Vogt, Koyanagi, Harada (VKH) Syndrome

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A case of Vogt, Koyanagi, Harada (VKH) syndrome is reported. A thirty six year old, housewife presented with diminished vision and headache for three months. She was found to have bilateral panuveitis with exudative retinal detachment, reducing her vision to light perception in each eye. She was treated with systemic and topical corticosteroids. Her retinal detachment resolved completely with restoration of vision to 6/9 in each eye.

INTRODUCTION

Alfred Vogt described a condition of chronic bilateral uveitis with premature whitening of hair and eye lashes, in 1906. Yoshizo Koyanagi in 1929 described exudative retinal detachment associated with vitiligo, alopecia, deafness and tinnitus. Enoshuke Harada in 1936 separately described exudative retinal detachment associated with bilateral exudative uveitis and cerebrospinal fluid (CSP) pleocytosis.

It was nearly two decades later when Bruno and McPherson in 1949 suggested that the clinical conditions described by Vogt, Koyanagi and Harada were basically the manifestations of the same syndrome complex and should be called as Vogt, Koyanagi, Harada Syndrome.1 We report a case of Vogt, Koyanagi, Harada (VKH) syndrome for the first time from Nepal.

CASE REPORT

A 36 years Hindu housewife from Dolkha district presented at Tribhuvan University, Teaching Hospital on 10th of August 1989 with history of ocular pain, redness and progressive diminution of vision in both eyes for last three months, more marked for the last two weeks. She had headache for three months and also complained of nausea with occasional vomiting and giddiness. The patient was not a known diabetic hypertensive. There was no history of skin or hair depigmentation. She had not missed her periods. There was no history of ocular trauma or surgery. The patient had resorted...

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frequent analgesic self medication for relief of headache. She was receiving topical corticosteroid and atropine since two weeks before presentation.

On examination the patient was able to perceive light with either eye and had faulty projection of light in either eye. Her intracocular pressure was 8 mm of Hg by applanation in both eyes. Circumcorneal congestion was present in both eyes. Cornea appeared normal except for multiple fine to medium sized keratic precipitates bilaterally; there were flare ++ and cells +++ in anterior chamber of both eyes. Iris was normal in colour and pattern and pupils were dilated and non-responsive to light (atropinised). Lenses were clear bilaterally and vitreous of both eyes showed cells ++++, more so posteriorly.

Fundus examination of the patient revealed hyperaemic, edematous discs with blurred margins bilaterally. Retinal veins were dilated. There was total retinal detachment with large bullae inferiorly in both eyes (Figure 1).

Figure 1

Fundus on presentation

Systemic examination did not show any evidence of generalised or localised edema. There was no evidence of alopecia, vitiligo or poliosis. She was normotensive with blood pressure recording of 110/80 mm Hg. Her hemoglobin was 13 G%. Total and differential white blood cells counts were within normal limits. Erythrocyte sedimentation rate was raised to 70 mm in the first hour. Rheumatoid factor and LE cells in the blood were negative. Serum proteins were found to be 8.5 G/litre and serum calcium 2.2 m equivalent/litre. Fluorescent Treponema antibody adsorption (FTA-ABS) was negative. Cerebrospinal fluid examination showed total cell count of 36 cells per cubic millimeter with 10% polymorphs and 90% lymphocytes, protein of 1.8 G/litre and chloride 120 m equivalent/litre. Her chest and skull X-ray were within normal limits. She was found to have neurosensory hearing loss on audiogram.

Fluorescein fundus angiography of the patient showed subretinal pooling of the dye and leakage from the disc (Fig. 2).

Figure 2a & b

Fluorescein Angiography

Vogt, Koyanagi, Harada syndrome was diagnosed because of presence of bilateral panuveitis with exudative retinal detachment in a woman of oriental stock.

The patient was started on tab. prednisolone 90 mg orally alternate days on 14th of August 1989. In addition, topical corticosteroid and atropine were continued. prednisolone was increased to 120 mg every other day on 18/8/1989, in view of non-
improvement.

On 26/8/1989 the patient was able to count fingers close to her face with either eye and retinal detachment appeared shallower in both eyes.

On 31/8/1989 she was able to count fingers at 1 meter with right eye and at 1.5 meters with the left. Her intracranial pressure was 14 mm Hg in either eye. Anterior chamber showed flare but no cells, in both eyes. Retinal detachment had settled further leaving behind a shallow detachment inferiorly in both eyes. She was free from headache.

The patient was allowed home on 31/8/1989 on prednisolone 120 mg alternate days, topical betamethasone 2 hrly and topical atropine 1% thrice daily. She failed to turn up for an early follow-up as advised and discontinued treatment a week after discharge from hospital.

Fundus examination revealed mild disc pallor, normal vessels with multiple pigmenetary mottling in both eyes (Figure 3). Fluorescein angiography repeated 3 months after the first one revealed multiple pigment epithelial changes in the form of window defects in both eyes (Fig. 4).

![Figure 4](image)

**Fluorescein angiography after 3 months**

Following readmission to the hospital she was put on tab. prednisolone 100 mg every alternate day. In addition topical betamethasone 2 hourly and atropine 1% thrice daily were also started for both eyes.

