

CASE REPORT

# Vogt-Koyanagi-Harada disease in prodromal stage misdiagnosed as Idiopathic Intracranial Hypertension: A Case Report.

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## Abstract

Vogt-Koyanagi-Harada (VKH) disease is an autoimmune disorder involving most commonly the nervous system, integumentary system, eye and ear. It causes bilateral granulomatous panuveitis associated with serous neurosensory layer detachments, papillitis and other complications. Advances in imaging modalities, especially optical coherence tomography (OCT) and fundus fluorescein angiography (FFA) that can evaluate pathognomic changes in the retina and choroid have immensely aided the diagnosis. Treatment with corticosteroids and, more recently, steroid-sparing immunomodulators have been found to be effective. We present a case of a young female who was misdiagnosed as a case of Idiopathic intracranial hypertension leading to significant visual morbidity. Though a diagnostic challenge especially in early-nonspecific prodromal phase, a good clinical history and evaluation, relevant laboratory workup and prompt and appropriate treatment can prevent irreversible visual loss and long-term ocular complications related to VKH syndrome.

**Key-words:** VKH, multisystem inflammatory disease, misdiagnosis, optical coherence tomography.

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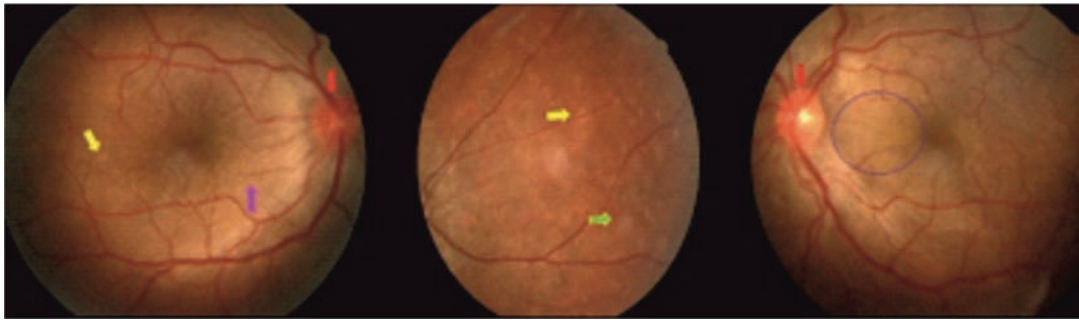
## Introduction

VKH syndrome is a rare T-cell mediated autoimmune disease involving multiple systems, most commonly nervous, ocular, auditory and integumentary system [1]. It is prone to be misdiagnosed and treated as other common disorders like Idiopathic Intracranial hypertension (IIH) especially in its prodromal stage, in view of headache and visual symptoms, which may lead to delay in early diagnosis and vision threatening complications.

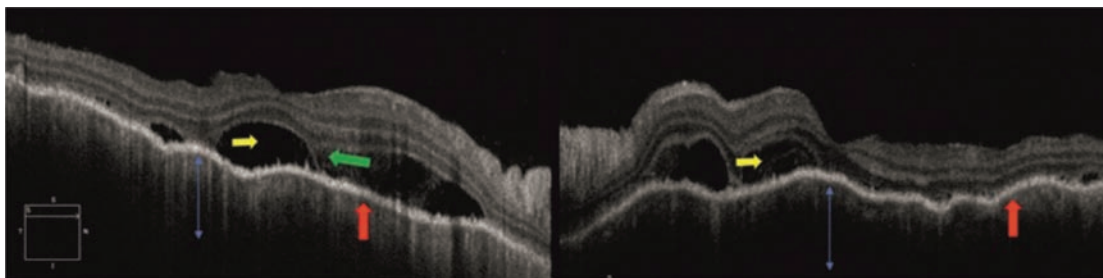
## Case Report

A 26-year-old lactating female had headache for the last 6 weeks and blurring of vision in both eyes for 4 weeks. The headache was moderate in intensity, central and bifrontal in location, throbbing in nature, and was associated with nausea and episodes of vomiting. It increased on bending forward and relieved on taking analgesics. Blurring of vision was insidious in onset, painless, gradually progressive. She also gave history of pulsating tinnitus in both ears for the same duration. There was no history of similar complaints in the past, or ocular trauma. There were no associated flashes, floaters, metamorphopsia, red colour desaturation, pain on eye movement, colored halos, joint pain, genitourinary problem, focal neurological deficit, skin rash, high risk sexual behavior or travel to

