Photo Essay

Unusual case of vitiligo reversal in Vogt–Koyanagi–Harada syndrome

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Key words: Corticosteroids, multifocal retinal detachment, vitiligo, Vogt–Koyanagi–Harada syndrome

A 26 year old male came to our clinic with sudden onset loss of vision. His presenting visual acuity was counting fingers close to face. Examination of anterior segment of eye revealed non pigmented keratic precipitates with corneal edema, mild anterior chamber reaction with sluggishly reacting pupils. Anterior vitreous face showed 1+ vitreous cells not associated with flare. Fundus examination revealed clear optical media with blurred optic disc and multiple serous retinal detachments involving fovea. Patient had a big vitiligo patch covering nasal and malar area. With a diagnosis of incomplete VKH syndrome, patient was started on high dose corticosteroid therapy (1.5 mg/KBW) to which patient responded favourably. Within a period of 1 week serous detachments found to be reduced and complete resolution was noticed by the end of 1 month. Vitiligo patch also showed signs of re-pigmentation and complete re-pigmentation was observed by the end of 2 months. We presume this is the first case to be reported from this region.

Case Report

A 26-year-old patient came to our clinic with sudden-onset loss of vision in both eyes. It was associated with severe headache and tinnitus. The patient had a history of upper respiratory tract infection associated with fever and headache, for which he was hospitalized for 5 days. His vision dropped to hand movements in the right eye and counting fingers close to face in the left eye. There was a patch of vitiligo along the mid-face extending up to the malar bone of face. No history of ataxia or any other cerebellar signs could be elicited. He had an episode of visual loss 3 years back which was recovered with medical therapy. On clinical evaluation, there was mild circumciliary congestion, nongranulomatous keratic precipitates, anterior chamber showed mild reaction and flare, poorly reacting pupils, and lens was clear. Anterior vitreous face showed 2 + cells, media were clear, and optic disc showed hyperemia with multiple serous elevations along macula in both eyes. Magnetic resonance imaging was done and was found to be within normal limits, cerebrospinal fluid (CSF) analysis revealed marked pleocytosis with lymphocytes 85%, neutrophils 15%, CSF sugar 56 mg%, and CSF proteins 78 mg%. Fundus fluorescein angiogram revealed multiple pinpoint hyperfluorescence which increased in size and intensity in later frames. Optical coherence tomography showed multifocal inner neuroretinal layer separation from outer hyperreflective layer which gradually reduced with treatment and became flat in 1 month [Fig. 1].

With a diagnosis of incomplete Vogt–Koyanagi–Harada (VKH) syndrome was established, we started him on high-dose oral corticosteroid therapy (2 mg/kg body weight) which was tapered and reviewed on weekly basis. However, oral corticosteroid therapy was maintained at 5 mg/day over a period of 6 months.

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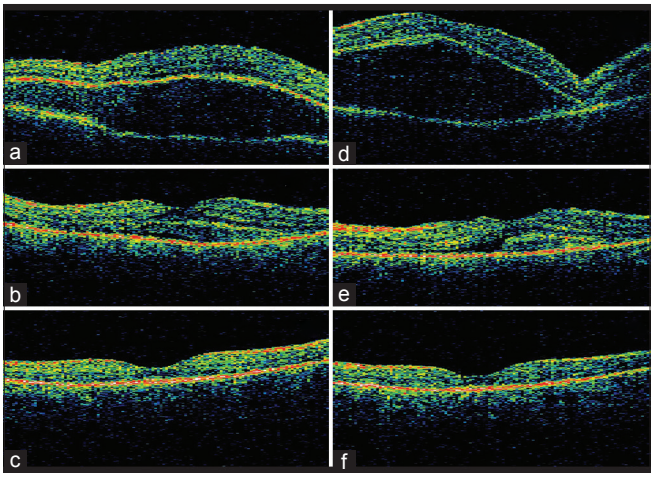

Figure 1: Exudative retinal detachment in optical coherence tomography at various points of follow-up getting reduced with systemic immunosuppressive therapy and attainment of normal macular contour finally. (a-c) Right Eye, (d-f) Left Eye

