International Medical Case Reports Journal

Dovepress
Taylor & Francis Group

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CASE REPORT

Intravitreal Corticosteroids in the Management of Refractory Macular Edema in Birdshot Chorioretinopathy

João Alves Ambrósio, Catarina Pestana Aguiar, Pedro Cardoso Teixeira, Vítor Miranda, João Chibante Pedro, Miguel Ruão

Ophthalmology Department, Unidade Local de Saúde Entre Douro e Vouga, Santa Maria da Feira, Portugal

Correspondence: João Alves Ambrósio, Ophthalmology Department, Unidade Local de Saúde Entre Douro e Vouga, Rua Dr. Cândido Pinho 5, Santa Maria da Feira, 4520-211, Portugal, Email alvesambrosio.joao@gmail.com

Introduction: Birdshot chorioretinopathy (BCR) is a chronic, bilateral posterior uveitis characterized by yellow-white fundus lesions and a strong association with HLA-A29. Visual decline, often due to cystoid macular edema (CME) and retinal atrophy, necessitates early immunomodulatory therapy. This case report describes the clinical course of BCR and highlights the role of intravitreal corticosteroids in managing inflammation and CME.

Case Report: A 54-year-old previously healthy male diagnosed with BCR based on clinical findings and a positive HLA-A29 test presented with refractory CME. Over 20 months, his best-corrected visual acuity (BCVA) and central foveal thickness (CFT) were monitored. Initial treatment included topical corticosteroids, methotrexate, and oral corticosteroids to address anterior chamber reaction, vitritis, diffuse retinal lesions, and vasculitis. Cyclosporine was added for persistent inflammation but discontinued due to a cutaneous reaction. Despite these efforts, CME persisted, necessitating intravitreal corticosteroids. BCVA in the right eye (OD) fluctuated between 20/20 and 20/30, while the left eye (OS) ranged from 20/20 to 20/40, with changes linked to treatment adjustments. Recurrent CME episodes were more pronounced in the OS, where CFT varied from 328 to 637 µm, while OD values ranged from 304 to 576 µm. Intravitreal dexamethasone and fluocinolone implants reduced CFT in both eyes, achieving stabilization at the final assessment (OD 341 µm, OS 347 µm).

Conclusion: This case illustrates the challenges of managing BCR with refractory CME. While systemic immunomodulatory therapy is foundational, intravitreal corticosteroids play a vital role in controlling CME and preserving visual function. Combining systemic and local therapies proved essential for disease control. Long-term monitoring and individualized treatment are critical in managing this chronic condition.

Plain Language Summary: Birdshot chorioretinopathy (BCR) is a rare eye condition that causes inflammation in the posterior segment of the eye and can lead to vision problems, often due to fluid buildup in the retina (called cystoid macular edema, or CME). Early treatment is crucial to prevent long-term damage to eyesight. This report describes how a combination of treatments helped a 54-year-old man manage BCR and maintain his vision. The patient, previously healthy, was diagnosed with BCR after experiencing symptoms such as vision changes and inflammation in both eyes. Initial treatments included medications like methotrexate and corticosteroids to reduce inflammation. Despite these efforts, fluid buildup (CME) persisted, affecting his vision. To address this, doctors used injections of corticosteroids directly into the eye, which significantly reduced the fluid and helped stabilize his vision. By the end of treatment, both eyes showed improvement in retinal thickness and overall visual function. This case highlights the importance of combining systemic (whole-body) and local (eye-specific) treatments to manage BCR effectively. It also emphasizes the need for ongoing monitoring and tailored therapy, as this is a chronic condition requiring careful management to preserve eyesight.

