ELSEVIER

# Early Manifestation of Vogt-Koyanagi-Harada Disease as Unilateral Posterior Scleritis

Noriko Kouda, Hiroshi Sasaki, Sachiko Harada,
Yoshihisa Yamada, Nobuo Takahashi and Kazuyuki Sasaki

*Department of Ophthalmology, Kanazawa Medical University, Uchinada, Japan*

**Purpose:** To describe a patient with an initial diagnosis of unilateral posterior scleritis who developed bilateral Vogt-Koyanagi-Harada (VKH) disease 12 months later.

**Case:** A 38-year-old woman was diagnosed with posterior scleritis in her right eye at successive examinations. The left eye showed no signs of ocular inflammation.

**Observations:** Her right eye developed a second attack of posterior scleritis 3 months after the initial diagnosis. Twelve months after the first incident, granulomatous uveitis and sunset glow fundus were observed in both eyes. At the same time, lymphocytosis was found in the cerebrospinal fluid, and she was HLA DR-4-positive. In addition, poliosis was noted.

**Conclusions:** The final diagnosis was primarily VKH disease with presenting signs and symptoms of unilateral posterior scleritis. We therefore recommend that when following a patient with posterior scleritis include VKH disease in the differential diagnosis. **Jpn J Ophthalmol 2002;46:590–593** © 2002 Japanese Ophthalmological Society

**Key Words:** Posterior scleritis, steroid therapy, Vogt-Koyanagi-Harada disease.

## Introduction

Vogt-Koyanagi-Harada (VHK) disease usually presents as bilateral panuveitis associated with meningeal irritation. Simultaneous onset in both eyes is common, and in unilateral cases, the second eye is affected within 2 weeks of the first.[1] Posterior scleritis is a relatively rare disease that occasionally shows signs, such as exudative retinal detachment and multiple pinpoint leakages in fluorescein angiography, similar to those of VKH disease. We describe a patient with an initial diagnosis of unilateral posterior scleritis who later developed bilateral VKH disease.

## Case Report

A 38-year-old woman had ocular pain with hyperemia in the right eye for 2 weeks. Her ophthalmologist prescribed topical steroids (betamethasone 0.1%) for the treatment of the conjunctivitis. She then experienced blurred vision and severe ocular pain in the right eye. About 1 month after the initial onset in November 1997, she came to the Kanazawa Medical University Hospital for ophthalmological consultation.

She had no history of ocular abnormalities or any subjective signs including meningeal irritation before the onset of her symptoms. Her corrected visual acuity was 0.6 OD and 1.0 OS, and her intraocular pressure was 12 mm Hg in the right eye and 14 mm Hg in the left eye. Dilation of the sclero-conjunctival vessels was present in the right eye. The anterior chamber showed no inflammatory findings, and the flare value was less than 5.0 PC (photon counts)/ms (Flare-Cell Meter, FC-1000, Kowa, Nagoya). The vitreous body of the right eye showed mild inflammation with (1+) cells.

Fundus examination revealed a serous retinal detachment including a macular lesion (Figure 1, left). The left eye showed no inflammatory findings. Fluorescein angiography of the right eye revealed multiple pinpoint leakages from the lesion in the early phase and serous retinal detachment and pooling of the dye in the late phase (Figure 1, top right). The left eye showed no sign of dye leakage. Indocyanine green angiography showed a filling defect in the choroid below the lesions in the right eye. Ultra-

Received: August 17, 2001
Correspondence and reprint requests to: Hiroshi SASAKI, MD, Department of Ophthalmology, Kanazawa Medical University, Daigaku-machi 1-1, Uchinada, Ishikawa, 920-0265, Japan



Published by Elsevier Science Inc.

0021-5155/02/$–see front matter
PII S0021-5155(02)00545-2

sound and optical coherence tomography showed thickening of the sclera and the choroid in the right eye (Figure 1, bottom right). Laboratory evaluations recorded a normal white blood cell count, negative rheumatoid factor, and a mild elevation of the erythrocyte sedimentation rate (15 mm/h). Cerebrospinal fluid showed no signs of lymphocytosis.

From these findings, she was diagnosed as having unilateral posterior scleritis, and prednisolone (PSL; 60 mg/day) was prescribed. The serous retinal detachment disappeared in 1 week, and the dose of systemic steroid was slowly tapered off.

After 3 months of the systemic steroid therapy at a dose of PSL 15 mg/day, she again complained of blurred vision in the right eye. Her corrected visual acuity was 0.5 OD. No signs of iritis were noted in either eye (flare value was less than 5.0 PC/ms), and the vitreous body showed mild inflammation (1+ cells) in the right eye. Fundus examination revealed a serous retinal detachment including a macular lesion. Her left eye still showed no inflammatory signs. These findings suggested a recurrence of posterior scleritis in the right eye, and methyl PSL, 1000 mg/day for 3 days, was prescribed. The serous retinal detachment promptly resolved, and the PSL was tapered off over a period of 6 months.

