Case Report

# A Tale of Two Syndromes: Vogt–Koyanagi–Harada Disease and Acquired Immunodeficiency Syndrome in a Nigerian Female

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## Abstract

A 54-year-old female Nigerian presented with a 1-week history of sudden deterioration of vision in both eyes. There was no antecedent history of ocular trauma, floaters, nor flashes of light. However, she gave a history of a febrile illness associated with headaches and malaise 2 weeks prior to onset of ocular symptoms. She is a known retroviral-positive patient on treatment with highly active antiretroviral therapy for the past 5 years but is not a known hypertensive nor diabetic. At presentation, the best corrected visual acuity was hand movement in both eyes. Anterior segment examination of both eyes revealed fine keratic precipitates on the corneal endothelium with flare and inflammatory cells in the anterior chamber and grade 1 nuclear sclerosis. Dilated binocular indirect ophthalmoscopy of both eyes revealed pink disks with blurred margins total exudative retinal detachments. An assessment of Vogt–Koyanagi–Harada syndrome in a patient with human immunodeficiency virus/acquired immunodeficiency syndrome was made. Bilateral exudative detachment resolved with improvement of her best corrected visual acuity to 6/9 in both eyes after systemic treatment with steroids.

**Keywords:** Vogt-Koyanagi-Harada, HIV/AIDS, Exudative retinal detachment, Vitiligo, Hearing deficit, Alopecia, HAART

## Introduction

Granulomatous inflammatory conditions involving the eyes typically present with mutton fat keratic precipitates, inflammatory cells in the anterior chamber, vitritis, panuveitis, and granulomas. These diverse etiology of granulomatous ocular inflammation which include tuberculosis, sarcoidosis, and Vogt-Koyanagi-Harada disease (VKH) may present with exudative retinal detachments. Detailed clinical evaluation and investigations are the bedrock of differentiating and arriving at the definitive diagnosis of these varying ocular granulomatous inflammatory conditions.[1-3]

The VKH syndrome is a granulomatous, multisystemic, autoimmune disease which affects pigmented tissue such as the eye, inner ear, meninges, skin, and hair which are populated by large numbers of melanocytes.[3] The common ophthalmic presentation of VKH is with bilateral diffuse panuveitis. This syndrome was first described by Koyanagi and then Harada and Vogt at different times; subsequently, the identified clinical signs were noted to involve the same spectrum; hence, the name VKH syndrome.[4] It typically occurs in Asians, people from the middle East, and native Americans but said to be quite rare in Africans.[5]

Nevertheless, a few cases have been documented in people of African descent including an African-American, two Nigerians and a Ugandan respectively.[6-8] The occurrence of VKH with human immunodeficiency virus/acquired immunodeficiency syndrome (HIV/AIDS) is rare and only one case in an Indian female has been documented in literature to the best of our knowledge.[9] This case report showcases the rare combination of VKH syndrome and HIV/AIDS infection in a female Nigerian.

Access this article online

Quick Response Code:

Website: www.nigerianjournalofophthalmology.com

DOI: 10.4103/njo.njo_18_21

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**Received:** 3 September 2021 **Revised:** 20 September 2021
**Accepted:** 15 January 2022 **Published:** 20 May 2020



**How to cite this article:** Babalola YO, Oluleye TS, Majekodunmi OI, Ijaduola MA. A Tale of Two Syndromes: Vogt–Koyanagi–Harada Disease and Acquired Immunodeficiency Syndrome in a Nigerian Female. Niger J Ophthalmol 2022;30:28-32.

## Case Report

A 54 year old woman presented via the accident and emergency department with a 1-week history of sudden deterioration in vision in both eyes which she noticed while driving. There was no antecedent history of trauma nor entry of any foreign body. She gave a positive history of a febrile illness associated with malaise and frontal headaches 2 weeks prior to the onset of ocular symptoms. There was no associated history of floaters, flashes of light, haloes, nor ocular pain. She has been using spectacles for reading in the last 5 years. There was no associated history of cough, weight loss, night sweats, nor neck stiffness. An onset of hearing deficit and tinnitus were noticed at the time of onset of ocular symptoms by the patient. She is a known retroviral positive patient diagnosed ten years ago on highly active antiretroviral therapy (tenofovir and lamivudine). She is being treated for peptic ulcer disease but she is not a known hypertensive or diabetic. She has no known allergies and has never received blood transfusions.

