Macular Hole Associated with Vogt-Koyanagi-Harada Disease at the Acute Uveitic Stage

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Key Words
Macular hole · Vogt-Koyanagi-Harada disease · Uveitis · Vitrectomy

Abstract
We describe a case with macular hole (MH) associated with Vogt-Koyanagi-Harada (VKH) disease. A 71-year-old Japanese woman presented with visual loss and headaches. The best-corrected visual acuity (BCVA) was 0.02 in the right eye (RE) and 0.1 in the left eye (LE). The patient was diagnosed with VKH based on circumferential choroidal detachments, multiple serous retinal detachments, and optic disc hyperemia. The multiple serous retinal detachments improved with high-dose corticosteroid therapy and gradual tapering. The BCVA was recovered to 1.2/0.7 in the RE/LE. Six weeks after the initial administration of steroid, vitreomacular traction was found by optical coherence tomography in the LE, which progressed to stage 4 MH with the BCVA of 0.2 in the LE. Twenty-three weeks after the initial treatment, vitrectomy was performed with the standard surgical procedures, including inner limiting membrane peeling around the fovea and air tamponade. The MH was closed successfully and the BCVA was 0.4 in the LE 5 weeks after the vitrectomy. This is the first report of a case with MH secondary to the acute uveitic stage of VKH. Successful closure of MH was achieved with the standard surgical intervention for an idiopathic MH. To conclude, at the early stage of VKH, there is a possibility of MH formation due to the rapid progress of vitreous traction following the inflammation, and the surgical procedure could be effective to resolve this secondary disorder.

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Introduction

Vogt-Koyanagi-Harada (VKH) disease is a rare multi-systemic inflammatory disorder which affects tissues containing melanin, including eyes, meninges, and skin [1]. The classic clinical course of VKH is divided into three stages: stage 1 (the prodromal stage) includes flu-like symptoms; stage 2 (the acute uveitic stage) includes auditory and neurological manifestations followed by visual disturbance associated with exudative retinal detachments and optic disc hyperemia, and stage 3 (the convalescent stage) can last for months or be chronic with sunset glow fundus, cutaneous hypopigmentation, and poliosis [1].

Macular hole (MH) secondary to uveitis is known to occur rarely [2], and only two cases with MH secondary to VKH disease at the convalescent stage have been reported in peer-reviewed literatures [3]. The following describes a case with VKH who developed a unilateral MH at the acute uveitic stage.

Case Report

A 71-year-old Japanese woman presented with visual loss accompanied by headaches and tinnitus. No history of ocular disorders was reported except for cataract surgery in the right eye (RE). The best-corrected visual acuity (BCVA) at the initial examination was 0.02 in the RE and 0.1 in the left eye (LE). Slit-lamp ophthalmoscopy revealed inflammatory signs in the anterior chamber and vitreous cavity in both eyes. Comprehensive fundus examinations showed circumferential choroidal detachments, multiple serous retinal detachments, and optic disc hyperemia (fig. 1a–c, fig. 2a). An increased number of mononuclear cells were detected in the cerebrospinal fluid and homozygous HLA-DR4 alleles were identified.

The patient was diagnosed with VKH disease based on the clinical findings, and high-dose corticosteroid therapy with gradual tapering was initiated 2 days after the presentation. The inflammatory signs, choroidal detachments, and serous retinal detachments disappeared within 2 weeks, and the BCVA was recovered to 1.2/0.7 in the RE/LE after 4 weeks (fig. 2b). Six weeks after the initial administration of steroid (i.e. in the gradual tapering period), vitreomacular traction (VMT) was found by optical coherence tomography (OCT) in the LE (fig. 2c). Two weeks later, the VMT progressed to MH (stage 4), and the BCVA dropped to 0.2 (fig. 2d).

Twenty-three weeks after the initial treatment, vitrectomy was performed with 23-gauge instruments. The standard surgical procedures for an idiopathic MH were employed, including inner limiting membrane (ILM) peeling around the fovea and air tamponade. The entire posterior vitreous detachment (PVD), which had been present preoperatively, was confirmed during the operation. After keeping a face-down position for 4 days, the MH was closed successfully. Five weeks after the vitrectomy, the BCVA was improved to 0.4 (fig. 2e). No recurrent inflammatory events associated with VKH have been observed subsequently.

Discussion

A case with MH secondary to the acute uveitic stage of VKH was treated successfully with vitrectomy. To our knowledge, this is the first report of MH which was developed at the acute uveitic stage of VKH. The serial OCT images indicate that the uveitic inflammation induced severe vitreomacular adhesion and PVD formation, which resulted in VMT syndrome and progressed to MH.
Kobayashi et al. [3] reported two cases both at the convalescent stage of VKH that developed MH associated with epiretinal membrane (ERM). Based on the electron microscopic findings of the excised membranes, they speculated that the retinal pigment epithelial (RPE) cells might have migrated/proliferated on the ILM at the earlier uveitic stage; then the MH was formed by adhering of the RPE cells to the vitreous cortex which eventually contracted. While the RPE migration/proliferation in ERM secondary to uveitis has been thought unusual, some investigations have shown its possibility. In a histologic review of 168 ERM samples, Clarkson et al. [4] reported that 1 out of 16 samples which contained RPE cells had a history of chronic uveitis. There is also an immunopathologic study of VKH which suggested RPE migration into the retina [5]. Although there are no available data indicating RPE migration in our case, RPE migration/proliferation could be a cause of the rapid PVD formation and severe VMT at the acute uveitic stage of VKH.

To conclude, at the early stage of VKH, there is a possibility of MH formation due to the rapid progress of vitreous traction, and vitrectomy with ILM peeling could be an effective treatment.

**Statement of Ethics**

The study and treatment protocol was approved by the local Ethics Committee of National Tokyo Medical Center.

**Disclosure Statement**

The authors have no conflicts of interest to disclose in this work.

**References**


M.M. and K.F. contributed equally to this work and should be considered equivalent authors.
Fig. 1. Fundus photographs, fluorescein angiograms, and indocyanine green angiograms of a case with VKH disease who developed MH. a Fundus photography at presentation showed circumferential choroidal detachments at the periphery, multiple serous retinal detachments, hyperemia and edema of the optic disc in both eyes. Fluorescein angiography (b) and indocyanine green angiography (c) detected multiple serous retinal detachments and inflammatory changes at the choroidal level. Images obtained at the early phase are shown on the left side and those obtained at the late phase are shown on the right.
Fig. 2. Serial OCT images of a case with MH secondary to VKH disease. a OCT demonstrated the choroidal folds and multiple subretinal fluids at presentation. b The choroidal folds and multiple subretinal fluids were resolved 4 weeks after the initial administration of steroid. VMT was first observed 6 weeks after treatment (c), and a MH was formed 2 weeks later (d). e Five weeks after vitrectomy (28 weeks after the initial treatment), successful closure of the MH was confirmed by OCT.