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Incomplete Vogt-Koyanagi-Harada in a 14-year-Old African American female with bilateral disc edema

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ABSTRACT

Purpose: To report an unusual case of incomplete Vogt-Koyanagi-Harada (VKH) in a 14-Year-Old African American female.

Observations: Here we present a 14-Year-Old African American Female with incomplete VKH who presented to the emergency department with a one-month history of malaise, fever, bilateral decreased vision and temporal headaches. At the time of presentation, she was found to have bilateral anterior uveitis and disc edema. The brain and orbit MRI were unremarkable, CSF analysis showed lymphocytic pleocytosis with negative cultures and gram stain. An extensive lab workup was unrevealing.

Conclusion: This is an unusual case of incomplete VKH in a young African American female presenting with anterior uveitis and disc edema without serous retinal detachments. Initiation of high dose oral prednisone promptly after diagnosis resulted in rapid improvement of symptoms and improvement in visual acuity with the resolution of the disc edema. This case highlights the importance of considering VKH in the differential diagnosis of disc edema associated with uveitis, even in the absence of serous retinal detachment.

1. Introduction

Vogt-Koyanagi-Harada (VKH) disease is a multisystem disease with auditory, neurologic, ocular, and dermatologic manifestations.[1,2] Although the true pathophysiology of the disease is not well understood, current leading theories suggest an autoimmune T-cell mediated process against melanocytes.[1] VKH classically presents as bilateral panuveitis as well as extraocular manifestations such as meningismus, alopecia, poliosis, vitiligo, and dysacousia.[1] The disease mainly affects pigmented individuals.[1] It has been reported to occur less frequently in African Americans when compared to other ethnicities, including Hispanics, Native Americans, Asians, and Middle Easterners.[1,4]

We present an unusual case of a 14-year-old African American female with incomplete VKH manifesting as bilateral anterior uveitis and disc edema.

2. Case report

A 14-year-old African-American female with no significant past medical history was referred to the emergency department (ED) for worsening headaches and vision loss. Her symptoms started one month prior to presentation with fevers (102.6 °F), sudden onset pounding bitemporal headache of 8/10 severity, body aches, paranasal sinus drainage and occasional cough. She was seen at an urgent care clinic and was started on amoxicillin/clavulanic acid. There was no lymphadenopathy or meningismus at that time. The patient underwent a CT scan of the head at an outside hospital, which was reported as normal. One week later, she started to notice bilateral eye redness. She was evaluated and found to have anterior uveitis and disc edema. She was started on topical steroids and cycloplegic eye drops. On follow-up visit, she was noted to have worsening disc edema and visual acuity and was referred to our ED.

On examination, visual acuity at distance was 20/40 in the right eye and 20/200–1 in the left eye with no improvement with pinhole. Pupils were equal, round, and sluggishly reactive to light with no afferent pupillary defect. Extraocular motility was full with intact visual fields to confrontation bilaterally. Intraocular pressures were 9 mmHg and 10 mmHg in the right and left eye, respectively. On slit-lamp examination, she had 1+ conjunctival injection in both eyes. The anterior chamber was deep with 3+ cell and inferior keratic precipitates in both eyes. She was noted to have bilateral posterior synechia with pigment on anterior lens capsule bilaterally. Dilated fundus exam showed bilateral disc

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edema, diffuse choroidal thickening, and fine macular exudates in the left eye (Fig. 1). Her blood pressure was 100/78 mmHg and weight was 62.4 kg (BMI 19.92).

MRI of brain/orbit and MRV were performed in the ED and were unremarkable except for enhancement of the optic disc bilaterally. The patient was admitted for further workup. Laboratory testing including CBC, CMP, ACE, ANCA panel, QuantiFERON gold, HLA-B27, ANA, Bartonella, and syphilis antibody were negative. Lyme IgG was positive; however, confirmatory western plot testing came back negative. Lumbar puncture showed lymphocytic pleocytosis with negative gram stain and cultures with elevated CSF opening pressure of 27 cm H_2O.

Optical coherence tomography showed intact foveal contour and significant peripapillary retinal nerve fiber layer edema in both eyes (Fig. 2). The patient was diagnosed with incomplete VKH and started on 60 mg of oral prednisone daily and continued on topical steroids.

