Case study

Unilateral Normotensive Angle Closure as a Presenting Feature of Vogt-Koyanagi-Harada Disease

ABSTRACT

We describe an atypical presentation of probable Vogt-Koyanagi-Harada Disease with unilateral angle closure and normal intraocular pressure in a young male. On Ultrasonographic Biomicroscopy, there was anterior rotation of ciliary processes with supraciliary effusion. Fundus Fluorescein Angiography showed early phase hyperfluorescent spots followed by leakage and pooling, typical of VKHD. The angle closure responded to intravenous methyl prednisolone followed by tapered oral steroids. It is necessary for the ophthalmologist to be aware of this presentation so that there is no delay in initiating the required therapy and the patient does not have to undergo various unnecessary procedures like laser iridotomies.

Keywords: Vogt-Koyanagi-Harada disease, Uveitis, Angle closure glaucoma, Ophthalmology

1. INTRODUCTION

The Vogt Koyanagi Harada disease (VKHD) is a multisystem autoimmune disease with bilateral posterior or panuveitis, with a female preponderance. Patients usually present with ocular symptoms like blurred vision, redness of conjunctiva and ocular pain. There have been a few reports of a rare presentation with acute angle closure glaucoma.[1], [2] We report a young Indian male who presented with unilateral angle closure, but with normal intraocular pressure (IOP), and responded to steroid therapy.

2. CASE DETAILS

A 40-year-old hypertensive male presented redness, blurred vision, and pain in his left eye, for 4 days. He had headache and raised blood pressure of 190/140 mmHg for which he was admitted to the hospital. His best corrected visual acuity was 6/6, N6 unaided in the right eye and 6/60, N8 improving to 6/12, N8 in the left eye with -2.00DS. He had no previous history of wearing glasses. On slit lamp examination, the right eye appeared normal (Figure 1a), while there was circumciliary congestion in his left eye, with shallow anterior chamber (Van Herick grade 1) and 2+ cells (Figure 1b). His intraocular pressure was 12- and 15-mm Hg in right and left eye respectively, on applanation tonometer. On gonioscopy, the angles of right eye were fully open with scleral spur visible over 360 degrees. The angles of left eye were closed over 360 degrees (Figure 1c). They opened to the pigmented trabecular meshwork in the superior and inferior angles on indentation. [Insert Figure1]

Figure 1:

