control of his venous hypertension, and the intraocular pressure responded to medical anti-glaucoma therapy.

Conclusions: Pulmonary hypertension may be associated with secondary open-angle glaucoma, choroidal engorgement and serous macular detachment, and should be considered in the differential diagnosis of elevated episcleral venous pressure. Management of ocular complications is challenging and requires a multi-disciplinary approach.

1. Introduction

Pulmonary hypertension encompasses a heterogenous group of disorders characterized by elevated pulmonary vascular resistance. As a consequence of chronic right ventricular dysfunction, orbital venous drainage may become impaired resulting in elevated orbital and episcleral venous pressure (EVP). Ocular complications are uncommon but may include secondary glaucoma, retinal vascular occlusion, choroidal effusion, and serous retinal detachment.[1–9] We describe herein a patient with severe pulmonary hypertension who developed secondary open-angle glaucoma and serous macular detachments resembling central serous retinopathy associated with choroidal engorgement documented with enhanced depth optical coherence topography (ED-OCT).

2. Case report

A 59 year-old Caucasian male with a history of pulmonary hypertension was referred for evaluation of bilateral blurry vision. Systemic medications consisted of furosemide 40mg, tadalafil 20mg, ambrisentan 5mg, pantoprazole 40mg, and potassium chloride 20meq all taken orally once daily. Best corrected visual acuity was 20/40 in the right eye and 20/50 in the left eye. Intraocular pressure (IOP) was 22 mmHg in the right eye and 28 mmHg in the left eye using timolol maleate 0.5% twice daily in both eyes and brimonidine tartrate 0.2% twice daily in both eyes. An afferent pupillary defect was noted in the left eye. There were no signs of proptosis or gaze restriction. Prominent dilated and tortuous episcleral vessels were present in both eyes (Fig. 1), and blood was noted in Schlemm's canal in the left eye. As illustrated in Fig. 2, retinal venous congestion and tortuosity was present in the left eye with severe glaucomatous cupping (top right panel) and nasal visual field depression (bottom right panel) with a mean deviation of −19 dB (24-2 SITA Standard, Humphrey Field Analyzer 3, Carl Zeiss Meditec, Dublin, CA). Bilateral macular thickening was visualized.

Fluorescein angiography revealed normal retinal vasculature, and indocyanine green angiochoriography (Fig. 3, top panel) revealed markedly enlarged and engorged choroidal vessels with pronounced hyper-fluorescence and congestion. ED-OCT (Spectralis, Heidelberg Engineering, Franklin, MA) confirmed bilateral serous macular detachments (Fig. 3, middle panel) consistent with central serous

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https://doi.org/10.1016/j.ajoc.2020.100878
Received 25 June 2020; Received in revised form 25 July 2020; Accepted 3 August 2020
Available online 18 August 2020

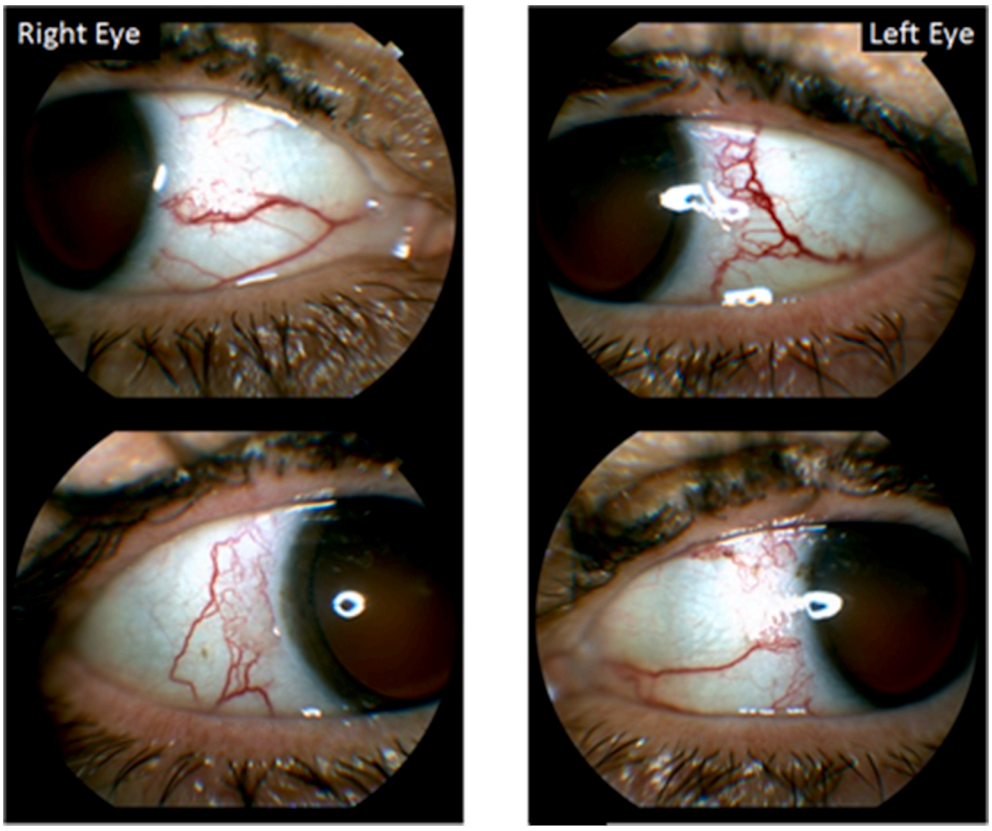

Fig. 1. External photos of both eyes illustrate engorged episcleral vessels.

