Barton AT, Francis ER, Feibelman P, Geary M, Booy D. Recurrent Vision Loss in a Patient with Giant Cell Arteritis while on High Dose Corticosteroids. *Brown Hospital Medicine*. 2024;3(2).
doi:10.56305/001c.92898

Case Reports

Recurrent Vision Loss in a Patient with Giant Cell Arteritis while on High Dose Corticosteroids

Andrew T. Barton[1], Elizabeth R. Francis, MD[1], Palmer Feibelman, MD[2], Meghan Geary, MD[1], David Booy, MD[2]

[1] Department of Medicine, Warren Alpert Medical School of Brown University,
[2] Department of Ophthalmology, Warren Alpert Medical School of Brown University

Journal of Brown Hospital Medicine
Vol. 3, Issue 2, 2024

Article Information

Keywords: Giant cell arteritis, Acute Vision Loss, Steroids, GCA, Vision loss

https://doi.org/10.56305/001c.92898

Submitted: September 22, 2023 EDT

Accepted: January 31, 2024 EDT

Abstract

Giant cell arteritis (GCA) is a chronic vasculitis that can lead to permanent vision loss. In this case we describe a patient with permanent monocular vision loss from GCA, managed with long-term glucocorticoids, who presented with elevated erythrocyte sedimentation rate and acute visual loss. This patient was promptly given intravenous steroids, with initial improvement, however, she again experienced acute vision loss on the fourth day of treatment. Given the rare nature of this recurrent GCA-related vision loss during treatment, a broader workup was pursued without an alternative cause identified. Temporal biopsy was not performed due to concern of a false-negative in the setting of chronic steroid use. In this report we discuss the long-term monitoring and management of GCA, recurrence in the acute treatment setting, and present evidence related to diagnosis of GCA in the context of long-term steroid use.

BACKGROUND

Giant cell arteritis (GCA) is a large vessel vasculitis commonly affecting the major branches of the aorta, especially the vertebral and carotid arteries with associated systemic symptoms and potential visual loss.[1,2] Permanent visual loss occurs in up to 20% of patients, consisting of partial or complete loss in one or both eyes.[3] Diagnosis relies on histopathologic evidence, typically in the form of temporal artery biopsy. Long-term management of GCA requires monitoring of inflammatory markers and clinical correlation and an individually tailored treatment regimen to avoid the serious consequences of recurrence. In this case, we present a patient with recurrent GCA despite chronic steroids, and recurrent acute vision loss during hospitalization.

CASE PRESENTATION

A 79-year-old female with a history of GCA causing complete vision loss in the left eye managed with long-term prednisone, as well as wet macular degeneration managed with consistent anti vascular endothelial growth factor (VEGF) injections, presented with acute vision loss of the right eye. Her left eye has been unable to perceive light since the initial presentation of biopsy-confirmed GCA nine years prior. She had been taking 1mg of prednisone daily for several years, with occasional brief increases based on regular erythrocyte sedimentation rate (ESR) levels. Over the past five years, her ESR had been between 13 and 25 mm/h (normal 0-30 mm/h). One month before presenting, her ESR was found to be 61 mm/h on a routine laboratory examination. Review of systems did not indicate recent infection, and this ESR elevation was attributed to GCA flare. Her prednisone dose was increased to 40mg daily, and then, after 11 days, increased to 60 mg daily.

One week after the second dose increase, she realized she had new blurry vision in the right eye and presented to the hospital. A non-contrast computed tomography (CT) scan of the head found no evidence of large vessel occlusion, or stenosis in the neck. She denied typical symptoms associated with GCA - fever, chills, unintentional weight loss, jaw claudication, headache, scalp tenderness, arthralgia, or proximal muscle pain. However, she noted she did not have these symptoms at presentation nine years prior. Vital signs were notable for blood pressure of 211/99. Ophthalmology, and rheumatology were consulted. Examination found good temporal artery pulses, without cordis, and a visual acuity of 20/150 in the right eye. Fundus examination of the right eye was notable for trace pallor of the optic disc, drusen, and normal vessels. No retinal ischemic lesions were noted. Her ESR in the ED was 10 mm/h with a C-reactive protein (CRP) of 1.64 mg/dL. Despite this atypical presentation and low ESR, her acute vision loss was attributed to GCA given her history, and she was started on 250 mg of intravenous (IV) methylprednisolone every six hours. Magnetic resonance imaging found no acute pathology in the brain and orbits. There was atrophy of the left optic nerve

consistent with chronic vision loss and some volume loss of the right optic nerve, but without focal lesion or abnormal enhancement. A temporal artery biopsy was not performed as her long-term prednisone course increased the likelihood of a false negative.