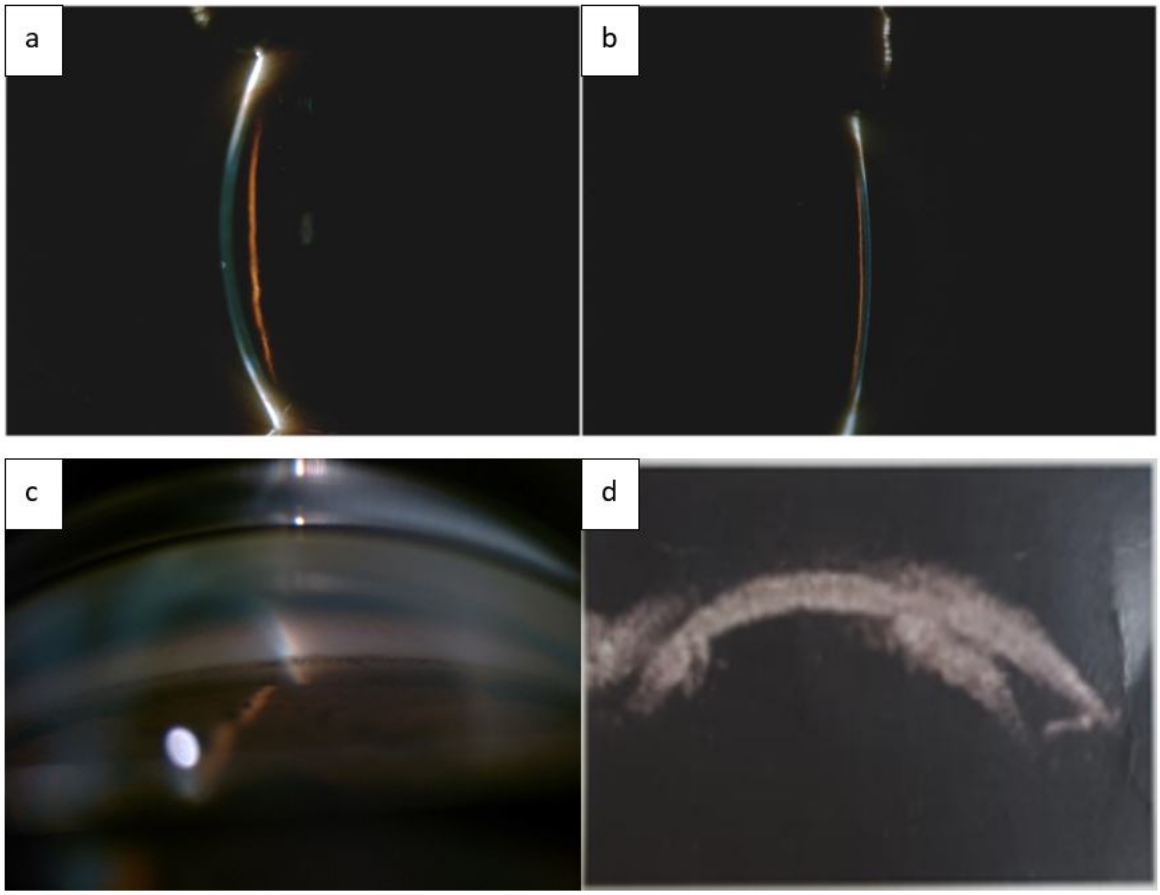

Figure 1: a. Slit lamp image of right eye; b. Slit lamp image of left eye; c. Gonioscopic view of left eye; d. UBM of left eye

On examination of posterior pole, the optic disc was hyperemic in both eyes with a cup disc ratio of 0.5:1 in the right and 0.4:1 in the left eye. Retinal folds were seen at the macula in the right eye and neurosensory detachments and retinal

folds were seen in the peripapillary area and near the superior and inferior arcuate vessels in the left eye (Figure 2a and 2b). Ultrasound Biomicroscopy (UBM) showed circumferential supraciliary effusion causing medial rotation of ciliary body and processes, leading to secondary angle closure, in the left eye (Figure 1d). B scan ultrasound in the left eye showed serous retinal detachment superior to the disc and increased chorioretinal thickness in some areas. Optical Coherence Tomography (OCT) confirmed the neurosensory detachment. The patient was given topical steroid and atropine along with anti-glaucoma medication in anticipation of rise in intraocular pressure as inflammation subsides. On follow-up, the circumciliary congestion and anterior chamber cells decreased. Fluorescein fundus angiography (FFA) showed few early hyperfluorescent spots with leakage at the superior temporal retinal artery (Figure2d) and few hyperfluorescent spots in the right eye (Figure 2c). As the patient presented very early in the course of the disease and treatment was initiated, classic FFA findings did not develop. [Insert Figure2]

Figure 2:

