A CASE REPORT : PROBABLE VOGT–KOYANAGI–HARADA SYNDROME: A RARE PRESENTATION IN A PAKISTANI MALE

Asna Tahir, Imran Ahmad, Hina Ikhtiar, Muhammad Usman Khan, Ansa Anam

[1]Department of Ophthalmology, MTI/Khyber Teaching Hospital, Peshawar - Pakistan

ABSTRACT

A 48-year-old male presented to us as an out-patient with a painless blurring of vision for five days. On examination, the visual acuity (VA) of his right eye was 6/36, while that of his left eye was 6/24. The rest of the systemic examination showed no notable findings except for fundus examination, which revealed multiple serous retinal detachments (SRD) confirmed on Optical Coherence Tomography (OCT). Treatment involved intravenous administration of 1 gram methylprednisolone followed by oral steroids. His condition significantly improved after three doses with a corresponding enhancement in visual acuity with a resolution of SRDs. This case report illustrates the efficacy of corticosteroids in the initial and successful treatment of Vogt-Koyanagi-Harada syndrome.

This article may be cited as: Tahir A, Ahmad I, Ikhtiar H, Khan MU, Anam A. A Case Report : Probable Vogt–Koyanagi–Harada Syndrome: A Rare Presentation In A Pakistani Male. J Med Sci July-September;32(3):281-283

INTRODUCTION

VKH is an autoimmune disease involving multiple systems. The occurrence of VKH disease fluctuates depending on geographical location and ethnic background, but generally, it is more prevalent among Asians. These epidemiological findings, along with the discovery of linked major histocompatibility complex (MHC) class II antigens suggest a genetic predisposition underpinning the disorder's pathogenesis. [1] It manifests as a non-necrotizing diffuse granulomatous uveitis, that commonly affects various structures like eyes, inner ear, central nervous system, and skin. [2] It is believed that VKH is due to a T cell (CD4+ Th1 lymphocyte) driven aggressive response to the melanin-producing melanocytes. Melanin has a significant function in maintaining normal vision Within the retina.

Ocular complications from this condition are numerous, and early diagnosis and management can have a great impact on decreasing morbidity. [3] In the early stages of the illness, patients may solely exhibit ocular symptoms, commonly with inflammation primarily affecting the choroid, possibly extending to the iris and ciliary body. [4] Early ocular signs of VKH include multifocal serous retinal detachments (SRDs) and choroidal thickening.

Correspondence
Dr. Imran Ahmad
Assistant Professor
Department of Ophthalmology, MTI/Khyber Teaching Hospital, Peshawar - Pakistan
Cell: +92-300-9333000
Email: doctorimranahmad@gmail.com
Date Received: 22/05/202?
Date Revised: 05/07/202?
Date Accepted: 04/03/2024

The revised diagnostic criteria of VKH syndrome [5] categorizes this syndrome as complete, incomplete, and probable VKH disease. These categories are based on the following;

1. Absence of past penetrating eye trauma,

2. No concurrent ocular conditions,

3. Bilateral uveitis,

4. Presence of neurological and auditory symptoms, and

5. Dermal manifestations such as vitiligo etc.

In complete VKH, criteria 1 to 5 must be fulfilled; for incomplete VKH, criteria 1-3 plus either 4 or 5 are required, and for probable VKH, only criteria 1-3, indicating isolated ocular disease, should be present. Evaluation of VKH includes taking a detailed ophthalmic and systemic history, followed by examination of both eyes along with relevant investigations to rule out any other causes.

CASE REPORT

A 48-year-old Pakistani man visited the outpatient department of Khyber Teaching Hospital, reporting painless blurred vision and floaters in both eyes, which began five days prior and had worsened progressively. He experienced no associated pain, watering, or sensitivity to light. His systemic examination revealed no notable findings, and he had normal blood sugar and blood pressure levels. His past medical and surgical histories were also unremarkable. The patient's previous ocular history indicated a period of diminished vision in both eyes. OCT of the macula was performed, which showed multiple exudative retinal detachments with intervening septae. (Figure:1)

On ocular examination, the visual acuity of the patient's right eye was measured at 6/36, while the left eye's visual acuity was 6/24. Pupils were round, regular, and responsive to light, with absent relative afferent pupillary defect (RAPD) observed. Extraocular movements were within normal limits, and no abnormalities were noted in the adnexa.

