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OCULAR MANIFESTATIONS IN VOGT KOYANAGI HARADA SYNDROME- A CASE REPORT

Ophthalomology

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ABSTRACT

Vogt Koyanagi Harada syndrome is a central nervous system disease characterized by a loss of immune tolerance to melanocytes in meninges, eyes , skin, hair, ears. It is known to affect vision and hearing. The immune reaction can start following an inciting viral disease. Following this inciting event, there is a T lymphocytes mediated non necrotizing granulomatous inflammation to melanocytes. The incidence of VKH depends on geographic location and ethnicity. It primarily affects pigmented races. Women are more affected than men. A majority of patients are in the second to fifth decade of life.

KEYWORDS

Vogt Koyanagi Harada Syndrome, Granulomatous Panuveitis, Autoimmune Diseases, Vitiligo,alopecia.

INTRODUCTION

Vogt Koyanagi Harada is a relatively rare central nervous system disorder which most commonly affects vision and hearing.

It was first described as an iridocyclitis by Alfred Vogt in 1906, later serous retinal detachment component was added by Yoshizo Koyanagi in 1926, Einosuke Harada added the integumentary symptoms.[1]

The disease presents with symptoms of loss of immune tolerance toward melanocytes in meninges, skin , hair, eyes and ears. Patients are usually in 3rd or 4th decade of life , women are more commonly affected with ratio of 3:2 as compared to men. [2]

It may be associated with DRB1*0405 and/or DRB1*0410 gene.[3]

In prodromal stage the patients present with dizziness, headache, eye pain, fever, photophobia. There may be history of a preceding viral illness.

In acute phase common symptoms are blurring vision and photophobia, this may be due to chorioretinal inflammation, exudative retinal detachment , acute angle closure or papillitis.

In chronic stage , hearing loss, depigmentation of skin, ciliary depigmentation are common manifestations. [1]

Our patient is a 39 year old female , presenting in acute stage with ocular manifestations.

Case Report

A 39 year old female patient presented with pain over Left Eye associated with decreased vision since 1 week. It was also associated with headache. Patient also gave history of weakness of muscles of right hand since 6 months.

Vision assessment with Snellen's chart showed that the vision in left eye was grossly decreased – counting fingers at 2 metres, whereas the right eye vision was normal (6/6). Near vision was grossly decreased in LE. Colour vision was also defective in LE.

On slit lamp examination, there was no evidence of anterior chamber reaction, but a shallow anterior chamber of grade 2 Van Herick's was observed in both eyes. Gonioscopy was done and angles found to be open. Pupillary reaction was found to be sluggish in the left eye, pupillary dilatation was achieved with Tropicamide eye drops. Fundoscopy of the left eye showed disc oedema, macula showed serous detachment, and hypopigmented choroidal lesions in the periphery. An OCT and FFA of both the eyes was performed. OCT of LE showed gross macular oedema with the presence of subretinal fluid. Fundus Fluorescein Angiography of LE displayed evidence of diffuse disc hyperfluorescence with pin point leaks throughout posterior pole , which increase in intensity and size in later frames. Right eye findings were normal. A diagnosis of Vogt Koyanagi Harada syndrome was made and treatment was started with intravenous Methyl prednisolone at 1g/day for 3 days. Electrolytes and blood sugars were regularly monitored, patient was discharged after 3 days with continued oral steroids .Vision improved within 1 week of treatment.

OCT scan of left eye showed presence of subretinal fluid.

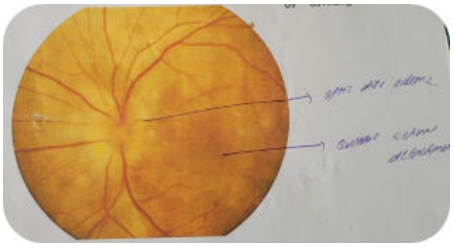


Figure 1 : Fundus Picture Of Left Eye Showing Disc Oedema And Bullous Retinal Detachment At Macula