The best corrected visual acuity at presentation was hand movement in both eyes with accurate light projection. Anterior segment examination of both eyes showed fine, fresh nonpigmented keratic precipitates on the endothelium in both eyes with flare and grade 2 inflammatory cells. Bilateral nuclear sclerosis grade 2 was present. The intraocular pressure measured by applanation tonometry was 10 mmHg in both eyes.

Ocular examination of the fundi with binocular indirect ophthalmoscopy revealed vitreous cells with blurring of the disc margins, shifting subretinal fluid with a total bullous, and exudative detachment bilaterally. No retinal breaks nor holes were identified. Bilateral exudative retinal detachment with retinochoroidal thickening was confirmed with ocular B-scan ultrasonography, as shown in Figure 1a & b.

The general and systemic examinations were essentially normal. An assessment of bilateral panuveitis in a known retroviral disease patient to rule out choroidal tuberculosis and posterior scleritis was made.

Various investigations were ordered to identify the possible etiology of the bilateral exudative retinal detachments. The erythrocyte sedimentation rate was 36 mm/hour which was slightly above the normal value of $\leq$20 mm/hour in females, whereas the $CD4^+$ count of 1011 cells/$mm^3$ was within the normal range of 500 to 1200 cells/$mm^3$. The viral load was 10 copies/mL which is <200 copies/mL and is interpreted to mean that our patient had undetectable virus levels indicating good response to HAART; hence, she cannot transmit the virus to others. The full blood count, fasting blood sugar, liver function test, chest X-ray, and Mantoux test were all within normal limits. These investigations ruled out the possibility of infective etiology such as tuberculosis and other granulomatous inflammations such as sarcoidosis.

Our patient was commenced on guttae dexamethasone 2 hourly both eyes, guttae tropicamide tds, tablets omeprazole 20 mg bd, mist magnesium trisillicate, and intravenous methyl prednisolone 1000 mg at 1 mg/kg body daily for 3 days. She was discharged home and subsequently commenced on tablets prednisolone 60 mg daily which was gradually tailed off during her follow-up clinic visits over 3 months.

She was given a referral to the ENT clinic for review by the otorhinolaryngologists who scheduled her for audiometry. The $CD4^+$ and viral load levels were also being monitored by the infectious disease physicians.

During the first 4 weeks of her weekly follow-up visit after discharge, the best corrected visual acuity remained at hand movement in both eyes with minimal resorption of the subretinal fluid. At the sixth week follow-up visit, post-commencement of treatment with steroids, the best corrected visual acuity was noticed to have improved to 6/24-1 and 6/18 in the right and left eyes, respectively.

She also complained of hypopigmented patches on her face and forehead associated with loss of hair around the temples 2

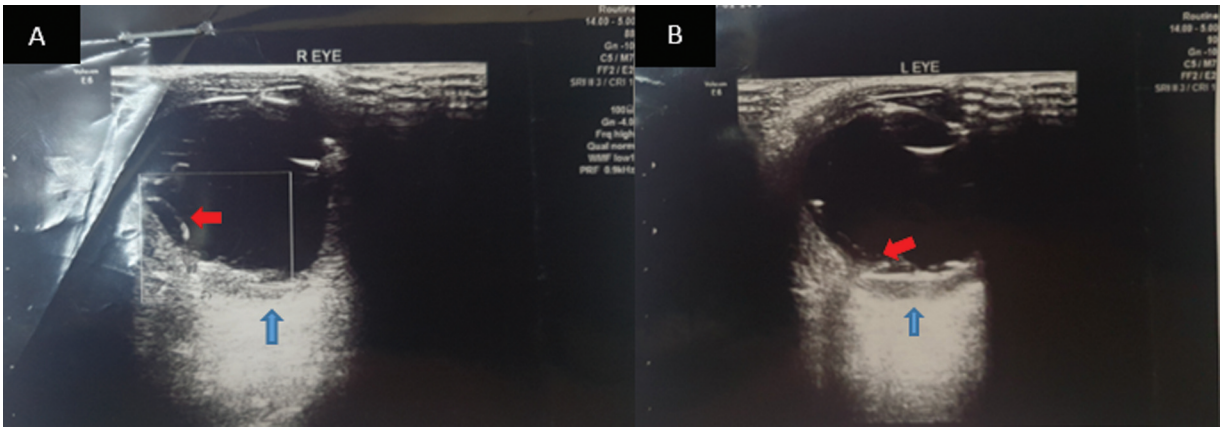


**Figure 1:** Ocular B-scan ultrasonography images of the right and left eyes at presentation showing subretinal fluid (red arrow) & retinochoroidal thickening (blue arrow).

