Case Report

A Unique Case of Acute Deterioration in Visual Acuity: Vogt-Koyanagi-Harada Disease

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Abstract

Vogt-Koyanagi-Harada (VKH) disease is a chronic autoimmune inflammatory disorder with multisystem involvement particularly involving the eye, skin, ear and brain. Our patient reported in the Ophthalmology OPD, CMH, Lahore, a 41-year-old female patient who presented with decreased visual acuity of 6/9 and 6/18 in her right and left eye respectively with intermittent tinnitus. A slit lamp examination revealed bilateral paramacular choroiditis, more pronounced in the left eye. Fundoscopy showed the 'sunset glow fundus.' Her OCT showed slight flattening of the foveal contour in the left eye with generalized foveal thinning while the right eye had parafoveal thickening with a foveal lamellar hole. FFA showed a serous detachment in her left eye. A diagnosis of bilateral incomplete Vogt-Koyanagi-Harada syndrome was made as she tested negative for infectious diseases. She was started on an oral course of steroids initially but after persistent episodic exacerbations ciclosporin 75mg twice daily was started and regular 2 weekly follow-ups were advised. The mainstay of treatment for Vogt–Koyanagi–Harada disease is systemic corticosteroid therapy but the refractory cases get additional immunosuppressants like ciclosporin which was required in our patient as well.

Keywords: Autoimmune, Fovea, Visual acuity.

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Introduction

Vogt-Koyanagi-Harada (VKH) disease is a chronic autoimmune inflammatory disorder with multisystem involvement, particularly in the eye, skin, ear and brain. Ophthalmologic manifestations include iridocyclitis, serous retinal detachments, diffuse choroidal swelling and optic disc hyperemia.[1] The disease is reported to be T-cell mediated against melanocytes, triggered by a viral infection in susceptible individuals.[2] It has 4 stages: prodromal, acute inflammatory, chronic and recurrent.[3] The disease is usually bilateral involving the eyes simultaneously, but we report a case where the ocular findings were initially unilateral and predominant in the left eye before involvement of the right eye.

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Case

This is a case of a 41-year-old female patient, presenting to the Ophthalmology OPD of Combined Military Hospital (CMH) Lahore, who is a diagnosed case of hypertension, hypothyroidism and asthma and first presented with an acute decrease in visual acuity

(VA.) She had a VA of 6/9 in the right eye and 6/18 in the left eye improving to 6/6 in the right and 6/5 in the left with a pinhole. She was initially examined under a slit lamp and had 1+ AC cells in the right and occasional cells in the left eye. The pressure was 16 mmHg in both eyes. An area of serous detachment was noted in the left eye as well. Vitreous cells were seen in both eyes occasionally. She was found to have bilateral paramacular choroiditis, more pronounced in the left eye.

She was advised Optical Coherence Tomography (OCT,) Fundus Fluorescein Angiography (FFA) alongside a battery of blood work as mentioned in Table I along with their respective results. Additional imagining includes Chest X-ray and Ultrasound Abdomen. Table 01 shows the results of the investigations ordered.

Table 1: *Investigations carried out on the patient*

Investigation	Result
➢ Blood Complete Picture a) Hemoglobin b) Total Leukocyte Count c) Platelets	 11.3g/ dL 5.92 x 10^9/L 200 x 10^3 / *u*L
➢ ESR	45 mm / hour
➢ CRP(Q)	12 mg/L
➢ Blood Urea ➢ Serum Creatinine ➢ Serum Sodium ➢ Serum Potassium ➢ Serum Calcium	3.4 mmol/L 82 *u*mol/L 141 mmol/L 4.0 mmol/L 9.0 mmol/L
➢ Gene Xpert	Mycobacterium Tuberculosis DNA not detected
➢ Serum Uric Acid	4 mg/dL
➢ Serum RA	5 IU/mL
➢ Brucella : Serum IgM ELISA Serum IgG ELISA	 Negative
➢ Toxoplasma: Serum IgM & IgG ELISA	 Negative
➢ Serum ACE Levels	4.5 IU/L

