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Case Report

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Macular Hole Related Retinal Detachment in a Case with Myelinated Retinal Nerve Fiber Layers

Yang C and Chen S*

Department of Ophthalmology, Changhua Christian Hospital, Changhua, Taiwan

***Corresponding author:**

San-Ni Chen,
Department of Ophthalmology,
Changhua Christian Hospital,
Changhua, Taiwan, E-mail: 108562@cch.org.tw

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1. Introduction

Myelinated Retinal Nerve Fiber Layers (MRNFL) are retinal nerve fibers with a myelin sheath. This relative rare condition is usually benign without specific symptoms, but in some cases ocular findings including axial myopia, amblyopia, and strabismus are noticed [1]. However, to our knowledge, few case of Retinal Detachment(RD) combined with macular hole in MRNFL has been reported. Many of the vitreous-retina structural characteristics of RNFL myelination have also not yet been described. Herein, we describe the clinical course, fundus photography, Optical Coherence Tomography(OCT), and the surgical highlight of this eye with both MRNFL and RD.

2. Report of Case

A 48-year-old female was referred to the retina service because of sudden onset blurred vision in the right eye. Her right eye was amblyopic and highly myopic since she was a child. She was otherwise healthy and asymptomatic. The Best Corrected decimal visual acuity (BCVA) of her right eye was 0.02. Color fundus photography of the right eye taken on Aug 3, 2020 (Figure A) showed a macular hole and RD with nasal aspect spared. Extensive MRNFL inside and outside the vascular arcade was also noted. An OCT image showed a macula hole and RD (Figure B).

Due to the severity of condition and compromised vision, vitrectomy to reattach the retina was arranged. During the operation on Aug 25, 2020, after core vitrectomy, triamcinolone acetonide assisted cortical vitreous peeling was performed, however the vitreous was tightly adherent that only the posterior cortical vitreous could be removed till the inner margin of MRNFL. While doing indocyanine green (ICG) assisted internal limiting membrane (ILM), the peeling could only be done till the inner margin of MRNFL. No ILM could be found or stained in the MRNFL area. On the other hand, the ILM was noted to be thin, fragile, tightly adherent, and became shredded while peeling, thus a whole sheet of ILM flap could not be obtained to cover the macular hole and the ILM around the macular hole was left unremoved and inserted into the macular hole. Air-fluid exchange with drainage retinotomy at superior peripheral retina and laser retinopexy around the drainage site was performed. Silicon oil was instilled at the end of surgery for retinal tamponade.

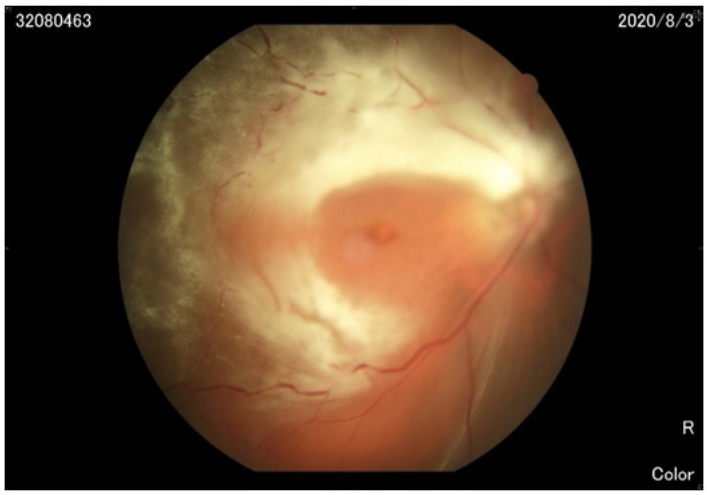

Figure A

The postoperative course was smooth, unless steroid glaucoma with intraocular pressure up to 45 mmHg was noted 1 week later. After discontinue topical prednisolone acetate eye drop and anti-glaucomatous agent, her intraocular pressure returned to normal. The axial length measured at this time was 29.11 mm with the BCVA revered to 0.1. Post-operation color fundus photos of the right eye on Aug 26, 2020 (Figure C) and showed he retina well attached. The OCT image taken on the same day showed attached retina and sealed macular hole (Figure D).

