Bilateral Iris depigmentation, transillumination defects and hypotony in Vogt–Koyanagi–Harada disease

Mohit Garg, Dipankar Das, Harsha Bhattacharjee, Kanika Godani, Madhusmita Mahapatra, Riddhi Raichura

Key words: Iris, uveitis, vogt koyanagi harada syndrome

Vogt–Koyanagi–Harada (VKH) disease is a potentially sight-threatening condition that affects the pigmented tissues of the body. The disease is diagnosed based on meticulous anterior and posterior segment examination augmented by the use of investigations. But several cases are misdiagnosed as anterior uveitis leading to inadequate management. Such patients have a very grave prognosis and not much can be done at that stage for their benefit. We report such a case which was inadequately managed, eventually leading to irreversible vision loss. We also outline rarely documented findings in a case of late presentation of VKH, namely, iris depigmentation and transillumination defects with ocular hypotony.

A 32-year-old female presented to the clinic with complaints of gradual progressive diminution of vision in both eyes for the past five years. She had been diagnosed with chronic anterior uveitis elsewhere and was treated with multiple courses of topical steroids over the past years. The patient also gave a history of frequent severe headaches one year back treated as migraine. On presentation, her visual acuity was hand movement in the right eye (OD) and no perception of light in the left eye (OS). Intraocular pressure by Goldmann’s applanation tonometry was 14 mmHg in OD and 12 mmHg in OS. Slit-lamp biomicroscopy revealed complicated cataract with posterior synechiae in both eyes, with mutton-fat keratic precipitates and pigments on the lens capsule. In the left eye, there was neovascularization of the iris. Gonioscopy revealed closed angles with no angle structures visible. B-scan ultrasonography was advised which revealed increased choroidal thickness in both eyes (OU). The patient was advised cataract surgery in the right eye under guarded visual prognosis. Synechiolysis with cataract surgery and intraocular lens (IOL) implantation was done for the right eye and the visual acuity improved to 6/24 after 1 month. The patient then reported after a year. Posterior segment evaluation revealed a normal-looking posterior pole with vitreous cells. The anterior segment examination revealed peripheral iris atrophy and transillumination defects in OU [Fig. 1] with mutton-fat keratic precipitates OU [Fig. 2]. IOP was 8 mmHg in the right eye and 7 mmHg in the left eye. A diagnosis of progressive iris atrophy OU was added to the previous diagnosis. The patient was started on topical steroids and asked to review back after two weeks. The patient was lost to follow up and reported after another year with complaints of diminution of vision in the right eye. The visual acuity was counting fingers in OD and no reception of light in OS. IOP was 7 mmHg in OD and 8 mmHg in OS. Anterior segment evaluation revealed peripheral iris depigmentation with transillumination defects OU. There were also mutton-fat keratin precipitates in OU. Posterior segment evaluation of the...
Figure 1: Slit-lamp biomicroscopy image showing peripheral iris depigmentation (black arrow) and mutton-fat keratic precipitates (white arrow)

right eye revealed vitreous haze with a sunset glow appearance. B-scan ultrasound was done to measure the retina-choroidal thickness which was 2.05 mm in OD and 2.55 mm in OS. Indocyanine green angiography and fundus fluorescence angiography were planned but could not be done due to media haze. All these signs prompted the treating physician to make a diagnosis of Vogt–Koyanagi–Harada disease and the patient was started on systemic steroids in tapering dose.

Discussion

Very few cases have been reported in the literature of patients with VKH disease presenting with bilateral iris depigmentation and hypotony. All these reported cases were inadequately treated for the disease in the acute phase. The case we reported was also not diagnosed as VKH previously and was never treated with oral steroids and immunosuppressants.

The primary ocular manifestation of VKH is diffuse thickening of the uveal tract caused by non-granulomatous inflammation, more commonly involving the posterior uveal tract. Matsuda et al. demonstrated a close relationship of lymphocytes and melanocytes in VKH disease. The stromal inflammation of long duration is postulated to cause eventual stromal atrophy and transillumination defects. As the iris root is more aggressively involved in inflammation, the depigmentation begins peripherally.

Ocular hypotension is postulated to be due to the involvement of both pigmented and non-pigmented ciliary epithelium during the acute phase. This eventually leads to atrophy leading to ocular hypotension in the patients.

In this case, the patient was earlier diagnosed as a case of chronic anterior uveitis and complicated cataract OU. The patient received topical steroid treatment on multiple occasions without detailed posterior segment documentation and investigations. The diagnosis was delayed, leading to under-treatment and very little could be done to preserve the vision of the patient at this stage. It was only after a uvea specialist was consulted that a diagnosis of VKH disease was made and appropriate therapy was initiated.

Even though diagnostic ambiguity is encountered rather frequently in medical science, a detailed clinical examination with relevant investigations can help accurately diagnose the disease. VKH has shown to have a very good visual prognosis when treated aggressively with systemic steroids or immunomodulatory therapy in the acute stage. When inadequately treated or misdiagnosed, irreversible damage to the photoreceptors occurs and the patients have a grave visual prognosis. In this case, if a detailed posterior segment examination would have been done by the comprehensive ophthalmologist with an early referral to a uveitis clinic, the prognosis would not have been so grave. Counselling about the disease process and follow-up schedule is an indispensable part of treatment and can prevent treatment failure due to loss of follow up.

Peripheral iris depigmentation and atrophy with ocular hypotension are late presenting features of VKH disease. A careful slit-lamp examination and fundoscopy must be done to rule out VKH disease. Proper diagnosis and prompt referral...
followed by aggressive immunomodulatory therapy can help preserve the visual acuity of such patients.

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.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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