

IMAGING HIGHLIGHTS

Value of MRI in diagnosis of giant cell arteritis

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Abstract

Giant-cell arteritis is an inflammatory disease of blood vessels most commonly involving large and medium arteries of the head, predominately the branches of the external carotid artery. It is a form of vasculitis. Giant-cell arteritis of the temporal artery is referred to as “temporal arteritis”. Giant cell arteritis is rare among Asians. Diagnosis is based on American College of Rheumatology (ACR) classification criteria for giant-cell arteritis with gold standard temporal artery biopsy.¹ Blindness is a feared complication, mostly caused by anterior ischaemic optic neuropathy. High-resolution magnetic resonance imaging (MRI) has started to play a role in the diagnosis of giant cell arteritis. Evidence of arteritis involving the superficial temporal arteries with mural thickening and even the ophthalmic arteries can be seen on MRI, which further increase the diagnostic confidence prior to biopsy.^{2,3} Optic perineuritis is a rare association with giant cell arteritis. MRI is required for diagnosis especially for differentiation from optic neuritis.⁴ We report a case of giant cell arteritis with optic perineuritis to demonstrate the role of MRI in the diagnosis.

CASE REPORT

A 76-year old Malay gentleman presented with 10 days history of severe throbbing headache. Pain was most severe bitemporally, and 3 days prior to admission he developed intermittent jaw pain and claudication associated with sudden and painless bilateral visual loss. There was no fever or myalgia. He had no known medical illness and was well prior to presentation. Examination revealed a fully conscious patient. Visual acuity was no perception to light bilaterally. His pupils were dilated and poorly reactive to light with bilateral pale and swollen optic discs (Figure 1). Cornea and anterior chambers were normal and there was no ophthalmoplegia. Clinically, eye findings indicated bilateral anterior ischaemic optic neuropathy. The superficial temporal arteries were distended and tense bilaterally. His ESR and C-reactive protein (CRP) were elevated with values of 64mm/hour and 6.05 mg/dL respectively. He was anaemic with haemoglobin level of 10.4 g/dL.

MRI of the orbit showed bilateral perineural optic nerves thickening and enhancement in keeping with optic perineuritis (Figure 2). This was associated with minimal surrounding retro-

orbital fat streakiness. The optic nerves themselves did not exhibit abnormal enhancement post contrast. The globes and extraocular muscles were unremarkable. Bilateral superficial temporal arteries exhibited significant mural thickening (ranging from 1-1.5mm) and enhancement in the pre and post gadolinium fat-suppressed VIBE (volume interpolated breath-hold) T1-weighted image (Figure 3). Fat-suppressed T2-weighted image showed hyperintensity representing inflammation of the fat surrounding these vessels (Figure 4).

MRI of the brain only revealed non-specific white matter changes with no regional infarction. Biopsy of right temporal artery revealed intimal thickenings with infiltration of inflammatory cells and a few granulomas consisting of histiocytes and giant cells consistent with the diagnosis of giant cell arteritis.

He was started on oral high dose corticosteroids. There was gradual reduction in the ESR and CRP levels. His headache and both optic nerves swelling improved though his vision remained poor. Unfortunately, four months after presentation, patient succumbed to death due to overwhelming sepsis from pneumonia.