Twelve months after the initial onset, the patient complained of bilateral visual disturbance and ocular pain. Ocular examination revealed granulomatous iridocyclitis with mutton-fat keratic precipitates, Koeppe's nodules, and 3+ cells and 2+ flare (56.9 PC/ms OD and 23.6 PC/ms OS) in the anterior chamber. A mild inflammation of 1+ cells was found in the vitreous body of both eyes. Fundus examination revealed bilateral "sunset glow fundus" (Figure 2). At the same time, lymphocytosis was found in the cerebrospinal fluid and the patient was HLA-DR4-positive. She also developed poliosis.

She was then treated with systemic steroid and the anterior granulomatous iridocyclitis rapidly disappeared. From these findings, she was diagnosed as having VKH disease.

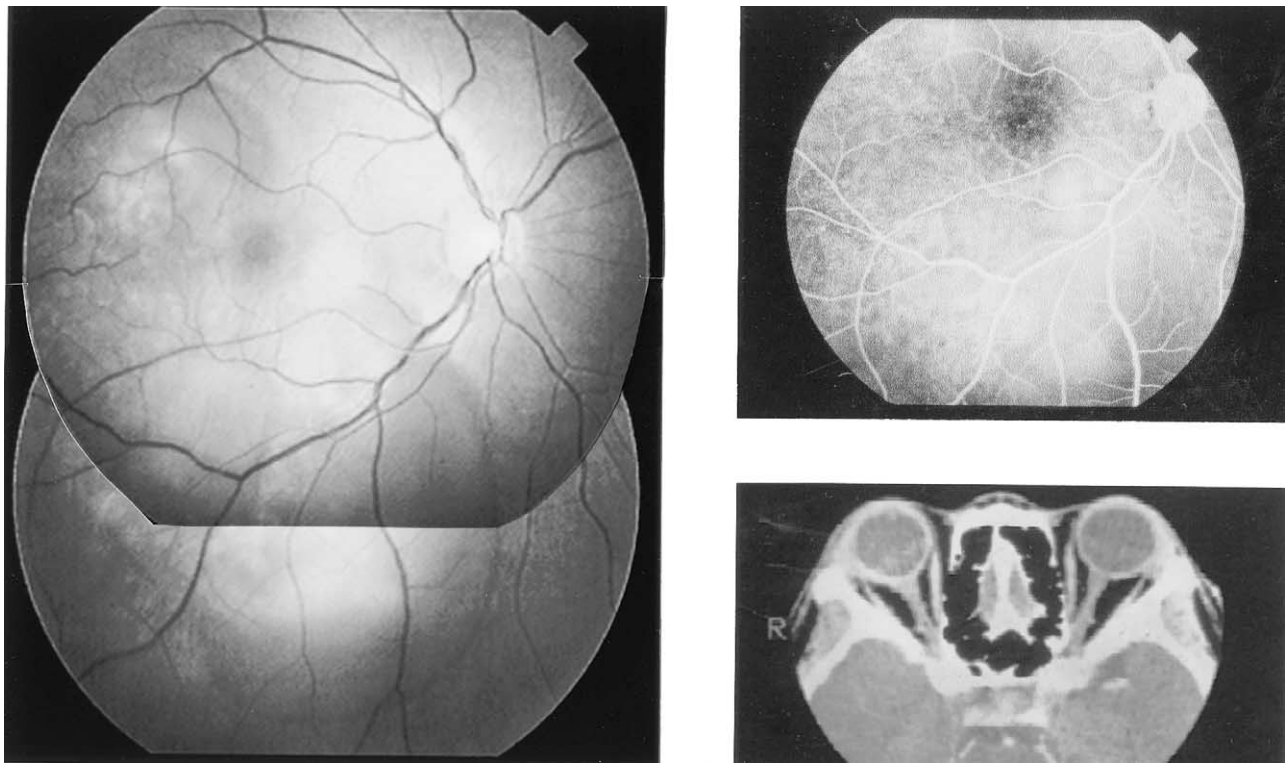


**Figure 1.** Left: Serous retinal detachment, including a macular lesion in the right eye, was seen at the first visit of a 38-year-old woman initially diagnosed as having unilateral posterior scleritis. Top right: Late-phase fluorescein angiogram at first visit of same patient, showing multiple pinpoint leakages and pooling of the dye in the right eye. Bottom right: Optical coherence tomography showing thickening of the sclera and the choroid in the right eye and no signs of posterior scleritis in the left eye at first visit.