Keywords: autoimmune diseases, white dot syndromes, chorioretinitis, HLA-A29 antigen, birdshot chorioretinopathy

Received: 22 December 2024
Accepted: 3 March 2025
Published: 30 March 2025

Introduction

Birdshot chorioretinopathy (BCR) is a chronic intraocular inflammatory disease of unknown origin, accounting for 0.6–1.5% of all uveitis cases.[1,2] Its hallmark ocular features include bilateral vitritis and distinctive yellow-white, hypopigmented choroidal lesions, typically occurring without severe anterior segment inflammation.[3,4] As a presumed organ-specific autoimmune condition, BCR exhibits the strongest known association with a human leukocyte antigen (HLA) class I allele. It is strongly linked to HLA-A29, which is present in 98% of BCR cases compared to only 7% of the general population.[4–6] BCR typically manifests in the fifth decade of life, with a slight predominance among females.[3,4] In most patients, the disease follows a chronic and progressive course. The condition often leads to visual decline, primarily due to cystoid macular edema (CME) and retinal atrophy. Recurrent episodes of inflammation in chronic uveitis, if left untreated or inadequately managed, can result in damage to surrounding tissues, further impairing vision.[7] The treatment of noninfectious uveitis focuses on managing inflammation, preventing and addressing complications, achieving disease quiescence, protecting adjacent structures, and preserving or enhancing vision.[8] Current management strategies for chronic noninfectious uveitis of the posterior segment include systemic, local, or topical corticosteroids, either alone or in combination with systemic immunosuppressants. Topical corticosteroids are generally ineffective for intermediate or posterior uveitis due to poor intraocular penetration, while systemic treatments are often limited by significant side effects.[9] The ideal treatment for posterior uveitis, in the absence of systemic disease, targets the disease site while minimizing systemic exposure. It should involve a low corticosteroid dose to reduce ocular side effects, provide long-lasting effects to decrease dosing frequency, and be easy to administer to minimize patient inconvenience and procedure-related side effects. Intravitreal drug delivery systems offer a promising approach to achieving these objectives.[7,9–12]

The 0.19 mg fluocinolone acetonide implant (FA; Iluvien, Alimera Sciences, Hampshire, UK) is a slow-release (0.2 mcg/day), intravitreal, non-biodegradable corticosteroid implant that provides sustained efficacy for up to three years. In 2019, the FA implant was approved in Europe for preventing relapse in recurrent non-infectious uveitis affecting the posterior segment of the eye.[11] Another intravitreal corticosteroid implant approved in Europe for the treatment of posterior segment non-infectious uveitis is the 0.7-mg dexamethasone extended-release implant (Ozurdex; Allergan Pharmaceuticals Ireland, Westport, Ireland), which provides sustained delivery of dexamethasone for up to six months.[12] Recent studies have demonstrated a significant reduction in the recurrence of posterior uveitis following treatment with corticosteroid implants.[7,10–12]

This case report explores the clinical course of BCR and evaluates the role of intravitreal corticosteroids in controlling inflammation and managing macular edema. The long-term management approach is particularly noteworthy, as it required tailored treatment adjustments to accommodate the patient's evolving clinical needs—an aspect that is not often explored in the current literature on this topic.

The case report complies with the guidelines for human studies and was conducted ethically in accordance with the World Medical Association Declaration of Helsinki. Institutional approval was not required. The patient was informed of the delicate nature of his ocular condition and provided written informed consent for the publication of this report.

Case Report

We present the case of a 54-year-old previously healthy male with a one-week history of bilateral myodesopsia. Clinical examination revealed mild anterior segment inflammation, vitritis, yellowish-white lesions scattered throughout the posterior segment, and peripheral vasculitis. A diagnosis of birdshot chorioretinopathy (BCR) was suspected and later confirmed by a positive HLA-A29 test. The patient's clinical course, best-corrected visual acuity (BCVA), and imaging findings—including retinography (Topcon TRC-NW8 non-mydriatic retinal camera) and optical coherence tomography (OCT; Spectralis®, Heidelberg Engineering) with central foveal thickness (CFT) measurements—were closely monitored over a 20-month follow-up period (Figure 1).