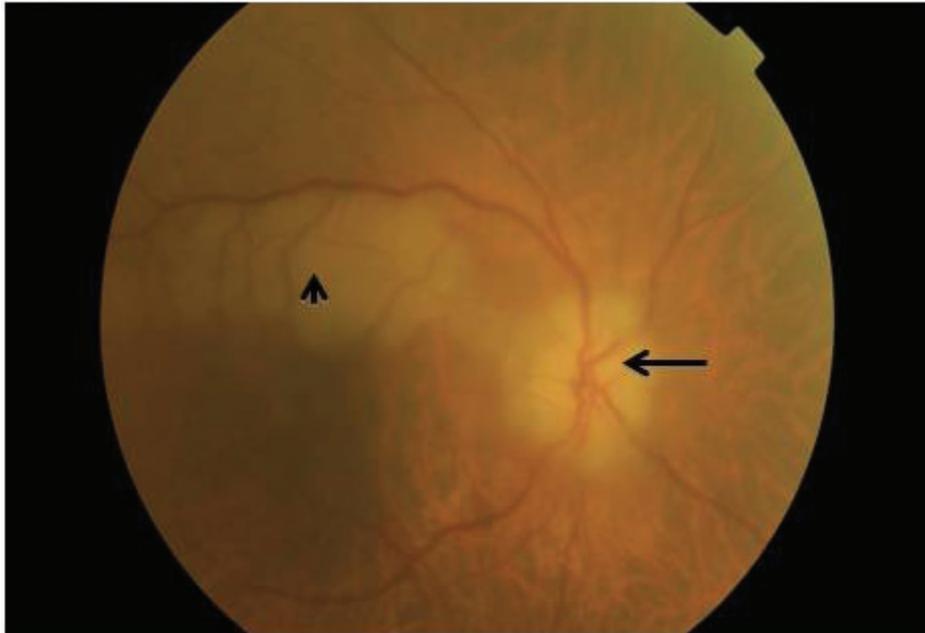


Figure 1. Fundus photo of the right eye shows a pale and swollen optic disc (arrow) with an area of pale deep retina (arrowhead) at superior macula.

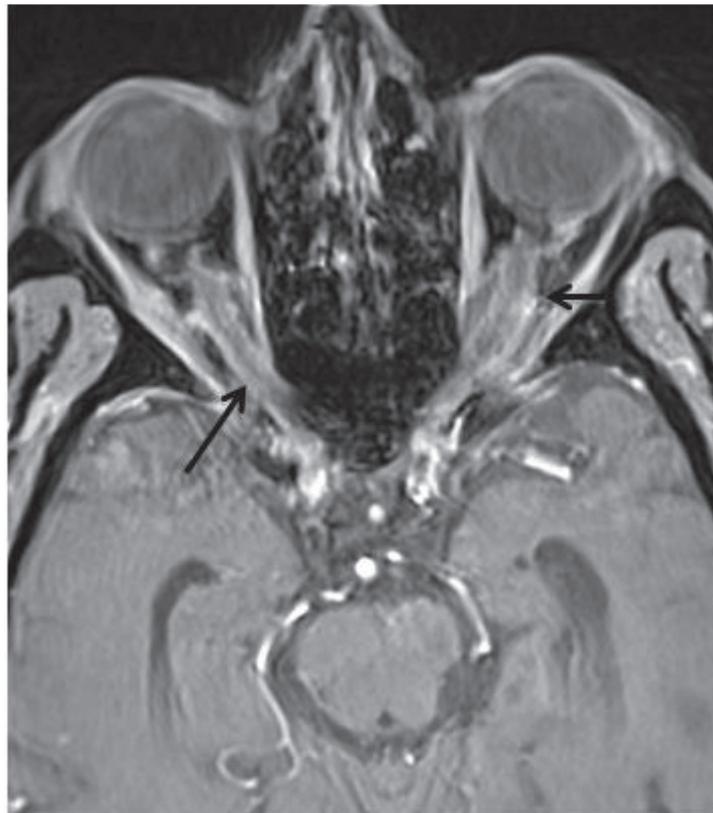


Figure 2. Post gadolinium fat suppressed VIBE T1-weighted in axial plane showing bilateral optic perineural enhancement (arrows) consistent with optic perineuritis.