weeks prior to this clinic visit, as shown in Figure 2. Dilated binocular indirect ophthalmoscopy revealed some resorption of the subretinal fluid with shallow retinal detachment [Figure 3a,b]. Optical coherence tomography of the macula performed after commencement of corticosteroid therapy as the subretinal fluid resolved showed few intraretinal cystic spaces with atrophy of the retinal pigment epithelium and loss of the ellipsoid zone bilaterally which was more pronounced in the left eye [Figure 4a & b].

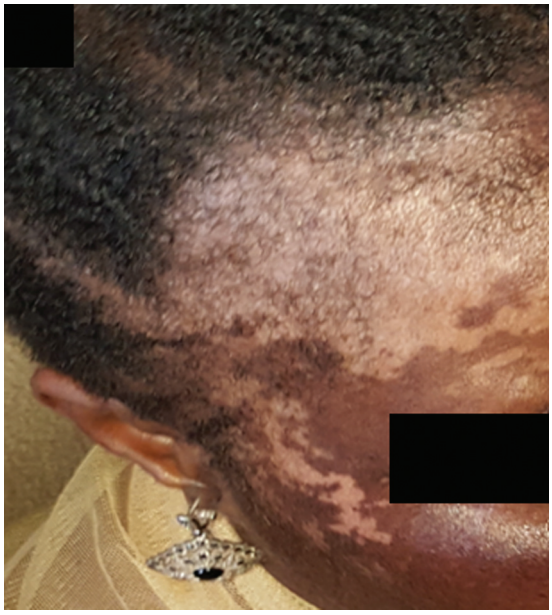

**Figure 2:** Hypopigmented vitiligo patches are seen on the face and forehead with areas of alopecia along the frontal hairline and temples of the patient.

The visual acuity progressively improved over the next 3 months of follow-up and by the fourth month postcommencement of treatment best corrected visual acuity in both eyes had improved to 6/9 bilaterally. The anterior segment was essentially quiet bilaterally with intraocular pressures of 13 mmHg by applanation tonometry in both eyes.

Fundus examination revealed cup disk ratio of 0.3, mild temporal "disc" pallor, resolution of vitritis, attenuated vessels, few hyperpigmented spots at the macula with total resorption of the subretinal fluid, and an orange, sunset glow appearance of both fundi, as shown in Figure 5. Oral prednisolone was gradually tailed off. From 60 mg alternate days over 6 months.

As at the last follow-up visit, her ocular status is stable with best corrected visual acuity maintained at 6/9 in both eyes. She is regular on her HAART and being followed up by the infectious disease unit.

## Discussion

Our index patient, a known retroviral infection on HAART presented with a 1-week history of deterioration ofvision in

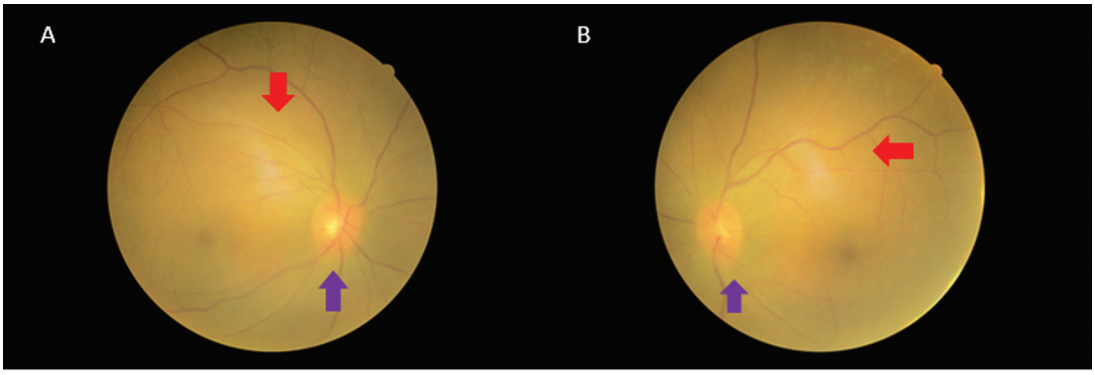


**Figure 3:** Fundus photographs of both eyes showing blurring of the disc margins more pronounced in the left eye (purple arrow), resolving subretinal fluid (red arrow) and hazy media presumably due to vitritis.

