Management of a rare presentation of Vogt-Koyanagi-Harada disease in human immunodeficiency virus/acquired immunodeficiency disease syndrome patient

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Vogt-Koyanagi-Harada (VKH), a multisystem autoimmune bilateral panuveitis with systemic manifestations, is uncommon in immunocompromised patients such as human immunodeficiency virus (HIV)/acquired immunodeficiency disease syndrome (AIDS). We report a rare presentation of VKH in a 45-year-old HIV-positive female on highly active antiretroviral therapy (HAART) who presented with a history of recurrent panuveitis. A diagnosis of probable VKH was made based on ocular and systemic signs and symptoms. She was treated with topical and systemic steroids with close monitoring of CD4 counts and viral loads. After inflammation control, complicated cataract was managed surgically under perioperative steroid cover. VKH in HIV/AIDS has not been reported earlier. This case shows that significant inflammation

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can be seen even in HIV/AIDS patients on HAART with VKH in spite of moderate CD4 counts. Management is a challenge considering the systemic risks with long-term use of steroids.

Key words: Acquired immunodeficiency disease syndrome, cataract surgery, complicated cataract, human immunodeficiency virus, Vogt-Koyanagi-Harada

Vogt-Koyanagi-Harada (VKH),[1,2] a bilateral granulomatous panuveitis with various systemic manifestations, is relatively uncommon even in immunocompetent individuals, and its occurrence in immunocompromised patients such as human immunodeficiency virus (HIV)/acquired immunodeficiency disease syndrome (AIDS) has not been reported. Here, we are reporting a rare presentation of VKH in a patient with HIV/AIDS, its course, and challenges in medical and surgical management.

Case Report

A 45-year-old Indian female presented to us with a history of recurrent redness of both eyes and gradual diminution of vision associated with headache and gradual changes in skin pigmentation since the previous 2 years. She was treated as bilateral panuveitis with topical steroids elsewhere. She was known HIV positive since 10 years and was on highly active antiretroviral therapy (HAART) (efavirenz 600 mg, lamivudine 300 mg, and tenofovir 300 mg). Her nadir CD4 counts were 87 cells/μl and CD4 count at the time of presentation was 170 cells/μl. General physical examination showed vitiliginous patches over face, back, and upper and lower limbs [Fig. 1a and b].

Her best-corrected visual acuity (BCVA) was hand movements in the right eye and 6/60 in the left eye. Anterior segment examination revealed active anterior segment inflammation with complicated cataract in both eyes with corneal edema in the right eye [Fig. 2a and b]. Applanation tonometry revealed an intraocular pressure (IOP) of 50 mmHg in the right eye and 10 mmHg in the left. Gonioscopy showed synechial angle closure in the right and narrow angles in the left eye. Ultrasound B-scan of both eyes showed attached retina with other evidences of vitreous inflammation and choroidal thickness 2.1 mm in the right eye and 1.9 mm in the left eye.

Secondary angle closure glaucoma was treated with yttrium-aluminum-garnet (YAG) peripheral iridotomy in both eyes, and patient was started on antiglaucoma medications (AGM). Post-YAG, dilated fundus evaluation revealed a panuveitis with sunset glow fundus. The patient underwent tests to rule out other possible causes of panuveitis such as chest X-ray, Mantoux tests ELISA toxoplasma, syphilis, cryptococci, and serum angiotensin-converting enzyme which were all within normal limits. A diagnosis of probable VKH with secondary glaucoma and complicated cataract was made. She was started on a tapering course of topical and systemic steroids along with AGM with constant monitoring of her CD4 counts and systemic status under care of a HIV physician. At 1-month follow-up, IOP and inflammation were under control, and her systemic status was stable. At 2-month follow-up, inflammation was under control, but right eye had progressively developed total intumescent cataract and she underwent phacoemulsification with acrylic hydrophilic intraocular lens implantation. Postoperatively, she had fibrinous inflammation, which was managed with topical and systemic steroids. Similarly, at 3-month follow-up, left eye complicated cataract was managed by a similar procedure under steroid cover**.** Optical coherence tomography of both eyes done at this visit revealed foveal thickness of 156 μ in the right eye and 152 μ in the left eye. At last follow-up of 1 year of initial presentation, BCVA was 6/7.5, N6 in both eyes [Fig. 3a and b] with inflammation under control [Fig. 4a and b]. Her CD4 count was 284 cells/μl while the patient was on regular HAART.

