

HSV or HLA?

Jordan R. Conger, MD
Resident Physician
Department of Ophthalmology
Gavin Herbert Eye Institute, University of California Irvine

ABSTRACT

Purpose: To present a case of HLA-B27 related acute anterior uveitis initially thought to be of Herpes Simplex Virus (HSV) origin.

Case: A 41-year-old male with a history of recurrent genital HSV infections presented with acute anterior uveitis and concomitant corneal edema. The corneal findings were thought to represent HSV endotheliitis. His clinical course did not improve with oral antiviral medications. The patient developed a thick fibrinous membrane in the anterior chamber resulting in hand motion vision, which improved with oral steroids. Work-up for uveitis resulted in HLA-B27 positivity. The case was complicated by optic nerve head edema; a rare finding that can be present in cases of HLA-B27 related uveitis.

Conclusion: Establishing an etiology for acute presentations of uveitis can be challenging. Reevaluating the patient's response to therapy and adjusting management accordingly is critical to ensure successful care.

HISTORY/EXAMINATION

A 41 year-old male presented to the emergency room for right eye pain and decreased vision. The left eye was asymptomatic. The patient had a past medical history of a recurrent genital herpes simplex virus (HSV) infection and was on a prophylactic dose of acyclovir (400mg oral two times daily). On exam, vision was found to be 20/125 (down from a baseline of 20/20) and intraocular pressure was 17mmHg. The cornea was notable for 1+ microcystic edema (MCE), central trace Descemet-folds (D-folds), but staining was negative for dendrites or pseudodendrites. The anterior chamber (AC) had 2-3+ cell with few inferior keratic precipitates (KPs). The iris was notable for posterior synechiae (PS) temporally with poor dilation. The left eye was quiet but notable for old inferior corneal neovascularization (KNV) along the limbus from 6 to 8 o'clock. The patient was started on a treatment dose of oral acyclovir at 400mg 5x/day, topical prednisolone forte 4x/day (QID), and topical cyclopentolate 3x/day (TID) in the right eye only.

The patient was seen the next day in clinic where vision was found to have improved to 20/100, IOP 14, but still with significant anterior chamber inflammation. The prednisolone forte was increased to 6x/day.

The patient followed back up in clinic 1 week later. Vision was found to be hand motion (HM) and IOP was 21. The corneal MCE was improving and there was resolution of the D-folds. The anterior chamber had 3-4+ cell with a thick fibrinous membrane in front of the pupil. B-scan ultrasonography was negative for retinal detachment or vitritis. Acyclovir was switched to valacyclovir 1000mg TID and prednisolone forte was switched to difluprednate QID. A uveitis workup was initiated. 60mg of oral prednisone was started the next day after negative syphilis testing and a negative chest x-ray.

Two days after starting oral prednisone, the patient returned to clinic for repeat examination. Vision improved to 20/50 with resolution of the thick fibrin membrane in the anterior chamber and broken PS (Figure 1). The uveitis panel returned positive for HLA-B27. Unfortunately, the patient was not able to tolerate the oral steroids so this was tapered off. Difluprednate was continued, but the valacyclovir was discontinued and the patient was restarted on his prophylactic acyclovir.

One week later, the patient returned to clinic with complaint of seeing brief flashes of light lasting roughly 10 seconds in duration each episode. On exam, vision had decreased to 20/70. The AC cell continued to decrease. The posterior exam was notable for grade 3 optic nerve head edema. Optical coherence topography of the nerve fiber layer (OCTnfl) showed significant thickening (Figure 2). Given the new optic nerve head edema, the patient was restarted on oral prednisone 20mg per day. On follow up exam, the nerve head edema was improving (Figures 3) and the vision improved to 20/20 with resolution of the transient visual obscurations (TVOs). The patient was referred to rheumatology for further workup of the HLA-B27 positive testing.

DISCUSSION

Given this patient's history and presentation, there was initial concern for an HSV-related process. Supporting findings include the corneal changes consistent with an endotheliitis in the affected eye and old KNV in the unaffected eye. Additionally, the patient had a prior history of recurrent genital HSV (with sporadic adherence to prophylactic acyclovir). HSV is also a common cause of unilateral anterior uveitis. However, the patient was found to have a normal IOP on presentation, whereas HSV usually causes an ocular hypertensive response. The other notable finding was the presence of PS, which could have indicated prior episodes of inflammation. The patient denied a prior history of similar symptoms, suggesting possible mild phases of anterior uveitis that were not clinically symptomatic to the patient, another finding inconsistent with HSV.

