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LETTER

Visual Recovery with Iloprost Added to Corticosteroids in a Case of Giant Cell Arteritis

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ABSTRACT

Introduction: To date, corticosteroids remain the cornerstone treatment of ocular involvement of GCA, and no other treatment has proven to be effective in this setting. We herein report on a unique case of GCA with ocular involvement worsening despite high dose corticosteroids and recovering with intravenous iloprost.
Case report: A 70-year-old man presented with acute vision loss in his left eye related to anterior ischemic optic neuropathy. The diagnosis of giant-cell arteritis was confirmed by a temporal artery biopsy. Despite intravenous pulse methylprednisolone for 3 days then oral prednisone at 60 mg/day, the patient developed from day 5 fluctuating vision loss in the right eye, related to ocular ischemia by occlusion of the ophthalmic artery, and responsive to hyperhydration. Iloprost, an analog of prostacyclin PGI2, was then administered intravenously for 5 days and resulted in a stable improvement in visual acuity in the right eye.
Conclusion: This case highlights the potential role of vasodilatator agents in giant cell arteritis with ocular involvement.

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KEYWORDS
Giant cell arteritis; ocular complications; iloprost; corticosteroids; anterior ischemic optic neuropathy

Report of a case

A 70-year-old man was referred for an acute vision loss in his left eye (day 0) in the context of headache, jaw claudication, episodes of transient diplopia and a body weight loss of 2 kg (usual body weight of 63 kg) evolving since 2 weeks. Best-corrected visual acuity (BCVA) measured with Snellen charts was 20/20 in the right eye and "light perception" in the left eye with a papillary edema on fundus examination (Figure 1) suggestive of anterior ischemic optic neuropathy (AION). Erythrocyte sedimentation rate at the first hour was 72 mm (N < 50) and C-reactive protein titer was 50 mg/L (N < 5). Intravenous pulse methylprednisolone was administered at day 1 at 500 mg/day during 3 days according to EULAR recommendations for the management of large vessel vasculitis,[1] in addition to aspirin at 75 mg/day without visual improvement (Figure 2). The diagnosis of giant-cell arteritis (GCA) was confirmed by a temporal artery biopsy performed on day 2. On day 5, after switching to oral prednisone 60 mg/day, BCVA in the right eye dropped to 20/80 without any fundoscopic sign of AION. Markers of inflammation were normalized. Fluorescein and indocyanine green angiograms showed delayed choroidal filling, while magnetic resonance and Doppler ultrasound imaging of the orbits revealed very severe flow disturbances of the ophthalmic and central retinal arteries and inflammation of the ophthalmic arteries (Figure 3). Intravenous steroids (500 mg bid for 3 days) were repeated but the right eye's BCVA kept fluctuating then declined on day 8. Hyperhydratation (2 L of normal saline daily) was started on day 10 to ensure an optimal volemia and a better perfusion of the ischemic right eye. BCVA increased then dropped again when normal fluid intake was resumed. Following a multidisciplinary consultation meeting, iloprost (an analog of prostacyclin PGI_2) was considered. After explaining the experimental nature of the treatment, written consent was obtained from the patient and the medication was administered on day 15 for 5 consecutive days (1.5–2 ng/kg/min intravenously following a titration phase over 6 hours/day). BCVA quickly increased to 20/20 in the right eye and remained stable. Tocilizumab (an anti-IL6R antibody) was then started, and corticosteroids were tapered. At the last evaluation (month 8), under subcutaneous tocilizumab 162 mg/week and prednisone 5 mg/day, the patient was still in clinical and biological remission with a right BVCA of 20/25. However, sequelae were found on Humphrey Visual Fields and Optical Coherence Tomography testing: the inability to reach back a 20/20 vision in the right eye could thus be explained by visual field defects and a slight thinning of the retinal nerve fiber layer. As for the visual acuity in the left eye, it stayed at light perception despite the resolution of the papillary edema.

Discussion

We herein report on a unique case of GCA with ocular involvement worsening despite high dose corticosteroids and recovering with intravenous iloprost.

