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Asymptomatic Birdshot Chorioretinopathy in a Non-Caucasic Woman

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Abstract

Objective: To report a case of Birdshot chorioretinopathy with positive HLA-A29 tyífication in an asymptomatic, non-caucasin patient, and to review the current literature regarding the disease.

Observations: A 50-year-old asymptomatic woman was referred due to abnormal fundus findings in her myopia control. Her fundoscopy revealed rounded and oval hypopigmented peripapilar lesions interspersed with normal areas in both eyes. Fluorescein angiography showed macular filtration and late perivascular hyperfluorescence, while the optical coherence tomography confirmed the presence of macular edema with intraretinal cysts in both eyes. On the other hand, the electroretinogram had amplitude and latency alterations in cone responses bilaterally. HLA-A29 typification was carried out to confirm the suspected diagnosis. After discarding any possible latent infections, mycophenolate was initiated.

Conclusions: Birdshot chorioretinopathy should be suspected even in non-caucasic patients. It is important to remember that patients might be asymptomatic. Corticosteroid sparing treatment is effective to control the progression of this entity.

Keywords

Chorioretinopathy; Retinochoroidopathy; Birdshot; HLA-A29; Uveitis

Introduction

Birdshot chorioretinopathy (BCR), also known as Birdshot retinochoroidopathy or vitiligous chorioretinitis, is a chronic and bilateral posterior uveitis with characteristic findings in the eye fundus [1]. Up to 96% of cases have been related to HLA-A29 [2].

The clinical presentation is variable, being the most frequent symptoms diminished visual acuity (68%), floaters (29%), nyctalopia (25%), dyschromatopsia (20%) and photopsia (17%) [3]. A characteristic finding of BCR is that, although patients manifest subjective diminished visual acuity (VA), their best corrected visual acuity (BCVA) remains almost unaltered [4]. A significant proportion of patients may present asymptomatic or with minimal clinical alterations in initial stages of the disease, so the level of suspicion must remain high in order to avoid diagnostic delays [5].

Physical examination may reveal an anterior chamber within normal limits, while the posterior segment usually presents with vitritis and hypopigmented, rounded or oval, cream-colored and bad-defined choroidal lesions [1]. These tend to be bigger than 300 micrometers and may eventually be confluent, which is seen as a big hypopigmented area. Additional findings include retinal vasculitis, macular edema and choroidal neovascular membranes [6].

The objective of this case report is to present an asymptomatic, non-caucasic patient with BCR and to evaluate the best approach for these types of patients.

Case Report

A 50 year-old woman with no previous medical or surgical history was referred to the retina expert due to abnormal findings at the eye fundus during routinary myopia control. She was completely asymptomatic.

At physical examination, her BCVA was 20/25 in the right eye (OD) and 20/20 in the left eye (OS). Photomotor reflexes were normal OD and OS and there was no relative afferent pupillary defect. Biomicroscopy revealed clear corneas, ample anterior chamber and clear eye lenses in both eyes. A small pterygium and superficial leucomas were found OD. Fundoscopy was notable for posterior vitreous detachment, 2/6 excavation of the papilla and rounded and oval hypopigmented lesions of peripapillary distribution interspersed

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with normal findings OD and OS (Figure 1).

Computerized visual field (CVF) showed a small central scotoma OD and paracentral scotoma OS. Fluorescein angiography was notable for macular filtration and late perivascular hyperfluorescence in the main arcades (Figure 2). Optical coherence tomography (OCT) confirmed the presence of macular edema with the retinal cysts OS and OD (Figure 3). The electroretinogram (ERG) revealed amplitude and latency alterations in cone responses in both eyes. Due to suspected BCR, general laboratory blood work (hemogram, PPD, RPR and biochemicals) and a complete computerized tomography was carried out to discard any latent infection or metabolic alterations that may need to be treated before starting immunosuppressive treatment. HLA-A29 typification resulted positive.

Given the absence of symptoms, mycophenolate 500 mg every 12 hours was initiated, and we abstained from the use of corticosteroids as a first-line treatment. The patient has been under regular follow-up and has remained completely asymptomatic with notable improvement in laboratory and imaging tests.

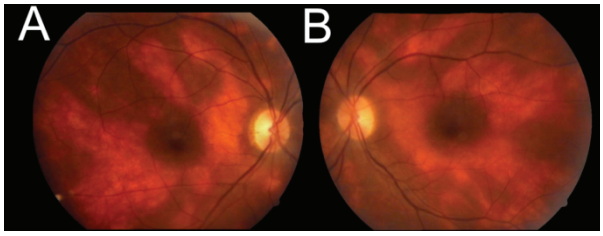

Figure 1: (A) Fundus of the right eye and (B) fundus of the left eye. Both reveal the presence of rounded hypopigmented lesions of peripapillary distribution mixed with normal areas.

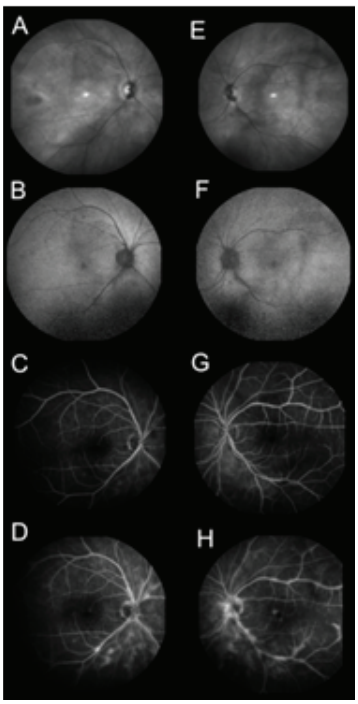

Figure 2: (A) right eye infrared (B) right eye autofluorescence (C) early fluorescein angiography of the right eye (D) late fluorescein angiography of the right eye with perivascular hyperfluorescence in the main arcades (E) left eye infrared (F) left eye autofluorescence (G) early fluorescein angiography of the left eye and (H) late fluorescein angiography of the left eye which is also notable for perivascular hyperfluorescence in the main arcades.