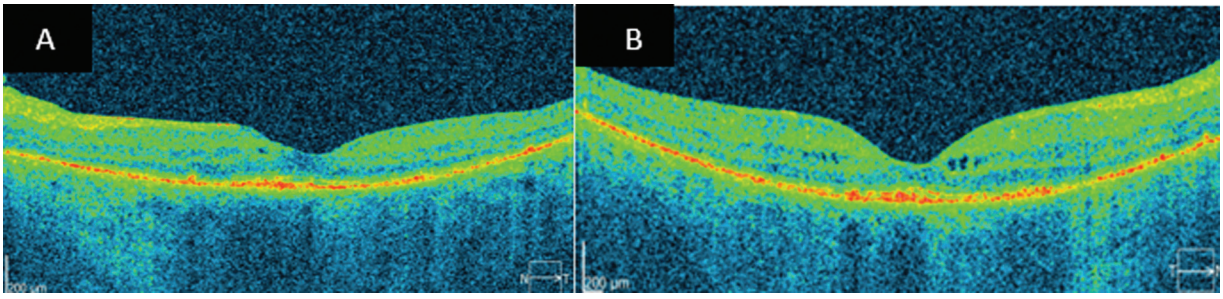


**Figure 4:** Optical coherence tomography scans of the right and left eye after commencement of corticosteroid therapy as subretinal fluid resolved showing few intraretinal cystic spaces, disruption of the ellipsoid zone and retinal pigment epithelium atrophy more pronounced in the left macula.

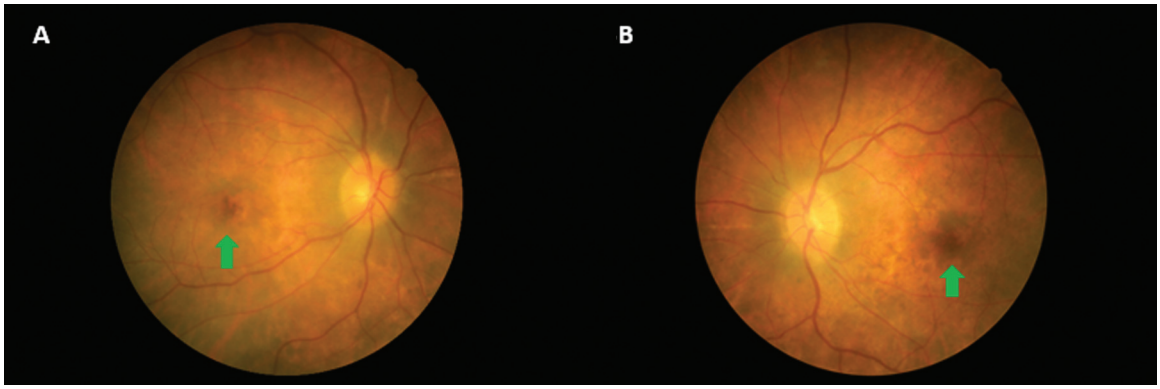


**Figure 5:** Fundus photographs of both eyes showing orange, sunset glow appearance, retinal pigment epithelial and atrophic changes (green arrow) at the macula after resolution of the exudative retinal detachments.

both eyes. Clinical features of bilateral panuveitis represented by fresh keratic precipitates, anterior chamber cells, vitreous cells, and bilateral exudative retinal detachment were present in both eyes. Systemic and ocular investigations performed ruled out inflammatory conditions such as tuberculosis and posterior scleritis that could present in a similar pattern, hence, the delay in diagnosis of VKH. The appearance of vitiligo and alopecia aided in clinching the diagnosis of VKH in subsequent clinic visits.

