


CASE REPORT

An atypical presentation of giant cell arteritis: Fatigable signs to anterior ischemic optic neuropathy and choroidal infarction

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Key Clinical Message

Prompt diagnosis and treatment of GCA are crucial to preserve vision. Because of this, new-onset ptosis or diplopia in elderly patients should warrant consideration of GCA, even in the absence of “classic” features, systemic symptoms or elevated inflammatory markers.

Abstract

Giant cell arteritis (GCA) is a vision-threatening, ophthalmic emergency that classically presents with new-onset headaches, scalp tenderness, systemic symptoms, visual disturbances, and elevated inflammatory markers. We describe an atypical presentation of GCA in an 87-year-old patient with fatigable ptosis and diplopia, with subsequent anterior ischemic optic neuropathy and choroidal infarction.

KEYWORDS

blonde fundus, choroidal infarction, diplopia, giant cell arteritis, ptosis

1 | INTRODUCTION

Giant cell arteritis (GCA) is an immune-mediated vasculitis of the elderly that primarily affects medium and large-sized arteries. The onset of GCA is usually insidious and can present with a wide spectrum of manifestations, including but not limited to new-onset headache, scalp tenderness, jaw claudication, fever, and fatigue.¹ Prompt diagnosis and treatment are necessary to prevent permanent vision loss. The most common, vision-threatening manifestation of GCA is arteritic anterior ischemic optic neuropathy (AAION), although GCA-associated retinal and choroidal ischemia have also been reported. Ptosis and diplopia, although less common and serious than their blindness-inducing counterparts, can be premonitory

symptoms of GCA.² In this article, we report the case of an 87-year-old woman who initially presented with diplopia and fatigable ptosis suggestive of ocular myasthenia gravis (OMG), but eventually developed bilateral AAION and choroidal ischemia secondary to GCA.

2 | CASE HISTORY AND EXAMINATION

An 87-year-old Caucasian woman with a history of transient ischemic attack, hypertension, and hyperlipidemia presented to the neuro-ophthalmology service at our institution for a 4-week history of right-sided ptosis and pain behind her right eye that radiated down her neck. She also

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endorsed a 2-week history of intermittent, binocular horizontal diplopia, and a mild headache the day prior that resolved with over-the-counter lidocaine cream and heat patches. She denied any current headaches, scalp tenderness, jaw claudication, or fever. On initial evaluation, the patient's best corrected visual acuity (BCVA) was 20/40 in the right eye and 20/60 in the left eye. She demonstrated fatigable ptosis on the right, as well as esotropia and left hypertropia. Based on these findings, OMG was suspected and serologic work-up was done, which later returned within normal limits.

Three days later, the patient returned with sudden loss of the inferior half of her visual field in her right eye and narrowing of her visual field in her left eye that she reported occurred overnight. She also stated that the day prior, she experienced diplopia, headache, and mild temporal tenderness that resolved. Her BCVA at that time was 20/400 in the right eye and 20/50 in the left eye. Humphrey® Visual Field Analyzer testing revealed an inferior altitudinal defect in the right eye (Figure 1) and generalized depression in the left eye. Fundal examination showed diffuse pallor with normal retinal vasculature bilaterally and choroidal ischemia that was more prominent in the left eye (Figure 2). The patient was subsequently referred to the emergency department for stroke evaluation and GCA work-up and presented to an outside hospital.

There, magnetic resonance imaging (MRI) of the brain and orbits revealed no optic nerve enhancement, optic nerve sheath enhancement, or acute intracranial abnormalities. Erythrocyte sedimentation rate (ESR) was normal at 34 mm/h (age-adjusted reference range 0 to 48.5 mm/h), but C-reactive protein (CRP) was found to be elevated at 5.5 mg/dL (normal ≤ 0.3 mg/dL). The patient was subsequently discharged without treatment from the outside hospital and advised to follow up at our institution the next day.

