You can C-ME after Uveitis

Abstract:
Approximately 50% of uveitis patients will present with vision loss secondary to cystoid macular edema[1]. Two patients with uveitis present with a constant decrease in BCVA over the course of one month.

I. Case History

- Patient demographics:
  - *ML: 60 year old male African American
  - *RJ: 52 year old, male African American
- Chief complaint:
  - *ML: left sided headache, injected OS, blurred vision inferiorly, OS, nausea, photophobia
  - *RJ: Red, painful eye (8/10) OD, photophobia, blurred vision OD
- Ocular/Medical History:
  - *ML: Pigment Dispersion Syndrome, Hypertension
  - *RJ: Recurrent Orbital Pseudotumor OD, Ocular Hypertension, Hypertension
- Medications:
  - *ML: cetirizine, cyclobenzaprine, olopatadine
  - *RJ: Amlodipine, timolol, sildenafil, hydrochlorothiazide

II. Pertinent Findings

*ML:

Visit 1

*Visual Acuity cc: 20/20 OD, **20/25-2 OS**
*PERRL, (-)APD, FROM OU, pain on levoversion, no diplopia or jaw claudication
* SLE
AC: deep/quiet OD, 3+ cells/2+ flare OS, (-)KPs
L: 2+ diffuse pigment on lens OS
Macula: clear OU, (-)CME

Visit 2 (2 days later)

*Visual Acuity cc: 20/20 OD, **20/20 OS**
*PERRL, (-)APD, FROM OU, (-)pain on eye movement
* SLE
AC: deep/quiet OD, 2+ white cells, 2+ pigmented cells, 1+ flare OS, (-)KPs

Visit 3: (5 days later)

*Visual Acuity cc: 20/20 OD**, 20/40 OS**
AC: deep/quiet OD, 1+ white/pigmented cells OS, tr flare OS (-)KPs
Macula: clear OD, **(+)CME OS**

Macular OCT
OS: central subfield thickness: **404 um**
(+)cystic spaces

Visit 4 (11 days later)

*Visual Acuity cc: 20/20 OD**, 20/50 OS**
* SLE
AC: deep/quiet OD, 1+ cells, (-)flare, (-)KPS
Macula: clear OD, **(+)CME OS**

Macular OCT
OS: central subfield thickness: **462 um**
(+)cystic spaces

*RJ:

Visit 1:

*Visual Acuity sc: **20/60 OD,** 20/20 OS
*PERRL, (-)APD, FROM OU
* SLE
C/S: moderate diffuse injection OD, clear OS
AC: 3+ cells OD, deep/quiet OS
Macula: clear OU, **(-)CME**

Visit 2 (3 days later)

*Visual Acuity sc: **20/50 OD,** 20/20 OS
* SLE
C/S: tr injection OD
AC: 1-2 cells OD
Macula: **(+)CME OD**

Macular OCT
OD: central subfield thickness: **506 um**
(+)cystic spaces

Visit 3 (5 days later)

*Visual Acuity sc: **20/60 OD,** 20/20 OS
* SLE
C/S: tr injection OD
AC: 1-2 cells OD
Macula: **(+)CME OD**

Macular OCT
OD: central subfield thickness: **543 um**
(+)cystic spaces

Visit 4 (9 days later)

*Visual Acuity sc: **20/25-2 OD, 20/20 OS**
* SLE
C/S: tr injection OD
AC: deep/quiet OU

Macula: **(+)CME OD**

Macular OCT
OD: central subfield thickness: 359 um
(+)cystic spaces

Visit 4 (14 days later)

*Visual Acuity sc: **20/20-1 OD, 20/20 OS**
* SLE
C/S: white/quiet OU
Macula: **(+)CME OD**

Macular OCT
OD: central subfield thickness: 300 um
**(+)minimal cystic spaces

III: Differential Diagnosis

Primary:

- Cystoid macular edema secondary to uveitis

Others:

- Ischemic optic neuropathy
- Optic neuritis
- Choroidal neovascularization membrane
- Granulomatous uveitis

IV: Diagnosis:

Cystoid macular edema (CME) is a major cause of reduced vision in patients with uveitis. In fact, CME occurs in approximately 50% of all patients presenting with uveitis [1]. One out of four patients with anterior uveitis develop CME. [1]. During a uveitic episode, the human body responds by releasing inflammatory mediators. These mediators, via the cyclooxygenase and lipooxygenase pathway, include leukotrienes, thromboxanes, prostacyclins, and prostaglandins. Other mediators include TNF-α and IL-1 β [2], which are specific cytokines that rupture the blood-retinal barrier. By disrupting the blood-retinal barrier, specifically the tight junctions of the zonula occludens, adherens and the macula adherens the integrity of the retinal-pigmented epithelial layer becomes compromised. The RPE serves as a barrier for the retina and maintains fluid exchange between the vitreous cavity and choroid [2] via the enzyme carboanhydrase. Due to the dysfunctional Mueller cells of the retina and the inability to stay dehydrated, the macular layers become thickened and edematous ultimately leading to necrosis of these layers if left untreated.

IV: Treatment and management

The presence of decreased visual acuity occurs several weeks after the onset of acute anterior uveitis as seen with the patients above. Patients with intermediate or posterior uveitis present sooner and more frequently with CME[2]. Patients with cystoid macular edema can be diagnosed via fluorescein angiography and/or optical coherence tomography (OCT). On an OCT, diagnosis is made visually by noting

cystic, hyporeflective spaces separated by hyperreflective bridges. On FA, the dye pools in the outer plexiform layer forming a hyperfluorescent petalloid or honeycombed appearance. [1]. After confirmation of cystoid macular edema, treatment varies depending on etiology and the persistence of uveitis and CME. If CME is present with a posterior uveitis, steroid injections periocularly, intravitreally or systemically has been proven to be effective [3]. In addition, anti-vegf and immunomodulatory drugs have been proven to reduce the inflammatory process and increase best corrected visual acuity. In cases of mild CME, topical NSAIDs along with topical steroids are effective [3]. Another viable option is vitrectomy, which removes much of the inflammatory debris from the vitreous. The incidence of visual improvement following CME after uveitis is promising. The estimated 6 month incidence of ≥2 lines VA improvement was 52% [4]. ML was referred to a retinal specialist for intravitreal steroid injection. However, ML's visual acuity improved to 20/25 OS while waiting to be seen by the retinal specialist in one week. The retinal specialist recommended to continue prednisolone and acular qid OS. RJ was prescribed prednisolone and acular qid OD that improved vision 5 lines in four weeks.

V: Clinical Pearls

If a patient complains about reduced vision after a uveitic episode, consider an OCT or FA to rule out cystoid macula edema. If CME is present, consider initiating a topical NSAID and steroid regimen before referring patient to retinal specialist. If no improvement with 1-2 weeks, consider a referral.

References

[1]Karim, Rushmia. "Interventions for the Treatment of Uveitic Macular Edema: A Systematic Review and Meta-analysis." *Clinical Ophthalmology* June (2013): 1-36. *DovePress Journal*. Web.

[2]Okhravi, Narciss, and Susan Lightman. "Cystoid Macular Edema in Uveitis." *Ocular Immunology and Inflammation* II (2003): 29-38. Web.

[3]Guex-Crosier, Y. “The Pathogenesis and Clinical Presentation of Macular Edema in Inflammatory Diseases.” *Documenta Ophthalmologica* 97 (1999): 297-309. Web.

[4]Levin, Marc H., and Maxwell Pistilli. "Incidence of Visual Improvement in Uveitis Cases with Visual Impairment Caused by Macular Edema." *Ophthalmology* (2013): n. pag. Web.