Case Report

A Misleading Presentation of Vogt – koyanaji – Harada Disease

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Abstract

Purpose: To report an unusual clinical presentation of Vogt – koyanaji – Harada disease.

Design and methods: A 36 years-old Iraqi woman presented to Neurology department with chronic headache and bilateral disc edema, neuro-imaging was normal, a provisional diagnosis of idiopathic intracranial hypertension was made, aspiration of cerebrospinal fluid (CSF) was done. The CSF pressure was normal and the patient did not respond to conventional treatment. After 3 weeks, the patient developed drop of vision in the right eye, the patient referred to the Ophthalmology department for evaluation of the visual problem. On examination, vision was counting finger 3 meters right eye and 5 meters left eye. There is bilateral granulomatous uveitis both eyes, bilateral disc edema, and multifocal exudative retinal detachment in both eyes. Optical coherence tomography (OCT), and posterior fundus photography document the presence of multifocal retinal detachment.

Keywords: VKH, optic disc edema, granulomatous.

Introduction

Vogt - Koyanaj- Harada (VKH) disease is a chronic autoimmune systemic disease that target the melanocytes in eye, ear, skin and brain [(1)]. It is commonly affects females between 20-50 years of age. It is more common in Asian, Hispanic and Native American [(2, 3)]. VKH disease forms about 2.2 % of cases referred to uveitis clinic [(4)]. At presentation, the extraocular features forms about 60% and more than 90% of these features is meningism[(4)]. Other systemic features include tinnitus, hearing defects, CSF pleocytosis and vitiligo[(2)].

The commonest ocular presentation is panuveitis (more than 90% of cases)[(5)]. The main target in the posterior part of the eye is the choroid presented clinically as multifocal choroiditis with exudative retinal detachments[(5, 6)]. Optic disc edema is presented as part of posterior segment inflammation[(7)]. Involvement of optic nerve and vestibular nerve are uncommon in the course of the disease[(8)]. Diagnosis of VKH disease diagnosis is mainly clinical depending on the presence of bilateral granulomatous uveitis with exudative retinal detachment documented by optical coherence tomography of retina and fundus fluorescein angiography[(6)].

While the main treatment is systemic and local steroids, other immunosuppressive drugs are used as a long-term regime to control the disease[(5)].

Complicated cataract and glaucoma are the main complication followed by post-inflammation retinal atrophy, optic atrophy, ocular hypotony [(9)].

Case report

A 36 years old lady presented to the Neurology department with severe headache and bilateral optic disc edema considered as papilledema, neuro-imaging was normal. A provisional diagnosis of idiopathic intracranial hypertension was made and the patient given treatment. After 3 weeks, the patient referred to the Ophthalmology department because of drop of vision in the right eye. On examination, the visual acuity was counting fingers 3 meters in the right eye and 5 meters in the left eye. Further questioning revealed complain of tinnitus in the right

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ear prior to presentation to eye clinic. Pupillary examination was normal. Slit lamp examination showed bilateral anterior and posterior uveitis with few mutton fat keratic precipitates at the back of the inferior half of each cornea.

Dilated fundus examination revealed bilateral optic disc edema, abnormal macular appearance, multiple areas of retinal whitening distributed through the posterior pole.

Fundus photography (VISUCAM 524 ZEISS fundus camera) showed swelling of optic nerve, patchy swelling of the posterior pole around the optic disc separated by elevated white lines. Retinal folds are more apparent in green and blue colored images.

Optical coherence tomography (ZEISS HD 5000 OCT) showed multiple areas of subretinal fluids with maximum thickness between 700-900 um at macular area. There is no enhanced depth imaging facility (EDI) in hour OCT system to measure choroidal thickness.

Fundus fluorescein angiography is not done at presentation because the patient is generally tired.

CSF examination in her records documents 50% cell, 98% of cell are lymphocytes. All general investigation was normal. The patient put on systemic steroids (1mg/kg) with Azathioprine tablet 50 mg two times daily because of delayed onset of action.

After 3 weeks on treatment, patient visual acuity improved to 6/36 in right eye and 6/24 in the left eye. The photophobia disappeared, there is clearance of anterior uveitis, the multiple subretinal fluid has reduced to small subfoveal film of fluid.

On 3 months follow up visual acuity was 6/12 both eyes with nearly normal retinal examination, optical coherence tomography and colored fundus imaging showed no abnormal findings.