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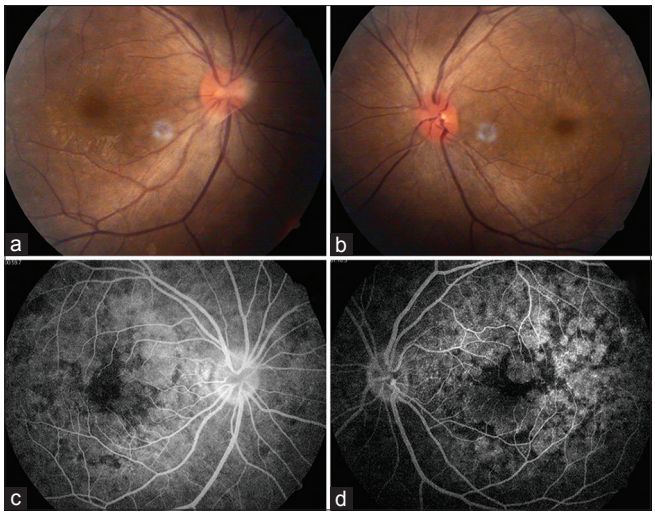

Figure 2: (a and b) Fundus photograph showing bilateral macular elevation with loss of foveal reflex, (c and d) arteriovenous phase of fundus fluorescein angiogram showing multiple hyperfluorescent leaks with hypofluorescent choroidal scars, suggestive of remission of Harada syndrome

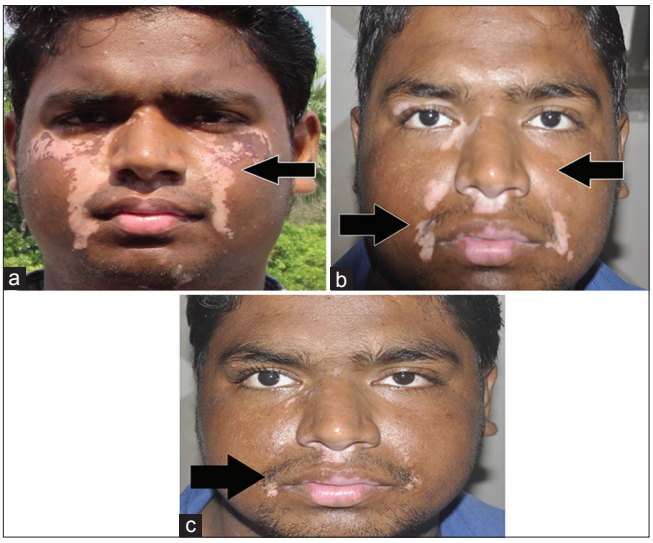

Figure 3: Regimentation of vitiligo patches along the mid-facial area of face. (a) at the time of presentation, (b) at 1 month follow up, (c) at 2 months follow up

On treatment, leaks got reduced, but there was spotted hyperfluorescence due to staining of choroidal scars and hypofluorecscence due to blockage by retinal pigment epithelium (RPE) migration just like salt and pepper fundus; this marked the healing choroidal and RPE inflammation [Fig. 2]. We observed vitiligo patch present over facial aspect which showed signs of repigmentation on treatment. Further, vitiligo reversal was noted over malar area and lateral aspect of nose after 1 month of commencement of treatment, and the process was complete after 2 months of commensal of corticosteroid therapy [Fig. 3].

Discussion

VKH syndrome is still an enigmatic condition to most ophthalmologists. It affects all age groups from 5 to 80 years.[1] The prevalence of disease in uveitis cohorts is 1.4%–3.5% in India.[2] Etiology of the disease is mostly genetic, but the actual mode of inheritance is not yet revealed completely. Integumentary findings such as vitiligo occurs as late clinical manifestation of VKH syndrome; however, recurrence of intraocular inflammation marks simultaneous presence of acute uveitic state and late integumentary findings.[3] Vitiligo is characterized by autoimmune destruction of melanocytes and resulting in the absence of melanin from epidermis. Immunohistochemical analysis has shown marked infiltration of helper T1 cells in epidermis along with increased CD4/CD8 and interleukin 2 (IL-2) expression and amount of expression correlates positively with degree of amelanosis.[4] Immunosuppressive therapy with steroids and various other immunomudulators has found to downregulate various pro-inflammatory cytokines such as IL-2, IL-3, IL-4, IL-5, interferon gamma, tumor necrosis factor alpha, and granulocyte necrosis factors; conversely, it leads to upregulation of IL-10 and thus inhibits unchecked destruction of melanocytes by helper T1 cells.[5] Vitiligo reversal in VKH syndrome following systemic immunosuppression has been mentioned in only one previous study.[6] This paper supports the observation by author of the article aforementioned.

Conclusion

Vitiligo reversal may be considered as favorable prognostic indicator to corticosteroid therapy in VKH syndrome. Repigmentation of vitiligo correlates well with remission of disease process. However, any conclusion is difficult to arrive with a single case.

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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