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Vogt-Koyanagi-Harada disease in a patient with extreme anisometropia

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ABSTRACT

Purpose: To report Vogt-Koyanagi-Harada (VKH) disease in a patient with extreme anisometropia.

Observations: A 56-year-old woman was referred to our hospital. Her past medical history was significant for amblyopia in the right eye. At the initial visit, decimal best-corrected visual acuity (BCVA) was 0.03 (Snellen equivalent 5/160) in the right eye and 0.03 (Snellen equivalent 5/160) in the left eye, and axial length was 28.44 mm and 22.36 mm, respectively. Anterior chamber inflammation was seen predominantly in the right eye with fibrin exudates. Swept-source optical coherence tomography demonstrated choroidal thickening and folds predominantly in the left eye. Additionally, serous retinal detachment (SRD) was much more evident in the left eye than in the right eye. Subfoveal choroidal thickness (SCT) was 417 μm in the right and over 800 μm in the left eye. Cerebrospinal fluid examination revealed lymphocyte-dominant hypercellularity. Based on these findings, we diagnosed the patient with VKH disease and treated her with a high-dose systemic corticosteroid. One month after the initiation of treatment, SRD in both eyes fully resolved, and SCT decreased to 105 μm in the right and 311 μm in the left eye.

Conclusions and Importance: The marked discrepancy in axial length between the right and left eyes might contribute to the different severity of inflammation in VKH disease.

1. Introduction

Vogt-Koyanagi-Harada (VKH) disease is a bilateral uveitis caused by an autoimmune response against melanocytes and characterized by ciliochoroidal effusion, serous retinal detachment (SRD), and choroidal thickening.[1] Lymphocytes, macrophages, and epithelial cells infiltrate the choroid and provoke an immune response targeting melanocytes in the choroidal stroma, resulting in choroidal thickening.[2] Previous studies have reported a higher prevalence of VKH disease in pigmented races than in Caucasians due to significantly higher choroidal melanin content.[3,4] Although choroidal volume is known to be correlated remarkably with axial length,[5] the relationship between axial length and severity of inflammation in uveitis remains poorly understood. Herein, we report VKH disease in a patient with extreme anisometropia (see Figs. 1–3).

2. Case report

A 56-year-old woman was referred to the University of the Ryukyus Hospital. The patient had been aware of visual impairment for one week before the initial visit, and together with the onset of ocular symptoms, headache, tinnitus, and gray hairs appeared. Her past medical history was significant for amblyopia in the right eye since childhood. The decimal best-corrected visual acuity (BCVA) at the initial visit was 0.03 (Snellen equivalent 5/160) in the right eye and 0.03 (Snellen equivalent 5/160) in the left eye. The axial length was 28.44 mm in the right eye and 22.36 mm in the left eye. Slit-lamp examination revealed inflammatory cells and keratic precipitates in the anterior chambers bilaterally, whereas fibrin exudates only in the right eye. Hyperemia was more severe in the right eye. The laser flare meter showed more inflammation in the right at 188.5 pc/ms than in the left eye at 118.5 pc/ms. Anterior-segment optical coherence tomography (OCT) demonstrated a similar degree of ciliochoroidal effusion in four directions at the superior, temporal, inferior, and nasal points in both eyes. On swept-source OCT, the right eye showed only localized SRD and mild choroidal thickening, whereas the left eye showed severe SRD with fibrin, marked choroidal thickening, and choroidal folds. Subfoveal choroidal thickness (SCT) was 417 μm in the right and over 800 μm in the left eye. Fluorescein angiography showed multiple granular hyperfluorescence, dye pooling, and optic disc hyperfluorescence in both eyes. Indocyanine green