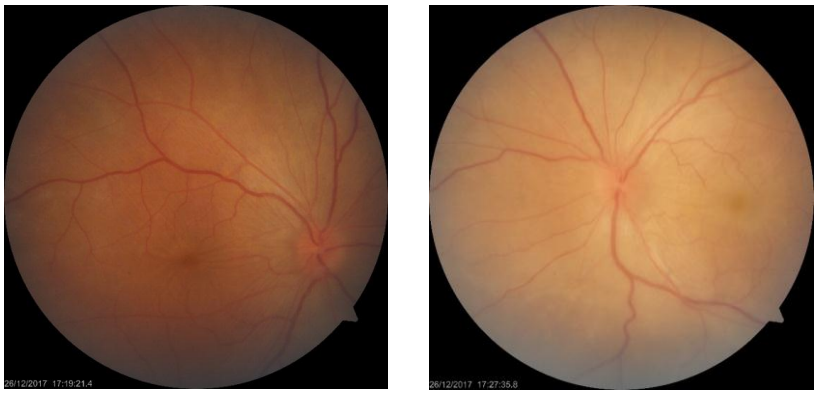

Figure 1. Colored fundus images of right and left eye showing optic discs and macular edema

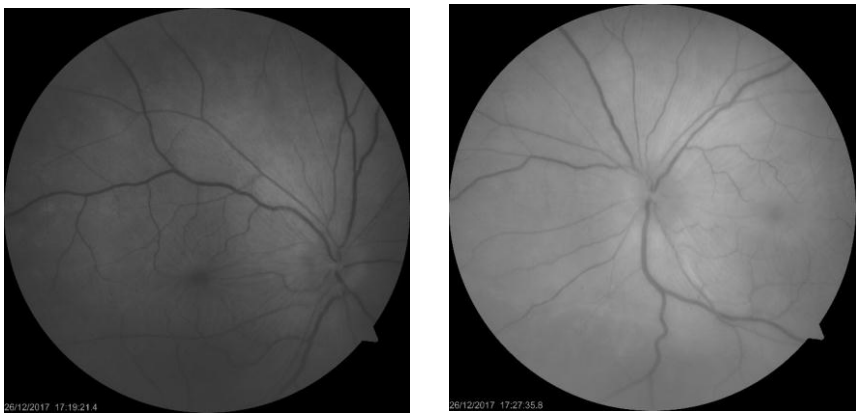

Figure 2. Red free fundus images demonstrated severity of edema and secondary retinal striations