Discussion

A diagnosis of probable VKH was made in our patient according to revised diagnostic criteria of VKH.[3,4] Our patient had ocular features suggestive of the same and was on chronic reactivation phase. VKH, a multisystem autoimmune bilateral panuveitis, is uncommon in immunocompromised patients such as HIV/AIDS.

VKH reflects autoimmunity against melanocytes.[2,5] Studies show that predominant infiltrating cell in the choroid is T lymphocyte with a larger proportion of helper cell (CD4+) than cytotoxic cells (CD8+) along with Class II major histocompatibility complex molecules. The infiltrating CD4+ T cells recognize human melanocyte antigens. Furthermore, cytokines such as interleukin-7 (IL-7), IL-21, and IL-23 stimulate CD4+ T lymphocyte to produce IL-17, which in turn leads to the development of uveitis in VKH. The triggering factor for this process is still unknown.

HIV mainly targets CD4 T lymphocytes, which are necessary to initiate immune response.[6] A decline in CD4 counts leads to progressive immune deficiency, mainly cell-mediated immunity. A possible hypothesis is that there could have been an immune dysfunction since they seem to be acting on similar cell lines. It might be just a coincidental occurrence of VKH in a patient with HIV/AIDS. Even this is a rare occurrence of an immune-based disease in an immunocompromised individual and significant as it poses a therapeutic challenge considering the need for long-term use of systemic steroids and/or immunomodulators.

Uveitis in HIV patients without any obvious clinical ocular opportunistic infections (OIs) could be drug induced, immune recovery, or HIV *per se*, which is a diagnosis of exclusion.[7] There was no clinical evidence of any ocular OI or immune recovery uveitis[8,9] in our patient. Significantly, our patient had intense inflammation even with moderate CD4 counts.

Vitiliginous patches have also been described in patients with HIV without uveitis due to possible mechanism of destruction of melanocytes by autoimmune-mediated mechanism due to release of protein from dying CD4 cells.[10] In our patient, the depigmentation was seen as part of a convalescent phase of VKH along with sunset glow fundus.

Our patient was a known case of HIV/AIDS on antiretroviral therapy who developed ocular and systemic inflammatory features very characteristic of VKH. Considering that the patient at presentation was in Stage III immunosuppression,[11] with CD4 counts of 170 cells/μl, it was interesting to note that she presented with significant inflammation with VKH which is a unique feature. This possibly indicates that in such scenario of noninfective uveitic entities in patients with HIV/AIDS, the inflammatory response resembles the underlying uveitic

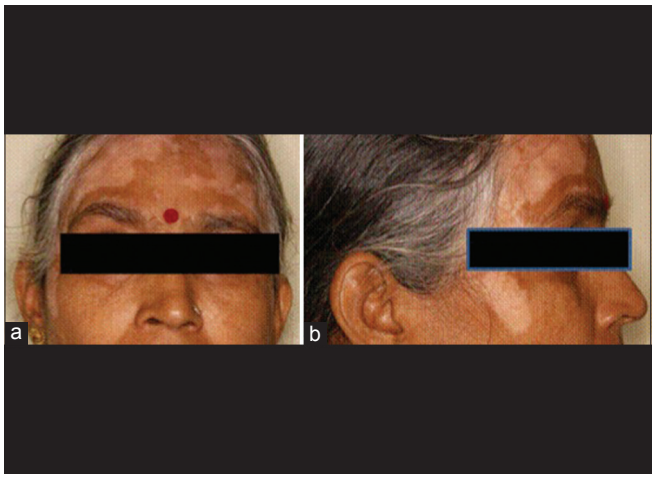

Figure 1: (a and b) External photographs showing vitiliginous patches over forehead and face