The following morning, she reported worsening vision with BCVA at counting fingers in the right eye and no light perception in the left eye. Dilated fundus examination revealed new-onset optic disk edema in both eyes consistent with AAION, with confirmation on retinal nerve fiber layer (RNFL) analysis from Spectralis®-OCT. Fluorescein angiography (FA) (Figure 3) showed delayed transit times in the choroidal phase between 16 and 26 s bilaterally (normal ≤ 15 s). Optical coherence tomography (OCT) imaging of the macula confirmed choroidal infarction bilaterally, as well as paracentral acute middle maculopathy in the left eye (Figure 4).

She was subsequently admitted to our institution's hospital for initiation of IV steroid therapy (methylprednisolone 250 mg, four times daily), after which she was transitioned to oral prednisone 60 mg per day and discharged with a steroid taper. Temporal artery biopsy showed granulomatous panarteritis consistent with a diagnosis of GCA.

3 | OUTCOME AND FOLLOW-UP

On subsequent follow-up 2 weeks later, the patient's BCVA was 20/60 in the right eye and no light perception in the left eye. Inflammatory markers were improved but she continued to have a persistent right inferior altitudinal defect, which prompted reinitiation of prednisone 60 mg daily. She was instructed to follow up in the clinic in monthly intervals with plans to gradually taper her prednisone as tolerated. At these subsequent visits, the patient's BCVA fluctuated between 20/60 and 20/40 in her right eye and remained no light perception in her left eye. OCT images of both eyes showed gradual but expected progression of optic nerve atrophy from her GCA event. Inflammatory GCA markers have remained within normal limits since discharge.

At her most recent visit, 6 months after her initial event, the patient's BCVA was 20/50 in the right eye and no light perception in the left eye. She endorsed "fuzzy and dim" vision and was found to have visually significant cataracts in her eyes, likely secondary to her prednisone treatments. OCT images and visual field testing at this time demonstrated stable optic nerve atrophy in both eyes and persistent inferior altitudinal defect in her right eye. Prednisone was decreased to 5 mg daily, and the patient was scheduled for right eye cataract surgery. She will continue to follow up with our institution for close monitoring.

4 | DISCUSSION

In this article, we discuss an atypical presentation of GCA in an 87-year-old woman who presented with a 4-week history of right-sided ptosis and a 2-week history of binocular horizontal diplopia with subsequent vision loss. It is estimated that 16%–20% of patients with GCA experience permanent loss of vision.^{3–5} Although the exact pathophysiology of GCA remains unknown, one hypothesis is that dendritic cells in the arterial adventitia release cytokines in response to unknown "danger signals," causing vascular inflammation, remodeling, and subsequent occlusion.³ Classic presenting symptoms are new headaches or changes to characteristics of pre-existing headaches, jaw claudication, abrupt vision changes, and systemic symptoms such as fever. Less common symptoms are limb claudication, asymmetric blood pressures, or vascular bruits. New-onset ptosis or diplopia are also atypical for GCA but should be differentiated from other etiologies like levator dehiscence, decompensated euthyroidism, myasthenia gravis, and Graves' disease if clinical suspicion is high.

In a multicenter, retrospective study, Ross et al found that patients with diplopia from GCA were more likely

OD Three in One

Central 30-2 Threshold Test

Fixation Monitor: Blind Spot
 Fixation Target: Central
 Fixation Losses: 0/18
 False POS Errors: 0/12
 False NEG Errors: 0/10
 Test Duration: 11:15
 Fovea: <0 dB

Stimulus: V, White
 Background: 31.5 asb
 Strategy: FASTPAC
 Pupil Diameter:
 Visual Acuity:
 Rx: +3.25 DS