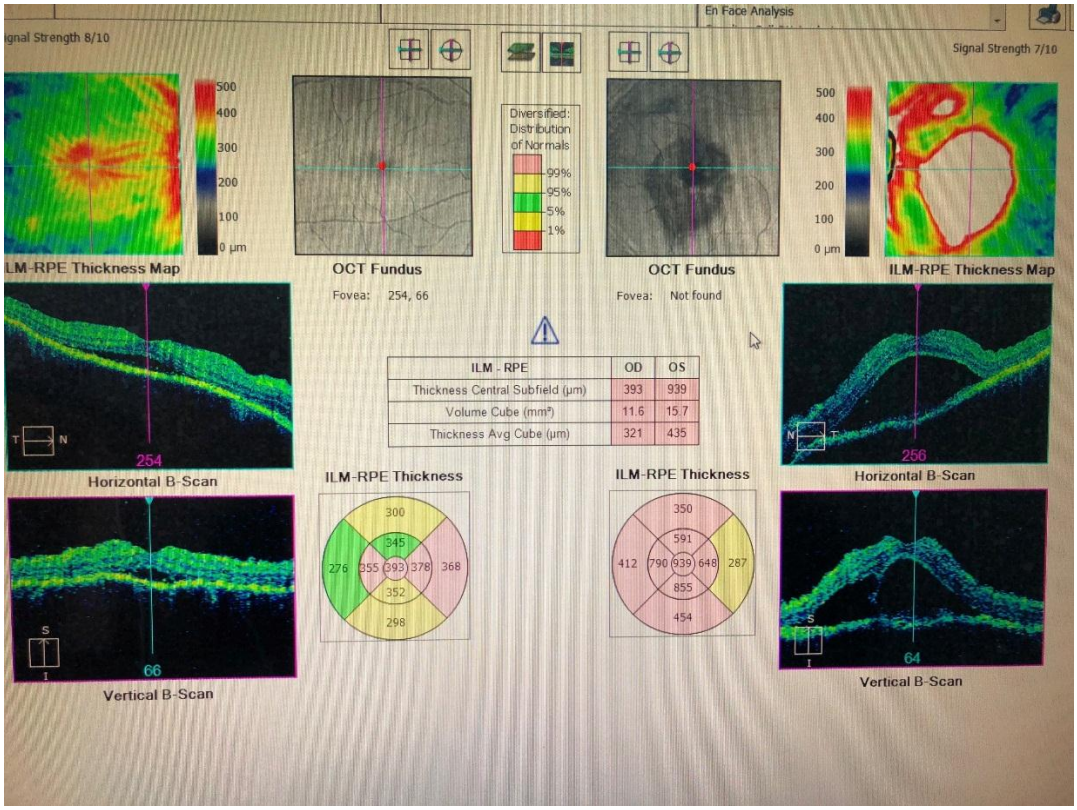

Figure 3. Optical coherence tomography of both eyes before treatment showing areas of subretinal fluids

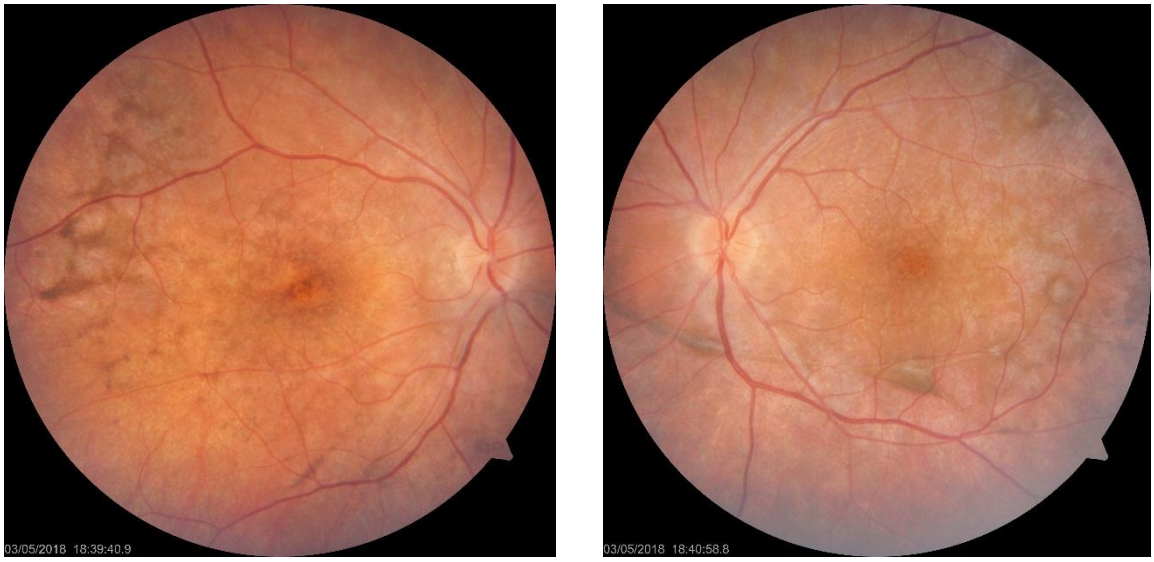

Figure 4. Colored fundus images of right and left eyes show resolution of optic discs and retinal edema with typical sun setting fundus after treatment

