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Vogt-Koyanagi-Harada Syndrome

Sharmeen Akram and Khabir Ahmad

ABSTRACT

Vogt-Koyanagi-Harada (VKH) syndrome is a rare multisystem disease of melanocyte containing organs. It is characterized by diffuse granulomatous inflammation involving various organs including eye. VKH syndrome is usually sporadic, but some familial cases have also been reported indicating a hereditary basis. VKH is not associated with mortality but it may result in long-term complications such as decreased vision associated with cataract, glaucoma and choroidal neovascularization. For successful outcomes, early aggressive treatment using systemic steroids with gradual tapering is essential. This report describes a case of VKH syndrome in a 26-year-old male of Pakistan origin who was successfully treated with systemic steroids. The case is briefly contextualised within wider literature.

Key Words: Steroids. Visual outcome. Vogt-Koyanagi-Harada syndrome. Uveoencephalitis. Melanocytes.

INTRODUCTION

Vogt-Koyanagi-Harada (VKH) syndrome or uveoencephalitis is a rare systemic disease of melanocyte-containing organs.¹ In the eye, VKH presents as granulomatous panuveitis with exudative retinal detachment. It is also often associated with skin, auditory and neurologic manifestations. It is found in Asian, Middle Eastern, Hispanic, and Native American populations and is more common in females.² The diagnosis of VKH syndrome is based on the revised criterion set by an International Committee on Nomenclature which has defined 3 categories of disease: complete, incomplete and probable VKH.³ Treatment involves topical and systemic steroids and immuno-suppressants.

We report a case of VKH syndrome with a brief literature review.

CASE REPORT

A 26-year-old male, diagnosed with VKH in an eye clinic in Sydney, Australia, came to our clinic for follow-up. He presented there with bilateral red eye and decreased vision—right more than the left. There was no history of any neurological complaints. On examination, he was found to have bilateral anterior uveitis with exudative retinal detachment in the right eye. There were no auditory, neurological or dermatological symptoms or signs present at the time of initial presentation. Figure 1 shows the optical coherence tomography (OCT) report of his right eye at presentation. Fluorescein fundal angiography (FFA) was done as well at that time but the

report is not available. A series of blood tests (e.g. complete blood count, angiotensin converting enzyme, antinuclear antibody, enzyme-linked immunosorbent assay) were done to rule out other causes of uveitis. He was put on oral (120 mg/day) and topical steroids.

Three months after, he was diagnosed with VKH, he came to our clinic while still on a tapering dose of steroid (prednisolone) - 15 mg/day. While his vision had improved on medication, he still had complaints of decreased vision, metamorphasia and glare during night driving, and had recently developed decreased hearing in both ears.

On examination, the best-corrected visual acuity in his right eye was 20/40. The left eye had a 20/20 vision. He had an intraocular pressure of 10 mmHg in both eyes. Anterior segment of both eyes was normal. Examination of the posterior segment showed a dull macular reflex in the right eye with no retinal detachment. The left eye was normal. OCT and FFA three months after the initiation of treatment (Figure 1 b and c) showed resolution of exudative retinal detachment. Amsler Grid showed wavy lines. He was referred to a skin specialist but had no skin signs. His audiogram showed sensorineural hearing loss in the high frequency range in both ears. According to the criterion set by the International Committee on Nomenclature,⁴ the case met the first four criteria and was diagnosed as incomplete VKH (Table I). The dose of steroid (prednisolone) was increased to 60 mg/day and tapered over the next 3 months. His best corrected vision in the right improved to 20/20 from 20/40. His near vision is N/6 in both the eyes. Hearing improved and metamorphasia also decreased slightly. The only remaining complaint has been the glare during night driving 6-month post-treatment while on maintenance dose of steroids 5 mg/day. A year later, glare also resolved. Only slight metamorphasia is present but does not interfere with his daily life. He is not taking any medicine for the last four months.

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Table I: VKH syndrome: diagnostic criteria and treatment.

A. Diagnostic criteria
1. Absence of history of penetrating ocular trauma or surgery preceding the initial onset of uveitis.
2. Absence of clinical or laboratory history of other type of ocular disease
3. Bilateral ocular involvement.
4. Presence of neurological/auditory findings.
5. Presence of cutaneous findings not preceding onset of ocular disease or neurological manifestations.
Presence of criteria 1-5 (Complete VKH), Presence of criteria 1-3 plus 4 or 5 (Incomplete VKH), Presence of criteria 1-3 (Probable VKH).
B. Prognostic factors
Age, timing of intervention, vision within 1 month of treatment.
C. Treatment options
Steroids, immunosuppressant, immunoglobulins.

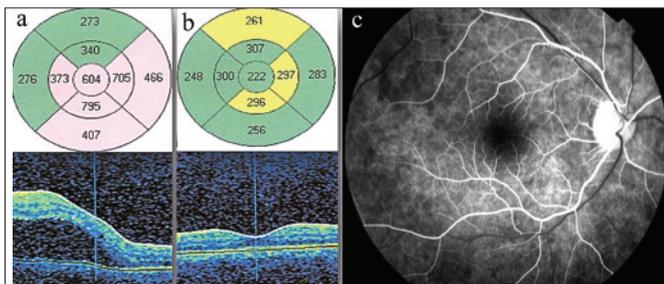


Figure 1 (a,b,c): (a) OCT right eye pre-treatment, (b) OCT right eye 3 months after the initiation of treatment, (c) FFA right eye 3 months after the initiation of treatment.