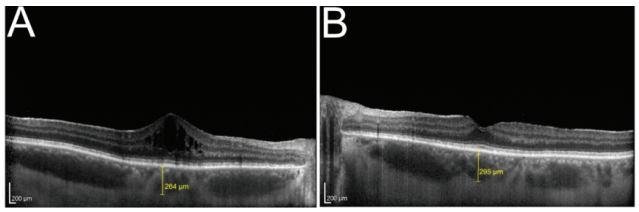

Figure 3: (A) optical coherence tomography of the right eye and (B) optical coherence tomography of the left eye. The main findings include the presence of intraretinal cysts in both eyes, bigger in the right eye.

Discussion

BCR is a disease that predominantly affects women of European families, between the fifth and sixth decade of life. It accounts for 6%-8% of patients with posterior uveitis [7]. The objective of this case report was to present a non-Caucasian, asymptomatic patient and describe the outcome with corticosteroid sparing treatment, while evaluating different treatment options in these types of patients.

Complimentary tests usually help with the diagnosis in the early stages of the disease. Fluorescein angiography is useful to evaluate the presence of active vasculitis, macular edema and choroidal neovascular membranes. On the other hand, indocyanine green angiography is relevant due to its higher sensitivity when compared to fluorescein angiography [6,8]. OCT is useful in evaluating the presence of macular edema, neovascular membranes, retinal atrophy and disruption in the photoreceptor layer. These last two findings have been related to lower visual acuity and worse prognosis in the long term. Finally, autofluorescence may reveal hypofluorescent areas that are easily noticeable in contrast to common fundoscopy, in which hypopigmented areas may be invisible in the early stages of the disease [6].

BCR diagnosis is eminently clinical, with a sensitivity of 97.5% and a 100% specificity[9]. Diagnostic criteria include (1) bilateral findings; (2) three or more characteristic lesions, defined as cream-colored, rounded and irregular choroidal lesions; (3) minimal anterior chamber inflammation, defined as less than 1+ cells (SUN scale); and (4) vitreous inflammation of no more than 2+ (NEI/SUN score) [5]. Exclusion criteria include the presence of keratic precipitates, posterior synechia and the concomitance with other inflammatory, infectious or neoplastic pathologies that may cause multifocal choroidal lesions.

It is important to notice that, although it is the most strongly related human disease with a HLA allele, HLA-A29 positivity is not considered as a diagnostic criteria. This is due to the possibility of HLA-A29 negative BCR and that up to a 7% of asymptomatic caucasians present this allele [2]. Therefore, HLA-A29 study should be considered as confirmatory rather than a diagnostic test. In our case, HLA positivity was useful for confirmatory purposes in order to start immunosuppressive treatment in a patient with no symptoms but characteristic signs of BCR.

Differential diagnosis should include infectious and non-infectious diseases, such as tuberculosis, syphilis, uveal lymphoma and sarcoidosis [8]. This last entity is the most important in cases of HLA-A29 negativity [6].

Systemic corticosteroids are the first-line treatment for BCR while the possibility of initiating immunosuppressive treatment is evaluated (bridge therapy). Corticosteroids are useful for acute inflammation, but there is no evidence that disease progression is affected [4]. Moreover, corticosteroid monotherapy doses are unacceptably high and secondary effects are common [5]. In our case,

the patient was completely asymptomatic, so we abstained from using them in order to escalate directly to immunosuppressive treatment, avoiding possible complications and side effects of corticosteroids.

Immunosuppressive treatment, also referred to as corticosteroid sparing treatment, may include antimetabolites (methotrexate, azathioprine or mycophenolate), T-cell inhibitors (cyclosporin) or the use of intravenous immunoglobulins. The objective of this treatment is to preserve visual function, minimize high-dose corticosteroid side effects and to prevent the progression of the disease [4]. In our experience, we had excellent results with the use of mycophenolate, evidenced in the persistent absence of symptoms and the improvement in the patient´s exams.

Another treatment option is the use of biological agents. There is evidence that the use of tumoral necrosis factor (TNF) inhibitors is successful both in short-term and long-term results in patients with posterior uveitis (infliximab, adalimumab) [4,10]. Tocilizumab (interleukin-6 inhibitor) treatment was recently reported to be successful in patients with refractory disease to TNF inhibitors plus corticosteroid treatment [8]. Treatment demonstrated to improve visual acuity, inflammation and reduce macular edema, with minimal side effects.

Local intravitreal injections with corticosteroids are useful for disease exacerbations, especially when only one eye is affected. Nonetheless, up to 50% of patients present serious side effects such as intraocular pressure elevation and rapidly developing cataracts [4].

BCR is a chronic disease that tends towards progression, so patients should be regularly controlled in order to evaluate treatment effectiveness. Ophthalmological evaluation, added to the use of images (fluorescein angiography, indocyanine green angiography and OCT), ERG and CVF is the most appropriate measure, despite the poor correlation between symptoms, physical examination and image results [5].

Conclusions

In conclusion, this case supports the positive outcome in the treatment of an asymptomatic patient that presented with signs of BCR. Clinical findings and high clinical suspicion were essential for the early diagnosis and prompt treatment. HLA-A29 typification is only useful as a confirmatory test and should be considered in non-caucasic patients. It is important to keep in mind the frequent necessity of immunosuppressive treatment. Finally, in order to evaluate the treatment response and disease progression, regular ophthalmological evaluation is necessary.

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