At the follow-up visit 3 weeks after the discharge, vision improved to 20/20 in the right eye and 20/25 in the left eye. Intraocular pressures were normal in both eyes. The anterior chamber was deep with 1+ cell bilaterally. The dilated fundus exam was notable for bilateral disc edema.

At the subsequent follow-up, 6 weeks after the discharge, vision remained 20/20 in the right eye and 20/25 in the left. The anterior chamber was deep and quiet in both eyes. The dilated fundus exam revealed blurred disc margins. The oral prednisone dose was decreased to 40mg. Topical steroids were reduced to twice daily use. The oral steroids were tapered slowly over the subsequent follow-up visits and methotrexate was started. At the 4 months follow up-visit, dilated fundus exam showed sharp and pink nerves without disc edema (Fig. 3).

At the final visit, 6 months after the discharge, vision measured 20/20 in both eyes. The anterior chamber was deep and quiet in both eyes. The dilated fundus exam revealed sharp disc margins with healthy rims; her methotrexate dose was 25 mg/week.

3. Discussion

Vogt-Koyanagi-Harada (VKH) is an autoimmune disease mainly targeting the melanin-containing cells in the ears, skin, eyes and meninges.[2] The diagnosis is made clinically based on the criteria established at the First International Workshop on Vogt-Koyanagi-Harada disease.[3] For the diagnosis of complete VKH disease presentation should include all of the following 5 criteria and for the diagnosis of incomplete VKH disease presentation should include criteria 1–3 as well as either 4 or 5:

1. No prior history of surgery or penetrating ocular trauma before the onset of uveitis
2. No laboratory or clinical evidence suggesting other ocular pathology
3. Bilateral ocular involvement (depending on the stage of the disease, either a or b must be met)
   a) Early disease manifestations:
      - Evidence of diffuse choroiditis: focal subretinal fluid, or bullous serous retinal detachment
        OR
      - Characteristic FA findings AND diffuse choroidal thickening
   b) Late disease manifestations:
      - History of prior uveitis matching the above description AND evidence of ocular depigmentation (sunset glow fundus, Sugiura sign)
      - AND other ocular manifestations such as recurrent/chronic anterior uveitis, RPE clumping/migration, depigmented chorioretinal scar
4. Neurologic/Auditory findings (meningismus, tinnitus, CSF pleocytosis)
5. Cutaneous findings (Alopecia, poliosis, vitiligo)[2,3]

Our patient met the first 4 criteria but lacked integumentary changes and was diagnosed with incomplete VKH.

VKH is most commonly seen amongst individuals with pigmented skin such as Asians, Native Americans, Middle eastern and Hispanics.[1] However, the disease is relatively uncommon in Africans and Caucasians.[1] VKH often presents in the third to fifth decade of life and it is more predominant in females.[1] Our patient is a 14-year-old African American female, which makes her an unusual case of VKH given her age and ethnicity. Wang et al. looked at the demographics of patients with VKH and in most case series African Americans comprised less than 10% of the cases.[4]

In the acute uveitic stage, VKH most commonly presents with bilateral, symmetric panuveitis, optic disc hyperemia, optic disc edema, choroidal thickening and various degrees of serous retinal detachment (SED).[2] Choroidal involvement in this stage also affects the retinal pigment epithelium, which in turn leads to collection of subretinal fluid and SED.[2] However, isolated disc edema, as in our patient, without serious retinal detachments is unusual. It is important to consider that subclinical SEDs can be missed on clinical examination. Attia and colleagues looked at ocular imaging characteristics of patients with acute VKH without evident serous retinal detachment clinically.[5] Almost half of the patients in their series were referred with a diagnosis of primary optic nerve disorder.[5] Thirty out of 36 eyes had optic disc swelling while OCT showed subclinical SED in 50%.[5] Our patient had disc edema on