Examination of the conjunctiva, cornea, and lens revealed bilateral clarity. In the anterior chamber, +1 cells were seen. A Fundus examination revealed multiple serous retinal detachments (SRDs) bilaterally. All baseline systemic investigations including complete blood count (CBC), C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), and Mantoux test for tuberculosis (T.B) were also well within normal ranges.

Since we planned to start the patient on intravenous steroids, we checked contraindications for IV steroids by monitoring random blood sugar, fasting blood sugar, and glycated hemoglobin (HbA1c). All of which were well within the normal ranges. After doing all the necessary investigations, we started our patient on intravenous (I/V) steroids (methylprednisolone 1 gram) therapy for his uveitis.

On presentation to OPD, his VA in the right eye was 6/36 and 6/24 in the left eye. After the administration of methylprednisolone, his visual acuity of the right eye was found to be 6/36, whereas the VA of the left eye improved to 6/18. On encountering these results, we administered methylprednisolone again with which his VA further improved and was reported to be 6/18 and 6/12 in the right and left eye, respectively. Carrying on with the same pattern, after three doses of I/V steroids his vision improved to 6/9-1 in the right eye, and 6/12 in the left eye. After improvement in the vision, the patient was discharged on oral steroids in a tapering manner along with azathioprine therapy. The patient was advised of total compliance of medications along with regular follow-ups.

DISCUSSION

Probable VKH is a rare autoimmune disease. Arevalo JF, Lasave AS, and Gupta V et.al mentioned in their article that VKH has been reported in a wide variety of populations such as with female predominance and typically in the age range of 20 - 50 years. [6] In our case, the patient was a 48-year-old Asian male. VKH typically affects females more frequently, with a male-to-female ratio of 1:2 as stated by Greco A, Fusconi M, Gallo A, Turchetta R et.al in their publication.[7]

The clinical progression of VKH is categorized into 4 phases. The patient presented to the OPD during the uveitic phase with bilateral redness in the eyes, and painless decreased vision more pronounced in the right eye, and reported seeing floaters in both eyes. Bilateral posterior uveitis was also observed along with multiple serous retinal detachments in both eyes. Until the last follow-up, this patient did not experience a recurrence.

The diagnosis of this condition primarily relies on clinical evaluation. This patient met the criteria of Probable VKH.[5] Treatment options include steroids, immunosuppressants, and immunoglobulins. For a favorable visual prognosis and to prevent inflammation recurrence, it is advised to continue treatment initially for at least six months. [4] Initially, the patient was administered high-dose steroids, with subsequent gradual reduction in dosage.

CONCLUSION

VKH, a multi-system disorder, is primarily a clinical diagnosis. Aggressive corticosteroid therapy is the cornerstone of treatment during the acute phase, often supplemented with immunosuppressive agents because of the