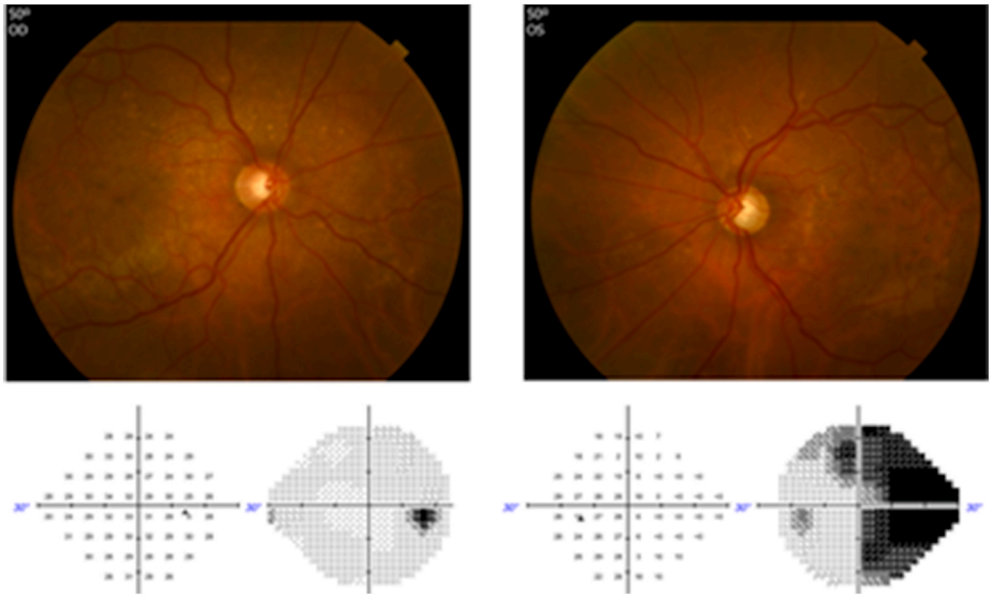

Fig. 2. Fundus photos (top panels) and visual fields (lower panels) of both eyes illustrate moderate glaucomatous cupping and visual field loss in the left eye.

retinopathy. Enhanced depth OCT imaging revealed choroidal thickening in both eyes. Computerized tomography (CT) of the brain and orbit, CT angiogram of the head, and cerebral angiography were unremarkable. Magnetic resonance imaging was not performed due to the presence of a cardiac pacemaker. Neuro-ophthalmologic consultation revealed no evidence of orbital apex syndrome or carotid-cavernous fistula.

Treatment was initiated with latanoprost 0.005% once daily in both eyes and methazolamide 50mg orally twice daily. The patient was referred for cardio-pulmonary consultation. Echocardiography and Swan Ganz catheter monitoring revealed an increased mean systolic pulmonary artery pressure of 67 mmHg, and more aggressive systemic diuresis and vasodilation was recommended including treprostinil, a prostacyclin analogue administered via intravenous pump for systemic vasodilation. After one year of follow-up the IOP improved to 17 mmHg OU, and there was considerable resolution of serous macular fluid in

