

# SÍNDROME DE VOGT KOYANAGI HARADA

## VOGT KOYANAGI HARADA SYNDROME

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### ABSTRACT

**Case reports:** Case 1. A female patient with visual loss and migraine who was found to have an exudative, bilateral retinal detachment. Steroid therapy was commenced initially and then cyclosporin-A added. Nine months later, her vision was 10/10 in the right eye and 9/10 in the left eye.

Case 2. A female patient with unclear vision who was found to have an exudative, bilateral retinal detachment. Systemic steroid therapy was given and ultimately her vision was 9/10 in the right eye and 8/10 in the left eye.

**Discussion:** Vogt Koyanagi Harada syndrome is a bilateral granulomatous panuveitis associated with neurologic and dermatologic disorders. When a patient has exudative multifocal and bilateral retinal detachment and systemic symptomatology, the possibility of this disease needs to be suspected. Good visual prognosis is possible if early treatment is given (*Arch Soc Esp Ophthalmol* 2008; 83: 385-390).

**Key words:** Bilateral panuveitis, Harada syndrome, serous detachment, uveomeningoen, Vogt Koyanagi Harada syndrome.

### RESUMEN

**Casos clínicos:** Caso 1. Mujer con pérdida visual y cefalea. Presentaba un desprendimiento de retina exudativo bilateral. Se inició tratamiento corticoideo y posteriormente se añadió ciclosporina-A. Tras nueve meses, su visión es de 10/10 en ojo derecho y 8/10 en izquierdo.

Caso 2. Mujer con visión borrosa. Mostraba vitritis con desprendimiento seroso bilateral. Tras iniciar tratamiento corticoideo permanece estable con visión de 9/10 en ojo derecho y 8/10 en izquierdo.

**Discusión:** El Síndrome de Vogt Koyanagi Harada es una panuveítis bilateral acompañada de alteraciones neurológicas y cutáneas.

La sospecha ante desprendimientos de retina exudativos, multifocales y bilaterales con sintomatología sistémica es fundamental para el diagnóstico. Su tratamiento precoz y mantenido es la base de una buena evolución.

**Palabras clave:** Panuveítis bilateral, síndrome de Harada, desprendimiento seroso, uveomeningoencefalitis, síndrome de Vogt-Koyanagi.

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## INTRODUCTION

Vogt-Koyanagi-Harada syndrome (VKH) is a multisystemic idiopathic disorder characteristically affecting individuals with higher levels of pigmentation, preferably young women (1). It is a diffuse bilateral granulomatous panuveitis that is accompanied by neurological, auditory and dermatological alterations. It has been reported that a positive blood test for the HLA-DRB1\*0405 haplotype increases the suspicion of having the disease. Its pathogenesis has been linked to immunological disorders whose target cells are melanocytes.

Two cases of VKH that were diagnosed and treated in our centre are reported below.

## CASE REPORT

### Case 1

A 28-year-old woman with a history of «multiple evanescent white dot syndrome» in both eyes (fig. 1) for the last seven years and who has responded well to oral steroids. She came into the emergency room of our hospital for a bilateral decrease in visual acuity and headache lasting for 15 days. The eye examination gave a visual acuity of 3/10 with inflammatory reaction in the anterior chamber and bilateral exudative retinal detachment (fig. 2). High-dose oral steroid treatment (80 mg of predni-

sone) was started, with a good initial response. One month after treatment a relapse of the foveal retinal detachment is seen in the left eye, so cyclosporine was added (starting dose: 4 mg/kg/day). After nine months of checkups, a visual acuity of 1 is seen in the right eye and 8/10 in the left with pigment mobilization in four quadrants and subretinal fibrosis (fig. 3).

### Case 2

A 45-year-old woman, with no relevant ophthalmologic history. She came into the emergency room for nausea, blurred vision and pain in both eyes. An eye examination showed a visual acuity of hand movement in both eyes with inflammation in the anterior chamber and vitritis with serous detachment affecting the posterior pole of both eyes (fig. 4).

She was referred to our department for evaluation and treatment. After starting high-dose steroid treatment (200 mg of intravenous prednisone that was gradually decreased every three days starting from day three to 160 mg, 120 mg, 80 mg and 60 mg, and then switched to oral prednisone), a fluorescein angiography and OCT are done. The patient remained stable during the first year of post-treatment check-ups, with a visual acuity of 9/10 in the right eye and 8/10 in the left eye (fig. 5). Funduscopy showed generalized atrophy of the pigmentary epithelium with pigment mobilization (fig. 6).