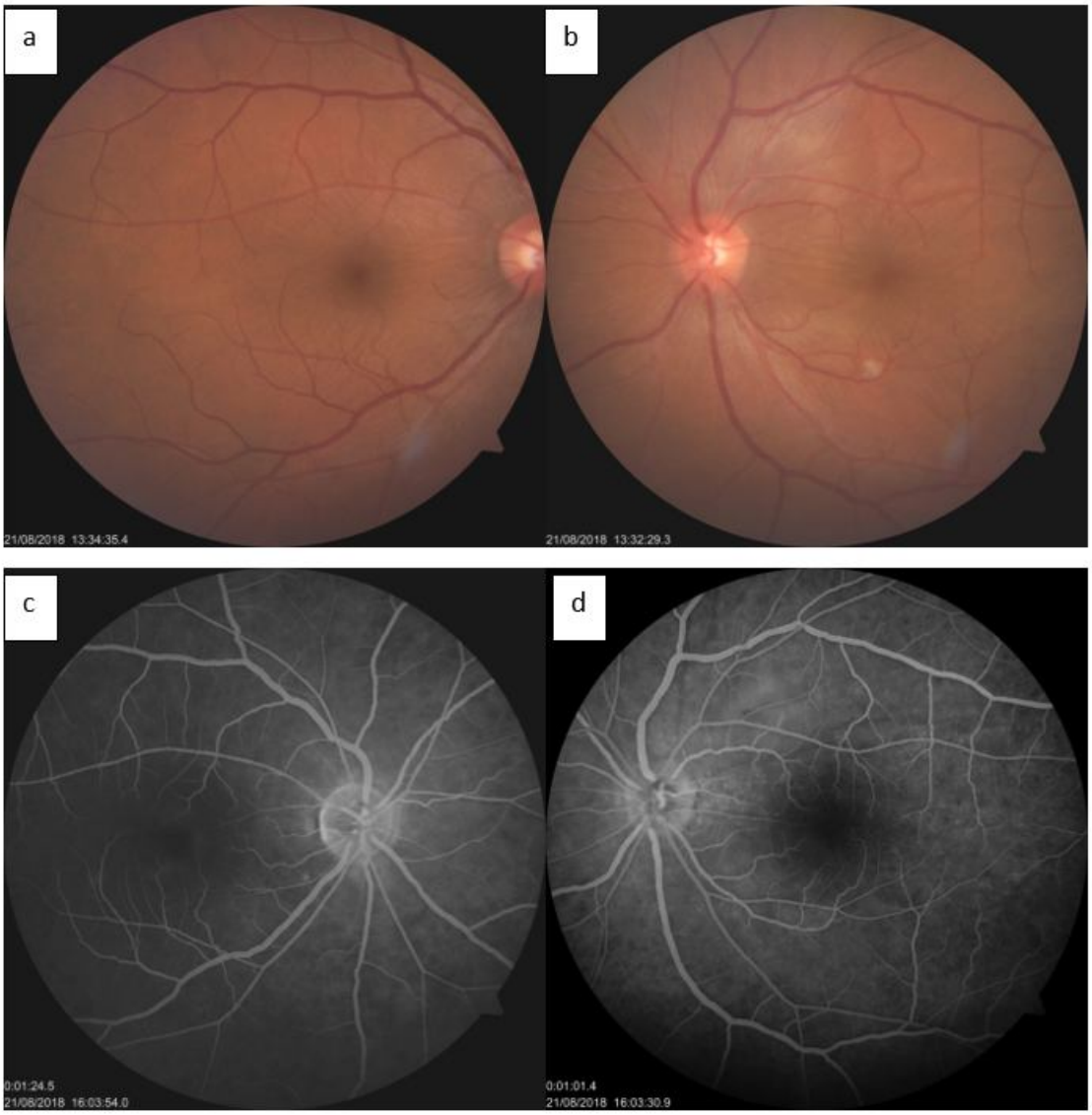

Figure 2: a. Fundus photograph of right eye; b. Fundus photograph of left eye; c. FFA image of right eye; d. FFA image of left eye

As the angles of the right eye were getting progressively narrower, with only the pigmented trabecular meshwork visible, he was started on topical atropine in the right eye, as well. He was prescribed intravenous methyl prednisolone, 1 g/day for 3 days, followed by oral steroids with tapering over 6 months.

On successive follow-up examinations, there was evidence of resolving neurosensory detachment and resorption of subretinal fluid. On his last visit, his visual acuity was 6/9 in both eyes unaided, with bilateral deep anterior chamber and open angles, and resolved inflammation. There were multiple subtle retinal pigment epithelium (RPE) changes in both eyes.

3. DISCUSSION

The list of differential diagnoses may include Vogt-Koyanagi-Harada disease, hypertensive choroidopathy, uveal effusion syndrome, central serous chorioretinopathy, multifocal choroiditis, posterior scleritis, masquerade (lymphoma, leukemia, carcinoma, metastasis).

Vogt-Koyanagi-Harada disease (VKHD) is a granulomatous autoimmune disease with a bilateral panuveitis. It occurs more frequently in races like Hispanics and Asians. It has a female sex predilection, more often in the second to eighth decades of life.[3] There may be a genetic susceptibility, with increased risk among those with HLA genotype DRB1 *0405.[4] An autoimmune reaction to melanocytes or their tyrosinase related peptides plays a role in the pathogenesis.[5] Its evolution has been described in the following phases : prodromal, uveitic, convalescent and recurrent.[3]

Patients usually present with blurring of vision, redness, ocular pain in both eyes. They might have a preceding history of headache, neck stiffness, earache, vertigo. There have been reports of VKHD presenting with bilateral acute angle closure glaucoma due to anterior segment inflammation and ciliary effusions.[1], [2] Our case is the first reported to have presented with unilateral angle closure with a normal intraocular pressure. Open angles at presentation in the fellow eye and presence of multiple serous neurosensory detachments in both eyes prompted a UBM and FFA which confirmed the diagnosis as probable VKHD.[6] Adequate response to steroid therapy was seen in the form of resolution of neurosensory retinal detachments, opening of angles and deepening of the anterior chamber. In a retrospective analysis of 486 patients with VKHD, eight patients had presented with bilateral narrow angles, six of them women. They were misdiagnosed as acute angle closure glaucoma, but the raised intra ocular pressure did not respond to anti glaucoma medication. One patient underwent enucleation of one eye after failed trabeculectomies, before they were referred to a uveitis clinic. The intraocular pressure was controlled after starting the patient on systemic steroids as for VKHD.[7] This case is in

concordance with other reported cases where the angle closure glaucoma did not respond to laser iridotomies but improved on starting steroid therapy.[2], [7]

4. CONCLUSION

Vogt-Koyanagi-Harada disease may present unilaterally or asymmetrically. Despite the presence of angle closure, intraocular pressure is relatively normal. Fluorescein Angiography and Ultrasound Biomicroscopy are useful to confirm the diagnosis.

CONSENT

Written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

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