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A Case Report of Probable Vogt Koyanagi Harada Syndrome: A Rare Case

Dr.M.Sahana[1],Dr.M.Harika[2] ,Dr.K.Vikasini[3],Dr.L.J.Sandhya vali[4].
Post graduate, Assistant Professor,Post graduate, Professor, Department of Ophthalmology, Guntur medical college. Guntur, Andhra Pradesh,India.

Abstract:
Vogt Koyanagi syndrome is bilateral granulomatous panuveitis with or without extra ocular manifestation[1]. It is autoimmune reaction against melanocyte containing tissues mediated by cellular immune responses[4]. A 46 year old female came to ophthalmology OPD with chief complaint of gross diminution of vision in both eyes left>right since 1 month sudden in onset rapidly progressive in nature. On detailed ophthalmological and neurological and dermatological examination patient was started on pulse therapy of IV steroids which showed improvement in vision followed by tapering of oral steroids with initiation of non steroidal immunosuppressive agents.

Key Words: *Vogt koyanagi harada syndrome, steroids*

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I. Introduction

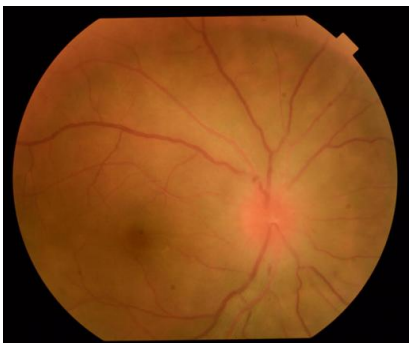
Vogt Koyanagi syndrome is bilateral granulomatous panuveitis with or without extra ocular manifestation.Vogt koyanagi syndrome characterised by chronic severe anterior uveitis, alopecia,poliosis,perilimbal vitiligo with cutaneous vitiligo and dysacusia [1][2][3].Harada syndrome characterised by bilateral exudative uveitis with pleocytosis of cerebrospinal fluid [1][3].

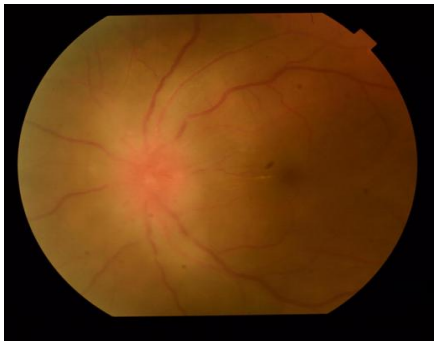
II. Case Report

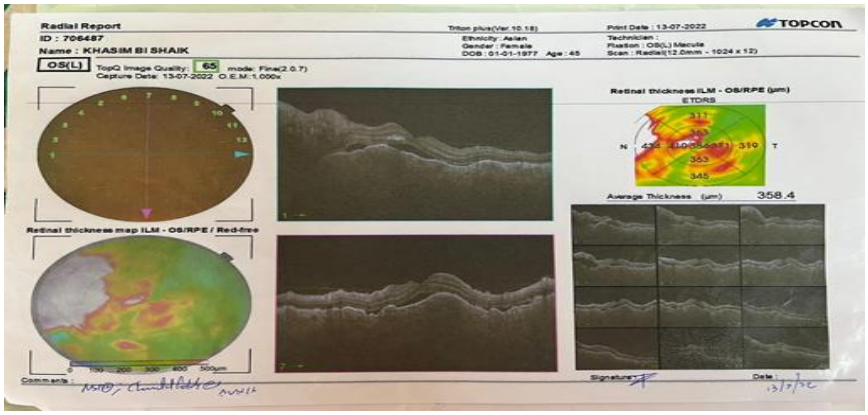
A 46 year old female came to ophthalmology OPD with chief complaint of gross diminution of vision in both eyes [left eye >right eye] since 1 month which is sudden in onset rapidly progressive in nature. Associated with redness & watering in both eyes which resolved on usage of topical medication. Not associated with tinnitus,hearing loss,vertigo,poliosis,alopecia, neck stiffness,fever,limb weakness. No history of intraocular surgery,intraocular injection,trauma.No history of weight loss,joint pains,back pain.No history of chronic usage of any medication

Ocular examination:

BCVA-RE-6/60 ; LE-Counting finger 2mts. Extra ocular movements are free full painless in all gazes in both eyes.On slit lamp examination both eyes showed pharmacological mydriasis with pigments in circular pattern seen on clear lens s/o broken synechiae and remaining details within normal limits .On fundus examination both eyes showing hazy media due to vitritis,optic disc is hyperaemic and edematous, with engorged veins and altered foveal reflex with background showing multiple sensory retinal detachments along superior and inferior arcade.B scan shows vitreous echogenicities with choroidal thickening. On OCT thickened choroidal folds with multiple neurosensory retinal detachments noted. FFA showed multiple pinpoint leaks in early stage with increased hyperfluorescence in late stages.All blood investigations are within normal limits







Patient was treated with IV methyl prednisolone 1gm once daily for 3 days followed by initiation of high dose oral steroids. After one week of oral steroids patient's best corrected vision improved to 6/6 and fundus examination showed decreased vitritis and disc edema and patient was continued on slow tapering of oral steroids and initiation of non steroidal immunosuppressive agent methotrexate 15mg once weekly. Tapering of steroids done for every 2 weeks at rate of 10mg. When patient was on 20mg/day dosage patient experienced blurring of vision with fundus showing presence of active choroiditis suggestive of relapse. So dosage of oral steroids was increased to 40mg/day

III. Discussion

Vogt Koyanagi syndrome is bilateral granulomatous panuveitis with or without extra ocular manifestation. USG will present diffuse choroidal thickening with low to medium reflectivity, serous retinal detachments, vitreous opacities without posterior vitreous detachments and scleral or episcleral thickening [6][7].OCT will demonstrate the presence of subretinal fluid. In the presence of subtle choroidal folds, they will be detected as corrugation of the RPE/choroid with choroidal thickening [2][5]. Multiple septae creating compartments or pockets of fluid in the outer retinal may be seen. Typically the inner retina inward to the external limiting membrane is normal. There is increased choroidal thickness in acute stage.

Diagnostic criteria[4]:

1. No history of penetrating ocular trauma or surgery
2. No clinical or laboratory evidence suggestive of other ocular diseases
3. Bilateral ocular involvement
4. Neurological and auditory findings
5. Integumentary findings-alopecia,poliosis,vitiligo

Complete VKH- all above criteria 1-5 must be present
Incomplete VKH-criteria 1-3 with either 4 or 5 must be present
Probable VKH-isolated ocular disease; criteria 1-3 must be present
In acute phase IV high dose corticosteroids 1g methyl prednisolone for 3 days followed by oral dose of 1-1.5mg/kg/day for minimum of 6 months.Topical steroids and cycloplegic agents are used for anterior

uveitis. Steroid resistant cases may require immunosuppressive agents. Immunosuppressive agents cyclophosphamide, methotrexate, cyclosporine can be used. Biological agents like infliximab,daclizumab,INF-2a can be given as alternative agents for long term therapy

IV. Conclusion

VKH is usually a rare disease entity which may not be always associated with systemic manifestations. So if any case presents with sudden onset bilateral vision loss we should keep in mind that there is possibility of probable VKH syndrome which on prompt management can helps to save the vision and halt the progression of disease. We must follow up the case at regular intervals to look for any relapse during steroid tapering and also after completion of steroids for relapse or recurrence of disease.

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