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Journal of
Ophthalmology and Eye Disorders

ISSN 2831-3216

Case Report

Birdshot Chorioretinopathy Treated With Adalimumab And Methotrexate From Onset: The Value Of Regular Angiographic Follow-Up.

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Running title: Angiographic follow-up of Birdshot

Abstract

Purpose: Birdshot chorioretinopathy (BSCR) is a chronic, bilateral, auto-immune, posterior uveitis. Through this BSCR case, we investigated the value of angiographic follow-up, after starting immunomodulatory therapy (IMT).
Case report: Upon BSCR diagnosis, a 46-year-old male patient started IMT (adalimumab + methotrexate). At 8 months, we observed clinical quiescence. Angiographically, we observed complete disappearance of the typical choroidal lesions within the first year, but retinal vasculitis persisted to be present up to 2 years to date. Therefore, we continued IMT.
Conclusions: This BSCR case report shows that, even while treated with intensive IMT with adalimumab from onset and despite apparent clinical remission, regular angiographic follow-up revealed incomplete remission after 2 years, especially the retinal vasculitis component. Therefore, we recommend cautiously tapering of IMT and regular angiography for monitoring.

INTRODUCTION

Birdshot chorioretinopathy (BSCR) is a chronic, bilateral, auto-immune, posterior uveitis.(1) The initial stage of the disease course is characterized by vitreous floaters, retinal vasculitis and choroidal lesions. In the more advanced stages cystoid macular edema, vascular attenuation, retina pigment epithelium changes, optic nerve atrophy, and seldom subretinal neovascularization, become evident. The conventional treatment is immunomodulatory therapy (IMT) through a step-up approach: after starting with oral corticosteroids, the subsequent step is steroid sparing IMT. Given the known chronic course of BSCR, there is a relative indication for the early use of IMT.(2) If left untreated, the visual acuity (VA) of patients may decline to ≤ 20/200 over 10 years in 16-22% of the patients.(3) However when using IMT, VA stabilises or improves in 79-89%.(4)

CASE REPORT

Disease onset

A 46-year-old man first presented at the emergency department of the Rotterdam Eye Hospital (REH) with blurred vision, floaters and photopsia in the right eye. His symptoms had been progressive for one year. Eye examination revealed a VA of 20/20 in both eyes. Two small retinal defects were detected, which were treated with laser. No signs of inflammation were observed. During the follow-up visits, he complained of persisting floaters in the right eye, but only a Weiss ring was newly reported. Symptoms were assigned to a posterior vitreous detachment and dry eyes caused by coexisting Meibomian gland dysfunction. Since he experienced much burden from the floaters, he enquired about undergoing laser vitreolysis at another hospital. During that consultation, signs of vitritis were observed in both eyes and the patient was referred to the uveitis service of the REH.

