

Multifocal Choroiditis and Panuveitis in a Patient with Von Willebrand's Disease**Shinji Makino**

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Abstract: We present a case of multifocal choroiditis and panuveitis in a 69-year-old woman with von Willebrand's disease. Vitreous hemorrhage, anterior chamber inflammation and iris neovascularization were observed in both eyes. Fundusoscopic examination revealed multiple retinochoroidal lesions. To our knowledge, no previous report has described inflammatory ocular disease in a patient with von Willebrand's disease. We discuss the possible mechanisms for this association.**Keywords:** multifocal choroiditis and panuveitis, von Willebrand's disease.

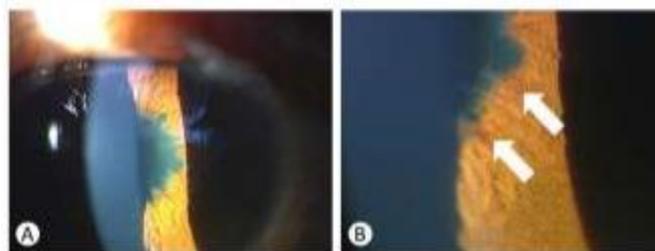
INTRODUCTION

Multifocal choroiditis and panuveitis (MFCPU) is a bilateral chronic uveitis characterized by multiple punched-out chorioretinal lesions [1-6]. Von Willebrand's disease (VWD) is a hereditary coagulation disorder characterized by an abnormality or deficiency of von Willebrand factor (VWF), which promotes platelet adhesion and aggregation and also acts as a carrier of coagulation factor VIII [7]. Ocular involvement in patients with VWD has rarely been reported [8, 9]. To our knowledge, no previous report has described inflammatory ocular disease in a patient with VWD. Herein, we report a case of MFCPU in a 69-year-old woman with VWD.

CASE REPORT

A 69-year-old woman complaining of blurred vision in both eyes was referred to our hospital. She had

a 40-year history of VWD, and her son also had VWD. Upon initial examination, her best-corrected visual acuity (BCVA) was 0.8 in the right eye and 0.7 in the left eye. Intraocular pressure (IOP) was 15 mm Hg in the right eye and 28 mm Hg in the left eye. Slit-lamp examination showed cortical opacities in both lenses. Fundus examination showed no abnormalities. Two months after the initial visit, the BCVA in the left eye had decreased to 0.2 due to mild vitreous hemorrhage. One month later, a vitreous hemorrhage occurred in the right eye. Although this vitreous hemorrhage improved after two months, an anterior segment examination revealed anterior chamber cell inflammation and flare in both eyes. Iris neovascularization with posterior synechiae also was observed in the right eye (Fig-1). The IOP was 17 mmHg in the right eye and 24 mmHg in the left eye.

**Fig-1: Slit lamp photographs of the right eye**

Note posterior synechiae (A) and iris neovascularization (B, arrows).

One month later, posterior synechiae had developed in both eyes, and cortical opacities had progressed in both lenses. The patient's BCVA had

deteriorated to 0.02 in the right eye and 0.07 in the left eye. Therefore, a pars plana vitrectomy with lensectomy was performed in the right eye (Fig-2).

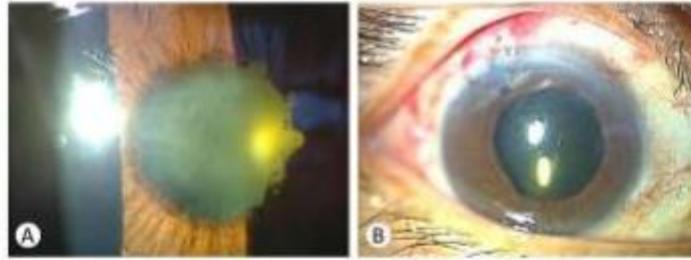


Fig-2: Pre- (A) and post-operative (B) photographs of the right eye

A postoperative fundus examination showed multiple retinochoroidal lesions in the right eye (Fig-3).



Fig-3: Fundus photographs of the right eye
Note multiple retinochoroidal lesions detected in the right eye.

Fluorescein angiography revealed these lesions as clearly defined hyperfluorescent spots (Fig-4). Retinal ischemia was not detected.

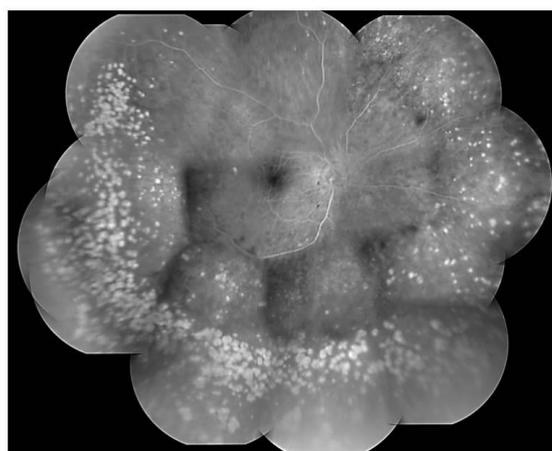


Fig-4: Fluorescein angiography of the right eye
Note hyperfluorescence corresponding to retinochoroidal lesions.