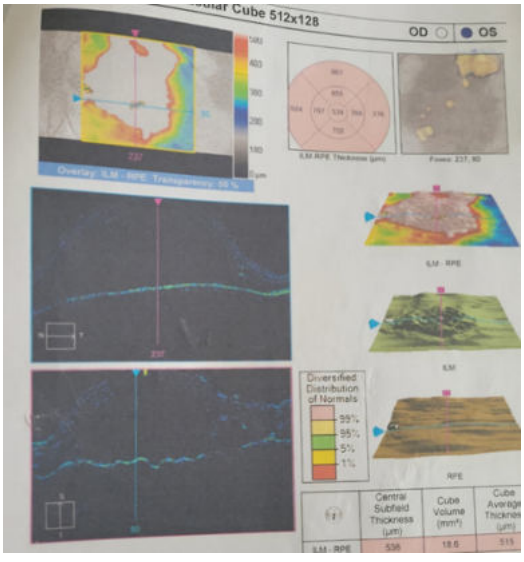
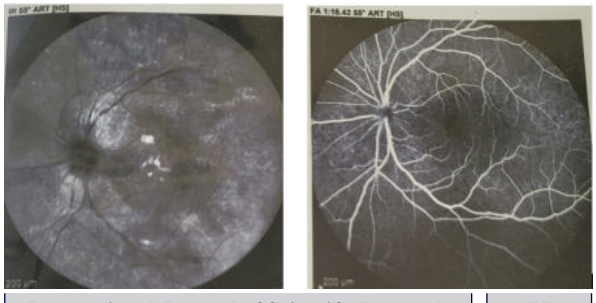


Figure 2 : OCT of left eye showing macular oedema



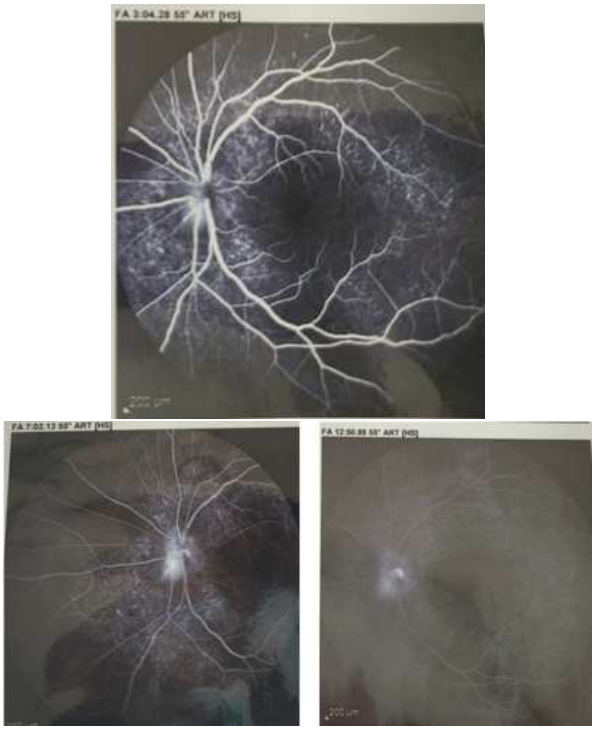


Figure 3 : Eye Showed Diffuse Disc Hyperfluorescence With Pin Point Leaks Throughout Posterior Pole , Which Increase In Intensity And Size

DISCUSSION :

Diagnosis is made if patients have any three signs among the following : bilateral chronic iridocyclitis, posterior uveitis with or without sunset glow fundus, neurological signs such as tinnitus, neck stiffness, nerve palsies, cutaneous signs such as vitiligo, poliosis, alopecia. Complications may be due to ciliary body inflammation presenting as cataract, cystoid macular oedema, acute angle closure glaucoma, choroidal neovascularization, choroidal neovascular membranes, retinal detachment , subretinal fibrosis (especially in paediatric patients) and ocular hypertension. [4]

Patients are treated with intravenous pulse therapy of Methyl Prednisolone (1g/day for 3 days) or oral Prednisolone 1-1.5 mg/kg/day.

Tapering and stopping treatment may incur recurrences. In chronic VKH , immunomodulatory therapy , especially cyclosporine is found to be effective.

Adalimumab is found to be effective in complicated cases and when steroids are ineffective.

Infliximab, methotrexate is useful in paediatric patients. [5] Intravitreal triamcinolone and bevacizumab are used as an adjunct in chronic stages of the disease.[1]

Our patient was treated with oral steroids in tapering dose, and later IV MP at 1g/day for 3 days. This was followed by similar disturbance in Right eye 6 months later.

CONCLUSION :

VKH syndrome requires regular and close monitoring. Early diagnosis with prompt treatment will help in visual recovery. Treatment with corticosteroids with slow tapering after 6 months is found to be associated with better visual recovery, whereas suboptimal dose of steroids can cause persistent inflammation. Better visual prognosis is seen when patient presents early and with good visual acuity.

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