On continued IV methylprednisolone, her vision improved to 20/30 in the right eye on the third day of hospitalization. However, the next morning she again experienced acute blurry vision, only being able to perceive hand motion. Repeat CTA was normal, and fundoscopic exam was unchanged. As her ESR and CRP were low while on the fourth day of IV methylprednisolone, broader diagnostic testing was pursued. Magnetic resonance angiography found no abnormal wall enhancement, and further lab work found normal complement levels, antinuclear antibodies, antineutrophilic cytoplasmic antibodies, anti-Smith antibodies, anti- DNA antibodies, as well as negative human immunodeficiency virus (HIV) and syphilis tests. With other imaging and laboratory testing continuing to be negative, GCA was still the presumed diagnosis. The next day, her vision improved again to 20/40 in the right eye. A prednisone taper was begun with plans to transition to abatacept outpatient. She was discharged with close rheumatology follow-up.

DISCUSSION

Giant-cell arteritis (GCA) is a vasculitis of large-sized vessels. While vessel inflammation may be widespread, the symptoms of GCA are usually related to inflammation of the cranial branches of arteries originating from the aortic arch.[3] This disease is almost exclusively found in those over 50 years of age, with women making up two thirds of those affected.[4]

This patient presented with vision loss without systemic symptoms, as is the case for approximately 20% of patients with GCA related vision loss.[5] The pattern of vision loss in GCA is painless and monocular, however, permanent binocular vision loss can develop with delayed diagnosis or treatment.[6,7] Anterior ischemic optic neuropathy is the most common mechanism GCA's ocular manifestations, followed by occlusion of the central retinal and cilioretinal arteries, ocular ischemic and infarction syndromes, and posterior ischemic optic neuropathy.[2]

The diagnosis of GCA relies on histopathology or imaging, however, this evidence can be challenging to obtain in the setting of a remitting and relapsing course, especially in the setting of long-term steroid use. Evidence on the effects of chronic steroids on temporal artery biopsy are mixed. Achkar et al. found similar positivity rates of temporal artery biopsy between untreated and corticosteroid-treated patients overall. However, they noted decreasing positivity rates with longer duration of treatment.[8] Whereas Allison et al. found a dramatic decrease in temporal artery positivity if the patient had been treated with corticosteroids for more than a week.[9] In another case, a patient was found to have positive temporal artery biopsy after six years of treatment with therapeutic doses of steroids.[10] As this patient had completed nine years of oral prednisone and received high-dose IV methylprednisolone during her hospitalization, temporal artery biopsy was not pursued due to uncertainty surrounding its reliability. It should also be noted that normal ESR at GCA presentation is not uncommon, with some studies recording rates as high as 22.5%.[11,12]

Management of GCA requires an initial prednisone dose of at least 40 to 60 mg with pulsed intravenous methylprednisolone given to patients with visual loss.[3] Diagnosis of a GCA flare in this case was challenging given that the patient's response to standard management was ineffective. Specifically, she suffered vision loss after her chronic outpatient steroid regimen was up titrated from 1mg to 40mg of prednisone upon noting an increase in serum ESR on routine lab work and then to 60mg of prednisone 11 days later. And, she had a recurrence of visual loss despite receiving three days of IV methylprednisolone at 1000 mg daily. The literature notes that an event while on steroids is considered unlikely.[3] Though, in a retrospective study of 245 patients with GCA, Aiello et al. found that 13% of patients with visual deficits can develop additional vision loss after beginning therapy.[13] This, in concomitance with negative imaging, lends support for GCA flare in this case.

GCA flares are common, with the highest rates at the time of steroid withdrawal. The risk at the time of steroid withdrawal has been observed to be close to 50%.[14,15] There is no set duration that a patient should continue on steroids after diagnosis, instead an individualized duration with frequent ESR checks is recommended.[14] On average, GCA patients will remain on oral steroids for two to three years.[16] Proven et al. observed that the median duration to reach 5mg of prednisone per day was 7.5 months and that, among their 87 patients who were followed to discontinuation of glucocorticoid therapy and permanent remission of GCA, the total median dose of prednisone was 6.47 grams.[12] Though glucocorticoids are considered the cornerstone therapy for GCA, the harmful systemic effect of long-term steroid use[17] has led to the emergence of alternative therapies for GCA including immunosuppressants and biologics, with tocilizumab considered a strong option.[18] In this same vein, our patient who has required eight years of prednisone at 1mg daily and recently suffered GCA flare with transient right visual loss, will transition to biologic therapy.

With high rates of recurrence and imperfect diagnostic tools available to clinicians, a high suspicion is necessary in the long-term care of patients with a history of GCA, while maintaining an awareness of other causes of acute monocular vision loss, such as retinal detachment,

arterial occlusion, infection, and other inflammatory conditions.[19]

Conflicts of Interest/Disclosures

The authors have no conflicts of interest to declare.

Corresponding Author

Andrew Barton
Warren Alpert Medical School at Brown University
Providence, RI 02903
Email: andrew_barton@brown.edu

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