Recurrent Anterior Uveitis in Patients With Vogt-Koyanagi-Harada Syndrome

Vogt-Koyanagi-Harada (VKH) syndrome is a bilateral granulomatous panuveitis with central nervous system, auditory, and integumentary manifestations. Typically, ocular involvement is a panuveitis associated with choroidal lesions, exudative retinal detachments, and optic nerve inflammation. Treatment requires high-dose systemic corticosteroids, usually for a prolonged period, often with additional immunosuppressive agents.

Resolution without visually significant sequelae may occur, but complications including cataract, retinal pigment epithelial disturbances at the macula, and choroidal neovascular membrane formation are not infrequent. Vogt-Koyanagi-Harada syndrome has been reported to recur, typically as a recurrent anterior uveitis (AU) with or without recurrent posterior involvement. It has been suggested that such syndrome recurrence is directly related to the failure to prescribe adequate corticosteroid therapy in the initial phase of the syndrome.

We report the manifestation, initial treatment, and clinical course of 3 patients with VKH syndrome who subsequently developed recurrent AU without further posterior segment involvement. Patients were identified from those attending the uveitis service of Moorfields Eye Hospital, London, England.

Report of Cases. Case 1. A 17-year-old Iranian woman attended the Casualty Department having a 2-day history of decreased vision in the left eye. Her visual acuity was 20/30 OD and counting fingers OS. Ophthalmic examination revealed bilateral AU and serous retinal detachments. Treatment was started with topical corticosteroids. The patient was referred to the uveitis service of Moorfields Eye Hospital.

Two days later, her visual acuity had decreased to 20/120 OD and 20/200 OS. Fundoscopy revealed large bilateral serous retinal detachments, vitritis, choroidal lesions, and intensely congested optic nerves. A diagnosis of VKH syndrome was made and treatment with 80 mg/d of prednisolone was started.

Disease resolution occurred rapidly and her corticosteroid dose was steadily tapered down to 20 mg/d over the next 3 months, and then gradually down to 15 mg/d over the following 2 months. At this point she suffered a relapse of posterior segment disease with recurrence of serous retinal detachments, and the dosage of prednisolone was increased to 80 mg/d. This was slowly tapered to 0 over 1 year, after which she stopped taking oral corticosteroids. At that time her visual acuity was 20/20 OD.

Since then, her posterior segment has remained quiet, but she has suffered recurrent AU, with 7 acute episodes occurring between October 1991 and January 2000. These have been successfully treated with topical corticosteroids and mydriatics, without the need for further systemic medication. Visual acuity remained stable at 20/20 OD until June 2000, when a choroidal neovascular membrane developed that has steadily reduced her visual acuity to 20/120 OD, despite therapy with pericocular corticosteroids and oral prednisolone, and repeated photodynamic therapy.

Case 2. A 54-year-old white woman was seen in the Casualty Department with a 2-day history of blurred vision in both eyes that was associated with headaches. Visual acuity was 20/200 OU. Ophthalmic examination revealed bilateral serous detachments and choroidal lesions. A diagnosis of VKH syndrome was made and combined treatment with 80 mg/d of oral prednisolone and 1 g twice daily of mycophenolate mofetil was started. Her condition improved rapidly over the following month while the corticosteroid dose was tapered to 40 mg/d; unaided visual acuity was 20/30 OU 8 weeks later. The corticosteroid dose was tapered to 0 gradually over the next 9 months.

The posterior segment has remained quiet since, but she has subsequently had 2 episodes of AU, which responded well to topical corticosteroid treatment alone. Visual acuity remains 20/30 OU.

Case 3. A 41-year-old Hispanic woman was seen in the Casualty Department with a 2-week history of a red aching eye and blurred vision with accompanying headache and aural pain. Visual acuity was 20/120 OD and 20/60 OS. Ophthalmic examination disclosed bilateral panuveitis. Combined treatment was started with topical corticosteroids and a cycloplegic agent.

