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CASE SERIES

Magnetic resonance imaging findings in giant cell arteritis

NM D'Souza[1], ML Morgan[2], SJ Almarzouqi[2] and AG Lee[1,3,4,5]

[1]Department of Ophthalmology, Baylor College of Medicine, Houston, TX, USA

[2]Department of Ophthalmology, Houston Methodist Hospital, Houston, TX, USA

[3]Departments of Ophthalmology, Neurology, and Neurosurgery, Weill Cornell Medical College, Houston, TX, USA

[4]UTMB Galveston, TX and the UT M.D. Anderson Cancer Center, Houston, TX, USA

[5]Department of Ophthalmology, The University of Iowa Hospitals and Clinics, Iowa City, IA, USA

Correspondence: AG Lee, Department of Ophthalmology, Blanton Eye Institute, Houston Methodist Hospital, 6560 Fannin Street, Scurlock Tower 450, Houston, TX 77030, USA
Tel: +1 713 441 8823; Fax: +1 713 790 6441.
E-mail: AGLee@HoustonMethodist.org

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Abstract

Purpose **Giant cell arteritis (GCA) is a systemic vasculitis that affects medium-to-large-caliber arteries. Early diagnosis and treatment is essential as involvement of the ophthalmic artery or its branches may cause blindness. Radiographic findings may be variable and non-specific leading to delay in diagnosis. We conducted a review of the literature on neuroimaging findings in GCA and present a retrospective case series from tertiary-care ophthalmic referral centers of three patients with significant neuroimaging findings in biopsy-proven GCA.**

Methods **Retrospective case series of biopsy-proven GCA cases with neuroimaging findings at the Department of Ophthalmology, Blanton Eye Institute, Houston Methodist Hospital between 2010–2015 were included in this study. Literature search was conducted using Google Scholar and Medline search engines between the years 1970 and 2015.**

Results **We report findings of optic nerve enhancement, optic nerve sheath enhancement, and the first description in the English-language ophthalmic literature, to our knowledge, of chiasmal enhancement in biopsy-proven GCA. We describe four main categories of neuroimaging findings that may be seen in GCA from our series and from past cases in the literature.**

Discussion **It is essential that clinicians be aware of the possible radiographic findings in GCA. Appropriate and prompt treatment should not be delayed based upon these findings.**

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Introduction

Giant cell arteritis (GCA) is a systemic vasculitis affecting medium- and large-caliber arteries.[1] Ophthalmic artery involvement may cause arteritic-anterior or -posterior ischemic optic neuropathy (A-AION or A-PION), cilioretinal, or central retinal artery occlusion, or choroidal ischemia with devastating vision loss. Early diagnosis and treatment with high-dose corticosteroids is crucial. Symptoms include headache, jaw claudication, fever, weight loss, polymyalgia rheumatica, and tender temporal artery.[1–3]

Atypical presentations may delay diagnosis.[2] Diagnosis is confirmed via superficial temporal artery biopsy (TAB) and neuroimaging is not typically necessary, but some patients in the United States frequently undergo these studies before evaluation. Unfortunately, imaging findings can be non-specific. In addition, orbital inflammatory signs and symptoms can occur, mimicking idiopathic orbital inflammatory syndrome or other infectious/inflammatory etiologies.[4–8] Furthermore, MRI abnormalities including T2 hyperintensity, diffusion-weighted imaging restriction, and gadolinium enhancement may occur in GCA-related AION or PION, broadening differential diagnosis.[2,4,7–10] Previously described MRI findings, noted in Table 1, include dural and perineural sheath enlargement, sheath and nerve enhancement, and extracranial and intracranial vascular changes.

We present neuroimaging findings in biopsy-proven GCA in three patients, review the literature on the subject, and outline four main categories of findings. To our knowledge, this is the first report of MR chiasmal enhancement in biopsy-proven GCA in the English-language ophthalmic literature.