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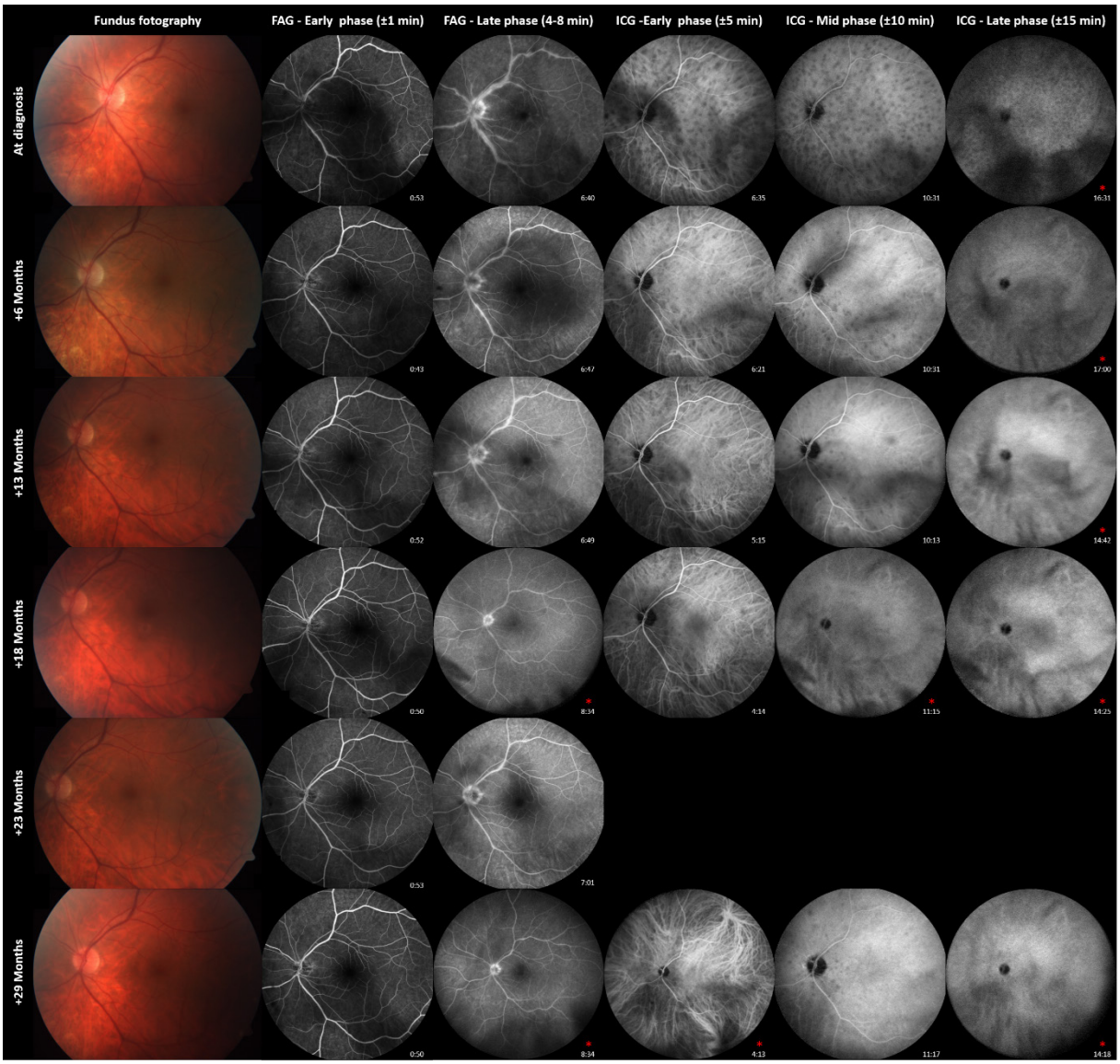
Received: 03-July-2025, Manuscript No. JOED - 4966 **; Editor Assigned:** 05-July-2025 **; Reviewed:** 29-July-2025, QC No. JOED - 4966 **; Published:** 09-August-2025, **DOI:** 10.52338/joed.2025.4966.
Citation: Mirjam E.J. van Velthoven, MD PhD. Birdshot Chorioretinopathy Treated With Adalimumab And Methotrexate From Onset: The Value Of Regular Angiographic Follow-Up. Journal of Ophthalmology and Eye Disorders. 2025 August; 12(1). doi: 10.52338/joed.2025.4966.

Diagnosis

Upon his first visit at our uveitis service, the patient reported to have bilateral symptoms. VA was 20/20 in both eyes. Slit lamp examination revealed 0.5+ anterior chamber cells in both eyes and vitreous haze of 0.5+ in the right eye and 1-2+ in the left eye. Fundus examination revealed peripheral yellowish choroidal lesions, vascular sheathing, venous dilatation, and calibre changes (**Figure 1**). We performed fluorescein (FA) and indocyanine green (ICG, using infracyanine after the second time, because of a suspected moderate allergic reaction to ICG) angiography. Although the macular OCT was unremarkable, FA showed retinal vasculitis and late-phase mild macular edema. ICG revealed extensive choroidal lesions. His electroretinogram showed decreased retinal function. We performed blood examination: the full blood count, liver and kidney function were normal. Additional testing did not show evidence for syphilis, tuberculosis, sarcoidosis, and other systemic auto-immune diseases. When the HLA-A29 typing result came back positive, we made the diagnosis BSCR.

Figure 1. Overview of sequential angiographic follow-up in BSCR, using the Topcon fundus camera and Spectralis Heidelberg for the angiographic images.



