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# CASE REPORT: SUCCESSFUL TREATMENT OF A PATIENT WITH VOGT-KOYANAGI- HARADA DISEASE.

Ophthalmology

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## ABSTRACT

We describe a 14 years old girl with the complaints of blurred vision in both eye and headache since 1 month. On Ocular examination the best corrected visual acuity was RE : CF 3 Metres , LE : CF 1 Metre. Intraocular pressure by applanation tonometry in right eye was 16mmHg and 12 mmHg in left eye. On examination anterior segment findings were unremarkable. On fundus examination with binocular indirect ophthalmoscopy, both eye showed disc edema with exudative retinal detachment in entire retina with greater severity in macula. Patient was diagnosed as probable VKH. Patient received high-steroid pulse therapy (intravenous methylprednisolone 1gm for 3 days) followed by oral prednisolone in tapering dose. Patient improved rapidly and significantly. After 1 week best corrected visual acuity was 6/12 in RE and 6/18 in LE.

## KEYWORDS

Vogt-Koyanagi-Harada , Prednisolone , successful treatment.

1. No history of penetrating ocular trauma or surgery preceding the initial onset of uveitis.
2. No clinical or laboratory evidence suggestive of other disease entities.
3. Bilateral ocular involvement in which either part a or b must be met, depending on the stage of the disease when the patient was examined
   a. Early manifestations of the disease
      (1) There must be evidence of a diffuse choroiditis (with or without anterior uveitis, vitreous inflammatory reaction or optic disk hyperemia) which may manifest as one of the following:
         (a) Focal areas of subretinal fluid
         (b) Bullous serous retinal detachment
      (2) With equivocal fundus findings, both of the following must be present as well:
         (a) Focal areas of delay in choroidal perfusion, multifocal areas of pinpoint leakage, large placoid areas of hyperfluorescence, pooling within the subretinal fluid and optic nerve staining (listed in order of sequential appearance) by fluorescein angiography
         (b) Diffuse choroidal thickening, without evidence of posterior scleritis by ultrasonography
   b. Late manifestations of the disease
      (1) History suggestive of prior presence of findings from 3a and either both 2 and 3 listed below or multiple signs from 3
      (2) Ocular depigmentation (either of following manifestations is sufficient)
         (a) Sunset glow fundus
         (b) Sugiura's sign
      (3) Other ocular signs (any of the following manifestations are sufficient)
         (a) Nummular chorioretinal depigmentation scars
         (b) Retinal pigment epithelium clumping and/or migration
         (c) Recurrent or chronic anterior uveitis.
4. Neurological/auditory findings (which may have resolved by the time of examination). (Any of the following manifestations are sufficient)
   a. Meningismus (malaise, fever, headache, nausea, abdominal pain, stiffness of the neck or back, or a combination of these factors. However, headache alone is not enough to meet a definition of meningismus)
   b. Tinnitus
   c. Cerebrospinal fluid (CSF) pleocytosis.
5. Integumentary findings (which do not precede the onset of central nervous system or ocular disease). (Any of the following manifestations are sufficient)
   a. Alopecia
   b. Poliosis
   c. Vitiligo.

*Complete Vogt-Koyanagi-Harada's disease–Criteria disease criteria* 1–5 must be present
*Incomplete Vogt-Koyanagi-Harada's disease–Criteria disease criteria* 1–3 and either 4 or 5 must be present
*Probable Vogt-Koyanagi-Harada's disease–Criteria disease isolated ocular disease-criteria* 1–3 must be present.

**Picture 1 : Revised Diagnostic criteria for VKH[2]**

## INTRODUCTION

Vogt–Koyanagi–Harada (VKH) is a multisystem autoimmune inflammatory disorder with ophthalmic, auditory, dermatologic, and neurologic manifestations. It occurs due to cell mediated responses against melanocytes.[2] The prevalence of this disease varies with ethnicity. VKH characteristically affects pigmented groups, such as Hispanics, Asians, people from the Middle East, and Asian Indians, but not the blacks of sub-Saharan African descent.[1] The extraocular manifestations appear in different phases of the disease and show ethnic variations.[1] VKH disease accounts for 8% of uveitis in Japan, 2.9% in the Middle East, 1.2% in Europe, and 1%–4% in the USA.[1] In a study from a referral eye care center in South India, the prevalence is reported as 1.4%–3.5%.[1] VKH is seen more frequently in an adult population, and women are more frequently affected.[1] VKH disease usually manifests as four distinct clinical phases: prodromal, acute uveitic, convalescent, and chronic recurrent.1 The prodromal phase features neurologic and auditory manifestations, including a headache, tinnitus, neck stiffness, and hearing loss. The acute uveitic phase shows diffuse choroiditis with exudative retinal detachment (RD) and optic disc swelling. A variable amount of anterior chamber and vitreous inflammation may be seen. In the acute phase, anterior uveitis seen in some patients usually presents as low-grade non-granulomatous anterior uveitis, while in the chronic recurrent phase, patients present with recurrent granulomatous anterior uveitis. "Sunset glow fundus," due to depigmentation of the choroid, is the hallmark of the convalescent phase. Recurrent episodes of anterior uveitis are characteristic of the chronic recurrent phase.