The treatment regimen included topical and systemic corticosteroids, methotrexate, cyclosporine, and intravitreal corticosteroid implants (dexamethasone and FA). Initially, topical dexamethasone was started. However, two months later, the patient's initial signs - recurrent bilateral anterior chamber reaction, vitritis, diffuse retinal lesions, and peripheral vasculitis - persisted, prompting the initiation of oral prednisolone at a dose of 60 mg. Three months after the initial presentation, methotrexate was

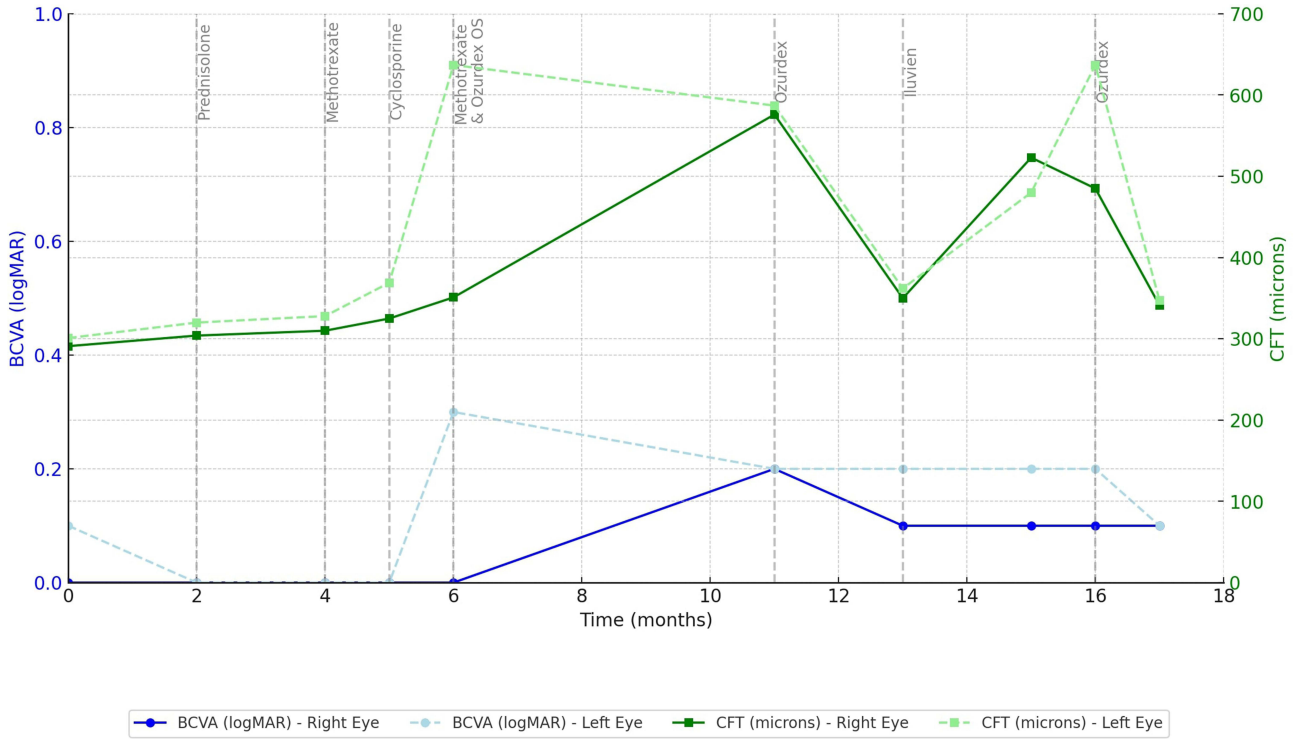

Figure 1 Visual acuity and macular changes over time.
Abbreviations: BCVA, best-corrected visual acuity; CFT, central foveal thickness; logMAR, logarithm of the minimum angle of resolution; OS, left eye. Unless otherwise specified, treatments were administered bilaterally.