Immune recovery posterior scleritis is described as a non-infectious occurring in retroviral-positive patients with cytomegalovirus retinitis or other intraocular infections. It occurs when there is a substantial increase in $CD4^+$ T-lymphocyte count. This diagnosis was ruled out in our patient, as the signs of unilaterality of disease, periocular pain, and the classic T-sign of diagnosis was absent and our patient had $CD4^+$ count values within normal limits.[10] Sympathetic ophthalmitis was also ruled out as a possible etiology of the exudative detachment, as there was no antecedent history of ocular trauma or surgery.

Vogt-Koyanagi-Harada syndrome has been classified into four stages namely the prodromal phase, the acute uveitic stage, the convalescent phase and the chronic/recurrent stage.[3] Our index patients' symptoms started with the headaches, febrile illness, malaise, and tinnitus in the prodromal stage before she subsequently presented to our retina unit in the acute uveitic stage with bilateral panuveitic features and exudative retinal detachment.

She had good response to intravenous methylprednisolone and subsequently oral prednisolone which was gradually tailed off over a 6-month period. Corticosteroid therapy is the main stay of treatment in VKH, though immune modulators such as azathioprine and methotrexate may be used especially in cases of patients with poor response to steroids.[3,6,7,9] Our patient had a good response to corticosteroid therapy with resolution of her ocular symptoms and improvement in the best corrected visual acuity from hand movement at initial presentation to 6/9, respectively, in both eyes.

Depending on the presenting clinical features, diagnosis of VKH may be classified as complete, incomplete, or probable VKH.[5,11] Our patient had a diagnosis of complete VKH as all the five defining features which include no preceeding history history of ocular trauma, lack of laboratory evidence of other uveitic entities, bilateral ocular involvement presenting as choroiditis, or reflected as bullous and serous retinal detachment. Other defining features in our patient were diffuse choroidal thickening on B-scan ultrasonography and sequelae of sunset glow fundi with retinal pigment epithelium clumping. Presence of neurologic and auditory findings of tinnitus and headaches and integumentary finding of both vitiligo and alopecia established the features of complete VKH thus, clinching the diagnosis.[11]

Sunset glow fundus is a clinical feature in the convalescent phase; a sequelae of exudative retinal detachment and depigmentation of the choroid are observed as a characteristic orange glow of the retina due to loss of choroidal melanocytes.[12] This was also the endpoint of the fundus changes in our patient after reabsorption of subretinal fluid with some pigmentation at the macula.

VKH is thought to be an autoimmune inflammatory condition that is mediated by T-lymphocyte cells that target melanocytes with a larger proportion of cells being of the helper $CD4^+$ cells variety in comparison with the cytotoxic $CD8^+$ cells.[3,5,13] The human immunodeficiency virus targets the $CD4^+$ cells which usually initiate immune response of the body.[14] Hence, it has been postulated that an immune dysfunction may be responsible for this occurrence as both VKH and HIV/AIDS act along similar cell lines of the $CD4^+$ and $CD8^+$ cell varieties.

Only one other case of VKH with HIV/AIDS has been reported in literature in which the patient was diagnosed with probable VKH in the chronic recurrent stage with bilateral cataracts, secondary angle closure glaucoma, sunset glow fundus, and vitiligo.[9] These authors also considered the fact that this coexistence in this patient may just be a rare incidence.

The $CD4^+$ count of our patient was 1100 c/mm, whereas her viral load was 10 copies/mL which were both within normal limits with no history or clinical signs suggestive of recent opportunistic infection or immunosuppression. In depth research is necessary to understand and correlate any associations between VKH and HIV/AIDS infection.

## Conclusion

VKH is a possible etiology of bilateral panuveitis in patients with retroviral disease. It is essential to thoroughly investigate and rule out other infective and inflammatory ocular conditions more commonly associated with HIV/AIDS to arrive at the diagnosis. Auditory deficit, alopecia, poliosis, and vitiligo in VKH are a sequelae of destruction of melanin-bearing cells due to the autoimmune response attack on the melanocytes and melanocyte-associated antigens. Though a rare association of HIV/AIDS, a high index of suspicion, detailed clinical history, and evaluation with appropriate laboratory work-up is essential for prompt and accurate diagnosis and management.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

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