From these findings, the patient was diagnosed with MFCPU. Six months later, a pars plana

vitrectomy with lensectomy was performed in the left eye. A postoperative fundus examination showed

multiple retinochoroidal lesions in both eyes (Fig-5).

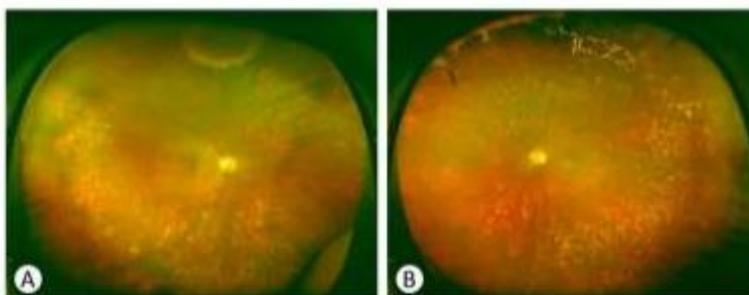


Fig-5: Fundus photographs of the (A) right and (B) left eyes
Note multiple retinochoroidal lesions in both eyes.

Although the patient's BCVA improved to 0.4 in the right eye and 0.3 in the left eye, the left IOP had increased to 35 mm Hg, and iris neovascularization

had developed in the left eye. An Ahmed tube shunt was inserted into the left eye (Fig-6).

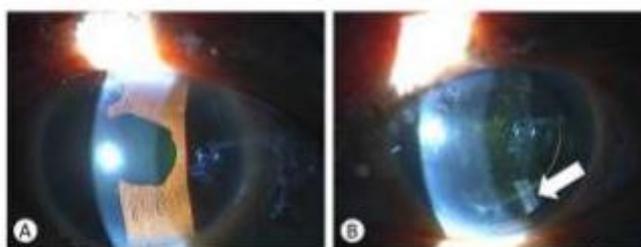


Fig-6: Slit lamp photographs of the (A) right and (B) left eyes

Note no iris neovascularization in the right eye and the Ahmed tube shunt inserted into the left eye (arrow).

Postoperatively, the left IOP reduced to 12 mmHg. Her BCVA was maintained at 0.7 in the right eye and 0.3 in the left eye during a one-year follow-up period.

DISCUSSION

To our knowledge, only two cases of retinal and vitreous hemorrhage associated with VWD have been reported [8, 9], although it is expected that such intraocular bleeding events might remain undiagnosed or unreported [9]. The present report describes bilateral vitreous hemorrhage in a VWD patient.

Neovascular glaucoma occurs when new fibrovascular tissue proliferates onto the anterior chamber angle, obstructing the trabecular meshwork. Retinal ischemia is thought to be the main stimulus. Uveitis without retinal ischemia is a rare cause of neovascular glaucoma. The present case is the first report of neovascular glaucoma secondary to MFCPU in the absence of retinal ischemia.

Furthermore, no previous reports have described MFCPU in a patient with VWD. We considered the possibility that VWF might play a role in the relationship between MFCPU and VWD. VWF is a large glycoprotein that is synthesized in the vascular

endothelium, and high observed levels of VWF reflect endothelial cell perturbation or damage. First, elevated factor VIII-VWF antigen levels are associated with vascular endothelial injury in several disorders, including scleroderma, Raynaud's phenomenon, polymyalgia rheumatic, and temporal arteritis. King *et al.* [10] evaluated factor VIII-VWF antigen levels in patients with serpiginous choroidopathy. They observed a mean factor VIII-VWF activity of $226 \pm 7.3\%$ in patients, compared with $107 \pm 28\%$ in a disease-free, age- and sex-matched control group. These findings suggest that in some patients, serpiginous choroidopathy represents an occlusive vascular phenomenon that involves choroidal circulation. In the present case, however, the VWF antigen level was 57.0%, and the VWF activity level was 28%. Second, Palmer *et al.* [11] evaluated VWF levels in patients with ischemic or non-ischemic retinal vasculitis. They reported significantly higher levels of VWF in patients with ischemic retinal vasculitis than in patients with non-ischemic retinal vasculitis. However, neither group of patients had VWF levels outside the normal range. In the present case, therefore, we suspect that the association of MFCPU and VWD most probably is coincidental rather than causative. Additional cases will need to be examined in order to characterize further

these rare and unusual associations between MFCPU and VWD.

DISCLOSURE

The authors have no conflicts of interest to disclose.

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