*Only the worst eye is presented. 1st column, color fundus photographs: At diagnosis (top row) a vitreous haze of 1-2+ was observed which completely disappeared at 13 months (3rd row). Birdshot choroidal lesions are not evident on the color images. 2nd-3rd column, fluorescein angiography (FA), early (2nd) and late (3rd) phase: At diagnosis (top row), FA shows retinal vasculitis and optic disc hyperfluorescence. Vasculitis decreased over time but did not disappear completely at 23 and 29 months (bottom rows). 4th-6th column, indocyanine green angiography (ICG), early, mid and late phase respectively: at diagnosis (top row) multiple hypocyanescent spots (birdshot choroidal lesions) are seen (late phase), which fully disappeared around 18 months (4th row). At 29 months some residual hypercyanescent dots in the peripapillar area are seen in mid to late phase and are considered scarred lesions. The frames marked with a red * are widefield images (102°). *There was no ICG at 23 months, due to unavailability of infracyanine.*

Treatment and disease course

At the first presentation at our uveitis service, the patient started with prednisolone eye drops. Within one month, once the additional systemic work-up was unremarkable, and after consultation with an immunologist of an affiliated Erasmus Medical Centre, he started with a loading dose of 80 mg subcutaneous adalimumab followed by 40 mg doses biweekly, methotrexate 7.5 mg weekly and folate acid, without oral prednisolone. Since we diagnosed a latent hepatitis B, he also started with entecavir prophylactically. Subsequently we saw the patient at regular intervals at our uveitis service. We tapered the prednisolone eye drops rapidly. The patient reported a slow decline in visual symptoms, and we observed a decrease in uveitis activity. Clinically, signs and symptoms disappeared over a period of 8 months, including fading of the birdshot lesions, vascular sheathing, and calibre changes. We considered the BSCR clinically quiescent at that time (Figure 1). To meet the patient's need for reassurance, angiographic follow-up at 6-monthly intervals was performed (at 6, 13, 18, 23 and 29 months respectively). On ICG, the active Birdshot choroidal lesions had disappeared completely at 13 months. However, FA showed persistent retinal vasculitis through the second year of follow-up (Figure 1 – 2nd and 3rd column). Guided by these angiographic observations, we did not taper the IMT.

DISCUSSION

Our case demonstrates that even when clinical quiescence is present and, consequently, tapering of IMT is considered, angiographic follow-up is mandatory to evaluate retinal and choroidal subclinical signs of inflammation. Our case showed angiographic signs of inflammation in a BSCR patient for over a period of at least 2 years, despite aggressive treatment with adalimumab and methotrexate from onset. Based on our experience and knowledge regarding treatment of BSCR with IMT, we usually do not perform angiographic follow-up at regular intervals, only when treatment decisions depend on it, in line with the practice of other uveitis specialists.(5-9) In practice, we tend to continue with IMT in BSCR, without invasive diagnostic follow-up, to prevent a risk of early recurrence, or faster disease progression to an advanced stage. This exemplary case however revived our awareness to appreciate the value of performing invasive angiography in BSCR patients before tapering IMT.

Currently, the consensus is that IMT may be tapered in case of clinical quiescence after 24 months.(3) The rationale to start treatment with the combination of two immunomodulatory drugs, adalimumab and methotrexate from start, without prednisolone, is based on the outcomes of several major trials in uveitis.(6-8) In those trials clinical quiescence is the main reported outcome, unlike outcomes of angiography. Despite that, several reports mention the use of regular angiographic follow-up in clinical practice to monitor treatment effects of IMT in BSCR patients.(5-9) None of the aforementioned studies reported the discrepancies observed between clinical quiescence and angiography outcomes under treatment with adalimumab.

In conclusion, continued angiographic follow-up in this clinically recovered BSCR patient illustrated the chronic disease course even with intensive IMT from onset and the need for caution with tapering of IMT in BSCR patients. Further research is necessary to determine the role of angiographic imaging in follow-up of BSCR patients and in its role in determining the optimal moment to taper or discontinue IMT.

Declarations

Funding

Not Applicable

Conflicts Of Interest/Competing Interests

Not applicable

Availability Of Data And Material

Not applicable

Ethical Approval

The local Ethical Committee does not require official approval for the publication of single case reports.

Consent To Participate And For Publication

Written informed consent was obtained in which the patient agrees to publish the medical information and images included in this case report.

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