In a study by Rao et al, in an ethnically and geographically diverse group of patients, it is revealed that there are two clinical signs which are highly specific to VKH – exudative RD during the acute phase and sunset glow fundus during the chronic phase of the disease.3

Imaging technology has allowed a better visualization and understanding of the disease process in VKH. The first international workshop on VKH disease proposed criteria to make diagnosis.[4]

According to the revised diagnostic criteria ( Picture 1 ), VKH disease can be complete, incomplete, and probable. All the three categories have the absolute requirement of bilateral eye disease; however, a unilateral or delayed involvement of the other eye can occur in rare cases.[5-8].

Multiple treatment regimens have been tried for VKH disease, including intravenous, oral, regional corticosteroid, cyclosporine, antimetabolites, and alkylating agents.[1]

## CASE REPORT

A 14 years old girl belonging to lower middle socioeconomic status presented to us with the complaints of blurred vision in both eye and headache since 1 month.

On General examination BP: 110/70 mmHg, Pulse: 80 /min, Spot RBS – 95mg/dL .On Ocular examination the best corrected visual acuity was
**RE :** CF 3 Metres,
**LE :** CF 1 Metre

Intraocular pressure by applanation tonometry in right eye was 16mmHg and 12 mmHg in left eye.

On examination anterior segment findings were unremarkable. On fundus examination with binocular indirect ophthalmoscopy, both eye showed disc edema with exudative retinal detachment in entire retina with greater severity in macula.

OCT revealed both eye epiretinal membrane with elevated foveal contour with large subretinal fluid collection with multiple pigment epithelial detachment subfoveally and juxtafoveally. The central foveal thickness in right eye was 598 µm and 1265 µm. ( figure 1 and 2 )

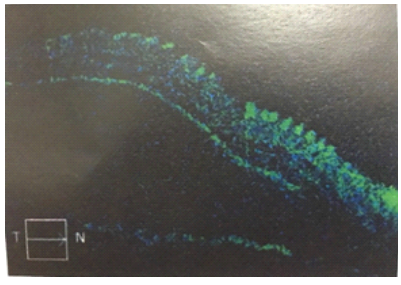


**Figure 1 : OCT picture of right eye on Day 1.**

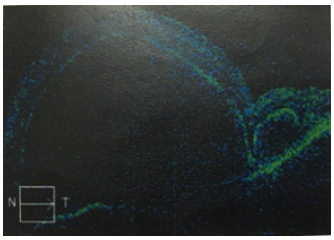

**Figure 2 : OCT picture of left eye on Day 1**

Ocular Ultrasound revealed bilateral anechoic vitreous cavity with choroidal thickening. ( figure 3 and 4 )

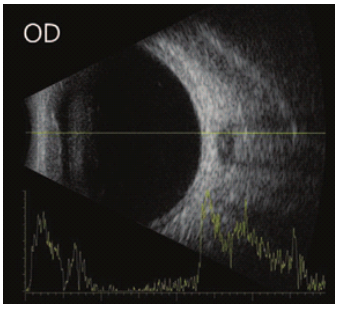


**Figure 3 : Right eye ultrasound showing choroidal thickening**

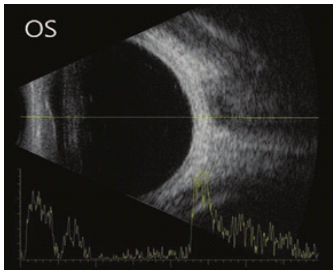


**Figure 4 : Left eye ultrasound showing choroidal thickening.**

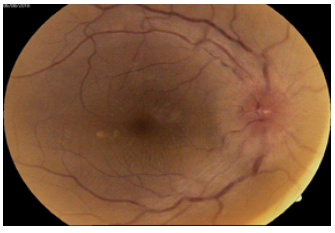

**Figure 5a and 5b : RE Fundus Photograph showing resolving Disc edema and serous detachment after pulse steroid treatment.**

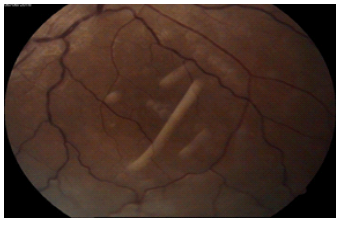

**Figure 5b**

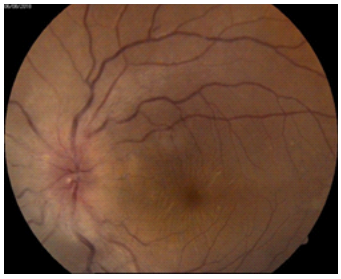

**Figure 6a and 6b : LE Fundus Photograph showing resolving Disc edema and serous detachment after pulse steroid treatment.**

