Unilateral Birdshot Chorioretinopathy in an Elderly Patient

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INTRODUCTION

Birdshot chorioretinopathy (BSCR) is a poorly understood bilateral posterior uveitis strongly associated with the human leukocyte antigen (HLA)-A29. Herein, we report an unusual case of unilateral BSCR in an elderly patient.

CASE

A 69-year-old male was referred for evaluation of focal hypopigmented choroidal lesions in the right eye. The lesions were unchanged when compared to fundus photography from a previous eye exam three years earlier; however, the patient complained of vague right eye discomfort prompting referral. Past ocular history was notable only for refractive error/presbyopia. Past medical history included hypertension, hyperlipidemia, and prostate cancer undergoing radiation. On exam, visual acuity was 20/20 bilaterally with normal intraocular pressures and no afferent pupillary defect. Slit-lamp biomicroscopy was notable for 1+ nuclear sclerotic cataracts. On dilated fundus exam, there were cream-colored choroidal lesions within the posterior pole extending to the mid-periphery of the right eye; no vitreous cells, disc edema, vascular sheathing or cystoid macular edema was present. The left eye fundus was unremarkable (**Figure 1**). Optical coherence tomography and fluorescein angiography were unremarkable bilaterally. On indocyanine green angiography, there were hypocyanescent choroidal lesions within the posterior pole and mid-periphery of the right eye, measuring ¼ to ½ disc diameter in size (**Figure 2**); left eye was unremarkable. Additional testing included a normal CBC, a non-reactive RPR/FTA-ABS, normal ACE/lysozyme, positive HLA-A29 and normal MRI orbits and brain. An electroretinogram (ERG) was unremarkable. A hematology consultation was obtained; however, given the normal CBC, the low suspicion for indolent lymphoma or Waldenström's macroglobulinemia, no further work-up was recommended. Given the

Figure 1. Color fundus photography exhibited hypopigmented choroidal lesions within the posterior pole of the right eye and a normal fundus appearance of the left eye.

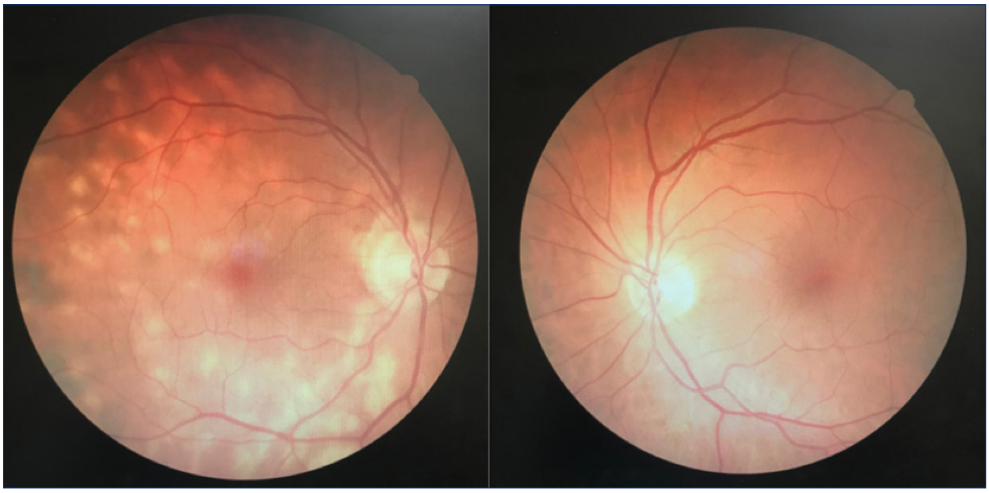
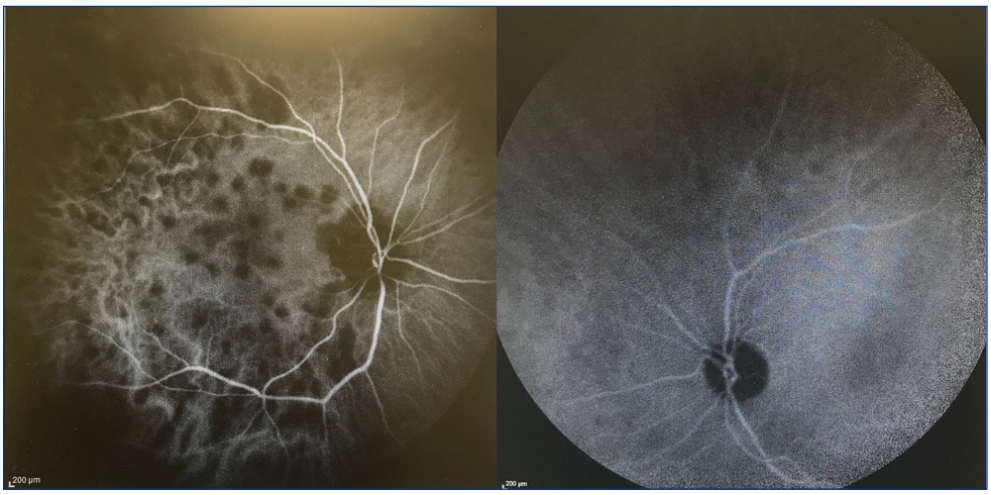


