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THE CLINICAL SPECTRUM OF VOGT-KOYANAGI-HARADA SYNDROME IN A TERTIARY CARE CENTER OF KUMAON REGION OF UTTARAKHAND

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ABSTRACT

Aim

To report the clinical spectrum, management, and outcome in typical cases of Vogt-Koyanagi-Harada Syndrome (VKH) in a tertiary care center in kumaun region of uttarakhand.

Materials and methods

It is a retrospective study of VKH patients attending OPD of department of ophthalmology G.M.C.Haldwani ,Uttarakhand. The diagnosis of VKH disease was established by "The revised Diagnostic Criteria for VKH disease established at the First international Workshop on Vogt-Koyanagi-Harada disease".

Results

58 eyes of 30 patients (age range 24-64 years) were included. The classification of the cases with regard to VKH disease was as follows: 3 eyes were classified as complete, 20 eyes as incomplete, and 35 eyes as probable VKH disease. Commonest extraocular manifestation noted in 17 cases was headache. The more common ocular presentations were vitritis (53 eyes) and exudative retinal detachment in the posterior pole (51 eyes), disc hyeperemia(37eyes) . All patients were managed on systemic steroids, and few with systemic immunosuppressive therapy. Majority of the participants had good visual outcome.

Conclusion

Early recognition and aggressive treatment of VKH disease result in good visual outcome in typical VKH cases.

KEYWORDS : Vogt-Koyanagi-Harada disease, Exudative retinal detachment, Uveitis.

INTRODUCTION

Vogt-Koyanagi-Harada (VKH) disease is defined as a bilateral granulomatous panuveitis with or without extraocular manifestations affecting young adults.

Originally VKH disease was classified as two separate entities: Vogt-Koyanagi syndrome , characterized by chronic severe anterior uveitis, alopecia, poliosis, cutaneous as well as perilimbal vitiligo (also known as Sugiura's sign), and dysacusia1,2,3. Harada's disease, characterized by bilateral exudative uveitis accompanied by pleocytosis of cerebrospinal fluid1,3

Since there is much overlapping of signs and symptoms between the two entities, in 1932 Babel suggested to call the entity Vogt-Koyanagi-Harada disease.

The incidence of VKH will vary depending on the geographic location and the ethnicity encountered. The disease primarily affects Hispanics, Japanese and heavily pigmented races, but interestingly the disease is rare among Sub-Saharan Africans, indicating that the amount of skin pigmentation alone is not the sole etiologic factor in the pathogenesis of VKH syndrome.4 Identification of association with major histocompatibility complex class II antigens, strongly

supports an underlying genetic predisposition in the pathogenesis of the disorder. There is an association with HLA-DR1 and HLA-DR4 (subtype 0405).5,6

The disease appears to affect women more frequently than man, but no specific sex predilection has been established.

In Japan it accounts for 6.8% to 9.2% of uveitis cases7,8, meanwhile in the United States it hovers around 1%-4%. The majority of the cases found are around the second and fifth decades of life. Women have been reported as being more affected than men; but this will vary depending on the population studied.

The aim of this study is to analyze demographics, clinical characteristics treatment response in VKH patients referred to a tertiary eye care center at Kumaon Region uttarakhand.

Materials and Methods

It is a retrospective study of VKH patients attending OPD of department of ophthalmology G.M.C.Haldwani ,Uttarakhand. The clinical data of 30 patients diagnosed with VKH were reviewed for patient demographics, clinical signs, investigations, treatment received, and visual outcome. All patients underwent a complete ophthalmic examination, including measurement of best corrected visual acuity (BCVA), slit-lamp examination, applanation tonometry, dilated fundus examination with scleral indentation, colour fundus photography, and ocular ultrasonography (if the view of the fundus was not clear). External examination evaluated the presence of poliosis and vitiligo.

