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Case Report

Changes in Choroidal Circulation in a Case of Retinitis Pigmentosa Complicated with Macular Hole Before and After Vitreous Surgery

Kazufumi Tanaka and Ryuya Hashimoto*

Department of Ophthalmology, Toho University Sakura Medical Center, Sakura, Chiba, Japan

***Corresponding Author:** Ryuya Hashimoto, Department of Ophthalmology, Toho University Sakura Medical Center, Sakura, Chiba, Japan.

Received: February 20, 2024; **Published:** March 04, 2024

Abstract

Background: Changes in the choroidal blood flow (CBF) of patients with advanced retinitis pigmentosa (RP) complicated by a full-thickness macular hole (FTMH) were evaluated using laser speckle flowgraphy (LSFG) before and after vitreous surgery.

Case: A 48-year-old Japanese female who was diagnosed with advanced RP presented with worsening and blurred vision in the left eye. Optical coherence tomography (OCT) at the initial visit revealed an FTMH in the left eye. The results of the LSFG demonstrated that the mean blur rate (MBR) in the macular lesion, which indicates macular blood flow, was lower in the affected eye (9.1 AU) (arbitrary units [AU]) than in the unaffected eye (10.6 AU). We performed a 25-gauge vitrectomy and ILM peeling using a hemi-inverted ILM flap technique in the left eye. Postoperative follow-up showed successful closure of the FTMH, with an increased in the macular blood flow to 11.8 AU and 13.2 AU at one and six months post-surgery, respectively along with improved visual acuity.

Conclusion: Increased choroidal circulation was observed in a case of RP complicated by an FTMH after 25-gauge vitrectomy and the hemi-inverted ILM flap technique. The results suggest that a closer FTMH using the hemi-inverted ILM flap technique may improve macular blood flow and visual acuity.

Keywords: *Retinitis Pigmentosa; Macular Hole; Choroidal Blood Flow; Laser Speckle Flowgraphy; Hemi-Inverted ILM Technique; Vitrectomy*

Introduction

Retinitis pigmentosa (RP) is a genetically inherited disorder of the retina that causes photoreceptor and retinal pigment epithelium (RPE) degeneration, with a global prevalence of approximately one in 4,000 individuals [1]. Macular complications are notably more prevalent in RP patients than in the general population, and the most frequently observed macular abnormalities in RP patient are cystoid macular edema (CME), epiretinal membrane (ERM), and vitreomacular traction (VMT) [2-4]. Additionally, full-thickness macular hole (FTMH) is a significant complication in RP patients, with a prevalence ranging from 0.5% to 4.5% [2-4].

Regarding the pathogenesis of RP, several studies have hinted at the involvement of impaired ocular blood flow in the pathogenesis of RP. Previous studies using laser doppler velocimetry have found that retinal blood flow is decreased in patients with RP compared with their corresponding age-matched controls [5,6]. Regarding choroidal blood flow, Falsini., *et al*. found that subfoveal blood flow, as assessed

by laser Doppler flowmetry, was diminished in individuals with RP, and was associated with the functionality of central cones, determined via focal electroretinograms (FERGs) [7]. However, to the best of our knowledge, changes in choroidal circulation in cases of retinitis pigmentosa (RP) complicated with FTMH using laser speckle flowgraphy (LSFG-NAVI®; Softcare Co. Ltd., Fukuoka, Japan) before and after vitreous surgery have yet to be elucidated.

LSFG is a non-invasive quantitative method that is very useful for measuring and assessing optic nerve head [8-10] and choroidal blood flow [11-13] based on the changes in the speckle pattern of the reflection of laser light from the fundus photo [14]. The mean blur rate (MBR) is a parameter of blood flow speed.

Herein, we demonstrate a change in macular blood flow in a patient with advanced RP complicated by FTMH using LSFG before and after vitreous surgery.