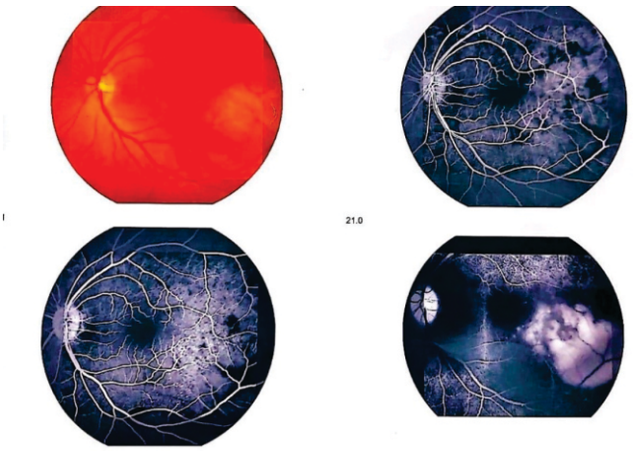

Figure I. *Findings on Fundus Fluorescein Angiography – Left eye shows initial choroidal hypoflourescene with late staining of choroiditis patch with accumulation of dye under serous detachment. The right eye shows normal choroidal/retinal hemodynamics. Both blood-retinal barriers are intact.*

She was treated with a topical sterile ophthalmic suspension of tobramycin and dexamethasone. Initially, she responded slightly to the topical treatment and her VA improved in her left eye to 6/9. She was started on ATT empirically, due to significant clinical suspicion for tuberculosis but was stopped after she tested negative. She had a normal Chest X-ray and abdominal ultrasound. She also had normal calcium levels and serum Angiotensin Converting Enzyme (ACE) levels, so there was a lower level of suspicion for sarcoidosis as well. Initial fundoscopy findings in FFA are shown in Figure I. OCT showed slight flattening of the foveal contour in the left eye with generalized foveal thinning while the right eye had parafoveal thickening with foveal lamellar hole. FFA showed an area of speckled hyperfluorescence in the temporal macula with pruning of serous fluid above it, pointing to a serous detachment in her left eye. She had complained of tinnitus previously. There was, however, no headache, deep eye pain or skin changes like vitiligo noted. As she was negative for a systemic inflammatory or infective disease, a diagnosis of bilateral incomplete Vogt-Koyanagi-Harada

syndrome was made. She was started on an oral course of steroids initially but after persistent episodic exacerbations ciclosporin 75mg twice daily was started and regular 2 weekly follow-ups were advised. She had been advised to start on Ophthalmic suspension of Prednisolone Acetate in acute vision deterioration episodes as well. She was intermittently been treated with intraocular steroid injections with significant improvement and remissions noted.

Discussion

Diagnosis of Vogt–Koyanagi–Harada disease relies on the development of panuveitis. The disease starts unilaterally and quickly involves the other eye. There is posterior uveitis, hyperemia and edema of the optic disk, multiple retinal detachments and the disease progressing to the anterior chamber eventually, thus the panuveitis. The diagnostic criteria include no history of penetrating ocular trauma or surgery before the onset of uveitis, no evidence of existing eye disease, bilateral ocular involvement and extraocular manifestations like headache, dizziness, deep orbital pain, and tinnitus.[4] An incomplete disease would have a lesser degree of involvement.

Our patient presented in the acute inflammatory stage of the disease with extensive foveal involvement in the left as well as the right eye later on. There was serous retinal detachment in her left eye. The differential diagnosis of Vogt–Koyanagi–Harada disease includes systemic chronic granulomatous diseases like tuberculosis, sarcoidosis, and ocular Lyme disease as well as infiltrative conditions of the eye like intraocular lymphoma, uveal lymphoid infiltration, metastatic carcinoma, systemic ophthalmia, posterior scleritis, and uveal effusion syndrome.[5] Some literature has documented its evidence post-COVID-19 as well.6 T.B was high up on the list considering the disease is endemic in our country and very widespread. Hence, the patient was started on ATT for a short duration. But through extensive systemic examination and workup, all the other causes were excluded.

The mainstay of treatment for Vogt–Koyanagi–Harada disease is systemic corticosteroid therapy but the refractory cases get additional immunosuppressants like ciclosporin. This was the case with our patient as well. In most cases, aggressive treatment with steroids and non-steroidal immunosuppressive agents is required. [7]

Cases of VKH disease are usually bilateral involving both the eyes but just like our patient whose initial presentation was unilateral, a few cases have been reported of only unilateral disease, as in a 20-year-old, female patient, FFA was diagnostic in this case as well. [8]

Conclusion

In conclusion, we present a rare case of incomplete Vogt–Koyanagi–Harada disease that was timely picked and diagnosed. Refractory initially to systemic steroid therapy was treated additionally with immunosuppressant and the patient responded well with adequate recovery.

Conflict of interest: *None*

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Authors Contribution

MM: Conceptualization of study
SN: Literature Search
DB: Statistical Analysis
SN: Data Collection and Analysis
MM: Writing of Manuscript
SN: Drafting, Revision
All authors are equally accountable for accuracy, integrity of all aspects of the research work.