Jpn J Ophthalmol
Vol 46: 590–593, 2002

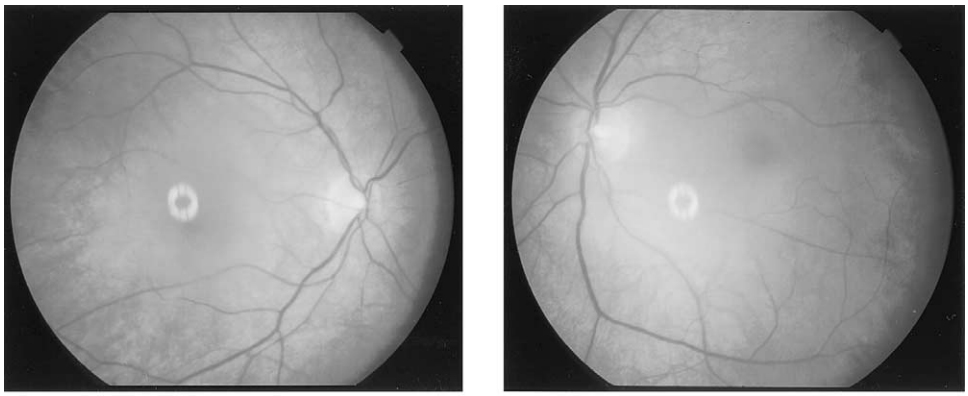


**Figure 2.** Fundus photographs 12 months after initial visit, with both eyes showing "sunset glow fundus." Final diagnosis was Vogt-Koyanagi-Harada disease.

## Discussion

VKH disease is one of the major forms of endogenous uveitis in Japan that usually involves both eyes, and in most patients, the involvement of the second eye occurs within 2 weeks.[1] There are only three reports of unilateral VKH disease.[2–4] Foster et al reported a 4-year-old boy with unilateral VKH disease, and even though there was no inflammation in the unaffected eye, ultrasonography revealed thickening of the choroids in both eyes.[2] Our case first presented with posterior scleritis and was diagnosed as having unilateral posterior scleritis because she had ocular pain, thickening of the sclera, and serous retinal detachment with multiple pinpoint leakages in fluorescein angiography in only 1 eye. It was not until 12 months later that she developed bilateral VKH with "sunset glow fundus."

VKH disease should be differentiated from posterior scleritis. In most cases of VKH disease, some signs are detectable in the fellow eye during fundus examinations, including fluorescein angiography and ultrasonography. The long interval for the full manifestation of VKH disease in our patient was therefore unexpected. Because 12 months later this patient showed lymphocytosis in the cerebrospinal fluid, positive HLA-DR4, poliosis, and bilateral "sunset glow fundus," we diagnosed this case as having VHK disease.

In retrospect, we suggest that this case was primarily VKH disease from the onset. The difficulty in the diagnosis may have been due to the subclinical level of inflammation in the uninvolved left eye which eventually led to the sunset-glow fundus. It can also be argued that the high dose of systemic steroid used initially may have delayed the involvement of the left eye. Whether systemic steroid delayed the onset in the second eye is, however, unknown, as there have been no reports of such an effect. The effect, if any, must be small because in almost all cases, VKH disease finally is bilateral, even though a high dose of systemic steroids is administered at the onset in the first eye.

Watanabe et al reported bilateral posterior scleritis concurrent with VKH disease in a patient.[5] However, this was not very likely in our case because VKH disease usually occurs as posterior serous retinal detachment or papillitis, and it does not exhibit granulomatous iridocyclitis or sunset-glow fundus as an initial sign, both of which were seen in our case.

We conclude that our case was VKH disease with early onset posterior scleritis in 1 eye and with a low-grade, subclinical ocular inflammation in the other eye. Thus, it appears that some cases of VKH disease may show severe serous retinal detachment unilaterally.

## References

1. Sugita S. Vogt-Koyanagi-Harada disease. Jpn J Ophthalmol 1978;22:9–35.
2. Forster DJ, Green RL, Rao NA. Unilateral manifestation of the Vogt-Koyanagi-Harada syndrome in a 7-year-old child. Am J Ophthalmol 1991;111:380–382.
3. Matsuura T, Tanaka T. A case of unilateral Harada's disease. Ganka Rinsho Iho (Jpn Rev Clin Ophthalmol) 1992;86:2445–2448.

4. Hosokawa M. A case of Harada's disease accompanied by unilateral acute glaucoma as an initial sign that remained unilateral throughout the clinical course. Nihon Ganka Kiyo (Folia Ophthalmol Jpn) 1997;48:1134–1137.

5. Watanabe K, Kata T, Hayasaka S. Concurrent bilateral posterior scleritis and Vogt-Koyanagi-Harada disease in a patient with positive rheumatoid factor. Ophthalmologica 1997;211: 316–319.