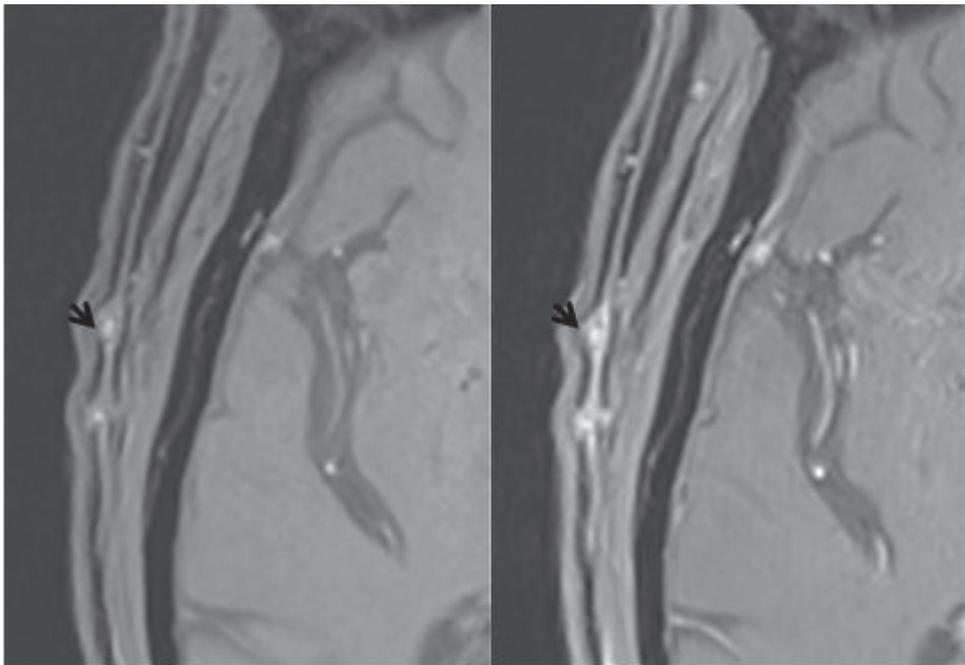


Figure 3. Pre (A) and post gadolinium (B) fat suppressed VIBE T1-weighted images show mural thickening and enhancement of the right superficial temporal arteries (arrows) (note in this sequence the flowing blood is bright in the pre gadolinium image).

DISCUSSION

Giant cell arteritis typically affects individual more than 50 years old. It is rare in Asians with

reported incidence to be 20 times less common as compared to Caucasians.⁵ Clinical presentations include severe headache with localized scalp tenderness, fever, jaw claudication, painful

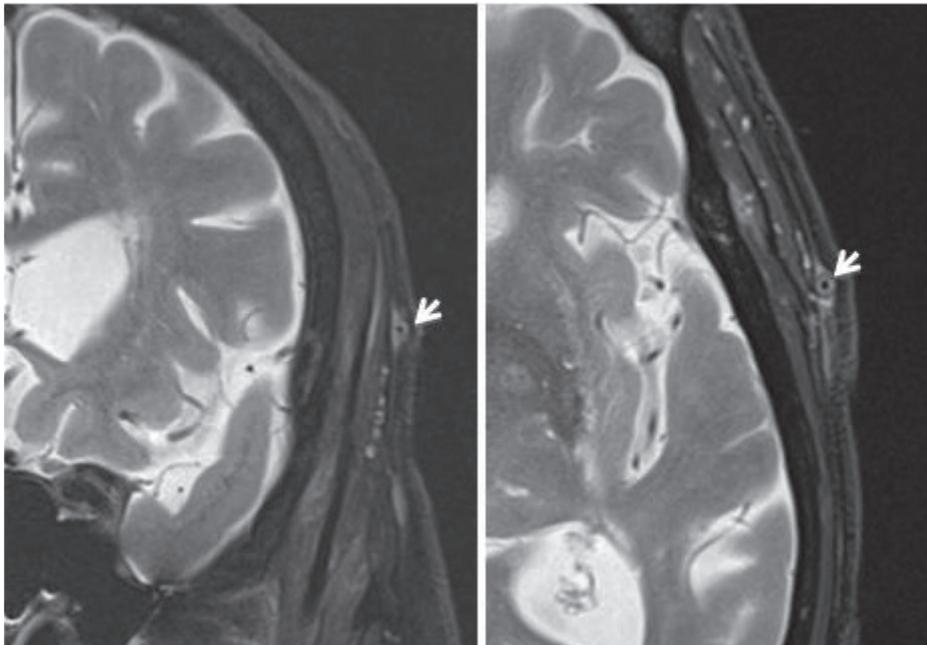


Figure 4. Fat suppressed T2-weighted images in coronal (A) and axial (B) planes show hyperintensity representing inflammatory changes surrounding the left superficial artery. The mural hyperintensity also indicate inflammation of vessel wall seen in arteritis.

tongue, anaemia with decreased temporal artery pulse. Our patient has typical presentation of an elderly with headache, jaw pain, and high ESR. The granulomatous inflammation of the temporal artery in histology confirmed the diagnosis of giant cell arteritis. Our patient also had bilateral visual loss. Visual loss occur in 10-60% of patients with giant cell arteritis.⁶ Most common cause of blindness is anterior ischaemic optic neuropathy resulting from ischaemia of the posterior ciliary arteries supplying the optic nerve head followed by central retinal artery occlusion accounting for less than 10% of cases.⁷ Other ocular manifestations include posterior ischaemic optic neuropathy and extra-ocular muscle palsies.