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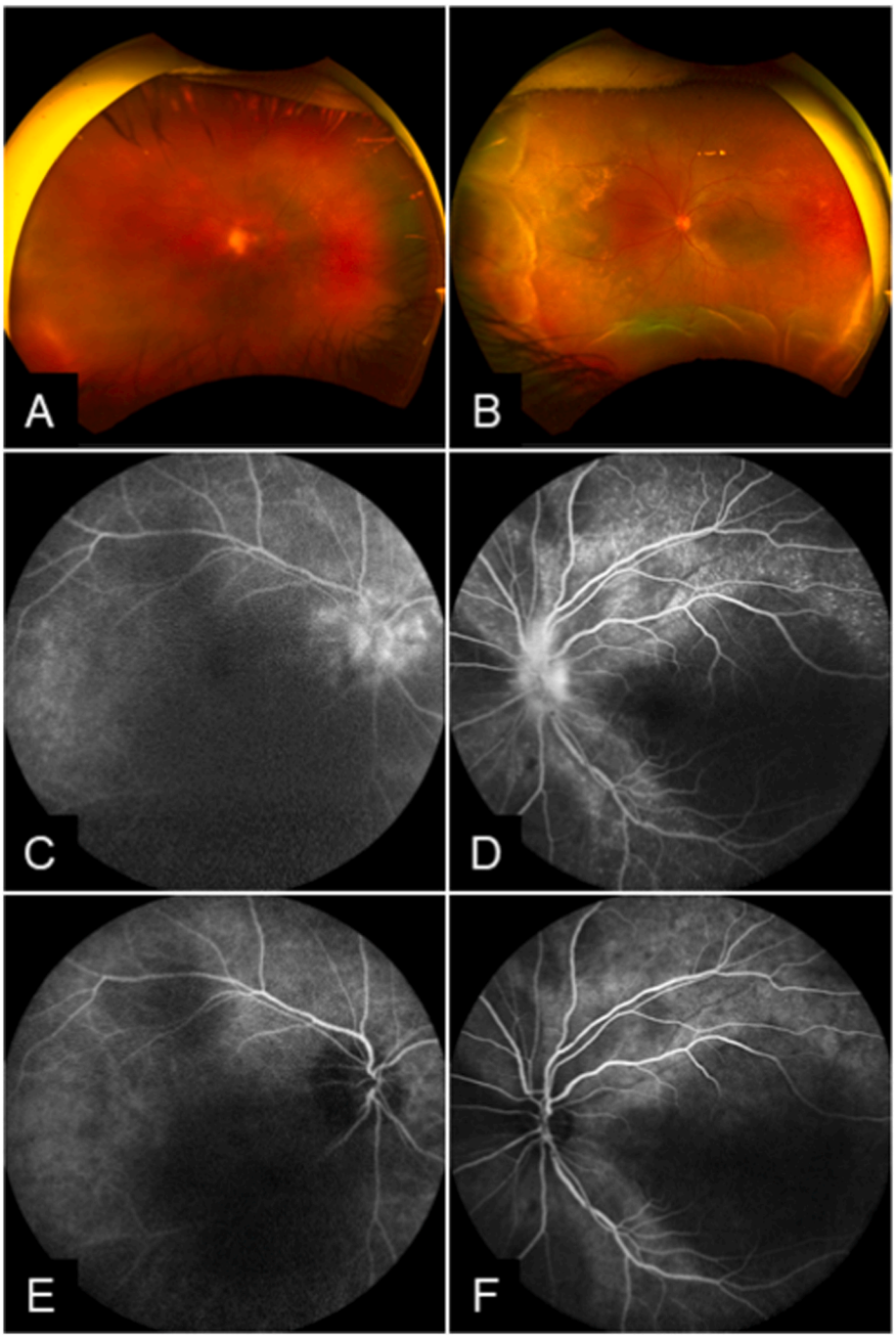

