Progression of Unilateral Birdshot Chorioretinopathy to Bilateral Disease

Kim, Isaac1; Huang, Natalie2; Swan, Robert2
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
- Fundoscopy reveals cream-colored or hypopigmented, round or oval, choroidal lesions that may occur years after symptoms
- Other clinical signs include vitritis without snowballs or snowbanking, 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 lesion
- Laboratory work-up remarkable for positive HLA-A29
- Presented to our clinic 5 months later; repeat fluorescein angiography of the left eye showed scattered peripheral hypocyanescent spots in the late phase
- Diagnosis of unilateral birdshot chorioretinopathy was made
- Systemic therapy with cyclosporine A and mycophenolate mofetil has been associated with long-term control of inflammation
- Extensive improvements seen 3 months later (B) while off systemic therapy (A) and another 3 months later (D).
- Near complete resolution of inflammation observed two months after initial symptoms of floaters and blurry vision.

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
- Local therapy with dexamethasone implant is an effective alternative

References

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