Optic perineuritis is rarely associated with giant cell arteritis with few reported cases previously.^{8,9} It is caused by inflammation in the surrounding optic nerve sheath. Compared to anterior ischaemic optic neuropathy, isolated optic perineuritis is mostly asymptomatic and is usually diagnosed incidentally by MRI. Optic perineuritis can also result in blindness. Our patient had clinical suspicion of anterior ischaemic optic neuropathy causing his blindness, but presence of bilateral intraorbital optic perineural enhancement and thickening with streakiness in the surrounding retro-orbital fat indicate associated optic perineuritis on MRI. If the thickening is more marked, this will produce the “tram-track sign” on the axial image and “doughnut-sign” on the coronal scan, mimicking optic nerve meningioma.⁴ Optic perineuritis is by no mean specific for giant cell arteritis and can occur in other conditions like neurosyphilis and inflammatory orbital pseudotumor.

The MRI of our patient also showed mural enhancement and thickening with luminal narrowing of the superficial temporal arteries on pre and post gadolinium T1-weighted images. The associated hyperintensity in the surrounding fat and wall of the affected vessels indicate oedema or inflammatory changes on T2-weighted image. These findings were consistent with previous reports.² We scanned the patient using a fat-suppressed 3D VIBE (volume interpolated breath-hold) T1-weighted image in which flowing blood within vessels are already hyperintense in the pre gadolinium scan, thus mural thickening and luminal narrowing can already be appreciated. Post gadolinium scan depicted the avid mural enhancement.

Apart from aiding diagnosis, MRI was also found to be useful in follow up after steroid treatment. The resolution in the degree of mural

thickening and enhancement on MRI were found to correlate well with laboratory remission.¹⁰ It may also be of value in localizing the best site for temporal artery biopsy thus resulting from less false negative biopsy due to skipped lesions.²

In conclusion, even though temporal artery biopsy is still the gold standard, MRI has much added value in increasing the confidence of giant cell arteritis diagnosis especially in our Asian population where this condition is rare.

DISCLOSURE

Conflict of interest: None

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REFERENCES

1. Hunder GG, Bloch DA, Michel BA, *et al.* The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. *Arthritis Rheum* 1990; 33:1122
2. Thorsten AB, Oliver W, Markus U, Jens T, Dieter S, Mathias L. High-Resolution MRI in giant cell arteritis: Imaging of the wall of the superficial temporal artery. *AJR* 2005; 184:283-7.
3. Julia G, Thomas N, Markus U, Wolf AL, Peter V, Mathias L, Thorsten AB. Involvement of the ophthalmic artery in giant cell arteritis visualized by 3T MRI. *Rheumatology* 2009; 48(5): 537-41.
4. Purvin V, Kawasaki A, Jacobson DM. Optic perineuritis: clinical and radiographic features. *Ach Ophthalmol* 2001; 119(9):1299-306.
5. Pereira LS, Yoon MK, Hwang TN, *et al.* Giant cell arteritis in Asians: a comparative study. *Br J Ophthalmol* 2011; 95:214-6.
6. Jonasson F, Cullen JF, Elton PA. Temporal arteritis. *Scot Med J* 1979; 24:111-7
7. Cullen JF. Occult temporal arteritis: A common cause of blindness in old age. *Brit J Ophthal* 1967; 51:513-25.
8. Karuppannan B, Wan Hazabbah WH, Wahid Abdullah SW, Zunaina E, Naik VR. Total ophthalmoplegia, optic perineuritis and central retinal artery occlusion secondary to giant cell arteritis: a case report. *Int J of Ophthal* 2009; 9(12): 2269-72.
9. Nassani S, Cocito L, Arcuri T, Favale E. Orbital psuedotumor as a presenting sign of temporal arteritis. *Clin Exp Rheumatol* 1995; 13:367-9.
10. Bley TA, Markl M, Schelp M, *et al.* Mural inflammatory hyperenhancement in MRI of giant cell (temporal) arteritis resolves under corticosteroid treatment. *Rheumatology* 2008; 47:65-7.