The diagnosis of VKH disease was made according to the clinical presentations and it was confirmed by fundus fluorescein angiography (FFA) in all cases. Additional investigations such as B-scan ultrasonography and spectral domain optical coherence tomography was done in selected cases. A history of trauma was ruled out in all cases. Investigations were done to rule out tuberculosis (Mantoux skin hypersensitivity test, PA view of chest X-ray) and sarcoidosis (serum angiotensin converting enzyme).

We used revised diagnostic criteria for Vogt-Koyanagi-Harada disease by Read et al. for classifying the study participants into complete, incomplete, and probable VKH9. All patients with associated diseases such as tuberculosis, sarcoidosis, syphilis or trauma were excluded. All the patients were treated with systemic corticosteroid therapy. Topical corticosteroids and immunosuppressive therapy were used in selected cases. Statistical analysis was done using the SPSS version 12 software.

Results

Thirty patients were included in the study. Age group of the subjects ranged between 24-64 years (mean 41.7 years). 18 patients were male and 12 patients were female. 28(93%) patients had bilateral presentation and 2(7%) patient had unilateral VKH. The clinical type

of VKH disease was classified as complete in 3 patients, incomplete in 20 patients, and probable in 12 patients.

Clinical presentation was posterior uveitis in 20 eyes and panuveitis in 38 eyes. 26 eyes (45%) had acute VKH disease, 20 eyes (36%) had chronic presentation, and 12 eyes (19%) had recurrent uveitis. (Table 1)

Diminution of vision and headache and were the commonest acute presentations followed by integumentary changes in chronic cases. Sensorineural hearing loss was present in 2 cases (Table 2).. The ocular findings of slit-lamp examination were circumciliary congestion in 30 eyes (53%), keratic precipitates in 17 eyes (30 %) with the presence of anterior chamber flare and cells (Table 3). Fundus examination revealed vitritis in 53 eyes (92%), optic disc edema involvement in 40 eyes (69%), exudative retinal detachment in 51 eyes (88%),(Table 3). FFA was done in all patients, which revealed multiple hyperfluorescent dots at the level of retinal pigment epithelium, optic disc leakage in and late accumulation of fluorescein in the subretinal space. Low- to medium- reflective diffuse thickening of the choroid was demonstrated by B scan ultrasonography and was most marked in the juxtapapillary region. Optical coherence tomography (OCT) findings in VKH in active state were multi-lobular serous retinal detachments, intra-retinal edema, subretinal septae

Table 1: Demographic and clinical characteristics of VKH patients

Demographic variables	No. (%)
Mean Age (yrs)	41.7(24- 64)
Gender	
Male	18(60%)
Female	12 (40%)
Laterality	
Bilateral	28(93%)
unilateral	2(7%)
Anatomical classification	
Panuveitis	38 eyes (65.5%)
Posterior	20eyes (34.4%)
Clinical types	
Probable	35 eyes (60.8%)
Incomplete	20 eyes (35%)
Complete	3 eyes (5%)
Course	
Acute	26 eyes (45%)
Chronic	20 eyes (36%)
Recurrent	12 eyes (19%)

Table 2: Incidence of various systemic manifestations

Systemic manifestations	No. (%)
Meningismus	12(40%)
Headache	17(58%)
Integumentary changes	1(5%)
Tinnitus	4(15%)
Sensoryneural Haring loss	2 (6%)

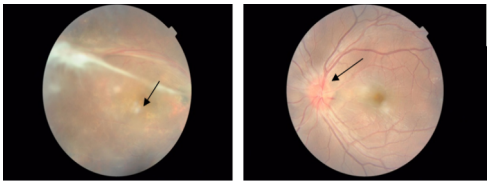
Table 3: Clinical findings

Anterior segment manifestations	No. (%)
Circumciliary congestion	30 eyes (53%)
Keratic precipitates (KP)	17eyes(30%)
Flare	39 eyes (68%)
Cells	40 eyes(70%)
Posterior segment manifestations	No. (%)
Vitritis	53eyes(92%)
Disc hyperemia	37eyes (64%)
Disc edema	40 eyes (69)
Retinal detachment	51 eyes(88%)