Fig. 1. Photographs at the initial visit. Widefield color fundus photographs of right (A) and left eyes (B) demonstrate mild disc edema in both eyes, poor transparency due to anterior chamber inflammation in the right eye, and bullous retinal detachment on the nasal and inferior side in the left eye. Fluorescein angiography of the right (C) and the left eyes (D) reveals multiple granular hyperfluorescence and optic disc hyperfluorescence. Indocyanine green angiography of the right (E) and the left eyes (F) shows scattered hypofluorescent spots and blocked fluorescence due to the serous retinal detachment in both eyes. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

angiography revealed scattered hypofluorescent spots bilaterally. Cerebrospinal fluid analysis revealed pleocytosis with lymphocyte predominance. Tuberculosis, syphilis, and collagen diseases were ruled out with blood testing. Based on these findings, we diagnosed the patient with a complete type of VKH disease.[6]

Accordingly, we started steroid pulse therapy (intravenous methylprednisolone 1000 mg/day for three days) followed by oral prednisolone 50 mg/day (approximately 1 mg/kg body weight/day) with gradual tapering for six months. Fourteen days after the initiation of medications, anterior chamber inflammation, ciliochoroidal effusion, SRD, and choroidal folds resolved. Thirty-three days after the initiation of medications, decimal BCVA improved to 0.1 (Snellen equivalent 20/200) in the right eye and 1.0 (Snellen equivalent 20/20) in the left eye. BCVA in the right eye did not improve further because of amblyopia. SCT decreased to 105 μm in the right and 311 μm in the left eye. Thereafter, no recurrent inflammation was observed in the anterior and posterior segments of the eyes.

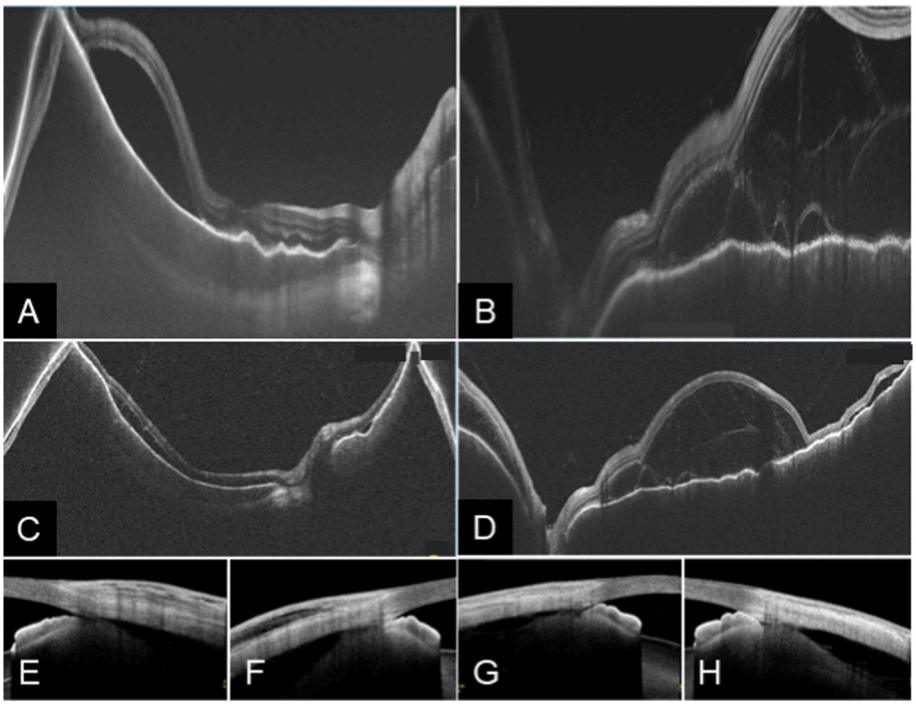

Fig. 2. Horizontal optical coherence tomography (OCT) images through the foveal center of the right (A) and the left eyes (B). OCT shows choroidal folds in both eyes, localized serous retinal detachment (SRD) in the right eye, and severe SRD with fibrin in the left eye. Choroidal thickening of the right was much less than that of the left eye. Subfoveal choroidal thickness was 417 μm in the right and more than 800 μm in the left eye.
Widefield OCT of the right (C) and the left eyes (D) also show localized SRD and mild choroidal thickening in the right eye and severe SRD with fibrin and marked choroidal thickening in the left eye. Anterior-segment OCT demonstrates ciliochoroidal effusion at nasal and temporal points of the right (E), (F) and the left eyes (G), (H), respectively.