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*These authors contributed equally to the work

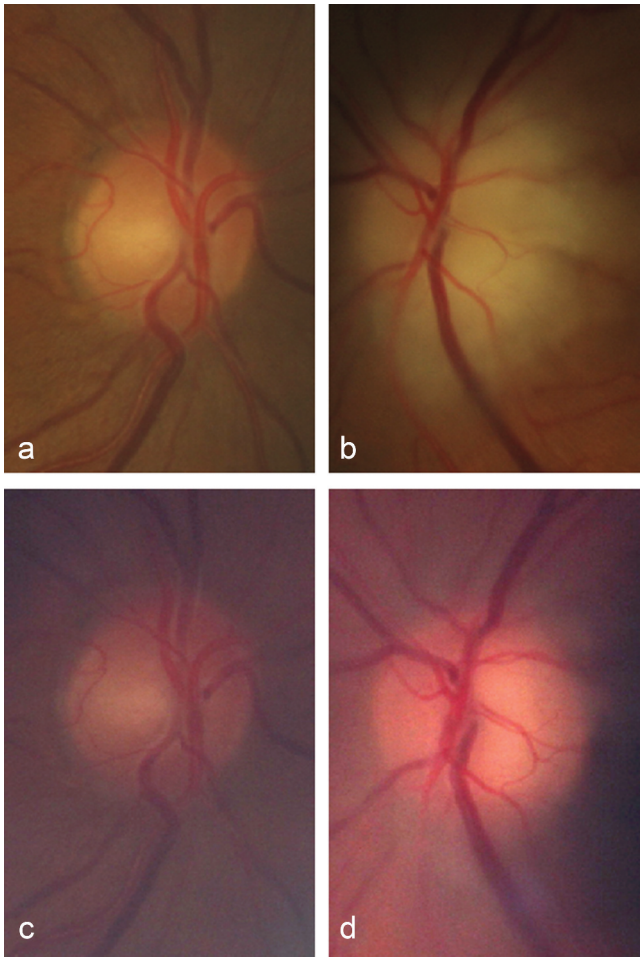

Figure 1. Fundus photographs of the optic nerve heads at presentation (a & b) and after the treatment with corticosteroids and iloprost (c & d). Note the papillary edema in the left eye (b) that resolved with time (d), while no changes were found in the right eye (a & c).

GCA is the most common form of vasculitis in elderly patients, with permanent visual loss from AION being one of its most feared complications.[2] Despite adequate treatment, functional recovery remains poor and some patients may even worsen during the first 2 weeks after steroid therapy initiation.[3] Thus, there are important unmet needs in the management of GCA-related ocular complications.

Ischemic complications of GCA are related to granulomatous inflammatory infiltration of the media, as well as to intimal hyperplasia and remodeling of the vessel wall ultimately occluding the vascular lumen.[2] In spite of targeting the inflammatory cascade leading to giant cell formation, corticosteroids don't seem to completely counteract ischemia from intimal hyperplasia and vascular remodeling.[4] Furthermore, there is a latent period between the start of corticosteroid therapy and the disease control.[5]

There is, therefore, a rationale for using a vasodilator in ischemic complications of GCA. Iloprost is a synthetic analog of prostacyclin PGI_2 whose pharmacological characteristics include not only arteriolar and venous vasodilation, but also activation of fibrinolysis and inhibition of leucocyte migration and platelet production, adhesion and aggregation. Steigerwalt *et al.* have used a similar molecule, prostaglandin E_1, in addition to steroids in 2 cases of AION, but their results are difficult to interpret as both medications were used simultaneously.[6] In our patient with a particular manifestation of GCA (ocular ischemia by occlusion of the ophthalmic artery), visual recovery was achieved after hyperhydration and introduction of iloprost, while steroid therapy alone could not prevent worsening. Hence, iloprost might be an interesting adjuvant to corticosteroids in the treatment of a sight-threatening condition. Larger studies are still necessary to better evaluate its use.

