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PRESUMED UNILATERAL VOGT-KOYANAGI-HARADA DISEASE-CASE REPORT OF A RARE CLINICAL ENTITY.

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Abstract :
In this retrospective report, we present a case of unilateral Vogt-Koyanagi-Harada (VKH) disease. This patient was evaluated with clinical, ophthalmological and laboratory examinations. The response following corticosteroid administration was evaluated. She had the characteristic clinical features of VKH involving only one eye, including disc edema, multiple sub retinal yellow lesions and exudative retinal detachment. This case indicates that the clinical and angiographic features were typical of VKH disease despite the unilateral involvement. Reports of unilateral VKH are meagre and we here report one of this rarity.

Keyword :
Fundus Fluorescein Angiography, Ultrasonography, Vogt-Koyanagi-Harada Disease.

Introduction:
Current diagnostic criteria for VKH disease4 requires the presence of bilateral ocular involvement. However, unilateral or delayed involvement of the fellow eye can occur in rare cases3. The diagnosis of VKH disease is essentially clinical and angiographic: exudative retinal detachment during the acute disease is very specific and FA typically reveals numerous punctate hyper fluorescent areas at the level of the retinal pigment epithelium in the early stage of the FFA followed by pooling of dye in the sub retinal space.

Case Presentation: A 50-year-old female (Fig. 1) presented to us with decreased visual acuity (VA) in the left eye (LE) for 1 month duration which was insideous in onset, gradually progressing and painless loss of vision. There was no history of penetrating ocular trauma or surgery. There was no history of headache,loss of consciousness, no history of flu like symptoms in the past, no history of hearing loss and no history of drug intake except the antihypertensive medications. She was hypertensive since past six months and was under antihypertensive medications. At the initial visit, the best corrected visual acuity (BCVA) in the RE was 6/9 and in the LE it was CFCF (counting finger close to face) .

Fig 1: Patient profile

The anterior segment examination of right eye was normal (Fig. 2) with IOP of 20 mm Hg by Goldmann applanation (GAT). Examination of the left eye revealed lids and eyelashes to be normal, Conjunctiva was clear, there were few keratic precipitates on back of cornea, Anterior Chamber revealed cells 1+, flare 1+, Pupil showed grade II RAPD. On dilatation: posterior synechiae and festooned pupil was seen (Fig. 3), Minimal lens changes and few cells (1+) in the anterior Vitreous face were noted. IOP in the LE was 20 mm Hg.(GAT)

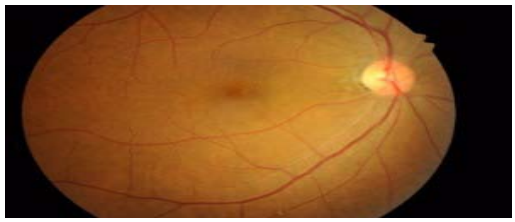

Fig 4:Fundus picture of RE

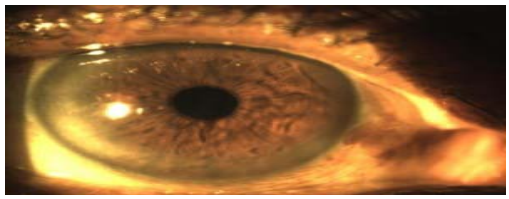

Fig 2: slit lamp picture of RE

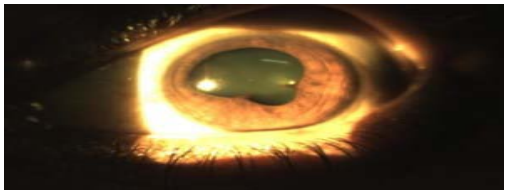

Fig 3: slit lamp picture of LE

Fig 5:Fundus picture of LE

RE fundoscopy was normal(Fig. 4) but fundus examination of the LE showed hyperemia and edema of optic nerve head, multiple subretinal yellow lesions in posterior pole associated with serous detachment(Fig. 5). A Fundus Fluorescein Angiography (FA) revealed angiographic features typical of VKH disease in the LE:, focal areas of delayed choroidal perfusion and multifocal areas of pinpoint leakage with late pooling and leakage of dye in the subretinal space, optic disc hyperfluorescence and its late staini (Fig. 6)