Materials and methods

Retrospective case series of biopsy-proven GCA with neuroimaging findings at the Blanton Eye Institute, Houston Methodist Hospital between 2010–2015 were included in this study. Literature search was conducted using Google Scholar and Medline search engines between 1970 and 2015, limited to English-language and full papers. Keywords included 'magnetic resonance imaging', 'giant cell arteritis',

Table 1 CT and MRI findings in GCA

Report[ref.]	*Age/sex and symptoms*	*Imaging findings*	*Treatment*	*Outcome*	*TAB results*
Nielsen and Eriksen[17]	75/M; Visual loss OS	XR: left orbit and maxillary sinus blurring (transitory edema)	Oral prednisone 80 mg	Improved	Positive
Laidlaw *et al*[6]	66/F; Proptosis, ophthalmoplegia	CT: proptosis	Oral prednisolone 80 mg	Resolved after 4 months	Positive
Chertok *et al*[4]	67/F; Ptosis, elevation deficit OS	CT: enhancing mass in superior rectus	Oral prednisolone	Improved	Positive
Nassani *et al*[18]	69/F; Bilateral proptosis	MRI: irregular, thickened sheaths, intraconal abnormal signal intensity	Oral prednisone 50 mg/day	Improvement after 40 days	Positive
Lee *et al*[9]	82/F; bilateral visual loss (NLP OS, 20/400 OD)	MRI: bilateral optic nerve enhancement	IV methylprednisolone (1000 mg/day) followed by oral prednisone taper	Visual function unchanged	Positive
	86/F; Acute, bilateral visual loss (counting fingers 1 m OD, NLP OS)	MRI: bilateral enhancement of optic nerves	Oral steroids	Visual function stabilized	Positive
	80/M; Visual loss OS followed by loss OD	MRI: mild bilateral optic nerve enhancement	IV methylprednisolone (1000 mg/day) followed by oral prednisone taper	—	Positive
Joelsen *et al*[19]	69/M; Double vision, LR palsy OD	MRI: dural thickening/enhancement, LR/temporalis enhancement	Prednisone	Full resolution	Positive
Lee *et al*[7]	70/M; Proptosis, visual loss OD, pain	MRI: bilateral infiltrative lesions	Steroids and cyclophosphamide	No light perception OD, 20/100 OS	Positive
	69/M; Diplopia, proptosis	MRI: enhancement of orbit and optic nerve	Steroids	Improved after 5 weeks	—
	72/F; Pain, proptosis, injection OD	MRI: abnormal signal in medial rectus OD	IV steroids	Improved after 5 weeks	Positive
Morgenstern *et al*[8]	83/M; NLP OD, 20/50 OS	MRI: bilateral enhancement optic nerve sheath and orbital fat	IV methylprednisolone	Vision stabilized	Positive
Cockerham *et al*[20]	75/F; Pain, visual loss OD, proptosis, atypical presentation	CT: intraconal (superior ophthalmic fissure) mass	External beam radiation (2000 cGy every 2 weeks)	Visual function restored after 1 month	Positive
Reddi and Volbracht[10]	83/F; Visual loss OD, classic presentation	MRI: enhancement of sheath, retrobulbar fat with inflammatory orbital pseudotumor	IV solumedrol 1 g/day Prednisone 60 mg/day	Improvement in visual function	Negative
Liu and Chesnutt[2]	83/F; Headache, blurred vision OD, acuity defect, elevated ESR, no fundus findings	MRI: bilateral perineural optic nerve enhancement	IV methylprednisolone	Loss of light perception	Positive
	68/F; Visual loss OS to NLP	MRI: perineural enhancement of left optic nerve	Prednisone 60 mg/day	Vision stabilized	Positive
Hittinger, Berlis and Pfadenhauer[5]	78/M; Visual loss, proptosis OS	MRI: diffuse orbital inflammation	Prednisolone 100 mg/day	Improvement visual acuity OS	Positive
Liu and Miller[23]	67/M; Visual loss OS	MRI: bilateral optic nerve sheath enhancement	IV methylprednisolone 1 g to prednisone 60 mg	Unknown	Positive
Current Series	60/M; Visual loss OS, diplopia	MRI: increased signal intensity proximal left optic nerve, subtle optic nerve enhancement	IV methylprednisolone 1 g/day to prednisone 1 mg/kg	Improved visual acuity OS	Positive
	83/M; Visual loss OS, orbital fullness	MRI: enhancement of left optic nerve sheath	Prednisone 60 mg/day	Stabilized acuity OS	Positive
	63/M; Visual loss OD progressing to bilateral loss with only LP intact	MRI: enhancement and enlargement of perineural sheath bilaterally and chiasm	IV steroids, low-dose prednisone	Stabilized at counting fingers 5 feet OD; 20/320 OS	Positive

'temporal arteritis', 'radiographic findings', 'enhancement', 'orbital findings', and 'orbital inflammation'. In each case, findings on TAB considered to be indicative of GCA included intimal thickening and proliferation with fragmentation of the internal elastic lamina, necrotizing vasculitis, mononuclear cell predominance, and granulomatous inflammation. Magnetic resonance imaging (MRI) was accomplished with a 3-T MRI scanner.

