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Sudden Vision Loss

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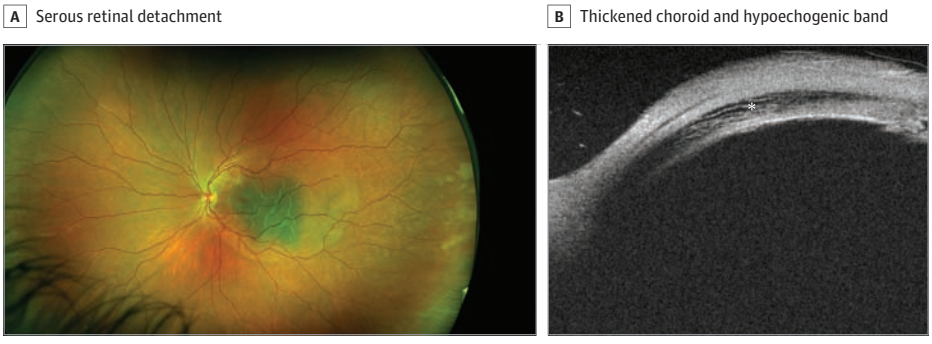

Figure 1. A, Serous retinal detachment in the left eye. B, Ultrasound biomicroscopy of the left eye showed a thickened choroid and a hypoechogenic band in the area of choriocapillaris congestion (asterisk).

A 37-year-old woman was referred for evaluation of a retinal detachment in her left eye. She noted a sudden vision loss in her left eye 3 days prior to presentation. Her Snellen visual acuity on presentation was 20/20 OD and 20/200 OS, and intraocular pressure was normal in both eyes. Anterior segment examination revealed shallowing of the anterior chamber in the left eye. Posterior examination results demonstrated a retinal detachment in the posterior pole with shifting fluid and no identifiable retinal break (**Figure 1**A). No vitreous cell was noted. Findings of imaging testing were notable for thickened choroid with a hyporeflective band on ultrasound biomicroscopy (Figure 1B).

She had a medical history of idiopathic pulmonary arterial hypertension (World Health Organization group 1 pulmonary hypertension) and was currently undergoing evaluation for a bilateral lung transplant. She was currently taking macitentan, an endothelial receptor antagonist; subcutaneous treprostinil, a continuous prostanoid infusion; and riociguat, a guanylate cyclase stimulator. She noted increased dyspnea, exertional hypoxemia, and peripheral edema in the preceding weeks; her diuretic therapy was adjusted and oxygen therapy was added. Chest, abdomen, and pelvis computed tomography demonstrated an ovarian cyst and cardiomegaly. Echocardiography showed findings of severe pulmonary hypertension and severe right ventricular dysfunction.

WHAT WOULD YOU DO NEXT?

A. Perform pars plana vitrectomy for retinal detachment repair

B. Begin high-dose prednisone

C. Work with pulmonary hypertension specialist to optimize pulmonary hypertension management

D. Start eplerenone therapy

CME Quiz at jamacmelookup.com

Diagnosis

Serous retinal detachment due to increased systemic venous pressure in the setting of pulmonary hypertension

What to Do Next

C. Work with pulmonary hypertension specialist to optimize pulmonary hypertension management

Discussion

Pulmonary arterial hypertension (PAH) is a rare condition characterized by pulmonary vasculature remodeling causing right heart failure. Ocular manifestations occur as a result of increased systemic vascular pressure, with distention of the internal jugular vein, cavernous sinus, and ophthalmic veins leading to increased episcleral venous pressure (EVP).[1] Idiopathic pulmonary hypertension accounts for about 10% of all cases of PAH, and most ocular complications are associated with this subtype.[2] Medical management attempts to restore vascular homeostasis. Multiple drug classes are used, including phosphodiesterase type 5 inhibitors, endothelin receptor antagonists, guanylate cyclase, prostaglandins, diuretics, and oxygen. Lung transplant is considered for appropriate patients who are worsening despite maximal medical therapies.

This patient presented with serous retinal detachments. In PAH, increased systemic venous pressure leads to increased hydrostatic pressure in the choroid and choriocapillaris, leading to occlusion of the choriocapillaris blood flow. Ocular manifestations include uveal effusion, choroidal detachment, exudative retinal detachment, and retinal vein occlusion. Glaucoma has also been reported as a rare complication due to angle-closure glaucoma, neovascular glaucoma, and elevated EVP.[3-8]

Aggressive management of PAH and right heart failure is the mainstay for treatment of ocular manifestations along with intraocular pressure–lowering medications. Surgical repair of exudative retinal detachments is not indicated and can lead to iatrogenic complications (option A). Early studies of eplerenone (option D) for treatment of CSCR showed promising results, but further randomized clinical trials demonstrated no benefit of eplerenone.[9] While VKH can be considered in this patient and treated with prednisone, the lack of intraocular inflammation makes this diagnosis highly unlikely. Oral steroids (option B) would have been the treatment choice in a patient with VKH, but the lack of other features of VKH, including anterior chamber cells and vitreous cells, makes this diagnosis less likely. In this patient, there was evidence for worsening PAH and right heart failure with worsening dyspnea and peripheral edema in association with development of ocular complications.

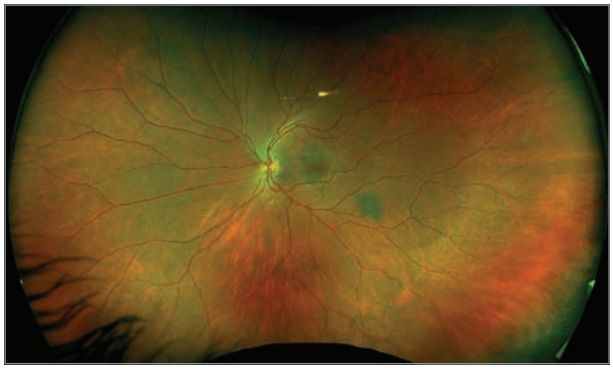

Figure 2. Fundus photograph at follow-up visit demonstrating resolution of the serous retinal detachment.

Patient Outcome

The patient was given supplemental oxygen, diuretics were adjusted, and PAH therapies were changed from riociguat to sildenafil, while macitentan was continued and treprostinil was increased. She subsequently underwent bilateral lung transplant for her pulmonary hypertension and subjectively reported immediate vision improvement on extubation from the transplant and further improvement in her vision in the subsequent days. On her repeated examination 20 days later, her serous retinal detachment and anterior segment narrowing had resolved (**Figure 2**). The combination of her transplant and change in medications decreased her systemic arterial pressure, leading to reduced EVP and ultimately resolution of her serous choroidal detachments. Her best-corrected visual acuity improved to 20/30 OS without any ophthalmic intervention.

ARTICLE INFORMATION

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Additional Contributions: We thank the patient for granting permission to publish this information.

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