A week later, the patient's vision significantly worsened to HM with the development of a dense fibrotic membrane in front of the pupil. This finding is uncharacteristic of HSV, especially while on a treatment dose of acyclovir. However, with the prior history of recurrent genital HSV already prophylactically being treated with acyclovir, this new ocular process could have represented a different strain of virus that was resistant to acyclovir. Thus, the acyclovir was switched to

valacyclovir. Meanwhile, a full uveitis workup was initiated, as the disease process appeared to be less likely of HSV origin.

The uveitis panel resulted in a positive test for HLA-B27, which better correlated with the presentation and clinical course. HLA-B27 related acute anterior uveitis presents almost exclusively in a unilateral fashion¹. Due to the severity of the inflammation, PS has been reported to occur in up to 13% of patients during the active phase of the disease, which explains the PS noted on presentation²⁻³. Fibrinous exudate in the anterior chamber may block or occlude the pupil, as seen in our case that led to HM vision¹. However, an unusual finding in this case was the development of optic nerve head edema. While the most typical ocular manifestation of HLA-B27 is anterior uveitis, posterior segment involvement during phases of acute anterior uveitis has been reported in the literature. In one study that looked 166 patients with HLA-B27 uveitis, 17.4% had posterior segment involvement. Papillitis occurred in 82.7% of those patients^{1,4}.

The patient's disease process responded well to oral prednisone, but, unfortunately, the patient was unable to tolerate high doses. After the development of optic nerve head edema, the patient was recommended to restart oral prednisone at a lower dose with close follow up. This resulted in improvement in visual acuity and an objective reduction in edema on OCTnfl.

CONCLUSION

Finding a cause for acute anterior uveitis is often difficult. This case demonstrates how important it is to reevaluate the patient's findings when the disease process does not respond appropriately to therapy. Although the initial corneal findings and past medical history were concerning for HSV, the development of the thick fibrinous membrane triggered concern for a different etiology. Furthermore, even with the finding of a positive HLA-B27 test, the clinical course was atypical with the development of optic nerve head edema. Therefore, it is important to keep a working differential diagnosis at all times.

REFERENCES

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FIGURES

Figure 1. External photograph of the right eye shows an irregular pupil secondary to posterior synechia.