Case Presentation

A 48-year-old Japanese female who was diagnosed with advanced RP presented with worsening and blurred vision in the left eye. She was referred to the Toho University Sakura Medical Center in Sakura, Japan, for a detailed eye examination.

At the initial visit, her best-corrected visual acuity (BCVA) in the right and left eyes was 20/20 and 20/50, respectively. Funduscopic examination revealed advanced RP, including peripheral retinal atrophy with bone spicule-shaped pigmentation in the mid-periphery, attenuation of the retinal vessels, and atrophy of the optic nerve head in both eyes (Figure 1A and 1B). Optical coherence tomography (OCT) showed thinning of the outer nuclear layer and an obliteration of the demarcation between the ellipsoid zone and the retinal pigment epithelium in both eyes (Figure 1C and 1D). Additionally, an FTMH was observed in the left eye. Goldman visual fields revealed tunnel vision in both eyes (Figure 1E and 1F).

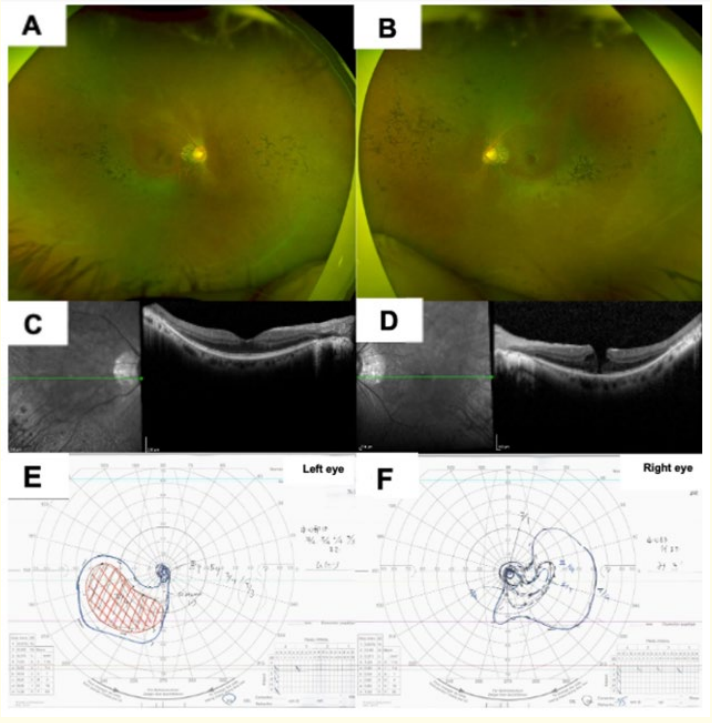

Figure 1: *Color fundus photography and optical coherence tomography findings and Goldman visual field at the initial visit. Color fundus photography in the right (A) and left (B) eyes. Optical coherence tomography (OCT) findings in the right (C) and left (D) eyes. Goldman visual field in the left (E) and right (F) eye.*

We measured the macular blood flow in both eyes using LSFG. LSFG composite color images of both eyes are shown in figure 2A and 2B. The results demonstrated that the MBR in the macular lesion, which indicates macular blood flow, was lower in the affected eye (9.1 AU) (arbitrary units [AU]) than in the unaffected eye (10.6 AU).

