Vogt Koyanagi Harada – A Case Report

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Introduction

VKH is characterized by severe bilateral panuveitis with serous retinal detachment with or without signs of meningeal irritation and auditory disturbances, also called uveomeningitic syndrome. VKH is difficult to diagnose and requires an astute clinician to tie together the seemingly unrelated signs and symptoms. There is no single test to make the diagnosis, and hence the diagnosis is purely clinical.

Case Report

A 53 yr old woman presented with bilateral diminution of vision of one month duration. Visual loss was progressive, rapid and more pronounced in left eye than right. She complained of floaters in both eyes since 1 week. There was no history of ocular pain or redness, nor was there any history of ocular trauma or surgery. There was no history of diabetes, however she was a known hypertensive on treatment.

General examination showed areas of hypopigmentation over face, around lips, arms, and fingers (Fig. 1 and 2). The right eye had a visual acuity of 6/24 improving with pinhole to 6/12. The vision in her left eye was 2/60 not improving further with pin hole.

Ocular Findings

Anterior segment examination of the right eye was normal except for poliosis on eyelashes.

Fundus examination showed the Media to be relatively clear. Retina appeared elevated inferior to the disc involving macula suggestive of exudative retinal detachment (Fig. 3a).

Optical coherence tomography showed serous retinal detachment (Fig. 3b).

Anterior Segment examination of left eye, showed 2+ flare and 2+ cells, posterior synechiae at 12 clock and 6 clock and cataractous changes in the lens. The vitreous showed 3+ cells. (Fig. 4a)

Posterior segment evaluation of left eye showed 2+ vitreous opacities (Fig. 4b). Details could not be visualized due to hazy media. Intraocular pressure was normal in both eyes.

Ultrasound B scan of right eye showed choroidal thickening as well as localized retinal detachment. Left eye showed opacities in the vitreous.

A detailed general examination was performed which was normal.

ESR was 54mm/hr. and CSF study was also normal. Audiometry showed bilateral sensorineural hearing loss (Fig. 6)

Based on clinical criteria in International uveitis study, the diagnosis of Vogt Koyanagi Harada Syndrome was made.

She was managed with systemic steroids. A high dose of IV Methyl prednisolone 1 gm /day was administered for 3 days. On the third day she developed glucose intolerance and was treated with insulin and continued on oral prednisolone 60 mg x 3 months along with topical steroids, non steroid anti inflammatory drops and cycloplegics in the left eye.

On follow up after one week, vision improved in right eye to 6/12 (PH 6/9) and in left eye 2/60 (PH 6/18). Retinal examination of right eye showed a remarkable reduction in exudative retinal detachment. Vitreous opacities had cleared in left eye enabling the view of disc and vessels.

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On follow up after 1 month, vision in right eye improved to 6/6 and left eye to 6/18.
Retinal examination in both eyes showed clear media with multiple areas of RPE defects. Exudative RD had resolved in the right eye.
OCT findings confirmed the same (Fig 7a & b).
She presented with a relapse after 4 months with diminution of vision this time more pronounced in right eye. vision in right eye was 3/60 (PH 6/18) and that in the left eye 6/18 (PH 6/12). Retinal examination of right eye showed vitreous opacities and fundus finding suggestive of exudative retinal detachment (Fig. 8).
FA done showed pinpoint leaks and pooling of dye in late phase in right eye (Fig. 9 a, b, c). Left eye showed leakage of dye from disc
OCT confirmed the findings (Fig. 10a, b)
She was managed with oral prednisolone 40 gm/day and Tab azathioprine 50 mg BD and is being followed up.

Discussion
VKH is an autoimmune process in which the immune system mistakenly attacks one's own tissues.
Because of varied clinical manifestations the American Uveitis society adopted the following diagnostic criteria:
1. No history of ocular trauma or surgery
2. At least 3 out of 4 criteria
   a. Bilateral chronic iridocyclitis
   b. Posterior uveitis –multifocal exudative retinal and RPE detachments and disc hyperaemia
   c. Neurological signs of tinnitus, neck stiffness, cranial nerve or CNS dysfunction
   d. Cutaneous findings of alopecia, poliosis or vitiligo
VKH presents with diverse manifestations categorized by prodromal, uveitic, convalescent and recurrent phase.
Fluorescien angiography, ultrasonography, lumbar puncture and other studies substantiate the diagnosis especially in atypical cases.
Fluorescien angiography is very characteristic and reveals hyperfluorescence at the level of RPE with pooling of the dye into subpigment epithelial or subretinal spaces delineating the serous retinal detachment.
The goal of therapy is to suppress the initial inflammation with early aggressive use of systemic corticosteroids followed by a slow taper over 3 – 6 months. This results in rapid recovery of vision however full recovery of vision is not likely because of secondary side effects like cararact. Cytotoxic agents are preferred when steroids are contraindicated. Cases with prominent CNS feature and in cases resistant to steroids, intravenous immunoglobulins are used.
Patients adequately treated with high dose steroids have fair visual prognosis with nearly 2/3 of them retaining 20/40 or better vision.
Fig. 3a. Fundus photograph of right eye showing inferior exudative retinal detachment

Fig. 3b. OCT of right eye showed a serious retinal detachment

Fig. 4a. Slit lamp examination of left eye showed 2+ flare and cells with a posterior synechiae at 12 O’clock and a cataractous lens.

Fig. 4b. Slit lamp biomicroscopic evaluation of the vitreous cavity of left eye showing a cataractous lens and plenty of vitreous opacities.

Fig. 5a & b Scan ultra sonogram of both eyes show diffuse choroidal thickening, localized retinal detachments and vitreous opacities

Fig. 6. Audiometry reading: Pure tone audiogram showing bilateral sensorineural deafness.
Fig. 7a and b. OCT evaluation in both eyes at 1 month follow up showed resolution of Retinal detachment and a normal retinal contour.

Fig. 8 Fundus photograph of right eye taken during a relapse 4 months after the initial episode showing exudative retinal detachment.

Fig. 9 a,b & c Fluorescin fundus angiography during the recurrence showing pinpoint leaks with progressively increasing leakage and late pooling of days.
Figure 10a & b  OCT during recurrence showing presence of exudative retinal detachment in both eyes

References