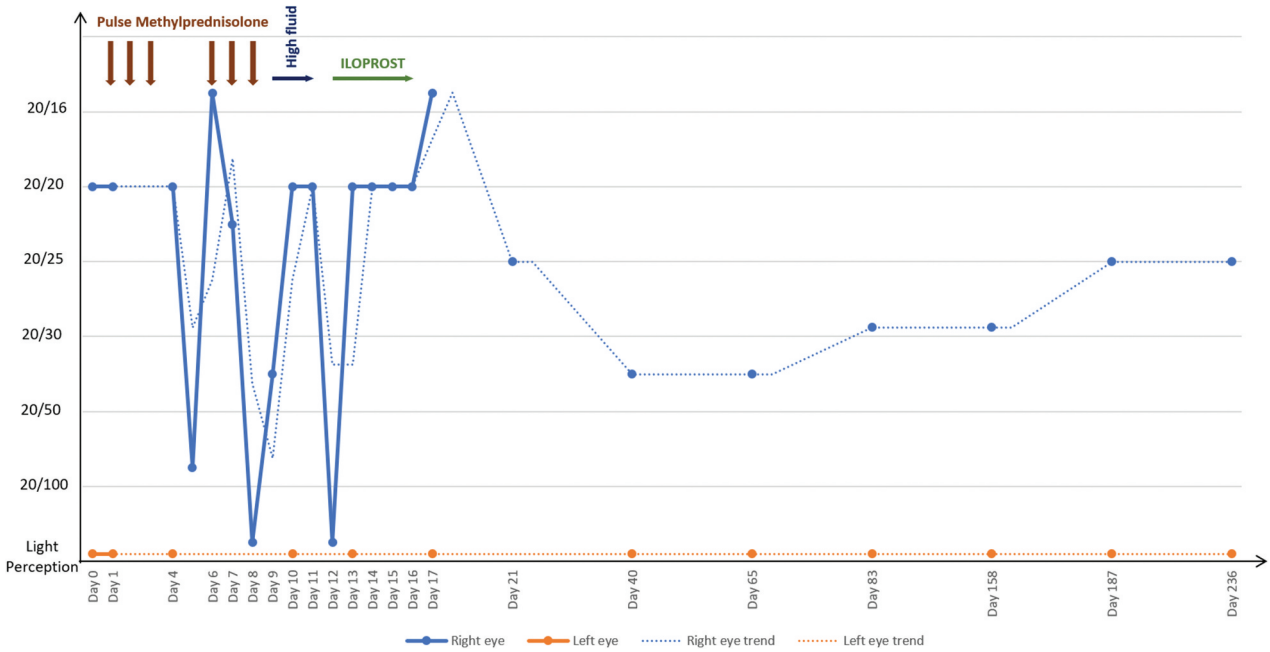

Figure 2. Evolution of the both eyes' visual acuities (Snellen chart) with treatment modulation over time.

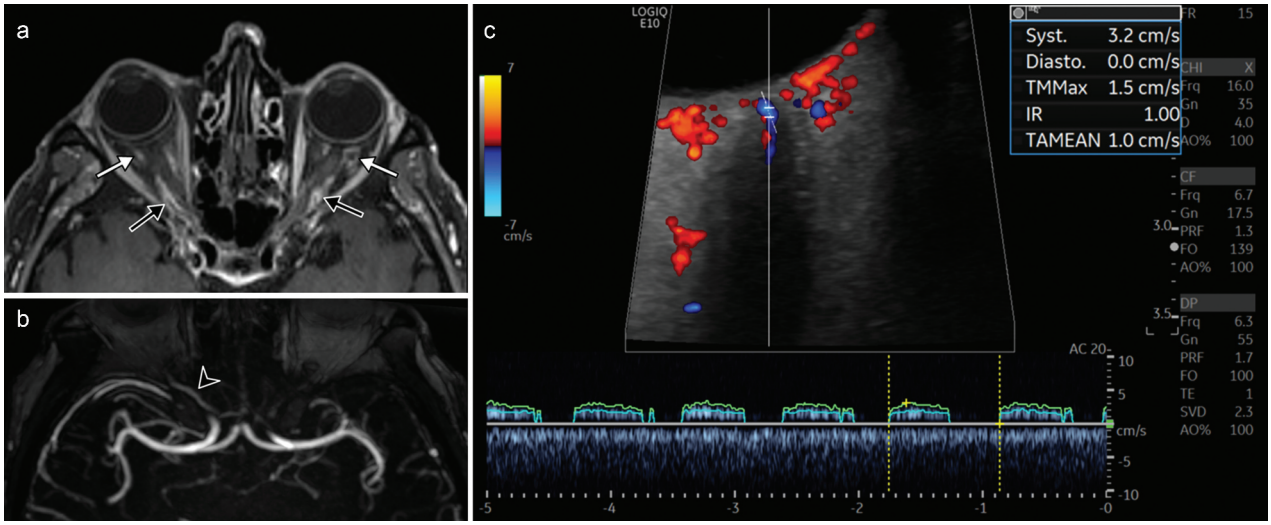

Figure 3. Orbital imaging features. (a) Contrast-enhanced fat-suppressed vessel-wall MRI showing bilateral thickening and enhancement of ophthalmic arteries' wall (black arrows) and peripapillary enhancement (white arrows) very suggestive of Giant Cell Arteritis (b) Magnetic Resonance Angiography showing an occlusion of the left ophthalmic artery and an inflammation of the right one (black arrowhead) (c) Color-Doppler flow imaging ultrasound of the right central retinal artery showing a decrease of peak systolic velocity and an increase of the resistive index suggestive of ocular suffering.

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Authors' contributions

Study concept and design: TS, NA, CVC
Acquisition of data: TS, NA, AL, CD, VV, MMF, CVC
Analysis and interpretation of data: TS, NA, AL, CD, VV, MMF, CVC
Drafting the manuscript: TS, NA
Critical revision of the manuscript for important intellectual content: TS, NA, AL, CD, VV, MMF, CVC

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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