

A Case of Recurrent Vogt-Koyanagi-Harada Disease Successfully Treated with Adalimumab in Young Female Adult Patient

Dear Editor,

Vogt-Koyanagi-Harada (VKH) disease is characterized by diffuse, bilateral, granulomatous panuveitis that can be associated with auditory, neurological, and cutaneous manifestations. Early and aggressive systemic corticosteroids are the primary initial therapy; however, despite proper treatment using corticosteroids and immunosuppressants, a number of patients experience recurrent attacks or steroid-associated complications [1]. Adalimumab, an anti-tumor necrosis factor α monoclonal antibody, is indicated for several inflammatory conditions that may be associated with intraocular inflammation. In this case report, we describe a 26-year-old female with recurrent VKH disease that could not be controlled with a steroid and immunosuppressant. Adalimumab treatment alone was successful in achieving remission without long-term recurrence.

A 26-year-old Korean female presented to our clinic with one week of bilateral blurred vision that had initiated eight months prior. The patient's uncorrected visual acuity was 20 / 22 (0.9) in the right eye and 20 / 30 (0.67) in the left eye. There were bilateral 1–2+ anterior chamber (AC) cells without keratic precipitates. Fundus exam showed a sunset-glow appearance with diffuse Dalen-Fuchs nodules, suggesting VKH disease (Fig. 1A). Optical coherence tomography showed mild choroidal thickening without subretinal fluid (Fig. 1B). Systemic workup was non-revealing. After maintenance treatment with oral prednisone (initiated with 30 mg/day and tapered weekly to 20 and 10 mg/day) and methotrexate 15 mg/wk for 1 month, inflammation resolved, and the patient recovered vision in both eyes. During the follow-up period, the patient suffered frequent

and intense relapses of anterior uveitis in both eyes when the dose of prednisone was reduced below 10 mg/day despite maintenance of methotrexate 15 mg/wk. A few years after initiation of systemic corticosteroid and multiple failed attempts of slow tapering, the patient complained about adverse effects such as weight gain, lethargy, depression, and gastrointestinal problems. Despite addition of mycophenolate mofetil 1,000 mg twice a day, the patient had persistent anterior chamber inflammation with posterior synechiae (Fig. 1C) and progressing chorioretinal atrophy threatening the posterior pole (Fig. 1D). In the following months, the patient developed tinnitus and vitiligo, indicating the diagnosis of complete VKH.

At that time, we started treatment with adalimumab (Humira; Abbvie, Chicago, IL, USA) 40 mg subcutaneously every 2 weeks, with resolution of anterior chamber inflammation 1 week after the first administration. During adalimumab treatment, itching eczematous lesions were

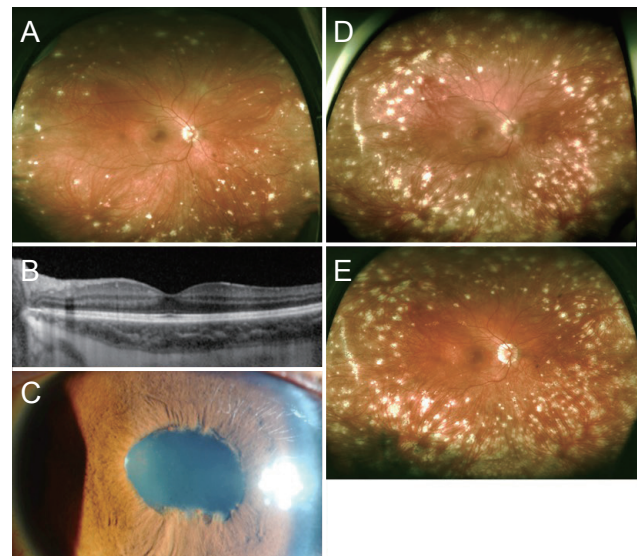


Fig. 1. (A) 11 months before the treatment of adalimumab. Color fundus photography showing sunset glow fundus changes with diffuse Dalen-Fuchs nodules, consistent with Vogt-Koyanagi-Harada disease. (B) Optical coherence tomography showing mild choroidal thickening without subretinal fluid. (C) Ant seg photo showing posterior synechiae on lens before adalimumab treatment. (D) One month before the treatment of adalimumab. Color fundus photography showing aggravated chorioretinal atrophy threatening the posterior pole. (E) 15 months after the treatment of adalimumab. Color fundus photography showing stopped progression of chorioretinal atrophy.

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noted over the whole body and were consistent with seborrheic dermatitis; these lesions resolved after 2 weeks of topical steroid cream. Sequential cessations of the oral steroid, mycophenolate mofetil, and methotrexate were possible. The patient has been on adalimumab monotherapy 40 mg biweekly for 15 months and 40 mg monthly for 6 months with best-corrected visual acuity 20 / 20 (1.0) in both eyes without signs of active uveitis or systemic symptoms. The fundus abnormality stopped progressing after the treatment with adalimumab (Fig. 1E). No other adverse events have been noted during adalimumab treatment.

To the best of our knowledge, this is the first case of recurrent VKH syndrome in Korea to be successfully treated with adalimumab. Adalimumab has been reported to be successful in controlling ocular inflammation with non-infectious uveitis [2]. Couto et al. [3] showed adalimumab to be safe and effective in controlling intraocular inflammation in 14 patients with recurrent VKH disease. Jeroudi et al. [4] reported successful use of adalimumab in a case of refractory pediatric VKH, with rapid resolution of inflammation and favorable visual outcome.

High-dose systemic corticosteroids have been the mainstay of initial treatment for VKH disease. But prolonged use of systemic corticosteroids can lead to complications such as Cushing syndrome, hyperglycemia, adrenal suppression, and psychological problems. Although our patient showed improvement with a high-dose systemic corticosteroid, she had significant difficulty in tapering the steroid and suffered corticosteroid-associated complications. Immunosuppressive agents can be added as adjunctive therapy in refractory VKH or in patients who cannot tolerate long-term corticosteroids. Despite these treatment options, refractory cases have been reported; in the present case, adalimumab treatment was an adequate alternative. In adalimumab treatment in general, complications such as malignancy, inflammatory neurologic disease, opportunistic infections, reactivation of latent tuberculosis, and congestive heart failure may occur [5]. However, in this case, no significant adverse events were observed except mild skin eruptions that were easily controlled with dermatologic treatment.

In conclusion, this case suggests that treatment with adalimumab could be an effective and safe therapeutic op-

tion for controlling inflammation and vision preservation in patients with recurrent VKH disease.

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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