Age: 87

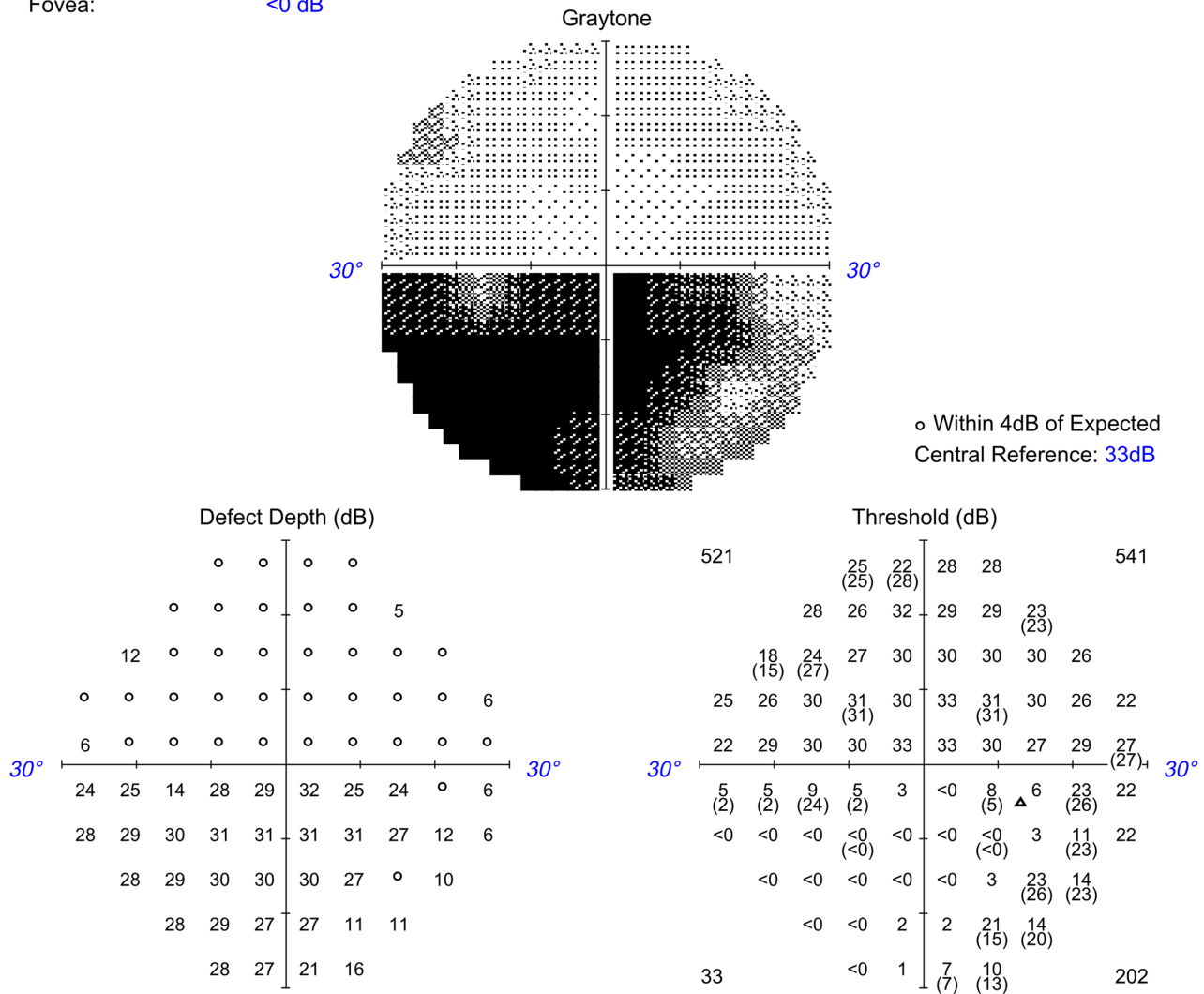


FIGURE 1 Stimulus Size V with Humphrey® Field Analyzer demonstrating an inferior altitudinal defect in the right eye.