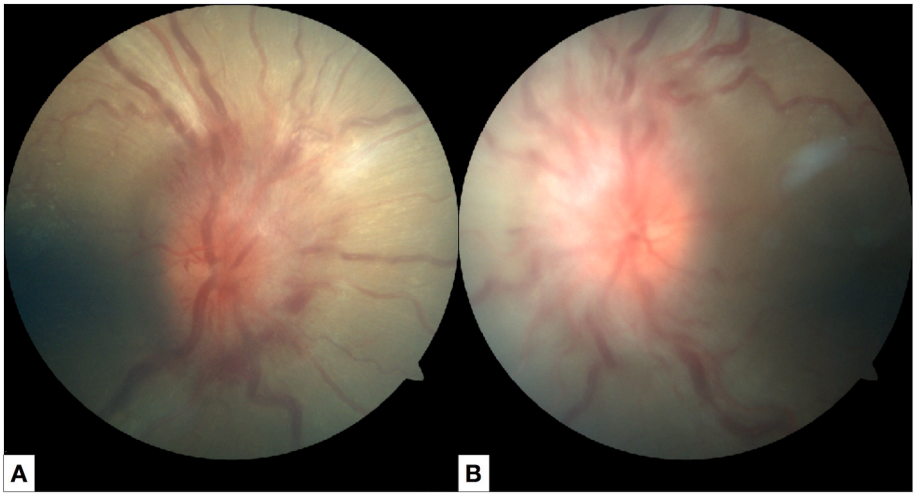

Fig. 1. Title: Fundus Picture at Presentation. Legend: Fundus picture at initial encounter of the right (A) and left (B) eye showing significant disc edema.

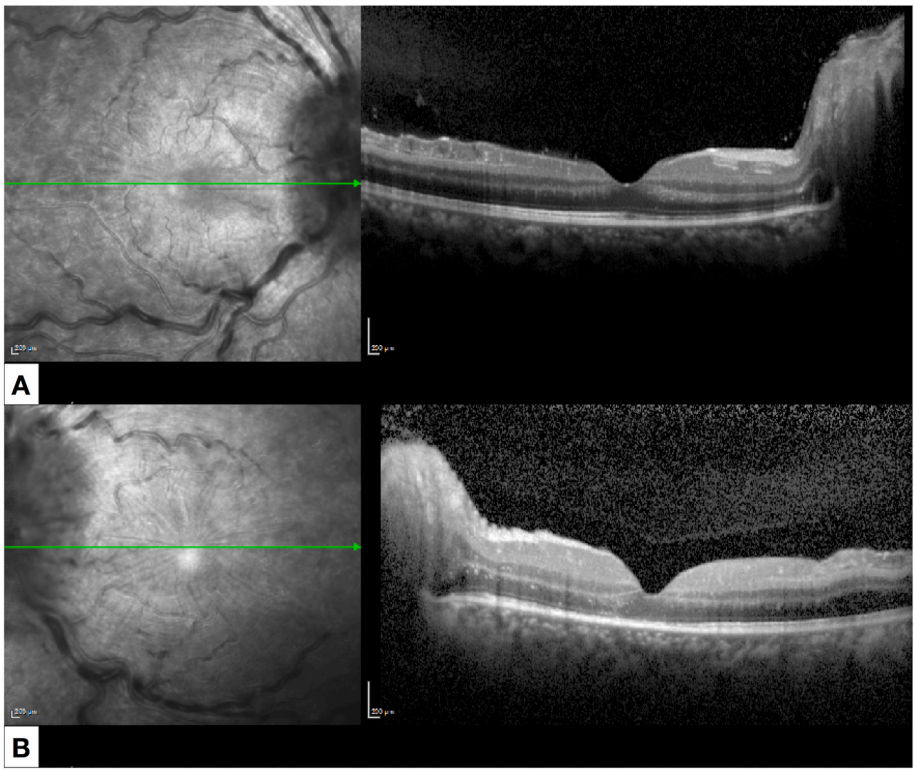

Fig. 2. Title: Spectral-Domain Optical Coherence Tomography. Legends: Spectral-domain optical coherence tomography (OCT) of the right (A) and left (B) eye demonstrating preserved foveal contour and retinal nerve fiber layer edema nasally in both eyes.