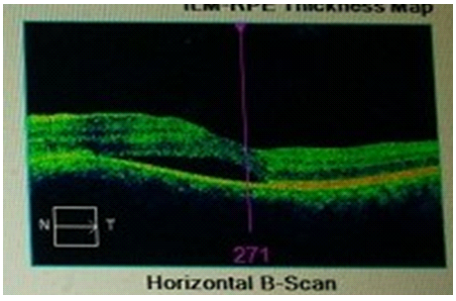
Table 4: Medical management of VKH syndrome

Drugs used	No. (%)
Topical corticosteroids	52eyes (90%)
Oral steroids	30 patients (100)
Intravenous methyl prednisolone	23 patients (76.6%)
Immunosuppressive	2 patients (6%)

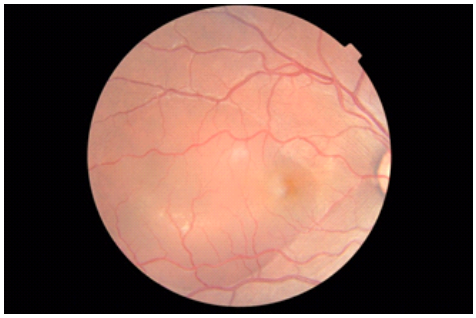
Figures 1- 4 indicate illustrative clinical findings observed in VKH patients



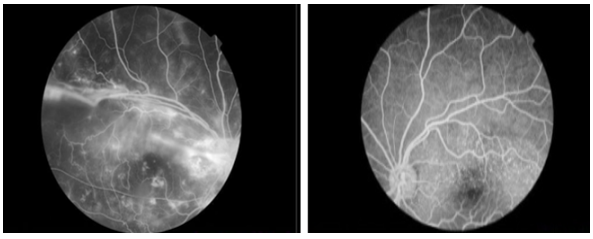
1) A). Serous retinal detachment 1) B). Disc Hyperemia



2)Serous Retinal Detachment on Optical Coherence tomography



3)Multiple serous detachment on fundus picture



4) Fundus Flouroscine angiography showing multiple pin point hyperflouroscent areas with leakage.

DISCUSSION

Vogt-Koyanagi-Harada disease (VKH), a multisystem autoimmune disorder targeting predominantly melanocytes, affects organs such as eye, skin, inner ear and meninges. In the eye, the disease affects the uveal tract, manifesting as granulomatous panuveitis. Acute presentation is characterized by the presence of exudative retinal detachment with preservation of the choriocapillaris. In the chronic stage, the disease is characterized by the presence of Dalen-Fuchs nodules and focal chorioretinal atrophy with loss of retinal pigment epithelium.10 In our study we found that males are more likely to develop VKH disease than females, whereas Soon-Phaik Chee et al. have reported equal distribution in both sexes.11,12,13,14,15

The early and aggressive use of corticosteroids in acute VKH disease prevented the appearance of the complete spectrum of the disease in the current study, as shown by Rajendram et al.15 Systemic steroid therapy was given for longer periods of time along with immunosuppressive therapy.

The diagnosis of VKH disease is based on history and clinical findings with supportive evidence from ancillary tests, including fundus fluorescein angiography, which showed classical presentation such as multiple pin-point hyperfluorescence in the early phase at the level of the RPE, with classical pooling of the dye in the late phase in all acute presentations.

OCT was used when the media was clear to document the changes and B scan was used in all acute cases and also in cases of hazy media, where we could not visualize the fundus to monitor the response to treatment.

All our patients were treated with systemic steroid therapy.

In summary, the clinical features of VKH presented to tertiary care center of kumaun region India are similar to the study except for male predominace of VKH conducted by reema bansal et.al16. Extra ocular manifestations were not noticed in acute cases with aggressive management with systemic steroids and immunosuppressive therapy. Chronic and recurrent cases had extraocular manifestations at the time of presentation in majority of the cases. Early and aggressive therapy resulted in good visual outcome in majority of our patients.

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Competing interests
The authors declare that they have no competing interests.

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