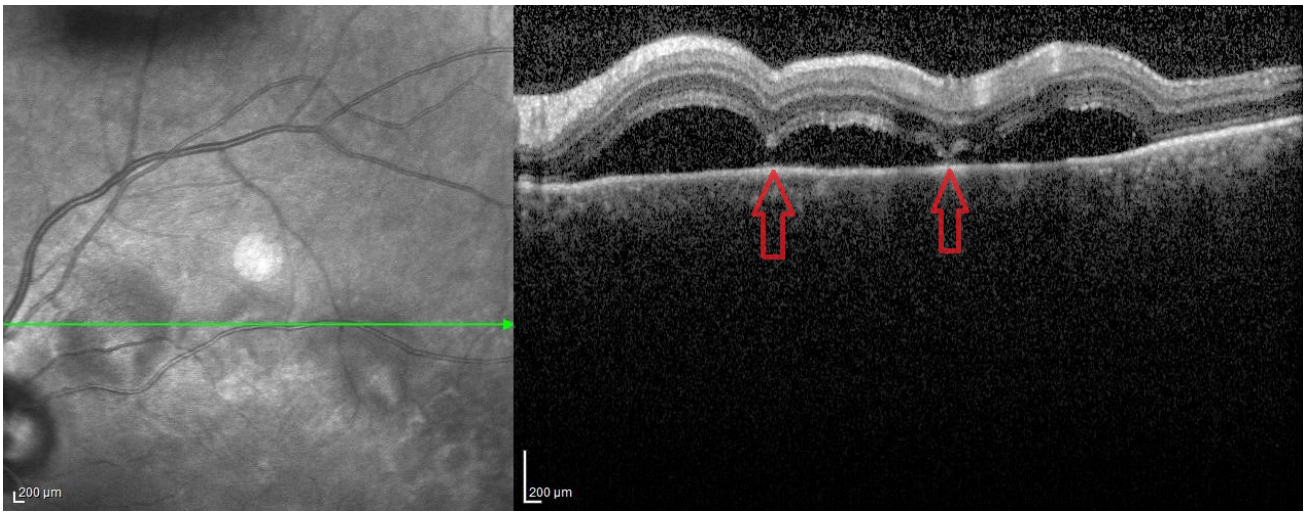

Figure 1: Optical coherence tomography (OCT) depicting multiple serous retinal detachments (SRDs) with septa (red arrows).

recurrences on tapering steroid therapy. Although there is a risk of substantial visual impairment, prompt recognition and treatment can mitigate ocular complications. VKH can lead to vision-threatening ocular complications. Consequently, regular and long-term follow-up with an Ophthalmologist is essential.

REFERENCES

1. Cunningham Jr ET, Rathinam SR, Tugal-Tutkun I, Muccioli C, Zierhut M. Vogt-koyanagi-Harada disease. Ocular Immunology and Inflammation. 2014 Aug 1;22(4):249-52.
2. Tugal-Tutkun I, Smit DP, Abu El-Asrar AM, Herbort Jr CP, Thorne JE. Vogt-Koyanagi-Harada Disease. Ocular Immunology and Inflammation. 2024 Apr 20;32(4):363-6.
3. Golzarri MF, Cheja-Kalb R, Concha-Del-Río LE, Gonzalez-Salinas R, Arellanes-García L. Risk factors for subretinal fibrosis in patients with Vogt Koyanagi Harada syndrome. Ocular Immunology and Inflammation. 2022 Feb 17;30(2):265-9.
4. Attia S, Khochtali S, Kahloun R, Zaouali S, Khairallah M. Vogt–Koyanagi–Harada disease. Expert Review of Ophthalmology. 2012 Dec 1;7(6):565-85.
5. Read RW, Holland GN, Rao NA, Tabbara KF, Ohno S, Arellanes-Garcia L, Pivetti-Pezzi P, Tessler HH, Usui M. Revised diagnostic criteria for Vogt-Koyanagi-Harada 6disease: report of an international committee on nomenclature. American journal of ophthalmology. 2001 May 1;131(5):647-52.
6. Arevalo JF, Lasave AF, Gupta V, Kozak I, Al Harbi MB, Al Rushood AA, Al Dhibi HA. Clinical outcomes of patients with Vogt–Koyanagi–Harada disease over 12 years at a tertiary center. Ocular immunology and inflammation. 2016 Sep 2;24(5):521-9.
7. Greco A, Fusconi M, Gallo A, Turchetta R, Marinelli C, Macri GF, De Virgilio A, de Vincentiis M. Vogt–koyanagi–harada syndrome. Autoimmunity reviews. 2013 Sep 1;12(11):1033-8.