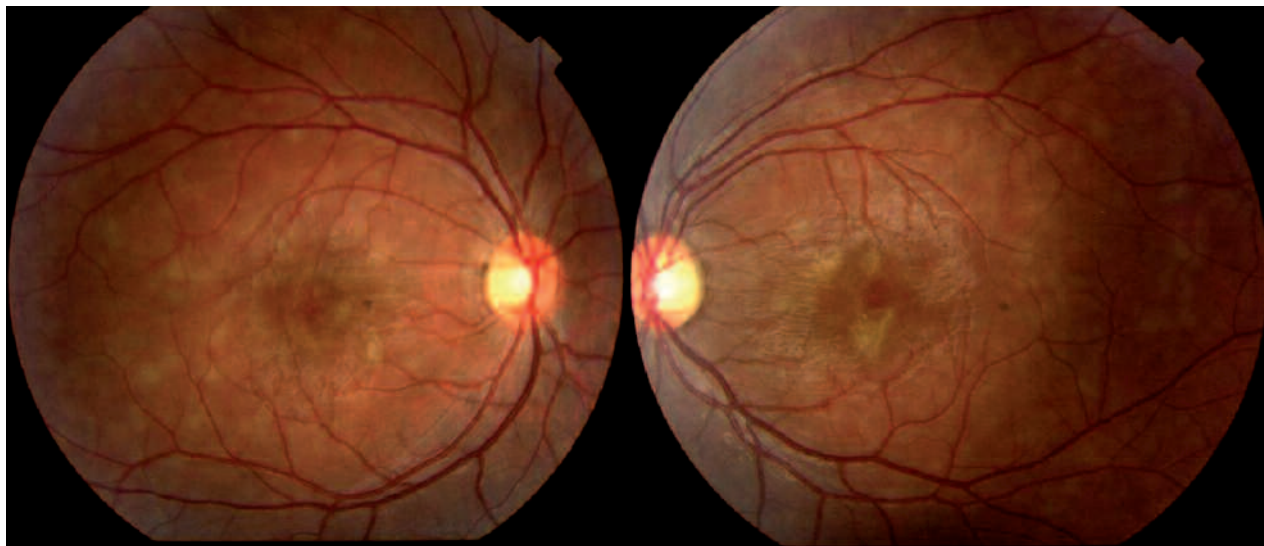


Fig. 1: Bilateral multiple evanescent white dot syndrome.