prescribed, with the dosage gradually increased while prednisolone was tapered. During the first four months of treatment, BCVA remained stable at 20/20 in both eyes (OU), and macular OCT revealed no significant abnormalities. At the five-month follow-up, mild intraretinal cysts were detected, prompting the initiation of cyclosporine at a dose of 200 mg as a replacement for methotrexate. However, the patient discontinued cyclosporine due to a cutaneous reaction and lymphocytosis. By the six-month follow-up, BCVA in the left eye (OS) had decreased to 20/40, with macular OCT revealing significant macular edema (CFT OS: 637 microns). Methotrexate was then reintroduced and maintained throughout the follow-up period. Despite these interventions, the persistent inflammation and CME necessitated intravitreal corticosteroid therapy. Consequently, a 0.7-mg dexamethasone intravitreal implant (Ozurdex) was administered in the OS. Unfortunately, the patient missed the next scheduled appointment and was evaluated five months later. He reported initial symptom improvement followed by a recurrence that prompted re-evaluation. At the 11-month follow-up, BCVA was 20/30 in OU, with CFT measuring 576 µm in the right eye (OD) and 587 µm in the OS. Given the likely improvement following the Ozurdex implant in the OS, bilateral dexamethasone implants were prescribed. Two months later, BCVA improved to 20/25 in the OD and 20/30 in the OS, while macular edema resolved, with CFT decreasing to 350 microns in the OD and 362 microns in the OS. As a result, bilateral Iluvien intravitreal implants were considered two months after the Ozurdex. At the 15-month follow-up, there was a slight worsening in BCVA to 20/25 in the OD and 20/30 in the OS, accompanied by an increase in CFT to 523 µm in the OD and 480 µmin the OS. At this time point a reevaluation was decided and one month later was a slightly improvement of the CFT in the OD to 485 µm, but a worsening in the OS to 637 µm, prompting a new proposal to bilaterally Ozurdex intravitreal implant. At the final follow-up period there was a stabilization of the CFT (OD 341 µm, OS 347 µm) and BCVA (OD 20/25, OE 20/25). Considering that the Iluvien implants were not removed before the new Ozurdex injection, it is essential to continue monitoring this patient, as long-term cumulative corticosteroid adverse effects may develop.

Figure 2 illustrates the OCT evolution throughout the follow-up period, highlighting the fluctuations in macular edema. Additionally, the formation of an epiretinal membrane (ERM) is noted in OU starting from the 11th month of follow-up.