Figure 2. Indocyanine green angiography exhibited prominent hypocyanescent choroidal lesions within the posterior pole extending into the mid-periphery of the right eye; the left eye was unremarkable.



patient's asymptomatic status, excellent visual acuity and lack of inflammation, the patient was observed.

DISCUSSION

Birdshot chorioretinopathy classically occurs in middle-aged Caucasian individuals and presents as bilateral choroiditis in the presence of ≤1+ anterior vitreous cells, ≤2+ vitreous haze and ≥3 peripapillary ill-defined choroidal lesions.[1,2] Although our patient's exam and imaging are similar to previously reported cases[3,4], the monocular aspect of this case is atypical and warranted evaluation for an alternate etiology, including sarcoidosis and syphilis. Primary intraocular lymphoma was also considered but was deemed unlikely in the setting of absent vitritis and unremarkable serology.

In 2013, Zucchiatti et al. reported 18 cases of HLA-A29 positive uveitis; four patients exhibited unilateral disease.[5] Three of these four patients presented with keratic precipitates and were subsequently diagnosed as Fuch's uveitis, granulomatous panuveitis and HSV panuveitis, respectively. The fourth patient was diagnosed as scleritis and displayed no retinal abnormality. The asymptomatic nature of our patient's retinopathy suggests a mild BSCR variant, which may underlie the unilaterality of the findings.

Birdshot chorioretinopathy can result in vision loss secondary to cystoid macular edema and slow progression of chorioretinal atrophy.[1,4] Therefore, early diagnosis and identification of complications is crucial to insure timely treatment with intraocular or systemic immunosuppression.[6] Patients should be monitored for progression of disease with repeat ocular imaging, electroretinogram (ERG) and/or visual field testing as progression often occurs despite lack of inflammation on exam.[6] This case suggests that BSCR should remain a consideration in HLA-A29 positive individuals who present with unilateral creamy peripapillary choroidal lesions with or without inflammation.

References

1. Cunningham, E., Levinson, R., Denniston, A., Brézin, A. and Zierhut, M. (2017). Birdshot Chorioretinopathy. Ocular Immunology and Inflammation, 25(5), pp.589-593.
2. Levinson, R., Brezin, A., Rothova, A., Accorinti, M. and Holland, G. (2006). Research Criteria for the Diagnosis of Birdshot Chorioretinopathy: Results of an International Consensus Conference. American Journal of Ophthalmology, 141(1), pp.185-187.
3. Teussink, M., Huis in het Veld, P., de Vries, L., Hoyng, C., Klevering, B. and Theelen, T. (2016). Multimodal imaging of the disease progression of birdshot chorioretinopathy. Acta Ophthalmologica, 94(8), pp.815-823.
4. Young, M., Fallah, N. and Forooghian, F. (2015). Choroidal degeneration in birdshot chorioretinopathy. Retina, 35(4), pp.798-802.
5. Zucchiatti, I., Miserocchi, E., Sacconi, R., Bandello, F. and Modorati, G. (2013). HLA-A29-Positive Uveitis: Birdshot Chorioretinopathy, What Else. Case Reports in Ophthalmology, 4(3), pp.287-293.
6. Minos E, Barry RJ, Southworth S, Folkard A, Murray PI, Duker JS, Keane PA and Denniston AK. Birdshot chorioretinopathy: current knowledge and new concepts in pathophysiology, diagnosis, monitoring and treatment. Orphanet J Rare Dis. 2016 May 12;11(1):61.

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