Progression of Unilateral Birdshot Chorioretinopathy to Bilateral Disease

Kim, Isaac[1]; Huang, Natalie[2]; Swan, Robert[2]

1. SUNY Upstate Medical University, College of Medicine, Syracuse NY 2. SUNY Upstate Medical University, Department of Ophthalmology & Visual Sciences, Syracuse NY

Introduction

- Birdshot chorioretinopathy is a rare, chronic, recurrent, bilateral, posterior uveitis with a strong genetic association with HLA-A29
- Most prevalent in middle-aged Caucasian population with a slight female predominance
- Pathogenesis is unclear; there are several proposed theories
- Symptoms include decrease of central vision, floaters, photopsia, dyschromatopsia, and nyctalopia
- Fundoscopic exam reveals cream-colored or hypopigmented, round or oval, choroidal lesions that may occur years after symptoms
- Other clinical signs include vitritis without snowballs or snow-banking, cystoid macular edema, optic disc edema, and retinal vasculitis

Case Presentation

- 67-year-old man with past medical history significant for Alzheimer Disease presented to an outside office with new floaters and intermittent blurry vision in the left eye for 8 weeks
- Visual acuity, pressures, pupils, and slit lamp exam were unremarkable with no afferent pupillary defect in either eye
- Fundus examination of right eye was normal; left eye showed mild vitreous haze, peripapillary vasculitis with small hemorrhage, and a hyperpigmented lesions
- Laboratory work-up remarkable for positive HLA-A29
- Presented to our clinic 5 months later; repeat fluorescein angiography of the left eye showed periphlebitis, papillitis, and cystoid macular edema
- Indocyanine Green of the left eye showed scattered peripheral hypocyanescent spots in the late phase
- Diagnosis of unilateral birdshot chorioretinopathy was made
- Systemic therapy with mycophenolate mofetil and cyclosporine were started but discontinued two months later due side effects
- Local therapy with intravitreal dexamethasone implant was used
  - 10th month visit – OS 11th month visit – OD
- Near complete resolution of inflammation observed in both eyes

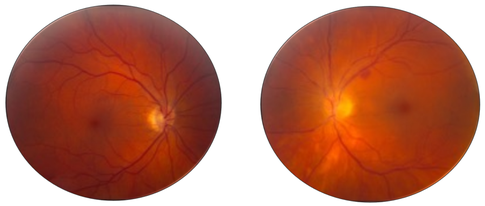

Figure 1. Color fundus photo at presentation to outside office.
Mild vitreous haze, peripapillary vasculitis, small hemorrhage, and hyperpigmented lesions seen in the left eye.

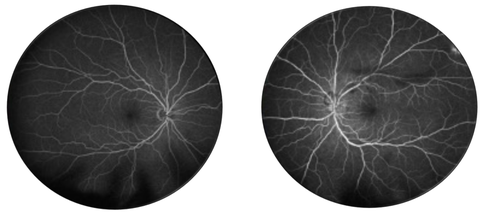

Figure 2. Fluorescein angiography photo at presentation to outside office
Blockage of fluorescence in the area of hemorrhage seen in the left eye.

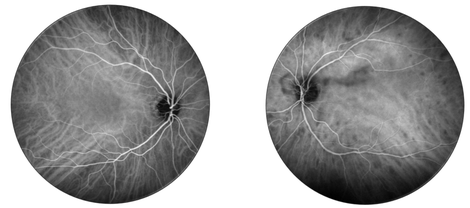

Figure 3. Indocyanine green at initial visit to our clinic.
Scattered hypocyanescent spots in the periphery of the left eye 7 months after initial symptoms of floaters and blurry vision.

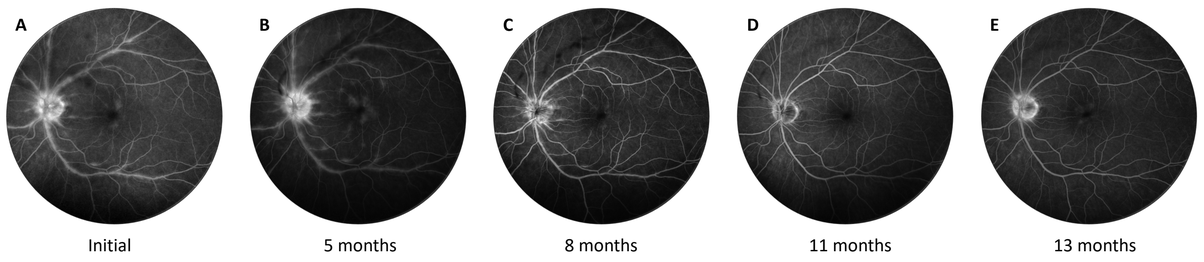

Figure 4. Fluorescein angiography of left eye
- Extensive hyperfluorescence along the veins and optic nerve (A) with worsening in the perifovea while two months into systemic therapy (B).
- Improvements seen 3 months later (C) while off systemic therapy.
- Near complete resolution of inflammation six weeks after local therapy (D).

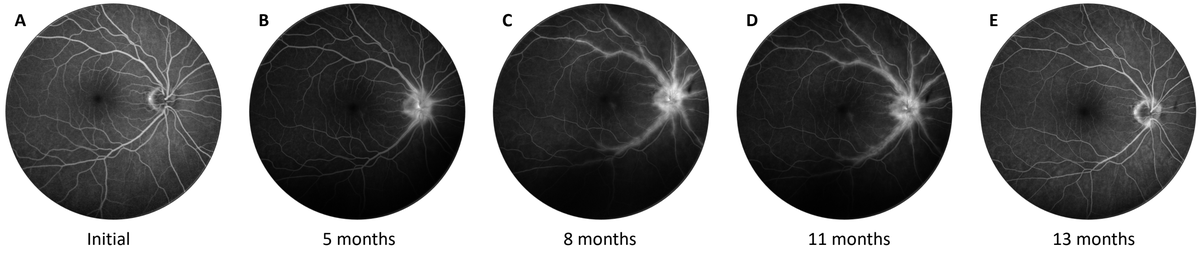

Figure 5. Fluorescein angiography of right eye
- Normal perfusion of right eye at initial visit 7 months after initial symptoms (A).
- Slight hyperfluorescence of the optic nerve noted while two months into systemic therapy (B), progressing to show extensive hyperfluorescence of the veins and optic nerve 3 months later while off systemic therapy (C) and another 3 months later (D).
- Near complete resolution of inflammation observed two months after local therapy (E).

Discussion

- A case of BSCR manifested unilateral initially and eventual bilateral involvement with retinal vasculitis as the most prominent clinical feature
- Systemic therapies often include immunomodulatory agents; combination regimen of cyclosporine A and mycophenolate has been associated with long-term control of inflammation[1]
- Local therapy with dexamethasone implant is an effective alternative[2]

References

1. Cervantes-Castañeda, R. A., Gonzalez-Gonzalez, L. A., Cordero-Coma, M., Yilmaz, T., & Foster, C. S. (2013). Combined therapy of cyclosporine A and mycophenolate mofetil for the treatment of birdshot retinochoroidopathy: a 12-month follow-up. *British Journal of Ophthalmology*, *97*(5), 637-643.
2. Walsh, J., & Reddy, A. K. (2017). Intravitreal dexamethasone implantation for birdshot chorioretinopathy. *Retinal Cases and Brief Reports*, *11*(1), 51-55.

This study was funded in part by unrestricted grants from Research to Prevent Blindness, Inc. New York, New York and Lions District 20-Y1, Syracuse, New York. No other significant financial interests or relationships to disclosure

SUPPORTED BY Research to Prevent Blindness