DISCUSSION

VKH has been reported in diverse populations (e.g. Asian, Arab, Hispanic, and Native American) with female preponderance. In this case, the patient, though diagnosed in Australia, was an Asian of Pakistani origin, the typical age range for this syndrome is 20 - 50 years. However, rarely the disease may present in children as well. VKH is more common among females, with a female to male ratio of 2:1.²

The disease involves various melanocyte containing organs including the eyes, ears and central nervous systems. Though the exact etiology of the disease is unknown, as was in this case, an autoimmune cause has been reported in literature. There is mainly a T-cell mediated immune response against the autoantigens such as uveal autoantigen, TRP1, TRP2, MART 1, PMel-17/gp100, KU-MEL-1, PAX3 on melanocytes. The changes noticed in the immune system of VKH patient include reduction in the number and activity of regulatory T-cells and decreased apoptosis of lymphocytes.⁵ There is an increased expression of major histocompatibility complex type II (MHC II). This autoimmune pathogenesis is supported by the fact that VKH is associated with HLA-DR4 and HLA-Dw53, of the allele subtypes, the most important are HLA-DRB1*0410 and HLA-DRB1*0405.⁶ In this case, the association for HLA was not worked up. Other possible etiologies identified as triggering factors include viral infection, trauma,

tumors and certain medications. No such correlation was identified in this case.

As shown in Table I, the diagnosis of this condition is based on the revised criterion set by an International Committee on Nomenclature which has defined 3 categories of disease: complete VKH (all 5 criteria), incomplete VKH (criteria 1,2,3, 4 or 5), and probable VKH (criteria 1,2,3).⁴ The clinical course of VKH is divided into four clinical phases, prodromal phase present with headache, mild fever, photophobia, and symptoms and signs of meningoencephalitis. Lumbar puncture at this stage may show lymphocytic pleocytosis, proteins and increased cerebrospinal pressure. This phase lasts for a few days to weeks with variable severity and is noticed in only 50% of the patients. This patient did not give the history of this phase or did not notice this phase.

This is usually followed by the uveitic phase which lasts a number of weeks. It is this phase in which most patients come for medical consultation.² This patient also presented to the clinic in the uveitic phase, with bilateral red eye and decreased vision-right more than the left. Bilateral blurred vision is the most common presenting complaint (in 70% of cases) in this phase.² Ocular signs during this phase have been described as bilateral posterior uveitis with retinal edema, papillitis, serous retinal detachments, iridocyclitis, mutton-fat keratic precipitate and iris nodules and raised IOP. This case presented with bilateral posterior uveitis with serous retinal detachment in the right eye. Otolaryngeal symptoms also usually present in this stage. This patient noticed decreased hearing and on testing was found to have neurosensory hearing loss, which responded to the treatment. Literature describes immunomediated sensorineural hearing loss which is bilateral, though may be asymmetrical and is reversible with treatment.⁷ Other otolaryngeal symptoms such as tinnitus, dizziness and vertigo were not reported by this patient. The convalescent phase reported in 80% of patients is associated with tissue depigmentation such as vitiligo and poliosis, this follows the uveitic phase. Then comes the recurrent phase which shows signs of recurrent uveitis and ocular complications such as cataract, glaucoma, retinal detachment, subretinal membranes and fibrosis.⁸ This patient did not have a recurrence till the last follow-up.

The diagnosis of this condition is mainly clinical and though no specific lab tests are present, tests done in routine for uveitis are ordered to help in differential diagnosis which includes conditions like sympathetic ophthalmia, sarcoidosis, primary intraocular B-cell lymphoma, posterior scleritis, and uveal effusion syndrome. OCT findings pre-treatment and 3 months after treatment and FFA findings 3 months after

treatment are shown in this case report. Indocyanine green angiography (ICGA) has been shown to be the most useful imaging modality to monitor response to the treatment but it is currently not available in our setting.

Treatment should be early and aggressive with high-dose oral corticosteroids; there is no advantage of intravenous therapy over oral one.⁹ Early aggressive treatment, young age, and good vision within one month of treatment are associated with a better visual prognosis (Table I). Treatment is associated with complete reversal of auditory symptoms which was also the case in this patient. It is suggested to continue the treatment for 3 months with tapering over 6 months to a year for good visual prognosis and to prevent recurrences. This patient was started on high dose steroids and then gradually the dose tapered. Patients not responding to steroids or not tolerating its side-effects can be treated with immunomodulatory agents,¹⁰ such as cyclosporine, tacrolimus, azathioprine, mycophenolate mofetil, cyclophosphamide, or chlorambucil. Recent evidence indicates that virtually all the patients with VKH can be successfully treated with immunomodulatory agent and now there is a trend to early immunosuppressive therapy in VKH.

A limitation of this case report is that we could not get the complete information regarding initial assessment such as FFA at the time of presentation, dose and frequency of topical steroids prescribed and vision within one month of treatment (a prognostic factor) as the patient was diagnosed in another country.

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