Case reports

Case 1

A 60-year-old Caucasian male patient presented with acute-onset headache, vision loss OS, diplopia, eye pain, and weight loss. Past medical history included hypertension and dyslipidemia. Erythrocyte sedimentation rate (ESR) was 36 mm/h (normal <20) and C-reactive protein was 3.76 (normal <0.8 mg/l). Visual acuity was 20/40 OD and counting fingers (CFs) at 2 feet OS. There was a relative afferent pupillary defect and disc edema OS with diffuse depression, with a mean deviation of −2.13 dB on visual field testing. Slit lamp examination (SLE) revealed 1−2+ nuclear sclerotic cataract (NSC) OU consistent with 20/40 vision. MRI revealed increased signal intensity on the proximal left optic nerve with subtle enhancement (Figure 1a). TAB indicated findings consistent with resolving GCA. After intravenous (IV) methylprednisolone 1 g/day for 3 days and then oral prednisone (1 mg/kg) visual acuity improved to 20/80 OS.

Case 2

An 83-year-old Caucasian male patient presented with sequential loss of vision OU, complaining of orbital fullness OS. Past medical history included diabetes mellitus, hypertension, and ischemic heart disease. ESR was 5 mm/h. The patient was seen by outside ophthalmologist who initiated oral prednisone 60 mg/day.

Visual acuity was no light perception (LP) OD and CFs at 1 foot OS. Pupils were sluggishly reactive OU. SLE revealed brunescent NSC with no view of the posterior segment OD. Dilated fundus examination (DFE) showed 1+ disc pallor OS with CDR of 0.6. TAB findings were consistent with GCA, whereas MRI of brain and orbits revealed left optic nerve sheath enhancement (Figure 1b). Vision remained unchanged after steroid treatment.

Case 3

A 63-year-old Caucasian male patient presented with rapid visual loss to LP OU. Past medical history included spinal stenosis, cataracts, and hypercholesterolemia. Pupils were poorly reactive with light-near dissociation OU. SLE was normal with mild nuclear cataract OD and posterior chamber intraocular lens OS. DFE showed disc edema and fluorescein angiogram confirmed mild leakage of discs OU without choroidal involvement.

MRI showed enhancement and enlargement of the perineural sheaths of both optic nerves and the chiasm (Figure 1c). TAB demonstrated findings consistent with GCA. After treatment with IV steroid therapy followed by oral prednisone, vision improved from LP OU to CFs OD and 20/320 acuity OS. Four months after presentation, the patient's vision was stable at CFs at 5 feet OD and 20/320 acuity OS.

Discussion

Neuroimaging is not usually required in patients with typical presentations of GCA and when performed is generally normal.[11,12] However, some patients have already undergone imaging before neuro-ophthalmic evaluation and these studies may be abnormal.

We report four main imaging findings in GCA from our cases and the literature (Table 1):

1. Non-specific orbital enhancement.
2. Optic nerve parenchymal enhancement.
3. Perineural sheath enhancement.
4. Optic chiasmal enhancement. To our knowledge, ours is the first report of MR chiasmal enhancement in biopsy-proven GCA.

Other important MRI findings in GCA include those involving the vascular supply not only extracranially but also intracranially, particularly vessel wall enhancement of the intradural ICA.[13] MRI findings may hold some diagnostic value in distinguishing between A-AION as in GCA and in non-arteritic AION, in which they are typically normal, but may include a small optic nerve and chiasm volume and rarely increased proton density, STIR signal, and gadolinium enhancement.[14–16] Differential diagnosis for these MRI findings can lead to inappropriate testing and delay diagnosis and treatment. Finding 1 may suggest orbital inflammatory disease, whereas Finding 2 may suggest inflammatory or demyelinating etiologies (eg, multiple sclerosis, neuromyelitis optica, optic neuritis). Finding 3 may point to inflammatory (eg, sarcoid, granulomatous disease), infiltrative optic perineuritis, or sheath neoplasm (eg, optic nerve sheath meningioma). Finding 4 may suggest inflammatory, infiltrative, or demyelinating chiasmopathy.[2,5,7–12,18–22] Clinicians should be aware of the radiographic findings in GCA and, although further testing may be required to exclude alternative etiologies, diagnosis and treatment should not be unnecessarily delayed.

