

CASE REPORT

Vogt-Koyanagi-Harada syndrome presenting as bilateral simultaneous acute angle closure

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Abstract

Presentation of bilateral simultaneous angle closure in a patient must alert one to look beyond the common “knee-jerk” diagnosis of acute primary angle closure (APAC). We describe a case with bilateral simultaneous acute angle closure with signs of anterior chamber inflammation with intraocular pressure of 60 mmHg. Based on the clinical details and ultrasonic findings of peripapillary choroidal thickening and supraciliary fluid, a diagnosis of Vogt-Koyanagi-Harada (VKH) disease was made. Our case highlights that every angle closure is not a primary angle closure requiring laser peripheral iridotomy. Detailed clinical examination with appropriate investigations could prevent misdiagnosis of APAC in VKH.

Introduction

Acute primary angle closure (APAC) is an ocular emergency which is traditionally managed with laser peripheral iridotomy (LPI). However, it rarely presents as an acute attack in both the eyes simultaneously. Such a presentation must always alert one to look beyond the common “knee-jerk” diagnosis of APAC. Bilateral acute angle closure (AAC) has been reported due to topical mydriatics in predisposed individuals, anticholinergic drugs, anticonvulsants like topiramate, following general anesthesia and spherophakia.^[1-3]

Vogt-Koyanagi-Harada (VKH) disease is a bilateral, chronic intraocular inflammation associated with neurological and cutaneous abnormalities including headache, tinnitus, vitiligo, poliosis, and alopecia.^[4,5] Decreased vision with headache is the most commonly presenting complaint and anterior chamber/vitreous cells are the most common sign of acute VKH.^[5] Association of glaucoma/ocular hypertension with VKH has been studied across Asian/Western population with reported prevalence of 10–62%.^[6-8] Rarely, it can manifest as an AAC.^[9-11] A 10-year retrospective review of VKH patients by Yang *et al.* revealed 8 (0.08%) cases which were misdiagnosed as APAC.^[9]

We describe a rare presentation of VKH as AAC along with a short literature review.

Case Report

A 52-year-old female presented with complaints of headache, decreased vision, and redness in both eyes for 1 week. Ocular examination at presentation revealed best-corrected visual acuity (BCVA) of 6/36 OU and intraocular pressure (IOP) of 52 and 62 mmHg on Goldmann applanation tonometry in the right and left eye, respectively. Both eyes showed mild ciliary flush with corneal edema, diffuse granular keratic precipitates (kps), mild flare, and shallow anterior chamber, with sluggishly reacting pupils [Figure 1a]. Gonioscopy revealed bilateral closed angles in all four quadrants.

Fundus examination revealed blurring of the nasal optic disc margin [Figure 1b] with shallow retinal detachment in the inferior periphery in both eyes. There was no vitritis. In view of the suspicion of VKH (elderly female, inferior exudative detachment, and disc edema), B-scan ultrasonography (USG) was done which revealed choroidal thickening (2.1 mm) in

the peripapillary area [Figure 1c]. Ultrasound biomicroscopy (UBM) showed supraciliary fluid with closed angles in both the eyes [Figure 1d].

Keeping bilateral AAC in mind, she was questioned about drug intake in the past. There was no history of psychotropic drug intake, recent general anesthesia, or snakebite.

She was started on oral acetazolamide, timolol, and brimonidine eye drops in both eyes. IOP reduced to 36 mmHg in both eyes.

Based on the presence of bilateral granulomatous iridocyclitis, bilateral exudative retinal detachment, and ultrasonic findings of peripapillary thickening, we made a presumptive diagnosis of VKH disease and started the patient on oral steroids 1 mg/kg body weight in combination with topical steroids betamethasone 1 hourly and atropine 1% eye drops 3 times a day. General physical examination, as well as neurological, dermatological, and auditory examinations for VKH syndrome, was all normal.

Within 3 days of therapy, BCVA improved to 6/9 in both eyes, IOP was 10 mmHg, with clear cornea and open angles [Figure 2a]. Fundus showed resolution of the exudative detachment [Figure 2b] and the disc margins were clear. UBM showed marked reduction in supraciliary fluid [Figure 2c, arrow].

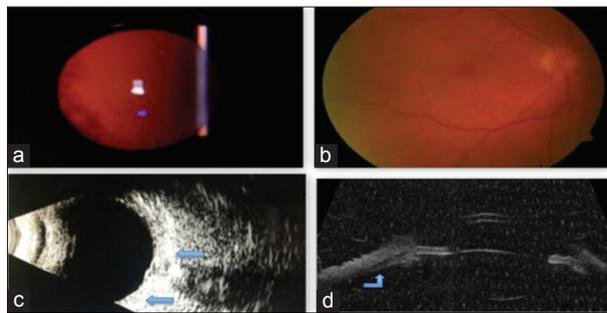


Figure 1: (a) Clinical photograph of the patient under retroillumination showing keratic precipitates in the inferior half of the cornea. (b) Right fundus photograph of the patient showing media clarity Grade II with blurring of the disc margins. (c) B scan ultrasound of the patient showing peripapillary thickening. (d) Ultrasound biomicroscopy showing anterior bowing of peripheral iris with fluid in supraciliary space