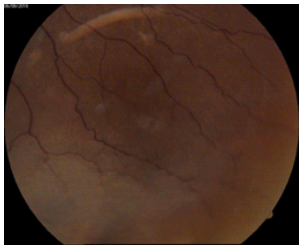

**Figure 6b**

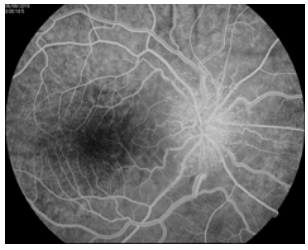

**Figure 7a**

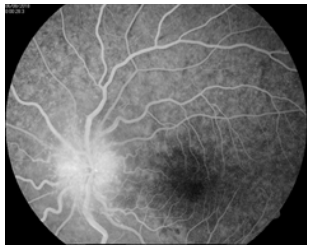

**Figure 7b**

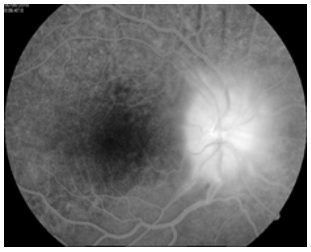

**Figure 8a**

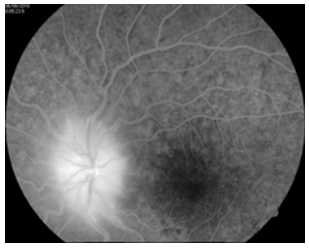

**Figure 8b**

A fundus fluorescein angiography after 3 days of Pulse steroid therapy showed optic disc hyperfluorescence with numerous pinpoint foci of leakage at the level of retinal pigment epithelium ( RPE ) in early phase ( figure 7a and 7b ) and subsequent disc leakage in late phase (figure 8a and 8b). It also shows hypofluorescent lines corresponding to choroidal folds. There are also multiple RPE window defects.

Blood tests including markers of inflammation and autoimmunity were negative as were the serological testing for the diagnosis of infectious pathologies. Chest xray was within normal limit.

MRI brain with optic cuts was done which showed BE choroidal thickening.

CSF analysis including opening pressure was normal except mild pleocytosis ( 10 cells per cubic mm ).

Based on above findings and according to the revised diagnostic criteria for VKH , patient was diagnosed as probable VKH.

High-steroid pulse therapy (intravenous methylprednisolone 1,000 mg for 3 days) was given followed by oral prednisolone in tapering dose. After 1week best corrected visual acuity was 6/12 in RE and 6/18 in LE.

The OCT showed significant decrease in central foveal thickness to 350 µm in RE and 354 µm in LE.( figure 9 and 10 )

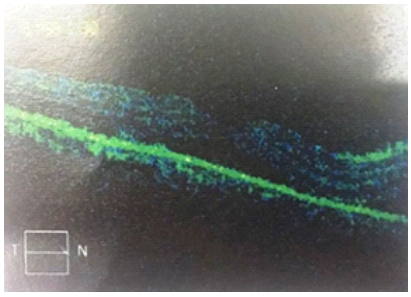


**Figure 9**

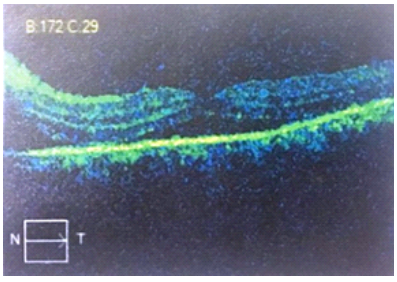


**Figure 10**

## DISCUSSION

The VKH disease is a multisystemic disorder that involves the eyes, ears, skin, hair and meninges.[2] The treatment of VKH disease should begin at the earliest stage because the prognosis is better if treatment is started then.[2] Thus it is critically important that the diagnosis of VKH be done accurately and as early as possible. High dose (1-2 mg/kg/day) oral prednisolone ,tapered over 3-6 months is the usual treatment which may be preceded by intravenous methlyprednisolone pulse therapy ( 500- 1000mg /day ).[9] Topical steroids and cycloplegics are used for anterior uveitis.[9] Steroid resistant patients may require immunosuppressives like cyclosporine, azathioprine and mycophenolate.[9]. Our patient though presented a bit late ( after one month of symptom onset ) successfully responded to intravenous pulse therapy followed by oral steroids and has not required immunosuppressives so far ( 2 months since the initiation of treatment.)

## CONCLUSION

This case presents successful treatment of VKH with systemic steroids which reveals that though it is rare condition a timely diagnosis and rational treatment may recover the patient significantly.

## ACKNOWLEDGENT

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## Conflict of Interest

The author has no disclosure.

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