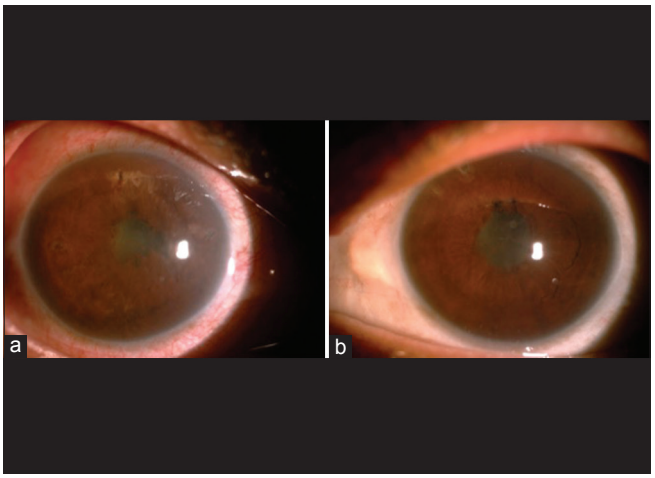

Figure 2: (a and b) Slit-lamp photographs of both eyes showing festoon-shaped pupil with complicated cataract

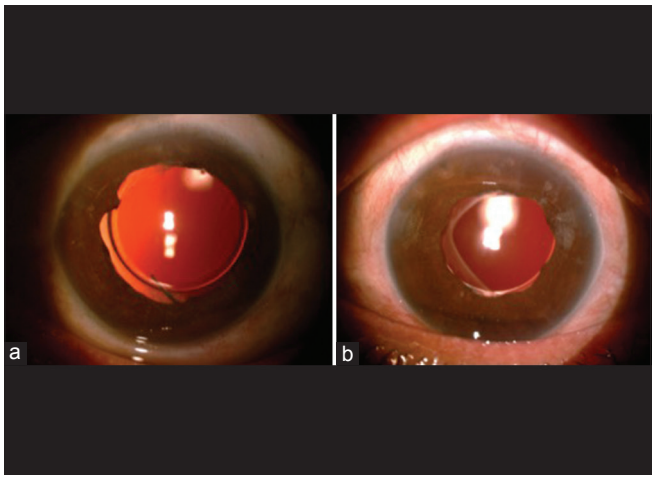

Figure 3: (a and b) Slit-lamp photographs of both eyes at the last follow-up showing a quiet eye within the bag posterior chamber intraocular lens

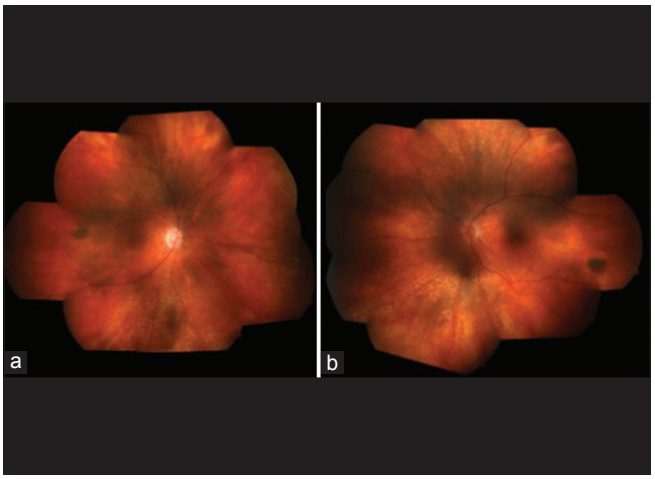

Figure 4: (a and b) Montage fundus photograph of both eyes showing a typical sunset glow appearance and numerous small atrophic and depigmented lesions in peripheral fundus

entity in spite of moderate CD4 counts or systemic status of the patient. Hence, the challenges lie not only in the identification of the disease but also in the judicious use of steroids in the management by close monitoring of CD4 counts, viral loads, and systemic status of the individual under care of a HIV care physician. A disease like VKH may need long-term steroids and/or immunomodulatory therapy and is known for its recurrences.

Medline search did not reveal any report of VKH in a patient with HIV/AIDS, and to our knowledge, this is the first case of VKH in HIV patient being reported. Interestingly, our patient had significant inflammation during the course of the disease and during postoperative stage in spite of moderate CD4 counts. This case represents an uncommon/rare manifestation of an autoimmune disease occurring in a HIV/AIDS patient with significant inflammation in spite of moderate CD4 counts.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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