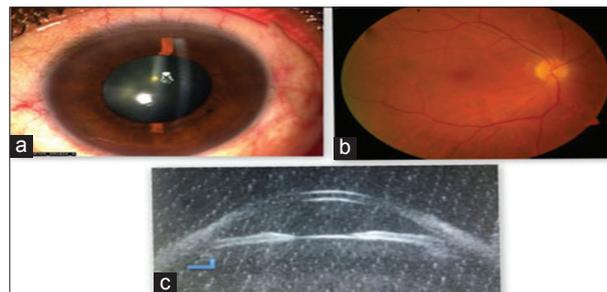


Figure 2: (a) Anterior segment of the left eye after 2 weeks of corticosteroid therapy showing disappearance of keratic precipitates. (b) Fundus photograph showing disappearance of disc edema. (c) Ultrasound biomicroscopy shows disappearance of supraciliary fluid at 6 weeks

All antiglaucoma treatment could be stopped with controlled IOP, and immunosuppression therapy for VKH was initiated.

Discussion

Secondary angle-closure glaucoma due to supraciliary fluid causing anterior rotation of the ciliary body may present as an AAC attack.^[1-3] Prompt recognition of the condition is imperative for optimum management. It is important to understand that this condition requires steroids and cycloplegics in contrast to the usual treatment of pilocarpine and LPI in primary angle closure. The latter treatment would prove to be disastrous due to exacerbated inflammation and further anterior movement of the iris-lens diaphragm.

In our patient, despite the presentation of a bilateral AAC, the recognition of granulomatous inflammation in anterior chamber and increased choroidal thickening with exudative retinal detachment supported the diagnosis of VKH. Prompt management resulted in resolution of the attack. We have previously reported two patients presenting with bilateral AAC, who were subsequently found to have acute retinal necrosis^[12] and tubercular granuloma,^[13] respectively, illustrating that posterior segment inflammatory diseases can have unusual anterior segment manifestations.

Previously published reports on AAC in VKH found it to be more common in females with associated uveal effusion. Rathinam *et al.* reported mildly elevated IOP with maximum of 27 mmHg in their series of three patients who did well with steroids.^[11] Yang *et al.* retrospectively found 8 eyes of 486 eyes VKH misdiagnosed as an angle closure where LPI was performed in three eyes which worsened the course until the correct diagnosis was established.^[9] UBM was performed in two patients at 3 weeks, which showed fluid in supraciliary space. Our patient had 60 mmHg of IOP with evident supraciliary fluid on UBM and choroidal thickening on USG. The presence of bilateral kps with disc edema in an elderly female raised the suspicion of VKH.

Glaucoma has been reported to be one of the most common ocular complications of VKH. Pandey *et al.* found 15.8% prevalence of glaucoma in retrospective review of 448 eyes diagnosed with VKH over 23 years.^[7] The most common mechanism was open-angle glaucoma in 46 eyes (64.8%), angle closure in 21 eyes (29.6%), and of combined mechanisms in the remainder (4 eyes, 5.6%). Only nine cases of 448 eyes presented with angle closure (four at presentation and five during follow-up) that was managed with LPI. UBM in these cases revealed supraciliary fluid in only 5/9 (55%) cases.

Bilateral AAC is a rarely seen ocular condition. Various reported etiologies are drug induced (sulfonamides, phenylephrine, ephedrine, and botulinum toxin), snakebite, uveal effusion, microspherophakia, and VKH.^[1-3,7] Although we did consider drug induced secondary angle closure, literature search revealed no reports of selective serotonin reuptake inhibitors (SSRI) to be implicated in the pathogenesis and our

patient used SSRI in the past. Mean duration of the onset of raised IOP was 71 days, but earliest has been reported within 1 week of drug intake with topiramate.^[2] The presence of granulomatous kps, optic disc staining, and inferior exudative detachment tilted the diagnosis toward VKH disease.

Our case highlights the fact that every angle closure is not a primary angle closure requiring LPI. This is especially important in countries such as India and China where PACG comprises half of primary glaucoma encountered and there may be an increased tendency to go ahead with LPI. In VKH, an autoimmune response is generated against uveal melanocytes, leading to uveal effusion. Secondary angle closure is due to inflammation as well as forward rotation of ciliary body and not pupillary block. Pilocarpine and LPI in such a scenario would increase the dilatation of uveal blood vessels and the resultant angle congestion would be likely to complicate the condition further.

Auxiliary investigations such as UBM and USG had a major role in reaching diagnosis in such atypical presentation. However, to order these investigations, a high index of suspicion is required. Our case seeks to highlight the possibility of VKH presenting as bilateral simultaneous AAC in the presence of intraocular inflammation which responded well to topical and oral steroids, avoiding LPI.

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