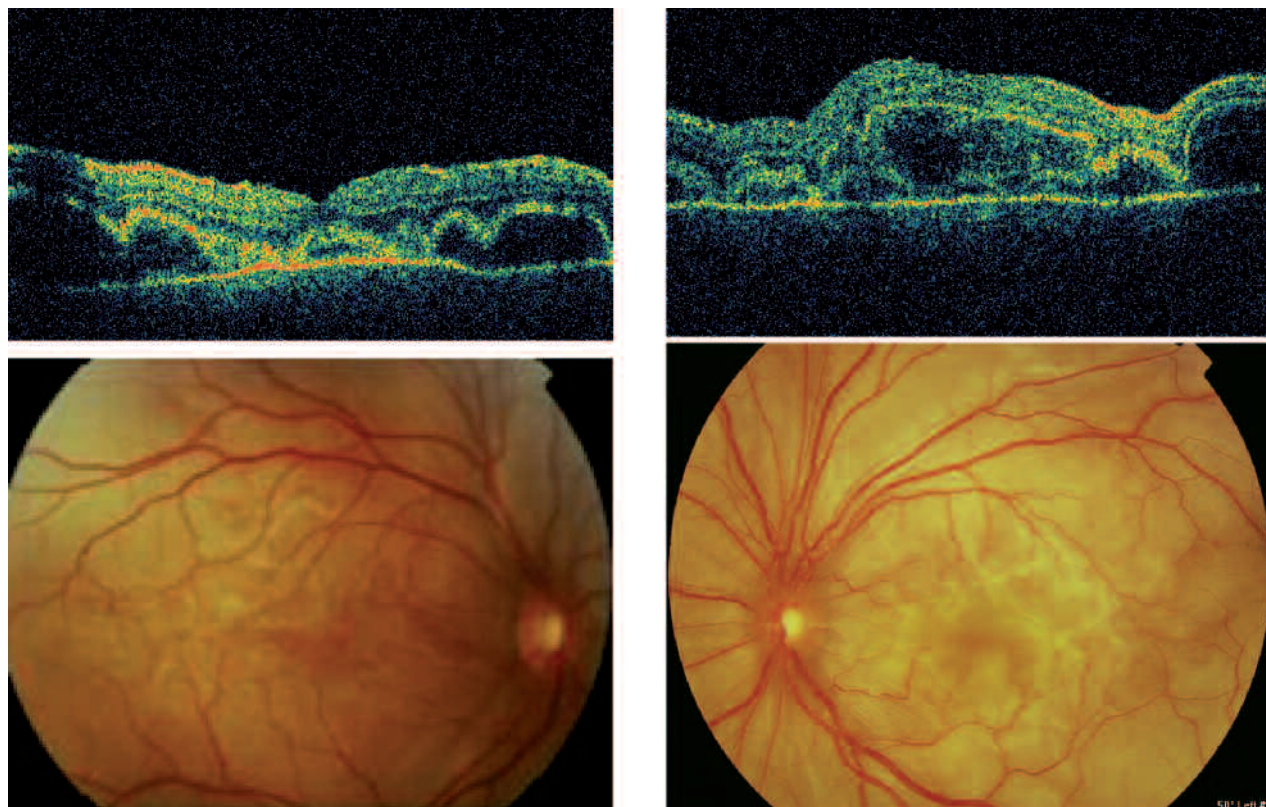


Fig. 2: Biomicroscopic image and optical coherence tomography of the bilateral exudative retinal detachment.

## DISCUSSION

Diagnosis of VKH is basically clinical, and a differential diagnosis from sympathetic ophthalmia, primary B-cell lymphoma, posterior scleritis and

uveal effusion syndrome is necessary. We can divide the progress of the disease into four clinical stages: prodromic, ophthalmologic, convalescent and chronic recurrent.

In the prodromic stage, patients suffer from headaches, nausea and vertigo, and pleocytosis is found in the cerebrospinal fluid. Then a few days later the ophthalmologic stage begins with blurred vision or central scotoma, bilateral in 80% of cases. The most characteristic finding is multifocal exudative retinal detachment. In the fluorescein angiograph (FAG), a color leak can be seen from the choroid to the subretinal space in multiple areas, with no color escape in the retinal vessels. Neovascularization in these patients is produced both by the sustained inflammation as well as the choroidal ischemia. The FAG is essential for differentiating between both processes and for treating them correctly.

In the convalescent stage, depigmentation of the tissue occurs, with recurring uveitis and ophthalmologic complications appearing in the chronic recurrent stage.

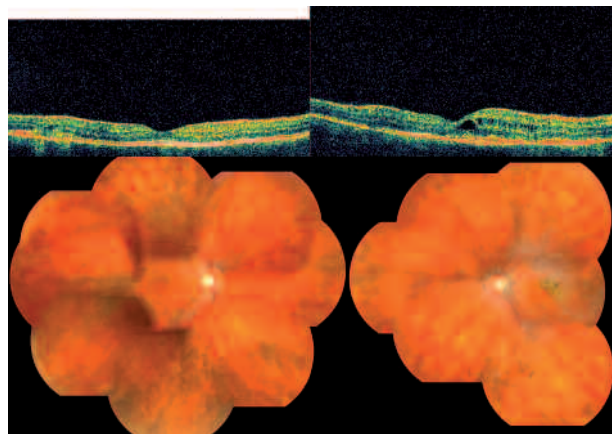


Fig. 3: Pigment mobilization in four quadrants and subretinal fibrosis in the left eye.

