

Atypical Unilateral Birdshot Chorioretinitis in a Hispanic Female

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A 62-year-old female patient was evaluated for gradual vision loss, floaters, and photopsia in her left eye. A left fundus examination revealed vitreous cells, hypopigmented lesions, and retinal vasculitis, and a workup revealed a positive HLA-A29 serology, all of which findings were consistent with birdshot chorioretinopathy. The patient was treated with oral prednisone and mycophenolate mofetil, which resulted in the adequate control of her uveitis. Even though unilateral cases are exceedingly rare and do not meet the established research criteria, this case highlights the importance of not minimizing the significance of birdshot lesions in the differential of patients with unilateral multifocal chorioretinitis. [*P R Health Sci J* 2023;42(2):180-182]

Key words: Birdshot chorioretinopathy, Hispanics, HLA-A29, Posterior uveitis

Birdshot chorioretinopathy (BSCR) is an autoimmune, most commonly bilateral, form of posterior uveitis with a distinct clinical presentation (1). It is characterized by the presence of multiple hypopigmented, ovoid-shaped, choroidal lesions that are usually clustered near the optic disc and in the inferonasal mid-periphery (1,2). This disease has a strong association with the HLA-A29 serotype, which has been reported in nearly 100% of the patients with BSCR; it is a supportive finding of the diagnosis, but it is not a required characteristic for the disease's diagnostic criteria (1,2).

It (BSCR) represents 6 to 7.9% of posterior uveitis cases, has a slight female predominance, and most commonly occurs in patients from 35 to 70 years of age (1). The differential diagnosis of BSCR includes infectious disease-associated posterior uveitis, such as syphilis and tuberculosis (1). All patients with posterior uveitis should be worked up with a treponema-specific test as luetic uveitis can mimic most uveitis entities yet represents a treatable and curable form of posterior uveitis (3). Tuberculosis screening is also advised (1,3).

Birdshot chorioretinopathy is commonly described as a bilateral disease (1,2). However, unilateral forms of the disease are known to exist, although they are exceedingly rare (3,4). Cohort studies by Gasch et al. and Thorne et al. have reported prevalences of unilateral disease of 1.7% and 5%, respectively (3,4). Conversely, unilateral BSCR do not fulfill the BSCR diagnostic criteria for research established by Levinson et al., potentially excluding these cases from the medical literature (2). Although they do not comply with these diagnostic criteria, unilateral cases are undoubtedly BSCR, and therapy with immunosuppressive medications leads to a better prognosis (5). We present the case of a 62-year-old Puerto Rican female with unilateral BSCR, intending to create awareness of this lesser-known variant (5).

Case Report

A 62-year-old Hispanic female patient presented to the clinic with gradual vision loss, floaters, and photopsia in her left eye. She had no history of recent travel or systemic illnesses, and her review of systems was unremarkable. The physical examination revealed a corrected visual acuity of 20/25 in the right eye and 20/20 in the left eye. The intraocular pressure was normal in both eyes. A slit-lamp examination revealed minimal lenticular changes and deep and quiet anterior chambers and was otherwise unremarkable.

A dilated fundus examination was within normal limits in the right eye. The left fundus revealed 2+ vitreous cells, perivenular sheathing, and subtle hypopigmented lesions in the inferonasal mid-periphery. Neither eye had macular edema. A workup revealed a positive HLA-A29 serology. Additional corresponding tests, including a chest X-ray, magnetic resonance imaging of the brain (with contrast), a fluorescent treponemal antibody absorption test, an antinuclear antibody test, an antineutrophil cytoplasmic antibodies test, and a purified protein derivative test, were all negative. Baseline fluorescein and indocyanine green angiograms were performed; however, the patient suffered an anxiety attack during the procedure and it was halted, not allowing for adequate angiographic images. A comprehensive

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physical examination by a board-certified rheumatologist did not reveal any systemic evidence of infectious or autoimmune disease.

An assessment of unilateral BSCR was made, and the patient was treated with mycophenolate mofetil 1.5 g (po, bid) and prednisone 60 mg (po, qd), which made the disease inactive. Subsequently, while continuing the mycophenolate mofetil, the prednisone was slowly tapered over several months, during which tapering, a disease flare occurred at the prednisone dose of 7 mg. The flare was treated by increasing the prednisone dose back to 60 mg until the patient's BSCR became inactive and subsequently tapering the prednisone to 8 mg. Subsequent trials of tapering (at 12 and 36 months) the prednisone below a dose of 8 mg while on the concurrent treatment with mycophenolate mofetil were unsuccessful. At 49 months follow-up, cystoid macular edema was detected in the left eye upon tapering the prednisone to 7 mg. Upon the performance of fluorescein and indocyanine green angiograms following the emergence of the cystoid macular edema, the left eye revealed the presence of perivascular hyperfluorescence and hypocyanescent spots, respectively, all compatible with the diagnosis of active BSCR (Fig. 1A, 1B, and 1C). The option of adding adalimumab as a second steroid-sparing agent was offered to the patient; however, she declined. She preferred to continue on mycophenolate mofetil 1.5 g (po, bid) and prednisone 8 mg (po, qd) on a chronic basis.