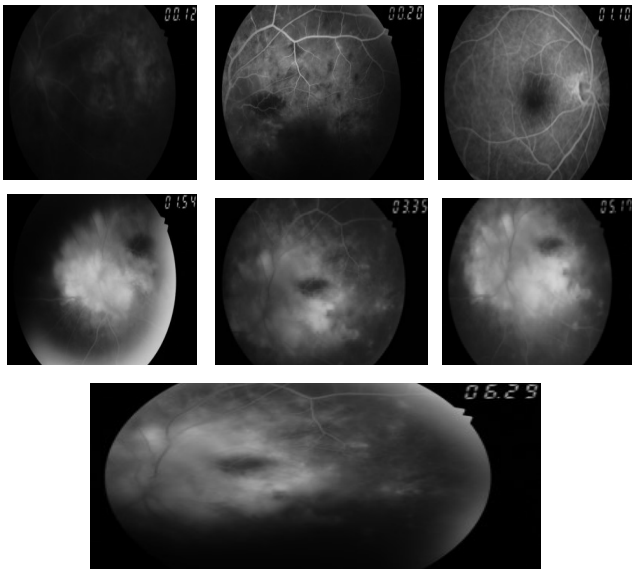

Fig. 6: Serial FFA picture

Optical coherence tomography was done for the patient. The OCT of the RE was normal (Fig. 7) confirming the unilateral involvement of the disease. LE OCT was not possible as the patient was unable to fix at the target.

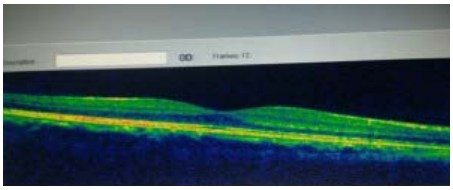

Fig 7: OCT RE

Ultrasonography of LE showed moderate homogenous echoes seen extending in an irregular manner over the ONH and also along the posterior pole, with presence of choroidal thickening and no evidence of posterior scleritis. A battery of investigations were done. Complete hemogram showed the following results:TC- 12,500 cells/cumm, DC -neutrophils 58%, lymphocytes 39%, eosinophils 4%, monocytes 1%.ESR- 24mm/40mm.Chest x ray showed clear lung fields and mantoux was negative. TORCH test was proved to be negative. Usg abdomen was normal. Renal function tests were normal. Serum calcium and ACE level were done and were found to be within normal limit. VDRL and VCTC was also negative. Neuroimaging did not show any significant findings and was found to be normal. Thorough systemic examination revealed a mild sensory neural deafness (Fig.9) and she had hypopigmented lesions over the hand probably vitiligo(Fig. 8). From the clinical, ophthalmological and angiographic picture a diagnosis of presumed unilateral VKH disease was made. The close differential diagnosis3 such as Sympathetic ophthalmitis, Posterior scleritis, Sarcoidosis, APMPPE, Uveal effusion syndrome, Intraocular B cell lymphoma and Cat scratch disease were thought for and their probability were ruled out. Sympathetic ophthalmitis was ruled out as there was no history of penetrating injury in the other eye. There was no severe ocular pain, unilateral T Sign in B Scan so Posterior scleritis was ruled out. Uveal effusion syndrome should present as subacute chronically progressive, relapsing and remitting course with bilateral presentation which was not found in this case. APMPPE presents with sudden loss of central vision following viral prodrome with multiple placoid yellow lesions and early hypo and late staining, which was not so in this case. There was no H/O neurological symptoms, vision was affected, imaging did not show lesions with blocked fluorescence with late staining suggestive of Intraocular B cell lymphoma. Moreover complete hemogram was normal. Sarcoidosis was ruled out as the ACE levels were normal, no hilar lymphadenopathy and fundus did not show any characteristic candle wax drippings.