3. Discussion

We presented a case of VKH disease in a patient with a marked discrepancy in axial length between the right and left eyes, showing a significantly different severity of inflammation between the eyes, both in the anterior and posterior segments. The immune response of CD4-positive T cells targeting melanocytes in the choroidal stroma leads to VKH inflammation.[1] Inomata et al.[7] performed autopsies on four eyes of two patients in the convalescent stage of VKH disease and uncovered lymphocytic infiltration of the choroid and a marked decrease in melanocytes. In VKH disease, the choroid is the primary site of inflammation, resulting in SRD, choroidal thickening, and obscuration of the choroidal lumen structure, whereas structures comprising melanocytes like the iris and ciliary body are also inflamed. In addition, inflammation of the choroidal stroma gradually extends into the retinal pigment epithelium (RPE) and outer retina.[8]

The characteristic feature of our case was the divergence of inflammatory findings between the anterior and posterior segments; inflammation of the posterior segment was more prominent in the left eye, whereas inflammation in the anterior segment was more intense in the right eye. Fernández-Vigo et al.[9] reported that the volume of the ciliary body is larger in the myopic eye than in the normal eye. The larger ciliary body in the right eye might induce more intense inflammation in the anterior segment. The morphology of the ciliary body may determine the difference in the severity of anterior-segment inflammation between the eyes.

Moreover, a marked difference was observed between the anterior and posterior findings in each eye. According to a study, melanin distribution in the RPE is significantly reduced in eyes with long axial length[10]; however, another study suggested no correlation between axial length and choroidal melanin.[11] Huang et al.[12] reported that in VKH disease, RPE cells are attacked, and melanin is changed to form focal RPE melanin accumulation. The area of focal RPE melanin accumulation significantly correlates with the formation of the sunset-glow fundus. In our case, the lower melanin content in the RPE of the right eye might relate to relatively less SRD.

In terms of the relationship between axial length and choroidal volume, a negative correlation between SCT and axial length has been shown, and both choroidal luminal and stromal areas are reduced in eyes with long axial length.[5] In particular, the choroidal luminal structure is more reduced relatively in longer axial length eyes than in shorter axial length eyes.[13] Furthermore, Wu et al.[14] proposed a state of choroidal circulatory insufficiency in severely myopic eyes. They reported that the outer retinal layer is thinned due to poor choroidal circulation, resulting in a compensatory increase in deep retinal vessels of myopic eyes.[14] In our case, the right eye had a longer axial length, suggesting that the choroidal stroma was reduced and inflammatory cells were difficult to be supplied due to inadequate blood flow. This might result in milder inflammation of the posterior segment in the right eye, and left-right differences in SRD. Interestingly, Horie et al.[15] reported that the choroid does not thicken in the area with patchy atrophy or large peripapillary gamma zone in eyes with sympathetic ophthalmia accompanied by pathologic myopia. The degree of choroidal thickening after the onset of VKH disease may differ markedly depending on the original choroidal thickness, consistent with our results. In addition, in the more myopic eye of a patient with anisometropia, choroidal thinning and choroidal circulatory insufficiency have been reported when compared to the other eye.[16] These facts also support the finding that the posterior ocular inflammation in the right eye was milder in this case. The anatomical and pathophysiological differences between the anterior and posterior segments might cause the difference in inflammation between the right and left eyes in VKH disease.