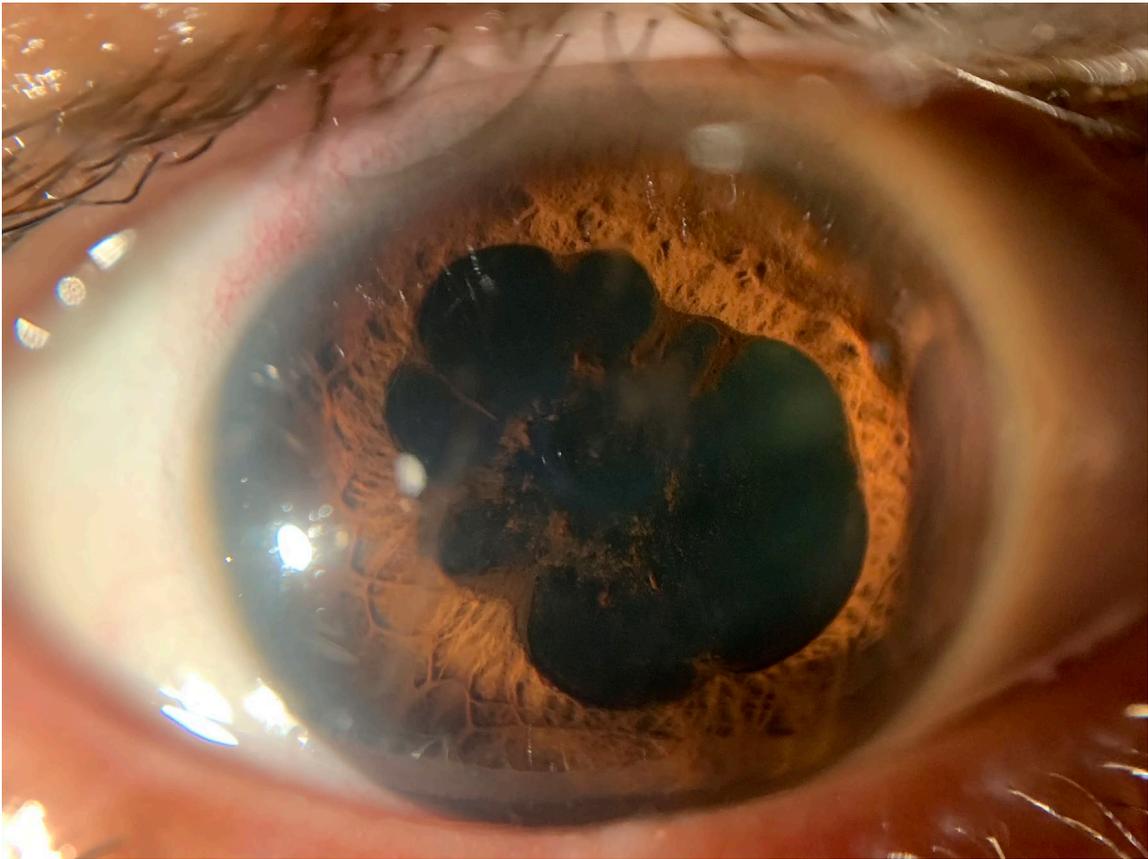


Figure 2. OCTnfl of the right optic nerve head showing edema.

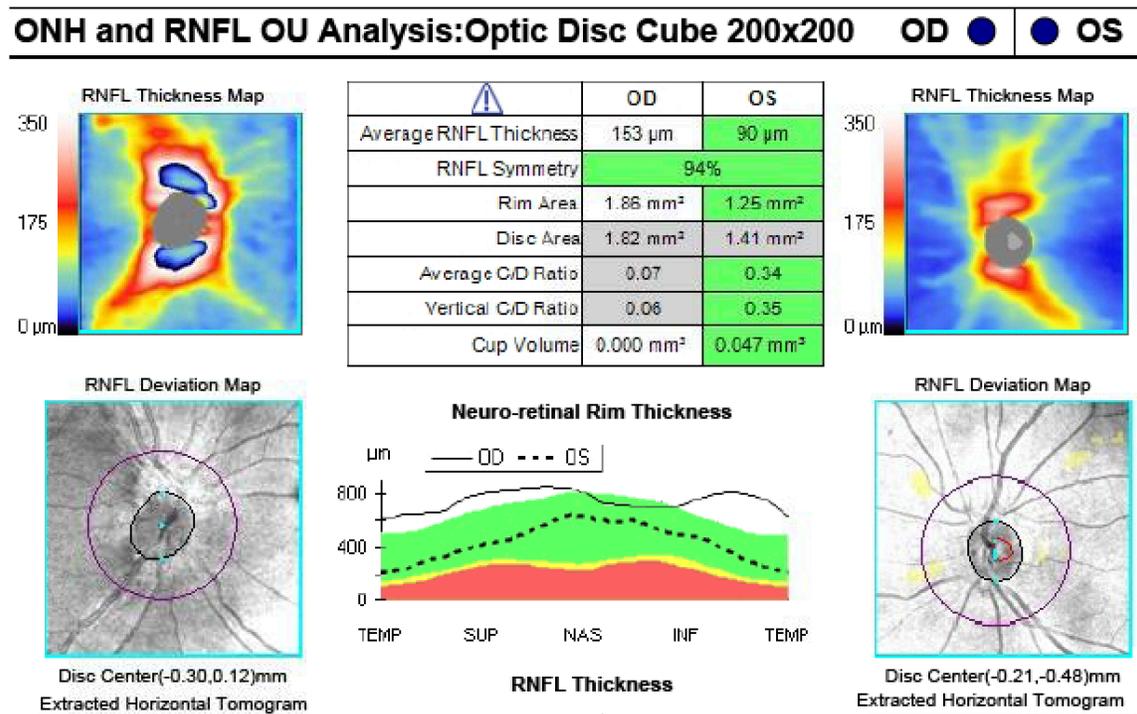


Figure 3. OCTnfl of the right optic nerve head showing improvement in edema.