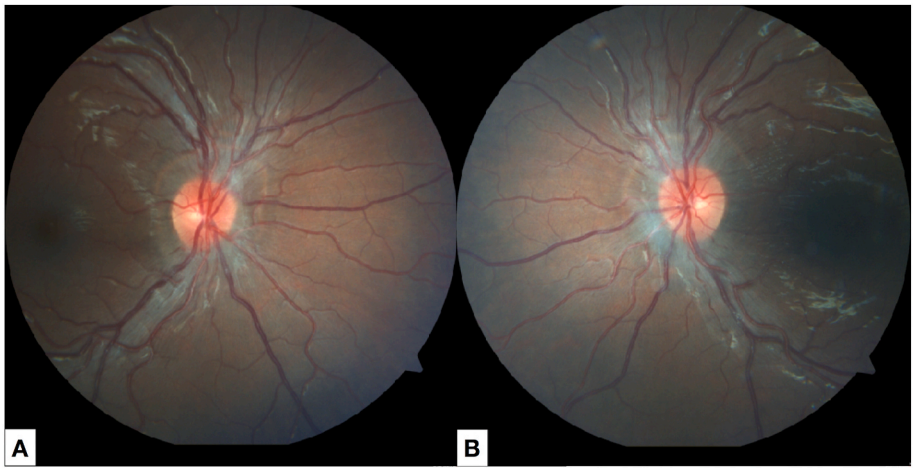

Fig. 3. Title: Follow-up Fundus Picture. Legend: Follow-up fundus picture of the right (A) and left (B) eye showing resolution of the disc edema in both eyes 4 months after starting high dose oral steroids.

exam without SED clinically or on OCT. Yang et al. reported a similar case to ours of probable VKH in a 40-year-old Korean woman with bilateral disc edema.[6] To our knowledge, this is the second case report of VKH presenting with bilateral disc edema without SED, and the first in an African American adolescent.

Nakao et al. evaluated the features that are associated with the presence of disc edema in VKH patients.[7] They have found that older patients >50 years old are more likely to present with disc edema.[7] In addition, a smaller cup to disc ratio was associated with the presence of disc edema.[7] In their study, they have also showed that eyes that present with disc edema are likely to have persistent visual field defects after resolution.[7] Upon resolution of the disc edema in our patient, she was noted to have a small cup to disc ratio of roughly 0.2 and no subjective visual field defects.

Early onset VKH in patients less than 14 years old can be aggressive. In one study, despite medical therapy and cataract surgery, visual outcomes were significantly worse in children compared to adults.[8] However, in a more recent report by Abu El-Asrar and colleagues, the overall prognosis of VKH in children remains favorable.[9] In this report, authors concluded that other prognostic factors such as initial presentation, delay in treatment, recurrence of inflammation and the type and tapering length of steroids played a more important role than age of the patient.[9]

The key to the management of VKH is early detection and systemic corticosteroid in the acute uveitis phase.[2] Adequate and prompt treatment leads to a lower rate of recurrence, changes the course of fundus

depigmentation and reduces the extraocular manifestations of the disease.[2] The recommended course of treatment consists of a high starting dose of prednisone (1–2mg/kg/day) followed by a 6 months slow taper.[2] Early termination of corticosteroid therapy is associated with worse visual prognosis and increased risk of recurrence.[2] In cases of recurrent inflammation after appropriate steroid taper, immunomodulatory therapy should be considered.[2]

In our case, the CSF opening pressure was elevated and it cannot be ruled out as the cause of the disc edema. However, the presence of uveitis, CSF pleocytosis and the prompt response to oral steroids point to VKH as the cause of the disc edema rather than the mildly elevated intracranial pressure. According to a study by Norose et al. opening pressure in most cases of VKH is normal and ranges from 8 to 21 cmH_2O.[10] However, Inomata and Kato report that opening pressure could be slightly elevated (20–25 cmH_2O) in some cases of VKH.[11]

There are numerous other causes of disc edema and uveitis in the pediatric population. Bilateral disc edema can be caused by intracranial hypertension (primary and secondary causes),[12–14] optic neuritis,[15] leukemic infiltration of the optic nerve,[16–18] and compression by tumors at the sella.[19] Pseudo-papilledema caused by anomalous optic nerves, optic nerve drusen, and retinal nerve fiber layer myelination represents another diagnostic consideration.[14] Bilateral uveitis in children occurs in the setting of juvenile idiopathic arthritis (JIA), pars planitis/intermediate uveitis, sarcoidosis, toxoplasmosis (congenital/acquired), and acute retinal necrosis (HSV/VZV).[20]

In summary, we present an unusual case of incomplete VKH in a 14-year-old African American female presenting in the acute uveitic stage with disc edema, anterior uveitis, and mildly elevated CSF opening pressure. This case highlights the importance of considering VKH in the differential diagnosis of disc edema associated with uveitis, even in the absence of serous retinal detachment.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

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