Discussion

The research criteria for the diagnosis of BSCR include bilateral disease, the presence of at least 3 peripapillary "birdshot lesions" inferior or nasal to the optic disc in 1 eye, low-grade anterior segment intraocular inflammation, and low-grade vitreous inflammatory reaction (2). However, this patient presented with unilateral low-grade vitreous inflammation, hypocyanescent birdshot lesions, and HLA-A29 positivity. Therefore, according to Levinson et al., this patient would not meet the research criteria for the disease due to her unilateral presentation (2). However, the research diagnostic criteria for BSCR are not meant to be 100% sensitive (2). For instance, in Thorne et al.'s cohort, 2 patients did not present with bilateral disease; this highlights the fact that unilateral BSCR cases exist in the medical literature (4).

Patients with BSCR may suffer irreversible central vision loss from recurrent cystoid macular edema and autoimmune-mediated retinal dysfunction, the latter of which may occur even in the absence of macular edema (5). Treatment with systemic immunosuppressors has been associated with a decreased risk of developing cystoid macular edema and the reversal of peripheral visual field constriction in BSCR (5,6). Treatment with systemic corticosteroids alone does not render such benefits as treatment with systemic immunosuppressors does (5). Therefore, it is ideal to identify all cases of BSCR, including unilateral ones, to provide adequate therapy and management.

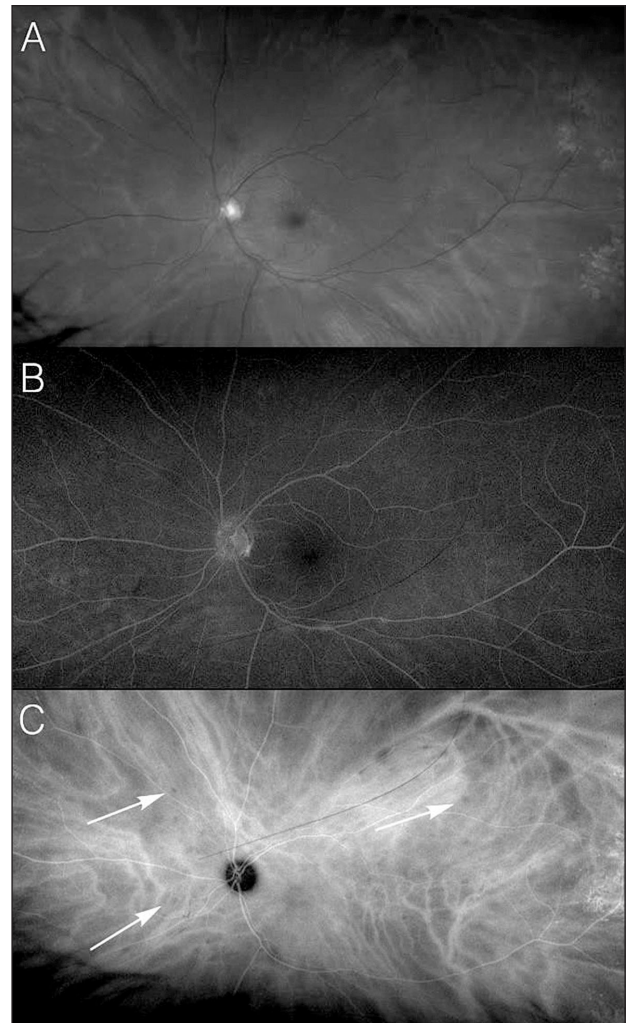


Figure 1. Ultra-widefield fundus imaging of the left eye on presentation. A) Color photographs show several subtle hypopigmented lesions in the inferonasal periphery. B) A late-phase intravenous fluorescein angiogram revealed mild perivenular leakage. C) Late-phase indocyanine green angiography revealed multiple hypocyanescent spots in the nasal and superotemporal mid-periphery (arrows).

Resumen

Presentamos el caso de una paciente de 62 años evaluada por la pérdida gradual de visión, flotadores y fotopsia en su ojo izquierdo. El examen del fondo del ojo izquierdo reveló células vítreas, lesiones hipopigmentadas y vasculitis retiniana. Un estudio reveló una serología de HLA-A29 positiva, lo que era compatible con una coriorretinopatía en perdigonada (birdshot). La paciente fue tratada con prednisona oral y micofenolato de mofetilo, lo que permitió un control adecuado de su uveítis. Aunque los casos unilaterales son extremadamente raros y no cumplen con los criterios de investigación establecidos, este caso subraya la importancia de tener en cuenta el "birdshot" en el diferencial de los pacientes con coriorretinitis multifocal unilateral.

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