to experience systemic symptoms (i.e., fever and scalp tenderness) and have elevated inflammatory markers.⁶ Similarly, Ing and colleagues reported that patients with diplopia and GCA were more likely to be older, experience jaw claudication and vision loss, and have higher ESR, CRP, and platelet levels than their GCA-negative counterparts with diplopia.⁷

Concomitant ptosis and diplopia in GCA are even less common, but have been reported in the literature. Hiraoka et. al described a case of GCA in a 78-year-old man whose presenting symptoms were fever, right temporal tenderness, and ptosis of the upper lid in the right eye.⁸

He was treated with 1 g intravenous methylprednisolone for 3 days followed by high-dose prednisolone (1 mg/kg of weight) with improvement in ptosis and diplopia 3 weeks after the initiation of therapy. Mantero and colleagues also described a case of GCA in a 77-year-old man who presented with an isolated left third nerve palsy, fatigable ptosis, and fever.⁹ This patient also received steroid therapy (50 mg daily for 1 month and subsequent slow taper) with complete resolution of his neurological symptoms 3 months later.

Similar to Mantero et. al's case, a correct diagnosis of GCA was delayed in our patient due to the lack

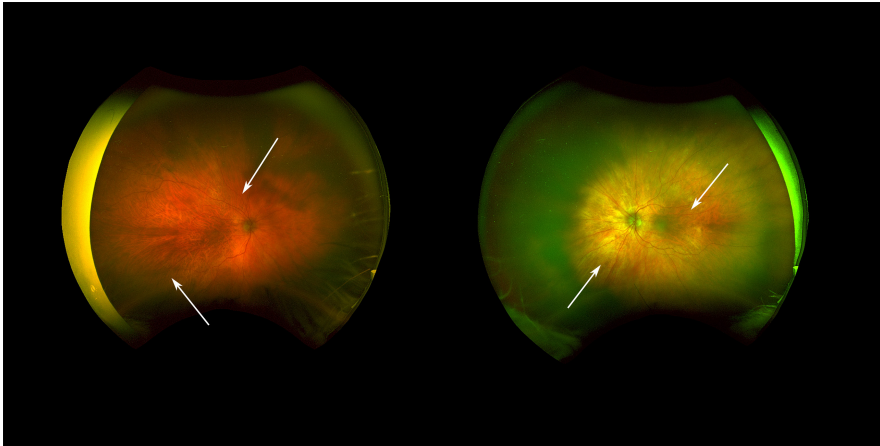


FIGURE 2 Optos fundus photographs of both eyes showing gross choroidal ischemia bilaterally (arrows).

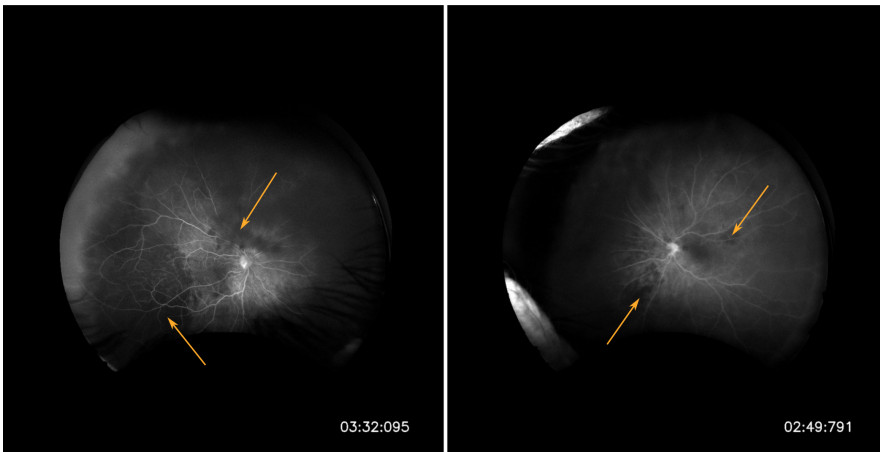


FIGURE 3 Optos fluorescein angiography (FA) imaging demonstrating delayed transit time, perfusion deficits, and choroidal ischemia bilaterally (arrows).