https://doi.org/10.2147/IMCRJ.S513752

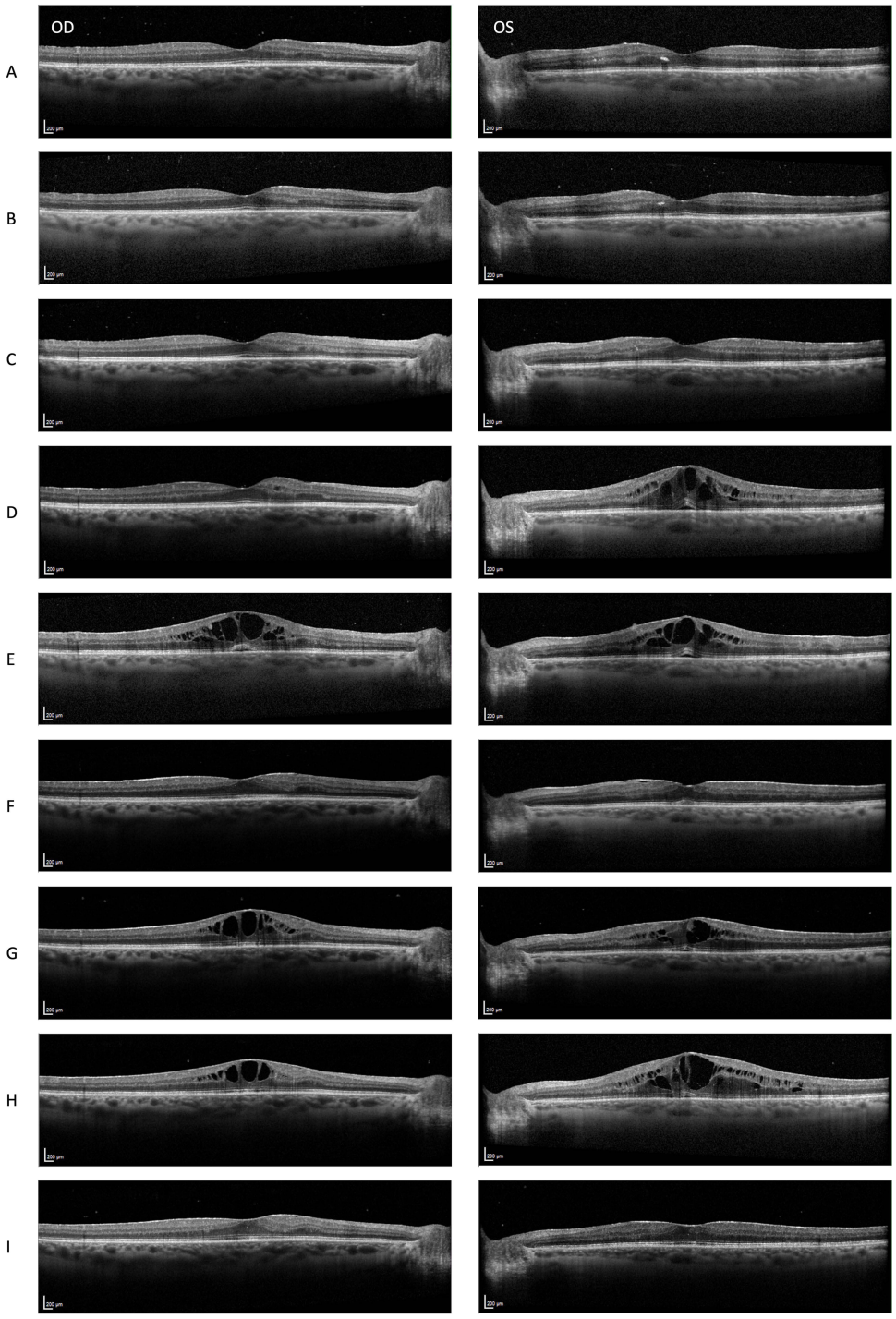

Figure 2 Optical coherence tomography progression in both eyes. OD: right eye; OS: left eye. (**A–I**) represent follow-up at 2, 4, 5, 6, 11, 13, 15, 16, and 17 months, respectively.

No complications were reported during the follow-up period. The patient developed ocular hypertension in the OD prior to the administration of any intravitreal implants, likely due to the inflammatory process. It was successfully managed with antiglaucomatous eye drops and resolved before the corticosteroid implants were administered, with no significant elevation of intraocular pressure (IOP) during the subsequent period. Additionally, there were no changes in the Lens Opacities Classification System (LOCS) measurements throughout the follow-up.

Discussion

BCR is a chronic inflammatory condition that can potentially threaten vision. This case highlights the sustained management of CME following treatment with corticosteroid intravitreal implants, an aspect that is not often explored in the current literature on this condition. The therapeutic efficacy of local ocular corticosteroids in non-infectious posterior uveitis is well established.[7,9–14] Intravitreal implants have demonstrated significant effectiveness in controlling intraocular inflammation, often reducing the need for systemic therapy. Localized treatment is particularly advantageous in BCR, as the disease is confined to the eyes.[1,3–6] This study evaluates the use of both dexamethasone and FA intravitreal implants in managing patients with BCR.

Previous studies have reported a significant incidence of cataract progression and glaucoma associated with corticosteroid use. However, in this case, IOP-lowering drops were required only temporarily and were not related to the corticosteroid intravitreal implant. Moreover, no cataract development was observed.

As choroidal inflammation in BCR does not appear to respond adequately to monotherapy with slow-release, sustained-delivery intravitreal steroids, it is generally believed that systemic corticosteroids or conventional immunomodulatory therapy are necessary for comprehensive disease control. Consequently, our patient was maintained on methotrexate starting from the sixth month of follow-up.

Although local steroid therapy alone cannot fully control choroidal inflammation in BCR, it may still offer significant benefits by effectively managing retinal vascular leakage, preserving retinal function, and controlling CME.[14] This can help reduce the need for high doses of systemic therapy and their associated risks. Since monotherapy with Ozurdex or Iluvien does not appear to ensure complete disease remission, a combined approach integrating local and systemic therapies may represent the most effective strategy for managing these patients.

Conclusion

This case underscores the challenges of managing BCR, particularly refractory CME. Intravitreal corticosteroid implants effectively stabilized macular edema and preserved visual function, complementing systemic immunomodulatory therapy. The combined use of systemic and local treatments allowed for better disease control with a favorable safety profile, including no cataract development or significant intraocular pressure issues. Tailored treatment strategies and long-term monitoring remain essential for managing this chronic condition effectively.

Funding

The article processing charges for this publication were funded by Alimera Sciences Ltd.

Disclosure

The authors report no conflicts of interest in this work.

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