4. Conclusions

We experienced a case of VKH disease with extreme anisometropia,

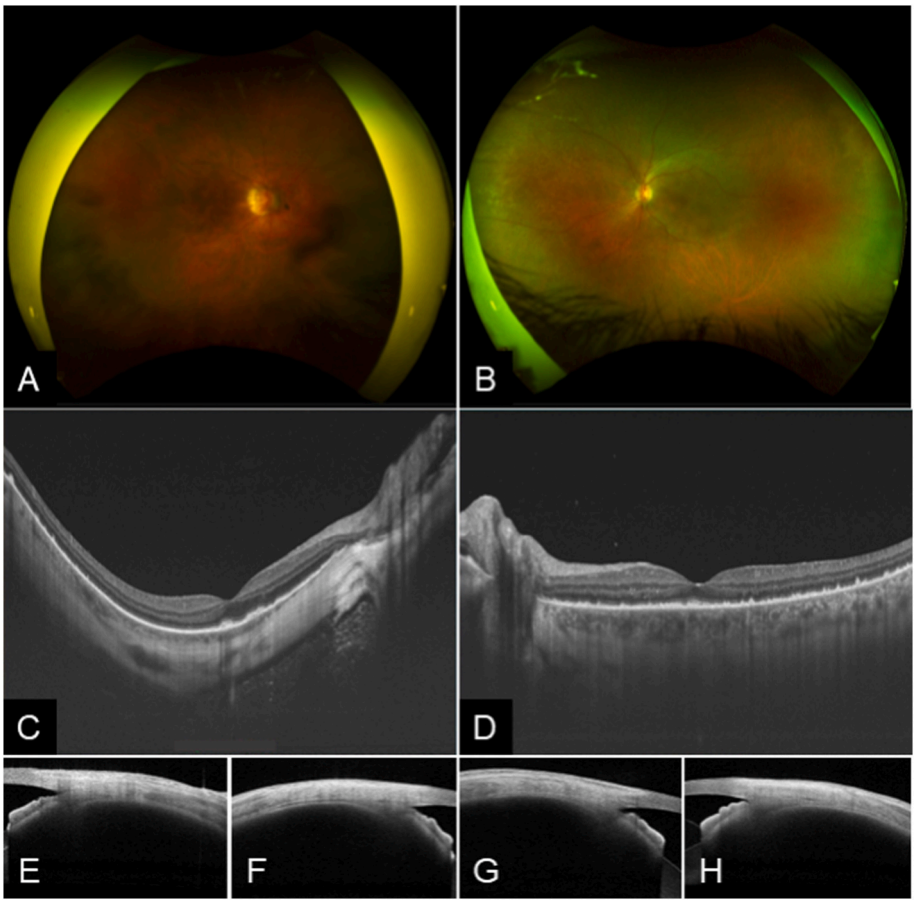

Fig. 3. Photographs at 33 days after the initiation of medications. Widefield color fundus photograph of the right eye (A) demonstrates resolution of disc edema and increased transparency due to the disappearance of anterior chamber inflammation. Widefield color fundus photograph of the left eye (B) shows resolution of disc edema and bullous retinal detachment. Horizontal optical coherence tomography (OCT) through the foveal center of the right (C) and the left eyes (D) show no serous retinal detachment, choroidal folds, and choroidal thickening. Subfoveal choroidal thickness decreased to 105 μm in the right and 311 μm in the left eye. Anterior-segment OCT reveals no ciliochoroidal effusion at nasal and temporal points of the right (E), (F) and the left eyes (G), (H), respectively. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

showing significantly different severity between the eyes in terms of anterior and posterior inflammation. Although we cannot deny the possibility of a coincidental event, differences in choroidal and ciliary body structures and the amount of RPE melanin content due to the difference in axial length may cause a discrepancy in inflammation between the right and left eyes.

Patient consent

The patient provided verbal consent for the publication.

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Authorship

All authors meet the current ICMJE criteria for authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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