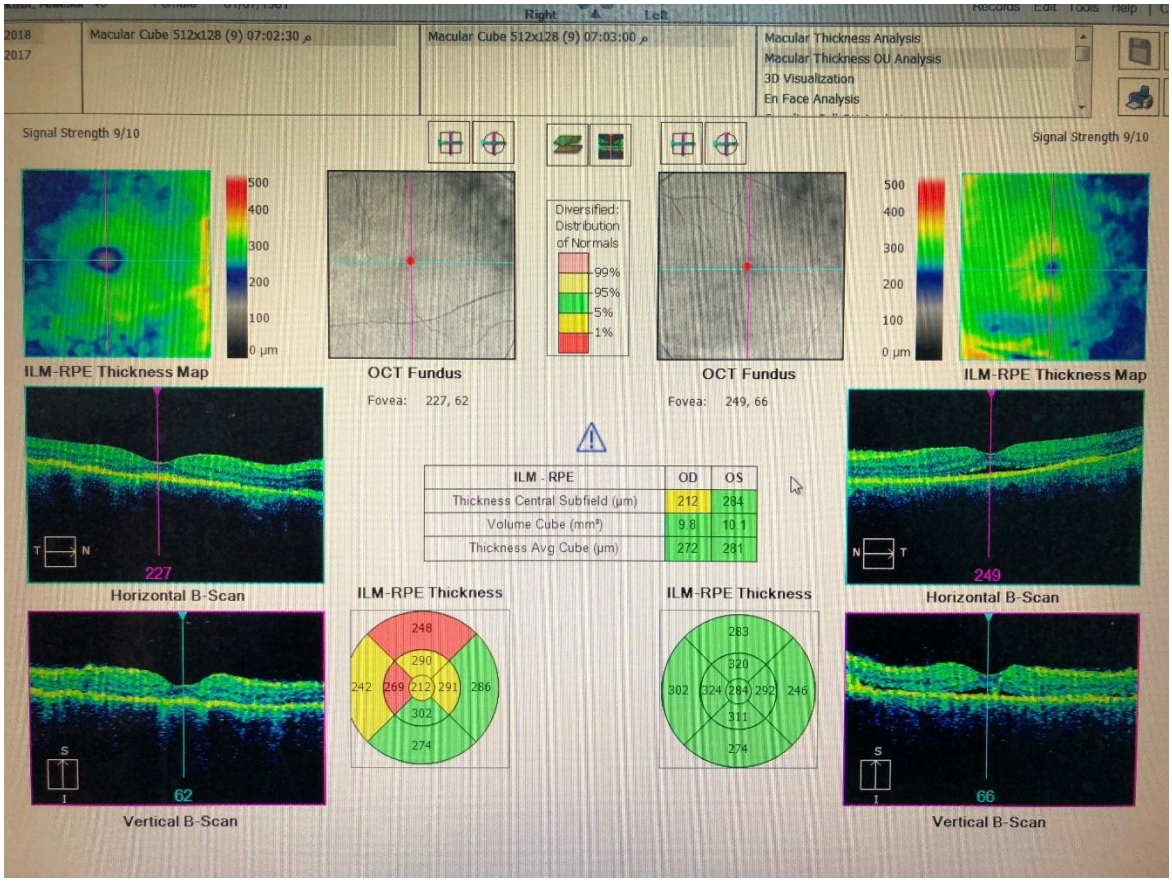

Figure 5. Optical coherence tomography of both eyes after treatment showing normal right eye image with scanty fluid in the left macula.

Discussion

Vogt koyanaji Harada disease is a sight threatening disease and a common cause of immune mediated panuveitis. Choroid, ciliary body and iris are the main sites of ocular inflammation[8]. The disease has variable clinical presentations, the diagnosis is clinical after exclusion of other similar presentations provided that there is no history of trauma[2]. Fundus flouresceine angiography and optical coherence tomography can aid the clinical diagnosis[9]. The main diagnostic ocular feature is inflammation of anterior and posterior segments of both eyes[1]. Bilateral granulomatous uveitis limits the differential diagnosis to very few diseases which may include sympathetic ophthalmitis (there should be a history of penetrating ocular trauma or surgery to one eye), posterior scleritis (unilateral, pain radiating to face jaw and awake patient from sleep), intraocular lymphoma (old age, associated neurological features, multifocal yellowish subretinal lesions), chronic serous chorioretinopathy (CSCR) is idiopathic disease affects middle aged males who are healthy characterized by areas of subretinal detachments but the vision is good with no optic disc edema or granulomatous uveitis[9]. systemic steroids are the main line of treatment other immunosuppressive drugs may combine with or replace steroids to control disease like methotrexate, azathioprine[2]. Sight threatening complications of VKH syndrome include glaucoma, cataract (amenable to treatment), choroidal neovascularization, and subretinal fibrosis[9].

Although systemic features of VKH disease form more than 60% of total symptoms and signs, bilateral granulomatous panuveitis is the commonest ocular sign and the key feature in the diagnosis of the disease[4, 5]. Headache and optic disc edema in both eyes present the patient to neurology department, normal neuroimaging in

middle aged female made the diagnosis of idiopathic intracranial hypertension (IIH), after cerebrospinal fluid (CSF) aspiration, CSF pressure were normal and medications to lower intracranial pressure didn't improve patient condition. After 3 weeks, the patient referred to ophthalmology department when blurring of vision of right eye started to develop. Finding of bilateral granulomatous panuveitis with multifocal choroiditis in both eye has proved the diagnosis. CSF pleocytosis is a strong neurological feature[4]

Conclusion:

VKH disease is an autoimmune disease that has various clinical features, bilateral optic disc edema presentation combined with headache may give other diagnosis to neurologists, we recommend detailed ophthalmological evaluation including visual acuity, visual fields for any optic disc edema especially if it is associated with drop of vision where papilledema is unlikely. Slit lamp examination may declare other ocular findings that may be presented together with optic disc edema which may help to reach the final diagnosis.

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