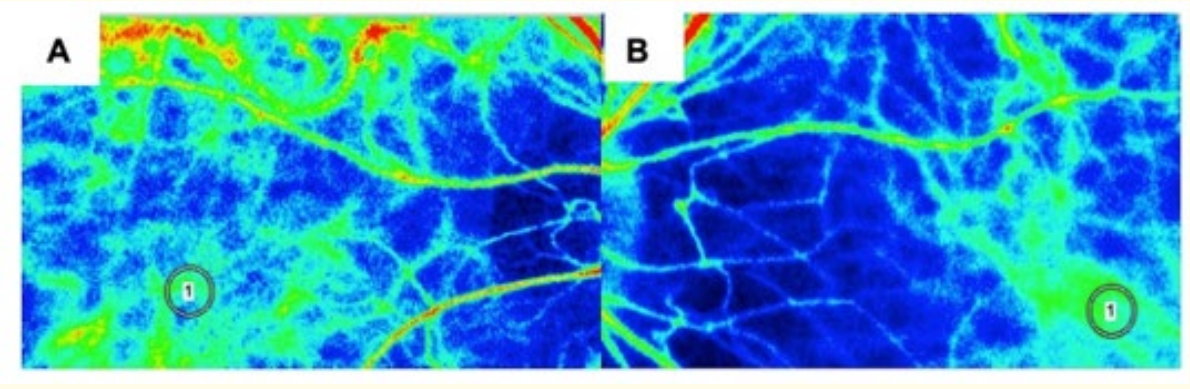

Figure 2: *Laser speckle flowgraphy (LSFG) color composite image at the initial visit. A LSFG composite color map in the right (A) and left (B) eye. Cooler color was observed in the left eye with full-thickness macular hole compared to that in the right eye.*

We diagnosed the patient with advanced RP complicated with FTMH and performed microincision vitrectomy with a 25-gauge instrument using a Constellation vented gas forced infusion and IOP control system (Alcon, Fort Worth, Texas), ILM peeling with hemi-inverted ILM flap technique and 20% SF6 gas tamponade on the left eye under local anesthesia via retrobulbar injection of 2% lidocaine hydrochloride (Xylocaine, 2.5 mL; AstraZeneca K.K., Osaka, Japan) and 0.75% ropivacaine hydrochloride (Anapeine, 2.5 mL; AstraZeneca K.K., Osaka, Japan). During surgery, the constellation vented gas forced infusion and IOP control system were set in two steps of different pressures: lower pressure (8 mmHg) and higher pressure (15 mmHg).

One month after vitreous surgery, OCT findings showed successful closure of the FTMH (Figure 3A), and the patient's BCVA in the left eye improved to 20/40. LSFG color maps of the left eye indicated that the number of warm colors was higher than that at the initial visit (Figure 3B), and the rate of macular blood flow was found to increase to 11.8 AU. Six months after surgery, the patient's BCVA in the left eye improved to 20/32, and a FTMH remained closed (Figure 3C). Macular blood flow further improved to 13.2 AU (Figure 3D).

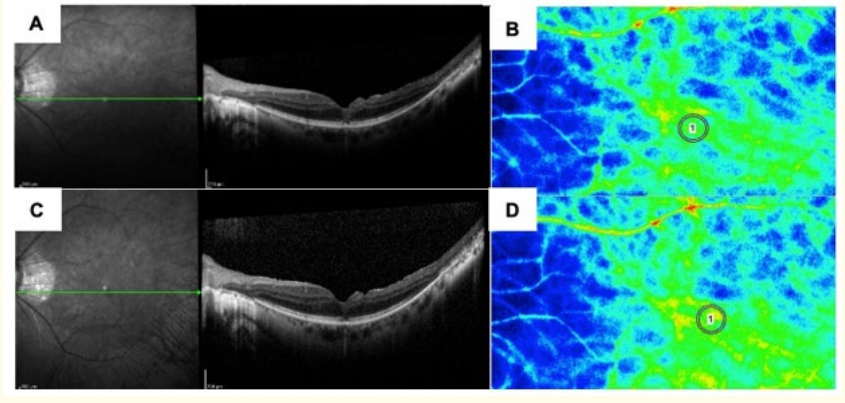

Figure 3: *Change in optical coherence tomography (OCT) findings laser speckle flowgraphy (LSFG) color image before and after surgery. A significant increase in the warmer colors was observed in LSFG findings after performed a closer full-thickness macular hole (FTMH). OCT and LSFG color map of the right eye one month after surgery (A, B). OCT and LSFG color map of the right eye six months after surgery (C, D).*

Next, we measured systolic blood pressure (SBP) and diastolic blood pressure (DBP) using an automated sphygmomanometer (HBP-9020; OMRON Healthcare, Kyoto, Japan), in the sitting position. At the same time, we also measured the rate of macular blood flow since there is a linear relationship between choroidal blood flow and ocular perfusion pressure (OPP) [15]. The mean blood pressure (MBP) was calculated from the SBP and DBP values using the following equation: MBP = 1/3 (SBP - DBP) + DBP [16,17]. The following equation was used to calculate OPP: OPP (sitting position) = (2/3 × MBP) - IOP [16,17].