On examination the next day, her visual acuity had decreased to 20/120 OU. She had developed bilateral serous detachments. She was diagnosed as having VKH syndrome and treatment with 80 mg/d of oral prednisolone was started. Disease resolution occurred quickly, and visual acuity improved to 20/30 OU over the next month. Oral corticosteroids had been tapered to 40 mg/d over this month, but posterior segment disease then recurred and the dosage of the oral corticosteroids needed to be increased to 80 mg/d once again. After this, the posterior segment remained quiet and her systemic corticosteroid therapy was tapered gradually down to 0 over 1 year.

Subsequently, she has had 4 episodes of AU, which have responded well to topical corticosteroid therapy alone; the posterior segment has remained inactive throughout. Visual acuity is 20/60 OU, due to increasing cataract formation.

Comment. In this case series, we report 3 cases of VKH syndrome in which patients had recurrent AU after syndrome quiescence had been achieved with the use of high-dose corticosteroids initiated rapidly after manifestation. In all cases, the recurrent AU responded well to combination therapy of topical corticosteroids and mydriatics, without the need for further systemic immunosuppression.

We thus confirm previous reports relating recurrent AU as a sequela of VKH syndrome. However, our case series differs from the cases previously reported because the recurrent AU in their patients proved resistant to topical cortico-
steroid therapy and led to further ocular complications and resultant poor visual acuity, whereas the cases reported herein responded well to topical corticosteroid therapy and had better visual outcomes.

One patient did develop a choroidal neovascular membrane formation 11 years after the initial diagnosis of VKH syndrome that has previously been reported to occur in 11% of affected eyes. It has been suggested that chronic AU may lead to inflammatory events in the retina and choroid that damage the retinal pigment epithelium and Bruch’s membrane, causing chorioretinal degeneration and choroidal neovascular membrane formation, based on the histopathologic condition. However, this was localized and unassociated with any other posterior segment changes.

The trigger for this recurrent AU remains unknown, but it cannot be purely a reflection of syndrome reactivation as the choroid and retina remained uninvolved in each case. Therefore, it seems probable that it may represent a secondary immune event. A similar phenomenon has been reported in patients with healed acute retinal necrosis, who may also demonstrate recurrent AU in the absence of posterior segment inflammation or recurrence of acute retinal necrosis. It has been suggested that the destruction of retinal tissue may release antigen that initiates an immune response to ocular tissue; similar to the sensitization to retinal S-antigen that follows panretinal photocoagulation. As this phenomenon does not occur in all patients with VKH syndrome, there may be the additional involvement of a genetic predisposition.

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Late Posterior Migration of Glass Intraocular Foreign Bodies

The treatment of intraocular foreign bodies (IOFBs) in the previtrectomy era was largely dependent on the source and composition of the material, associated ocular pathologic features, and intraocular location. With modern vitreoretinal surgical techniques, however, most glass IOFBs are removed despite their inert nature. Occasionally, the surgical complexity involved in their extraction must be weighed against the risks of leaving them in place. We describe 3 patients with retained glass IOFBs in which initially stable or encapsulated glass IOFBs subsequently migrated and induced further complications or visual symptoms. There are rare reports of glass IOFBs located initially in the posterior segment, migrating anteriorly, and producing additional anterior segment decompensation, such as illustrated in case 3 below. However, cases 1 and 2 are most unusual, as late migration caused additional retinal pathologic conditions.

We followed up 3 patients from 2 institutions with retained glass IOFBs for more than 6 months. The mechanism of injury, visual acuity, findings from the clinical examination, and fundus appearance were documented. Late migration necessitated surgical intervention in all 3 patients, although 1 patient refused surgery because of special circumstances.

Report of Cases and Results. Case 1. A 23-year-old man was struck with a glass beer bottle during an altercation and sustained a penetrating open globe injury with multiple glass IOFBs in the anterior and posterior segments of the left eye (Figure 1). The wound involved 7 mm of the cornea and an additional 22 mm on the scleral surface. All anterior segment glass IOFBs were removed (the largest measuring 7 × 6 × 3 mm) and the wound was repaired. One additional fragment with largest dimension measuring approximately 1 cm

Figure 1. Case 1. Preoperative axial computed tomographic scan obtained following initial injury, demonstrating 3 intraocular foreign bodies in the left eye.