While still in the hospital her vision dropped to 6/18 in either eye and fundoscopy revealed shallow detachment of retina in both eyes. Prednisolone was increased to 120 mg every alternate day. After two intakes of increased dose of prednisolone her vision was noted to be 6/9 P OD and 6/12 P OS. Anterior chamber reaction was less intense and keratic precipitates appeared flattening. Prednisolone was tapered to 100 mg alternate days. On 28/12/1989 Kps were absent in both eyes and there was minimal flare and no cells in anterior chamber with cells one plus in vitreous of both eyes. Fundus examination showed mildly pig disc with multiple pigmentary changes. The patient was discharged on 31/12/1989 with tapering doses of systemic and topical steroids. Her vision at the time of discharge was 6/9 in each eye.

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DISCUSSION

Vogt-Koyanagi-Harada (VKH) syndrome is more commonly seen in yellow oriental races between 30 to 50 years of age. There is bilateral chronic uveitis simultaneously. Where Harada's disease is predominant, the onset is usually sudden with serious progressive diminution of vision. In typical cases there is exudative retinitis first with vitreous opacities and exudative iridocyclitis with occasional retinal haemorrhages. Retinal detachment follows exudative retinitis leading to profound visual loss. Disc may be hyperaemisic and occasionally edematous.

Meningeal involvement is manifested by headache, nausea, vomiting, increased CSF pressure and pleocytosis of CSF.

Alopecia areata and poliosis are seen in 90% of the cases and vitiligo and temporary deafness in about 50% of the cases. Skin and hair changes start 3 weeks to 3 months after the onset of ocular symptoms. Hair become normal in 5 to 6 months in many cases. Vitiligo and deafness develop with ocular symptoms or soon after and last for 4 to 6 weeks. There may be nerve deafness of both high and low frequency on audiogram.

In about 30% of the cases there is complete visual recovery following resolution of the disease. Uveitis subsides, retinal detachment settles leaving irregular pigmented changes in the fundus. Yellowish subretinal nodules, Dalen-Fuchs nodules are hall mark of V.K.H. Sympathetic ophthalmia and sometimes white strands of scar tissue are left in choroid.

Permanent visual loss may result due to plastic iridocyclitis, secondary glaucoma due to occlusion of saccus pupillae, and phthisis bulbi. Secondary cataract may also develop. Following criteria for diagnosis of V.K.H. have been laid down by American Uveitis Society.

1. The patient should have no past history of ocular trauma or surgery.

2. One finding from at least three of the following groups of signs should be present.
   a. Bilateral chronic iridocyclitis.
   b. Posterior uveitis including exudative retinal detachment, forme fruste of exudative retinal detachment (disc hyperaemia or edema, subretinal macular edema) and 'sunset glow' fundus.
   c. Neurological signs of tinnitus, neck stiffness, cranial nerve or central nervous system symptoms or C.S.F. pleocytosis.
   d. Cutaneous findings such as alopecia, poliosis or vitiligo.

In the case reported, skin and hair changes were absent. However typical ocular findings being defect and manifestations of meningeal involvement were sufficiently convincing to make diagnosis of V.K.H. In difficult cases extracocular manifestations and C.S.F. pleocytosis help to differentiate V.K.H. from sympathetic ophthalmia, extracocular manifestation being rare in latter.

CSF analysis is also helpful to differentiate ocular changes of toxemia of pregnancy from V.K.H. where toxemia of pregnancy is attributed for headache, vomiting and exudative retinal detachment.

Steroids remain the mainstay in the treatment of V.K.H. Antiinflammatory have been used in resistant cases. Recurrence and chronicity in V.K.H. syndrome are well known. In our case too we found recurrence after the patient discontinued therapy. Recurrence can take place even when the steroid therapy is being tapered. Interestingly we found that our patient developed retinal detachment despite being on systemic steroids, though there was no detachment prior to reinstitution of systemic steroids.

The fact that she developed detachment inspite continued systemic and topical steroids may be due to inadequate dose of systemic steroids. Hence the saying 'steroids should be used soon enough, adequate enough and long enough to treat the case of V.K.H. syndrome'.
are borne out by our case.

A good and long follow-up in the case of V.K.H. syndrome can not be over emphasised as each recurrence may leave some permanent affect on the eye.

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REFERENCES


Oral Immunization Against Typhoid Fever in Indonesia with Ty21a Vaccine

When tested under conditions of moderate transmission of typhoid fever, a liquid formulation of the oral typhoid fever vaccine Ty21a had a protective efficacy of 96% in Egypt, and an enteric coated capsule formulation had an efficacy of 67% in Chile. We compared the two formulations under conditions of intense transmission of typhoid fever in Indonesia in a randomized, double-blind trial. 20,543 subjects (age range 3-44 years) received either three doses of enteric coated capsules containing placebo or live Ty21a reconstituted with phosphate buffer.

During 30 months of follow-up, the rate of blood-culture-positive typhoid fever among controls was 810/100,000 per year. Rates of typhoid fever were 379/100,000 per year for subjects who received the liquid formulation of vaccine and 468/100,000 per year for subjects who received enteric coated capsules. The protective efficacies of the liquid and enteric coated formulations were 53% and 42%, respectively. Neither formulation protected against infection with Salmonella paratyphi A. No major side-effects were noted, but the overall incidence of side effects was greater in the vaccine groups. Under conditions of intense transmission, Ty21a protected against typhoid fever; however, because Ty21a will not protect all individuals, there is a need for additional approaches to prevent the disease.