The OPP of the left eyes was 35.9 mmHg at the initial visit, 33.2 mmHg at the one-week follow-up, and 29.8 mmHg at the six-week follow-up, although choroidal blood flow increased after surgery.

Discussion and Conclusion

This study is the first to report increased choroidal blood flow in a patient with RP complicated by FTM after vitrectomy and the hemi-ILM inverted flap technique using the non-invasive LSFG method.

At the patient's initial visit, macular blood flow and BCVA were lower in the left eye (with FTMH) than in the right eye (unaffected eye). Previous reports have shown that macular MBR can decrease to 75% in patients with RP compared with control subjects, and MBR has been reported to be significantly correlated with VA and mean deviation (MD) values [18]. Regarding FTMH, another study demonstrated that subfoveal choroidal blood flow was significantly lower in the eyes of patients with stage 4 and stage 1a FTMH compared to the eyes of age-matched controls [19]. Although we have not compared the macular MBR with that of healthy individuals, this result is highly consistent with that of previous research.

Generally, the outer retina is the site of many insults, and the retinal pigmental pigment epithelium (RPE) undergoes some degree of apoptosis in RP patients [20]. Choriocapillary circulation contributes to the metabolism of RPE [21]. In fact, histological animal studies of photoreceptor degeneration have demonstrated that a gradual loss of photoreceptors with secondary reduction of the outer retinal oxygen consumption occurs [22]. Considering the above findings and the fact that macular choriocapillary flow area and parafoveal vessel density were lower in eyes with FTMH than in unaffected eyes [23], reduced metabolic demand in the outer retina may induce a decrease in the rate of macular blood flow in RP eyes complicated with FTMH.

A closer FTMH was performed using the hemi-inverted ILM flap technique. As a result, macular blood flow was found to gradually increase, along with an improvement in BCVA in the operated eye. Previously, Teng et al. showed that the macular choriocapillary flow area of eyes with FTMHs increases significantly compared to the preoperative values one month after vitreous surgery [23]. By contrast, another study using LSFG found that postoperative choroidal blood flow did not differ from preoperative blood flow in patients with FTMH and epiretinal membrane. Regarding the infusion pressure of the vented gas-forced infusion setting during vitreous surgery, we set the IOP to 15 mmHg; however, in a previous study using LSFG, the infusion pressure was set to 8, 20, or 40 mmHg during surgery [24]. Choroidal circulation is known to have a weak autoregulatory capacity, and the luminal area of the choroid is reduced in response to decreased OPP induced by IOP elevation [25]. Taken together, these findings suggest that the discrepancy between previous study using LSFG and the case reported herein may be related to differences in the settings of the intraoperative infusion pressure and the recovery level of choroidal blood flow. Additionally, from the viewpoint of metabolic demand at the outer retina, it is reasonable to posit that an increased outer retinal metabolic demand, along with an improvement in BCVA, may induce elevation of macular blood flow.

To summarize, in this case report, changes in the choroidal circulation in a case of RP complicated by FTMH after 25-gauge vitrectomy and treated using the hemi-inverted ILM flap technique are highlighted. The findings suggest that a closer FTMH using the hemi-inverted ILM flap technique may improve macular blood flow along with BCVA.

Patient Consent

Written informed consent was obtained from the patients' parent for publication of this case report and any accompanying images.

Acknowledgement

We thank editage author services for editing this manuscript.

Conflict of Interest

None.

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Volume 15 Issue 3 March 2024