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A Case of Probable Vogt-Koyanagi-Harada Disease

Olası Vogt-Koyanagi-Harada Hastalığı Olgusu

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Summary

The aim of this article is to present a rare form of Vogt-Koyanagi-Harada (VKH) disease. The complete and incomplete forms of VKH disease are more frequent than the probable form. A 58-year-old woman presented with bilateral painless blurred vision and headache. The patient's visual acuity was 1/10 for both eyes. The patient had bilateral iridocyclitis, optic disc swelling, serous retinal detachment and choroidal thickening. The examinations by specialists in otorhinolaryngology, neurology, dermatology, rheumatology and chest diseases revealed no abnormal findings. Cerebrospinal fluid examination was also normal. After the diagnosis of probable VKH disease, the patient was treated with systemic pulse corticosteroid and cyclosporine-A. After this treatment, the abnormal findings were resolved and her visual acuity was improved to 10/10 for both eyes. VKH patients should be followed closely for risk of disease recurrence and side effects of medications. Immunomodulatory and immunosuppressive drug therapies are necessary to prevent corticosteroid-associated complications as well as recurrence of VKH disease. *(Turk J Ophthalmol 2012; 42: 235-7)*

Key Words: Vogt-Koyanagi-Harada (VKH) Disease, uveitis, serous retinal detachment

Özet

Bu makalenin amacı Vogt-Koyanagi-Harada (VKH) hastalığının nadir bir formunu sunmaktır. VKH hastalığının tam ve tam olmayan formları olası formuna göre daha sıktır. Elli sekiz yaşında kadın hasta baş ağrısı ve iki taraflı ağrısız görme bulanıklığı ile başvurdu. Hastanın görme düzeyi her iki göz için 1/10 idi. Hastada; iki taraflı iridosiklit, optik disk ödemi, seröz retina dekolmanı ve koroidal kalınlaşma vardı. Kulak burun boğaz, nöroloji, dermatoloji, romatoloji ve göğüs hastalıkları muayeneleri normaldi. Beyin omurilik sıvısı incelemesi de normaldi. Olası VKH hastalığı tanısı konulduktan sonra, hasta sistemik puşe kortikosteroid ve siklosporin-A ile tedavi edildi. Bu tedaviden sonra anormal bulgular geriledi ve hastanın görme düzeyi her iki göz için 10/10' a yükseldi. VKH hastaları; hastalık tekrarlama riski ve tedavilerin yan etkileri açısından yakın takip edilmelidirler. İmmünmodülatör ve immünsüpresif ilaç tedavileri, hem kortikosteroid ilişkili komplikasyonların hem de VKH hastalığının tekrarlamasının önlenmesi için, gereklidirler. *(Turk J Ophthalmol 2012; 42: 235-7)*

Anahtar Kelimeler: Vogt-Koyanagi-Harada hastalığı, üveit, seröz retina dekolmanı

Introduction

Vogt-Koyanagi-Harada (VKH) disease, also known as uveomeningitic syndrome, is an idiopathic multisystem inflammatory disease affecting the eyes, auditory system, meninges and the skin.[1-7] It is a cytotoxic T cell-mediated autoimmune situation against melanocytes. VKH disease consists of a nonnecrotizing diffuse granulomatous inflammation of the uveal tractus.[8] VKH disease is common in pigmented people.[1-6] The ocular components of VKH disease are characterized by multifocal serous retinal detachment, choroidal swelling, and optic disc hyperemia in the acute stage.[8] According to the findings and involvement of organ systems, VKH disease is categorized as complete, incomplete and probable.[9,10] The diagnosis of VKH disease is based on history and clinical findings. The probable form of VKH disease is characterized by

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bilateral ocular involvement as defined above, no history of ocular trauma and ocular surgery, and no clinical or laboratory evidence suggestive of other ocular disease.[8,9]

High-dose pulse corticosteroid can improve the symptoms and signs of the disease in the acute stage. Recurrent episodes may be resistant to corticosteroids, so immunosuppressive agents such as cyclosporine, cyclophosphamide or azathioprine can be use in this stage of VKH disease.[1-7]

Informed consent was obtained from our patient and the study was approved by the ethics committee of our hospital.

Case

A 58-year-old woman presented with bilateral painless blurred vision and headache. Her initial visual acuity was 1/10 for both eyes. There was no history of ocular trauma and ocular surgery in our patient. The patient had bilateral iridocyclitis, optic disc swelling, serous retinal detachment and choroidal thickening. Laboratory and full clinical investigations were performed including fundus photography (FP), fundus fluorescein angiography (FFA), B-scan ocular ultrasonography (USG), orbital and cranial magnetic resonance imaging, cerebrospinal fluid examination, full blood examination (complete blood count, c-reactive protein, erythrocyte sedimentation rate, rheumatoid factor, etc). FFA showed prominent leakage of dye from the optic disc, mild dye pooling in the subretinal space, pin-point hyperfluorescence and macular edema (Figure 1). USG showed diffuse choroidal thickening with minor reflectivity, serous retinal detachment and vitreous opacities (Figure 1).