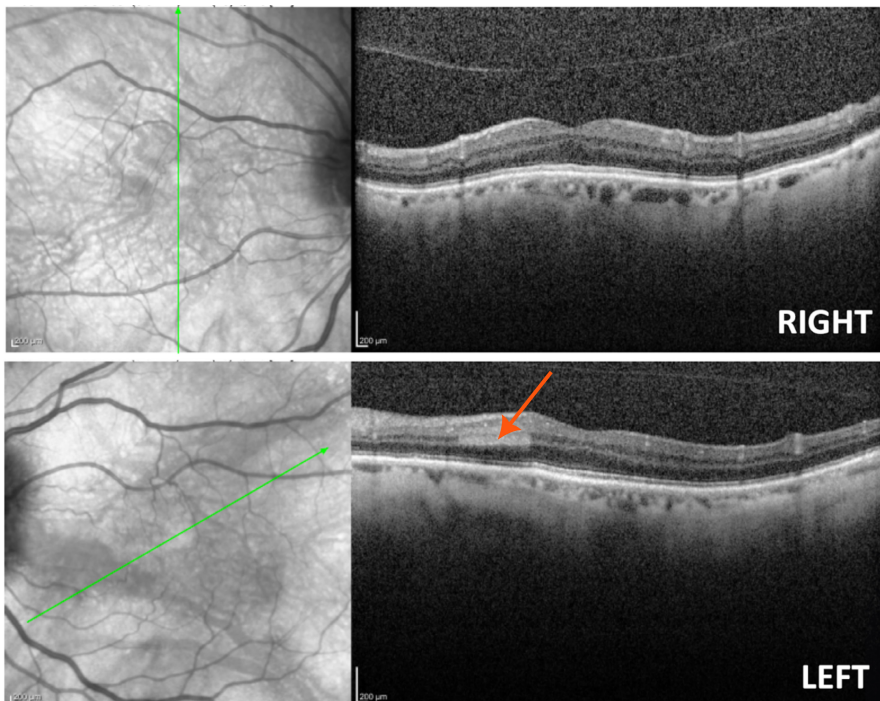


FIGURE 4 Optical coherence tomography (OCT) images of the macula significant for paracentral acute middle maculopathy in the left eye.

of characteristic systemic symptoms, unusual imaging findings, and equivocal inflammatory marker values. Although similar cases of choroidal ischemia from GCA

have been reported, these notably had disproportionate sparing of one eye¹⁰ or had only focal choroidal involvement.¹¹ Our case demonstrated diffuse prominent

choroidal ischemia bilaterally, with delayed FA choroidal transit times between 16 and 26 s. For reference, a normal FA choroidal phase would show dye entering the posterior ciliary arteries (and cilioretinal artery if present) within 15 s, with subsequent filling of the arteries 1–2 s later.¹² Also notable was the patient's 2-week history of transient diplopia preceding the diagnosis, which is atypical as one of the hallmarks of GCA is its rapidly progressive course.¹³

In summary, we report an unusual presentation of GCA mimicking OMG that eventually progressed to bilateral AAION and choroidal ischemia. GCA should be considered in elderly patients presenting with new-onset ptosis or diplopia, even in the absence of systemic symptoms or elevated inflammatory markers.

AUTHOR CONTRIBUTIONS

Meagan Shinbashi: Conceptualization; writing – original draft; writing – review and editing. **Summer Hakim:** Data curation; writing – original draft; writing – review and editing. **Elzbieta Mechel:** Writing – review and editing. **Mitul C Mehta:** Writing – review and editing. **Robert W Crow:** Supervision; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

Meagan Shinbashi, BS, Summer Hakim, BS, Elzbieta Mechel, MD, Mitul Mehta, MD, R. Wade Crow, MD have nothing to disclose.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable as no datasets were generated or analyzed during the writing of this article.

PATIENT CONSENT STATEMENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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