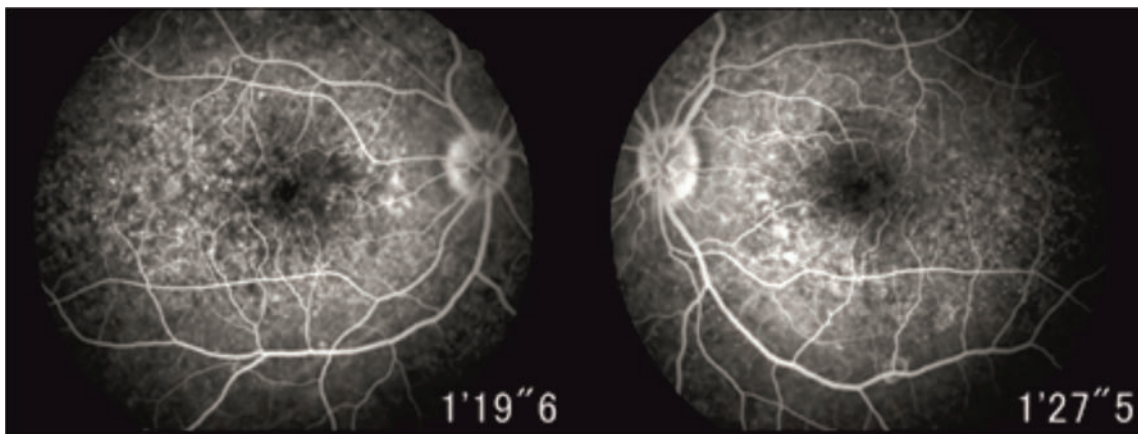
any endemic area or raising pets. There was no history of recent covid infection, vaccination, or oral contraceptive use. Patient had a normal delivery 5 months back. The pregnancy went uneventful. Patient was being treated at an outside centre as a case of Idiopathic Intracranial Hypertension since last 10 days and was prescribed Tab Acetazolamide 250 mg twice a day. Systemic examination was essentially within normal limits, except for mild terminal neck rigidity. The best corrected visual acuity ( BCVA) was 3/60 in right eye and 6/60 in left eye. Pupillary reaction was sluggish. Both eyes had hyperemic disc (right>left), multiple well defined hypopigmented subretinal fluid pockets in mid periphery. Right eye macula had a dull foveal reflex due to subretinal fluid (SRF) (**Figure 1**) [2]. Ultrasound- B scan revealed a thickened retinochoroidal complex. Both eye OCT imaging had large multiple serous retinal detachment with fibrin and membranous septae. Characteristic Bacillary layer detachment was noted. The retinal pigment epithelium (RPE) was thick undulated. Choroid thickness and choroidal vascularity index were increased (**Figure 2**) [3]. FFA revealed a leaky disc, multiple pinpoint leakage throughout the retina at level of RPE giving the characteristic Starry sky appearance. Pooling was noted in Subretinal spaces in late phase (**Figure 3**) [4]. Routine blood investigation were within normal limit. Antibody titres for Syphilis, Lyme's disease, Borrelia, TORCH was negative,



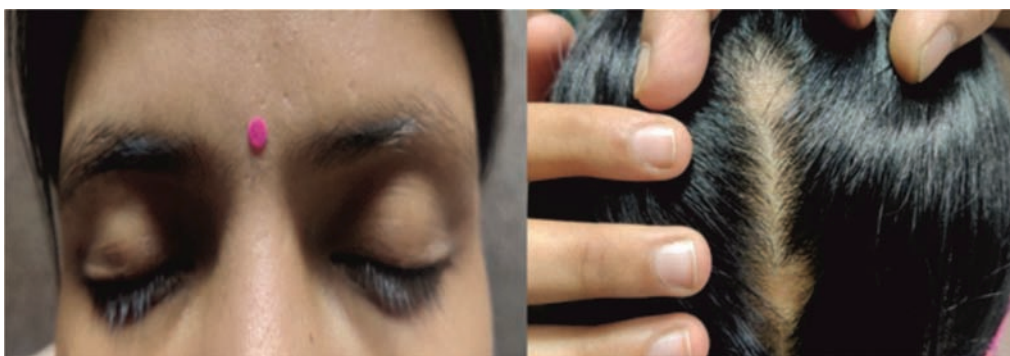
**Figure 1: Fundus Photograph of RE and LE: Hyperemic disc (red arrow), SRF at macula (purple), multiple focal neurosensory detachment (yellow)**



**Figure 2: OCT Macula of RE and LE: Thick Undulated RPE (red arrow), Large multiple serous retinal detachments and cystoid spaces with fibrinous fluid separated by membranous septae (yellow), bacillary layer detachment (green), Thickened choroid (blue)**



**Figure 3: FFA of RE and LE: Leaky disc and multiple pin point leaks: Starry sky appearance**



**Figure 4: Scalp alopecia and Eyebrow madarosis and poliosis**

Quantiferon TB gold test was negative.

Contrast MRI Brain had no significant abnormality. CSF had mild pleocytosis (15 cells, 98% lymphocytes), increased protein (110mg/dl) and normal sugar levels (75mg/dl). CSF gram stain, fungal stain, Cryptococcal Antigen, ZN stain, culture sensitivity, ADA, Gene Xpert tests were all negative.