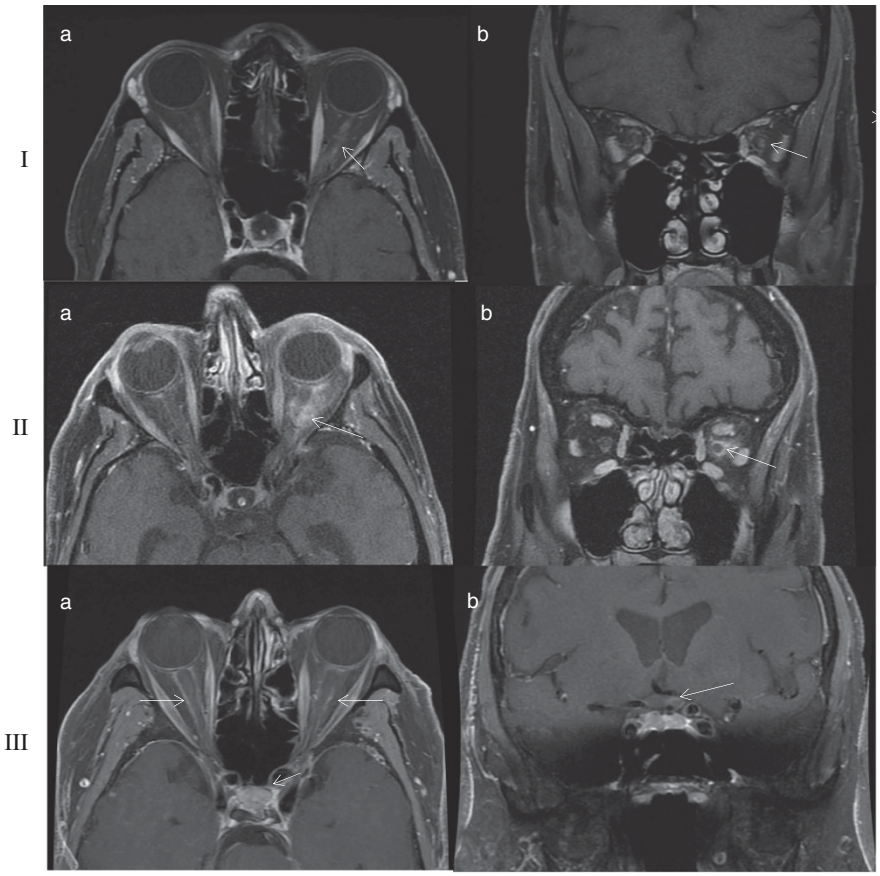

Figure 1 Contrast-enhanced axial (a) and coronal (b) post-gadolinium T1-weighted orbital magnetic resonance imaging with fat suppression. Case 1 (top), increased signal intensity at the proximal portion of left optic nerve with subtle optic nerve enhancement (arrows). Case 2 (middle), enhancement of left optic nerve sheath (arrows). Case 3 (bottom), enhancement and enlargement of the perineural sheath of both optic nerves, as well as the chiasm (arrows).

Summary

What was known before

- GCA is a vasculitic disorder that can lead to devastating visual loss if not treated promptly.
- TAB is the gold standard for diagnosis but neuroimaging may also have a role in atypical presentations.
- Neuroimaging findings in GCA are often non-specific and variable and can lead to unfortunate delays in diagnosis and treatment.

What this study adds

- Three new unique cases of GCA with variable neuroimaging findings that agree with past reports.
- The first report, to the authors' knowledge, of chiasmal enhancement in GCA.
- A review of the literature and outline of the four main categories of neuroimaging findings in GCA, based on the current series and past studies, that health professionals should be aware of.

Conflict of interest

The authors declare no conflict of interest.

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