Received: 20 February 2017 | Accepted: 18 April 2017

DOI: 10.1002/jgf2.89

IMAGES IN CLINICAL MEDICINE

Journal of General and Family Medicine Open Access WILEY

Bilaterally dilated episcleral vessels in patients with heritable pulmonary arterial hypertension

Meri Watanabe MD | Shinji Makino MD, PhD | Hiroto Obata MD, PhD

Department of Ophthalmology, Jichi Medical University, Shimotsuke, Japan

Correspondence
Shinji Makino, Department of Ophthalmology, Jichi Medical University, Shimotsuke, Japan.
Email: makichan@jichi.ac.jp

KEYWORDS: episcleral vessel, heritable pulmonary arterial hypertension, intraocular pressure

A 51-year-old woman complaining of blurry vision in her left eye for the previous 6 months was referred to our hospital. Her medical history was indicative of heritable pulmonary arterial hypertension (HPAH) since when she was in her 30s, which was confirmed on the basis of a mutation in the gene encoding bone morphogenic receptor 2 (BMPR2). The latest examination of the right heart catheterization (RHC) demonstrated pulmonary artery pressure (PAP) values of 43/23 (mean 32) mmHg, right atrial pressure (RAP) of 6 mm Hg, and pulmonary capillary wedge pressure (PCWP) of 8 mm Hg on treatment with bosentan (125 mg/d) and intravenous epoprostenol (43 ng/kg/min). She was clinically stable (World Health Organization (WHO) functional class II). During the initial examination, her best-corrected visual acuity was 1.2 in the right eye and 0.06 in the left eye. Intraocular pressure was 15 mm Hg in the right eye and 28 mm Hg in the left eye. Slit-lamp examination revealed bilaterally dilated port-wine-colored episcleral vessels, which had been present since when

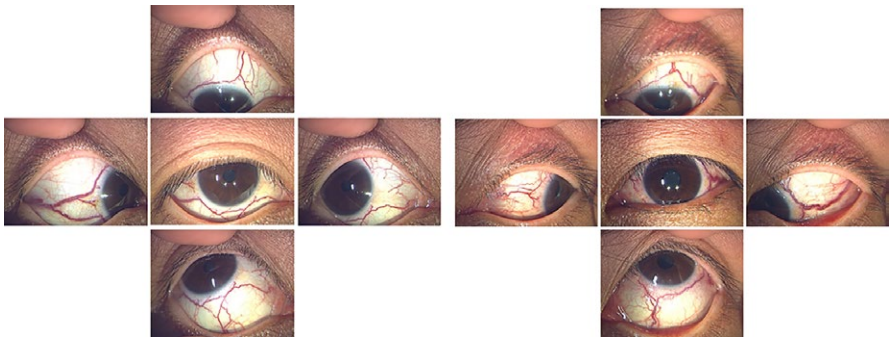

FIGURE 1 Slit-lamp photographs show bilaterally dilated port-wine-stained episcleral vessels in a 51-year-old woman

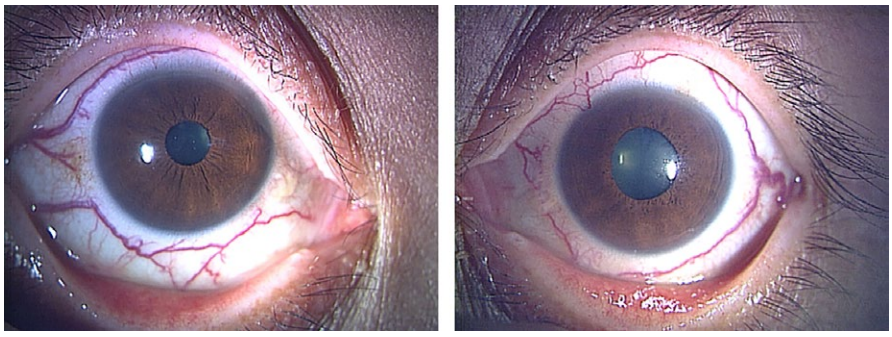

FIGURE 2 Slit-lamp photographs of her 25-year-old son

she was in her 20s (Figure 1). Fundus examination revealed optic atrophy in the left eye.

When we examined her 25-year-old son who is also diagnosed with HPAH, we similarly observed bilaterally dilated port-wine-colored episcleral vessels (Figure 2). His latest examination of the RHC demonstrated PAP values of 51/26 (mean 34) mm Hg, RAP of 4 mm Hg, and PCWP of 4 mm Hg on treatment with macitentan (10 mg/d), tadalafil (40 mg/d), and intravenous epoprostenol (98.2 ng/kg/min). He was clinically stable (WHO functional class II).

Heritable pulmonary arterial hypertension is a rare, fatal, autosomal dominant disease in which idiopathic obliteration of the pulmonary arterial capillaries results in a right heart failure. Only few cases with pulmonary arterial hypertension (PAH) and ocular manifestations of the anterior segment have been previously reported.[1,2] In particular, there was only one case report that PAH was diagnosed via its ophthalmic features.[1] Their clinical findings are similar to those in our cases. We were able to rule out any conditions known to cause dilated episcleral vessels, such as venous obstruction and arteriovenous shunts. Although the both cases were well managed by medications, we suggest that PAH-induced elevation of venous pressure contributed to episcleral vessels dilation. In addition, we speculate that increased episcleral venous pressure might inhibit normal aqueous outflow and increase the intraocular pressure in her left eye. However, the reason why it occurred on one side is unknown.

Although this present finding that was already diagnosed as PAH, we emphasize that symmetrically dilated port-wine-colored episcleral vessels may be observed in patients with a more common PAH sequela and should be considered in the differential diagnosis of the red eye. Accumulation of reports may be needed.

CONFLICT OF INTEREST

The authors have stated explicitly that there are no conflicts of interest in connection with this article.

ORCID

Shinji Makino http://orcid.org/0000-0002-0732-3254

REFERENCES

1. Faure C, Miocque S, Fleury L, Robert MP. Primary pulmonary arterial hypertension diagnosed via its ophthalmic features in an adult: diagnosis and therapeutic challenges. Br J Ophthalmol. 2012;96:459–60.
2. Saran BR, Brucker AJ, Bandello F, Verougstraete C. Familial primary pulmonary hypertension and associated ocular findings. Retina. 2001;21:34–9.

How to cite this article: Watanabe M, Makino S, Obata H. Bilaterally dilated episcleral vessels in patients with heritable pulmonary arterial hypertension. *J Gen Fam Med*. 2017;18:464–465. https://doi.org/10.1002/jgf2.89