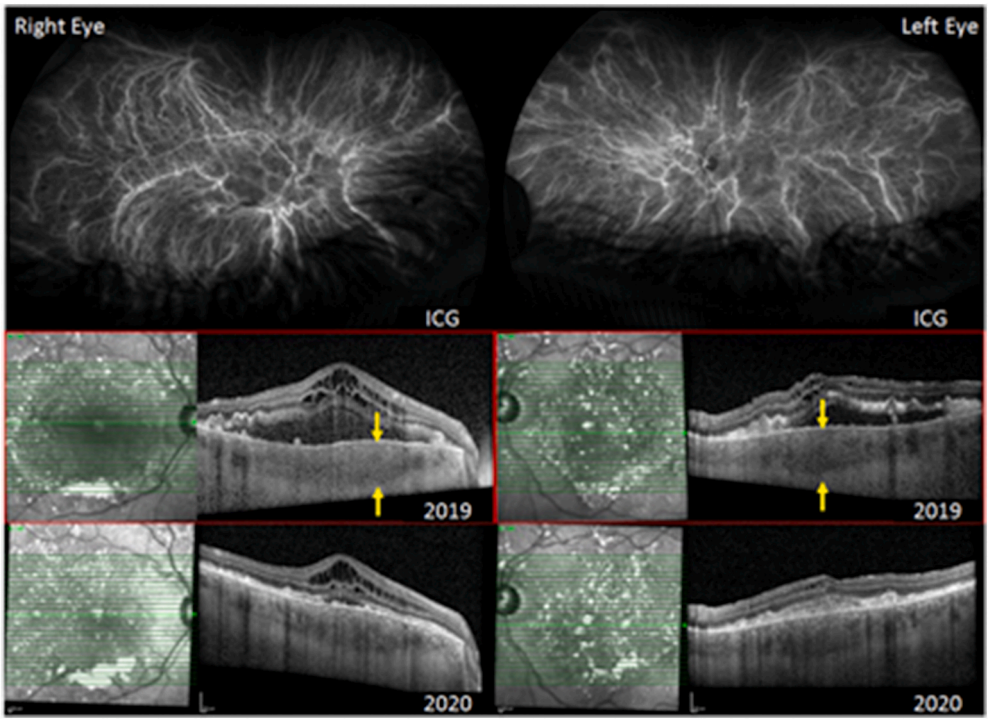

Fig. 3. Indocyanine green angiography (top panels) and enhanced depth optical coherence tomography (middle and lower panels) demonstrate subretinal fluid exudation and choroidal engorgement (yellow arrows) in both eyes. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

both eyes (Fig. 3, bottom panel) with stable vision of 20/50 OU.

3. Discussion

Pulmonary arterial hypertension (PAH) is characterized by remodeling of the pulmonary vasculature which results in elevated right ventricular systolic pressure. PAH is defined as a mean pulmonary arterial pressure of 25 mm Hg or greater at rest.[4,10] Chronically elevated right ventricular systolic pressure leads to right sided ventricular hypertrophy (RVH) in order to maintain cardiac output. Sequela of maladaptive RVH include distension of the internal jugular vein, cavernous sinus and superior and inferior ophthalmic veins.[10] Treatment is aimed at restoring vascular homeostasis and providing symptomatic relief. Medical therapy consists of prostaglandins, phosphodiesterase type 5 inhibitors, endothelian receptor antagonists, diuretics and steroids. Many of the medications have pleiotropic effects and are employed in concert.[4,5,10]

Our patient presented with bilateral submacular effusions with marked choroidal thickening that resembled central serous retinopathy. Other considerations in the differential diagnosis of choroidal thickening include Vogt-Koyanagi-Harada (VKH) disease, pachychoroid pigment epitheliopathy, and pachychoroid neovasculopathy. Elevated systemic venous pressure increases hydrostatic pressure in the choroid and choriocapillaris. When the vascular leakage rate exceeds the normal transscleral flow, it results in fluid accumulation in the choroid and in potential spaces such as the suprachoroidal and subretinal.[8] This can lead to occlusion of precapillary arterioles and obstruction of choriocapillaries blood flow. Uveal effusion, choroidal detachment, exudative retinal detachment and retinal vein occlusion has been described.[1–8] Furthermore, hypoxia induces vascular endothelial growth factor in ischemic tissues including the retina.[4] A multi-disciplinary approach is necessary to achieve cardiopulmonary hemodynamic balance. Our patient achieved improvement in subretinal fluid exudation and choroidal congestion with enhanced systemic vasodilatory therapy.

Few reports have described glaucoma as a consequence of PAH. Mechanisms include secondary open-angle glaucoma associated with elevated EVP, neovascular glaucoma, or angle-closure glaucoma.[4,5,7–9] As demonstrated in our patient, glaucoma due to elevated EVP may present with asymmetric damage. Medical therapy consists of topical and oral IOP-lowering therapy and simultaneous treatment of any associated underlying systemic condition. Although central serous chorioretinopathy has been reported in association with latanoprost use, administration of latanoprost in our patient achieved IOP reduction without concurrent worsening of visual acuity, macular edema or subretinal fluid.[13] In eyes refractory to conservative management, surgical intervention may be required with intraoperative measures to reduce the risk of intraoperative or postoperative choroidal effusion.[11,12]

4. Conclusions

PAH is a serious systemic disorder that may be associated with secondary open-angle glaucoma and serous macular detachment due to increased venous outflow resistance, and should be considered in the differential diagnosis of elevated EVP. To our knowledge this is the first report to demonstrate choroidal engorgement by ICG angiography and ED-OCT imaging. Management of ocular complications requires a multidisciplinary approach.

Patient consent

The patient who is the subject of this case report provided informed consent for publication of this report.

Disclosures and acknowledgements

Financial Funding/Support: P30EY014801 University of Miami core grant; Research to Prevent Blindness unrestricted grant, New York, New York; an unrestricted grant from Mr. and Mrs. Thorne B. Donnelley, Chicago, IL, and the Maltz Family Endowment for Glaucoma Research, Cleveland, Ohio.

Authorship

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

Declaration of competing interest

I Gupta: None.

L Haddock: None.

DS Greenfield: C. Consultant: Aerie Pharmaceuticals, Allergan Inc, Alcon Inc, Bausch + Lomb, Eyenovia, Galimedix, Pfizer Inc, Quark Pharmaceuticals, Santen.

Acknowledgements

None.

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