In view of above findings, a diagnosis of Incomplete VKH syndrome was made based on the Revised AUS guidelines [5].

Tab Acetazolamide was stopped. High dose Oral Prednisolone (1mg/kg) was started. Headache decreased in 4 days and visual recovery and normalization of OCT findings were noted within the first week. After 2 weeks, the patient was started on Tab. Azathioprine 50mg B.D. for 4 weeks, followed by 50mg O.D., which was continued with monitoring of blood counts and liver functions [6]. The steroid was then gradually tapered over 6 months. At 6 weeks the BCVA of both eyes was 6/6 with complete resolution of SRF, normalization of RPE and choroid morphology on OCT [7].

The patient on follows up, presented with patchy hair loss over scalp and madarosis, poliosis of lateral aspect of eyebrows, hence confirming our diagnosis of VKH syndrome (Figure 4). The patient at 12 months of follow up had stable vision, no recurrence or signs of extensive depigmentation on ocular examination or any other complications [8].

## Discussion

VKH syndrome is a multisystem inflammatory disease affecting mainly the nervous, ocular, auditory system and skin. It causes granulomatous posterior or panuveitis associated with serous retinal detachments, disc edema, with eventual development of a depigmentary sunset glow fundus with significant visual morbidity. Diagnosis involves the exclusion of other ocular diseases, no history of recent surgery or penetrating eye trauma, bilateral eye involvement with evidence of diffuse choroiditis, auditory (tinnitus), neurological findings (meningismus, headache), and dermatological findings including depigmentation, alopecia, poliosis, madarosis [5].

In prodromal phase, the nonspecific symptoms and ocular findings can be misdiagnosed with other entities like posterior uveitis, neuroretinitis, Benign intracranial hypertension, posterior scleritis or any other infective etiology. A thorough systemic and ocular examination especially dilated fundus examination can clinch the diagnosis. Imaging modalities like OCT and Fluorescein angiography are excellent diagnostic tools. Prompt treatment with steroids leads to excellent visual recovery. Immunosuppressants like azathioprine, mycophenolate mofetil, cyclosporine, methotrexate etc. are effective in cases of steroid intolerance or for long term therapy. OCT is helpful on subsequent follow up to gauge therapeutic response and track recurrences.

## Conclusion

VKH syndrome is an autoimmune disease with myriad of signs and symptoms which may be misdiagnosed with other entities including benign intracranial hypertension. A thorough history, systemic and ocular examination with relevant investigations will help to prevent its misdiagnosis, thereby helping the clinician to initiate prompt treatment in the form of steroids and immunosuppressants to prevent its dangerous complications.

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## References

1. Moorthy RS, Inomata H, Rao NA. Vogt-Koyanagi-Harada syndrome. *Surv Ophthalmol.* 1995 Jan-Feb;39(4):265-92.
2. Rao NA, Gupta A, Dustin L, Chee SP, Okada AA, Khairallah M, et al. Frequency of distinguishing clinical features in Vogt-Koyanagi-Harada disease. *Ophthalmology.* 2010 Mar;117(3):591-9. 599.e1.
3. Liu XY, Peng XY, Wang S, You QS, Li YB, Xiao YY, Jonas JB. Features of optical coherence tomography for the diagnosis of vogt-koyanagi-harada disease. *Retina.* 2016 Nov;36(11):2116-2123.
4. Fardeau C, Tran TH, Gharbi B, Cassoux N, Bodaghi B, LeHoang P. Retinal fluorescein and indocyanine green angiography and optical coherence tomography in successive stages of Vogt-Koyanagi-Harada disease. *Int Ophthalmol.* 2007 Apr-Jun;27(2-3):163-72.
5. Sakata VM, da Silva FT, Hirata CE, de Carvalho JF, Yamamoto JH. Diagnosis and classification of Vogt-Koyanagi-Harada disease. *Autoimmun Rev.* 2014 Apr-May;13(4-5):550-5.
6. Read RW, Yu F, Accorinti M, Bodaghi B, Chee SP, Fardeau C, et al. Evaluation of the effect on outcomes of the route of administration of corticosteroids in acute Vogt-Koyanagi-Harada disease. *Am J Ophthalmol.* 2006 Jul;142(1):119-24.
7. Hashizume K, Imamura Y, Fujiwara T, Machida S, Ishida M, Kurosaka D. Retinal pigment epithelium undulations in acute stage of vogt-koyanagi-harada disease: Biomarker for Functional Outcomes After High-Dose Steroid Therapy. *Retina.* 2016 Feb;36(2):415-21.
8. Potapova NV, Yeh S, Smith JA, Jirawuthivoravong G, Mahdi N, Chan CC, et al. Ocular Complications in Vogt-Koyanagi-Harada Disease. *Investigative Ophthalmology & Visual Science.* 2007 May 10;48(13):5145-5145.