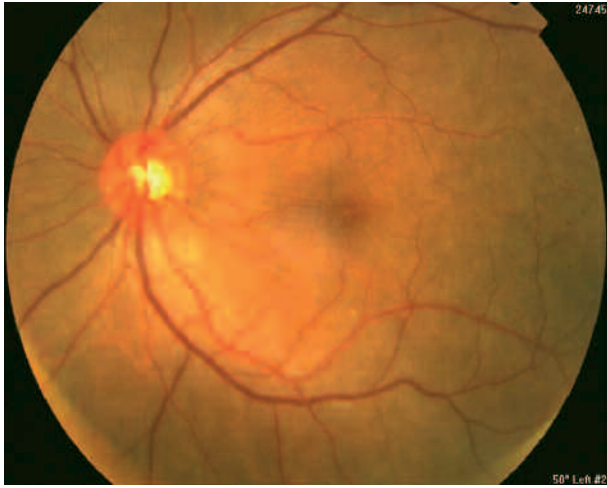


Fig. 4: Biomicroscopic image of bilateral exudative retinal detachments.

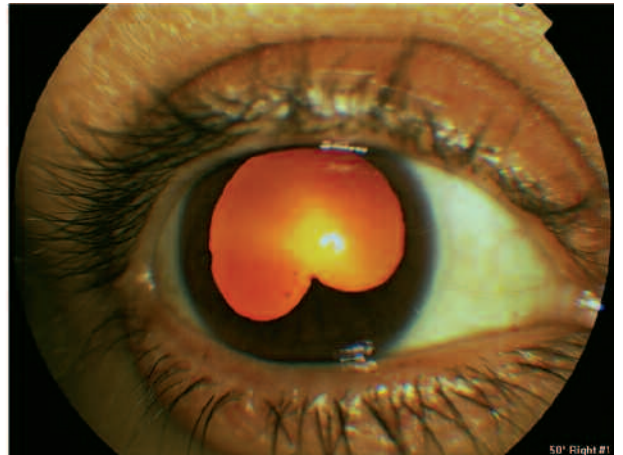
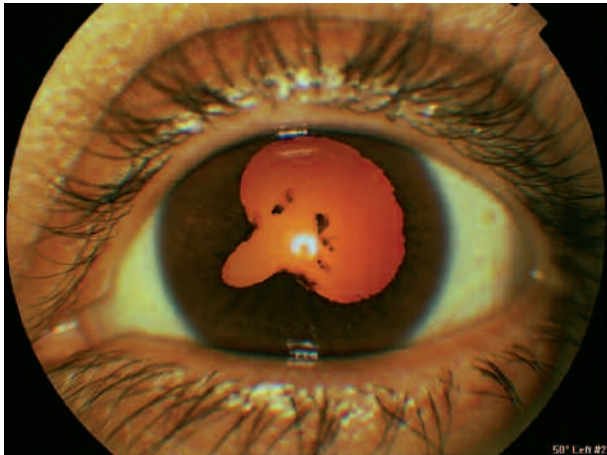


Fig. 5: Image of the patient's anterior segment after the first year of check-ups.

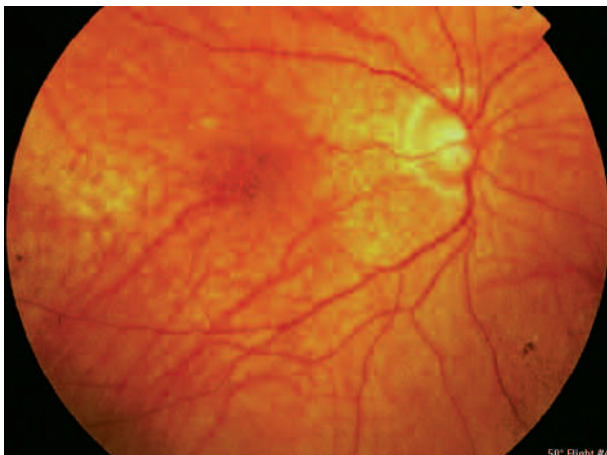


Fig. 6: Generalized pigment mobilization and pigment accumulation.

The diagnostic criteria for VKH are valid for a definitive diagnosis during the final stages of the disease, but not so during the initial stages, which is why it is necessary to review these (2).

KU=MEL=1 in the HLA=DRB1 has been described as a new auto-antigen for VKH, suggesting that it may be used for diagnosis and treatment of this disease (3).

The basic treatment comprises systemic and topical steroids. The initial dose, treatment duration and the pace for their withdrawal should be individualized for each patient (4,5).

The visual prognosis of the patients is general good if the diagnosis is made early and correct treatment is prescribed aggressively and maintained over the course of time.

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