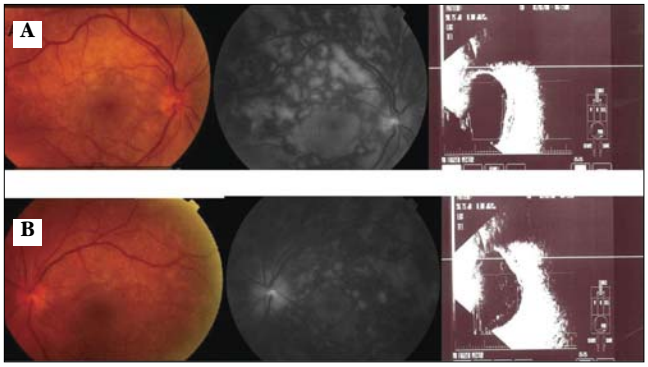

Figure 1. A (right eye), B (left eye) pre treatment. FP, FFA, ocular USG. FP showed diffuse choroiditis with optic disc edema and serous retinal detachment. FFA showed prominent leakage of dye from the optic disc, mild dye pooling in the subretinal space, pin-point hyperfluorescence and macular edema. USG showed diffuse choroidal thickening with minor reflectivity, serous retinal detachment and vitreous opacities

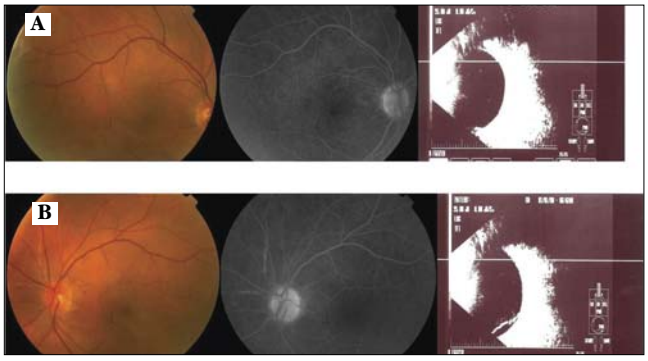

Figure 2. A(right eye), B(left eye) post treatment. FP, FFA, ocular USG. FP showed findings resolved with therapy. FFA showed less leakage of dye from the optic disc. USG showed localized posterior vitreous detachment

The examinations by specialists in otorhinolaryngology, neurology, dermatology, rheumatology and chest diseases revealed no abnormal findings. Blood tests and cerebrospinal fluid examination were also normal. Chest radiography and serum angiotensin-converting enzyme level were normal. We suspected the diagnosis of a probable VKH disease, thus the patient was treated with systemic pulse corticosteroid and cyclosporine-A. After 6-month systemic corticosteroid (3 days pulse corticosteroid) and 6-month cyclosporine-A treatments, the abnormal findings were resolved and her visual acuity was improved to 10/10 for both eyes (Figure 2). There were no treatment-related complications and recurrence during the 15-month (3 months without therapy) follow-up period. There were no ocular complications such as limbus depigmentation (Suguira sign) and no extraocular complications such as vitiligo, alopecia, poliosis, auditory dysfunction, and central nervous system damage findings during the 15 months follow-up period.

Discussion

VKH disease is an inflammatory multiorgan disorder. The pathogenesis and causes of the disease are still uncertain - it occurs more frequently in pigmented races such as Native Americans, Hispanics but is uncommon in Africans and Caucasians.[7] This information suggests that skin pigmentation alone is not an etiologic factor of the disease. Various investigators have suggested an underlying autoimmune and infectious process. Genetic link to HLA-DRB1, HLA-DR4 and HLA-Dw53 has been reported in recent studies, therefore, genetic analysis may be useful for monitoring the VKH disease and screening other family members.[11,12]

T cell-mediated autoimmune process directly attacks chorodial melanocytes in genetically predisposed people and this response may facilitated some infectious agents such as cytomegalovirus, herpes simplex virus by a mechanism of molecular mimicry.[1-7, 13,14] Our patient did not have any family members who had same symptoms or any infectious diseases for a six-month period. The differential diagnosis should include sarcoidosis, sympathetic ophthalmia, idiopathic uveal effusion syndrome, B-cell lymphoma, posterior scleritis and acute posterior multifocal placoid pigment epitheliopathy.[1-7] Sarcoidosis was excluded because of normal chest radiography, serum angiotensin-converting enzyme level and presence of exudative retinal detachment. Sympathetic ophthalmia was excluded because of no history of ocular trauma and ocular surgery. B-cell lymphoma was excluded because of rapid progression of bilateral involvement. Posterior scleritis was excluded because of painless blurred vision, USG features of case and normal rheumatological examination. Our patient's systemical examinations (otorhinolaryngology, neurology,

dermatology, rheumatology and chest diseases) and cerebrospinal fluid analysis were normal, therefore, her diagnosis was probable VKH.

The first treatment choice of VKH disease is systemic corticosteroids. High-dose pulse corticosteroid treatment has quick effects in the acute stage of the disease.[1-7] İmmunomodulatory and immunosuppressive drug therapies such as cyclosporine, cyclophosphamide or azathioprine are necessary to prevent recurrences and the corticosteroid-associated complications.[1-7] Pulse corticosteroid treatment was effective to achieve quick response in our patient. Oral corticosteroid therapy was started and tapered in 6 months. Cyclosporine-A treatment was started to prevent disease recurrence and the corticosteroid-associated complications.

Cataract, glaucoma, choroidal neovascularization and subretinal fibrosis are the most common complication of this chronic disease. There were no complications in our case. Her final visual acuity was 10/10 for both eyes.[15] Early diagnosis and appropriate treatment of VKH disease may protect vision of these patients.

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