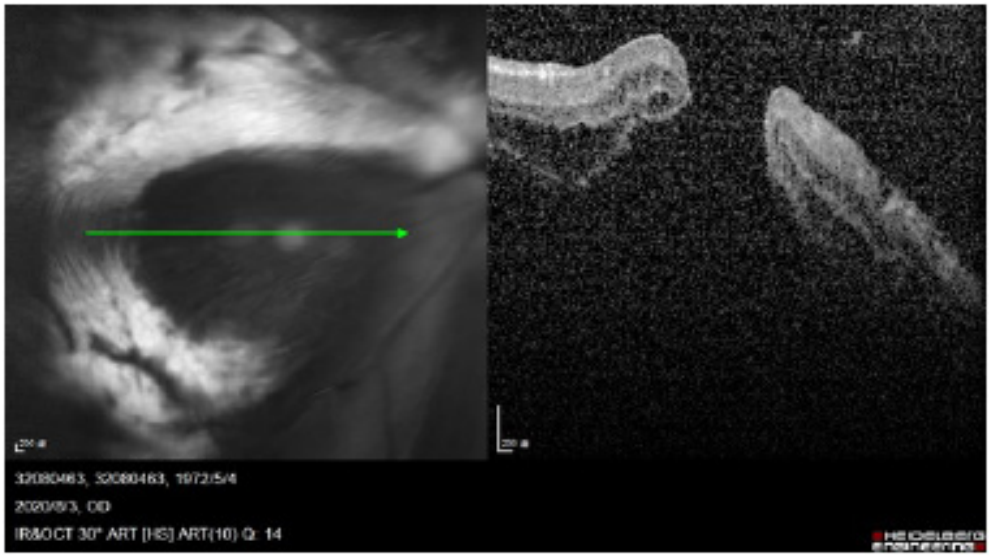

Figure B

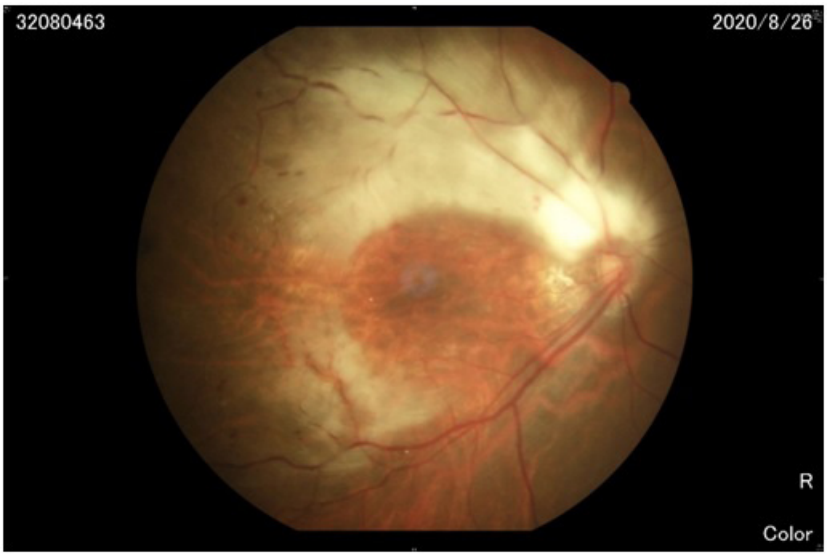

Figure C

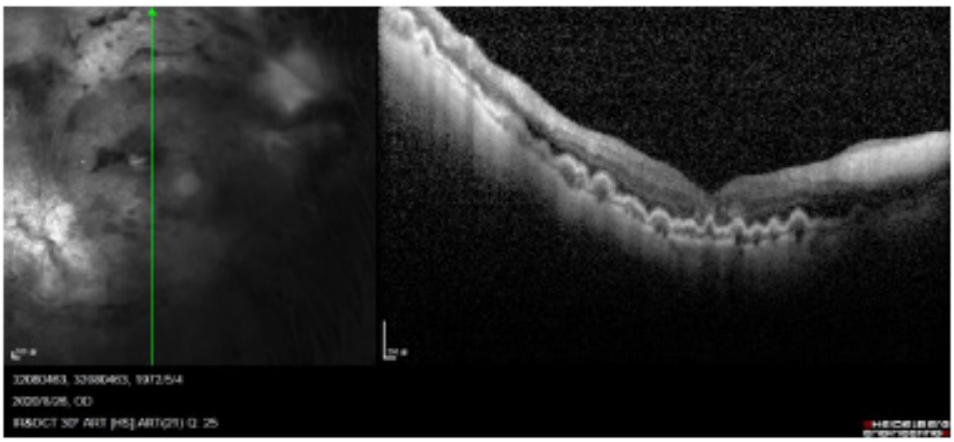

Figure D

3. Discussion

MRNFL are present in 0.57 to 1% of the population and can occur bilaterally in approximately 7% of affected patients [2]. Although often asymptomatic, this condition is sometimes complicated with myopia, strabismus, and amblyopia. More severe ocular diseases reported with this disorder include coloboma, macular aplasia, polycoria, keratoconus. Serious systemic abnormalities include dyscrania, especially oxycephalia, are also reported [3]. Genetic control of the disease are largely unknown and active areas of research. Familial cases of MRNF have been reported in either ocular syndromes only or combined with other systemic conditions [4]. In general fundoscopy, MRNFL appear as white, sharply demarcated patches on the surface of the retina that obscure the underlying retinal vessels. Red-free images and infrared images highlights the white appearance of the MRNFL. On the contrary, MRNFL appears dark in autofluorescence images. OCT images show thickened nerve fiber layers in areas of the MRNFL [5].

Chen et al. reported a case of RD and difficult artificial posterior vitreous detachment(PVD) was noted [6]. Minami et al. described a case with a macular hole with limited MRNFL located outside the macula [7]. However, no RD with macular hole has been reported in MRNFL, and the relationship between ILM and MRNFL remain unknown.

In our case, artificial posterior vitreous detachment could not be induced in the area of MRNFL due to firm vitreo-retinal adhesion in this area, which is compatible to previous reports. Nevertheless, artificial PVD only performed in area inside the inner margin of MRNFL seemed sufficient to improve the patient's visual outcome. As for the difficult peeling of ILM at MRNFL site, instead of tightly adherent ILM and MRNFL, we hypothesize that the ILM doesn't exist at the MRNFL area.

The nature of the ILM remained largely unknown. Kanavi et al. reported the immunoreactive for glial fibrillary acidic protein and neuron specific enolase [8], which indicated the neurogenic nature of this structure. We hypothesize the ILM to be a product of the retinal nerve fiber layer demyelination. Since the demyelination process didn't occur in MRNFL area, the absence of ILM in MRNFL is a corollary consequence.

The nature of MRNFL and ILM are waiting to be discovered. Further histology and biochemistry studies should be done for clarification.

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