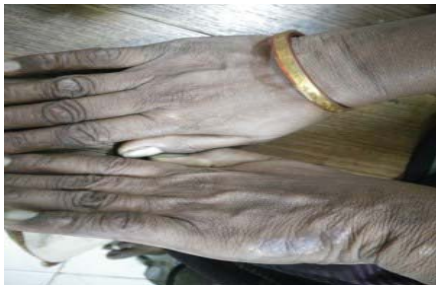

Fig. 9: Hypopigmented patches.

Fig 8 : Audiometry showing SNHL

Patient was treated with Intravenous Methylprednisolone 500mg BD for three days, followed by Tab. Prednisolone 60mg OD and Tablet Ranitidine 150mg BD. Patient is under regular follow up. At the end of 1 week after beginning oral steroids the fundus revealed resorption of sub retinal fluid (Fig. 9), but there was no improvement in the visual acuity. At the end of 1 month, the fundus of LE typically revealed the Sunset Glow fundus (Fig. 10) with a visual acuity of 3/60. The improvement of visual acuity was not marked despite treatment due to the presence of central nummular chorioretinal atrophic scar.

Fig 9: Fundus picture LE 1 week post treatment

Fig 10: Fundus picture LE 1 month post treatment.

Discussion: Current diagnostic criteria for VKH disease require the presence of bilateral ocular involvement; however, unilateral or delayed involvement of the fellow eye can occur in rare cases. The diagnosis of VKH disease is essentially clinical and angiographic: exudative retinal detachment during the acute disease is very specific and FA typically reveals numerous punctate hyperfluorescent areas at the level of the retinal pigment epithelium in the early stage of the exam followed by pooling of dye in the subretinal space. Differential diagnosis includes other causes of pan uveitis, such as sympathetic ophthalmia, uveal effusion syndrome, posterior scleritis, primary intraocular lymphoma, uveal lymphoid infiltration, acute posterior multifocal placoid pigment epitheliopathy and sarcoidosis. VKH disease may be differentiated from other causes by a complete history, review of systems and physical examination associated with a laboratory evaluation. In this case, all laboratory tests were negative and review of systems revealed mild sensorineural deafness and vitiligo, excluding other possible diagnoses. Despite the unilateral involvement, the clinical and angiographic features were typical of VKH disease. 5Systemic steroids are the therapy of this entity in order to control active inflammation and to prevent new inflammatory episodes. They are tapered slowly to prevent recurrence of the disease and to minimize the incidence and severity of extraocular manifestations. However, there are recent studies2 which support the evidence that first-line use of corticosteroid combined with immunosuppressive agents decreases the development of late complications and recurrence of the disease, improves long-term vision and facilitates more rapid tapering of steroids.

Our patient has been receiving systemic corticosteroid therapy for 4 weeks and is on constant follow up. In VKH disease, the 'sunset glow' fundus (depigmentary changes in melanocytes in the choroid) is an important finding that appears between 1 and 6 months after disease onset. The development of LE depigmentary changes in our patient, in comparison to normal fundus pigmentation in the RE, is one more clinical sign that supports the diagnosis of unilateral VKH. There are few reports on unilateral VKH in the literature (less than 10), and some of them are delayed forms of bilateral disease; our report adds one more case to this rare condition, with no clinical signs of the disease in the contralateral eye. Some of the published case reports1had a final BCVA less than 20/20 in the affected eye; our case highlights the importance of a correct diagnosis and treatment. In conclusion, our study presents a rare clinical variant of the VKH disease and enhances the fact that unilateral VKH disease is rare but should be promptly diagnosed and treated.

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